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Neha Madhok, Kathryn Scharbach and Sara Shahid-Saless

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In Brief

Hypospadias

Neha Madhok, MD
Kathryn Scharbach, MD
Sara Shahid-Saless, MD
Children's Hospital at Montefiore,
Bronx, NY

Author Disclosure

Drs Madhok, Scharbach, Shahid-Saless, and Adam have disclosed no financial relationships relevant to this article. This commentary does not contain a discussion of an unapproved/investigative use of a commercial product/device.

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Hypospadias, derived from the Greek words "hypo" meaning below and "spadon" meaning rent or hole, is defined by a urethral meatus that opens on the ventral surface of the penis proximal to the normal glanular location. Hypospadias is the most common malformation of the male genitalia and occurs in 1 in 300 male births, with some studies demonstrating a doubling of the rate in the United States between 1970 and 1993. Depending on the degree of anatomic defect, affected patients may be unable to urinate while standing and can experience issues regarding body image, sexual intercourse, and relation-

ships. Surgical repair is the only treatment.

A review of embryology is useful in understanding the nature of this defect. As the phallus elongates at 7 to 8 weeks gestational age, the urethral groove develops on the ventral surface of the penis. The two edges of the urethral groove fold together and fuse in a proximal-to-distal direction, forming the urethra, with the meatus located on the glans. Finally, the prepuce originates from the periphery of the glans penis and is complete by 14 weeks' gestation. Incomplete fusion of the urethral folds results in hypospadias, a urethral opening on the ventral surface of the penis or on the scrotum. The development of the male external genitalia depends on the presence of testosterone and its metabolites as well as a functional androgen receptor.

Despite extensive investigation, the cause of hypospadias remains unknown for most patients. Even in severe forms, the underlying cause is never identified in as many as 70% of cases. Although androgens clearly are critical for penile development, defects in androgen metabolism and the androgen receptor account for an extremely small subset of cases, implying that other factors are responsible for hypospadias.

Epidemiologic studies have identified some genetic, maternal, and fetal factors as well as environmental exposures associated with hypospadias. A genetic component in the development of hypospadias is suggested by a 14% recurrence rate in male siblings and an 8% recurrence rate in male offspring of men who have hypospadias. Maternal and fetal factors that increase the risk of hypospadias include

advanced maternal age, pre-existing diabetes mellitus, and poor intrauterine growth. Studies have failed to show any discernible risk associated with gestational diabetes; hypertension; pre-eclampsia; or maternal use of alcohol, tobacco, or other substances.

Exposure to "endocrine disruptors" has been postulated to contribute to the development of hypospadias. High concentrations of estrogenic environmental contaminants and antiandrogen compounds have been shown to impair penile development in animal models. In humans, epidemiologic studies have revealed an increased incidence of hypospadias subsequent to maternal diethylstilbestrol intake during early pregnancy. A higher incidence also has been observed in the male offspring of vegetarian women and has been suggested to be a result of an increased exposure to phytoestrogens.

Whether early exposure to progesterone increases the risk of developing hypospadias is controversial. Some studies suggest that progesterone taken for threatened abortion or as part of assisted reproduction techniques during the first trimester may be associated with increased rates of hypospadias. Progesterone taken for the purpose of contraception, however, has not shown the same association. Additional research is needed to establish the biologic link between hypospadias and these risk factors.

To classify hypospadias accurately, the exact anatomic location of the meatus should be described as well as the presence or absence of chordee, defined as ventral curvature of the penis. The meatal position is classified most accurately after correction of chordee into the following three categories: dis-

tal (glandular, coronal, subcoronal), middle (distal shaft, midshaft, posterior penile), or proximal (penoscrotal, scrotal, perineal). Approximately 60% of cases of hypospadias are distal, with the subcoronal position being the most common; 25% are midpenile; and 15% are proximal. The appearance of the foreskin often is described as a dorsal hood, that is, thin or absent on the ventral surface of the penis while abundant on the dorsal aspect. Of note, in the rare variant called megameatus intact prepuce, the foreskin may be complete and normally developed. In this case, the hypospadias may be revealed only at the time of circumcision after the dorsal slit is performed. If this situation arises, the circumcision should be stopped and a urologic evaluation requested.

The clinical presentation of hypospadias has a broad spectrum, depending on the severity. Children who have a mild form in which the meatus is located on the glans may have a completely normal urinary stream, whereas children who have the more severe proximal forms of hypospadias may have a narrow meatus with a weak, downward-angled stream. Affected boys may be unable to urinate standing upright, and patients who have severe lesions may have difficulties with fertility because abnormal deflection of ejaculate may prevent effective insemination. The presence of chordee may cause painful erections and, in severe cases, preclude vaginal penetration during intercourse.

Although hypospadias, particularly the distal and middle forms, usually is an isolated anomaly, it can be one of the features included in more than 200 syndromes. Associated malformations of the urinary tract are seen most often with proximal hypospadias, where a direct association exists between the severity of hypospadias and the frequency of an associated anomaly. Anomalies of the genital tract, such as

cryptorchidism and inguinal hernia, are most common, with an incidence of 8% to 10% and 9% to 15%, respectively. Other urinary tract anomalies, such as vesicoureteral reflux, ureteropelvic junction obstruction, pelvic or horseshoe kidney, crossed renal ectopia, and renal agenesis, occur in 1% of children who have distal and 5% of children who have proximal hypospadias.

The association of distal and middle forms of hypospadias with renal anomalies is no greater than that of the general population, making it unnecessary to perform renal ultrasonography or voiding cystourethrography in such patients. Proximal hypospadias, however, requires a more complete evaluation. Clinicians should suspect the possibility of an intersex condition if a child who has hypospadias has cryptorchidism and one or both testes are not palpable. In this case, karyotyping and ultrasonography of the urinary tract and internal genital organs should be performed.

The degree of hypospadias and presence of associated anomalies are useful in describing the forms of hypospadias and in determining surgical repair. Most importantly, because the foreskin is used in hypospadias repair, circumcision is contraindicated. Patients who have hypospadias should be referred for surgical evaluation in the first few weeks after birth. Such a timely evaluation addresses parental concerns and questions and establishes a relationship with the surgeon. However, the optimal time for hypospadias repair is at 6 months of age or shortly thereafter. Correction of any complications should be delayed until at least 6 months after the initial repair.

An effort should be made to avoid genital surgery around the toddler age, when toilet training and uncooperative behavior may be issues. Therefore, surgery that does not occur or is not complete during infancy should be delayed until the child is 4 years of age.

Today, outpatient surgery is the standard of care. A shortened hospital stay is believed to reduce the psychological impact and separation anxiety involved.

The goal of hypospadias surgery is to create proper anatomic placement of the urethral meatus with good cosmesis and function. More specifically, an optimal repair results in a straight penis with a slitlike meatus on the glans, leading to a forward-directed urinary stream and normal intercourse. Many surgical techniques are used, with the method of repair dictated by the anatomic variables, as outlined previously. Straightforward cases of hypospadias can be addressed with a single procedure; a staged approach may be necessary in more complex forms. Families should be informed about the probability of reoperation in the case of complications.

The factors that affect the success and complication rate of the operations include severity of the deformity, previous operation or circumcision, age of the patient, postoperative care, and experience of the surgeon. Early complications of hypospadias repair include infection, tissue necrosis in the operative area, development of a fistula or diverticulum, urethral or meatal stenosis, unacceptable cosmetic outcome, and functional complaints of micturition (spraying, dribbling, and deviation of urinary stream). Although there is a correlation between the short-term and long-term results of hypospadias repair, many complications do not appear for more than 1 year after surgery. In fact, some may not manifest for more than a decade. Later complications also include fistula, diverticulum, urethral or meatal stenosis, dissatisfaction with penile appearance, and functional complaints of micturition. Among the other poor long-term outcomes are erectile problems and difficulties with sexual performance. With some complications of hypospadias repair occurring so long after the surgery, the final outcome can

be evaluated only after the patient has reached adulthood.

Comment: Hypospadias is a defect in which the urethral meatus opens on the ventral surface of the penis proximal to its normal position. A much rarer

congenital defect is epispadias, which can affect both boys (~1 in 100,000) and girls (~1 in 500,000). In boys who have epispadias, the urethral meatus is on the dorsal surface of the penis, and in girls, the opening is displaced anteriorly, usually in the clitoris or between

it and the labia. Most cases of epispadias are associated with exstrophy of the bladder or, even more extreme, with cloacal exstrophy.

Henry M. Adam, MD
Editor, In Brief

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