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Bladder Exstrophy

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What is Bladder Exstrophy?

An abnormality of formation of the bladder and the bony pelvis. The bladder does not form into its normal round shape but instead is flattened and exposed on the abdominal wall. The pelvic bones are also widely separated. The remainder of the lower urinary tract may also be flattened and exposed, with abnormal formation of the prostate and penis. This congenital birth defect is seen in one of 10,000 to 50,000 live births. In a family with a child with exstrophy, the likelihood of a second child being born with exstrophy is one in 100. The risk of having a child with exstrophy is one in 70, if the parents have exstrophy. Major genetic studies are currently underway at Johns Hopkins involving the exstrophy-epispadias complex.

Diagnosis

Diagnosis can be made on careful repeated ultrasounds done before delivery, but usually the diagnosis is not made until the baby is born. The finding of the exposed bladder is typical.

Treatment

Advances in surgery in the last 15 years have allowed reconstruction of the penis and bladder so that a more "normal" and functional lifestyle can be maintained by the patient and family. Current state-of-the-art treatment for exstrophy involves reconstruction of the various aspects of the deformity (i.e. closing the bladder, repair of the penis, and prevention of urine leakage. This usually involves separate operations at various times in the life of the child to obtain the best results.

- **Initial Closure:** This is usually done soon after the baby is born. At this first operation, the pelvic bones are reformed into their normal ring shape (by a Pediatric Orthopedic surgeon), the bladder, abdominal wall, and posterior urethra are closed and the belly button is reconstructed. This procedure takes between 4-6 hours. Following surgery, the baby is placed with the lower legs in traction to prevent separating the pelvic bones. Babies are observed carefully and may be in the Intensive Care Unit to be monitored for the first day or two. The baby usually stays in the hospital for 3-4 weeks of healing. Antibiotics are given following the operation to prevent infection. The tube in the bladder is removed four weeks following surgery. The size of the bladder gradually increases over time. In very special circumstances with an excellent bladder plate and good sized penis, bladder exstrophy closure and

epispadias repair can be combined. However, this is only for very experienced exstrophy surgeons.

- **Epispadias repair:** This repair occurs around 6-12 months of age. Time of surgery and the extent of the surgery is dependent on the size of the bladder and the deformity of the penis. At this stage, the urethra on top of the flattened penis is closed and transferred to below the corporal bodies as in the normal penis.
- **Contenance Procedure:** At this time the control of urine leakage is repaired and then further enlargement of the bladder is preformed if necessary. The timing of this procedure depends totally on the capacity of the bladder and the child's emotional and developmental status. The child must "want to be dry" and able to participate in a voiding program.

Special Voiding Improvement Program for the Exstrophy - Epispadias Patient

Children with bladder exstrophy face a combination of medical and emotional challenges as they work with their urology team. Developing a continence management program is an ongoing process that is often stressful for children and their families. The voiding improvement program provides hands-on one-on-one assistance to the child and family before and after bladder neck repair.

The clinic is staffed by a specially trained, multidisciplinary team including a pediatric urologist, pediatric nurse practitioner, pediatric behavioral psychologist, and pediatric clinical nurse.

The Voiding Improvement Team also assists the pediatric urologist in the evaluation of readiness for bladder neck reconstruction to help the child and family prepare for the post-surgical work that will allow for favorable continence outcome.

After bladder neck reconstruction surgery we work with children and families using both behavior modification and muscle retraining procedures to teach the child, family, and the child's bladder musculature to function at their maximum potential for long-term continence.

After bladder neck surgery, frequent daily phone consultations occur until the child is voiding well and tube free.

Visit usually last one hour and may include the following treatment components:

- bladder ultrasound
- urine culture
- medication management
- biofeedback for bladder muscle retraining
- establishment of a voiding urine
- assessment of barrier to adherence
- nutrition education
- behavior therapy
- relapse prevention recommendations

Progress is evaluated at each visit and communicated to the pediatric urologist.

Follow-up

This, however, requires dedicated and intensive treatment and long-term follow up into adolescence and adulthood by the exstrophy team.

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