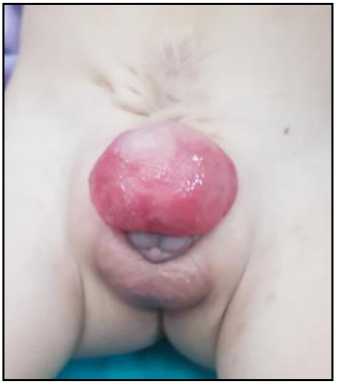
Indian Association of Pediatric Surgeons

Patient Information Sheet

BLADDER EXSTROPHY



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What is bladder exstrophy ?

Bladder exstrophy is a congenital abnormality that occurs when the skin over the lower abdominal wall does not form properly. The skin, subcutaneous tissue under the skin, muscles of the lower abdominal wall, including the pelvis bones have not fused in the midline. In addition, the urinary bladder is also open and lies exposed. In almost all cases the urethra is open up to the tip of the penis (complete epispadias). This condition is also referred to as the 'exstrophy-epispaias' complex.

What causes this problem and how common is it ?

It is a congenital anomaly and is quite uncommon. The reported incidence is variable; ranges from 1 in 10,000 to 50,000 live births. The exact cause for this anomaly is not known. In very rare instances the large intestine also opens into the open bladder; it is then referred to as the 'cloacal exstrophy'.

What are the symptoms ?

The bladder is open and exposed on the outside of the abdomen. In epispadias, the urethra also remains open. Urine is constantly coming out of the ureteric orifices in the open bladder and the child is constantly wet. The bladder mucosa is prone to injury and can bleed. In older children the bladder mucosa also undergoes 'metaplastic' changes due to constant exposure. Because the pelvic bones have also not fused the child develops a waddling gait. The ureters and kidneys also get dilated and urinary tract infection may occur.

When to see your doctor ?

When parents notice this at time of birth.

How it is diagnosed ?

It is diagnosed by clinical examination only. However, to study the effects of the open bladder blood tests to study kidney function, urine examination to rule out urinary tract infection and some x-rays maybe required.

What are the treatments available ?

Bladder exstrophy and epispadias are corrected by multiple operations over the first few years of life. The overall aim of treatment is to prevent any kidney damage and correct the abnormalities so that the child's urinary system and genitals work properly and look as normal as possible.

When it should be operated ?

The surgery depends upon clinical situation and associated anomalies of the child and treating team decides about optimal time of surgery. If neonate is able to tolerate, bladder closure is done in first stage.

Are there other alternative methods of treatment ?

Surgery is the only treatment option. The choice of procedure may depend on a number of factors including age, size of the bladder, condition of the bladder.

What does the operation involve ?

There are many procedures to repair bladder exstrophy. Some surgeons advocate a single stage operation; this is very major, requires a lot of tissue mobilisation and may take up to 12 hours. Others may prefer to do it in several stages as tolerated by the patients. Your doctor will discuss these in detail with you.

The parts of the reconstruction will include :

- Closure of the bladder
- Reconstruction of the anterior abdominal wall with or without closure of the pelvic bones
- Bladder neck reconstruction to provide a continence mechanism for urination
- Repair of the epispadias
- Reimplantation of ureters if vesico-ureteral reflux is a major problem (this is usually done later).

These procedures can be combined in various permutations and combinations.

Such surgeries should be done in well equipped hospitals with tertiary care services.

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

Immediately after surgery there will be about 4 or 5 catheters sticking out of the body. If the pelvic bones have been closed then there maybe restrictive dressings for several weeks. All patients require antibiotics and pain killers. Oral feeding is resumed within a week. Blood transfusions maybe required if blood loss is excessive.

Complications include mainly those related to the surgery eg tissue necrosis, wound infection, wound dehiscence and urinary tract infection. Some of these complications may require secondary operations.

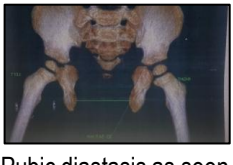
What is the outlook or future of these children ?

They do need a long follow up; in some cases life long support and treatments. Many urological problems like a small or non-compliant bladder may damage the kidneys and also cause incontinence. Such patients may require further treatment to increase the capacity of the bladder (augmentation cystoplasty) using parts of the stomach or intestines. These procedures have their own share of metabolic problems.

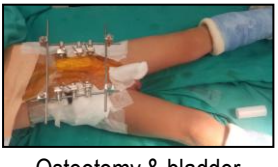
Often, despite best measures, continence may remain a problem. There are operative procedures designed to tackle this in the form of closure of the bladder neck and a stoma to perform clean intermittent catheterisation (CIC).

Sexual problems eg inadequate sexual performance due to dorsal chordee may affect the psychological well being. The sperm instead of coming out may go inside the bladder and lead to infertility.

In girls the outcomes are usually better.



Pubic diastasis as seen in exstrophy



Osteotomy & bladder closure in exstrophy