GENITOURINARY SYSTEM

HYPOSPADIAS

Hypospadias is the most common congenital anomaly of the penis.it occur in approximately 1 to 3 males per 1000 live births.

Definition: refers to a condition in which the urethral opening is located behind the glans penis or anywhere along the ventral surface of the penile shaft.

HYPOSPADIAS

PATHOPHYSIOLOGY

1. HYPOSPADIAS is a mild to severe malformation in which the urethra may open either near to where the normal meatus should be located or on the ventral surface of the penile shaft in one of several position: just behind the glans on the body of the penis.
2. an associated congenital chordee caused by a fibrous band of tissue that extends from the scrotum up the penis and pulls it vertically in an arc, may be present.chordee,however may occur as an isolated phenomenon. The ventral foreskin is frequently absent.

Clinical manifestation

Hypospadias can usually be observed at birth, nurse observe the voiding pattern of infant. Infant does not void normally fact is reported.

Emotional distress

The inability of the boy to stand to urinate: he must sit to void.

Intercourse difficult.

NURSING MANAGEMENT;

- IF the anomaly should be repaired after infancy but before the child is of school age.
- Surgery is generally performed at approximately 3 years of age, because penis has increased sufficiently in size and Child does not yet have anxiety caused by the fear of mutilation.

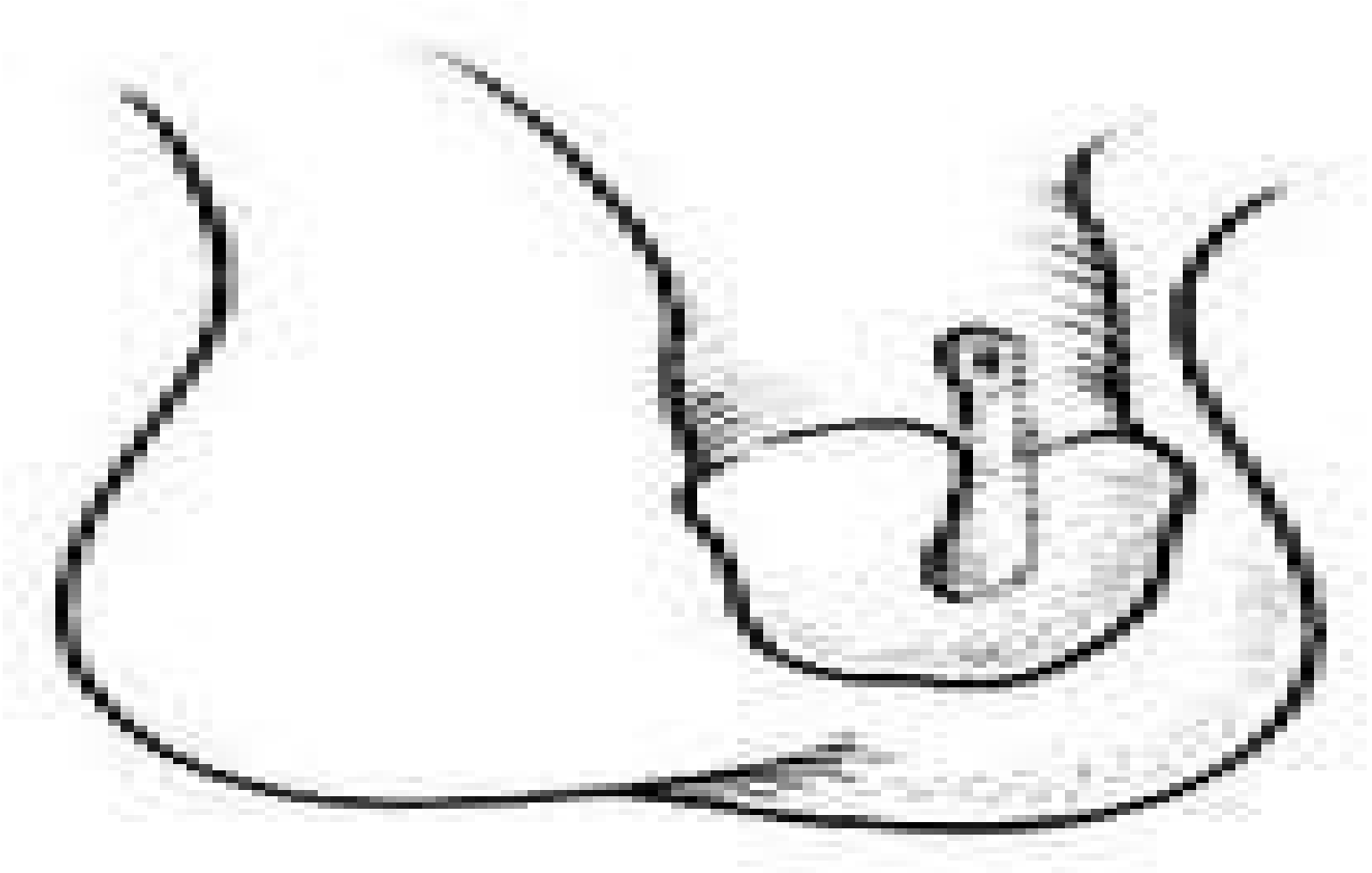
- The goals of surgical management for hypospadias with chordee are to straighten the penis,to move the meatus distally and to improve the penis cosmetically and functionally.
- Preoperative preparation of the parents and child for the type of surgery to be done

- Parents are disappointed if they expect the appearance of penis to be perfectly normal after surgery.
- Nurse must help parent and child releave there anxiety.
- Provide psychological support.
- Urethroplasty is done, a form of urinary drainage, such as a catheter or suprapubic tube is used to keep the area dry, to promote healing .
- Surgical area kept dry, free from infection.

EPISPADIAS

- **Definition; the urethra opens on the dorsal surface of the penis.in a mild defect, the urethra may lie just behind the glans.**
- Epispadias is a rare congenital (present from birth) defect located at the opening of the urethra.
- In this condition, the urethra does not develop into a full tube and the urine exits the body from an abnormal location.

EPISPADIAS



- **ETIOLOGY:**
- The causes of epispadias are unknown at this time. It may be related to improper development of the pubic bone.
- In boys with epispadias, the urethra generally opens on the top or side of the penis rather than the tip. However, it is possible for the urethra to be open along the entire length of the penis.

CLINICAL MANIFESTATION

- In males:
- Abnormal opening from the joint between the pubic bones to the area above the tip of the penis
- Backward flow of urine into the kidney (reflux nephropathy)
- Short, widened penis with an abnormal curvature
- Urinary tract infections
- Widened pubic bone

Conti…

- In females:
- Abnormal clitoris and labia
- Abnormal opening where the from the bladder neck to the area above the normal urethral opening
- Backward flow of urine into the kidney (reflux nephropathy)
- Widened pubic bone
- Urinary incontinence
- Urinary tract infections

INVESTIGATION:

- Blood test to check electrolyte levels
- Intravenous pyelogram (IVP), a special x-ray of the kidneys, bladder, and ureters
- MRI and CT scans, depending on the condition
- Pelvic x-ray
- Ultrasound of the urogenital system

TREATMENT

- Surgical repair of epispadias is recommended in patients with more than a mild case. Leakage of urine (incontinence) is not uncommon and may require a second operation.
- Surgery generally leads to the ability to control the flow of urine and a good cosmetic outcome.

- The primary goals of treatment of epispadias are to: lengthen and straighten the penis by correcting dorsal bend and chordee; and create functionality and cosmetically acceptable external genitalia with as few surgical procedures as possible.
- If the bladder and bladder neck are also involved, surgical treatment is required to establish urinary continence and preserve fertility.

- There are two popular surgical techniques that achieve these objectives. The first is the modified Cantwell technique, which involves partial disassembly of the penis and placement of the urethra in a more normal position.
- The second technique and most recent evolution of the modern epispadias repair is the complete disassembly of the penis into its separate components - two corpora cavernous and a single corpus spongiosum. Following disassembly, the three components are reassembled such that the urethra is in the most functional and normal position and dorsal chordee is corrected.
- Both techniques provide a straight urethra positioned on the underside of the penis, correction of chordee and an acceptable cosmetic result.

Ambiguous genitalia

- **Definition**: Ambiguous genitalia is a birth defect where the outer genitals do not have the typical appearance of either a boy or a girl.

Ambiguous genitalia

Increased androgen production results in ambiguous genitalia in newborn girls.

- **ETIOLOGY:**
- Pseudohermaphroditism. The genitalia are of one sex, but some physical characteristics of the other sex are present.
- True hermaphrodism. This is a very rare condition, in which tissue from both the ovaries and testicles is present. The child may have parts of both male and female genitals.

- Mixed gonadal dysgenesis (MGD). This is an intersex condition, in which there are some male structures (gonad, testis), as well as a uterus, vagina, and fallopian tubes.
- Congenital adrenal hyperplasia. This condition has several forms, but the most common form causes the genetic female to appear male. Many states test for this potentially life-threatening condition during newborn screening exams.

- Chromosomal abnormalities, including Klinefelter's syndrome (XXY) and Turner's syndrome (XO).

- If the mother takes certain medications (such as androgenic steroids), they may make a genetic female look more male.

- Lack of production of certain hormones can cause the embryo to develop with a female body type, regardless of genetic sex.
- Lack of testosterone cellular receptors. Even if the body makes the hormones needed to develop into a physical male, the body cannot respond to those hormones.

ETIOLOGY

- **Androgen insensitivity syndrome (AIS)** - a genetic condition characterised by the fetal tissue's insensitivity to male hormones. This affects genital development. For example, a newborn may have some of the female reproductive organs but also have testicles.

Sex chromosome disorders - instead of having either XX or XY sex chromosomes, a baby may have a mixture of both ('mosaic' chromosomes); or specific genes on the Y chromosome may be inactive; or one of the X chromosomes may have a tiny Y segment attached to it.

Maternal factors - the pregnant mother may have had an androgen-secreting tumour while pregnant, and the excess of this male hormone affected her baby's genital development.

INVESTIGATION

- Physical examination
- Hormone tests using blood, urine or both
- Genetic tests using blood, urine or both
- Ultrasound scan

Treatment

- **Parental counseling** - successful sex assignment and identity for the child depends largely on the attitude of the parents. It is important that both the mother and father are fully informed about their child's condition. Support groups may provide help in this area..
- **Surgery** - for example, an overly large clitoris may be trimmed, or a fused vulva separated, or undescended testicles relocated into the scrotum. However, surgical gender assignment depends heavily on what genital structures the surgeons have to work with

- The majority of babies with ambiguous genitalia have been brought up as girls. A few operations may be needed, usually begun in the child's first year. Further surgery might be required during adolescence. Some intersex support groups feel that surgery is not always the answer, particularly when the gender of the child is not clear. Others suggest that surgery should wait until the child is old enough to decide for themselves. However, most medical professionals advocate early surgical and hormonal intervention for the sake of clearly establishing the child’s gender and sense of belonging in society.

- **Counseling for the child** - the child needs to be informed and talked to about their diagnosis in a very careful way.
- **Hormone therapy** - during their teenage years, the child may need hormone supplementation therapy to help bring on puberty. A child with CAH will need to have daily hormone therapy.

Medication:

Hormone medications may help correct the hormonal imbalance. In some children, hormones may be administered shortly after birth and may be thFor example, in a genetic female with a slightly enlarged clitoris caused by a minor to moderate case of congenital adrenal hyperplasia, proper levels of hormones may shrink the tissue close to a normal sizee only treatment necessary.

Surgery

- In children with ambiguous genitalia, surgery may be used to:
- Preserve normal sexual function
- Create more natural-looking genitals
- The timing of surgery for ambiguous genitalia will depend on your child's specific situation. Many doctors prefer to postpone surgery done solely for cosmetic reasons until the person with ambiguous genitalia is mature enough to participate in the decision about gender assignment.

- For girls with ambiguous genitalia, the sex organs often work normally despite the ambiguous outward appearance. If a girl's vagina is hidden under her skin, surgery in childhood can help with sexual function later. For boys, surgery to reconstruct an incomplete penis may improve appearance and make erections possible