

Bladder Exstrophy

BIRTH DEFECT RESEARCH FOR CHILDREN



What is Bladder Exstrophy?

Exstrophy means to “turn inside out” and that is what happens when a child is born with bladder exstrophy. In this condition, the bladder and related structures are turned inside out. Typically the skin of the lower abdominal wall that normally covers the bladder does not form properly exposing the inside of the bladder to the external world.



Bladder Exstrophy



What other conditions may accompany Bladder Exstrophy?

Babies born with Bladder Exstrophy may have all or some of these other associated abnormalities:

Epispadias:

In boys, the urethra (tube that carries urine from the bladder and out of the body) may be short and split so that it opens on the upper surface rather than at the end of the penis. In girls, the opening occurs between a split clitoris and labia minora.

Dorsal Chordee:

In this condition, the penis is pulled against the abdomen in a slight downward curve.

Bladder malformations:

The bladder neck and sphincter (the ring of muscles that permit the bladder to open and close) may be missing, making it impossible for the child to achieve bladder control without surgery. In addition, the bladder may be unable to hold a normal amount of urine, and the ureters (the tubes that carry urine from the kidneys to the bladder) may be abnormally positioned.

Separation of the pubic bones:

In bladder exstrophy, the large bones of the pelvis that normally support the bladder and lower organs fail to join, leaving a wide opening. This causes the hips to rotate outward and the feet to point away from one another when the child walks or runs. This does not, however, prevent a child with bladder exstrophy from being able to move about normally.

Abnormal positioning of the anus:

The opening of the rectum is positioned closer to the scrotum or vagina. This does not usually cause problems with bowel function.

A lower belly button:

The belly button often cannot be seen and is typically removed when the bladder is surgically closed.

What causes Bladder Exstrophy?

Doctors do not know what causes Bladder Exstrophy, but they do know that it occurs at about the 4th or 5th week after conception.

How often does Bladder Exstrophy occur?

It occurs in 1:30,000 births in the general population, but some families have a greater likelihood of reoccurrence. The condition also occurs 2-3 times more often in males than females.

What treatments are available for Bladder Exstrophy?

Treatments will vary according to the individual situations; however, Bladder Exstrophy must be surgically repaired. Current methods involve a series of surgeries over a number of years, a process known as staged reconstruction. Surgeons will suggest strategies best suited to individual cases.

Within 24-48 hours of birth, surgery involving primary closure of the bladder is usually performed. Also at this time, separated pelvic bones are brought into place and the bladder is closed and returned to the pelvis.

Genital repair occurs at different times for boys and girls. In girls, the genitalia and the urethra may be repaired at the same time as the initial surgery on the bladder. In boys, genital repair is usually accomplished between one and two years of age.



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What is the prognosis for children with Bladder Exstrophy?

Individual surgical success varies, but recent studies show that even though many children require self-catheterization to empty their bladders, their quality of life was only slightly limited. Most children with bladder exstrophy had good self-esteem, but some children and their parents benefited from intervention from a multi-disciplinary team during different stages of childhood.

Fact Sheet by:

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