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Hypospadias in male infants – a review

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Abstract. – The external genitalia problems are cumbersome problems for both doctors and parents, as these abnormalities have a consequent impact on future generations. However, the affected young infants are unaware of the consequences due to immature emotional state. Further, the feeling of being different and inferior in affected young patients could give rise to negative emotions including depression, insecurity, anxiety, powerlessness, etc. These all factors collectively could cause a mental imbalance in the affected children. The present review article is focused on the latest updates in the area.

Key Words:
Hypospadias, Urethral meatus, Anatomical anomalies.

Introduction

The hypospadias is a common male infant congenital problem on external genitals of male[1]. The appearance of the genital shows distinct characteristics establishing distinct characteristics in comparison to normal and often leads to negative behavioral emotions in the developing child[2]. In appearance, the associated penile shortening and ventral curvature known as chordee are more likely to be present with proximal defects. Therefore, hypospadias is associated with variable anatomical anomalies of the penis, abnormal ventral urethral opening, chordee of the penis and hooded foreskin. However, it has been observed that these anomalies are not present in the same patient[3]. Further, a study from the USA in a recent past cited approximate incidence of hypospadias and found that 1 in every 250 male births was affected by hypospadias. Moreover, the incidence of hypospadias is greater in whites as compared to blacks[4]. The present review article would discuss all-important aspects of this pathological state including etiologies, treatment and post treatment care.

Etiologies for Hypospadias

Multiple etiologies have been cited in literature in the recent studies including genetic, endocrine, and environmental factors proving confirmed causative factors for hypospadias[5,6]. Hereditary plays an important role in the spread of hypospadias, as it is more commonly observed in the infants whose family history showed a similar pathological state in fathers[7]. Further, the prevalence of hypospadias in male children of fathers with hypospadias is reported to be 8%, and 14% has been recorded in the brothers of children with hypospadias. Further, the nature of the inheritance is more likely to be polygenic[8]. Several genes are involved in the sexual determination. Similarly, variable genes acts, at different levels of sexual maturation of the infants, are observed to be responsible for hypospadias and are influenced frequently by the environmental factors. At the level of phallus development, homeobox genes family, fibroblast growth factor (FGF) and related gene family have been reported to be involved[9]. Further, at the level of testicular determination, SOX9, DMRT1 and GATA4 are observed to be influenced by environmental factors and have been associated with various genital disorders including severe hypospadias[10]. Finally, at the level of androgen biosynthesis, mutations in the LH receptor gene and the 5α-reductase gene directly causes severe hypospadias along with cryptorchidism[11].

Endocrine and Environmental Factors

The resultant condition, in case of inability to utilize available androgen or decline of the available androgen, is hypospadias. Also, certain mutations in androgen receptor genes are also responsible for the severe forms of hypospadias[12]. On the other hand, defects in the synthesis of testosterone also result in both forms viz. mild hypospadias and severe hypospadias[13]. Further, 5-alpha reductase enzyme mutations are involved in the spread of hypospadias as they directly affect testosterone synthesis. Estrogenic activity is known to be associated with the penile development and a study in recent past suggested that it got influenced significantly by pesticides on fruits and vegetables, from plastic linings in metal cans, and by certain pharmaceuticals leading to mild hypospadias[8]. A growing body of evidence

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suggested that the development of hypospadias is the combined result of a genetic predisposition coupled with fetal exposure to an environmental disruptor. There is evidence that poor semen quality, testicular cancer, undescended testes and hypospadias, are symptoms of one underlying entity[14]. Further, environmental influences have been confirmed to disturb embryonal programming and gonadal development to cause hypospadias.

Associated Anomalies and Intersex

The most common anomalies associated with hypospadias are undescended testes 9.3%, inguinal hernias 9.1%, which increase in the severe form of hypospadias up to 30% of undescended testes and to 20% of inguinal hernias, respectively[15]. A severe form of hypospadias could be the presentation of disorder of sexual development (DSD), particularly when associated with undescended testis[16]. Associated persistent prostatic utricle is present in 20% of cases, and occasionally noted when catheterization of the urethra is attempted in patients with hypospadias. Bifid scrotum is also associated with severe hypospadias. Associated abnormalities of the upper urinary tract, such as pelviureteric junction obstruction, vesicoureteral reflux are rare, occurring in about 2% of patients with severe hypospadias; therefore, routine ultrasound scan is not necessary for the mild form of hypospadias.

Diagnosis

Hypospadias is typically diagnosed by a physical examination of newborn genitals by observation of distinct characteristics of the disease. The confirming common distinct characteristic of hypospadias is a dorsal hood of the foreskin with a glanular groove. Further, observation of penile curvature is another common characteristic noted for the diagnosis of hypospadias[8]. Moreover, a bifid scrotum and penoscrotal transposition are also commonly observed in hypospadias affected newborns.

Therapeutic Avenues for Hypospadias

Surgical Correction

Surgical correction of hypospadias is the prime treatment avenue available with concerned pediatric surgeon/urologist in the field. However, it remains a great challenge for pediatric surgeon/urologist. In the recent past, many technological advances have been made for the betterment of surgical option for the correction of hypospadias. The major technical advances that offered high accuracy and efficiency are the development of technology, which can preserve the urethral plate when incision is made in the urethral plate. Furthermore, technological advances have also allowed the dorsal midline plication to correct penile curvature. All these technological advances collectively contributed towards better functional and aesthetic outcomes[17]. The prime aim of hypospadias repair surgery is to create a straight penis by improvising its appearance as straight as possible (urethroplasty and meatoplasty)[18]. This operation helps the affected child in his future sexual intercourse, and also allows an acceptable cosmetic appearance too. Further, surgical correction also leads to urethral plate improvised vascularization along with a rich nerve supply, muscular backing and gland formation.

Role of Preoperative Hormone Stimulation

For better results of the surgical correction, urologists often use preoperative androgen therapy worldwide on hypospadias patients. The procedure involves an intramuscular injection of hormones viz. gonadotrophin or testosterone to the affected patients[19]. Tropical ointment of the hormones is also possible. This preoperative androgen therapy helps in enhancement of the size of the penis, thereby improving the blood supply for better surgical repair.

Time of Surgical Repair

The correct age of surgical repair has always remained the topic of debate and a lot of variations have been made in the past. At present, the most appropriate age as recommended by most of the surgeons is 6-18 months, trending toward earlier intervention[20]. This age has a dual advantage of an improved emotional as well as psychological result.

Surgical Techniques

Reconstruction of the urethra is possible in two ways during surgical correction of hypospadias viz. single-stage or a two-staged procedure. The preference or choice is made by the surgeon on the basis of location and type of hypospadias. Single step procedure is utilized for mild distal, mid shaft and proximal hypospadias[21]. On the other hand, two-staged repair is used for a more severe form of hypospadias[22].

Postoperative Management

The prime post-operative step is dressing, which helps in the maintenance of haemostatic pressure over the wound and keeps the phallus in an upright position. Moreover, the dressing also immobilizes the penis to minimize edema and prevent hematoma formation. Common dressing included silastic foam, a clean bandage, bio-oc-

clusive membrane dressing, and non-adhesive dressing. Further, the dressing is in such a way that it prevents the discomfort and distress to the patient. However, most dressings are bulky, hard to apply or remove, and might fall off in an active child. The second important post-operative management step is the urinary diversion. It helps in the prevention of urethral edema that in turn allows the neourethra to heal completely before contact with urine flow. The final step of post surgery management included post-operative analgesia and pain control. It could be achieved by caudal anesthetic block, epidural anesthetic or penile block. The common method is the utilization of a caudal anesthetic block to help decrease postoperative pain by bupivacaine 0.25% followed by oral or suppository diclofenac. Bladder spasm caused by catheters could be managed with Oxybutynin.

Conclusions

It is quite clear from the above literature that a lot of developments have been made for the betterment of young male patients suffering from the pathological state of hypospadias. However, there is still a scope for the development of better treatment avenues for the affected children.

Conflict of interest

The authors declare no conflicts of interest.

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