BOSTON CHILDREN'S HOSPITAL'S

Bladder Exstrophy Handbook

Boston Children's Hospital
Until every child is well™

Bladder Exstrophy Program

Table of Contents

ABBREVIATION GUIDE

ART	assisted reproductive techniques	MSRE	modern staged repair of exstrophy
BE	bladder exstrophy	RNC	radionuclide cystogram
BNR	bladder neck reconstruction	RUS	renal ultrasound
CBC	complete blood count	UDS	urodynamic study
CIC	clean intermittent catheterization	UPJ	ureteropelvic junction
CMG	cystometrogram	UTI	urinary tract infection
CPRE	complete primary repair of exstrophy	UVJ	ureterovesical junction
EEC	exstrophy-epispadias complex	VCUG	voiding cystourethrogram
KUB	kidney ureters bladder (abdominal x-ray)	VUR	vesicoureteral reflux

Boston Children's Hospital | Bladder Exstrophy Program

Until every child is well™

Dear families,

The Bladder Exstrophy Handbook has been created by the staff of the Bladder Exstrophy Program of the Department of Urology at Boston Children's Hospital to accompany you along your journey of care with us. The Bladder Exstrophy Program is composed of a dedicated interdisciplinary team of physicians, nurses, nurse practitioners, social workers, child life specialists, public health researchers and administrative staff. As an established and dedicated program within the Department of Urology, our interdisciplinary team strives to provide exceptional care for infants, children, adolescents and young adults with bladder exstrophy or other diagnoses within the exstrophy-epispadias complex (EEC).

This handbook provides easy access to useful information along the continuum of normal development, and potential medical and surgical care for your child with exstrophy. It is meant to assist, but not replace, the critically important partnership between our team and you and your child. Feel free to read it from cover to cover, or just review the sections that are important to you and to your child's care.

Our entire team welcomes your partnership as we strive to optimize the care and the experience for you and your child. Please know that our door is always open for your valuable suggestions and feedback.

— The Bladder Exstrophy Program
617-355-7796

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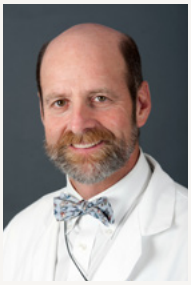

David A. Diamond, MD
Urologist-in-Chief
Associate Clinical Ethicist
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Our team

Meet the bladder exstrophy experts

A team of health care professionals that includes physicians, nurses, social workers and child life specialists will care for your child. Research coordinators and assistants are also important members of our team.

Physicians

The Department of Urology at Boston Children's Hospital is consistently ranked as one of the best pediatric urology departments in the country. Our physicians have decades of experience performing surgeries and managing the care of patients with bladder exstrophy. In addition to our Urology team, care of patients with bladder exstrophy is supported by experts in Anesthesiology, Radiology, Advanced Fetal Care and Orthopedics.

Nursing

In the Department of Urology, our nursing staff includes nurse practitioners (NPs) and registered nurses (RNs). The nursing staff works alongside our physicians to help manage the care of our patients with bladder exstrophy. Nurses provide patients and families with education and guidance to prepare for all aspects of care including surgery and homecare.

Our bladder exstrophy nurses have extensive experience with managing the care of patients of all ages and are available to answer questions regarding your child's condition. Our nurses work with the team to plan the biannual Bladder Exstrophy Support Group and annual Teen Picnic.

Social Work

We have a dedicated social worker for the Bladder Exstrophy Program. Our social worker helps families navigate any psychosocial issues surrounding major pediatric surgical reconstruction and living with a chronic medical condition. Social workers are available to help families find housing during surgery, work with the child's educational institution, provide emotional support to the family, and locate both hospital and community resources for the family. Social work staff is also present at the annual Teen Picnic and biannual Support Group and helps with the planning of these events.

Research

The Bladder Exstrophy Program has a dedicated research team in basic science and clinical research. In the basic science laboratory, we are investigating the genetics of the bladder exstrophy-epispadias complex. In clinical research, we are conducting research studies to improve the quality of life for patients with bladder exstrophy.

Child Life

Child life specialists help patients and families adjust to and understand their health care experience while at Boston Children's Hospital. They use developmental interventions such as play to educate and prepare patients for tests and procedures. They also work to enhance patient coping skills during times of potential or realized fear, pain and anxiety. This team helps to plan and facilitate events for the annual Teen Picnic and biannual Bladder Exstrophy Support Group.

Overview

What is bladder exstrophy?

Bladder exstrophy (BE) is a relatively rare combination of complex anatomical anomalies that occurs during early fetal development.

The disorder usually involves several organ systems in the body, including the urinary tract, the reproductive tract, the digestive system, the muscles and the skin of the lower abdominal wall and the muscles and bones of the pelvis. Some bones of the pelvis are shortened compared to normal pelvic bones. Orthopedic surgery colleagues are routinely involved in care. Bladder exstrophy is seen in approximately 1 in every 40,000 births.

For individuals born with bladder exstrophy:

- The inner surface of the urinary bladder is open on the lower abdominal wall.
- The bladder is not closed and not covered as it would be normally by muscle and skin, but rather the bladder is essentially inside out with the inner surface exposed. The inner lining of the urethra is exposed on the top surface of the penis in boys or between the separated right and left halves of the clitoris in girls.
- In the absence of normal closure of the bladder and urethra, there is no ability for the bladder to store urine, and urine constantly trickles onto the exposed inner surface of the bladder and surrounding skin. Following birth, this may cause irritation of the nearby skin prior to repair.

What is epispadias?

Epispadias occurs when the urethra fails to close normally, and the inner lining of the urethra lays flat and exposed on the surface of the penis. Patients with bladder exstrophy will also have epispadias, but epispadias may also be present on its own. A diagnosis of isolated epispadias is considered less severe and is also less prevalent than classical bladder exstrophy. The prevalence of isolated epispadias is believed to be 1 in 200,000 to 1 in 400,000 live births.

In isolated epispadias, the bladder is closed and covered by the lower abdominal wall muscles and skin. In some patients with epispadias, there may be bony pelvis abnormalities similar to, but not as severe as, that seen with bladder exstrophy.

Similar challenges with urinary continence and vesicoureteral reflux may also be present in epispadias because the inner lining of the normally closed urethra is exposed on the surface of the penis or between the separated halves of the clitoris. In boys, approximately one-half of the urethra (the half closer to the tip of the penis) is open and exposed. In girls, the exposed urethra is complete and extends up to the bladder.

What is cloacal exstrophy?

The most complex and severe manifestation of the exstrophy-epispadias complex (EEC) is known as cloacal exstrophy. Individuals with cloacal exstrophy have anatomical findings similar to those found in bladder exstrophy. In addition, however, there is an abnormal connection of the bowel (intestine) to the exposed bladder. This connection needs to be separated with the help of general surgery colleagues, typically on the first or second day of life with, in some cases, bladder closure at the same time.

Similar to bladder exstrophy, in cloacal exstrophy the umbilical cord exits the body lower than normal on the abdominal wall. More often in cloacal exstrophy there may be a large abdominal wall hernia just above the umbilical cord, called an omphalocele. This will require repair, typically on the first or second day of life, or in stages, depending on the size of the omphalocele.

In cloacal exstrophy, the pelvic bone abnormalities are often more pronounced relative to those seen in individuals with bladder exstrophy. The bones of the vertebral column (backbone) and the spinal cord are often abnormally developed. Individuals with cloacal exstrophy require evaluation and management by orthopedic and neurosurgery

specialists. There may be associated abnormalities of one or both kidneys such as impaired development, abnormal function or position, or a solitary kidney.

What causes bladder exstrophy?

While the exact cause is unknown, the most popular theory suggests overdevelopment of a normal structure known as the cloacal membrane, which is present during early fetal development. In normal development, the cloacal membrane is present temporarily and eventually dissipates during normal fetal growth. This allows for normal growth of the bladder, urethra and other pelvic organs, muscles and bones, and normal coverage of these structures by lower abdominal wall muscle, connective tissue and skin. Over-development and/or prolonged presence of the cloacal membrane may prevent appropriate tissue development and ingrowth and the joining together of the supportive lower abdominal wall structures. This also results in failure of bladder and urethral closure, as well as "herniation" of the open bladder and urethra and the associated abnormalities described above.

Who is affected by exstrophy of the bladder?

Bladder exstrophy is slightly more common in boys than girls. Some reports show a clustering of bladder exstrophy in families, suggesting an inherited (hereditary) factor. However, the chance for parents to have another child with exstrophy of the bladder is small (one percent or less).

Components of the urinary system and their functions

Two kidneys
A pair of purplish-brown organs located below the ribs toward the middle of the back. Their function is to:

- remove liquid waste from the blood in the form of urine
- keep a stable balance of body fluid volume
- keep a stable balance of salts and other substances in the blood
- produce erythropoietin, a hormone that aids the formation of red blood cells

Two ureters
Narrow tubes that carry urine from each of the kidneys to the bladder. Muscles in the ureter walls continually tighten and relax forcing urine downward, away from the kidneys. About every 10 to 15 seconds, small amounts of urine are emptied into the bladder from the ureters.

Bladder
A spherical shaped, hollow organ located in the lower abdomen. It is held in place by ligaments that are attached to other organs and the pelvic bones. The bladder's walls relax and expand to store urine, and contract and flatten to empty urine through the urethra.

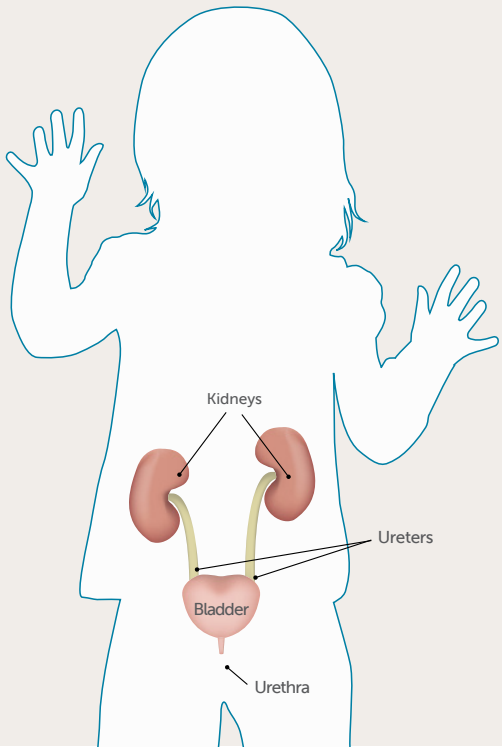
Two sphincter muscles
Circular muscles that help keep urine from leaking by closing tightly like a rubber band around the opening of the bladder.

Nerves in the bladder
Alert a person when it is time to urinate or empty the bladder, and help the bladder perform its function of storage and emptying.

Urethra
The tube that allows urine to pass outside the body. The brain signals the bladder muscles to tighten, which squeezes urine out of the bladder. At the same time, the brain signals the sphincter muscles to relax to let urine exit the bladder through the urethra. When all the signals occur in the correct order, normal urination occurs.

Ureteropelvic junction (UPJ)
The transition point from the renal pelvis to the upper ureter. This is a potential site of congenital obstruction to urine flow that leads to hydronephrosis (dilation of the renal pelvis).

Ureterovesical junction (UVJ)
The transition point from the ureter to the bladder. This is another potential site of congenital obstruction to urine flow.



How does the urinary tract work?

The body takes nutrients from food and converts them to energy. After the body has taken the food that it needs, waste products are left behind in the bowel and in the blood.

The urinary system keeps chemicals such as potassium, sodium and water in balance by removing a type of waste called urea from the blood. Urea is produced when protein is broken down in the body and is carried in the bloodstream to the kidneys, where it is removed.

Other important functions of the kidneys include blood pressure regulation and the production of erythropoietin, which controls red blood cell productions in the bone marrow.

Diagnosis

Detecting bladder exstrophy

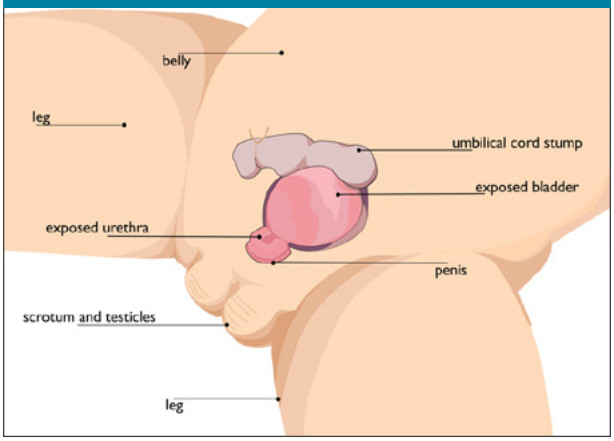
How is it diagnosed?

Bladder exstrophy may be diagnosed either immediately at birth based on typical physical findings, or during a fetal ultrasound or other imaging techniques such as magnetic resonance imaging (MRI).

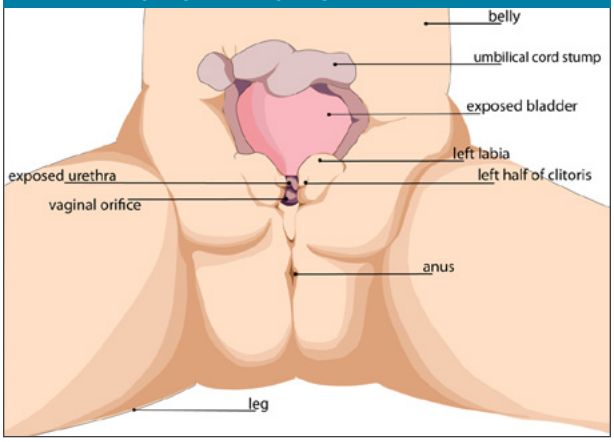
In most cases, exstrophy of the bladder is associated with the following:

- defect of the lower abdominal wall and visibility of the exposed inner lining (surface) of the bladder, which is usually bright pink
- shortened penis with the inner lining of the urethra exposed (epispadias) and open along the top surface of the penis in boys or in the surface of the body between the abnormally divided clitoris in girls
- abnormally separated (spread to the side) lower abdominal wall muscles
- displacement of the umbilical cord, lower than normal on the abdominal wall, usually immediately above the bladder
- umbilical hernia may be present (section of intestine protrudes through a weakness in the abdominal muscles), also called omphalocele
- absence of a normal appearing belly button
- anterior displacement of the anus, which opens on the skin closer to the scrotum in the boys or vagina in girls
- widened and shortened pubic and other pelvic bones
- outwardly rotated legs and feet in some boys and girls

BLADDER EXSTROPHY IN BOYS



BLADDER EXSTROPHY IN GIRLS



Illustrations by Sarah Dobbins

> “Approach your child’s treatments and path to health in small steps. Try not to feel totally overwhelmed by the diagnosis. It will be a lifelong issue. However, you can overcome the worries as you support your child. And remember: Attitude is everything.”

Parent of a 14-year-old boy with BE

Prenatal diagnosis

Prenatal care is an important aspect of health for every expectant mother and fetus. For the expectant family, the prenatal diagnosis of bladder exstrophy may allow time for learning about the condition and preparing for anticipated care. As the pregnancy progresses toward birth/delivery, it is helpful for the expectant family to access and use available health care professionals and resources, including this booklet and social support.

Boston Children’s offers a variety of resources for families, including the Advanced Fetal Care Center (AFCC). The AFCC provides support through prenatal counseling, treatment and follow-up for families with a confirmed or suspected fetal congenital anomaly. In addition, medical staff, social workers, child life specialists and chaplaincy services are available to support the expectant mother and her family. As part of family preparation and education, a tour of the inpatient floors will be offered to promote familiarity with the hospital environment.

Natural vaginal delivery is reasonable for a baby with epispadias or bladder exstrophy but is not ideal for a baby with cloacal exstrophy. When there is a prenatal diagnosis of bladder exstrophy, your obstetrician may recommend scheduled induction of labor for a more predictable delivery. In instances of prenatally diagnosed cloacal exstrophy, your health care team will likely prefer a scheduled caesarian section for delivery of the baby. The AFCC staff will collaborate with all health care team members to determine the optimal time for delivery and initial surgical intervention.

Regardless of the diagnosis, your newborn baby will be carefully examined and assessed by the neonatology team upon delivery. Shortly thereafter, the pediatric urologist will see your baby to confirm the diagnosis and provide recommendations for initial care. A more detailed plan for management, including the timing and technique for initial surgical repair, will also be determined. Optimally, there will be an opportunity for parents and newborn to begin bonding, and breastfeeding may also be possible, both before and after initial surgical repair.

Although further surgery beyond the initial repair is often required, the majority of individuals with bladder exstrophy do achieve urinary continence and adequate bladder function, as well as normal sexual function. After your baby’s birth, the pediatric urology team will work with you throughout the baby’s care.

Treatment

Treatment and care for bladder exstrophy

Treatment for exstrophy of the bladder begins at birth. Most important immediate care involves tying off the umbilical cord with a soft tie ("umbilical tape") and covering the bladder and urethra with soft, transparent material such as a transparent adhesive covering in order to protect the bladder and urethral surface from the diaper.

Beyond the immediate concerns for the health and well-being of any newborn, care of a patient with bladder exstrophy encompasses a range of surgical procedures and tests. Your child's primary care physician, pediatric urologist and the urology health care team will work together in order to develop the specific plan of treatment for your child.

The most important goals in the care of a boy or girl with bladder exstrophy are to:

- preserve normal kidney function
- develop adequate bladder function and promote urinary continence
- provide acceptable appearance and function of the external genitalia
- ensure that your child has a typical childhood, supported by physical and psychological health

Possible procedures for infants

There are several different approaches to initial repair for the boy or girl with bladder exstrophy, and institutions around the world approach the timing and/or technique for initial and definitive repair of bladder exstrophy in different ways.

Timing

Initial reconstructive surgery may be performed either within the first 2 or 3 days of life ("immediate") or at approximately 6 to 12 weeks of age ("delayed"). The "delayed" repair is preferred at Boston Children's. Delaying the initial closure may be the preference of some pediatric urologists based on practice or based on the specific anatomy of the baby. For example, some urologists prefer delaying initial surgery if the anatomy is too small for safe repair, especially if the baby is born prematurely.

Technique

There are three basic options for initial repair of bladder exstrophy. These options can be performed immediately or delayed depending on the anatomy and the preference of the physician and family.

Complete Primary Repair of Exstrophy (CPRE)

With the CPRE approach, the bladder is closed and the epispadias is repaired at the same time. Children who undergo CPRE will likely require additional surgery during later years to manage urinary incontinence and vesicoureteral reflux (VUR).

Modern Staged Repair of Exstrophy (MSRE)

This strategy involves three surgeries for reconstruction of bladder exstrophy:

- At the initial surgery, or 1st stage of this staged repair, the bladder and only the proximal urethra, the part of the open urethra nearest the bladder (partial epispadias repair), are closed/repaired.
- The 2nd stage involves completion of the epispadias repair.
- The 3rd stage is directed both at improving continence with bladder neck reconstruction (BNR) and eliminating VUR with bilateral ureteral reimplantation.

Urinary Diversion

This strategy involves creating a purposeful and functional connection between the urinary and intestinal tracts. Although there are several variations, the primary objective is to divert urine from the urinary bladder into the bowel (large intestine/colon) for temporary storage and then allowing for passage of the urine by emptying per rectum with the stool. Passage, at regular intervals, is under the control of the child. The bladder and urethra are closed but are "dry" and are neither exposed to the urine nor necessary for handling of the urine.

Complete Primary Repair of Exstrophy (CPRE) approach

The initial surgery for children with bladder exstrophy may combine the first two stages of the modern staged repair of exstrophy: closure of the bladder and the entire urethra (complete epispadias repair). Called Complete Primary Repair of Bladder Exstrophy (CPRE), this technique combines the goals of the first two stages of the staged reconstruction into a single operation, and may create an environment that allows more normal bladder function (cycling or filling and emptying) to begin earlier, which could optimize bladder growth and development, and also result in normal appearance of the external genitalia.

The operation consists of closure and internalization of the bladder, closure of the urethra (epispadias repair), repair of external genitalia and repair/closure of the lower abdominal wall muscles and skin. Some newborns may require osteotomy (see Osteotomy section below) to help bring the pelvic bones closer together. (Some centers prefer osteotomy in all newborns). Patients may stay in the hospital for 1 to 4 weeks of healing and will be carefully observed during this time. Antibiotics are given after the surgery to prevent infection, and the urinary tube in the bladder is usually removed several weeks following surgery. This approach has resulted in adequate urinary continence and voiding in some patients, therefore making it unnecessary to perform formal BNR to treat incontinence. Still, almost all boys and girls with bladder exstrophy will require surgery for treatment of vesicoureteral reflux, which is present in nearly all with bladder exstrophy (See Elementary School section.) The belly button is also reconstructed at the time of CPRE.

Although the results of complete primary repair have been promising, parents should expect the possibility of

Developmental Stage

Infant 0–12 months

All infants are unique and develop at their own pace. Talk to your infant's primary health care provider if you have any questions or concerns.

Typical Development

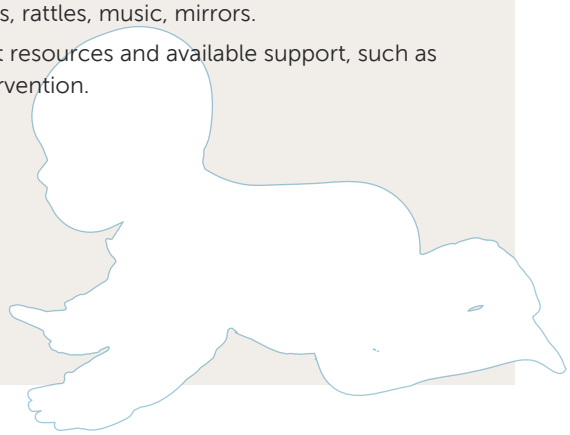
- Beginning to sit, roll, crawl, stand and walk
- Beginning to "coo," babble and use first words
- Starting to eat breast milk, formula and solids
- Learning from their environment (5 senses: hearing, smelling, seeing, feeling and tasting)
- Beginning to develop trust with and attachment to primary caregivers

Impact on Development

- Physical challenges: restriction of motion (traction or spica cast) and/or tubes
- Sensory overload: doctor's appointments and hospital stays may be overwhelming (new people, sounds, lights, smells)
- Trust and attachment challenges: large number of caregivers and interactions associated with pain (procedures, tests)

What You Can Do

- Keep routines/schedules as consistent as possible.
- Be aware of too much or too little light, music, touch, smell and encourage "positive touch".
- Promote coping/soothing techniques (swaddling, massage, rocking, sucking).
- Speak up/advocate as parents/guardians regarding pain management (use of sucrose, comfort holds) and infant's likes/dislikes: YOU know your infant the best.
- Encourage play.
- Developmentally appropriate toys include mobiles, aquariums, rattles, music, mirrors.
- Ask about resources and available support, such as early intervention.



> “Our child’s diagnosis has made us more aware of people and their differences. Watching (our son) go through his issues has made others feel if (he) can go through his issues and be so wonderful, they can step up and take on and accomplish anything they set their minds to.”
>
> *Parent of a 19-year-old boy with BE*

further surgery as their child gets older. All of these babies require temporary immobilization and traction (see below) in order to stabilize the initial repair and optimize healing. In addition, after surgical repair, lifelong follow-up care is necessary, with particular attention paid to the child’s development, growth and overall health, particularly bladder and kidney function.

In some boys and more often in girls, successful complete primary repair will eliminate the need for BNR later in life. (Bladder neck reconstruction is the 3rd stage of the modern staged repair.)

Complete Primary Repair of Exstrophy (CPRE) approach: Delayed vs. Immediate

Delayed Complete Primary Repair of Bladder Exstrophy (CPRE) is the preferred treatment at Boston Children’s.

We believe that delaying CPRE beyond the first 2 or 3 days of life may have several advantages for the child and the family unit:

- Delay presents an opportunity for normal bonding between the baby and parents before the initial reconstructive surgery and lengthy recovery period that follows.
- Time between birth and initial repair allows for growth, development and additional maturation of organs (including the bladder and genitalia) and systems prior to the complex surgery, and makes the anesthesia and surgery safer.

Osteotomy (pelvic)

When a child with bladder exstrophy is born, several physical abnormalities typically exist. In nearly all cases, the pubic bones are widened or spread away from the midline (middle of the body). In some cases, the legs and feet may be slightly outwardly rotated. Given these abnormalities, a pediatric orthopedic surgeon who specializes in treating abnormalities of bone and muscle will evaluate and help to care for your child.

An osteotomy is a surgical operation in which a bone is cut to shorten, lengthen or change its alignment. With bladder exstrophy, where the pubic bones are spread widely apart in the front, just above the genitalia, the osteotomy procedure helps partially correct the abnormalities of the pelvic bones by bringing these bones closer together. This creates a more normal support of the soft tissues of the pelvis, which may also help with overall healing and potentially, eventual urinary continence.

This surgery, performed by an orthopedic surgeon, helps to decrease the tension placed on the pelvic bones and soft tissues at the time of initial exstrophy closure. In this way, it increases the chances for successful healing of the closure. Past experience suggests a benefit of improved outcome and eventual continence for those boys or girls who have osteotomy at the time of bladder exstrophy repair.

Newborn babies who undergo immediate repair (2 or 3 days of life), when the pelvis is still malleable (pliable) due to the mother’s production of the hormone relaxin, may not require osteotomy. If the closure is performed after 2 or 3

> “Normal (regular) life is still much more prominent than the occasional challenge. The kids will have the same interests and experiences as any other child. Also, we learn how remarkably resilient we are. The kids and the parents will be able to handle the challenges ahead.”
>
> *Parent of a 9-year-old girl with BE*

days of age, this hormone has decreased, and an osteotomy will need to be performed. Osteotomy is also recommended for all reoperations for previously failed closures and for almost all babies with cloacal exstrophy.

Umbilicoplasty or “belly button surgery”

In patients born with bladder exstrophy, the belly button (umbilicus), which is the stump of the umbilical cord, is attached to the upper portion of the bladder—an abnormally low location on the abdominal wall. The umbilical stump must be removed during the initial reconstructive surgery.

Research has found that the belly button is an important aesthetic landmark for most children and adolescents. To create a more normal appearance, umbilicoplasty, or “belly button surgery,” is often performed. This plastic surgery is often combined with the initial closure or performed separately, later on in a child’s life. It is usually performed under general anesthesia and may be combined with other surgical procedures. In most cases, there is no additional surgical scar as a result of the procedure, as the primary incision is made at the planned site of the belly button itself.

Traction

After the initial surgery, your child will be partially immobilized, placed in “traction” for several weeks. This helps stabilize the complex repair and optimizes healing in the most important period of time. There are several options:

- **Modified Bryant’s traction** consists of supporting both legs by gently supporting the legs straight upward toward the ceiling, away from the bed, with the legs bent at the hips as the baby lies on his or her back.
- **Buck’s traction** is used to immobilize position and align the legs and hips in a straight line with the child lying flat on the bed.
- At many institutions, including Boston Children’s, the preferred technique for immobilization is the “spica” cast. With this technique, a plaster-type cast is applied to the lower abdomen, pelvis and legs. The **spica cast** is custom-fit and placed by the orthopedic surgery team immediately after initial repair is completed. The baby’s skin and body are protected inside the cast with padding. The spica cast allows for close physical contact between you and your baby, and may allow your baby to leave the hospital for home earlier while he or she is healing in the cast.

All of these types of traction allow for close physical contact with parents and for breastfeeding if desired.

There are other approaches to osteotomy and postoperative immobilization that involve more extensive incisions in the pelvic bones and more elaborate external fixation devices. It is possible that alternative approaches may be favored at your institution or by your child’s orthopedic surgeon.

> “It made me stronger. Our marriage benefited from it. It brought my wife and I together to ‘team up‘ to support our daughters.”

Parent of a 9-year-old girl with BE

Modern Staged Repair of Exstrophy (MSRE) approach

MSRE consists of three surgical components:

- the initial repair (bladder closure and partial epispadias repair)
- completion of the epispadias repair (in boys)
- bladder neck reconstruction with bilateral ureteral re-implantation

Stage 1: Initial Closure

During this first operation, the bladder and proximal (closest to the bladder) urethra, and the abdominal wall are closed. The belly button is reconstructed, and an osteotomy is sometimes performed (the pelvic bones are reformed to aid in the repair). Whether or not the infant needs an osteotomy is dependent on how soon after birth the operation is performed. (With certain physical factors, or if the initial closure takes place three days or more after birth, then osteotomy is felt to be necessary.)

After surgery, your baby will be placed with the lower abdomen, pelvis and legs in traction/ immobilization in order to aid the healing of the pelvic bones and the repair. Patients may stay in the hospital for three to six weeks of healing and will be carefully observed during this time. Antibiotics are given after the surgery to prevent infection, and the urinary drainage tube in the bladder is usually removed four weeks following surgery. As the size of the bladder increases over time, your child will be ready for the next stage, the epispadias repair.

Stage 2: Epispadias Repair

Epispadias is a congenital (present from birth) defect in the formation of the urethra that is always associated with bladder exstrophy.

In boys, the proximal part of the urethra close to the bladder is closed along with the bladder at the first stage of MSRE. The distal (closer to the tip of the penis) open and exposed inner lining of the urethra, on the top surface of the penis, was left untouched at the first stage and is now closed at the second stage of the MSRE (epispadias repair).

In girls, the urethra is usually closed along its entire length at the first stage of MSRE.

The surgical procedure used to correct epispadias reconstructs the missing portion of the urethra and restores the appearance of the external genitalia. The exact surgery used differs according to the complexity of each individual case. Because of the varying anatomy of each child with bladder exstrophy, surgical procedures will not only have different outcomes but also have different effects on continence. This repair occurs around six months of age, but the time and extent of the surgery are dependent on the size of the bladder and the condition of the urethral tissue (urethral plate).

During this surgery, the urethra is repositioned to its normal placement. After the surgery, the dressings will remain in place for several days, and the diverting urinary catheter (suprapubic cystostomy tube) will typically be removed after several weeks. As with any surgical procedure, postoperative care will be individualized as needed.

> “Friends, school, sports—you would never know from the outside looking in that our child was any different.”
>
> *Parent of a 9-year-old boy with BE*

Stage 3: Bladder Neck Reconstruction (BNR)

Performance of this stage of the MSRE depends on sufficient growth of the bladder as judged by the volume of urine it can hold (capacity). Before making the decision to proceed with this surgery, your child's bladder anatomy and function may be evaluated by one or more of several studies. These may include a bladder and urethra anatomy x-ray (voiding cystourethrogram (VCUG)), urodynamic study (UDS) and/or exam under brief general anesthesia; bladder capacity may be evaluated in this setting as well. Bilateral ureteral reimplantation is almost always performed with BNR. This procedure is usually performed between the ages of five and 10. (See Elementary School section for more details.)

When necessary, following initial repair with the CPRE technique, BNR and bilateral ureteral reimplantation are performed in similar fashion for boys and girls in order to improve urinary continence and eliminate vesicoureteral reflux.

Hernias (inguinal)

A hernia occurs when part of the intestine protrudes (pokes out) through a weakness in the abdominal muscles. A soft bulge is seen underneath the skin where the hernia has occurred. In children, a hernia usually occurs in either the groin area (an inguinal hernia) or the belly button area (an umbilical hernia). For children with bladder exstrophy, inguinal hernias are common due to associated lower abdominal wall muscle and connective tissue abnormalities. Hernias develop often within the first few months of life or at some point during infancy.

During a hernia operation, your child will be placed under general anesthesia. A small incision is made in the area of the hernia (lower abdominal wall). The intestine, when present in the hernia defect, is replaced back into the abdominal cavity. The muscles are then stitched back together to strengthen the area.

This procedure is usually performed as outpatient surgery, which means that children who have an inguinal hernia surgically repaired can often go home the same day they have the operation. Your medical team will provide you with more detailed post-operative instructions.

Possible Procedures for Toddler/Preschool Children

Ureteral Reimplantation

Vesicoureteral reflux (VUR) is abnormal backward flow of urine from the bladder up to the kidney(s). A ureteral reimplant is a surgical procedure that corrects VUR by repositioning the ureters within the bladder so that the urine flows only in the normal direction, from kidney to bladder. (The ureter is that tube that carries the urine from the kidneys to the bladder.) If the ureter is connected to the bladder incorrectly, it can let the urine flow backwards to the kidneys, potentially causing damage to the kidney(s).

During the procedure, the doctor will reposition the ureter so that it's connected to the bladder correctly. The surgery usually does not exceed a few hours. Your child will have one or more drainage tubes after the surgery that will need to stay in place for a few days. In most cases, children are released from the hospital two to four days after the surgery.

Developmental Stage

Toddlerhood/Preschool 1–5 years

All children are unique and develop at their own pace. Talk to your infant's primary health care provider if you have any questions or concerns.

Typical Development

- Jumping and climbing
- Magical thinking
- Short attention spans
- Increasing language skills: receptive language six months ahead of verbal
- Fear of bodily injury
- Temper tantrums and negative behavior are normal
- Establishing routine behaviors
- Increasing independence: getting dressed, etc.
- Using play to learn, express self and work out fears

Impact on Development

- Regression: (e.g., using pacifier during hospitalizations, using diaper after being potty trained)
- Fear of strangers heightened due to large number of caregivers when hospitalized and interactions associated with pain (procedures, surgery)
- Increased stress and acting out when routines are not followed
- Feeling loss of control (during hospitalizations or tests—can't eat, etc.)

What You Can Do

- Keep routines and schedules consistent.
- Set limits and offer appropriate choices (e.g., "You have to take your medicine. Do you want to do it by yourself or do you want mommy/daddy to help?").
- Promote positive coping and soothing techniques (e.g., taking deep breaths).
- Speak up/advocate as parents/guardians regarding pain management and your child's likes/dislikes. YOU know your child the best.
- Give concrete explanations and one-word descriptions for hospital visits and medical procedures.
- Encourage play: developmentally appropriate toys include bubbles, cause-and-effect toys, pop-up books, lights, etc.; provide opportunities for medical play (play doctor with your child so he/she can "play out" what they have experienced or fear).

Possible Procedures for Elementary and Middle School Children

Bladder Neck Reconstruction (BNR)

The term "bladder neck reconstruction" refers to a surgical procedure that reconstructs the anatomy of the connection between the bladder and the urethra or bladder neck. The goal of this procedure is to control urine leakage and potentially enlarge the bladder as a result.

Typically, in bladder exstrophy, this area is too wide/ open and is unable to retain urine within the bladder. The bladder neck is the lowest part of the bladder, the site where urine leaves the bladder and enters the urethra on its way to the surface of the body. The primary goal of this surgery is continence or dryness and freedom from urine leakage. This is done by narrowing the channel of urine flow in the area of the bladder neck.

The best candidates for BNR had a successful primary closure and, over time, developed a good-sized bladder with good bladder growth. (If your child's medical team used the immediate CPRE approach at birth, BNR may not be necessary.) Bilateral ureteral reimplantation is almost always necessary at the time of BNR, in order to make room for bladder neck reconstruction surgery and in order to eliminate VUR.

Bladder neck reconstruction surgery carries some risks, including:

- persistence of incontinence
- inability to pass urine (urinary retention)
- persistence of vesicoureteral reflux (VUR)
- infection in the urine and/or kidney(s)

In some children, additional surgery may be necessary to ensure good kidney function and urinary control. Procedures may include enlarging the bladder so that it may hold larger volumes of urine to help keep your child dry. This is known as bladder augmentation (described in the next section).

An alternate technique for emptying the bladder is clean intermittent catheterization (CIC)- and/or an alternate route-continent catheterizable conduit- may also be necessary.

The timing of BNR depends to some degree on your child's bladder capacity, as well as his or her emotional and developmental status. With BNR, it is important that both the child and the family are prepared for urinary continence, as the bladder voiding training before and after the surgery is rigorous and requires time and effort to be successful. Your child should be able to participate in a bladder-training program before and after surgery. The training begins six months before surgery. Your medical team will review the specifics of your voiding program. Refusal or difficulties to follow aspects of the training program, such as timed voiding, hydration, or medication, could indicate that your child is not ready for this surgery.

Developmental Stage

Elementary School
6–10 years

All children are unique and develop at their own pace. Talk to your child's primary health care provider if you have any questions or concerns.

Typical Development

- entering school and forming relationships with peers
- beginning to develop a sense of control over their environment
- understanding relationship between things and ideas, seeking out knowledge
- devising new games and new rules
- using play to learn, express self and work out fears

Impact on Development

- regression (e.g., baby talk)
- increased stress and anxiety surrounding painful procedures, tests and illness (due to their ability to now ask questions)
- anxiety about body alteration, fear of not being well again
- feeling left out/ different due to time away from routines, school, family, friends

What You Can Do

- Keep routines and schedules consistent (school, family time, etc.); consider home tutoring if needed.
- Set limits and offer appropriate choices.
- Promote positive coping and soothing techniques.
- Give concrete explanations and simple descriptions.
- Reassure your child that the condition is not their fault.
- Maintain open communication with your child and encourage him/her to ask questions.
- Encourage your child to take an active role in the treatment of their illness (e.g., asking their doctor questions).
- Encourage play by making developmentally-appropriate toys, games and crafts available.
- Act out what they are experiencing with medical play.

Bladder Augmentation

Bladder augmentation is a surgical procedure that increases the capacity of the bladder, allowing it to hold more urine. Bladder augmentation may be recommended to help treat otherwise irreversible forms of incontinence, protect the kidneys from high-pressure urine storage or, at times to assist in eliminating vesicoureteral reflux.

With bladder augmentation, the current standard of care is to use a small portion of the child's intestine (bowel) as a patch that is placed on the bladder. During the surgery, the top of the bladder is opened, and a small patch of stomach, small or large intestine is used to make the bladder larger.

The operation increases the room inside the bladder (bladder capacity) and decreases the pressure within the bladder in order to make storage of urine safer. This, in turn, allows the kidneys to drain better.

Bladder augmentation is chosen as a treatment plan for a patient only after a thorough physical exam and appropriate testing is performed.

Patients who undergo bladder augmentation must learn to place a catheter (a small, flexible, plastic tube), or urinary tube, into their urethra (self-catheterization). They must also not have any other bowel or urethral disease.

Developmental Stage

Middle School 11–13 years

All children are unique and develop at their own pace. Talk to your child's primary health care provider if you have any questions or concerns.

Typical Development

- Scientific thinking
- Expressing more complex feelings
- Believing in one's own competence
- Developing sense of self-image, self-development and gender
- Forming peer relationships and social comparisons
- Engaging in school, schedules and routines
- Asserting independence
- Beginning to understand sexual development, puberty

Impact on Development

- Regression (sleep with stuffed animal)
- Challenging the need to rely on parents when it is more normal to want more independence
- Considering long-term effects of illness and impacts on their future
- Feeling increased stress due to time spent away from normal routines at school, as well as friends and family
- Thinking of the impact on adolescent and future adult life (especially in terms of sexuality)

What You Can Do

- Provide appropriate choices.
- Provide opportunities for control.
- Encourage/promote/provide attainable goals and responsibilities.
- Give appropriate jobs.
- Respect reasonable privacy.
- Encourage increased participation in care and independence (e.g., alone time during doctor appointments).
- Encourage continued focus on educational/future goals.
- Give HONEST answers when questions are asked; if you don't know, say you don't know.
- Begin age-appropriate sex education.

> “It is a life of ups and downs, but the ups will outweigh the downs. Exstrophy is only a part of who they are and with the many obstacles they will go through, they are extremely amazing individuals.”
>
> *Parent of 14- and 16-year-old boys with epispadias and bladder exstrophy*

Continent Catheterizable Conduit (urostomy)

A urostomy is a surgically created alternate opening for access to the urinary system that is made when long-term drainage of urine through the urethra is not possible. With a continent urostomy, the goal is to give your child control over urine flow from their body.

The Mitrofanoff principle, named after the surgeon who developed this idea, creates a continent urostomy, or stoma, which is an opening on the skin (typically in the lower abdominal wall) in order to create a connection between the skin surface and the bladder. The appendix is often used for this purpose. The urine empties from the bladder by inserting a catheter into the small opening (stoma), which is usually on the lower abdomen just above the pubic area. Urine will not flow out of the stoma unless a catheter is inserted.

You and/or your child will typically need to catheterize approximately four or more times a day to drain the bladder. In this manner, your child will have control over their urine output.

Around the time of such surgery, after initial healing and during your child’s time in the hospital, the nurses will help perform and teach catheterization and care. However, it is important that you and your child become involved in the care as soon as possible.

Clean Intermittent Catheterization (CIC)

When a child is unable to void (urinate) successfully or completely, an alternate approach is available: self-catheterization. Clean intermittent catheterization (CIC) may be performed either through the reconstructed urethra or via an alternate route such as a continent catheterizable conduit.

If you are interested in hearing more about other children’s and parents’ experience with catheterization, please visit our Experience Journal.

Possible Procedures for Adolescents and Young Adults

Reconstructive surgery

Reconstructive surgery for a child, adolescent or young adult with bladder exstrophy may help revise, smooth or hide the abdominal wall or other surgical scars. Your urology care team may seek the advice and expertise of a plastic surgeon for the optimal care of your child. Four basic types of surgical procedures are most commonly used.

Scar revision

The surgical procedures that children with bladder exstrophy undergo may result in scarring on their abdominal and genital areas. The degree of improvement that can be achieved with scar revision will depend on the severity of scarring and the type, size and location of the scar. If scar revision is indicated, either your pediatric urologist or plastic

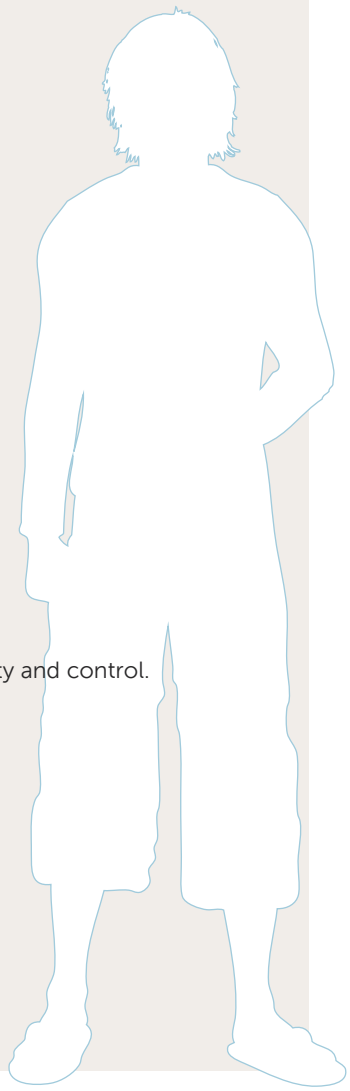
Developmental Stage

High School 14–18 years

All adolescents and young adults are unique and develop at their own pace. Talk to your adolescent's primary health care provider if you have any questions or concerns.

Typical Development

- Focusing on body image (how they compare to their peers)
- Establishing and maintaining peer relationships
- Concern with what others think
- Wanting/asserting independence
- Feeling "invincible"
- Testing values
- Preparing for adult lifestyle and thinking about future plans
- Developing new relationships with parents
- Developing sexually and possibly becoming interested/engaging in sexual activity



Impact on Development

- Becoming more aware of their illness and impact on the future
- Decreasing adherence to medical regimens (e.g., catheterizing, taking medicines)
- Denying their illness
- Defining self by disease, instead of as a person
- Becoming more "body conscious" in sexual encounters; may delay sexual activity

What You Can Do

- Encourage and provide privacy when possible.
- Promote healthy personal hygiene choices (nutrition, exercise, etc.), independence, mobility and control.
- Encourage normal peer/family interactions.
- Encourage continued focus on educational/future goals.
- Give HONEST answers when questions are asked; if you don't know, say you don't know.
- Be an advocate or role model.
- Continue to set limits, yet allow for appropriate control and separation.
- Discuss nuances of sex and address concerns your teen may have.
- Utilize resources available (e.g., clinicians, age-appropriate sex education classes).

surgeon will recommend the scar revision techniques to achieve the best results for your child's specific needs. In addition, umbilicoplasty ("belly button surgery") may be performed during adolescence. Nowadays, most infants have this procedure performed during initial closure, but it can also be performed later in development. These procedures are optional and may not be necessary for your child; all patients are assessed on a case-by-case basis.

Monsplasty

The mons pubis is the area over the middle of the lower abdominal wall, just above the genitalia, where the pubic bones meet. This area is composed of soft tissues such as connective tissues and skin. Some bladder exstrophy patients may experience significant scarring and/or depression of the tissues in the mons pubis area after their surgeries. A monsplasty is a surgical procedure that removes scar tissue present, flattens the area of the mons pubis and joins hair-bearing skin for a more normal appearance.

Vaginal Reconstruction

A narrowing at the skin surface, known as vaginal stenosis, may develop after reconstruction of a girl with bladder exstrophy. This narrowing may occur with changes in vaginal tissue. If necessary, doctors can address the issue through a relatively minor surgical procedure called a vaginoplasty. In rare instances, a more extensive reconstructive surgical procedure involving reconstruction of the vaginal canal may be necessary.

Vaginoplasty is usually performed during high school years or later in young adulthood.

Penile Reconstruction

Boys require extensive reconstruction of the urethra and penis at the time of initial repair regardless of whether the initial repair is performed using the CPRE technique or the MSRE approach. Some boys will require surgical revision of the penis (penile reconstruction) later in life in order to repair or revise surgical scars that remain from the original surgery and/or to straighten or lengthen the penis. This procedure may be performed at any time, if needed, but is typically performed in either adolescence or young adulthood.

Developmental Stage

Young Adult

All young adults are unique and develop at their own pace. Talk to your young adult's primary health care provider if you have any questions or concerns.

Typical Development

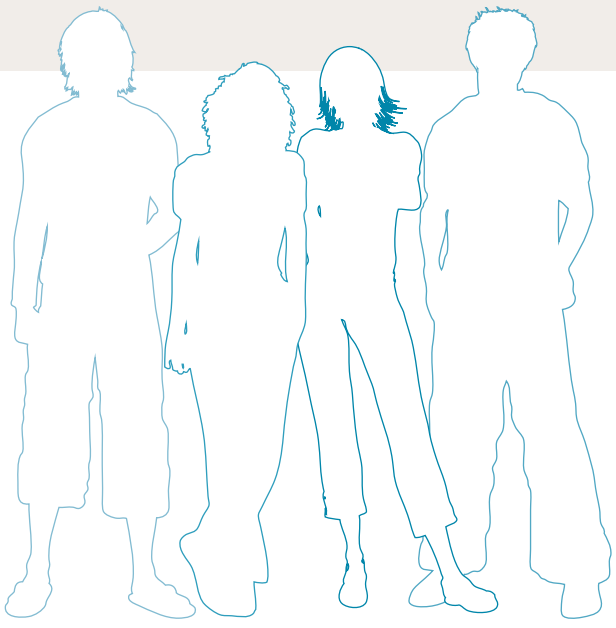
- Transitioning from high school to college/job/workforce
- changing environment
- Establishing long-lasting relationships and commitments
- Pursuing the formation of moral values
- Becoming sexually active
- Taking on more responsibility in their life
- Developing a sense of autonomy

Impact on Development

- Becoming more accepting of illness
- Increasing self-management and independence

What You Can Do

- Encourage autonomy.
- Consider advance preparation for independent living (e.g., select single dorm room, talk with employer).
- Consider communication with professors and employers regarding conditions.



Tests

Diagnostic tests and labs

Diagnostic tests for bladder exstrophy patients are periodic and may span the entire time your child receives care at Boston Children's.

Renal Ultrasound (RUS)

In a renal and bladder ultrasound, (RUS), sound waves are passed through the body. When sound waves "bounce off" the tissues of an organ, the waves can be changed into pictures by a computer. These "snapshots" of the bladder and kidneys are used to check for problems and growth. This test is painless and does not require an injection.

Plain abdominal x-ray (KUB)

An x-ray of the abdomen, often referred to as a KUB (kidneys, ureters and bladder), is needed to investigate these internals structures of the body. Similar to a renal ultrasound, x-rays are painless and require your child to remain still while the technician takes a series of photos.

MRI of pelvis

A pelvic MRI (magnetic resonance imaging) scan is an imaging procedure that uses magnets and radio waves to create a photo of the area between the hip bones (the pelvic area). During this procedure, your child/adolescent will need to remain motionless in the MRI machine for approximately 30 to 60 minutes. In some instances, sedation or general anesthesia may be necessary.

Voiding cystourethrogram (VCUG)

A VCUG is a test that uses a special x-ray technology called fluoroscopy to observe the urinary tract and bladder during filling and voiding. Contrast fluid is instilled into the bladder through a catheter. Imaging obtained from the procedure is used to assess the presence of residual urine after voiding.

DMSA renal scan

A DMSA (dimercaptosuccininc acid) renal scan is a nuclear medicine test that allows your child's doctor to see the size, shape, function and position of the kidneys. It can detect scarring caused by frequent infections. To perform the scan, a small amount of liquid is injected into your child's veins. The liquid goes to the kidneys where it is processed, and then images are obtained with a camera.

Radionuclide cystogram (RNC)

A radionuclide cystogram (RNC) is a nuclear medicine test used to check if your child has vesicoureteral reflux. Fluid is instilled into the bladder through a catheter and images are obtained with a camera.

> “They will do everything that other kids do. Bladder exstrophy is just part of who they are.”
>
> *Parent of an 8-year-old boy with BE*

Renal MAG3 Scan/MAG 3 with Lasix Renal Scan

A renal MAG3 (mercapto-acetyl-triglycine) scan is a nuclear medicine test that shows how the kidneys are functioning and how well they are able to drain fluid into the bladder. The test can show if there is any obstruction of the kidney.

Urodynamics study (UDS)

A urodynamics study (UDS) is a test to assess how well a child's bladder and urethral sphincter are working at collecting, holding and releasing urine. During the test, a thin, flexible tube (catheter) is placed into the bladder and the bladder is filled slowly with a warm saltwater (saline) solution to measure urine flow and bladder pressures.

The study may consist of one or more of these four individual parts:

- urinary flow rate (uroflow)
- urethral pressure profile (UPP)
- cystometrogram (CMG)
- electromyogram (EMG)

The combination of tests used in the urodynamic study will depend on your child's condition.

Lab Work

Complete blood count (CBC)

A complete blood count (CBC) measures the number of red, white and other blood cells. A CBC can be done for many reasons, such as to check for infection or a low red blood cell count (anemia).

Electrolytes

An electrolytes blood test measures the amount of sodium, chloride, potassium and bicarbonate found in the blood.

Creatinine

A creatinine level test is a blood test that measures how well the kidneys are filtering wastes from the body.

Urinalysis

Urinalysis is a common diagnostic test that is used to determine if a patient has a urinary tract infection (UTI). It consists of a laboratory examination of urine for various cells and chemicals, such as red blood cells, white blood cells, infection, or excessive protein.

Urine Culture

Urine culture is a common diagnostic test used to detect the presence of an infection and identify organisms in a urine sample that can cause urinary tract infections.

Tips

Preparing your child

Talking with your child about upcoming procedures or surgeries can be a source of stress and anxiety. You may be concerned about how to best approach these topics with your child as well as when and how to have these discussions.

We understand that every child is a unique individual. Each child's age, past hospital experiences, temperament and coping style can affect how he or she copes with upcoming procedures or surgery.

Children and adolescents are often present when future procedures or surgeries are discussed with the team. They are often encouraged to participate in these discussions at a level appropriate to their age. Therefore, we encourage you to discuss these plans with your child before returning to the hospital. It will help your child prepare for the experience and give him or her time to process information previously heard and any new information. This will help build and maintain trust between you and your child.

Here are some tips which you may find helpful:

- When talking with your child, start by using a frame of reference such as, "Do you remember when we last went to the hospital and the doctor and nurse spoke with us about a surgery you need? Well, it's time we start talking together about when this will happen."
- Remind your child who will be staying with him or her with reassurances that he or she will not be alone during this experience.
- Keep explanations simple and wait for your child to ask questions. This way you will learn what is important to him or her.
- Be sure your child is told at a time when you will be available to answer questions. Some families would prefer to avoid a discussion at bedtime. Others, depending on their child, find bedtime a time when their child is comfortable having a sensitive discussion. If so, revisit the topic in the morning. A child needs time to process this type of information at his or her own pace and it helps if you are there to answer questions.
- If your child asks a question that you cannot answer or do not feel comfortable answering, a good way to respond is, "That's a great question. Let's write it down so we do not forget it. We can ask the doctor or the nurse."
- If your child is younger and needs concrete reminders about the surgery or procedure, consider taking your child to your room and pack your suitcase together. This is a concrete way to let your child know that you are going to the hospital too.
- You know your child the best. Do not hesitate to contact a member of the team if you want to think together about approaches that are best for your child and family.

Age-Specific Tips

Parents often ask for information about how and when to talk to their child. There are many ways to help children prepare for a procedure, surgery or inpatient stay in the hospital. A child's personality, language development and ability to comprehend information can affect understanding of procedures, surgeries and other reasons for coming to the clinic or hospital. Previous experiences can also influence their response. Since children develop at different rates and have personalities and coping styles unique to them, these guidelines may not describe your child exactly or accurately, but may be helpful in guiding you.

Newborns to 2-years-olds

When your child is very young, concentrate on preparing yourself for the hospital. If you feel at ease, your child is usually able to sense this and react in the same way.

> “Our child’s diagnosis is just one part of the family dynamic. When we are going through a surgery, it teaches us how we all need to work together to support each other.”
>
> *Parent of a 9 year-old boy with BE*

2- to 3-years-olds

At this age, children do not understand time in the same way older children and adults do. Consider telling your child in simple language about his or her operation or procedure one to two days before going to the hospital. Playing with your child and incorporating the upcoming hospital events in your play together will be a concrete way to help your child at this age.

3- to 6-years-olds

At this age, children are beginning to learn about the days of the week and develop a sense of time. However, it is difficult for a child to understand why an operation or procedure is necessary. Your child may worry that they did something wrong. Consider reassuring your child that the hospital stay is about having something fixed and is never a punishment. Use simple, short explanations. We would recommend telling a 3- to 4-year-old child about an operation or procedure one to two days before going to the hospital and a 5- to 6-year-old child three to five days ahead of time. You may want to remind your child about the last visit with the team when these decisions may have been made. Again, playing with your child to help them understand is concrete and fun for you both.

7- to 11-years-olds

There is great variability in maturity within this age group. However, elementary-age children are able to understand the reason for an operation or procedure and have typically participated in discussions with the team. For younger children in this age group, you may want to tell your child of plans a week before going to the hospital if they have not asked questions or discussed their situation since their last visit with the team. This will give your child plenty of time to ask questions and to talk about any worries or concerns.

12-year-olds to adults

At this age, it is best to include your child, teen or young adult from the very beginning of any discussions. Encourage your child to ask questions and to talk about any worries or concerns. Most children are struggling for independence from their parents while at the same time seeking their support. Your child may have valuable suggestions about what works best for him or her. Consider an approach which encourages this valuable feedback about how you can best help them through the hospital stay or procedure. This is also a good time to ask your child if they would like to speak to a young adult mentor trained in our Family to Family Program, who has experienced a similar surgery or procedure.

FAQs

Frequently asked questions

Q: How many surgeries will my child need for bladder exstrophy?

It depends on where your child is treated. Here at Boston Children's Hospital, we use a method called complete primary repair of exstrophy (CPRE) to treat the condition with a single surgery not long after a child's birth.

CPRE allows the bladder to be closed and the epispadias to be repaired at the same time. The CPRE operation includes closure and internalization of the bladder (moving it inside the body), closure of the urethra (epispadias repair), repair of the penis in boys or the external genitalia in girls and repair/closure of the lower abdominal wall muscles and soft tissues. (See Infancy section for more details.)

Q: Will my child need bladder neck reconstruction if she undergoes CPRE?

The answer to this question will be based on your child's ability to have normal dry periods between voiding/bladder emptying (urinary continence). Your child's bladder capacity and his or her ability to stay dry—referred to as a dry interval—help to determine the need for a bladder neck reconstruction (BNR).

In the modern staged repair of exstrophy (MSRE) approach, which is a three-step surgery, BNR is planned on all patients. In the single-surgery complete primary repair of exstrophy (CPRE) approach, however, BNR is performed only on patients who have not achieved satisfactory urinary continence.

So, determining whether or not your child will need BNR after CPRE depends on many factors, most important of which is how his or her ability to be dry has developed following the initial CPRE surgery. Based on our experience and that of other institutions, about three-quarters of boys and one-half of girls may eventually need bladder neck reconstruction following CPRE.

Q: Will my child have the ability to be appropriately dry?

One of our major goals when treating children with bladder exstrophy is to help your child get total control of her bladder. Achieving continence may be challenging, but it's generally attainable in all cases.

Many children do well with the practice of good health habits, such as adequate fluid intake and a regular voiding program. If your child undergoes the CPRE approach to initial surgical management of bladder exstrophy, he or she may not need bladder neck reconstruction to help attain continence (as described above).

However, some children may need help with emptying their bladder. These children may use a technique called clean intermittent catheterization (CIC). This involves the passage of a soft pliable catheter via either the urethra or a catheterizable conduit with a well-hidden lower abdominal wall stoma (a surgically constructed tube/opening connecting the bladder to the skin surface). This allows for safe and painless emptying of urine from the bladder.

Although it is uncommon, some children with bladder exstrophy may need bladder augmentation in order to increase the bladder's capacity for holding urine.

Q: Will my child's genitals look "normal"?

In addition to working to provide your child with total control over his or her bladder, we also aim to ensure that your child's genital area has a satisfactory cosmetic appearance. Genital repair is performed at different points in care or at different ages for boys and girls, and may depend on whether your child's doctor opts for single surgery (CPRE) or the three-part surgery (MSRE).

> “First thing—breathe. You are in the best place for your child’s medical needs. It will be hard, but you will get through it with great support.”
>
> *Parent of a 18-month-old boy with BE*

Q: If I have another child, will he or she also have bladder exstrophy?

The risk of a sibling being born with bladder exstrophy is very low—less than 1 percent.

Q: Will my child be able to play sports?

Definitely. In most cases, your child should be able to enjoy a normal, active childhood. The one difference is that he or she will potentially have to take special care regarding urinary continence. Even children with bladder exstrophy who are normally continent during the day may have “stress incontinence” or incontinence while running, jumping, coughing or engaging in any other activity that puts stress on the bladder. This may cause your child to have small to moderate urinary leakage, and he or she may need to wear a pad while being active. However, this doesn’t mean that your child cannot play and enjoy sports.

Q: What are the expectations regarding my child’s quality of life?

This is a question unique to each family. Our interdisciplinary team welcomes the opportunity to discuss any of your concerns. We expect that your child will be able to participate in any and all of life’s joyful moments and childhood activities. Your child’s experience should be much the same as any other child, with the understanding of and appreciation for the fact that every individual’s life is special and unique.

Q: Will my child be able to have children of their own?

Almost all boys with exstrophy produce healthy sperm. However, some may have difficulty fathering children through traditional sexual intercourse. The potential difficulty is in the delivery of the sperm to the egg. Difficulties may be a result of backward flow of semen during ejaculation (also known as retrograde ejaculation), the inability to ejaculate at all or low semen volume. The good news is that, if necessary, couples may be able to use Assisted Reproductive Techniques (ART), such as intrauterine insemination and in vitro fertilization.

If girls with bladder exstrophy have a problem with sex and infertility, it’s most often a result of an anatomical concern. In some, the vaginal opening may be too narrow and may require surgical enlargement. When necessary, this minor procedure should allow for normal sexual intercourse and achieving pregnancy.

Some women with bladder exstrophy may develop uterine prolapse, in which anatomical support for the uterus is lacking and the uterus may protrude into the vagina during the later stages of pregnancy. If this happens, a woman may need surgery, but she can still have babies by Caesarean section (C-section), so that her urinary continence isn’t affected during vaginal delivery.

Resources

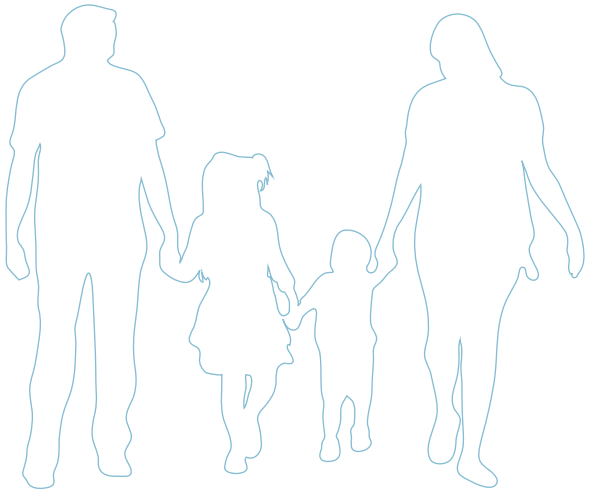
Information, education & support

At Boston Children's

Boston Children's Support Group for Children with Bladder Exstrophy and their Families

The support group was founded at Boston Children's in 1992. The group holds an event where children, adolescents, young adults and their families can come together, learn from each other and receive the kind of peer support that comes hand-in-hand with those who have shared similar experiences.

The Department of Urology at Boston Children's has pioneered efforts in the support and psychosocial development of patients with bladder exstrophy. The support group is a combined effort with our surgical programs, nursing, child life specialists, social workers and hospital volunteers.



We provide education and interaction among patients and families with:

- parent-to-parent networking
- guest speakers
- teen and young adult networking/activities
- medical play intervention using real or toy medical supplies to help children express feelings and gain understanding of medical experiences

The support group meets twice a year, is free for families and includes breakfast and lunch. Children have scheduled activities and meet in adjoining rooms relative to where the parents meet.

Support group topics include:

- complete primary repair of exstrophy
- clinical guidelines for care
- the development of the bladder exstrophy clinic
- urodynamics
- bladder neck reconstruction
- strategies for helping children to cope with bladder exstrophy
- catheterization
- updates on clinical research projects

A typical meeting may also include one or more of the following:

- guest speaker or panel discussion
- networking and group discussions
- group activities

During the meetings, children and teens actively participate in developmentally appropriate expressive and therapeutic activities to encourage peer relationships and normalization. Opportunities are provided for open discussions about emotions, illness and hospitalization.

Teen Chat

Recognizing the unique concerns and needs of patients with exstrophy, the center provides a monthly support dinner group meeting. Past chat topics have included transitioning to college, maximizing self-care and independence, navigating the challenges of relationships with family and friends, and psychosexual health and education.

Teen and Young Adult Picnic

Every year, the center provides an evening for all teens and young adults to gather together for sharing and activities. As the summer comes to a close and transitions are anticipated for the fall, the picnic provides a wonderful reminder that teens and young adults are members of a caring and supportive network.

Family-to-Family Program

Families and patients benefit from speaking to others receiving care in the Bladder Exstrophy Program. The nursing coordinator and social worker are able to match you with a family who is eager to listen and to answer any of your questions, which may include how they managed challenges associated with hospitalizations, surgeries, separation from family and navigating school.

Beyond our world-class bladder exstrophy care, Boston Children's Hospital offers numerous services that touch the lives of our patients and families. These services include:

Case Management Center

The case management team provides care coordination services, including discharge planning for post-hospitalization services. Our utilization management case managers support appropriate use of and access to health care resources at the hospital. This includes work with insurance companies to facilitate coverage of medically necessary care. The center's dedicated group of nurses and support staff has advanced degrees in specialty areas such as public health, nursing and business.

Chaplaincy

Our chaplaincy offers spiritual companions to children and families of all traditions and to those with no religious affiliation.

Child Life Services

Child life specialists possess expertise in child development and are available to help children and families cope with illness and hospitalization.

Hale Family Center for Families

This multi-service resource center offers a variety of programs designed to enhance your family's experience.

Interpreter Services

Boston Children's interpreter services arranges for interpreters in more than 35 languages to assist you at any point along the continuum of care.

Clown Care

The Big Apple Circus Clown Care is a community outreach program of the Big Apple Circus that brings the joy and excitement of classical circus clowning to the bedsides of hospitalized children five days a week, 50 weeks a year.

Pawprints

Pawprints is our therapy dog visitation program, which provides hospitalized children and their families a healthy diversion from the usual hospital routine and an opportunity for social interaction.

Social Work

At Boston Children's, clinical social workers and resource specialists are available in every area of the hospital to help you and your child or adolescent face a broad range of psychosocial issues and challenges.

For a comprehensive list or for more detailed information about these resources, visit bostonchildrens.org/familyresources.

Learn more about Boston Children's services and resources at bostonchildrens.org/familyresources

Additional Resources

National Resources

Association for Bladder Exstrophy Community
bladderexstrophy.com

Youth Rally
rally4youth.org

United Ostomy Associations of America, Inc.
ostomy.org

The Bedwetting Store
bedwettingstore.com

Massachusetts and Regional Resources *

Massachusetts Department of Public Health
Division for Children and Youth with Special Health Needs
mass.gov/dph/specialhealthneeds
800-882-1435, 617-624-6060

Connecticut Department of Public Health
Children and Youth with Special Health Care Needs
ct.gov/dph
800-505-7000
(Information Line to connect to appropriate state region)

Maine Department of Health and Human Services
Division of Population Health/Children with Special Health Needs
maine.gov
207-287-5357

New Hampshire Department of Health and Human Services
Special Medical Services
dhhs.nh.gov/DCBCS/BDS/sms/index.htm
603-271-4488

New York Department of Health
Children and Youth with Special Health Care Needs (CYSHCN) Program/Community, Family, and Minority Health/Special Health Care Needs
health.ny.gov
518-473-7016 or your local county health department

Vermont Department of Health
Children with Special Health Needs
healthvermont.gov/family/cshn/cshn.aspx
800-660-4427, 802-863-7338

**Please check your state public health department for resources should your family reside in other regions of the country or world.*

Thomas Vincent

Overcoming bladder exstrophy

On a warm June morning, a District of Columbia tour guide stops in front of the Korean War Memorial. Pointing out the 19 statues erected in tribute to soldiers who gave their lives in the conflict, he explains that the impressive seven-foot sculptures are meant to represent the 38th parallel, the demilitarized zone separating North and South Korea. When he turns to ask the group why a reference to the 38th parallel would only contain 19 statues, 16-year-old Thomas Vincent immediately offered a theory.

Thomas, who at the age of 16, was in the nation's capital to meet his senators and congressional representative as a patient representative of Boston Children's Hospital, has always been the portrait of an over-achieving student. When not busy with homework, chances are you could find Thomas practicing for any of the three sports he played for his school. If he's not on the field or the court, you might find him tutoring other students or at Spanish club. At first, second and even third glance, he is healthy and vital—the last person you might believe required extensive surgery only hours after he was born.

Dreary start to a bright future

When Thomas was born early Thanksgiving morning in 1996, the obstetrician and pediatrician-on-call at the Vincent's local hospital immediately knew something was wrong. "I just heard them say, 'We have a problem here' and they took him away," Thomas' mother Jean explains. "It was terrifying."

Looking at the newborn Thomas, doctors could see part of his bladder was exposed, and they soon diagnosed him with bladder exstrophy, a rare congenital condition where part of a newborn's bladder protrudes out of the body. (Or, as Thomas has gotten used to explaining, "it basically means my insides were on the outside.")

Although bladder exstrophy can sometimes be diagnosed in utero through an ultrasound, not all technicians know what to look for. So, when Thomas was delivered with his bladder exposed, his doctors were as surprised as his parents.

Within hours, a transport team whisked the newborn to Boston Children's—with his father following in his car—where Thomas would become only the second patient at Boston Children's to undergo a complete primary repair of exstrophy (CPRE).

Thomas 2013 (photo by Matt Parker)

The procedure—developed by former Boston Children's physician Michael Mitchell, MD, in 1989—not only placed Thomas's bladder back inside his abdomen, but also repaired anomalies to his urethra and reset his hips. By performing CPRE, Thomas' surgeons, W. Hardy Hendren, MD, and Joseph G. Borer, MD, were able to address all of these issues in a single procedure, rather than spreading the operation across three separate surgeries as other treatment methods require.

It took Hendren and Borer more than 12 hours to complete the CPRE, lasting from Thanksgiving afternoon to 4 a.m. the next morning. In the end, the procedure was a success. A day later, Jean was able to join her husband in Boston and set eyes on her son for the very first time. To give his organs time to heal, Thomas' doctors placed the newborn in traction, and it would be another three-and a-half weeks before Jean was able to hold her son.

> “Having a condition like this doesn’t mean a child can’t lead a full, active life. Thomas is living proof of that.”

No sign of slowing

Fast-forward to 2015, Thomas’ senior year in high school: you would never know he is managing with a congenital bladder condition. Like most bladder exstrophy patients, he’s had other operations, but none have slowed his drive or spirit. Some have been planned, including surgery to enlarge his bladder and install a stoma to help drain it. Others were unanticipated, including surgery to repair a bladder rupture from a sports injury.

Regardless of the time he’s spent in hospitals, Thomas has barely skipped a beat in his busy life. At school, he is a member of the National Honor Society, has played for his school’s basketball, soccer and tennis teams since his freshman year and was named captain of the soccer team last fall. He is also an intern/research assistant at a science and technology firm in Hampton, NH.

“When you throw in homework, it’s a lot,” he admits.

Thomas also isn’t shy about speaking up and sharing his experiences with other patients. When he first attended Youth Rally, a weeklong summer camp for teenagers with bladder and bowel conditions, he was so engaged with other campers he was mistaken for a counselor. This summer, Thomas will officially take on that leadership role. “Growing up, everyone says this is a rare condition, and a lot of kids think no one knows what they’re going through,” he says. “But Youth Rally’s motto is ‘you are not alone’, which is something that really resonates with me. I take it to heart.”

The missing 19 statues

Back in Washington, D.C., Thomas was sure that the answer had something to do with the soldiers’ reflections being visible in the memorial’s reflective wall, and when he voiced his answer, the tour guide confirmed his theory. Later that afternoon when Thomas spoke with Senators Kelly Ayotte and Jeanne Shaheen and Congresswoman Carol Shea-Porter, he was enthusiastic and engaged, displayed a seriousness of thought not normally associated with boys his age. Watching him in this scenario, it was easy to see why Boston Children’s invited Thomas and his parents to D.C. to represent the hospital at Family Advocacy Day, an annual event that brings families and legislators together to discuss health care reform.

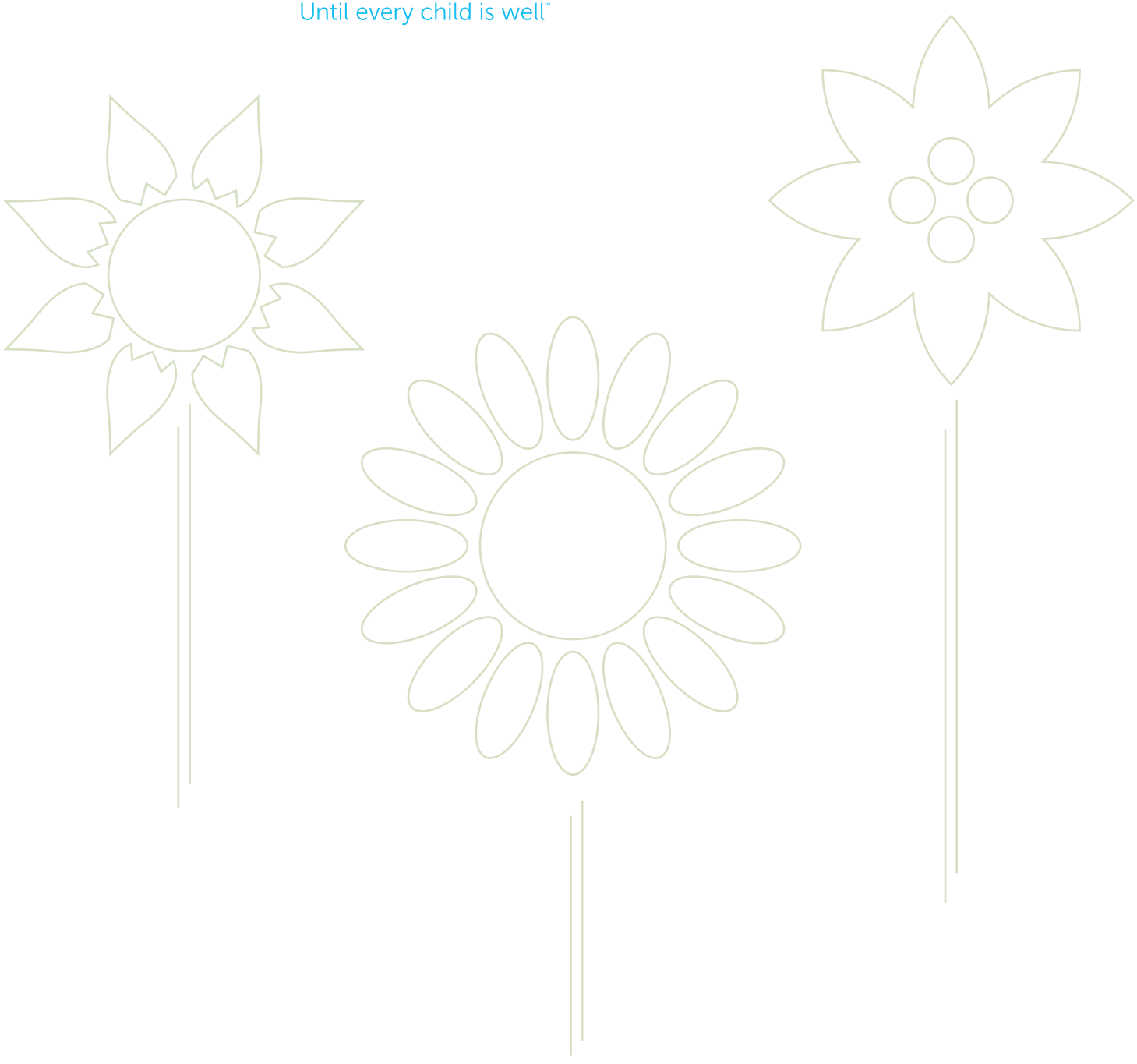
Looking to the future

Thomas will soon graduate from high school and will attend Duke University where he plans to study biomedical engineering and pursue a 5-year master’s degree or apply to medical school, or both, he says.

Though he may face more surgery in the future, given his track record, it’s unlikely to slow him down. As his father Maurice says, “There are different levels of severity for bladder exstrophy, but having a condition like this doesn’t mean a child can’t lead a full, active life. Thomas is living proof of that.”

Boston Children's Hospital
Until every child is well™

Bladder Exstrophy Program



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