ORIGINAL RESEARCH PAPER

Paediatric Surgery

MANAGEMENT OF MICROPENIS IN CHILDREN

KEY WORDS: Micropenis, surgery

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ABSTRACT

Congenital and acquired micropenis in children are managed by medical and surgical intervention. The goal of management in these patients is to restore a functional penis size in order to allow normal standing micturition,psychological well being, improve patient quality of life and to ensure satisfying sexual intercourse in future . We treated 45 childrens with micropenis by medical and surgical measures with excellent results both in short and long term follow up.

PENIS DEVELOPMENT

During embryologic development, gender remains indifferent until approximately seventh week of gestation. From the eighth week onward, male and female developmental pathways diverge. In males, maternal chorionic gonadotropins from the placenta stimulate growth and development of the testicular Leydig cells, which eventually produce their own testosterone for further development (1).The development of the external genitalia in males is dependent on the conversion of testosterone to the active component dihydrotestosterone (DHT).Testosterone is converted to DHT locally by 5 α-reductase and then directly acts on androgen receptors to initiate cell signaling pathways [1,2].

The genital tubercle enlarges to form both the shaft and glans of the penis. The urogenital folds fuse along the midline to become the penile urethra and the glans invaginates to create the glandular urethra.The labioscrotal folds fuse to create the scrotum. Complete penile differentiation should be complete by the end of the first trimester. Penile length and size increase during development in proportion to other fetal development (5). The average penile length increases by a mean of approximately 2 cm between 14 weeks and term. Penile elongation in utero is complete by androgens produced by the fetus. After the first trimester, the fetus depends on his own hypothalamic-pituitary axis for gonadotropin production. Any abnormality resulting in hypogonadism in utero can result in an underdeveloped penis, and therefore congenital or 'true' micropenis (5,6).

After normal development, there is an LH surge at birth which stimulates an increase in testosterone production and penile growth. This surge only lasts about 12 h and subsequently hormone levels drop quite low. Slowly, gonadotropin and androgen levels begin to rise and peak again. This rise in levels lasts for approximately 6 months and allows for continued penile growth (7). Ongoing penile growth occurs throughout development, without the need for surges, but rather as a result of normal growth. At puberty, the HPG axis gets activated and stimulates testicular testosterone production, which subsequently leads to further penile growth into adulthood (7,8).

Penile length

Normal penile length

Accurate measurement of penile length is important for both clinical and academic purposes. For this reason, a standard practice for measurement has been defined to maintain consistency. Rather than measuring a flaccid penis, which offers limited clinical significance, stretched penile length (SPL) measurement is considered best practice for adult men as it most closely replicates normal erect penile length (9). SPL is the maximum length of the penis while stretched, measured from the base of the penis, under the pubic symphysis to the tip of the glans and is thought to approximate erect penile length within 10% (). For accurate clinical assessment, SPL needs to be measured by a clinician.

Micropenis

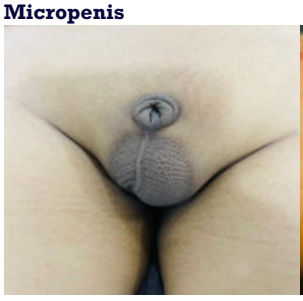

Fig 1. (Pre Operative)

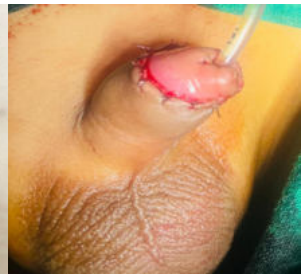

Fig 2. (Post Operative)

True micropenis is defined as a normally formed penis that has an SPL that falls below two standard deviations of normal for a patient's age and race ().Wiygul outlined the mean SPL in children as well as the diagnostic length for micropenis at each age (13). Normal values for preterm infants born between the 24th and 36th week of gestation can be calculated using the formula: (0.16 × weeks of gestation) – 2.27 (14).

Micropenis develops as a result of a central or local hormonal imbalance during fetal development. True micropenis is a congenital anomaly and is different from acquired penile length abnormalities such as buried penis or trapped penis. Based on the etiology of the hormonal dysfunction, micropenis can be divided into three broad categories: hypogonadotropic hypogonadism, hypergonadotropic hypogonadism, and idiopathic. Other, less common causes of micropenis have been documented . Disorders of sexual differentiation may present with micropenis, although hypospadias is more common (13). The coexistence of micropenis and hypospadias is termed "microphallus" (1).

Causes of Micropenis- Hypogonadotropic hypogonadism
Kallman's syndrome
Prader-Willi syndrome
Laurence-Moon syndrome
Bardet-Biedl syndrome
Rud's syndrome
Other pituitary hormone deficiencies
Hypergonadotropic hypogonadism
Anorchia
Poly-X syndromes (ex: Klinefleter)
Gonadal dysgenesis
LH-receptor defects
Noonan's syndrome
Trisomy 21
Robinow's syndrome
Laurence-Moon syndrome

Bardet-Biedl syndrome
Defects in testosterone action
GH/IL-GH deficiency
Androgen receptor defect
5-a Reductase deficiency
Fetal Hydantoin Syndrome
Developmental abnormalities
Aphallia
Cloacal exstrophy
Idiopathic
Other congenital malformations

Medical management-

Testosterone-

All 45 childrens between 2 years to 15 years underwent medical management with testosterone gel and injection testesterone enanthate monthly and hCG per week. In cases of true micropenis, the goal of treatment should be restoration of a functional penis size in order to improve body image and self-esteem, allow normal standing micturition . In children with true micropenis, the first step in management is always the least invasive, which includes the application of exogenous testosterone.

Multiple studies have explored intervention with testosterone replacement either early during development and/or at pubarche. If insufficient penile growth is not achieved with replacement, multiple courses of replacement can be considered without significant reduction in stature (12,13,14,15). In 2013, the beneficial effects of hormonal therapy on penile growth in children with micropenis was confirmed. In this study, prepubertal children were treated with 25 mg of exogenous parenteral testosterone enanthate once a month for 3 months, and pubertal or postpubertal children were treated with intramuscular hCG once a week for 6 weeks. Exogenous administration of hormone replacement in these boys resulted in a significant increase in SPL and suggests that these treatments could be the primary form of therapy for micropenis in paediatric patients (15). In addition to exogenous testosterone, topical applications have been studied in the micropenis population. Early studies revealed a 150% improvement in penile length with minimal side effects when DHT was applied locally (16).

The administration of exogenous testosterone in childhood does not compromise ultimate penile length increase in adulthood, however, the long-term effects of testosterone administration in childhood are still not fully understood and long-term data are needed [17]

Surgical management

Penile augmentation

Different types of injectable materials have been used for penile augmentation including liquid silicone, polyacrylamide, hyaluronic acid and mineral-oil (35,37). However, there is a significant risk of foreign body reaction, swelling, penile distortion, granulomas and need for removal (36). Autologous fat grafting has been described to increase penile length and girth, which has no foreign body reaction as it is derived from the body's own tissues. This is a much less invasive procedure compared to flap reconstruction or V-Y advancements. To obtain a fat graft, fat is liposuctioned from areas of excess, placed in 10 mL syringes and then centrifuged for 3 min at 300 g. The superior oil layer and lower aqueous layers are removed and the middle adipose layer is collected as the purified fat graft. The fat is transferred into smaller syringes for injected in multiple layers to improve fat graft survival. This technique has been refined by Sydney Coleman in recent years (21,22). The autologous fat graft will lose 20–80% of its volume over the first year of engraftment, and thus multiple procedures are sometimes necessary to achieve the desired result (23,25). Panfilov described his technique for penile augmentation with fat grafting in 88 patients. Incisions are made radially through the frenulum preputial and approximately 40–68 cc of fat is injected between the superficial penile fascia and the profunda, down to the root of the penis. The average length and circumference increase was 2.39 and 2.65 cm respectively, after 12 months (35). In one patient, the penis gained 3 cm in length at 6 months, but due to fat graft resorption, the stable length was 2 cm at 7 years. This technique can be combined with suspensory ligament release to further increase length. In Panfilov's series approximately 1/3rd of patients had the ligament release in addition to autologous fat grafting. Penile augmentation with fat grafts also increase the weight of the penis, which can itself increase the length by 2–3 cm (19). Dermal fat grafting has also been described to increase girth and length of the penis, which may have better fat retention and decreased contour irregularities at the expense of a larger donor-site scar (20).

Suspensory ligament release

The suspensory ligament anchors the penis to the pubic symphysis and while providing support, acts as the mobile point for the penis during erection. This attachment prevents the penis from moving further outward and creates an arched angle to the penile base (27). The suspensory ligament is composed of the suspensory ligament proper and the arcuate subpubic ligament that attaches the tunica albuginea to the midline of the pubic symphysis. Surgical release of this ligament changes the acute angle of the penis to the pubic symphysis to an obtuse angle which allows the penis to lie in a more dependant position and therefore gives the perception of lengthening (27). Division of the suspensory ligament, with or without bulking agent, fat pad excision or V-Y plasty is the most widely accepted surgical technique for penile elongation (28). All of our patients underwent this procedure without any untoward effects. The suspensory ligament can be accessed through a V-Y incision or a subcoronary circumcision technique (26). Complete release of the corpora from the pubic ramus has been described to further increase length, but is associated with significant risk to the neurovascular bundles of the penis, causing denervation and devascularisation of the penis (29).

small testicular prosthesis (30). Srinivas *et al.* have described the V-Y advancement with subsequent silicone sheath insertion between the pubis and the released suspensory ligament to prevent recurrence (31). After the inverted V-Y incision and suspensory ligament release, a silicone sheath from a penile prosthesis was inserted in the soft tissue defect created between the base of the penis and the symphysis pubis. They achieved a lengthening of 2.5 cm at 6 months. Dermal fat grafts have also been described to fill this space[31].

V-Y advancement

Penile elongation using a dorsal V-Y incision in the congenital or acquired short penis was first described over 40 years ago. A dorsal V-shaped incision was made, combined with partial detachment of the crura from the pubic ramis, which were then re-approximated in the midline and the dorsal incision was closed as a V-Y advancement flap.[36] The V-Y incision and subsequent V-Y advancement is commonly used in conjunction with a suspensory ligament release. The incision is typically an upside down V, which is closed in an upside-down Y-shape, which lengthens the dorsal skin by bringing lateral tissue to the midline. The flap is distally based, and poor wound healing, flap dehiscence and distal flap loss can occur if the flaps blood supply is compromised during dissection (26). Bulging of the penoscrotal transition can also occur, which can be treated using bilateral Z-plasties (19).

Suprapubic lipectomy

Suprapubic lipectomy has been performed to increase perceived penis length, particularly for patients with a buried penis. In these patients, weight loss does not always reduce

the problem of a large overhanging fold, or mons pannus. These folds can cause problems with hygiene, directing the urine stream and sexual function (32). Removal of the skin and fat concealing the penis can be performed as a suprapubic lipectomy or limited panniculectomy. The skin is removed as a trapezoid incision. This technique increases the exposed penile length. The shaft skin can be closed with a skin graft taken from the lateral thigh, or from the removed mons pubis skin to avoid a donor site [33]. If there is need for further length, release of the suspensory ligament can be performed along with the suprapubic lipectomy[34). If the buried penis is secondary to cicatrix post circumcision, which is more common in children but can present in adults, either Z-plasties or removal of the entire penile skin with skin grafting and vacuum assisted closure with a negative pressure wound dressing can be performed. Suction lipectomy, or liposuction, is considered inadequate to treat a buried penis unless it is used in conjunction with suprapubic lipectomy. [34]

There was no evidence of cartilage extrusion, erectile dysfunction or urethral damage at a mean follow-up of 6 months.

CONCLUSIONS

All childrens complaining of short penis need to be clinically assessed for evidence of true micropenis. Patients should first be treated conservatively with testosterone therapy. There are no current guidelines on the best surgical management for children with micropenis. Multiple surgical techniques have been developed each with their own limitations. In our study ,All childrens were treated with Suspensory ligament release with VY Advancement Flap.No child hasdeveloped any complication with highest parent satisfaction in lshort and lond term follow up.

Footnotes

Conflicts of Interest: The authors have no conflicts of interest to declare.

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