Proximal hypospadias: current management

ESSAY
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By
Ahmad Mohammad Abd Al Rahman
M.B.B.Ch.

Supervisors

Prof. Dr. AMR MOHAMMAD AL-SADEK NOWIER

Professor of Urology
Ain Shams University - faculty of medicine

Dr. SAMIR SAYED MUHAMMAD AZAZY

Lecturer of urology
Ain Shams University - faculty of medicine

Faculty of Medicine
Ain Shams University
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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

3β-HSD	3 beta-hydroxysteroid deshydrogenase
5α-DHT	5 alpha dihydrotestosterone
17β-HSD	17 beta-hydroxysteroid-dehyrogenase
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
GPPS	Genital Penile Perception Score
HCG	*Human Chorionic Gonadotropin*
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
LH	*Luteinizing Hormone*
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)**