Faculty of Medicine
Ain Shams University

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جامعـة عيـن شمــس

Recent surgical advances in management Of Hypospadias

An Essay

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التقنيات الجراحية الحديثة لعلاج الاحليل السفلى

رسالة

توطئة للحصول على درجة الماجستير في الجراحة العامة

مقدم من

الطبيب/ محمود شعماش إسماعيل

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INTRODUCTION

Hypospadias is one of the most common congenital defects affecting the external male genitalia ***(Hinman and Baskin, 2008).*** The incidence is about 1: 250 male newborns, although its incidence seems to be increasing **(*Kraft et al., 2010).***

Hypospadias is defined as an insufficient development of the urethral fold and the ventral foreskin, with or without penile curvature. The urethral opening is located more proximally anywhere between the tip of the penis and the perineum ***(Baskin and Ebbers, 2006).***

Most cases of hypospadias have an unknown etiology, which is probably a mix of monogenic and multifactorial forms, implicating both genes and environmental factors ***(van der Zanden et al., 2012).***

Several chromosomal abnormalities and malformative syndromes include hypospadias, from anterior to penoscrotal forms. More recently, CXorf6 and ATF3 have been reported to be involved. Besides these genomic and hormonal factors, multiple substances found in the environment can also potentially interfere with male genital development because of their similarity to hormones ***(Kalfa et al., 2009).***

Hypospadias classification is based on the position of the

meatus, within three categories: distal or anterior hypospadias with the meatus on the glans penis, at the corona, or subcoronal. Mid-penile hypospadias with urethral opening located on the distal penile shaft, midshaft, or on the proximal penile shaft. Proximal or posterior hypospadias have a penoscrotal, scrotal, or perineal urethral meatus location ***(Subramaniam et al., 2011).***

Hypospadias surgery in history started by partial resection of glans to locate the orifice more centrally, and they leave proximal hypospadias as incurable. Then, many have contributed to development of modern hypospadias repair. More than 300 different types of repairs have been described in medical literature, famous surgeons and their techniques like Mathieu, Snodgrass, and Hadidi ***(Abdelrahman et al., 2011).***

Historically, the ideal age for genital surgery is between 6 and 12 months of age. This age range appears to insulate most children from the psychological, physiological, and anesthetic trauma associated with hypospadias surgery ***(Kass et al., 1996).*** Recent data prefer to perform the repair at the age of 4 months in infant boys with an adequately sized phallus and without medical problems ***(Kraft et al., 2011).***

The goals in management of hypospadias repair are creating a straight penis, reconstructing slit-like meatus at the tip of penis, creating a urethra of adequate length and uniform

caliber, symmetry in appearance of glans and penile shaft, projectile stream and normalization of erections, and thereby imposing confidence in the child. These goals can be achieved by meatoplasty and glanuloplasty, orthoplasty, urethroplasty, scrotoplasty and skin cover ***(Bhat, 2008).***

Decision making in hypospadias repair is no longer determined by meatal location only, as in the past, but also by severity of penile curvature and appearance of the incised urethral plate. Severe curvature, requiring plate transection or an unhealthy incised plate are uncommonly encountered ***(Ghnnam, 2010).***

The techniques of hypospadias surgery continue to evolve. Tubularized incised plate repair has been the mainstay for distal hypospadias. In cases of proximal hypospadias, one-stage repairs such as the Duckett repair or the Koyanagi repair have been well established, while two-stage repairs remain important alternatives ***(Hayashi and Kojima, 2008).***

The repair of complications after hypospadias repair is still an open problem. The majority of patients with failed hypospadias repair require surgical reconstruction to fully resurfacing the glans and penile shaft ***(Barbagli et al., 2012).***

Several techniques of providing vascularized soft tissue cover to the neourethra have been described. They include de-epithelialized skin, corpus spongiosum, dartos fascia, and tunica vaginalis. Recent data suggest that tunica vaginalis may have an edge over dartos fascia for soft tissue coverage of the neourethra ***(Dhua et al., 2012).***

AIM OF THE WORK

To review updated literature regarding etiology, presentation, and advances in surgical repair of hypospadias, stressing on complications and how to deal with.

EMBRYOLOGICAL CONSIDERATIONS

Normal development of External Genitalia:

1. Indifferent Stage:

In the third week of development, mesenchyme cells originating in the region of the primitive streak migrate around the cloacal membrane to form a pair of slightly elevated cloacal folds. Cranial to the cloacal membrane the folds unite to form the genital tubercle. Caudally the folds are subdivided into urethral folds anteriorly and anal folds posteriorly,

In the meantime, another pair of elevations, the genital swellings, becomes visible on each side of the urethral folds. These swellings later form the scrotal swellings in the male and the labia majora in the female (Figure: 1) ***(Sadler, 2005).***

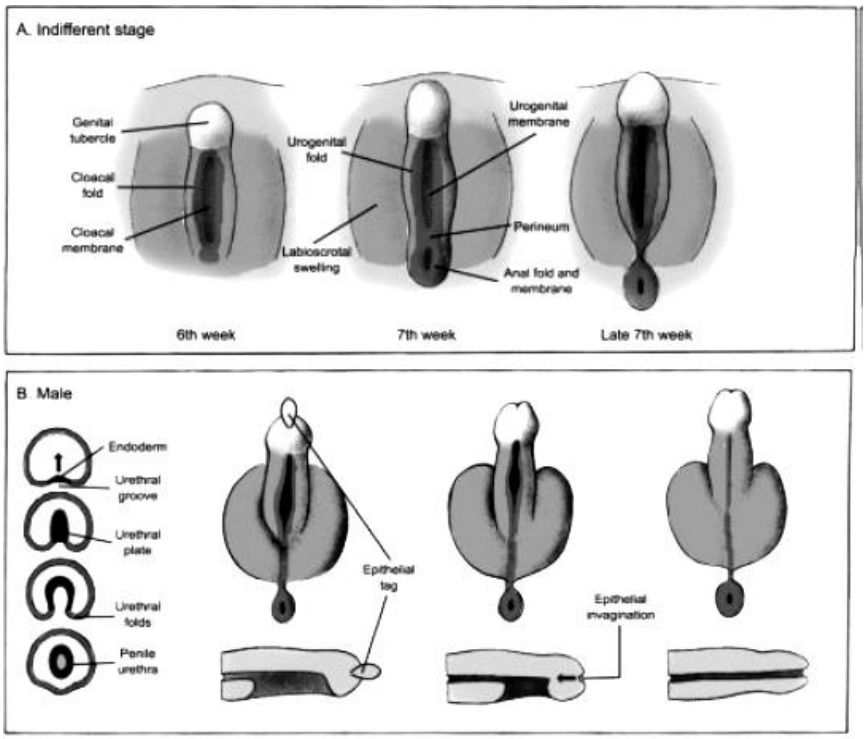

Figure 1: Cloacal division into the anterior urogenital sinus and the posterior anorectal canal, the portion of the cloacal folds ***(Park, 2002).***

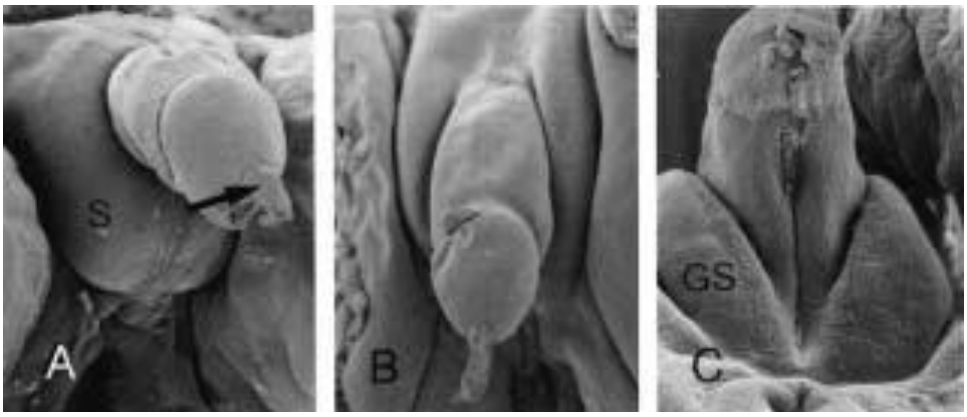

Figure 2: (A) Genitalia of a male fetus at 14 weeks, showing fusion of the scrotal swellings (S). Arrow, epithelial tag, (B) Genitalia of a female fetus at 11 weeks dorsal view & (C) Genitalia of a female fetus at 11ventral views. The genital tubercle at this stage is longer than in the male (A), and the genital swellings (GS) remain unfused ***(Sadler, 2005).***

2. External Genitalia in the Male:

Development of the external genitalia in the male is under the influence of androgens secreted by the fetal testes and is characterized by rapid elongation of the genital tubercle, which is now called the phallus. During this elongation, the phallus pulls the urethral folds forward so that they form the lateral walls of the urethral groove. This groove extends along the caudal aspect of the elongated phallus but does not reach the most distal part, the glans. The epithelial lining of the groove, which originates in the endoderm, forms the urethral plate,

At the end of the third month the two urethral folds close over the urethral plate, forming the penile urethra. This canal does not extend to the tip of the phallus. This most distal portion of the urethra is formed during the fourth month, when ectodermal cells from the tip of the glans penetrate inward and form a short epithelial cord. This cord later obtains a lumen, thus forming the external urethral meatus,

The genital swellings, known in the male as the scrotal swellings, arise in the inguinal region. With further development they move caudally, and each swelling then makes up half of the scrotum. The two are separated by the scrotal septum ***(Sadler, 2005).***

Starting in the 4th month, the effects of dihydrotestosterone on the male external genitalia become

readily apparent. The perineal region separating the urogenital sinus from the anorectal canal begins to lengthen. The labioscrotal folds fuse in the midline to form the scrotum, and the urethral folds fuse to enclose the penile urethra. The penile urethra is completely enclosed by the 14th week. The formation of the distal glanular urethra may occur by a combination of two separate processes-the fusion of urethral folds proximally and the ingrowths of ectodermal cells distally. It is generally thought that the stratified squamous lining of the fossa navicularis results from an ingrowth of surface ectoderm as far proximally as the valve of Guerin ***(Kurzrock et al, 1999).***

Embryology of the Urethra:

The origin of the urethral plate is an outgrowth from the walls of the cloaca and urogenital sinus. Development of the urethra begins at the 4^{th} week**,** when the urethral plate is recognizable as a thickening of the anterior wall of the endodermal cloaca. A urethral groove is established by the development of the urethral folds on the ventrum of the phallic portion of the urogenital sinus on either side of the urethral plate. These folds are covered by surface epithelium and is named the primary urethral groove**.** Asecondary urethral groove develops at the 8^{th} week as a result of disintegration of the roof of the primary groove. Continuation of this process eventually establishes the definitive urethral groove (Figure: 3) ***(Stephens et al, 1996)***

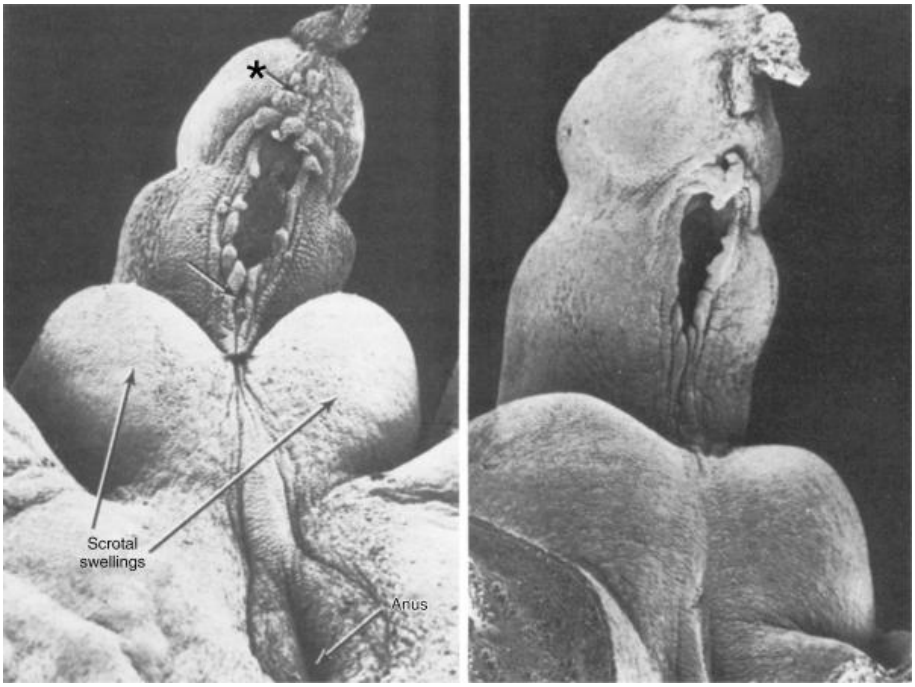

Figure 3: Development of the urethral plate and its canalization ***(Park, 2002).***

The proximal portion of the glanular urethra forms shortly there after and is thus derived from the urethral plate (endodermal origin). The distal portion of the glanular urethra is formed by lamellar ingrowth of the surface epithelium (ectodermal origin), which grows toward the distal extent of the urethral plate, becoming stratified squamous epithelium at the completion of development. This classic "ectodermal ingrowth theory"for development of the distal glanular urethra has recently been challenged by the "endodermal differentiation theory",

According to these investigators the urethral plate extends to the tip of the phallus and maintains patency and continuity throughout urethral development; therefore, the epithelium of the entire urethra originates from the urogenital sinus (endoderm). Sections of the distal glanular urethra showed no evidence of ectodermal tissue ingrowth ***(Kurzrock et al, 1999).*** (Figure: 4)

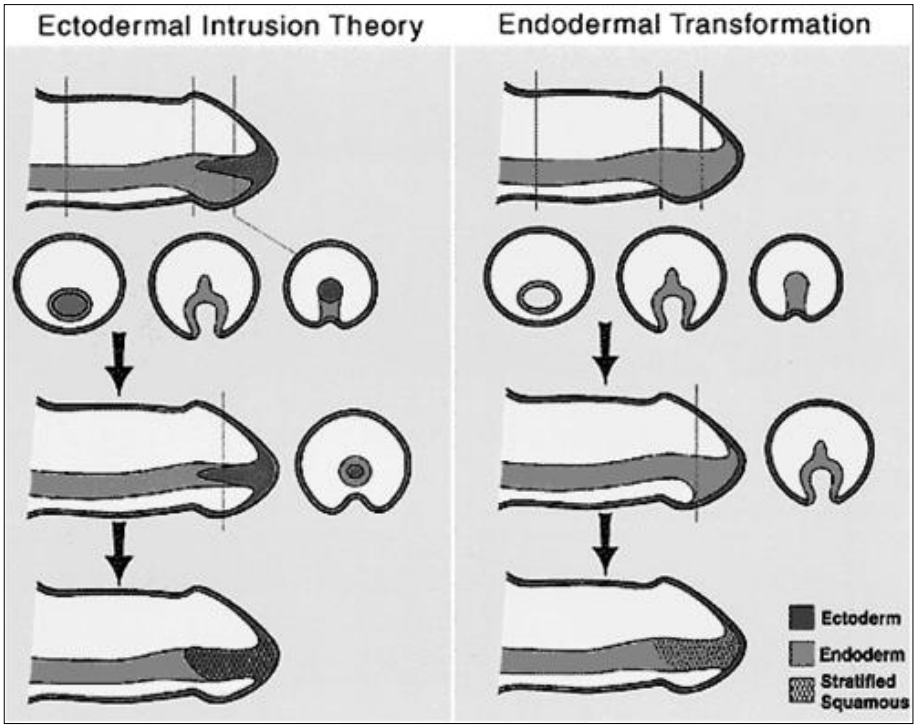

Figure 4: Embryology of the urethra ***(Retick and Borer, 2002).***

ANATOMICAL CONSIDERATIONS

Neurovascular Anatomy:

A study was made in the department of urology in the University of California, where a fetal penis at 33 weeks of gestation with distal shaft hypospadias was serially sectioned and compared to 10 normal human fetal specimens at 8 to 32 weeks of gestation.

Blood vessels were localized by the presence of red corpuscles and immuno-histochemical staining with Von-Willebrand's factor and factor VIII.

3D computer reconstructions of the nerves, corporeal bodies, tunica and urethra of the hypospadiac and normal fetal penises were compared.The results were equal except at the region of the abnormal urethral spongiosum and glans, the hypospadiac and normal penises showed no difference in neuronal innervations, corpora cavernosa and tunica albuginea architecture and blood supply.

The most striking difference was in vascularity. In the hypospadiac penis factor VIII, immunostaining revealed huge endothelial lined vascular channels filled with red blood corpuscles. In contrast, the normal penis had well defined small capillaries around the urethra that fanned out into the glans. Vascularity was also extensive under the urethral plate.

Nerve distribution in the abnormal glans was less extensive than in the normal penis.

In the hypospadiac penis, the abortive urethra is surrounded by an extensive vascularity, so isolating the urethral plate from this vascular nourishment by extensive mobilization under the urethral plate is not wise ***(Baskin et al, 1998).***

Anatomy of the Penis and Urethra:

The penis, the male copulatory organ, consists of an attached root in the perineum and a free normally pendulous corpus or body completely enveloped by coverings.

The root of the penis: The root of the penis comprises the three masses of erectile tissue in the urogenital triangle; the two crura and the bulb of the penis, firmly attached to the pubic arch and perineal membrane respectively ***(Sinnatamby, 1999).***

The crura of the penis: Each crus penis starts behind as blunt elongated round process, attached firmly to the everted edge of the ischiopubic ramus and covered by ischiocavernosus, anteriorly; it converges toward its fellow. Near the inferior symphysial border, the two crura blend sharply down and forward to become the corpora cavernosa ***(Sinnatamby, 1999).***