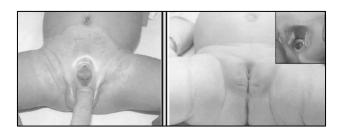


Indian Association of Pediatric Surgeons Patient Information Sheet

EPISPADIAS



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What is epispadias?

It is a developmental anamoly. Normally the urethra opens at the tip of the penis. In epispadias, which is a congenital absence of upper wall of urethra, the urethral meatus opens on the dorsum of the penis in boys. There are 3 main types depending on the position of the urethral meatus: penopubic (near the pubic bone), penile (on the shaft of the penis), and glandular (on the head of the penis). In girls the pubic bones are not joined to a variable extent and the clitoris is seen in two halves.

Associated abnormalities include diastasis of the symphysis pubis (pubic bone), renal agenesis, and ectopic pelvic kidneys. It is almost always associated with exstrophy of the urinary bladder in both boys and girls.

What causes this problem and how common is it?

The exact cause is not known. It is not a genetic defect and could not have been preventable by taking extra care or avoiding something. It is not possible to pickup on antenatal ultrasound scan, (unless assosciated with exstrophy bladder). It is not a condition passed on from father's or mother's side. It occurs in 1 out of 1,17,000 male births. It is very rare in females.

What are the symptoms?

The symptoms depend on the severity of epispadias.

- Symptoms common to all varieties include cosmetic, passing urine on abdomen, future sexual dysfunction.
- Distal cosmetic, passing urine on abdomen, future sexual dysfunction
- Proximal epispadias is associated inability to control urinary leak, cosmetic, passing urine on abdomen, future sexual dysfunction
- In assosciation with exstrophy bladder symptoms are dictated by exstrophy bladder.
- In boys the penis is short, flat and curved upwards (dorsal chordee)
- In girls the clitoris is split and the urethra opens in between

the split clitoris.

 Associated vesico-ureteral reflux may cause recurrent urinary tract infection.

When to see your doctor?

The attending pediatrician will have seen it at birth and a family counselling with a pediatric surgeon should be scheduled when all caretakers can be attending.

How is it diagnosed?

Its a visual diagnosis.

Diagnostic evaluation is required if there is associated urinary incontinence. In this situation the anatomical and functional aspects of the upper and lower urinary tracts need to be evaluated using blood tests and imaging studies.

Urodynamic studies are also required in the assessment of the lower urinary tract.

What are the treatments available ?

Surgical reconstruction is the only and proper way of treatment.

Are there any alternatives to surgery ?

No.

What does the operation involve?

The operation involves correction of the various subsegments of this condition :

- Bring the urinary opening to the tip of the penis
- · Bring the urinary tube on the under surface of the penis
- Correct the upward bend of penis (correct dorsal chordee)
- Try and lengthen the shaft of penis
- In proximal epispadias bladder neck may need to be repaired
- To achieve this, surgery will involve general and regional anesthesia
- In girls the urethra is short and the repair is less technically challenging than in boys. The split clitoris needs to be brought together at the same time.

 When associated with bladder exstrophy the epispadias is repaired along with the exstrophy. The exstrophy repair is more complex.

What are the possible complications / what happens after the operation?

- Child will have a catheter for 7 10 days
- · Child will be prescribed antibiotics and pain killers
- Constipation should be avoided
- If the bladder neck has also been repaired then the tubes may need to remain in position for up to 3 weeks.
- There is a small risk of re-surgery if needed.

What is the outlook or future of these children?

These children should have a normal urinary and sexual function. Cosmetic appearance can be excellent if expectations are pragmatic. In conditions with severe deformity, where incontinence is also present, partial continence or assisted voiding may be needed to stay dry.

Associated vesico-ureteral reflux may need long term management with both medications and/or surgical intervention.

Some boys may have retrograde ejaculation leading to infertility.



Dorsal chordee – the penis is bent upwards



Urethral orifice – peno-pubic location