

Proximal hypospadias: current management

ESSAY

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TABLE OF CONTENTS

Page Number	Title
5	Table of Figures
9	List of Tables
10	Aim Of The Work
11	Abbreviations list
13	Introduction
17	PENILE COVERINGS AND ITS BLOOD SUPPLY
27-68	AN OVERVIEW ON PROXIMAL HYPOSPADIAS
27	1. Embryology
33	2. Etiology
36	3. Epidemiology
37	4. Anatomical abnormalities
38	5. Associated anomalies
39	6. Ambiguous external genitalia
43	7. Classification
47	8. Peri-operative special considerations
	i. Karyotype
	ii. Radiologic evaluation
	iii. Laboratory investigations
	iv. Diagnostic surgical intervention
	v. Hormonal manipulation
	vi. Timing of surgery
	vii. Hemostasis
	viii. Optical magnification
	ix. Tissue handling
	x. Neo-urethral stenting
	xi. Dressing
	xii. Post-operative medications
56	9. General principles of hypospadias repair
	i. Orthoplasty

	ii. Urethroplasty
	iii. Meatoplasty and glanuplasty
	iv. Scrotoplasty
	v. skin coverage
69-108	ONE STAGE URETHROPLASTY
69	1. Transverse preputial island flap (Duckett, 1981)
81	2. Vertical preputial island flap (VPIF)
82	3. Onlay island flap (OIF)
92	4. The tubularized incised plate (TIP)
96	5. The Koyanagi repair
102	6. Lateral based flap (LB flap)
105	7. The lateral-based onlay flap technique (LABO)
109	THE STAGED REPAIR
122-140	POST-OPERATIVE FOLLOW UP AND COMPLICATION
124	1. Assessment of adequate neourethral caliber and micturition function
	i. Neourethral Calibration
	ii. Uroflowmetry
125	2. assessment of the cosmetic appearance of the penis
131	3. Assessment of the Sexuality and psychosexual life
133	4. Complications
	i. Bleeding and hematoma
	ii. Edema
	iii. Infection
	iv. Penile torsion
	v. Urethral stent-related problems
	vi. Fistula
	vii. Meatal stenosis
	viii. Urethral stenosis

	ix. Persistent chordee
	x. Balanitis xerotica obliterans
	xi. Urethral Diverticulum
	xii. Dehiscence
144	REPORT DEMERDASH HOSPITAL EXPERIENCE OF REPAIR PROXIMAL HYPOSPADIAS CASES IN YEAR 2012
149	Summary
154	References
	Arabic Summary

TABLE OF FIGURES

Page Number	Title	Figure Number
19	Mid-shaft cross-section of the penis	1
22	Preputial blood supply	2
23	Vasculature of the outer and inner preputial layers	3
23	blood supply of the penile skin and prepuce and anastomosis between axial penile artery and the dorsal artery	4
24	Arterial blood supply of the erectile tissues	5
26	Penile anatomy in cross-section	6
27	Relation of the hindgut and cloaca at the end of the 4 th week	7
29	The comparative development of the female and male external genitalia	8
30	Detail in transverse section of the urethral groove, urethral folds, and urethral plate	9
31	The mechanism of the glandular urethral formation	10
32	Formation of prepuce	11
34	Testosterone biosynthesis and its defects	12
42	External genitalia of 46XX female with congenital adrenal hyperplasia at Demerdash hospital	13
43	Hypospadias classification	14
44	A case of proximal hypospadias with hypoplastic urethra at Demerdash hospital.	15

44	Hypospadias classification according to the position of the corpus spongiosum division	16
46	GMS scoring criteria	17
46	Representative photographs of the GMS scoring criteria	18
52	Urinary diversion using urethral stent	19
57	Degloving of the skin of the penile shaft in a case of penoscrotal hypospadias at Demerdash hospital.	20
58	Midline dorsal placcation	21
58	Nesbit technique	22
59	TAP technique	23
61	The urethral plate elevation technique	24
63	Penoscrotal transposition	25
64	Glenn-Anderson repair (scrotal rotational flaps)	26
65	Inguinal based groin flaps technique	27
66	The Z-scrotoplasty procedure	28
68	Mucosal collar	29
70	Modified Asopa or (Hodgson XX)	30
73	Tubularized Island flap with glans channel hypospadias repair	31
73	Tubularized preputial island flap	32
75	Two-layer closure of the neourethra and Hayashi's modifications of anastomsis between neourethra and native urethra	33
78	Patel (2005) modification of tubularized TPIF repair	34
80	Aoki (2008) Modification of TPIF	35

	for severe hypospadias	
82	Vertical preputial island flap	36
83	Onlay island flap	37
85	The double onlay preputial flap technique	38
86	Modified double face onlay island preputial skin flap with augmented glanuloplasty	39
87	The inlay-onlay flap	40
88	Onlay island flap after Lengthening the urethral plate with a double flap technique	41
89	Lengthening urethral plate with inner preputial skin grafts	42
90	Macedo and Srougi modification of OIF (The 'three-in-one' technique).	43
91	Onlay on tunica albugina	44
95	Proximal tubularized incised plate repair	45
98	Koyanagi flap	46
99	Emir's modification of koyanagi technique	47
100	Hayashi (2001) Modification of Koyanagi repair	48
101	Hayashi (2007) Modification of Koyanagi repair	49
104	The lateral based flap technique	50
106	The lateral-based onlay flap technique	51
112	Harvesting buccal mucosa	52
113	Final appearance of the buccal graft at the end of first stage	53
114	Staged repair using Byar's flap	54
115	Two-stage preputial graft	55

119	Inverted U first-stage Bracka procedure	56
120	Dorsal Dartos Flap as a Bed for Preputial Skin Graft in Primary Staged Hypo-spadias Repair	57
124	Uroflowmetry curve	58
131	HOPE-score reference pictures	59
145	An example on one stage repair of proximal hypospadias at Demerdash hospital.	60
146	an example on 1 st stage repair using Bayr's flap combined with bilateral orchiopexy at Demerdash hospital	61
147	an example on staged repair at Demerdash hospital complicated with post operative glans dehiscence	62
148	an example on staged repair at Demerdash hospital with satisfactory result	63

LIST OF TABLES

Page Number	Title	Table Number
17	Stretched penile length in normal males according to age	1
17	The urethral width in healthy uncircumcised boys.	2
39	Classification of disorders of sex development (DSD)	3
126	Standards of normal cosmetic appearance HOPE-score	4
127	The HOPE scoring system	5

Aim of work

To review the different reconstructive surgical techniques which are used in the management of proximal hypospadias and to highlight the new procedures including perioperative considerations, postoperative follow up and complications. Also to report Demerdash hospital experience regarding cases of proximal hypospadias over the last year.

Abbreviations list

<i>3β-HSD</i>	3 beta-hydroxysteroid deshydrogenase
5 α -DHT	5 alpha dihydrotestosterone
<i>17β-HSD</i>	17 beta-hydroxysteroid-dehydrogenase
AMH	anti-mullerian hormone
AR gene	Androgen receptor gene
ATF3 gene	AMP-dependent transcription factor 3 gene
BXO	Balanitis xerotica obliterans
CAIS	complete androgen insensitivity syndrome
DES	Diethylstilbestrol
DHCR7	7-dehydrocholesterol reductase
DHEA	Dehydroepiandrosterone
DSD	Disorders of Sexual Development
DTT	Dichlorodiphenyltrichloroethane
EDC	Environmental Disrupting Chemicals
ERb2	Estrogen Receptor beta 2
Fr.	French
GMS score	Glans, Meatus and Shaft score
GPSS	Genital Penile Perception Score
HCG	<i>Human Chorionic Gonadotropin</i>
HFGS	Hand–Foot-Genital syndrome
HOPE	Hypospadias Objective Penile Evaluation
HOSE	Hypospadias Objective Scoring Evaluation
HOX genes	Homeobox genes
HOXA13	Homeobox A13
HPPS	Hypospadias Penile Perception Score
LB flap	Lateral Based flap

LABO	Lateral-Based Onlay flap
LBD	Ligand-Binding Domain
<i>LH</i>	<i>Luteinizing Hormone</i>
MIS	Mullerian Inhibiting Substance
MRI	Magnetic Resonance Imaging
MURCS	mullerian aplasia, renal aplasia and cervico thoracic somite dysplasia
OIFA	Onlay Island Flap on Albuginea
OIF	Onlay Island Flap
P450scc	P450 side-chain cleavage
PAIS	partial androgen insensitivity syndrome
PDS	Polydioxanone sutures
PPPS	Pediatric Penile Perception Score
RNA	Ribonucleic acid
SLO syndrome	Smith-Lemli-Opitz syndrome
SRY	Sex-determining Region Y
STAR	Steroidogenic Acute Regulatory protein
TAP Technique	Tunica Albuginea Plication technique
TDF	Testis-Determining Factor
TIP repair	Tubularized Incised Plate repair
TPIF	Transverse Preputial Island Flap
VPIF	Vertical Preputial Island Flap
WAGR syndrome	Wilms tumour, Aniridia, Genitourinary abnormalities, and Mental Retardation syndrome
WT1 gene	Wilms Tumour 1 gene

Introduction

Hypospadias is an association of three anatomical anomalies of the penis: an abnormal ventral opening of the urethral meatus which can be located anywhere on the ventral aspect of the penis, an abnormal ventral curvature of the penis (chordee) and an abnormal distribution of the foreskin around the glans with the ventrally deficient hooded foreskin. The chordee is a common finding but not a constant one. **(Mouriquand et al, 1995)**

Hypospadias is the most common congenital anomaly of the penis, affecting 0.4–8.2 of 1000 live male babies (10–15% of which are proximal hypospadias). **(Giannantoni, 2011)**

Present systems for categorizing hypospadias are based on the location of the external meatus. The assessment is carried out prior to surgery or after penis straightening procedures. **(Orkiszewski, 2012)**

The meatal position can be classified as anterior or distal (glanular, coronal, subcoronal), middle (midpenile), or posterior or proximal (posterior penile, penoscrotal, scrotal, perineal). **(Leung and Robson, 2007)**

Ectopically placed urethral meatus on the ventral aspect of the penis and proximal to the normal site is the mainstay element of the diagnosis which may be associated with ventrally deficient prepuce, ventral curvature of the penis (Chordee), torsion of the penis or Penoscrotal transposition. **(Snodgrass, 2012)**

Approximately 5–10% of boys with hypospadias have cryptorchidism rising to about 50% in those with the severe perineal or penoscrotal forms and these severe forms are also associated with a persistent prostatic utricle in 20% of cases. **(Wilcox and Mouriquand, 2008)**

The simultaneous occurrence of hypospadias with cryptorchidism increases the likelihood for disorders of sexual development (DSD). **(Snodgrass, 2012)**

The improvement of pediatric anesthesia allows hypospadiologists to operate upon the child at a younger age, the best between 6-18 months, giving the advantages of better healing and sparing the child of carrying the mental trauma of surgery to adulthood. **(Snodgrass, 2008)**

The goals in management of hypospadias repair are creating a straight penis, creating a urethra of adequate length and uniform caliber, reconstructing slit-like meatus at the tip of glans penis, symmetry in appearance of glans and penile shaft and projectile stream, these goals can be achieved by orthoplasty, urethroplasty, meatoplasty, glanuloplasty, scrotoplasty and skin cover. **(Bhat, 2008)**

Orthoplasty includes any procedure which aims to correct the ventral curvature of the penis. Straight penis is the first requirements for a successful repair. Complete penile curvature correction should be tested on table before proceeding for the urethroplasty. **(Bhat, 2008)**

Urethroplasty means Reconstruction of neourethra. It can be performed in a single stage or in a two-stage procedure. **(Wilcox and Mouriquand, 2008)**