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REVIEW ARTICLE

Tips and Tricks in the Management of Megameatus Intact Prepuce Variant of Hypospadias: A Review Article

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ABSTRACT

Introduction: Hypospadias is congenital anomalies with various spectrum and variants, presenting with abnormal urethral meatus short of the native tip of the glans. megameatus intact prepuce is a rare variant of glanular hypospadias seen in about 3-6 % of hypospadias cases. The main features of megameatus intact prepuce are large urethral external meatus with deep with deeply grooved glans penis and the preputial skin is usually present and intact with no chordae. Seven named procedures for megameatus intact prepuce were discussed.

Methodology: We use the following search engines for out study where we found the following articles and reviews as follows in Scopus search, we found 26 articles, in AJOL we there were 7 articles and in PubMed Central Search we found 32 articles, then in Google scholar the search revealed 388 articles, 45 articles were reviewed and summarised and 7 established procedures were discussed.

Results: Seven named established procedures for megameatus intact prepuce variant of hypospadias were summarized and discussed and include Pyramid Procedure, Glanular Approximation Procedure, Tubularised Urethral Plate Urethroplasty, Tubularised Incised Urethral Plate Procedure, Mathieu Technique, Cutaneous advancement procedure and The Inframeatal Vascularised Flap Procedure.

DISCUSSION: The Surgical procedures for repair of megameatus intact prepuce is technically challenging not only for the surgeon but the anesthethiogist alike because the patients are paediatric with small organs, therefore appropriate instruments and proper timing of the surgical intervention is paramount. Duckett and Keating et al in their study identified the first challenge is to identify those patients who truly need surgical repair. In a study by Marc Cendron et al they found out that the features identified as important in the decision-making process were (1) meatal opening close to or below the coronal margin; (2) A deep glans cleft; (3) a wide, splayed-out glans; and (4) an abnormal urinary stream if witnessed. in a study by Duckett JW, Keating MA, Hatch DA et at and Austin Hill G, Wacksman J they reported a satisfactory outcome with the various described surgical procedures.

Conclusion: The tip and tricks of the surgical procedures of megameatus intact prepuce rare variant of hypospadias is a review article to aimed to improve on the surgical outcome including normal conical appearance of the glans, a urethral meatus of normal caliber, and a single, normal urinary stream with no symptoms and to minimise the complications such as urethrocutenous fistula as extracted from the expert's vast experience and modifications of the existing techniques, in our study we summarised the experts seven established techniques and the modifications to achieve the desired objectives.

Keywords: Tip and Tricks, Surgical Management, Megameatus intact prepuce

INTRODUCTION

Hypospadias is congenital anomalies with various spectrum and variants, presenting with abnormal urethral meatus short of the native tip of the glans, it can occur with or without ventral chordae or hooding, megameatus intact prepuce is a rare variant of glanular hypospadias seen in about 3-6 % of hypospadias cases.1'2 The aim and the scope of our study is to provide the tips and tricks of the surgical techniques of managing megameatus intact prepuce variant of hypospadias, and the objectives of our studies to describe the established techniques of this rare variant of hypospadias and to help surgeons get acquainted with the procedures based on the experts descriptions and modifications. The main features of megameatus intact prepuce (MIP) are large urethral external meatus with deep with deeply grooved glans penis and the preputial skin is usually present and intact with no chordae, and usually no effects in micturition or sexual physiology. 3'4 However, quality of life and psychosexuality may be affected, although patients who present with the more enlarged forms of the meatus may present with a splayed urinary stream.

There is myriad of surgical techniques used in repair of megameatus intact prepuce (MIP) variant of anterior hypospadias with numerous and various modifications of some original techniques with the sole aim to improve the cosmetic outlook of the child's penis to minimise and most case to prevent the consequential psychological trauma the patients suffered if left untreated. ⁷¹⁸¹⁹

Many named procedures for megameatus intact prepuce include Pyramid Procedure, Glanular Approximation Procedure (GAP), Tubularised Urethral Plate Urethroplasty (TUPU), Tubularised Incised Urethral Plate (TIP) Procedure, Mathieu Technique, Cutaneous advancement procedure and Inframeatal advancement flap. 1011

The tips and tricks of the surgical procedures for megameatus intact prepuce variant of hypospadias are the maneuverers and the precautions taken to ensure a successful surgical procedure which can be technically challenging, with maximal possible cosmetic and functional outcome while minimising the complications rate, they are the extract of the experts' advice wit track records and experience in the management of this rare congenital anomaly and occasionally acquired.¹²

The distinct anatomic features of MIP have led to the emergence of several techniques specifically intended to achieve good cosmetic and functional results that were not achievable with standard technique for hypospadias repair (e.g., perimeatal-based flaps and MAGPI).^{13*22}

Another characteristic that makes MIP a surgical challenge is that most of the affected patients have been circumcised, the penile skin is usually thin and scarred, and are thus lacking prepucial and dartos tissue to be used for reconstructive procedures. However, a study by Snodgrass proved that circumcision does not affect the results of TIPU in MIP. 14'23

METHODOLOGY

We use the following search engines for out study where we found the following articles and reviews as follows in Scopus search we found 26 articles out of which 23 were written in English, in AJOL we there were 7 articles and in PubMed Central Search we found 32 articles, then in Google scholar the search revealed 388 articles as at 19th May 2023 by 8:21 pm our local time, the 45 relevant articles and reviews were summarised and 7 established procedures were discussed, tips and tricks extracted and presented in this manuscript, we use megameatus intact prepuce, surgical management of megameatus intact prepuce and tips and tricks in the surgical management of megameatus intact prepuce as our key words.

RESULTS

In our study we reviewed the seven established procedures that were described by the experts with various modifications and tips and tricks of the named procedures were discussed with the aim of minimizing the complications and improving the outcome and presented as follows:

PYRAMID PROCEDURE:

This is one the early surgical procedure for the repair of megameatus intact prepuce, Duckett and Keating described it in $1989\ ^{13^{\circ}15}$

Technique: Patient under general anesthesia draping and asepsis done, the prepuce is retracted and the megameatus exposed at this stage an appropriate sized well lubricated catheter should inserted, then a Tennis racket incision is made with size 15 or 10 blade around the edges of megameatus up to the level of the corona besides the groove of the glans, here one must be careful not to be too deep to avoid unnecessary spongy tissues bleeding, then glanular wings can be made with deepening the incision around the edges of the glanular groove, however the distal urethral groove is left flat dorsally, with the assistant helping with hemostesis then a small wedge is taken from the ventral tissue and the distal urethra is sutured in continuity with the urethral plate up to the apex of the pyramid, now the urethra and the glanular wings are Tubularised over a stent to form the neourethra, there is a modification which involves

the creation of a vascularized second-layer tissue flap to minimise risk of fistula formation, then glanuloplasty is done Skin cover is completed with approximation of the glanular flap. 13'15'16

A dressing is applied over the area to protect it while it heals usually using Vaseline gauze or povidone iodine instilled on the wound before application of the dressing and penis is kept in the anatomical position to avoid the pressure of the stent on the stiches, The patient is monitored for a few hours following the procedure to ensure that there are no complications. Prophylactic antibiotics and analgesia may be prescribed to help manage any discomfort during the recovery period. 17120

GLANULAR APPROXIMATION PROCEDURE:

This was described by Elbatarny et al in 2011 as well as M. Sanal et al reported an excellent outcome in 7 cases with GAP. $^{10\cdot16}$

Glans approximation procedure (GAP) is generally used for a MIP ventral hypospadias variant with a wide and deep glandular groove or a wide meatus. Even though GAP is relatively easy to perform, acquiring the gland's conical shape is difficult as the proximal gland is very small. 51718.

Technique: Patient under general anesthesia routine draping and aseptic technique with WHO protocol observed, the prepuce is retracted this will reveal the meatus appropriate sized catheter inserted, then a U- Shaped incision is made around the megameatus and the urethral plate, and subsequently the glanular wings are raised, there after the urethra plate is assessed and the excess urethral plate is trimmed to make it the same diameter with the proximal urethra, in some patients there maybe need to deglove the penile skin by 1 cm which will allows for harvesting the intermediate layer from island flap in uncircumcised patients or from the dartos fascia or Bucks fascia in circumcised patients this tissue will be used to reinforce the repair to eliminate the tension on the meatus, using vicryl 4/0 - 6/0 is used to do the urethroplasty and this is followed by laying the intermediate layer with interrupted sutures and finally the glanular wings are sutured together and the penile skin is sutured after circumcision. 17:18:19

The main limitations of the GAP are unequal size of neo-urethra due to non-mobilization of glanular wings and superimposition of glanular suture lines without interposing tissue layer may lead to fistula, and again it is most suitable in glanular MIP only. But those who were dissatisfied with the results of the application of other available techniques to the MIP variant, they modified GAP by adding the mobilization of glanular wings for equal size neourethra with normal urethra and tension free

urethroplasty; and interposing tissue layer dorsal dartos layer to prevent fistula. 20'21

TUBULARISED URETHRAL PLATE URETHROPLASTY:

Surgical correction of MIP variant of hypospadias in the era of increased cosmetic awareness is justified. Excellent functional and cosmetic results are obtained with TUPU with few modifications such as spongioplasty, prepuce preservation, and frenuloplasty because of the availability of well-developed spongiosum and wide urethral plate in these patients. Therefore, it should be the preferred procedure in all cases of MIP.²²²³

Technique: This involves the making of U incision around the urethral plate, with the prepuce retracted to exposure the megameatus, this is done in such a way that the base of the u incision is encircles the megameatus and the inner prepuce incised, then the urethral plate and the corpus spongiosum are now mobilized with the glanular wings raised, and the excessive urethral plate is trimmed, the urethral plate is Tubularised and spongioplasty is performed to cover the urethral plate, then glanular flaps are used to cover the neourethra, prepucioplasty can be done if the prepuce needs to be intact, or circumcision can be done at this stage. 20124

Excellent functional and cosmetic results are obtained with TUPU with few modifications such as spongioplasty, prepuce preservation, and frenuloplasty because of the availability of well-developed spongiosum and wide urethral plate in these patients. Therefore, it should be the preferred procedure in all cases of MIP. ⁵¹20123

TUBULARISED INCISED URETHRAL PLATE PROCEDURE

The tubularized incised plate (TIP) urethroplasty or Snodgrass procedure has gained worldwide acceptance for distal hypospadias repair due to its low complication rate, good cosmetic result, and technical simplicity. As a result, several articles have been published concerning various aspects and subtle variations of this procedure, The tubularized incised plate (TIP) repair is based on an old principle of urethral plate tubularization, also known as the Thiersch-Duplay procedure. 12 Although a good concept, its main drawback was the limitation imposed by the width of the urethral plate. 25'26'27

Technique: The procedure involves adequate patient preparation then a circumscribed incision is made around the megameatus, and the penis is degloved this is followed by de-epithelialization of the urethra and parallel longitudinal incisions are then made approximately 6 to 8 mm apart to

separate the glans from the lateral margins of the plate and the wings of the glans developed and mobilized for subsequent glans tension free closure. After which the edge of the urethral plate is deeply incised from the meatus to the end of the plate, just below the tip of glans penis, 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. Then the corpus spongiosum and the plate is sutured over the neourethra, and a vascularized dartos fascia flap is used to cover the urethroplasty as a second layer thereafter the prepuce is divided and rotated on both sides to cover the second layer. ^{28'29'30}

Surgical principles are important, especially in hypospadias surgery. With that in mind, Snodgrass has recommended two-layer neourethra closure to decrease fistula formation in all types of hypospadias defects and the TIP technique is performed by longitudinal incision of the urethral plate to broaden the urethra. The occurrence of urethral stricture or urinary fistula is significantly reduced because the urethral plate is composed of muscle fiber tissue and vascular innervations with no annular anastomosis 31'32'33

MATHIEU TECHNIQUE

Mathieu technique, the proximal urethral flap is reversed to match the urethral orifice, which retains the distal urethral plate, without distal and proximal urethral anastomosis. This procedure reduces the occurrence of urethral stricture and meatal stenosis, and the operation is simple and safe. 2133

Technique: In this procedure the patient is prepared and under general anesthesia the penis is degloved using circummeatal incision and the urethral is mobilized. Then vertical incisions are made on either sides of the glans parallel to the urethral plate, the glanular wings will be dissected while ensuring preservation of the urethral mucosa and sub mucosa and the excess mucosa can be trimmed and then brought back together in the midline using fine absorbable sutures with the appropriate size Foley's catheter as a stent and subcutaneous tissue layer can now be placed over the neourethra can thereby providing a vascularized layer of tissue. The glanular wings are now put over the urethra as well as the urethra meatus and narrowing of the meatus can be prevented by ensuring that the wings are not sutured close to the meatus. 7'34'35

Duan et al found that in Mathieu technique, the proximal urethral flap is reversed to match the urethral orifice, which retains the distal urethral plate, without distal and proximal urethral anastomosis. This procedure reduces the occurrence of urethral stricture, and the operation was simple

and safe. Compared to the other two procedures, they found that there was a significantly longer operative time for the Mathieu technique. One of the 5 patients who underwent the Mathieu procedure developed a urinary fistula while removing the urethral catheter at 2 weeks after surgery. The postoperative complication occurred early, which may be related to the reversed proximal urethral flap tissues being too thin, leading to poor blood supply. 36137138

THE CUTANEOUS ADVANCEMENT PROCEDURE

The Cutaneous Advancement technique has also been used for MIP but that may not be suitable for distal penile MIP, and the glanular defect remains uncorrected, the procedure is described by S. Docimo in 2001 for repair of both iatrogenic and primary megameatus intact prepuce variant of hypospadias. 35138

Technique: Adequate pre-operative preparation with informed consent obtained, patient under general anesthesia in a supine position patient draped and the operation site prepadressed. {operation site marking is done with an indelible marker, then a circummeatal incision is done and adequate hemostasias is secured the penis is then degloved using blunt dissection with gauze and, glanular wings are now raised, if there is redundant mucosa at this stage is reduced, taking care not to over trim it, then the urethral is Tubularised over an appropriate size well lubricated stent then the inferior based frenulum flap is developed which is well vascularized and the skin de-epithelialized and advanced over the neourethra then the glanular wings are approximated after achieving adequate hemostesis and marking sure the neourethral meatus is so tight to avoid future meatal stenosis, and wound is dressed. 39'40

Post operatively patient is given adequate analgesia, and prophylactic antibiotics, then stent and wound care instructions are written.

THE INFRAMEATAL VASCULARISED FLAP PROCEDURE

The Inframeatal Vascularised Flap Procedure was first described in 1994 by Retik AB et al and was modified in 2018 by Marc Cendron et al and was published by frontiers in pediatrics on 14th March 2018 with an effort to improve the outcome while minimising the risk of complications. 41'44

Techniques: Adequate preoperative preparations with sterile urine culture result and informed consent obtained the child is placed under general anesthesia in a supine position, routine draping and asepsis techniques in place, and following standard WHO sign-in, time-in protocol followed, the incision

site is marked out with an indelible marker, then a circumcision incision is done followed by adequate hemostesis, The penis is degloved using blunt dissection with gauze, on the ventral aspect the incision can be carried out after retracting the prepuce in uncircumcised patient, however if the boy is already circumcised then penile degloving can be performed ventrally after circumscribing the megameatus. Stay sutures are then placed below the megameatus and laterally so that it will allow for mobilization of the mega urethra, the stay sutures makes it easier for the dissection and the mobilisation of tissues while minimising the risk of cutting too deep, there after two vertical incisions are done in the glans parallel to the urethral plate so as to raise the glanular wings, sometimes if the bleeding is much one can use a timed tourniquet which must be removed every 20 to 30 minutes, after the urethral plate is freed from the glans, redundant edges of the mucosa can now be to be reduced and then placed back together in the midline over an 8 F or 6F Foley urethral catheter depending on the size of the urethra using vicryl 6 0. Interrupted stitches are then placed, then a layer of subcutaneous tissue can then be brought over the reconstructed urethra to provide an interposed vascularized layer of tissue glanular wings are then reconstructed over the neourethra and wound is dressed, and the penis is placed in an anatomical position to reduce the stress on the suture lines and that improves the outcome. 42'43'45

DISCUSSION

The Surgical procedures for repair of megameatus intact prepuce is technically challenging not only for the surgeon but the anesthethiogist alike since the patients are paediatric with small organs, therefore appropriate instruments and proper timing of the surgical intervention is paramount. Duckett and Keating et al 13 in their study identified the first challenge is to identify those patients who truly need surgical repair. In a study by Marc Cendron et al 2 they found out that the features identified as important in the decision-making process were (1) meatal opening close to or below the coronal margin; (2) A deep glans cleft; (3) a wide, splayedout glans; and (4) an abnormal urinary stream if witnessed. For those who display these features, in a study by the following authors Duckett JW, Keating MA {13}, Hatch DA et al 19 and Austin Hill G, Wacksman J et al 20 they reported a satisfactory outcome with the various described surgical procedures. In the Marc Cendron et al ² series the found out that 25 of the 481 hypospadias repairs carried out over the last 10 years had the diagnosis of MIP variant of hypospadias (5%). They outlined

the following surgical outcomes to include: (1) normal conical appearance of the glans, (2) a urethral meatus of normal calibre, and (3) a single, normal urinary stream with no symptoms.

anatomic characteristics of the hypospadias variant present a unique challenge to surgeons. The dissection of the wide meatus and urethral plate may result in thin glanular wings that are more prone to dehiscence and urethral fistula formation. The TIPU technique allows for better dissection of the glanular wings because the lateral aspects of the urethral plate are not dissected as they are with the GAP and pyramid procedure, leaving thicker glanular wings for glanular reconstruction. But the TIPU is required only when the urethral plate is narrow. Whereas in MIP, there is wide urethral plate and hence no need of incision here. Hence, we opted for TUPU because 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 TUPU restore the normal urethral and penile anatomy, 5,7,23

The controversies continue whether to operate these patients or not, however Duckett and Keatingwere were among the first to recognize the distinct surgical challenges presented by the MIP variant. Dissatisfied with the results of the meatal advancement and glanuloplasty (MAGPI) and perimeatal-based flap procedure, they described the "pyramid procedure." Yet another technique designed to overcome the challenges of a wide, deep glanular groove and a noncompliant fish mouth procedure is the glans approximation procedure (GAP). 1318

Another characteristic that makes MIP a surgical challenge is that most of the affected patients have been circumcised, the penile skin is usually thin and scarred, and are thus lacking prepucial and dartos tissue to be used for reconstructive procedures. However, a study by Snodgrass proved that circumcision does not affect the results of TIPU in selected patients with MIP. 713

The anatomic characteristics of the MIP hypospadias variant present a unique challenge to surgeons. The dissection of the wide meatus and urethral plate may result in thin glanular wings that are more prone to dehiscence and urethral fistula formation. The TIPU technique allows for better dissection of the glanular wings because the lateral aspects of the urethral plate are not dissected as they are with the GAP and pyramid procedure, leaving thicker glanular wings for glanular reconstruction. But the TIPU is required only when the urethral plate is narrow. $20^{\circ}23$

Surgical reconstruction of megameatus involves distinct challenges, as the distal urethra is widely



splayed, with a deficiency of spongiosum resulting in very thin, relatively immobile tissue on the ventral portion of the urethral meatus, therefore a variety of techniques have been described for and applied to megameatus repair, including the 'pyramid' procedure and tubularized incised plate (TIP) urethroplasty, Yuval Bar Yosef et al 37 routinely use aspects of each technique in their approach, which is based on patient-specific anatomy and The surgeon who attempts to repair hypospadias in a circumcised boy is therefore faced with the additional challenges that stem from local scarring following secondary healing processes as well as limited availability of skin and dartos tissue. 7'36'37 In a study by Shouxing Duan et al they found out that there were no significant differences in intraoperative bleeding, hospital postoperative analgesia rate, or cure rate among the three groups (P>0.05). Comparing the Mathieu repair with the TIP or Duplay procedures, statistically significant differences were observed in operation time (P0.05), and they found out that Urinary fistula occurred as early as 2 weeks after

surgery, and meatal stenosis occurred at 48 weeks after surgery. Figure 1 shows the Kaplan-Meier estimated overall complication-free survival. The primary estimated complication-free survival at 2, 3, 8, and 48 weeks was 96.0%, 92.0%, 87.8%, and 80.5%, respectively. Primary estimates of Mathieu, TIP, and Duplay complications were 80.0%, 84.6%, and 85.7%, respectively. ^{37'38'39}

COCLUSION

The tip and tricks of the surgical procedures of megameatus intact prepuce rare variant of hypospadias is a review article aimed to improve on the surgical outcome including normal conical appearance of the glans, a urethral meatus of normal caliber, and a single, normal urinary stream with no symptoms and to minimise the complications such as urethrocutenous fistula as extracted from the expert's vast experience and modifications of the existing techniques, in our study we summarised the experts seven established techniques and the modifications to achieve the desired objectives.



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