

## Surgical Options in the Management of Megameatus Intact Prepuce Review Article

Muhammad Ujudud Musa<sup>1\*</sup>, Abdulkadir Abubakar<sup>2</sup>, Bashir Yunusa<sup>2</sup> and Sadiq Abubakar<sup>3</sup>

<sup>1</sup>Department of Surgery, Federal Medical Center Katsina, Katsina State Nigeria

<sup>2</sup>Department of Surgery, Aminu Kano Teaching Hospital Kano, Kano State Nigeria

<sup>3</sup>Department of Surgery, Usman Danfodio University Teaching Hospital Sokoto, Sokoto State Nigeria

**\*Corresponding author:** Muhammad Ujudud Musa, Department of Surgery, Federal Medical Center Katsina, P.M.B 2121, Katsina State, Nigeria. Tel: +234-8036005365; Email: ujudud@gmail.com

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### Abstract

**Introduction:** Surgical options in the management of megameatus intact prepuce depends on various scenarios and its multifactorial hence the different surgical options with different outcomes, Hypospadias is a spectrum of different variants and megameatus intact prepuce (MIP) is a rare variant of glandular hypospadias seen is about 3-6% of cases of hypospadias.

**Materials and Methods:** We search the following sites Google Scholar, PubMed Central (PMC), PubMed, AJOL and EMBASE for articles on Megameatus intact prepuce, and the last search was on 21<sup>st</sup> July, 2019, using the keywords surgical management options of megameatus intact prepuce and the relevant articles were reviewed to extract the surgical options of managing megameatus intact prepuce variant of hypospadias from the experts.

**Results:** The Surgical Management of MIP is aimed at restoring function and cosmesis and several surgical techniques were described, including the glanular approximation procedure (GAP), the pyramid procedure, cutaneous advancement procedure, Tubularised incised plate (TIP), Tubularised Urethral Plate Urethroplasty (TUPU), Mathieu technique, and the subcutaneous frenulum flap with many modifications.

**Discussion:** MIP is rare and was first described by Juskiewenski and co in 1983, so many researchers reported various surgical options of treatment reporting different outcomes, in 2011 Elbatarny et al described GAP in seventeen patients with MIP over a period of 5years with an excellent outcome in 14 patients and 2 patients had a subjective score of 1 while one patient expressed dissatisfaction with the outcome, M. Sanal et al reported an excellent outcome in 7 cases with GAP.

**Conclusion:** The surgical management of options of megameatus intact prepuce are many with various modifications and outcomes and hence the need for a urologist to get acquainted with as many techniques as possible so as to achieve a desired goal in managing this category of patients with rare anomaly.

**Keywords:** Megameatus intact prepuce; Outcomes; Surgical Management Options

### Introduction

Surgical options in the management of megameatus intact prepuce depends on various scenarios and its multifactorial hence the different surgical options with different outcomes, Hypospadias is a spectrum of different variants and Megameatus Intact Prepuce

(MIP) is a rare variant of glandular hypospadias seen is about 3-6% of cases of hypospadias although this percentage is just a tip of an iceberg, due to the fact that some of the cases of megameatus intact prepuce may not present to the hospital, the diagnosis is missed initially or the case is not considered as clinically relevant or significant and hence under reported [1-3]. The term hypospadias is defined as an abnormal ventral urethral opening short of the normal glandular opening, the abnormal opening could be found along the penis, the scrotum or in the perineum and the more

proximal the opening the more severe is the hypospadias and the more the patient will present with ventral chordee, hooding and abortive corpus spongiosum and other associated anomalies such as inguinal hernia, undescended testis, congenital heart disease and other VERTALS associations [3,4].

There is paucity of review articles on this variant of hypospadias and its surgical management options, in 1983 the first description of Megameatus intact prepuce was done by Juskiewenski, et al. [2] and in 1989 Duckett and Keating described the presentation and surgical management of Megameatus intact prepuce named pyramid procedure [5]. In 2011 Elbatarny et al described modified Glanular Approximation Procedure (GAP) in surgical management of seventeen patients with Megameatus intact prepuce with an excellent outcome in fourteen patients with a subjective scores ranging from 2-3 [6]. The etiology of Megameatus intact prepuce is not yet fully understood with various theories put in place to explain it, however it is an interplay of the environmental, genetic and endocrinological factors that are found to be responsible for the development of Megameatus intact prepuce, the aim of treatment of MIP is to restore functional capability of the patient, to improve the cosmetic outcome and to eliminate the psychological disturbance to the patient, parents and relatives due to this abnormality [3,7].

The aim of this article is to review the surgical management options of these rare variant of hypospadias called megameatus intact prepuce.

### **Embryology**

As the genital tubercle enlarges and differentiates there is simultaneous elaboration of the male urethra, all these happened around the fifth and sixth gestational age under the influence of testicular androgens, the urethral groove deepens on the ventral part of the phallus and there is enlargement of the urethral folds which are located on either sides of the urethral groove, the urethral fold fused starting from the base of the phallus and moves distally to the glans, therefore hypospadias occurs from incomplete fusion of the urethral folds causing an incomplete urethra and incomplete or hooded foreskin. In the case of Megameatus intact prepuce variant of hypospadias, the formation of the prepucial skin is usually normal although the distal part of the urethra is wide open. Persistent splitting of the distal, urethra ventrally usually cause disunion of the glans of the phallus with consequent formation of an abnormally large meatus associated with a deep glans cleft, the glanular urethra generally become wide at the level of the corona and tapers downwards to become a normal caliber in the distal penile urethra [8-11].

### **Etiology**

The Etiology of Megameatus is not fully identified however

several theories were put through to explain that, there are basically three factors that are incriminated, they included environmental, genetic and endocrine. The environmental factors are found to increase the risk of developing MIP especially with the rising incidence of hypospadias in the past two decades this is evidenced by raised concerns of increased in utero exposure to progesterones, estrogens, or other antiandrogens such as phytoestrogens. However, despite the increased incidence of the anomaly, In a study of 130 boys with hypospadias 11 mothers gave a history of having ingested oral contraceptive pills inform of progestins, alone or in combination with estrogen, so also the exposure to the pesticides, fungicides and industrial pollutants and it is also found out that in early pregnancy Offspring of mothers treated with DES have been noted to have an increased risk of hypospadias, it is also reported that there is about fivefold increased risk of hypospadias in the boys conceived by *in vitro* fertilization, which might indicate disturbance of the maternal-fetal endocrine milieu by the progesterone therapy that accompanies assisted reproductive technology [12-14].

Genetic factors are found to be involved in the development of hypospadias since 7% of cases are seen in the first-, second-, or third-degree relatives. The probability that a male sibling of an affected boy will have a hypospadias is 9-17%. Hypospadias are equally transmitted through the maternal and paternal sides of the family, which is about 57-77%. Hypospadias have been described in over 200 syndromes. The two most well-known are the Wilms' tumor, aniridia, genitourinary malformations, and mental retardation (WAGR) and the Denys-Drash syndrome (genitourinary malformations and susceptibility to Wilms' tumor) [15,16]. The endocrine factors include Androgens and estrogens imbalance, causes various within the spectra of congenital penile anomalies such as hypospadias micropenis, and ambiguous genitalia, it is also found that reduced anogenital distance in boys with hypospadias occurs as a result of disruption of prenatal androgen exposure [17,18].

### **Clinical Features**

The diagnosis of Megameatus intact prepuce is mainly clinical, the pathology is found in the neonates during circumcision or later in life in non-circumcised boys whenever the prepuce is retracted, they don't present with chordee, they can present with a different variation in the appearance of the urethral meatus with dilated meatal opening or may present with a large fish mouth opening which may expand down or just below the coronal margin. The patients who present with the more enlarged forms of the meatus may present with a splayed urinary stream. No other urologic anomalies have been associated with MIP variant of hypospadias and no radiologic evaluation is needed in the absence of any other symptomatology, MIP can be classified into glanular, coronal, Subcoronal and distal penile [19-21].

## Material and Methods

We search the following sites Google Scholar, PubMed Central (PMC), PubMed, AJOL and EMBASE for articles on Megameatus intact prepuce, and the last search was on 21<sup>st</sup> July, 2019, using the keywords surgical management options of megameatus intact prepuce and the relevant articles were reviewed to extract the surgical options of managing megameatus intact prepuce variant of hypospadias from the experts, articles including case reports, case series, original research articles and review articles were used without restriction on the publication date and hence 43 relevant studies were appraised and summarized for the purpose of this study.

## Results

The Surgical Management of MIP is aimed at restoring function and cosmesis and several surgical techniques were described in the repair of Megameatus intact prepuce, including the Glanular Approximation Procedure (GAP), the pyramid procedure, cutaneous advancement procedure, Tubularised Incised Plate (TIPU), Tubularised Urethral Plate Urethroplasty (TUPU) and Mathieu technique with many modifications, these are some of the surgical treatment options described by various studies some of which are case reports, original review articles and some are case series.

### The Granular Approximation Procedure (Gap)

This entails making a U- Shaped incision around the megameatus and the urethral plate, then the glanular wings are raised with this the urethra plate can now be assessed and the excess redundant urethral plate is trimmed so as to make it the same diameter with the proximal urethra, in some cases there is need to deglove about 1 cm of the penile skin so as to allows for harvesting the intermediate layer from island flap in uncircumcised patients or from the dartos fascia or Bucks fascia in circumcised patients which will be used to reinforce the repair as well as to eliminate the tension on the meatus, usually vicryl 4/0 - 6/0 is used to do the urethroplasty over a Foleys catheter, 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 [22-26].

### The Tubularised Urethral Plate Urethroplasty (Tupu)

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 [27-30].

### The Pyramid Procedure

In this procedure a tennis racket incision is made around the edges of the megameatus beside the groove of the glans down to the level of corona, then the urethra is mobilized up to the apex of the pyramid, this is followed by deepening the incision on the edges of the glanular groove so as to develop a glanular wings from the urethral plate, while the distal urethral plate is left intact dorsally, then a small wedge is taken from the ventral tissue and the distal urethra is sutured in continuity with the urethral plate, now the urethra and the glanular wings are Tubularised over a stent to form the neourethra, then glanuloplasty is done [5,31-34].

### The Mathieu 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 [35-37].

### The Tubularised Incised Plate (Tip)

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 edges 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 so as to cover the second layer [38-41].

### The Subcutaneous Frenulum Flap (Scuff)

The Patient is prepared an informed consent obtained under general anesthesia, a circummeatal incision is done and the penis is

deglove, then glanular wings are raised and the redundant mucosa trimmed the urethra is tubularized over a 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 [42].

## Discussion

Megameatus intact prepuce is a rare variant of hypospadias which was first described by Juskiewenski and co [2] in 1983, so many researchers reported various surgical options of treatment reporting different outcomes, considering the fact that various surgical options have their own challenges and prospects, In 2011 Elbatarny, et al. [6] described modified Glanular Approximation Procedure (GAP) in surgical management of seventeen patients with Megameatus intact prepuce over a period of 5years, were they achieved an excellent outcome in 14 patients and 2 patients had a subjective score of 1 while one patient expressed dissatisfaction with the outcome, M. Sanal, et al. [10] reported an excellent outcome in 7 cases following glanular approximation procedure in managing their patients.in another study by George, et al. [25] in Children's Mercy Hospital in Kansas City, Missouri in a duration of 5-years were they reported an excellent outcome in all 37 consecutive cases of MIP using Glanular Approximation Procedure (GAP) however one of the patients had Urethrocutaneous fistula as a complication. Amilal, et al. [21] reported the outcome of Tubularised Urethral Plate Urethroplasty (TUPU) in 13 cases of MIP in a retrospective study between June 1996 - June 2015 were operated on 1026 cases of Hypospadias with 13 patients had an excellent result from Tubularised Urethral Plate Urethroplasty (TUPU). Won Hee, et al. [43] reported a ten years' experience with the treatment of 21 adults with hypospadias and 3 out of the 4 cases of MIP had Pyramid Procedure with excellent results, Marc Cendron [1] reported retrospective study of 481 cases of hypospadias repairs between 2007 to 2017 in the Boston Children's Hospital, Harvard Medical School, Boston, MA, United States and reported 25 cases of MIP out of which 10 patients Tubularization of the Urethral Plate Urethroplasty (TUPU) and 6 of the patients developed Urethrocutaneous fistula while 15 patients had Mathieu technique using a vascularized inframaleal flap with an excellent results. Yuval Bar-Yose, et al. [27] reported the outcome of 24 cases operated Tel-Aviv Sourasky Medical Center, Sackler Faculty of Medicine, Tel-Aviv University, Tel-Aviv, Israel, with good cosmetic and functional results in 20 patients with 4 patients plus 1 patient who underwent GAP and meatoplasty and 3 of the 11 patients who underwent TIP urethroplasty, of whom 2 developed a urethrocutaneous fistula. Steven GDocimo [42] reported 5 cases from University of Pittsburgh School of Medicine, Children's Hospital of Pittsburgh, Pittsburgh, Pennsylvania, USA using Subcutaneous Frenulum Flap (SCUFF) were two reoperative hypospadias repairs, one primary megameatus repair, and two

repairs of traumatic fistula and megameatus after circumcision. With an excellent outcome and one case of urethrocutaneous fistula

## Conclusion

The surgical management of options of megameatus intact prepuce which is the variant of hypospadias are many with various modifications and outcomes and hence the need for a urologist to get acquainted with as many as possible so as to achieve a desired goal in managing this category of patients with rare anomaly.

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