Hypospadias

Description/Etiology

Hypospadias is a congenital anomaly of the penis characterized by abnormal position of the urinary meatus (i.e., opening) on the ventral surface (i.e., underside) of the penis anywhere from the glans penis to the perineum. This type of genitourinary defect can reduce quality of life (QOL). Depending on the degree of defect, hypospadias can interfere with the ability to urinate while standing and in severe cases, with male fertility.

The etiology of hypospadias is thought to be multifactorial, including genetic factors, hormonal, and environmental influences during embryonic development. Hypospadias often occurs in association with chordee (i.e., ventral curvature of the penis) and dorsal foreskin hood. Additional genitourinary abnormalities (e.g., cryptorchidism [i.e., undescended testicles], inguinal hernia, hydrocele) can also occur. Severe cases are typically associated with chromosomal abnormalities. Diagnosis is generally made during the neonatal physical examination. Intersex evaluation—including ultrasound and chromosomal studies—should be performed when hypospadias is associated with cryptorchidism.

Surgery is the only treatment option for hypospadias. The goal of corrective surgery is proper placement of the urethral opening to allow urination while standing, enable full sexual function, and to make the penis look as normal as possible. The American Academy of Pediatrics (AAP) recommends that surgery be performed at 6–12 months of age. However, the surgery can be performed in infants as young as 4 months of age.

Facts and Figures

Hypospadias is the second most common male genital malformation, affecting 1:250 boys. It is more common in some geographic areas; for example, the prevalence is ~ 0.26:1,000 births in Mexico, 2.1:1,000 in Hungary, and 2.6:1,000 in Scandinavia. In the United States, it is more common in whites than in Hispanics or blacks. Based on the location of the urethral opening, hypospadias is classified as anterior or distal in 50–65% of cases, middle in 20–30%, and posterior or proximal in 10–30%. Hypospadias is seen in 8% of fathers of affected boys and 14% of male siblings, suggesting a genetic component. Although hypospadias often occurs in isolation, it is a component of more than 200 syndromes. Up to 16% of affected boys have additional congenital abnormalities (e.g., cryptorchidism [10–15%], inguinal hernia [9–15%], hydrocele [9–16%]).

Risk Factors

Risk factors include advanced maternal age, preexisting maternal diabetes mellitus, and poor intrauterine growth. Exposure to endocrine disruptors can contribute to hypospadias development. The role of early progestin exposure is controversial; progestins taken for threatened abortion or as part of assisted reproduction during the first trimester appears to increase risk, but contraceptive progestin use does not.

Signs and Symptoms/Clinical Presentation

› Urinary meatus is usually on ventral surface of the penis
› The affected toilet-trained child has trouble urinating while standing so that he usually sits down to urinate; abnormal urine spraying might be noted if urinating while standing
› Chordee can be present
**Assessment**

› **Patient History**
  - Assess for family history of hypospadias
  - Assess for history of infertility and assisted reproduction

› **Physical Findings of Particular Interest**
  - See Signs and Symptoms/Clinical Presentation, above

› **Laboratory Tests**
  - Karyotyping, molecular, and biochemical tests to assess for intersex conditions in affected boys with hypospadias and cryptorchidism

› **Other Diagnostic Tests/Studies**
  - Genital ultrasound, MRI, or CT scan to show other congenital anomalies sometimes associated with hypospadias (e.g., inguinal hernia, enlarged prostatic utricle)

**Treatment Goals**

› **Maintain Physiological Status Per Facility Protocol and Reduce Risk for Complications**
  - Follow facility pre- and post-surgical protocols if patient becomes a candidate for surgical repair of hypospadias; reinforce pre- and post-surgical education and verify parental completion of facility informed consent documents
  - Reinforce the treating clinician’s explanation of the surgical procedure and associated risks and answer parental questions
  - Following surgery, monitor vital signs and frequently assess procedure site; observe for postsurgical complications (e.g., bleeding, infection); immediately report abnormal findings to the treating clinician and treat, as ordered
  - Confirm that child is hemodynamically stable
  - Make sure that the child is comfortable; use comfort measures (e.g., pillows for positioning) and avoid irritation (e.g., from diaper) to the affected area
  - The child is normally comfortable because of local anesthetic, but an analgesic can be ordered for pain management

› **Provide Emotional/Psychological Support and Educate**
  - Assess anxiety level and coping ability of parents; educate and encourage discussion about hypospadias etiology, surgical correction, potential complications, and post-surgical care
  - Inform parents that penile swelling and redness at hospital discharge is normal and should diminish within 3 days
  - Explain that the dressing can fall off, and to apply prescribed antibiotic ointment should this occur
  - Tell the parents that the child should avoid excessive activity after surgery to avoid penile injury (e.g., if age-appropriate, avoid bicycles and gym bars)
  - Emphasize importance of infection prevention (e.g., do not bathe child until stent has been removed)

**Food for Thought**

› Investigators found that out of 74 adult patients who experienced complications of hypospadias before a surgical repair they had in childhood, over 50% experiences urethral stricture disease and 57% of these patients had to have repeat surgery (Hoy et al., 2017)

› Early surgical intervention is recommended for several reasons, including documented reduced postoperative discomfort in younger patients, minimal penile growth during first years of life, development of genital awareness at ~ 18 months, and avoidance of surgery during the “Terrible Twos” (characterized by uncooperative toddler behavior)

› Single-stage procedures for hypospadias repair are associated with high complication and reoperation rates of. Many experts believe that two-stage procedures—including Bracka two-stage graft repair—are preferred for treating proximal hypospadias. Researchers in India reported good results in terms of restoration of normal function and satisfactory cosmetic results with minimal complications in 30 patient who underwent procedure (Joshi et al., 2015)

**Red Flags**

› Infants with hypospadias should not be circumcised at birth; the foreskin should be preserved for use in later surgical repair

› Complications of hypospadias repair include hematoma, meatal stenosis, urethral stricture, urethrocutaneous fistula, urethral diverticulum, wound infection, and impaired healing

**What Do I Need to Tell the Patient’s Family?**

› Provide parents with instructions on home postoperative care; emphasize the importance of medication adherence (e.g., antibiotics, analgesics, anticholinergics)
Advise parents to inform the treating clinician if there is additional penile swelling, redness, or purulent discharge. Foul-smelling or cloudy urine can be an indication of infection.

Urge parents to contact the treating clinician for fever > 101°F/38.3°C

Reassure parents that results of surgery are typically good, both cosmetically and functionally; patients are able to stand to void, and sexual activity and fertility are generally satisfactory.

References


