UCSF PEDIATRIC UROLOGY

Laurence S. Baskin, MD
Hillary Copp, MD, MS
Michael DiSandro, MD
Anne Arnhym, CPNP
Angelique Champeau, CPNP

Epispadias and Exstrophy

What is bladder exstrophy/epispadias?

Bladder Exstrophy is a very rare abnormality that is present at birth (congenital). It occurs when the bladder does not fold and close and therefore the abdominal wall and the pelvic bones cannot close over the bladder. The baby is born with the bladder and urethra open on the abdominal wall. In its most extreme form there can be a defect in the urethra, bladder and bowel (cloacal exstrophy). Bladder exstrophy, also known as classic bladder exstrophy, is when the defect is limited to the bladder and the urethra. Epispadias includes only the urethra.

Bladder exstrophy is seen 1 in 30,000 live births. It is slightly more common in males than females and the risk of a family having more than one child with bladder exstrophy is 1 in 100. Children born to a parent who had bladder exstrophy have a risk of 1 in 70. Epispadias is less common, occurring in about 1 in 100,000 live births. Cloacal exstrophy occurs even less frequently, about 1 in 250,000 live births.

How is bladder exstrophy/epispadias diagnosed?

It is thought to occur around the 11th week of gestation. Frequently it is detected prior to birth by a prenatal ultrasound. Otherwise it is detected at the time of delivery.

How is bladder exstrophy/epispadias treated?

Bladder exstrophy is managed surgically. One technique is to close it in stages. First the bladder and bladder neck are closed, the bones of the pelvis are brought together, and the urethra is made. Secondary surgeries are required in the first year of life to reconstruct the penis and to manage inguinal hernias. In some instances the entire reconstruction including bladder, bladder neck, pelvic bones, and urethra are closed all in one surgery. Both these techniques require an experienced surgeon.

550 16TH STREET 5TH FLOOR, MAILCODE 1695, SAN FRANCISCO, CA 94143
415.353.2200 (Phone) • 415.353.2480 (Fax)

Both techniques will require the child to go to the intensive care unit (ICU) immediately after surgery for a few days. The child is then followed for a few more days in the regular (non ICU) part of the pediatric hospital before the child is ready for discharge. Hernia repairs will need to be performed later and often the child has vesicoureteral reflux, which may need to be corrected. Depending on the initial size of the bladder continence can be achieved up to 70% of the time. However, often these children require further surgeries to manage the bladder neck and treat incontinence. Often these children need to catheterize.

Epispadias occurs in males and females. It occurs in bladder exstrophy and without bladder exstrophy. In males it occurs when the penis does not properly form a tube out to the tip of the penis and the opening is along the shaft of the penis. It can occur from the tip of the penis all the way to the bladder and depending on the position of the opening it can be associated with urinary incontinence (urine leakage) and retrograde ejaculation (ejaculate going backward toward the bladder instead out the urethra). In girls it occurs when the urethra does not form a tube properly and is almost always associated with urinary incontinence that will require surgery to repair.

What happens after treatment of bladder exstrophy/epispadias?

Ongoing medical management is important for children born with bladder exstrophy/epispadias. Children with bladder exstrophy/epispadias can have normal lives with normal life expectancy.

Important Links:

Association for the Bladder Exstrophy Community

International network. Founded 1991. Mutual support for persons affected by bladder exstrophy including: parents of children with bladder exstrophy, adults, healthcare professionals and others interested in bladder exstrophy. Newsletter, literature, information and referrals, conferences, advocacy and directory of members.

Write:
Association for the Bladder Exstrophy Community
2901 W. KK River Parkway, Ste. 311
Milwaukee, WI 53215
Voice: 1-866-300-2222 or 414-385-7100
Website: http://www.bladderexstrophy.com
E-mail: admin@bladderexstrophy.com
Verified: 8/24/2012

See the next page for contact information.

Contact Information:

Laurence S. Baskin, MD
http://urology.ucsf.edu/people/laurence-s-baskin#

Hillary Copp, MD, MS
http://urology.ucsf.edu/people/hillary-l-copp

Michael DiSandro, MD
http://urology.ucsf.edu/people/michael-j-disandro

Appointments & Location

Mission Bay Benioff Children's Hospital (Surgical Admissions)
1975 4th Street
San Francisco, CA 94143
415.353.2200 (Phone)
415.353.2480 (Fax)

Children's Hospital & Research Center Oakland
747 52nd Street Ambulatory Care 4th
Oakland, CA 94609
510.428.3402 (Phone)

PEDIATRIC NURSE PRACTITIONERS

Anne Arnhym, CPNP
Certified Pediatric Nurse Practitioner
Anne.Arnhym@ucsf.edu

Angelique Champeau, CPNP
Certified Pediatric Nurse Practitioner
Angelique.Champeau@ucsf.edu