HYPOSPADIAS

A birth defect of the urethra in the male where the urinary opening is not at the usual location on the head of the penis.

Risk factor: Male

About Hypospadias

Hypospadias (Pic. 1) is defined as an anomaly involving the ventral (front) aspect of the penis (Pic. 2). These malformations mainly comprise of an abnormal ventral opening of the urethral meatus, an abnormal ventral curvature (Pic. 3) of the penis (chordee) and/or an abnormal distribution of the foreskin. The extent of the malformation varies. Such ectopic urethral openings (meatus) can be located at the tip of the glans penis (hypospadias sine hypospadias), glanular (Pic. 4), coronal, subcoronal, along the penile shaft, penoscrotal, scrotal, or perineal (Pic. 5). In some cases it is widely gaping and resembling the mouth of a fish. With an incidence of 1:300, hypospadias is one of the most common genital anomalies in male newborns. A stenosis (an abnormal narrowing in a tubular organ) is rather rare.

Hypospadias is classified into three grades, depending on the location of the meatus:

- first degree/mild (glandular or coronal urethral opening)
- second degree/moderate (in the penile shaft)
- third-degree/severe (urethral opening within the scrotum or perineum)

The mild forms, which account for the vast majority of hypospadias, is sometimes the most challenging in terms of the decision making process, because cosmetics is often the only real indication for treatment.

The cause of hypospadias is not known. Hypospadias is thought to result from failure of the urinary channel to completely tubularize to the end of the penis. Most often, it is the only abnormal finding, although in about 10% of cases hypospadias may be part of a syndrome with multiple abnormalities.

This condition does not limit newborn boys. But when they grow up a urinary opening that is not surrounded by glans tissue is more likely to “spray” the urine, which can cause a man to sit to urinate because he cannot reliably stand and hit the toilet. Also in cases when the hypospadias is accompanied by penis curvature, the sexual activity can be affected.

The goal of hypospadias repair is to build confidence in the child by creating a straight penis with a slit-like meatus at the tip of the glans and a urethra of uniform calibre and adequate length, reconstructing a symmetrical glans and penile shaft and achieving projectile stream and normal erection. This all can be only maintain with surgery.

In the management of hypospadias preoperative assessment is of prime importance which should include measurement of the size of the phallus, glans cleft (flat, incomplete, or complete), location and size of the meatus (type of hypospadias and meatal stenosis or mega-meatus), urethral plate width (<1 cm or ≥1 cm), type of chordee, prepuce (complete, incomplete, circumcised), penile torsion (clockwise, anticlockwise), shape of the scrotum (normal, penoscrotal transposition), and associated anomalies (undescended testes, inguinal hernia).

Timing of surgery is decided on the basis of anaesthesia's risk, penile size, and psychosocial development of the infant. The tolerance to anaesthesia is good at the age of 6 months. The difference in the penile length at one year and preschool age is 8 millimetre only. After 18 months the children enter a behavioural phase of development uncooperative for hospitalization. In the background of these facts, using microsurgical instruments and magnification, 6–18 months is the most suitable age for hypospadias repair. If the surgery is not performed during this age then the next window of surgery is preschool age (3-4 years) when the child starts
cooperating with treatment.

**Symptoms**

Hypospadias is usually diagnosed in the newborn nursery by the characteristic appearance of the penis. The urinary opening (“meatus”) is lower than normal, and most boys have only partial development of the foreskin, lacking the normal covering for the glans on the underside. The abnormal “hooded” foreskin calls attention to the condition. However, not all newborns with partial foreskin development have hypospadias, as some have a normal urinary opening with a hooded foreskin, which is called “chordee without hypospadias”.

**Associated diseases**

The most common associated defect is an undescended testicle, which has been reported in approximately 3% of infants with distal hypospadias and 10% of those having proximal hypospadias.

Hypospadias can be associated with other urogenital tract anomalies such as pelviureteric junction obstruction (a condition where blockage occurs at the junction where the ureter attaches to the kidney), vesicoureteric reflux (urine flows retrograde, or backward, from the bladder into the ureters/kidneys) and renal agenesis (the failure of an organ to develop during embryonic growth and development due to the absence of primordial tissue) which should be excluded by ultrasonographic scan in every hypospadias patient.

Proximal hypospadias with cryptorchidism (undescended testes), enlarged utricle, or penile size < 2.5 cm should be investigated for intersex disorders by ultrasonography, hormonal profile, and karyotyping.

**Complications**

Different operative interventions reflects the frustration of surgeons facing the high rate of complications that result from hypospadias surgery.

Problems that can arise include a small hole in the urinary channel below the meatus, called a fistula (Pic. 6). The head of the penis, which is open at birth in boys with hypospadias and is closed around the urinary channel at surgery, sometimes reopens. The new urinary opening can scar, resulting in meatal stenosis, or internal scarring can create a stricture, either of which cause partial blockage to urinating.

Most complications are discovered within six months after surgery, although they occasionally are not found for many years. In general, when there are no apparent problems after repair in childhood, new complications arising after puberty are uncommon. However, some problems that were not adequately repaired in childhood may become more pronounced when the penis grows at puberty, such as residual penile curvature or urine spraying due to glans dehiscence (the spontaneous opening).

These complications are usually successfully corrected with another operation, most often delayed for at least six months after the last surgery to allow the tissues to heal sufficiently before attempting another repair. Using modern surgical techniques, a normal appearing penis can usually be expected from hypospadias repair. Results when circumcision or foreskin reconstruction are done are the same, and so care-givers can choose whichever option they wish.

Another approach to the problem of frequent complications with hypospadias surgery is the use of antibiotic prophylaxis and postoperative treatment. Therefore, surgeons often administer prophylactic antibiotics, which entail administering one dose of antibiotics preoperatively. This is done in order to reduce the risk of a possible urinary tract infection (UTI) or wound infection related to the surgery or the malformation. Due to increasing prevalence of antibiotic resistance it is important to investigate whether the incidence of UTIs is higher in hypospadias patients and what actions should be taken regarding the antibiotic use associated with the operation.

**Risk factors**

It has been long recognized that age of the mother is one of the factors that defines a high-risk pregnancy in terms of reaching a healthy full-term pregnancy but also in terms of the potentiated risk for having a child with a congenital condition. In fact, age of the mother (> 40 years of age) is associated to hypospadias risk.

**Prevention**
Hypospadias cannot be prevented but it is possible to prevent the postoperative complications in several ways.

Boys with proximal hypospadias typically have small glans penis, making hypospadias repair a technically demanding operation. To improve outcomes, use of pre-operative hormonal stimulation (PHS) prior to surgery has been accepted as a relatively common practice among pediatric urologists and surgeons for decades despite the lack of high quality evidence to support it.

In severe and complicated cases there are several things, that can increased the success and prevent the complications. Histological studies show that androgens increase the number and density of blood vessels, supporting a theory of neovascularization (the formation of functional microvascular networks with red blood cell perfusion). Observational studies have shown that testosterone, dihydrotestosterone, and human chorionic gonadotropin have been associated with a temporary increase in penile length, glans circumference, and tissue vascularity. These changes have been hypothesized to improve outcomes of hypospadias repair.

How it can affect fertility

In cases of penile curvatures, sexual intercourse may be difficult. Penile angulation (curvature) not only causes potential sexual dysfunction, difficulty and pain during intercourse, or total coital incapacity, but also causes severe psychological problems.

Hypospadias places the urinary opening on the underside of the penis and this can prevent sperms from reaching the cervix. Surgical correction of hypospadias should solve the fertility problems, the curve of penis is straightened and ejaculation can flow properly forward.

Hypospadias is very often associated with cryptorchidism, which leads to impairment of sperm production in undescended testicles. Some studies also shows increase of antisperm antibodies in children born with hypospadias.

Prognosis

The severity of hypospadias is graded upon the position of the urinary meatus and the extent of ventral penile angulation. Severity rises as the distance of the displaced urethral opening increases from the normal position at the tip of the glans and with increasing penile curvature. Cryptorchidism and inguinal hernias are the most common extragenital anomalies, which are found in 7–10% of all hypospadias cases.

By 3D representation, the external genitalia are shown with a great clarity of structures and their anatomical relation which confirm the diagnosis of third-degree hypospadias. Therefore, the multiplanar representation of the genital area allows us to obtain a better understanding of hypospadias and to get proper parental counseling from pediatric surgeons, with the possibility to report the prognosis and future treatment. 3D ultrasounds are a very interesting tool for diagnosis and clinical evaluation in prenatal diagnosis of hypospadias.

After the confirmation and performance of successful surgery, men will not have any future consequences and can conceive child naturally.
Example of penis with hypospadias.

Uncircumcised human penis.

Associated curvature (40°) is confirmed by artificial erection.

Glanular hypospadias of penis.
Sources

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