Hypospadias

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Adapted from the Handbook of Pediatric Urology
Lippincott-Raven, 1997, Baskin, Kogan and Duckett

I. Introduction
A. Hypospadias is a congenital defect of the penis, resulting in incomplete development of the anterior urethra, corpora cavernosa and prepuce (foreskin).
B. Clinically, the hypospadiac urethral meatus does not cause significant urinary symptoms other than a urinary stream which may be deflected in a downward fashion.
C. Hypospadias is also associated with penile curvature and may result in infertility secondary to difficulty in semen delivery.

II. Embryology
A. At one month gestation the male and female genitalia are essentially indistinguishable.
B. Under the influence of testosterone the male external genitalia become masculinized.
C. By the end of the first trimester and the beginning of the 2nd trimester the penile urethra and accompanying prepuce are completely formed.
D. Abnormalities in this development can lead to hypospadias and associated penile anomalies.
E. In hypospadias, incomplete development of the glandular urethra does not allow the preputial folds to fuse.
F. Consequently, in hypospadias the foreskin is absent on the ventrum and there is excessive foreskin on the dorsal surface (dorsal preputial hood).

III. Classification
A. Hypospadias can be classified as to the location of the urethral meatus without taking into account penile curvature.
B. A more useful surgical classification is the location of the urethral meatus after penile straightening where:
  1. 50% of the patients will have anterior hypospadias with the meatus on the glans or subcoronal (Figure 1).
  2. 20% will have the urethral meatus on the penile shaft.
  3. 30% will have the meatus between the perineum and the penoscrotal junction.

IV. Incidence, Genetics and Etiology
A. Hypospadias occurs in ~one per three hundred live male births. Recent reports in Europe and the United States suggest that the rate of hypospadias is increasing.
B. There is a 14% incidence in male siblings and an 8% incidence in offspring.
C. Most cases of hypospadias do not have an identifiable cause. Rare cases can be attributed to abnormalities in androgen metabolism (Type II 5 'reductase deficiency) or defects in the androgen receptor. Unproven theories explaining hypospadias currently under study are genetic and environment factors such as endocrine disrupters or environmental toxins.
V. Associated Anomalies
A. Undescended testes occur in ~9% of patients with hypospadias.
B. An increased incidence of up to 30% in patients with penoscrotal or more severe hypospadias.
C. Inguinal hernias also occur in ~9% of patients with hypospadias.
D. A utricle or Mullerian remnant in the posterior urethra is found in a high percentage of patients with severe hypospadias.
E. Associated urinary tract anomalies are infrequent in patients with isolated hypospadias because the external genitalia are formed much later than the kidneys, ureter and bladder.
F. Patients with hypospadias and an undescended testicle or an inguinal hernia do not need further urinary tract evaluation such as an ultrasound or echocardiogram.
G. Patients who have hypospadias in association with other organ system anomalies such as a cardiac murmur, imperforate anus or pyloric stenosis require renal and bladder imaging with an abdominal ultrasound.
H. **Patients with severe hypospadias and undescended testes should be investigated for intersex with a karyotype and further endocrinologic work-up.**

VI. Treatment of Hypospadias
A. There are five basic phases for the successful reconstruction of the hypospadiac penis:
1) Creation of a normal urethral meatus and glans penis.
2) A straight penis.
3) A normal urethra.
4) Skin covering.
5) Normal position of the scrotum in relationship to the penis.

VII. Timing of Surgery
A. Hypospadias surgery is best performed between the ages of six months and eighteen months, prior to toilet training and during the
psychologic window when genital awareness has not been
recognized by the patient.
B. Outpatient surgery is now the standard of care. The majority of
hypospadiac defects can be repaired in a single-stage operation,
with severe cases requiring a staged procedure. Early hypospadias
repair with minimal hospitalization helps to avoid separation
anxiety and castration fears.

VIII. Anaesthesia
A. Hypospadias surgery is performed under general anesthesia.
B. A penile nerve block or a caudal supplementation is standard to
minimize post-operative discomfort.

IX. Hypospadias Operations
Hundreds of techniques to correct hypospadias have been
published in the medical literature.
Some of the standard repairs are listed below.
A. Meatal advancement and glanuloplasty procedure (MAGPI).
B. Mathieu procedure or Flip-Flap procedure.
C. Primary closure of the urethral plate with and without incision.
D. Onlay island flap procedure.
E. Transverse tubularized island flap.
F. Buccal mucosa grafts (taken from the inside of the cheek) can
be used for urethral replacement typically in a secondary procedure
when local vascular flaps have failed.
F. Straightening the penis can be performed with a dorsal tunica
plication procedure.

X. Complications of Hypospadias Surgery
A. Urethral cutaneous fistula.
• 1. Urethral cutaneous fistula consists of a communication
   between the new urethra and the penile skin, typically allowing
   the urine to come out through two separate holes.
• 2. Fistulas require operative closure ~ 6 months after the initial
   operation when tissue swelling has subsided.
B. Meatal stenosis
• 1. Strictureing of the new urethra which can occur anywhere along the urethroplasty, but most commonly occurs in the glans penis.

C. Urethral diverticulum
• 1. A large outpouching or ballooning of the urethra secondary to too large of a urethroplasty or an obstruction distally (i.e. meatal stenosis).

D. Superficial skin loss
• 1. A relatively common complication after hypospadias surgery and typically will heal spontaneously without the need for further surgical intervention by secondary penile skin granulation. This can be treated with local wound care and daily bathing.

E. Residual penile curvature. If severe reoperation with penile straightening is required.

XI. Postoperative Hypospadias Care
A. Anterior hypospadias repair such as the MAGPI are often done without the use of a drippy stent or catheter and require no special treatment.
B. More severe hypospadias requires the use of an indwelling drippy stent which is typically removed 7-14 days after surgery by cutting a stitch that secures the urethral catheter to the glans penis.
C. Prophylactic doses of antibiotics such as Bactrim or Nitrofurantoin are often prescribed while the stent is in place to keep the urine sterile.
D. Postoperative Symptoms Include:
• 1. Bladder spasms which can be treated with Ditropan (oxybutynin).
2. Urinary retention is uncommon, but can occur secondary to a stent malfunction such as blockage or kinking.
3. Postoperative pain is controlled with Tylenol and Tylenol and codeine elixir.

E. Dressings
1. The most common dressing used after hypospadias is a plastic dressing such as Tegaderm which is used with gauze to sandwich the penis onto the abdomen and is typically removed at home 2-3 days after surgery.

See Hypospadias Handout for Figures

References
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