Hypospadias

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Introduction

Hypospadias defines a congenital anomaly of the penis in which the urethral meatus opens proximal to the normal location. Most commonly, the meatus is located on the proximal glans or in the region of the coronal margin, but it can be found on the penile shaft, within the scrotum, or on the perineum. Typically, boys also have an incomplete dorsal foreskin and may exhibit ventral bending of the penis. Within the spectrum of related anomalies are boys with the urethral meatus properly located on the glans who have an incomplete foreskin and may also have ventral curvature, so-called *chordee without hypospadias*.

Etiology and Epidemiology

Hypospadias is considered an arrest in normal penile development occurring between the 9th and 20th weeks of gestation. Since elongation of the genital tubercle and fusion of the urethral folds are hormone-dependent events, disruption of normal masculinization is considered an underlying cause of hypospadias, although postnatal testing only rarely reveals detectable defects in testosterone production, 5 alpha-reductase II activity, or the androgen receptor [1].

Hypospadias is the 2nd most common anomaly in boys, found in approximately 1:150. The incidence increases in association with low birth weight, twins, and maternal age over 35 years. The likelihood that a boy will have hypospadias increases to approximately 10 % if his father or sibling is affected.

Although hypospadias can occur in association with syndromes, in most cases, it is an isolated anomaly.

Presentation

Hypospadias usually is visibly apparent during the newborn examination (Fig. 30.1) and sometimes is suspected on prenatal ultrasonography. The most obvious finding is the incomplete foreskin. Some practitioners refer to the appearance as a "natural circumcision" since the glans is visible, but the term is misleading and should not be used. When a dorsal foreskin is detected, examination next determines location of the meatus. The diagnosis is "chordee without hypospadias" when the meatus is in a normal position and hypospadias when it is found proximally. The glans also is incompletely fused in the ventral midline when the meatus is proximal.

A subset of hypospadias termed the megameatus intact prepuce variant occurs with a normally

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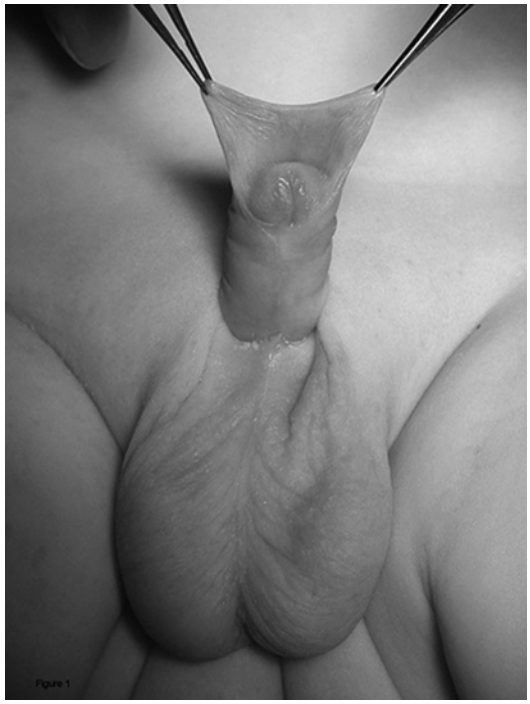

Fig. 30.1 Hypospadias is visible during newborn examination

formed foreskin (Fig. 30.2) and so may not be detected until newborn circumcision is performed or the foreskin becomes retractable later in life.

Physical examination should also note position of the testes, and the finding of both a penile anomaly and undescended testis indicates a possible disorder of sexual differentiation. Other findings may include a deep cleft in the midline of the scrotum and/or transposition of the scrotum alongside and even above the penile shaft in proximal hypospadias.

Some practitioners confuse a penoscrotal to perineal hypospadias with intersex disorders, but the diagnosis of disordered sexual differentiation is limited to cases with associated gonadal anomalies.

Evaluation

The diagnosis is established by physical examination. Radiologic testing of the urinary tract is not necessary, even with proximal hypospadias,

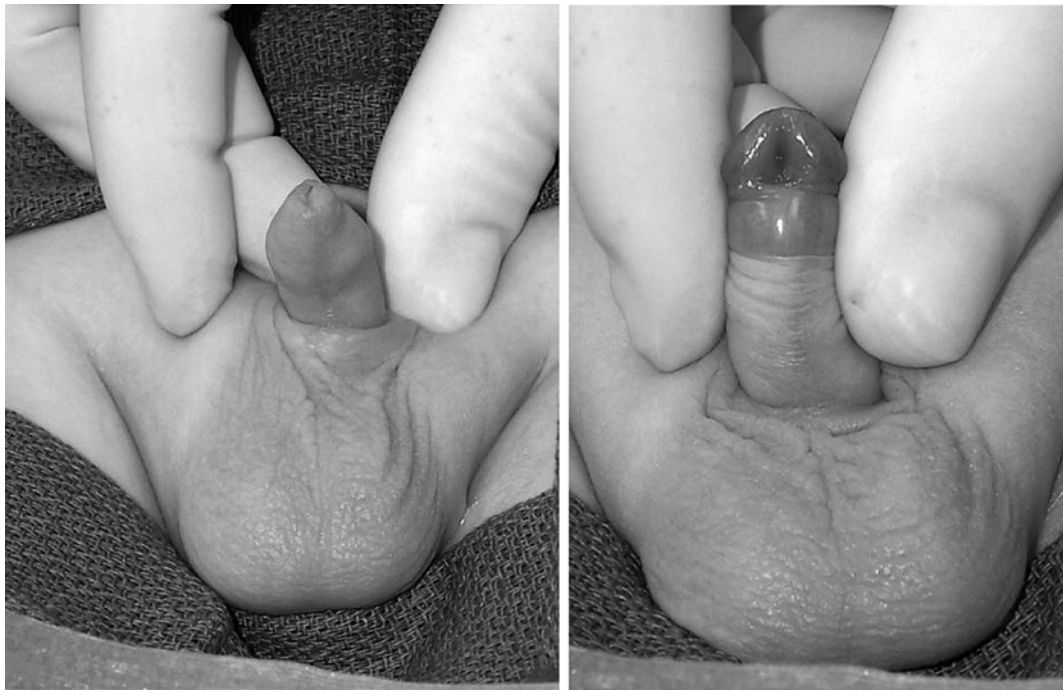

Fig. 30.2 Megameatus intact prepuce (MIP) hypospadias variant

since renal development occurs at 4 weeks whereas penile formation happens later beginning at 9 weeks.

A karyotype should be obtained in newborns with both an undescended testis and hypospadias. Incidence varies from approximately 15 % if the testis is palpable to nearly 50 % for proximal hypospadias with a nonpalpable testis [2]. Possible findings include mixed gonadal dysgenesis and true hermaphroditism, while bilateral nonpalpable testes should raise suspicion of female pseudohermaphroditism due to adrenal hyperplasia regardless of the extent of penile development. Otherwise, neither chromosome analysis nor endocrine studies are needed in infants with isolated hypospadias.

Reasons for Correction

Most often, hypospadias presents with a meatus on the proximal glans or at the corona. These patients will not have impaired fertility from the abnormal opening, but are likely to have difficulty aiming their urinary stream if left uncorrected. Normal voiding depends upon fusion of the glans over the urethra to direct the stream forward and focus it without lateral spaying. In addition, often a transverse web of skin is found just distal to the abnormal meatus that can defect the stream downward.

The dorsal foreskin also calls attention to the anomaly, not only in the newborn nursery but also in the locker room as boys mature. Hypospadias repair includes either circumcision or reconstruction of the foreskin to resemble a natural penis.

Ventral curvature is found in approximately 15 % of boys with distal forms of hypospadias and in over 50 % of those with the meatus on the proximal penile shaft or in the scrotum or perineum. Bending in distal cases most often is not so severe to cause difficulties with intercourse but in proximal hypospadias may preclude penetration.

Proximal cases additionally may have deep midline clefts in the scrotum and/or transposition of the scrotum higher than usual alongside the penile shaft. These findings combine with downward curvature of the penis to create a feminized appearance to the external genitalia.

Given these concerns and modern outcomes of surgical correction, repair is recommended for all but the most distal cases. A meatus that is located on the proximal glans but has 2 mm or more glans fusion ventrally does not present either a cosmetic or functional defect. If detected following circumcision, no additional repair is needed, but surgery to perform circumcision or foreskin reconstruction would still be advised for a dorsal prepuce.

Timing of Surgery

A 1996 position paper from the American Academy of Pediatrics suggested the optimal time for elective genital surgery including hypospadias repair is between 6 and 18 months, to avoid intervention after the time genital awareness begins. Anesthetic considerations delay elective day surgery until after 2 months in full-term, healthy infants. During this time, the normal postnatal testosterone surge stimulates penile growth to a size that remains constant in relation to overall body size until puberty. Today, many pediatric urologists proceed with correction anytime after 3 months of age in full-term, healthy boys. Referral can be done soon after birth, in part to allay family concerns about the condition since few parents are aware that hypospadias exists.

Preoperative hormonal stimulation with either injectable or topical testosterone may be recommended when the glans appears small, almost exclusively in those with proximal hypospadias.

Outcomes of Surgery

Today, the majority of operations are performed in infants in a single-stage, outpatient repair. Although several hundred techniques have been described since hypospadias repair began in the late 1800s, only a few are in common use today [3]. Most pediatric urologists repair distal cases by tubularizing the urethral plate (Fig. 30.3),

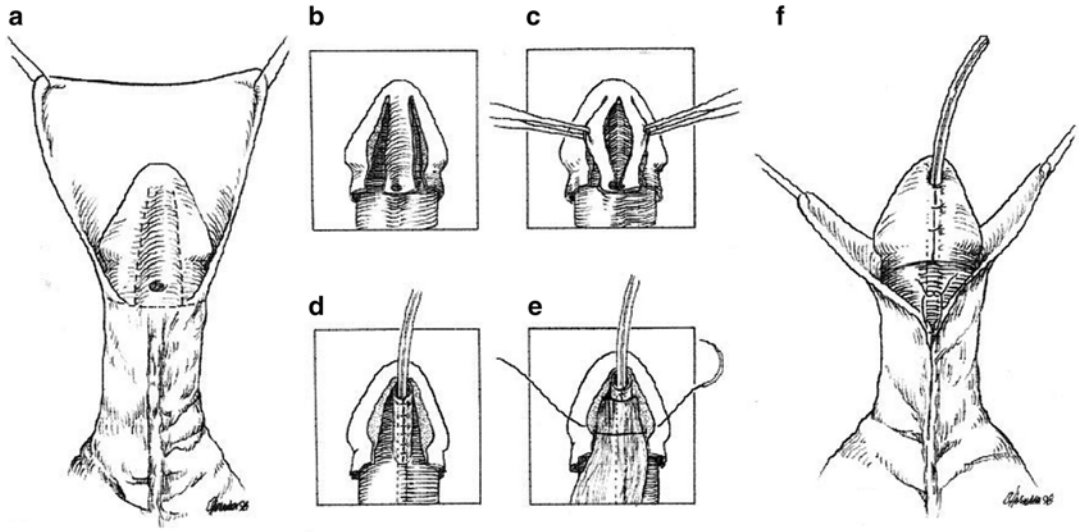

Fig. 30.3 Tubularized, incised plate hypospadias repair. (**a**) *Horizontal dotted line* indicating circumscribing incision approximately 2 mm proximal to the meatus. *Vertical dotted lines* indicate the junction of the urethral plate to the glans wings. (**b**) Urethral plate is separated from the glans wings, which are then mobilized laterally. (**c**) The key step of the operation is a deep, midline incision into the urethral plate extending from within the meatus to its distal margin, but not continuing into the glans apex. (**d**) The plate is tubularized over a small stent leaving a generous, oval meatus. (**e**) The neourethra is covered by a dartos flap, and then glansplasty begins at the coronal margin. (**f**) Glans wings, mucosal collar, and ventral shaft skin are closed

midline tissues extending from the meatus to the tip of the glans where the urethral opening should have developed. Skin flaps from the penile shaft or foreskin are not used as often now as in the past for distal cases, but preputial flaps remain an option for proximal hypospadias (Fig. 30.4). Two-stage repairs today are unusual, reserved in primary cases for proximal hypospadias with severe ventral curvature or for those reoperations in which the previously created neourethra needs to be replaced.

Following surgery, urinary diversion is most commonly maintained with a urethral catheter for approximately 7–14 days, depending upon the severity of the defect. In infants and young boys, this stent can be placed into diapers for drainage without a collection bag. Oral antibiotics are routinely prescribed during catheterization, and in older patients, anticholinergics may also be recommended to reduce bladder irritability.

The expected outcome from modern hypospadias surgery is a penis that looks normal or nearly so (Fig. 30.5). Tubularization procedures create a natural-appearing meatus and glans, and modifications added to flap procedures seek to duplicate this success. Parents can choose either circumcision or foreskin reconstruction in nearly all distal and many proximal cases. While some reports suggest a higher complication rate when preputioplasty is done, we found no difference in expected outcomes with foreskin reconstruction [4]. A recent study [5] used standardized questionnaires to compare parents' impressions of cosmetic outcomes following hypospadias repair using a tubularization procedure versus circumcision in otherwise normal penises, finding no differences.

There are very limited data concerning functional results of modern hypospadias surgeries. Innovation seeking to improve surgical techniques

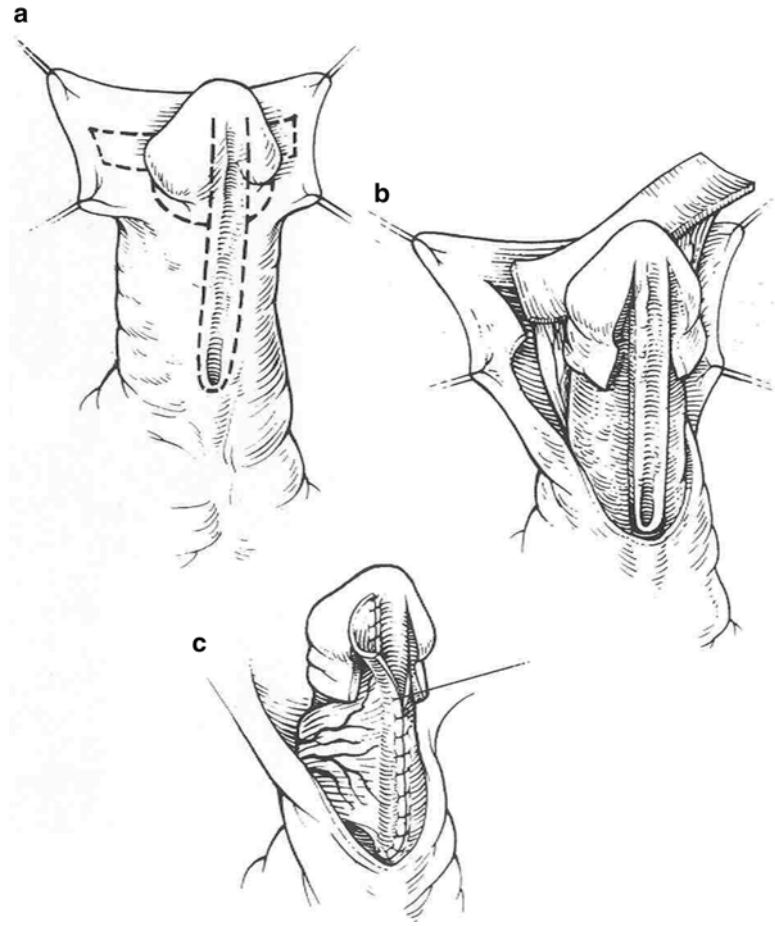

Fig. 30.4 Onlay preputial flap. (**a**, **b**) A rectangular flap of appropriate length and a width of approximately 8–10 mm is cut from the inner prepuce, maintaining its blood supply. (**c**) The flap is rotated ventrally and sutured to the urethral plate. The pedicle of the flap is used to cover the neourethral suture lines

generally has outpaced long-term follow-up, and infants undergoing the initial repairs of some modern techniques have not yet have completed puberty. One report [6] of teens and adults operated as children using skin flaps found most satisfied with their repairs, although minor complaints of spraying stream and post-void dribbling were common. Similarly, those who had experienced ejaculation reported they had to milk semen from the urethra. While there are no reports describing outcomes of straightening procedures after puberty, there are also no reports raising concern that recurrent curvature commonly develops.

A number of complications are possible after hypospadias urethroplasty. The most common is urethrocutaneous fistulas, which are usually small leaks found along the neourethra, often near the coronal of the glans. Meatal stenosis may result in a pinpoint stream and straining to void. Part or the entire repair can dehisce. After skin flap procedures, ballooning of the reconstructed urethra can occur during urination, with post-void dribbling. Urethral strictures may develop, most often at the juncture of a tubularized skin flap to the native urethra. Unless symptomatic obstruction requires intervention sooner,

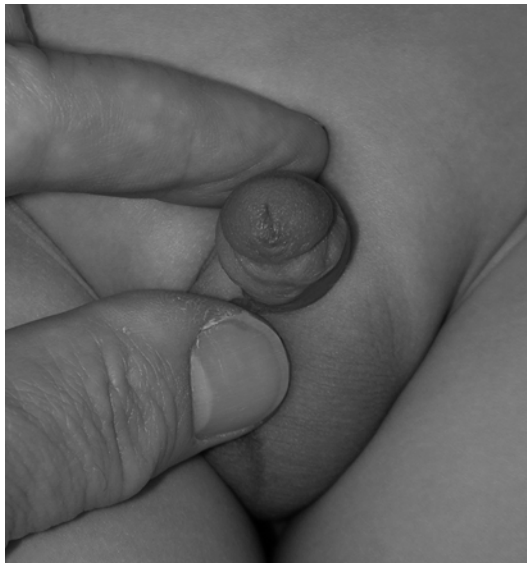

Fig. 30.5 Postoperative appearance following tubularized incised plate repair

it is routine to postpone further intervention to correct these problems for at least 6 months to allow tissues a period of recovery.

Circumcision in Newborns with Hypospadias

A common teaching is that circumcision must be avoided in newborns with hypospadias because the skin will be needed for urethroplasty. However, today, skin flap urethroplasty is much less common that in the past, and the reason to avoid newborn circumcision in most babies with hypospadias or chordee without hypospadias is that the Plastibell rings and Gomco clamps used to perform the procedures do not function properly when the foreskin is asymmetric and deficient ventrally. Accordingly, it remains best practice to defer circumcision until definitive repair of the hypospadias or other penile anomaly.

A subset of patients with hypospadias is born with a normal-appearing penis and complete foreskin. Knowledge that a urethral defect may be concealed beneath the foreskin has raised anxiety among practitioners who perform newborn circumcisions, resulting in aborted procedures when hypospadias is suspected. Consequently, some infants present to pediatric urologists with partial circumcision who have no hypospadias, but now require completion of circumcision under anesthesia. However, the foreskin is not needed to reconstruct these cases, and so infants with normal foreskins should undergo circumcision when desired without concern for the unusual concealed hypospadias [7].

When hypospadias is found unexpectedly after newborn circumcision, parents may suspect urethral injury during the procedure. Consequently, primary care providers should be aware of this condition to allay concerns.

Chordee Without Hypospadias

A dorsal foreskin with a normally positioned meatus is a common anomaly within the spectrum that includes hypospadias. Most often, the urethra is otherwise healthy and any apparent ventral curvature results from the relative deficiency of ventral prepuce and shaft skin. Correction involves either circumcision or foreskin reconstruction performed after 3 months of age as discussed above.

Chordee without hypospadias less often indicates a more significant penile defect with ventral curvature that requires straightening procedures or rarely a hypoplastic urethra that must be reconstructed.

The term “chordee” originally was used to describe fibrous scar-like bands thought to cause ventral curvature in hypospadias, although histologic studies of ventral tissues failed to demonstrate these fibrous bands. Additionally, “chordee” has been used synonymously for “curvature,” as in the diagnosis of chordee without hypospadias. However, the use of the same word to indicate either curvature or tissues thought to cause curvature leads to confusion, especially in families who encounter the term on the Internet, and so with the exception of the ICD-9 code 752.62 terminology of “chordee without hypospadias,” the use of the word should be discouraged.

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