

Modified Koyanagi Technique versus Two Stage Repair for Severe Forms of Hypospadias in Children

Thesis

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List of Abbreviations

AMH	:	Anti Mullerian hormone
DSD	:	Disorders of Sexual Differentiation
GMS	:	Glans meatus and shaft for hypospadias evaluation
GUMS	:	Glans and urethral plate separately
HOPE	:	Hypospadias Objective Penile Evaluation
MD	:	Mullerian Duct
MDR	:	Mullerian duct remnants
MIS	:	Mullerian Inhibiting Substance
PAIS	:	Partial Androgen Insensitivity syndrome
PSH	:	Penoscotal hypospadias
PU	:	Prostatic Utricle
SEM	:	Standard error of the mean
UGS	:	Urogenital Sinus
VV	:	Voided volume

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Introduction

Hypospadias is one of the most common congenital anomalies occurring in approximately 1 of 200 to 1 of 300 live births (**Cheng et al., 2002**).

Hypospadias, in boys, is defined as an association of three anomalies of the penis: an abnormal ventral opening of the urethral meatus that may be located anywhere from the ventral aspect of the glans penis to the perineum, an abnormal ventral curvature of the penis (chordee), and an abnormal distribution of foreskin with a “hood” present dorsally and deficient foreskin ventrally (**Mouriquand et al., 1995**).

The only treatment of hypospadias is surgical repair of the anatomical defect. The fact that more than 300 different operations are described in the literature is a testament that treatment has not been perfected or standardized (**Baskin et al., 2001**).

Determining the appropriate technique depends on several factors including meatal location, appearance of meatus and glans, presence or absence of chordee, quality of ventral skin coverage and quality of the intact urethra (**Zaontz et al., 2002**). |

A controversy exists regarding the optimum technique for repair of severe hypospadias (**Elhalaby, 2006**). It is one of the most challenging conditions to correct. The multiplicity of procedures that have been described over the years is indicative of the fact that no procedure has been universally acceptable or successful. Many have chosen to perform staged procedures since this has the advantage that the varied anatomical issues can be fixed sequentially with different aspects of the problem being tackled in time. A disadvantage of this approach is that by necessity patients undergo at least two and often more procedures. (**Elkassaby et al., 2013**).[†] |

For many years there was a consensus that severe cases of hypospadias are better treated with a planned 2 stage approach rather than a single stage procedure. There has been a growing interest in one- stage repair of all varieties of hypospadias including severe types (**Elhalaby, 2006**). A prerequisite for a one-stage repair is the presence of appropriate dorsal hood foreskin for a preputial based island flap and the adequacy of penile length for a one stage repair (**Upadhyay and Houry, 2004**).

Many authors believe that a planned one stage repair can offer comparable results and may spare the patients further surgical interventions. Even if there is a higher chance of secondary procedures (20-30%), approximately two thirds of the cases of hypospadias will be corrected with one intervention. Furthermore, a well-performed single-stage repair does not compromise availability of vascular tissue for subsequent procedures (**Upadhyay and Houry, 2004**).

In 1984, Koyanagi et al., reported meatal based foreskin flap repair for proximal hypospadias. He used the inner layer of the preputial skin for urethral tubularization; this technique can simply be described as a two-step hypospadias repair completed in one stage (**Elkassaby ~~M~~-et al, 2013-**). It combines a meatal based flap and a pedicle island flap into single procedure. It allows for excision of ventral midline chordee without jeopardizing the flap (**Hassan et al., 2011**). It had a relatively high-complication rate, in part, because no major attempt was made to preserve the blood supply of the skin flaps. A modification of the technique was described, in which the vascularity of the flaps, resulted in reduction of complication rate. The higher success rate of the modified Koyanagi technique is believed to reflect the impact of preservation of the lateral blood supply to the skin flaps and not to rely entirely on the microvasculature emanating from the region of the urethral meatus and its surrounding corpus spongiosum. (**Elkassaby et al,et al., 2013**).

Aim of the Work

This thesis aims to compare and discuss the functional and cosmetic outcome of modified koyanagi procedure as a one stage repair versus two stage repair for severe forms of hypospadias as assessed by GMS score (*Merriman et al., 2013*) over 6 months postoperatively.

Chapter I

Preoperative management

I. Diagnosis and evaluation of hypospadias

1) Diagnosis:

A. Antenatal:

Hypospadias is usually an isolated malformation but sometimes it is associated with other malformations or may be a part of a syndrome. Detailed analysis of genital morphology is the only reliable way of antenatal diagnosis. Ventral or lateral curvature of the penis, associated with its shortening are the main findings in 2D ultrasound. Meizner described a specific signal known as a tulip sign present in severe hypospadias that is corresponding to the presence of a short penis ventrally curved in association with penoscrotal transposition of a bifid scrotum. The introduction of 3D ultrasound allowed more detailed evaluation of the surface structures of the fetus (*Teresa et al., 2012*).

B. Postnatal diagnosis:

i) Symptomatology:

Clinical symptoms vary, and depend on the severity of the disease. Children with proximal hypospadias with penile curvature might not be able to void while standing. We do not know precisely what degree of penile curvature in children will inhibit sexual intercourse in adulthood or what the long term psychosexual outcome will be in these patients (*Giannantoni, 2011*).

ii) Clinical examination:

The description of hypospadias should include the following:

- Position, shape, and width of the orifice.
- Presence of an atretic urethra and division of the corpus spongiosum.
- Appearance of the preputial hood and scrotum.
- Penile size.
- Curvature of the penis on erection

(Stein et al., 2012).

2) Classification and Evaluation of hypospadias:

Many classifications of hypospadias have been defined and published. Hypospadias is usually classified according to the anatomic location of the urethral orifice:

- (A) Anterior or distal hypospadias
- (B) Middle shaft or intermediate (penile) hypospadias.
- (C) Posterior or proximal (penoscrotal, scrotal, or perineal) hypospadias.

(Stein, 2012).

The severity of hypospadias cannot be solely based on the meatal location assessed at the first consultation. Additional indicators of severity are the size of the penile shaft, the glans width, the amount of dorsal foreskin, associated scrotal abnormalities and age at initial presentation. Assessment of the urethral plate and the penile curvature (erection test) are the main indicators of severity, since preservation or section of the urethral plate is an essential step in the selection of the reconstructive technique *(Snodgrass, 2011)*.

Several authors tried to explain different methods of classification of hypospadias. In 2011, **Mouriquand** described: 1) hypospadias with a distal division of the corpus spongiosum with little or no ventral curvature; 2) hypospadias with a proximal division of the spongiosum with a marked ventral curvature

related to the poor development of the ventral tissues sitting in the triangle described above, and sometimes related to an asymmetrical development of the corpora cavernosa; 3) hypospadias cripple who already underwent several procedures leaving behind scarred tissues. (**Mouriquand, 2011**). In 2011, **Macedo** classified hypospadias according to the difficulty of reconstruction. In this respect, the aspect of the urethral plate and the need for dividing it are the two main factors to categorize a hypospadias. He would therefore distinguish hypospadias with a preservable urethral plate (most distal and midshaft hypospadias and some proximal ones) from those that need division of the urethral plate and therefore a more extensive reconstruction. Hypospadias cripple should be individualized as a third group. In 2011, **Snodgrass** stated that Hypospadias should be described as primary versus reoperative. Within both groups, meatal location at the time of urethroplasty most likely is the best means for consistent, reproducible classification. It cannot be agreed on the extent of hypospadias in the patient that preoperatively has a penoscrotal opening that is revealed to be only a distal shaft.

Authors' opinions differ a lot. Given this widely variable presentation, as well as anatomical nuances that make every case unique makes it difficult to describe hypospadias in a concise and standardized manner. In an effort to address the need for standardized criteria to classify the severity of hypospadias, the GMS hypospadias scale was developed, which was developed as a mean to qualitatively score the severity of hypospadias based on easily observable features of the glans (G), meatus (M), and penile shaft (S). Each of the three components is scored numerically on a scale of 1-4 with more unfavorable characteristics being assigned higher values. These values are then summed to determine the GMS total score. The lowest possible GMS score, therefore, is 3 and the highest score is 12 (**Merriman et al., 2013**).