FUNCTIONAL AND COSMETIC OUTCOME OF TWO-STAGE HYPOSPADIAS REPAIR: AN OBJECTIVE SCORING EVALUATION AND UROFLOWMETRY

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II. ABSTRACT

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Introduction

Hypospadias is characterized by an abnormally located urethral opening that could occur anywhere proximal to its normal location on the ventral surface of glans penis to the perineum and usually accompanied with chordee. Distal hypospadias is including glans, coronal and distal penile hypospadias. Proximal hypospadias is including proximal penile and penoscrotal hypospadias. It is the most common congenital anomaly affecting the penis (Wilcox & Ransley, 2000) with an incidence of 0.7% of male live births (Michael *et al.*, 2001). There have been many operations described for hypospadias involving many surgical subspecialties. This reflects the difficulty in getting optimum results from the surgery and implies that there is no gold standard technique for hypospadias repair (Arshad, 2005, Oztruk *et al.*, 2005). There is also no standardized objective method to assess the outcome of hypospadias repair until Holland *et al.* (2001) came with hypospadias objective scoring evaluation (HOSE).

Objective

The main objective of this study is to evaluate the functional and cosmetic outcome of patients who underwent two-stage hypospadias repair in Hospital Universiti Sains Malaysia and Hospital Raja Perempuan Zainab between January 1997 and December 2004, using HOSE (hypospadias objective scoring evaluation) and uroflowmetry and also to determine the factors that could influenced the outcome.

Methodology

This is an historical cohort study among hypospadias patients who have undergone two-stage hypospadias repair in Hospital Universiti Sains Malaysia and Hospital Raja Perempuan Zainab II between January 1997 and December 2004. Over the eight years 90 hypospadias patients underwent two-stage repair. Only 55 patients out of 90 patients (61.1%) with complete record and agree to participate were included in the study. They were examined to evaluate the functional and cosmetic outcome using HOSE: hypospadias objective scoring evaluation and uroflowmetry (if they were able to void volitionally and had no fistula). Five factors that may have influenced the outcome of hypospadias were studied, including type of hyposapadias, age at the completion of repair, duration between the first and the second-stage repair, techniques of hypospadias repair and surgeon.

Results

53 of the 55 patients were Malay, one Chinese and one Siamese. The age of patients at the time of the study ranged from 8 to 23 year-old (mean age 14.89 year). 35 patients (63.6%) had proximal type hypospadias (23 penoscrotal and 12 proximal penile) and 20 patients (36.4%) had distal hypospadias (12 distal penile, 7 subcoronal and one glannular) Four patients underwent circumcision in one to two years before hypospadias repair and two patients underwent previous unsuccessful hypospadias repair.

The types of operations performed were Bracka's two-stage procedure (37) and Byar's two-stage procedure (18). The complications encountered were urethralcutaneous fistula 17 patients (30.9%), followed by meatal stenosis 2 patients (3.6%), urethtal stricture one patient (1.8%) and wide meatal opening at subcoronal one patient (1.8%). Of the 17 patients with fistula, 9 underwent fistula repair and three had recurrence. Using the assessment criteria in HOSE, 34.5% had acceptable score and 65.5% had unacceptable score. The meatal openings were located at the tip of glans penis in 17 patients (30.9%), the meatal opening were vertical slit in 12 patients (21.8%), single urinary stream were obtained in 50 patients (90.9%), straight penis on erection were documented in 20 patients (36.4%) and there were no fistula in 44 patients (80%). Only 43 patients who were able to underwent uroflowmetry examination, in which 36 patients (83.7.0%) were considered normal, four patients (9.3%) as equivocal and three patients (7.0%) were obstructed. Only surgeon factor was found to have statistically significant influence on the outcome.

Conclusion

In conclusion, there seem to be a higher occurrence e of penoscrotal hypospadias in the Eastern side of Peninsula of Malaysia. HOSE and uroflowmerty are important objective tools to evaluate the functional and cosmetic outcome. The only factor that had a statistically significant influence on the outcome was the surgeon factor; other factors were found to be insignificant statistically.

III. ABSTRAK

HASIL PEMBEDAHAN MEMBAIKI HYPOSPADIAS DUA PERINGKAT DARI ASPEK FUNGSI DAN KOSMETIK: SATU PENILAIAN SKOR OBJEKTIF DAN UROFLOWMETRI

Pengenalan

Diantara ciri-ciri hypospadias ialah bukaan kencing yang tidak normal iaitu dibahagian bawah batang zakar di antara kepala zakar sehingga ke perineum dan biasanya berkaitan dengan zakar yang bengkok ke bawah. Hypospadias distal termasuk hypospadias kepala zakar, hypospadias takuk zakar dan hypospadias hujung zakar. Manakala proksimal hypospadias termasuk hypospadias pangkal zakar dan hypospadias penoskrotal. Hypospadias adalah kecacatan zakar sejak lahir yang paling kerap (Wilcox & Ransley, 2000) dengan kadar incidence 0.7% bagi setiap bayi lelaki hidup yang dilahirkan (Michael *et al.*, 2001). Banyak sekali pembedahan membaiki hypospadias yang telah dijelaskan secara bertulis dan melibatkan banyak pakar bedah dari pelbagai bidang kepakaran. Ini menunjukan yang pembedahan ini adalah sukar untuk mendapakan hasil pembedahan yang unggul dan tidak ada satu pun teknik pembedahan yang terbaik (Arshad, 2005, Oztruk *et al.*, 2005). Tiada cara yang objektif untuk menilai hsail pembedahan hypospadias sehingga Holland *et al.* (2001) muncul dengan skor penilaian objektif hypospadias (HOSE).

Objektif

Objektif utama kajian ini adalah untuk menilai hasil pembedahan hypospadias dua peringkat di Hospital Universiti Sains Malaysia dan Hospital Raja Perempuan Zainab II diantara Januari 1997 and Disember 2004.dari segi fungsi dan kosmetik dengan menggunakan HOSE: skor penilaian objektif hypospadias dan uroflowmetri. Juga untuk menentukan faktor-faktor yang boleh mempengaruhi hasil pembedahan ini.

Methodologi

Ini adalah kajian sejarah kohort dikalangan pesakit-pesakit hypospadias yang telah menjalani rawatan pembedahan di Hospital Universiti Sains Malaysia dan Hospital Raja Perempuan Zainab II diantara Januari 1997 dan Disember 2004. Selama lapan tahun tersebut, terdapat 90 pesakit manjalani pembedahan hypospadias dua peringkat Hanya 55 peaskit daripada 90 pesakit (61%) yang mempunyai fail yang lengkap dan bersetuju untuk menyertai kajian ini yang telah diambil kira dalam kajian ini. Mereka semua telah diperiksa dan ditemuduga untuk menilai hasil pembedahan hypospadias dari segi fungsi dan kosmetik dengan menggunakan HOSE dan manjalani ujian uroflowmetri [jika mereka boleh kencing sendiri dan tiada fistula (lubang kencing lain yang tidak normal)] Lima factor yang mungkin mempengaruhi hasil pembedahan hypospadias yang dikaji adalah jenis hypospadias, umur semasa selesai menjalani pembedahan hypospadias, selang masa di antara pembedahan peringkat pertama dan peringkat kedua, teknik pembedahan dan pakar bedah.

Keputusan

53 daripada 55 pesakit, adalah Melayu, seorang Cina dan seorang lagi Siam. Mereka berumur daripada lapan tahun sehingga 23 tahun (pukul rata 14.89 tahun)semasa kajian ini dijalankan. 35 pesakit (63.6%) mempunyai hypospadias proksimal (23 pesakit hypospadias penoscrotal dan 12 pesakit hypospadias pangkal zakar) dan 20 pesakit (36.4%) mempunyai hypospadias distal (12 pesakit hypospadias hujung zakar, 7 pesakit mempunyai hypospadias takuk zakar dan seorang hypospadias di kepala zakar). Empat pesakit telah berkhatan sebelum pembedahan membaiki hypospadias dan tiga lagi telah menjalani rawatan pembedahan hypospadias tetapi tidak berjaya.

Jenis pembedahan yang telah dijalankan adalah pembedahan Bracka dua peringkat (37 pesakit) dan pembedahan Byar dua peringkat (18 pesakit). Komplikasi pembedahan yang berlaku adalah fistula 17 pesakit (30.9%), diikuti oleh bukaan lubang kencing yang sempit 2 pesakit (3.6%), saluran kencing yang sempit seorang pesakit (1.8%) dan bukaan lubang kencing yang besar di takuk zakar seorang pesakit (1.8%). Daripada 17 pesakit yang mempuyai fistula, 9 pesakit telah menjalani rawatan fistula dan 3 daripadanya berlaku fstula lagi. Dengan menggunakan penilaian HOSE, 34.5% pesakit mempunyai hasil pembedahan yang boleh diterima dan 65.5% mempunyai hasil pembedahan yang kurang memberangsangkan. Tujuh belas pesakit (30.9%) mempunyai bukaan kencing di hujung kepala zakar, 12 pesakit (21.8%) mempunyai bukaan kencing berbentuk satu garisan, 50 peskit (90.9%) mempunyai aliran kencing tunggal, 20 pesakit (36.4%)

mempunyai zakar yang lurus semasa zakar ereksi dan 44 pesakit (80%) tidak mempunyai fistula. Hanya terdapat 43 pesakit yang boleh menjalani ujian uroflowmetri Daripada jumlah itu, 36 pesakit (83.7%) dianggap normal, tiga pesakit (7.0%) mempunyai saluran kencing yang sempit dan empat pesakit tidak tentu. Hanya faktor pekar bedah yang signifikan secara statististik mempengaruhi hasil pembedahan hypospadias.

Kesimpulan

Kadar hypospadias penoskrotal adalah tinggi di Pantai Timur, Semenanjung Malaysia. HOSE dan uroflometri adalah alat objektif yang penting untuk menganalisa hasil pembedahan membaiki hypospadias. Hanya faktor pakar bedah sahaja yang signifikan secara statistik mempengaruhi hasil pembedahan; faktor-faktor lain didapati tidak mempengaruhi hasil pembedahan ynag signifikan secara statistik.

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I dedicated this dissertation to my beloved wife,

Dr. Sharyla Azura and my son, Muhammad Ifran Syazwi.

Both of them had to bear my absence during my endeavor to be

a surgeon.

I also dedicated this dissertation to my late parent.

1. INTRODUCTION

Hypospadias is the most common congenital anomaly affecting the penis. Hypospadias deformity described as incomplete development of the urethra and presents with an abnormally proximally placed urethral opening on the ventral aspect of the penis or even in the perineum.

A band of fibrous tissue usually accompanied hypospadias that extends from the abnormal meatus to the glans and this band frequently shortens the ventral aspect of the penis. This band caused chordee that produced a downward curvature of the penis, noted during erection. The term *chordee* comes from Latin word chorda, meaning string and originally was used to describe the painful downward bending of the penis during erection caused by gonorrheal inflammation of the corpus spongiosum. Here, the fibrous tissue around and under the urethra causes the tunica albuginea to be inelastic and, therefore, restrict the growth of ventral urethra. This fibrous tissue, which is soft in small children, hardens over time and becomes a more fibrotic and inelastic tissue. However not every patient has all three associated anomalies; the hooded foreskin and chordee are common but not constant. Although in most cases hypospadias is not an urgent problem, it may create anxiety for the infant's parents. A good understanding of this condition will enable caregivers to address parents' concerns and questions confidently and accurately.

Hypospadias has a long history in medicine. It was first described in the second century by Galen, who gave the condition its name. The term hypospadias comes from the Greek words *hypo*, meaning below, and spadon, meaning opening. The earliest medical text describing hypospadias dates back to the Celsius (25 AD) and Galen (second century AD), with the description of hypospadias malformation. During the first millennium, the primary treatment for hypospadias was penile amputation distal to the meat. Since then, many have contributed to development of modern hypospadias repair.

Duple began the modern era in this field in 1874 by publishing a detailed procedure for urethra reconstruction. Now a days over 300 different types of repairs have been described in the medical literature. Although most reports have been in the last 60 years, most basic techniques were founded over the last century. Most of the procedures are multistage reconstruction; all consist of a first emergency stage that addresses correction of the stenotic meatus if required and a second-stage that eliminates the chordee and recurvatum, followed by urethroplasty (creation of a neo-urethra and its coverage).

There are numerous problems associated with multi-staged techniques, required multiple operations, often the meatus did not reach the tip of the glans or retract with time because of multiple tissue manipulations and repeating scarring, urethral stricture and/or fistula formation were frequent, and the final aesthetic result was poor.

The optimal repair for distal hypospadias remains controversial, with a variety of procedures reported to provide good results. The traditional goals of corrective surgery have been the correction of any associated chordee, the provision of a glandular meatus to enable voiding while standing and effective coitus. Recent advances in surgical techniques and a greater appreciation of long-term psychological consequences of a cosmetically unsatisfactory repair have led to the development of repairs that provide both a functional and cosmetically acceptable outcome. The goal of hypospadias surgery is a penis that is adequate both functionally and cosmetically normal. This requires a penis that is straight on erection with a vertically orientated meatus at the tip of the glans, thus promoting a single, coherent urinary stream.

The long-term complications of hypospadias surgery are well known and sadly too common. The incidence depends on the initial severity of the hypospadias, the type of operation and the skill and experience of the surgeon. The most common complication is fistula, occurring in up to 30% of patients. A fistula can present acutely immediately after the catheter is removed, or many years after repair.

Despite the many operative procedures described for correcting hypopsdias, there is no generally accepted system for assessing the surgical results. This lack of impartial method of documenting the results of hypospadias surgery has made the comparative evaluation of operative procedures inaccurate and subjective.

The ideal scoring system should incorporate important functional and cosmetic criteria, including relevant surgical complications, in addition to being reproducible and free from inter-observer error. This would allow an objective analysis of different repair techniques outcome so that a valid comparison can be made, facilitating surgical audit and a balanced evaluation of the outcomes of traditional and innovative surgical procedures for the repair of hypospadias

The main purpose of this study is to review the clinicopathological aspect of this deformity, to evaluate the treatment results in a standard objective scoring of the functional and cosmetic outcome of two-stage hypospadias repair procedures in two institutions over 8 years period (January 1997 – December 2004) using a Hypospadias Objective Scoring Evaluation (HOSE) and uroflowmetry. I would like to evaluate the factors that could influence the outcome. We are still lacking local data regarding the outcome of hypospadias repair. The only study regarding experience of hypospadias repair in Malaysia which was published internationally in *Br J Plast Surg*, 58, 2005 was written by Dato' Ahmad Ridzuan Arshad.

2. LITERATURE REVIEW

2.1 EPIDEMIOLOGY

The hypospadias incidence is 1 in 300 live male births, and appears to be increasing in frequency (Arshad, 2005, Wilcox & Ransley 2000, Duckett, 1998). Michael *et al.*, (2001) reported that the incidence of hypospadias was 0.7% of male live births (Michael *et al.*, 2001). In the United States, hypospadias affecting as many as 1 in 125 newborn males and before 1970, it occurred about once in every 250 live male births (Paulozzi *et al.*, 1997). The reason for the increase in prevalence of hypospadias in recent years is not known. Non-Hispanic whites are at a higher risk than other racial-ethnic groups for most types of hypospadias (Carmichael *et al.*, 2003).

Upon a retrospective review of the records from military treatment facilities by Micheal *et al.*, (2001), they found out that the incidence of hypospadias were 0.8% in white, 0.6% in black, 0.5% in Asian, 0.6% in Native American, and 0.6% in unknown. There was no significant difference between races, but the incidence of hypospadias in minorities was higher than previously reported.

In Malaysia, a retrospective study by Arshad (2005) involving 115 patients with hypospadias who were treated between September 1987 and December 2002, comprising eighty-four Malays (84/115), twenty-eight Chinese (28/115)

and three Indian (3/115). This study showed that hypopadias incidence is highest in Malays. However, this must be interprated within the context of the racial distribution of population in Malaysia.

2.2 PATHOPHYSIOLOGY

2.2.1 Embryology

Hypospadias is a congenital defect that is thought to occur embryologically during urethral development, from 8-20 weeks gestation. The external genital structures are identical in males and females until 8 weeks gestation, but genitals developed a masculine phenotype in males primarily under the influence of testosterone. As the phallus grows, the open urethral groove extends from its base to the level of the corona. The classic theory is that the urethral folds coalesce in the midline from base to tip, forming a tabularized penile urethra and median scrotal raphe. This accounts for the posterior midline urethra. The anterior or glanular urethra is thought to develop in a proximal direction, with an ectodermal core forming at the tip of the glans penis, which canalizes to join with the more proximal urethra at the level of the corona. The higher incidence of subcoronal hypospadias supports the vulnerable final step in this theory of development (Gatti *et al.*, 2003).

Baskin (2000) proposed a modification of this theory in which the urethral folds fuse to form a seam of epithelium, which is then transformed into mesenchyme and subsequently canalizes by apoptosis or programmed cell resorption. Similarly, this seam theoretically also develops at the glanular level, and the endoderm changes to ectoderm with subsequent canalization by apoptosis.

The prepuce normally forms as a ridge of skin from the corona that grows circumferentially, fusing with the glans. Failure of fusion of the urethral folds in hypospadias impedes this process, and a dorsal hood of prepuce results. On rare occasions, a glanular cleft with intact prepuce may occur; the megameatus intact prepuce (MIP) variant (Gatti *et al.*, 2003).

Chordee is often associated with hypospadias, especially more severe forms. This is thought to develop secondary to a growth disparity between the normal dorsal tissue of the corporal bodies and the attenuated ventral urethra and associated tissues. Rarely, the abortive spongiosal tissue and fascia distal to the urethral meatus forms a tethering fibrous band that contributes to the chordee (Gatti *et al.*, 2003).

2.2.2 Etiology

Several etiologies for hypospadias have been suggested. These etiologies include genetic, endocrine and environmental factors.

2.2.2.1 Genetic Factors

A genetic predisposition has been suggested by the eight-fold increase in incidence of hypospadias among monozygotic twins compared to singletons. This finding may relate to the demand of two fetuses for human chorionic gonadotropin (hCG) produced by a single placenta, with an inadequate supply during critical periods of urethral development (Gatti *et al.*, 2003).

A familial trend has been noted with hypospadias. Fathers of children with hypospadias had been affected in 7-10% of patients, and 10-20% of brothers of children with hypopadias are affected as well (Belman, 1997, Freell *et al.*, 2002, Gatti *et al.*, 2003). Familial patterns suggest a polygenic mode of inheritance, although recessive transmission has been implicated in the affected offsprings of cosanguineous marriages (Gatti *et al.*, 2003, Thomas, 2004).

However, the importance of familial factors should not be overstated as it is a sporadic anomaly in most boys, particularly those with distal or mid-shaft form of hypospadias (Thomas, 2004).

2.2.2.2 Endocrine Factors

A decrease in available androgen or inability to use available androgen appropriately may result in hypospadias. Aaronson *et al.* (1997) reported 66% of boys with mild hypospadias and 40% with severe hypospadias were found to have a defect in testicular testosterone biosynthesis. Mutations in the gene for 5-alpha reductase enzyme, which converts testosterone (T) to the more potent dihydrotestosterone (DHT), have been associated with hypospadias. Silver *et al.* (1999) found that nearly 10% of boys with isolated hypospadias had at least 1 allele affected with a 5-alpha reductase gene mutation. Although androgen receptor deficits, quantitative or qualitative, have been shown to result in hypospadias, this is thought to be a relatively in frequent occurrence and other factors are more commonly implicated.

2.2.2.3 Environmental Factors

The concept of endocrine disruption by environmental agents is gaining popularity as a possibe etiology for hypospadias and as an explanation for its increasing incidence (Gatti *et al.*, 2003). Endocrine disruption is also known as exposure to hormonally active agents in the environment that might be contributing to a declining reproductive health in humans, males in particular (Bukowski, 2001, Fisher, 2004, Landrigan, 2003).

Endocrine disrupting agents are synthetic or natural substances that can interfere with hormone systems by mimicking, blocking, or otherwise altering the normal action of hormones during critical periods of embryonic or fetal development (Landrigan, 2003). Human are continually in contact with endocrine-disrupting chemicals that are present in food and in the environment (Baskin *et al.*, 2001).

Endocrine-disrupting chemicals that inhibit androgen binding, androgen synthesis, and androgen-induced gene expression are known as antiandrogens. In theory, exposure to an antiandrogen during development of the urogenital tract can impede normal male differentiation, resulting in hyopspadias (Sharpe & Irvin, 2001).

Poor intrauterine growth is associated with hypospadias (Carmichael *et al.*, 2003, Fredell, 2003, Hussain, 2002, Gatti, 2001). Among small for gestational age infants in neonatal intensive care, hypospadias is ten times more common than in the general neonatal population (Gatti, 2001). Unknown factors, operating very early in gestation, are thought to disrupt both somatic growth and development of the urethra (Fredell, 2003, Gatti, 2001). A relative lack of androgen inducing hCG affecting smaller fetuses is one possible mechanism (Fredell, 2003).

2.3 DIAGNOSIS AND PHYSICAL EXAMINATION

The diagnosis of hypospadias is usually straightforward, relaying on physical examination alone (Wilcox & Ransley, 2000). Most forms of hypsopadias is immediately recognized during the physical examination of the newborn (Snodgrass, 2004). Quite often, the abnormality is identified lately during examination before a child circumcision. Very rarely, the ventral foreskin will be normal in appearance and the hypospadias will be noted later in life when the foreskin retracts or after a circumcision was performed. When a hypospadias is identified, an early referral should be made and circumcision should not be done (Elder, 1998).

In general, hypospadias itself produces no symptoms. Suspect hypospadias when there is incomplete formation of the foreskin or hooded prepuce (Snodgrass *et al.*, 2002). Look for associated features, such as a penile raphe that is displaced from the midline and a glans that tilted downwards (Snodgrass *et al.*, 2002).

2.3.1 Meatus Location

Locate the external meatus by inspecting the glans, shaft, scrotum and perineum closely. The meatus might appear to be pinhole-sized, but usually patent (Snodgrass *at el.*, 2002). The majority of hypospadias cases are glanular, coronal, or subcoronal (Sweet *et al.*, 1972, Paulozzi *et al.*, 1997). A blind dimple, or pit, might be seen in the glans at expected location of the meatus (Laura & Stokowski, 2004). If there is any doubt about the location of the urethral meatus, it may be necessary to observe the infant voiding or get history from mother. In many cases, the true meatal opening can only be determined at the time of surgery after correction of curvature (Baskin, 2004).

2.3.2 Prepuce

Normally, the prepuce of the newborn completely covers the glans and is adherent to the glans. Thus, the prepuce cannot, and should not be retracted (Cold & Taylor, 1999). A hooded prepuce refers to the appearance of the dorsal foreskin of the hypospadias penis where by the preputial tissue

is excessive on the dorsal surface, but absent on the ventral surface. This occurred because during fetal urethral development, dorsal preputial growth precedes ventral growth, and the preputial growth is interrupted by failure of fusion of the urethral folds (Baskin, 2000).

Megameatus intact prepuce (MIP) is another variant of hypospadias where the diagnosis might be missed until the foreskin is retracted for newborn circumcision (Retik, 2002). In the MIP type, the prepuce completely covers the glans; but there is an overly large urethral opening underneath.

2.3.3 Chordee

Chordee is the ventral or downward bowing or curvature of the penis that is more evident during erection. It can occur in isolation or in association with hypospadias. A minority of boys with hypospadias have significant chordee. On the other hand, most severe hypospadias is associated with chordee (Belman, 1997). Curvature of the penis occurs in approximately 15% of distal hypospadias and at least 80% of proximal hypospadias (Snodgrass & Warren, 1999).

2.4 CLASSIFICATION OF HYPOSPADIAS

Hypospadias classification depends on the position of the urethral meatus from the tip of the glans penis to the ventral surface (underside) of the penis, scrotum or penineum The majority of hypospadias is coronoglanular type, accounts for 80% of the cases (Sweet *et al.*, 1972, Paulozzi *et al.*, 1997). This is because distal penis is the region of the last stage of fetal urethral development (Snodgrass *et al.*, 2002). However, classification systems do not take into account the degree of penile curvature or other individual features or anomalies; thus, they do not always correlate directly with severity of the condition or extent of surgical repair that might be required (Baskin *et al*, 2001).

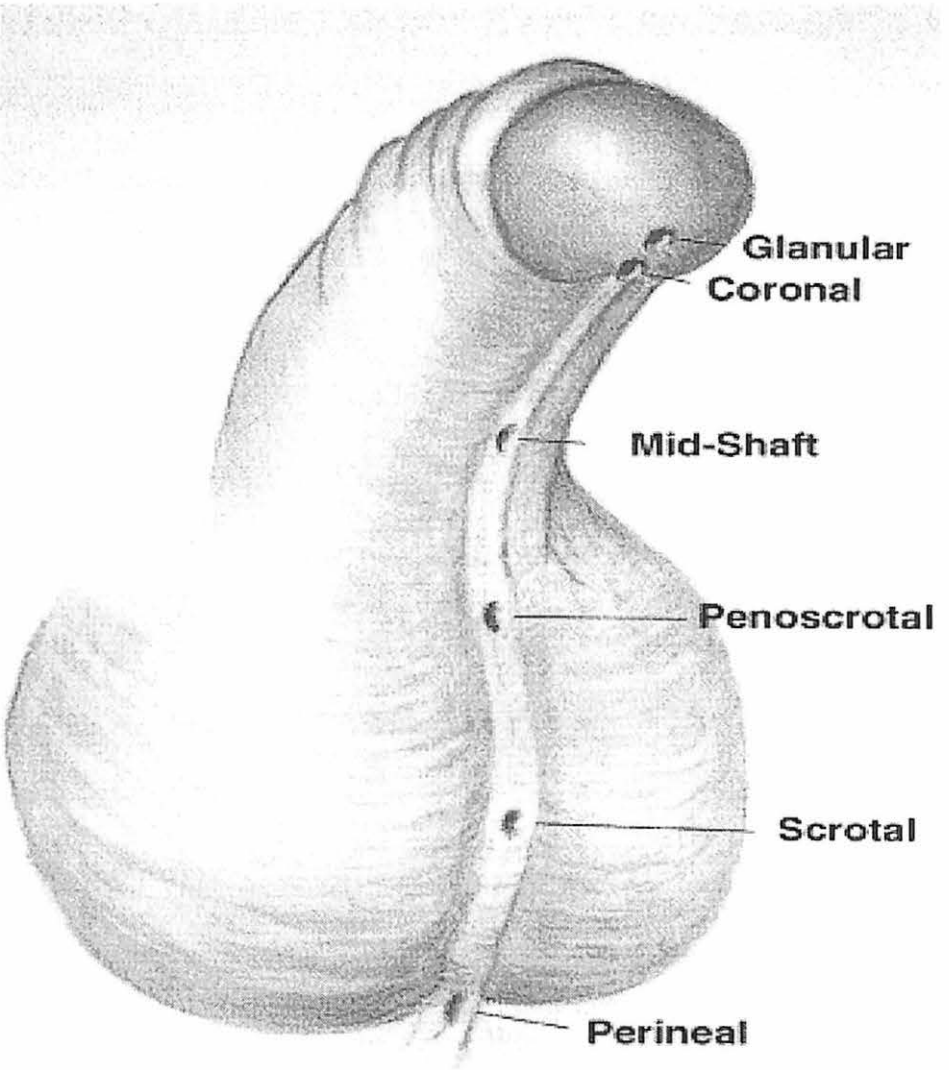

Figure 2.4: Classification of hypospadias. The position of the urethral meatus forms the basis of anatomic classification. Illustration by Peter Stone. (Reproduced with permission from the author, Laura A and Stokowski, 2004)

2.5 ASSOCIATED ANOMALIES AND CONGENITAL SYNDROMES

Several anomalies have been recognized in associated with hypospadias but they are very rare. Several urogenital defects are associated with hypospadias, as follows (Santenelli, 2002):

1. Cryptochidism (9%)
2. Inguinal hernia (9%)
3. Megalourethra, urethral fistulae, and hypoplastic testicles (reported less often)
4. Upper urinary tract defects (observed in association with proximally located hypospadias)

Hypospadias can be part of congenital syndromes such as Opitz syndrome (BBB syndrome), which can be recognized by midline abnormalities, including hypertelorism, cleft lip, heart defects, cryptorchidism, and bifid scrotum (Laura & Stokowski, 2004). Smith-Lemli-Opitz syndrome, caused by an inborn error of cholesterol synthesis, can present with hypospadias, in addition to microcephaly, cleft palate, syndactaly, and polydactaly (Snodgrass, 2002). Hypospadias is also seen in Wolf-Hirschhorn syndrome, a rare chromosomal disorder with multiple dysmorphic features and midline defects, and with Denys-Drash syndrome (Wilms'tumor) (Laura & Stokowski, 2004).

Patient with hypospadias and cryptorchidism need special attention. A retrospective study by Rajfer and Walsh (1976) reported the incidence of intersexuality in patients with hypospadias and cryptorchidisam; 45 such patients were identified. The purpose of their study was to identify patients who had evidence of intersexuality. Of the 45 boys, 11 (24.4%) had a unilateral undescended testicle, three (6.7%) were described as intersex, two (4.4%) had mixed gonadal dysgenesis and one (2.2%) was a true hermaphrodite. The remaining 34 boys had bilateral undescended testicles and 21 (62%) were felt to have an intersex disorder. This report highlights the need to consider and evaluate patients for intersex disorders who present with hypospadias and undescended testicles. Therefore, systemic evaluation is very important in every case of hypospadias.

2.6 THE SURGICAL REPAIR OF HYPOSPADIAS

2.6.1 Goals of Surgical Repair

Improvement in the surgical technique reflected positively on the results of hypospadias repair, not only the functional but also the aesthetic appearance. The abnormal appearance of genitalia may strongly interfere with the psychological development of patients and be a serious handicap for normal sexual and social life. Therefore, the present goals in hypospadias surgery are (Hinderer, 2000):

1. Normal orthostatic micturation with a straight and uniform urinary stream. This requires the reconstruction of a physiologically wide, regular, elastic and hairless urethra with normal growth potential and a transition from urethra to neo-urethra without stricture.
2. Aesthetic appearance of the genitalia with the meatus placed at or close to, the tip of the glans and with a normally distributed mobile skin cover with inconspicuous scars resembling a circumcised penis. The aesthetic appearance also requires the surgical correction of associated malformations of the scrotum and testes, and after puberty, of any penile hypoplasia present.
3. Normal sexual function with a straight erection of adequate strength.

2.6.2 History of the Surgical Procedures

Hypospadias has a long history in medicine (Laura & Stokowski, 2004). During the first millennium, the primary treatment for hypospadias was penile amputation distal to the meatus (Gatti *et al.*, 2003). Since then; many have contributed to development of modern hypospadias repair. Below are summarized some of the pioneers and development of the modern techniques which are divided into the following stages (Baran, *et al*, 2004);

1. **Correction of curvature deformity by chordee excision;**
    a. Mettuer in 1842
    b. Bouisson in 1861
    c. Duplay in 1874
    d. Devine and Horton in 1961
2. **Glanuloplasty;**
    a. Tunnelization techniques
        i. O-type orifice
            1. Nove-Josserand in 1897 to 1914
            2. Bevan in 1917
        ii. V-type flap
            1. Mustarde in 1965
            2. Hinderer in 1971
        iii. W-type flaps
            1. Cenetoglu et al. in 1992
    b. Splitting of glans into three triangular flaps
        i. Devine and Horton in 1961
    c. Splitting of glans into two lateral flaps by incisions made in middle or on two sides of the groove
        i. Mettauer in 1842
        ii. Bouisson in 1861
        iii. Thiersch in 1869
        iv. Duplay in 1874

v. Mathieu in 1932

vi. Duckett and Synder in 1981

vii. Snodgrass 1994

viii. Baran et al. in 2002

3. **Reconstruction of neourtehra;**

   a. Advancement of the urethra

      i. Beck and Hacker in 1897

      ii. Baran in 1982

      iii. Sensoz et al in 1997

   b. Local skin flaps (penile skin)

      i. Thiersch in 1869

      ii. Duplay in 1874

      iii. Bevan in 1914

      iv. Ombradanne in 1932

      v. Mathieu in 1932

   c. Free skin graft (inner surface of prepuce

      i. Devine and Horton in 1961

      ii. Hinderer in 1971

      iii. Bracka in 1989

   d. Distal skin flaps (prepuce; vascularized and island flaps)

      i. Hook in 1896

      ii. Toksu in 1970

      iii. Asopa and Asopa in 1984

iv. Hinderer in 1971

4. **Coverage of the skin defect on ventral surface of penis by skin flaps;**

   a. Scrotum

      i. Beck in 1897

   b. Prepuce

      i. Thiersch in 1869

      ii. Ombradanne in 1932

      iii. Byars in 1955

      iv. Devine and Horton in 1961

2.6.3 General Principles of Hypospadias Repair

There is no single surgical method that can be applied to all forms of hypospadias. This is clearly indicated by multitude of surgical techniques performed (Baran *et al*, 2004). There are three objectives of the surgical correction of hypospadias (Wilcox & Ransley, 2000);

1 The correction of the penile chordee

2 Reconstruction of the urethra

3 Cosmetically recreating the penis

2.6.3.1 Correcting Penile Chordee

Penile chordee can result from several factors, including abnormal tethering of the penile shaft skin onto the underlying structures, tethering of the urethral plate onto the corpora cavernosa, atretic corpora spongiosum tissue extending from the abnormal meatus to the glans, and abnormal flexion of the corpora cavernosa (Mouriquanad, *at el.*, 1995).

Correcting penile chordee may necessitate one or more of the following steps;

a) Degloving the penis.
b) Excision of the atretic and fibrous corpora spongiosum proximally and distal to the abnormal meatus.
c) If the penis is still having a ventral curvature it is then necessary to plicate the dorsal aspect of the tunica albuginea.

Gittes and McLaughlin (1974) described a technique to induce artificial erection intraoperatively, which allows the surgeon to assess immediately the adequacy of chordee correction. Rapid acceptance of this technique by the urological community led Glassman *et al.* (1980)

to postulate that no patient should have recurrent chordee in the future.

Baskin and Duckett (1994) reported their collective experience correcting penile chordee associated with hypospadias, they noted that in 87% of 1366 patients, once the penis had been degloved, no further corrective steps were necessary. Also13% of the patients required a plication of the dorsal tunica albuginea (a modification of Nesbit's procedure). Those boys who underwent a plication were reviewed, with a mean follow-up of 2.7 years; six (4%) had residual chordee and in only one (0.6%) was it necessary to proceed to further surgery. This study also stresses the importance of an artificial erection test to ensure that the chordee has been corrected during surgery.

Some surgeons do not advocate dissecting the urethral plate of the corpora, arguing that this procedure may devascularise the urethral plate for future neourethra (Wilcox & Ransley, 2000). This issue is unresolved, as no study has been done to compare these two teachniques.

2.6.3.2 Urethroplasty

There have been over than 300 different techniques of hypospadias repair described, but broadly they can be divided into four main groups:

1. Urethral advancement
2. Tubularizing the urethral plate
3. A vascularized pedicle flap
4. A two-stage free-flap repair

2.6.3.2.1 Urethral Advancement

Duckett and Snyder (1981) have originally described the Meatal advancement glanuoplasty and preputioplasty (MAGPI) procedure. The appropriate selection of patients is vital to the success of this procedure. These authors emphasized that the parameatal skin needs to be thick and pliable so that it can be mobilized safely off the urethra. It is important to ensure that the urethra can reach the tip of the glans, which can be confirmed if simple traction moves the meatus to the tip of the glans (Duckett & Snyder, 1981). A vertical incision is then closed transversely, so advancing the