Research Article

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Familial Infantile Inner Megaprepuce in Three Members of a Family

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Abstract

We present our reconstruction innovative unique technique for correcting the familial infantile inner megaprepuce in three cases, all being members of a family. Three members of a family, father and two identical twins, had infantile inner megaprepuce (IIMP). This was corrected using a technique combining penoscrotoplasty, prepucioplasty, inner megaprepuce partial or hemicircumcision and frenuloplasty. Father had IIMP as an index patient and his identical twins developed the same conditions. All three had undergone the same procedures uneventfully. We wish to describe a simple, safe, effective, minimal invasive, innovative technique in three patients from the same family with excellent, consistent, cosmetic, functional and patient/parental satisfaction results without complications. These simple techniques can be applied across a wide range of ages and other anomalies such as buried penis and penoscrotal transposition with penoscrotal web, etc. and is easy to be repeated by others.

Keywords: Congenital; infantile; familial; frenuloplasty; hemi-circumcision; meatoplasty; megaprepuce; partial circumcision; penoscrotoplasty; preputioplasty.

Introduction

Congenital abnormalities of the kidneys and the urinary tract are the most common sonographically identified malformations in the prenatal period [1]. However, it is not possible to diagnose IIMP prenatally or perinatally in the neonatal period as it is not congenital but appears later in infancy. True infantile inner megaprepuce (IIMP) is a striking obvious distinct emerging new unique entity characterized by a capacious preputial sac with extensive redundancy and ballooning of the inner prepuce like a preputial bladder, phimosis with preputial meatal stenosis, and a relative deficiency of penile shaft skin. Clinically it is difficult to hide or miss for diagnostic purpose but has challenging surgical management which we describe with simple, safe and very effective technique based on the precise embryo-pathological anatomy. IIMP should not be confused with a buried, concealed, webbed, trapped or micropenis. It has been reported as congenital megaprepuce [2] but we prefer to call it infantile inner megaprepuce as all babies are born normal at birth and the outer foreskin is normal. This condition is rare in itself and no cases of familial occurrence have so far been reported. We present a family of three including father and a set of identical twins having IIMP which may have genetic implications.

Case Histories

Case 1

Father, a 24-years-old male, born at term by normal vaginal delivery and had no concerns at birth. However, in infancy around 11 months of age developed painless gradually increasing ballooning of the foreskin associated with dribbling of urine and no definite stream. As the patient was wetting and worsening clinically, parents took him to their family general practitioner who referred the patient to a general surgeon in the nearby town. Preputial meatal dilatation was carried which allowed urine to be drained easily in the initial period but the condition started recurring again.

The patient was taken to a surgeon with urology special interest at district town. A dorsal slit procedure allowed to relieve tight phimosis, drained urine and decompress the swelling but cosmetic result was not so good due to hanging dog ears like wings and wrinkled redundant penile and scrotal skin.

The parents were concerned about the cosmetic appearance and the general surgeon referred him to one of his plastic surgical colleagues who did some flaps and tried to improve the cosmetic outcome at the age of 18 months. The outcome was certainly better than before but not satisfactory due to spraying of urine and poor cosmesis.

At the age of 4 years, the parents were still concerned about the appearance of the penis, spraying of the urine at micturition from the functional point and they were prepared to have normal circumcised appearance rather than such ugly situation despite three previous surgical interventions. Therefore, the parents brought him to us on a recommendation from one of our buried penis with scrotal transposition patient's parents in whom we have used this technique. This case was very much of the IIMP, but the challenge was that the patient has undergone several procedures earlier with scarring and the loss of tissue planes. The opportunity was to demonstrate that our technique is applicable, feasible and effective for revisional surgical case so we decided to go ahead with it using our innovative plastic pediatric surgical and urological unique technique.

Due to scarring at the previously operated site some oozing was more and the tissue planes were blurred as compared to a fresh case and took some more time. However, with patience and gentle dissection were able to separate the redundant inner megaprepuce which was completely excised and the outer prepuce skin was all preserved. The penile erection during anesthesia allowed us to accurately tailor the inner mucosal sleeve and outer preputial skin with subsequent closure using absorbable subcuticular stiches. The penoscrotal and median raphe were left untouched making it easy to dissect and repair in the middle and lower portion with well-preserved normal anatomical tissue planes.

The post operative period showed more pain, bladder spasms requiring anticholinergics, antispasmodics and muscle relaxants to control it, reactive swelling and extra hospitalisation as compared to early first-time surgery. The patient recovered well with satisfactory anatomical, functional and cosmetic outcome as compared to excellent outcome in a virgin case at younger age.

Case 2

The index patient got married, had first set of identical twins born which were completely normal at birth and initial infancy. However, at the age of 10 months both presented with con-

dition similar to their father and brought to us by parents. Twin1 had massive swelling below the tip of foreskin obscuring penis completely and appeared like another bladder. The patient cannot micturate with a stream, dribbled a little and needed to squeeze to drain urine out of the preputial bag.

On examination, infant had a scrotum continuous with a preputial sac filled with urine and the tip of the foreskin meatus at the top of it like a volcano effect (Figure: 1A).

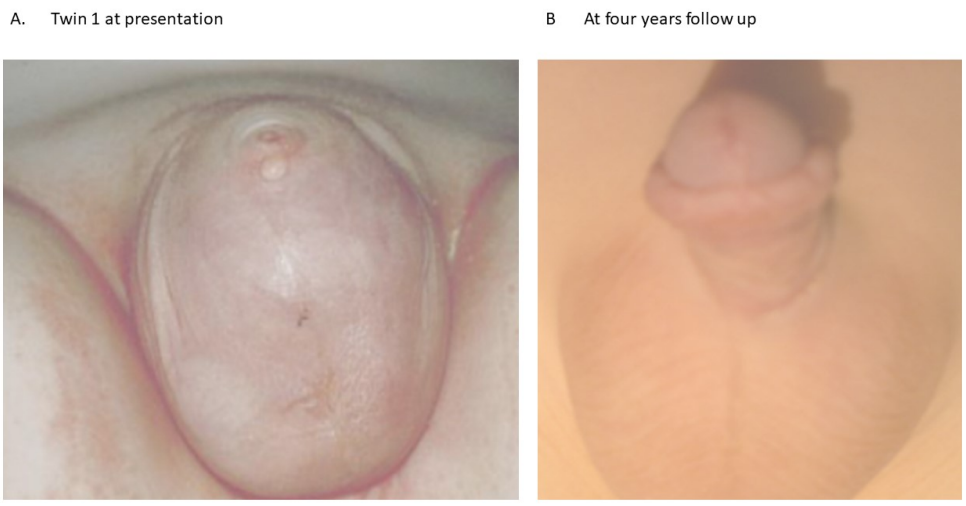

Figure 1: Twin 1 images A. at presentation B. at 4 years follow up

Clinically transillumination showed brilliantly transilluminating swelling filled with clear fluid (Figure: 2A). Examination under anaesthesia confirmed a massive bag of inner preputial sac with no visible but easily palpable normal penis and a narrow preputial opening expressing urine and few plaques of smegma. The operative findings were as displayed in (Figure: 2B).

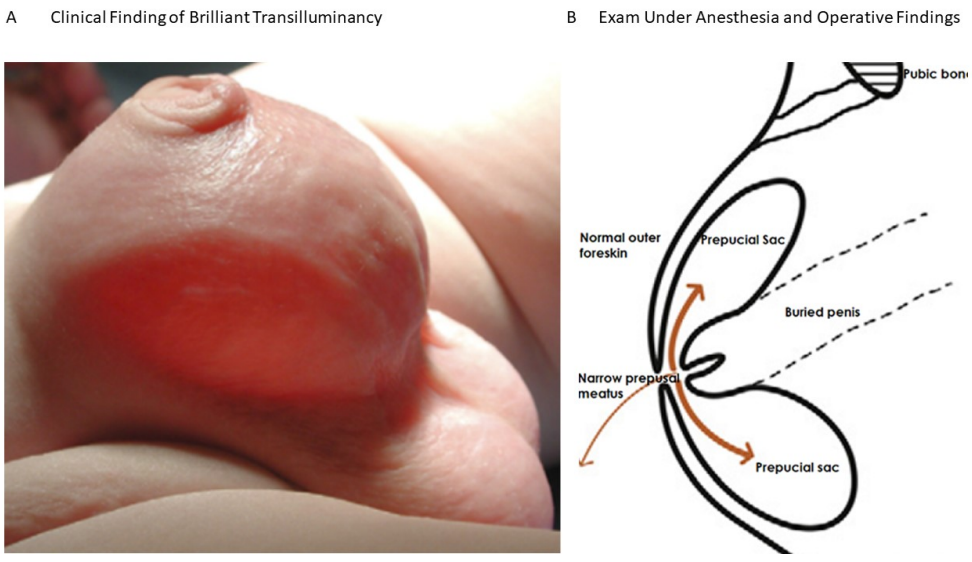

Figure 2: Clinical findings A-Transillumination test B Examination under anesthesia findings.

The operative procedures included penoscrotoplasty by a transverse penoscrotal incision made at the junction of penile and scrotal skin to be closed vertically later on (Figure 3A and 3C), The median raphe of the ventral midline incision starting from this transverse incision and extending up to the preputial meatus was made. It was continued as circumferential muco-cutaneous junction incision all around in the inverted racket form to divide inner and outer foreskin and relive

the preputial meatal stenosis as Y-V prepucioplasty. The dissection carried down to the level of Buck's fascia (Figure. 3B). The dartos was mobilized all round just below the glans to allow the normal outer preputial skin to slide back to mobilise the surrounding skin for separating secondary penoscrotal fusion and slide the outer skin back to clearly separate the penile shaft and the scrotum. Leaving a mucosal cuff just below the corona, the large and redundant inner preputial sac was excised. Hemostasis was confirmed, partial hemi-circumcision completed with frenuloplasty protecting the glans and the preputial branches of the blood vessels. The shaft of the penis which was of normal length was covered with the outer foreskin and dorsal penile skin using absorbable subcuticular stiches (Figure. 3D). In a true case of inner megaprepuce release of the penoscrotal junction by a transverse penoscrotal incision and then repairing vertically like Heine-Mikulicz pyloroplasty lengthens penis sufficiently in most cases but if it is associated with scrotal transposition, buried penis or other pathology we apply fixing sutures at penoscrotal junction ventrally and dorsal aspects but if it is pure inner megaprepuce, it is generally not necessary An 8-French silastic urethral stent was placed as a dripping stent with Cavicare dressing and double nappy for easy post-operative care at home in this day case surgery cases.

Post-operative period was uneventful, the dressing and stents were removed in one week. Post-operative follow-up showed excellent anatomical, cosmetic & functional result (Figure. 3E).

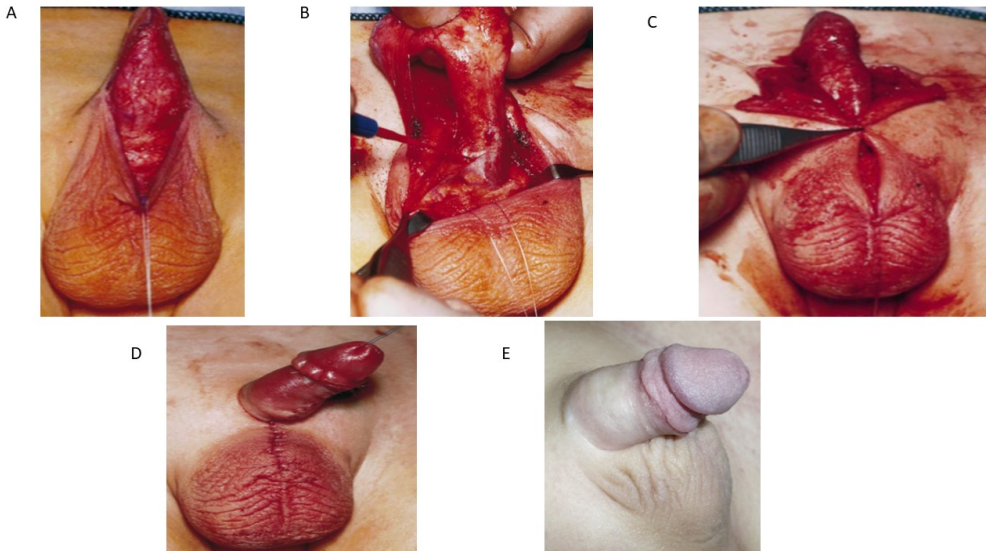

Figure 3: Operative findings – A. Transverse penoscrotal incision B. Median raphe and circumferential incisions C. Vertical closure of penoscrotal incision D. Post semi-circumcision, E. at follow up.

At the follow up at 4 years, the parents and patients were fully satisfied at the circumcised appearance with full length of penis exposed (Figure 1B).

Case 3

Twin 2 developed painless gradually increasing ballooning of the foreskin associated with dribbling of urine and no definite stream (Figure. 4A).

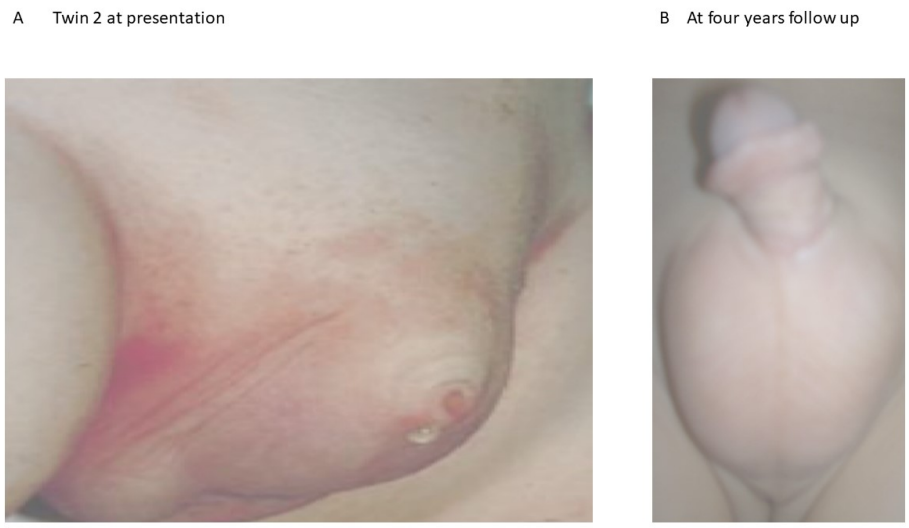

Figure 4: Twin 2 images A. at presentation B. at 4 years follow up.

On examination, infant had a scrotum continuous with a preputial sac filled with urine and the very tight phimosis at the tip of the massive swelling dribbling urine. On compression of the swelling urine with few specks of smegma can be expressed.

The patient had classic transillumination test positive clinically and had similar findings at examination under anesthesia to that of case 2 above (Figure 2A and 2B).

The patient underwent our special unique innovative technique described in the case 2 above as a day case together with his twin brother. The swelling was comparatively smaller and the redundant inner megaprepuce was less redundant and took less time. The procedure was uneventful and patient was discharged home the same day.

The post-operative period was uneventful and the silastic draining stent and Cavicare foam dressing was removed at one week as ward attendant. At 4 years follow up, the parents and patient were fully satisfied with cosmetic, functional and anatomical result with full length of penile shaft exposed and post circumcision appearance of the penis (Figure. 4B).

The parents and both twins were offered genetic counselling and investigations with sequencing of relevant genes but the request was declined as both boys were procedure and needle phobic, the family was very satisfied with the outcome, father did search on the genetic aspect of this condition and could not find any literature and especially in light of the present cost of genetic analysis.

Discussion

True IIMP is a unique condition which has been previously labelled as congenital megaprepuce (CMP) and erroneously classified as a type of buried penis [3]. It was first described in 1994 and named congenital megaprepuce [4]. However, the aetiopathogenesis and the long-term outcomes of correction of this condition remains to be established. Embryologically, the inner foreskin is equivalent to female vagina and have estrogen receptors with a potential to expand to enormous size as seen in massive hydrocolpos in cases of cloaca with narrow single outlet with partial obstruction and under the influence of estrogen hormone. IIMP may have similarities with distal preputial stenosis with phimosis causing partial obstruction under the influence of hormonal effects. The incidence is increasing probably due to exposure to estrogen in the fetal life.

It typically presents with massive swelling, difficulty voiding, often requiring manual expression of urine. Early surgical correction is recommended for parental cosmetic concerns to prevent complications of urinary stasis, infection, smegmaliths, etc and address parental concerns [5]. Considering it as simple phimosis and performing traditional circumcision should be avoided at all costs as it can create further deficiency of the outer foreskin and invite disaster with redundant inner foreskin lined with mucosa and may need complex revisional reconstructive surgery with poor cosmetic and functional results.

It is very essential to understand the difference between a full circumcision and a partial hemicircumcision. We consider a

hemicircumcision not in horizontal or transverse plane similar to that of traditional full circumcision but in the vertical plane separating the outer and inner preputial skin and resecting the inner foreskin only. Strictly speaking this is not a partial circumcision in the craniocaudal transverse direction but a vertical hemi-circumcision. That means to separate the outer skin and the inner megaprepuce at the junction of the mucocutaneous area circumferentially at the very tight phimosis at top and continue the separation in the vertical dimension. Continue separation till the subglanular groove level and then only resect the redundant sac of the inner megaprepuce preserving all deficient outer foreskin and penile shaft skin cuff. This is followed by stitching the inner megaprepuce mucosal cuff rim to the entirely preserved outer skin cuff margin at the base of the glans penis by subcuticular absorbable stiches for a perfect normal circumcision like appearance leaving the glans penis completely bare. This is acceptable to general public as circumcision which is being done for a variety of reasons. The problem with the partial circumcision in the transverse plane cutting both the inner and outer foreskin is that it will excise the outer skin excessively and then the distal shaft will have to be covered with inner megaprepuce mucosal cuff which does not give a good cosmetic and functional result as it secretes mucous and there are chances of malignancy to develop in the mucosal cuff covering penile shaft at a later date.

Since its introduction several surgical techniques have been described for its treatment in the quest for searching the best procedure. Surgical treatment is challenging and controversial. Different general, pediatric, urological and plastic surgical techniques have been described; however, optimum results have been difficult to achieve. Techniques such as preputial dilatation, dorsal slit, the inner preputial covering of the penis, Byar's flap, etc. have been reported with varying levels of unsatisfactory outcomes [6].

We certainly use Y-V preputioplasty for tight phimosis and multiple Y-V preputioplasty for the minimal invasive surgery for BXO cases and is one of the alternative options of a case of pure phimosis but in cases of IIMP phimosis is one of the elements to be addressed for which we have used this Y-V plasty at the junction of median raphe and the circumferential incision to correct the phimosis part at the top but then added partial hemi-circumcision for removing the excess redundant massive sac of inner megaprepuce and preserved normal but deficient outer skin to cover the penile shaft with skin cover as the length of penile shaft has increased following release of penoscrotal web at the penoscrotal junction by transverse incision and vertical closure. In order to address the redundant sac of inner megaprepuce, partial hemicircumcision of the inner megaprepuce only leaving the outer skin lined prepuce intact will allow covering the penile shaft with excellent cosmetic and functional result.

The principles in the IIMP reconstruction reported here include separation of penoscrotal fusion, follow the midline raphe to avoid any additional scar and correct the preputial stenosis using a Y-V plasty at the tip of the preputial stenotic opening and perform a partial hemicircumcision of the inner half of the mucosal layer only and perform frenuloplasty at the penile shaft-granular junction for covering the meatal branch of the preputial artery to complete the procedure. This technique provides the ability to create the appearance of a standard circumcision, giving the best cosmetic and functional appearance.

The beauty of this technique is virtually no chance for any urethral injury as this is a very superficial subcutaneous level surgery and the trick to avoid urethral injury is to mark the skin incisions transverse at penoscrotal junction and vertical along the median raphe first. Then start the transverse incision on the lateral aspects first and develop the subcutaneous plane of dissection and carry it medially and raise the subcutaneous plane around the median raphe followed by starting the vertical component of the incision at junction of T and gently carry it upwards till the mucocutaneous junction from where it is carried out circumferentially to separate outer preputial skin and inner megaprepuce mucosal sleeve. The outer skin is all preserved and the inner mucosal sleeve is resected leaving the small area just below the corona and the outer skin is attached to this mucosal cuff with subcuticular stiches to complete hemicircumcision resecting the inner megaprepuce and preserving the outer skin for a perfect post-circumscribed appearance. The key to avoid any urethral injury is to use a urethral stent and remain in subcutaneous plane all the time which avoids bleeding on one hand and the injury to strong deep Buck's fascia and the corpora underneath.

We traditionally use small silastic urethral stent and foam compression dressing for all our penoscrotal extensive surgery like hypospadias, inner megaprepuce, penoscrotal fusion with scrotal transposition, buried penis, etc as day care cases. This helps the post operative care at home by leaving dripping stent between two nappies to ease post-operative

care by parents/carer. In addition, it allows healing without any urine contamination and avoid any bleeding caused by urokinase to dissolve any blood clots at the healing interphase as a routine, The use of urethral stent is an optional one and can be done without a stent. However, if there is retention of urine in the immediate post operative period, catheterising may require another anesthesia and may be difficult.

Recently a systematic review of this rare challenging condition with a controversial surgical management indicated that some series have in the past included different variants of inconspicuous penis combining concealed penis, secondary acquired megaprepuce, penile transposition, and webbed penis. Techniques described in the last 26 years mostly with small number of patients and short follow-up [7]. We have, therefore, accurately given proper name as IIMP and corrected pure form of true IIMP using simple but effective technique.

Miniseries of cluster of familial IIMP with strong family history and recurrence in the offspring bring possible genetic influence. Congenital malformations often have a genetic background associated with a recurrence risk and may be part of a syndrome [8]. IIMP is not congenital in strict sense but similar to vascular anomalies and infantile hypertrophic pyloric stenosis which may have psycho-neuro-hormonal and environmental axis [9-10]. In most cases of IIMP, cause can be monogenic, but in our familial cases it may be caused by both genetic and environmental factors. In the case of IIMP, genetic predisposition is poorly characterised, is more common in monozygotic twins and in offspring of fathers with IIMP. Risk of affected second male child for IIMP could be 1 in 8 if no family history, 1 in 5 if one other male relative affected, 1 in 4 if two other male relatives affected.

Conclusion

Familial IIMP may have genetic implications. Early surgical correction is recommended to prevent complications. Our unique innovative minimal invasive technique of correction by combining four basic plastic surgical procedures of penoscrotoplasty to redefine distinct penoscrotal junction and release hidden penis, Y-V prepucioplasty to resolve preputial stenosis, partial hemicircumcision of the redundant massive bag of inner prepuce and preserve all outer prepuce skin to cover the shaft and frenuloplasty at shaft-glans junction is easy, safe, effective and can be applied to similar lesions.

Compliance with ethical standards

Acknowledgment

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Conflict of interest

The authors have no conflict of interest to declare. No funding source was involved in this study.

Ethical approval

All procedures performed on human participants were in accordance with the ethical standards of the institutional and national research committee and with the 1964 Declaration of Helsinki and its later amendments or comparable ethical standards.

Informed consent

Informed consent was obtained from the parents and all the relatives involved prior to all the procedures. Parents and all involved parties were informed about the procedure.

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