Bladder Exstrophy: Patient Care and Surgeon Education

FROM THE GUEST EDITOR

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As the most recent fellow representatives to the Society of Pediatric Urology, we were asked to serve as guest editors for this edition of Dialogues. The topic of this edition of Dialogues is bladder exstrophy. As one of the most anatomically complex abnormalities seen in pediatric urology, many questions persist about the ideal timing of surgery. Additionally, given the rarity of the disease, fellow and surgeon education remains a challenge.

We queried our current fellowship colleagues about their experiences in exstrophy education and recommendations for improvements in this area. Additionally we received contributions from prominent pediatric urology faculty and support staff at multiple academic institutions. This edition discusses the approach for the initial closure: immediate vs. delayed, multi-institutional vs. single institution surgical teams; overall patient care concerns from the point of view of Dr. Barry Duel (a pediatric urologist with a personal history of bladder exstrophy) and Bladder Exstrophy Support Group staff at Boston Children’s Hospital; and approaches to additional continence surgeries in this challenging population.

Being guest editors for Dialogues has been a wonderful experience that has allowed us to delve deeper into a challenging diagnosis. We appreciate the efforts from our fellowship colleagues and academic mentors in composing this edition.

FROM THE EDITOR

Elizabeth B. Yerkes, MD

Thank you to the Guest Editors and Congratulations on pulling the experts together to build this broad look at one of our ongoing challenges in Pediatric Urology. Very well done!

Although I enjoyed each of the contributions, I found the patient perspectives from patient and colleague Dr. Barry Duel and the insights from the Boston exstrophy support group to be particularly pertinent to modern, comprehensive delivery of care.

Successful primary closure is the common theme in the Point-Counterpoint contributions. With all of the modifications and refinements continually occurring in Staged Repair and Complete Primary Repair camps, these once contrary podiums on the grand stage of Pediatric Urology have scooted much closer together during my short-ish career. This evolution has been refreshing and also somewhat amusing to watch. These modern efforts to restore anatomy and function may now be more alike than divergent.

The insights of the recent fellows and the experiences of the consortium and institutional reconstructive teams are a reminder of the value of mentorship and collaboration throughout one’s career. We can hope for, but are unlikely to develop, a cure for bladder exstrophy, or any of our other complex anomalies, and we should continue to aspire to make life as normal and confident as possible for our patients, working together to enhance anatomical, functional and psychosocial outcomes.
The Multi-Institutional Bladder Exstrophy Consortium (MIBEC)

We asked the MIBEC group, “How have you benefited from being part of a Multi-Institutional Collaboration? Is it worth the cost? What lessons have you learned about collaborating across multiple institutions?”

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Introduction

Bladder exstrophy (BE) is within the spectrum of the exstrophy-epispadias complex with epispadias and cloacal exstrophy. Bladder exstrophy is the most common of these three entities with an incidence of approximately 2.15/100,000 live births. The manifestations of BE vary in severity and pose major challenges to the affected individual. The most important goals in the care of a child with bladder exstrophy are to preserve normal kidney function, promote adequate bladder function, attain urinary continence, provide the most natural appearance and function of the external genitalia, and achieve developmental milestones such that the affected individual can thrive in all psychosocial parameters.

Because BE is rare, many surgeons will see only a few of these children in their career, making it difficult to develop the surgical expertise necessary to provide optimal outcome. In 2012, we had a vision to reduce the variability in surgical care of patients with bladder exstrophy. We felt it would be helpful to bring several pediatric urologists with specific interest and expertise in the care of patients with BE together in order to make a meaningful, impactful and lasting advancement in the care of these children. The use of a “coach” was also felt to be critical.

While many hospitals treat bladder exstrophy using a planned three-stage surgical approach, our preferred technique is the Complete Primary Repair of Exstrophy (CPRE); combining bladder closure, bladder neck reconstruction and epispadias repair in one stage. CPRE is a technically challenging procedure, and our vision included creating a team of pediatric urologists dedicated to the care of bladder exstrophy to refine the CPRE technique, streamline follow up care and report patient outcomes. This was the impetus for the Multi-Institutional Bladder Exstrophy Consortium (MIBEC). The MIBEC is a novel surgical collaborative that is focused on refinement of CPRE and protocol driven follow up.

Process

Three alternately hosting institutions constitute MIBEC: Children’s Hospital of Wisconsin, Children’s Hospital of Philadelphia, and Boston Children’s Hospital. Dr. Michael E. Mitchell has served as both coach and participating surgeon. For a potential event (1-2 day site visit) all three institutions reserved two consecutive operating room days per month approximately six months in advance for potential CPRE. If additional cases present, days/events were added as needed. Traveling to the host site and electronic transmission of intraoperative audio and video facilitated observation, commentary and critique by collaborating surgeons. Video recording of surgery was used for real-time observation, teaching, and future analysis, editing and reviewing.

Technical aspects included the use of a commercially available video-camcorder, camera holding bracket and stabilizing arm, intravenous pole, image converter, cable for video image transmission to operating room wall monitors and remote site(s). Information technology and/or media service teams at each institution partnered to coordinate transmission and viewing of events.

Early experience

The MIBEC surgical collaborative was formed in the fall of 2012 and the first procedure was performed in February 2013. Early experience of the MIBEC has been published. In the first three years of the MIBEC, collaborators have participated in CPRE surgery for 32 consecutive patients with BE. There were no cases of dehiscence in this group. However, early in the series, several girls had significant bladder outlet obstructive complications following CPRE. The rate of bladder outlet obstruction in girls was increased from published reports. We noted a low complication rate in the boys following CPRE that was comparable to reports in the literature, and we have also observed early signs of continence and spontaneous voiding in some boys and girls. It will take several years to measure these outcomes. Video record of each procedure is reviewed and studied per respective patient and outcome.

Cost

Value in medicine has been defined as outcome divided by cost. Therefore, in order to know the true value of our efforts, we needed to determine the cost expended. MIBEC participants traveled to the host institution for observation and coaching during CPRE. Costs included: start-up (video equipment), visitor travel and lodging, and opportun-

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MIBEC (continued from previous page)

We defined the opportunity cost (OC) per day as the revenue that would have been generated if the surgeon was not traveling, and opportunity cost was calculated by annual collections of the surgeon divided by annual clinical days worked. Total OC = OC per day multiplied by the number of work days spent in travel and on-site observation.

Complete cost data was available for 20 of the first 23 site visits. The total cost of collaboration was $364,642 over 24 months. This figure is the sum of $25,518 for the hosting sites, and $23,971 for the visitors, plus opportunity costs. On average, each operating event cost $16,534, with $15,007 of that being opportunity costs. The most valuable commodity was time, with total opportunity cost of $315,153. Ultimately each MIBEC institution invested a significant sum to execute this collaborative experience. In order to know whether this consortium and coaching experience has been a valuable investment, we need to identify and measure specific quantifiable outcomes. An early outcome of significant positive economic impact is the absence of dehiscence in our cohort.

Conclusions

This collaborative effort has continued to accelerate the physician knowledge base by focusing on refining CPRE technique ultimately to benefit and optimize patient care and outcome for this rare condition.

Refinement of CPRE technique will continue with this unique model of continuing surgical education, without endpoint, for the betterment of patient, teacher and pupil. Collaboration not only increases the number of cases, hence experience for every surgeon by active participation, but it also promotes regular follow up with group analysis and discussion of every case. This collaborative model can be transferred to other rare, complex surgical procedures to maximize and share collective expertise, standardize/refine technique, and analyze outcomes to ultimately benefit patient care and outcome.

Future

Future considerations include long-term outcomes of upper urinary tract dilation, vesicoureteral reflux, additional surgical procedures, renal scarring, urinary continence, sexual function, and overall cosmesis and quality of life. Several surveys including those assessing quality of life and continence outcome will also be implemented.

References


Figure 2A. Two weeks post-operative photo after CPRE. Note the suprapubic tube at the umbilicus.
Figure 2B. Three months post-operative photo after CPRE. Note the location of the meatus at the glans.
Figure 3. Right kidney on renal ultrasound without hydronephrosis one month post-operative CPRE.
Figure 4. Left kidney on renal ultrasound without hydronephrosis one month post-operative CPRE.
Figure 5. VCUG via suprapubic tube one month post-operative CPRE. Note the tapered bladder neck (dashed white arrow) and the patent urethra during voiding phase (solid white arrows).
Everything You Always Wanted to Know About Your Exstrophy Patients (But Were Afraid to Ask)

We asked Dr. Duel, “What should pediatric urologists know from a patient perspective?”

Barry Duel, MD, Associate Professor of Urology, Director of Pediatric Urology, Cardinal Glennon Children’s Hospital, Saint Louis University School of Medicine

To begin, we need to consciously acknowledge that we and our patients inhabit completely different worlds which only intersect intermittently. I, however, live at that intersection, as the only pediatric urologist to also have been born with bladder extrophy. I’ve never made a secret of this, but I’d never widely discussed it until recently, as I became increasingly annoyed with the purely technical discussions we have year after year at our meetings. To that end, I’ve begun speaking out more, to help my colleagues understand what the patients won’t tell you.

First, these children suffer in silence. They’re worried about humiliation because of incontinence. They’re worried about their belly scars. They’re worried about whether they’ll be able to have sex, or find stable adult relationships, or become parents. The list goes on and on. They and their families ALL need psychological support whether they know it or not. This might be a mental health professional, an extrophy support group, or simply a Facebook page, like ABEC (the Association of Bladder Exstrophy Communities). These kids look, and are, completely normal except for their genitourinary systems. That’s very different from our patients with myelomeningocele, for instance. Most people probably wouldn’t be surprised to find out that the child in the wheelchair is incontinent, but extrophy has no obvious manifestations, so the child and family can choose how much information they share. In most cases, they keep the diagnosis a secret, out of fear that disclosure will result in ridicule; a fear that’s probably well-founded. It’s embarrassing to see telethons, marathons or colored ribbons for children with urologic anomalies or cancers. That’s why we don’t ever discuss an anomaly that causes incontinence and abnormal genitalia, In the end, we can really do a better job of taking care of our extrophy patients (and all of our patients) by just taking a few moments and trying to understand what these children experience outside of our few contacts with them. Once we do that, we may very well find that we as surgeons aren’t the best people to provide the rest of the care they’re silently begging us for. That’s when we need to put our egos aside, and get them the help they need to grow up to become the resilient, content adults they can all become.
Preface

Early one Saturday morning in November, a new family entered the room with hesitation. This moment of uncertainty quickly subsided as they were embraced by a group of seasoned patients, families, and staff. A 10-year-old boy smiled after he caught the eye of a peer whom he had met at a summer event for our young adolescent patients. The exuberant family of five, who are refugees from Somalia, entered with their hospital interpreter. A young adult patient arrived with her mother. She had worn continence pads until the age of 15 when she finally gained the courage to proceed with conduit surgery. These are the faces of our Bladder Exstrophy Support Group.

Our team consisting of nurses, child life specialists, a social worker, our medical director, a fellow and volunteers mingled among the group, which would grow to over 30 patients and their families. A total of 80 people from five different regional states arrived for this support group which has been sponsored for the last 23 years by the Boston Children’s Hospital Department of Urology’s Bladder Exstrophy Program. This support group is held in November and April of each year and invites patients and their families treated along the epispadias-exstrophy complex spectrum, including epispadias, bladder exstrophy, cloacal exstrophy, cloacal malformation, and associated anomalies.

Diverse socioeconomic, racial, cultural, and linguistic differences melt away as the singular commonality of meeting the challenges of these diagnoses is front and center. One mother, withdrawn before attending group had commented, “This is an experience no family should be without.” An adolescent boy with growing coping skills remarked with wide eyes, “I can’t believe everyone here has bladder exstrophy.” Another girl living with an unclosed bladder for the first five years of her life in a refugee camp came to her first group shy and tentative, but in subsequent groups, she has taken the lead in medical play and mentoring new or younger members. A family of a baby girl attended group for the first time with visible tears. They were embraced by a father of a college student, offering the reassuring words, “We got through this and so will you.”

The Bladder Exstrophy Support Group at Boston Children’s Hospital has been established for over 20 years with a team that consists of a social worker, nurses, and Child Life specialists, and families. We asked them, “What are the needs of patients/parents/families and what can you offer? What are some of the long term emotional or psychological effects that you see?”

Diane Price, MSW, Rosemary Grant, RN, BSN, Theresa Rankin, MS, CCLS, Laurel Anderson, MS, CCLS
Boston Children’s Hospital

Related Activities

Additional opportunities for peer support currently include an annual pre-adolescent event for boys and a summer picnic for our teens and young adults. A monthly evening Teen and Young Adult Chat was held for four years and is anticipated to be revitalized. Many of our pre-adolescent male patients have benefitted from making healthy connections with experienced patients as they consider and prepare for continence surgery including bladder neck reconstruction and creation of a continent urinary stoma.

Patients and families have also participated in related hospital programs including Family to Family, Teen Advisory Board, and department web casts. Our families are able to convey their experience,
Support Group Services (continued from previous page)

strength, and resilience to other families as well as to the medical team through these unique opportunities.

Lessons Learned/Next Steps

Our evaluations indicate high patient and family satisfaction with our support group activities, reflecting the transformative value of peer support, strong partnerships with providers, increased knowledge of medical, nursing, and psychosocial care, as well as development of stronger advocacy skills. Moreover, support group provides patients with a unique hospital experience that is removed from the ‘bedside’, providing comfort and connections with peers and, thus, strengthening coping and adjustment as challenges are met along the continuum of care.

However, our support group activities capture a small portion of our overall population of local, regional, national, and international patients. A critical challenge is determining how this quality of peer support and psychoeducation can be a universal standard of care. Under consideration is utilization of supervised Skype Family to Family outreach, moderation of teen and parent on-line chats, and enriched resources on our hospital webpage.

In Conclusion

The goals of any bladder extrophy program include the preservation of bladder and kidney health, successful surgical intervention, optimal medical and cosmetic outcomes, as well as achievement of milestones along the developmental spectrum. Our support group activities have provided patients and families ongoing valuable peer support and education. Likewise, the program has offered providers many opportunities to learn from patients and their families, resulting in richer partnerships. The strength of these relationships is particularly recognized during challenging junctures in care. We recommend consideration to include regular patient and family group support as one effective intervention to further achieve overall goals of the healthcare experience.

Support Group Services (continued from previous page)

“...patients have benefited from making healthy connections with experienced patients as they consider and prepare for continence surgery including bladder neck reconstruction and creation of a continent urinary stoma.”

POINT/COUNTERPOINT ON PRIMARY SURGICAL MANAGEMENT OF EXSTROPHY

Complete Primary Repair of Exstrophy (CPRE)

Point/Counterpoint:

We asked Dr. Mitchell, “What are the advantages to the Complete Primary Repair of Exstrophy (CPRE) as opposed to the Modern Staged Repair of Exstrophy (MSRE)?”

Michael E. Mitchell, MD, Professor of Urology, Children’s Hospital of Wisconsin, Medical College of Wisconsin

Exstrophy represents a unique reproducible spectrum of congenital deformation that is potentially ideally suited to surgical repair. As no specific gene has been associated with exstrophy to date, and approximately 50% of identical twins with exstrophy are discordant, the mechanism may be epigenetic. The problem seems to relate to premature loss of the infra-umbilical membrane which normally serves as a matrix for lower abdominal and pelvic development that results in ventral herniation of developing structures (i.e. colon, hindgut, lower urinary tract, genitalia). The exstrophy spectrum therefore is dictated by the timing in gestation of the event (i.e. 2-3 weeks cloacal exstrophy, 4-16 weeks complete exstrophy, 16+ weeks epispadias) (Fig. 1). Successful surgical repair is based on the assumption that restoring anatomy will facilitate normalization of functional physiology, and I am convinced that complete primary repair of exstrophy, CPRE, is the most logical and effective route to this end.

The exstrophic bladder is abnormal at birth, presumably because it has not had the appropriate stimulus for development (filling and emptying from week 20 of gestation). Fortunately, the potential for normalization seems to be present in the first few months after birth. Perhaps somewhat analogous is the observation that early valve ablation can correct the abnormal bladder of young patients with urethral valves. However, if ablation is delayed these bladder changes may not occur. Early establishment of cycling stimulus (dynamic stimulus) facilitates appropriate function. This is not a new concept, as John Lattimer, over 50 years ago, was committed to complete repair of exstrophy. Unfortunately, he was perhaps not aggressive enough in his dissection of the pelvic outlet and did not have intermittent catheterization as a tool to protect his patients in the early stages of high pressure voiding and obstruction after repair. As a result, some of patients developed hydronephrosis and renal failure. The reaction to this unacceptable result was to revert to primarily diversion of the exstrophy patients, or, in the case of a few brave souls such as Bob Jeffs, to conservatively correct exstrophy in stages. The latter approach reduced the chance of irreversible obstruction and delayed continence procedures. The prob-

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In reality the truth may be somewhere in between. Because of the spectrum of exstrophy (Fig 1), the potential for normal bladder function may depend, in part, on the timing of the causal event (severity) and relative potential for normalization of function. Our inclination is to get the bladder to cycle as soon as possible (first few months of life). However, even if this is achieved, it may be the inherent spectrum of the problem that ultimately dictates potential for bladder function; cloacal patients with lowest potential, the more typical classic exstrophy patients with fair to good potential, and penopubic epispadias patients with the best potential for functional (and cosmetic) results.

Timing of CPRE is potentially critical. We are presently trying to determine the optimal timing of CPRE through the Multi-Institutional Bladder Exstrophy Consortium, MIBEC (CHW, BCH, CHOP). Currently at the Children’s Hospital of Wisconsin, CPRE is performed in the first 3 months of life with bilateral ilium osteotomies and spica casting with average hospital stay of 7 days. In our reported Seattle CPRE experience 74% of the patients were voiding with continence, but only 29% achieved this with only one surgery. However, only one patient of 23 consecutive patients required augmentation to achieve dryness. We interpret this result as an indication that there is potential for the abnormal exstrophic bladder at birth to change with primary complete repair. These results may reflect in part the spectral nature of the problem.

The MIBEC initiative seems to be an important mechanism to: (a) expand surgical experience and cooperation through real time participation, (b) closely monitor, review and improve the CPRE technique, and (c) standardize and carefully study a surgical procedure and its consequence. Thus, by organizing, classifying and systematizing our knowledge about the details and specifics of the surgical repair of this very unique malformation we hope to maximize patient benefit. Is it possible to achieve, with one operation in the first few months of life, a genuine surgical cure such that a patient cannot appreciate that he or she ever had a problem?

References
Modern Staged Repair of Exstrophy (MSRE)

Point/Counterpoint:

We asked Drs. Gearhart and Di Carlo, “What are the advantages to the Modern Staged Repair of Exstrophy (MSRE) as opposed to the Complete Primary Repair of Exstrophy (CPRE)?”

The exstrophy-epispadias complex (EEC) is a rare spectrum of birth defects involving the genitourinary and gastrointestinal tracts, musculoskeletal system, pelvic floor, and bony pelvis. In classic bladder exstrophy (CBE), there is a lower abdominal wall defect exposing an open bladder and urethra, a wide diastasis of the pubic symphysis, and an epispadic urethral opening. These patients will undergo multiple reconstructive surgical procedures beginning with closure of the urethra (up to the midshaft in males), bladder and anterior abdominal wall, along with approximation of the pubic rami. The modern staged repair of exstrophy (MSRE) allows for a secure abdominal closure, reconstruction of functional and cosmetic genitalia, and urinary continence with preservation of renal function. Successful primary closure is of utmost importance as it is associated with improved bladder growth, imparts the greatest chance for eventual continence, decreased overall costs and decreased inflammation and fibrosis of the bladder.

MSRE includes a staged approach to this complex genitourinary reconstructive endeavor, starting with closure of the bladder and abdominal wall. Urethroplasty and feminizing genitoplasty is performed at the time of initial closure in females, while repair of the epispadias (which is essentially a midshaft epispadias repair) in males is typically performed at six months after primary closure. In select patients, bladder closure and epispadias repair can be combined. Bladder outlet surgery for continence is performed when an adequate bladder capacity is achieved and the patient is ready to be dry.

The principal behind the complete primary repair of exstrophy (CPRE) is that all components of exstrophy reconstruction be performed at one time – bladder closure with outlet procedure and genitoplasty. Numerous recent reports, however, have noted that continence is not achieved in the majority of patients even with a successful primary closure utilizing the “CPRE” technique without additional surgical procedures such as bladder neck reconstruction. Additionally, other necessary surgical procedures after CPRE, such as ureteral reimplantation to treat symptomatic vesicoureteral reflux as well as hypospadias repair to treat a hypospadiac urethral meatus after CPRE, demonstrate that this is approach is truly not a “complete repair.”

Besides a successful primary closure ensuring the greatest chance for eventual continence in this very unique and rare patient population, safety is of paramount concern. Numerous cases of penile loss (corporal body, glans, penile skin etc.) have been reported in the literature, necessitating further major reconstructive surgery to repair this devastating complication.

The timing of the primary closure is still in debate. Proponents of early bladder closure (closure during the first 72 hours of life) argue that prompt closure allows for sooner bladder cycling and improved bladder expansion. In addition, closure while the pelvis is still malleable may allow adequate reduction of the pelvic diastasis without the need for osteotomy. This is the authors’ approach when the bladder template is adequate for closure, as the first step in MSRE. Some studies have suggested that bladders that remain exstrophic for long periods of time are more likely to undergo pre-cancerous changes. Delaying bladder closure may be helpful in patients born with small bladder templates allowing the bladder template to develop prior to closure and thus increasing the likelihood of post-closure bladder growth. Additionally, delaying bladder closure may allow epispadias repair to be performed concomitantly rather than as an additional procedure.

Successful primary closure of exstrophy is crucial for eventual continence and optimal quality of life. MSRE allows for a carefully thought-out, safe, and stepwise approach to this complex reconstructive adventure, ensuring the best possible outcomes. More data is showing us that CPRE is actually not a complete solution to this rare congenital anomaly.
Searching for Continence In Bladder Exstrophy…

We asked Drs. Cain & Rink, “What procedures do bladder exstrophy patients need after initial closure (i.e. incontinence procedures, augments, reimplants, etc)? What is/are your preferred method(s)?”

Mark P. Cain, MD, FAAP, Professor and Chief, Pediatric Urology, Riley Hospital for Children at IU Health
Richard C. Rink, MD, FAAP, FACS, Robert A. Garret Professor of Urology, Pediatric Urology
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Despite many advances in Pediatric Urology over the last 40+ years, primary bladder exstrophy still remains one of the more difficult and devastating urologic birth defects. This stems primarily from the actual defect, which encompasses the bladder, sphincter mechanisms, abdominal wall, and pelvic bones to a varying degree in what is usually an otherwise “normal” newborn. We find this disease to be a continuous struggle in an effort to not only restore normal anatomy, but also more importantly create normal function: urinary continence with volitional voiding. We want to achieve this with a minimal number of surgical procedures while maintaining normal upper urinary tracts.

We have been asked by the editors to comment specifically about our experience regarding additional procedures for continence after initial closure, but since the initial operation frequently drives the results of future procedures, we will start with our approach to the primary closure. Following the results of the successful inter-hospital multidisciplinary team concept, we now view this as a non-urgent procedure after delivery, and have committed to a 2-surgeon team at our institution, usually at 4 - 8 weeks of life. Our approach, if the anatomy allows, leans toward that described by Mitchell et al, with a one-stage complete repair with pelvic osteotomies. Similar to many other so-called “one-stage repairs” in urologic surgery, further operations may certainly be required.

We find that one of the most difficult challenges is assessing the best timing for a further continence procedure. This decision is based on multiple factors – family and patient interest and resources, bladder volume and compliance based on videourodynamic studies, and patient age/maturity. The critical discussion with the family must include the potential need for intermittent catheterization and also the potential need for bladder augmentation if there is bladder deterioration after achieving adequate bladder outlet resistance. This latter is issue unfortunately is somewhat unpredictable. Our usual target age is between 4-6 years old, with the goal of achieving social continence by the time the child starts formal school.

We believe that preoperative evaluation should include: evaluation of baseline renal function with serum electrolytes and creatinine, nuclear renal scan and ultrasound; urine culture; formal videourodynamic to evaluate bladder volume, compliance, and presence of reflux; and patient/family participation in a pre-surgical evaluation clinic with the Surgeon and Nurse Practitioner, nursing staff, and any other perioperative services that would be included in the patient’s care. During this visit the postoperative expectations should be clearly defined, including the need for frequent office visits, and the potential long-term need and technique for catheterization to empty the bladder. In the situation of an extremely small bladder where augmentation may be required we have transitioned to an outpatient bowel preparation, and the family is instructed in the protocol during this visit.

Patients are admitted to the hospital the day of surgery. In addition to general anesthesia, we will routinely place an epidural catheter for postoperative pain management, since this patient group tends to have more significant and prolonged bladder discomfort. When possible our surgical approach is to use the Mitchell modification of the Young-Dees-Leadbetter (YDL) procedure to tubularize the bladder outlet while minimizing the reduction in bladder capacity.1 This is accomplished over an 8 Fr catheter, with closure in two layers. The bladder neck tubularization should be at least 2.5 cm, and we will sometimes augment the repair with a fascial or SIS wrap/sling, especially for redo repairs. We no longer use the formal YDL procedure with its triangular flaps and vest over pants closure. The goal being to provide a more funneled functional bladder neck reconstruction rather than any sudden caliber change or angulation. The ureters are usually moved caudally and reimplanted using a cross-trigonal technique and ureteral stents are left in place for 7-14 days until bladder edema has decreased. The bladder base is frequently smaller than usual, and we will accept a slightly shorter tunnel of 1.5-2 cm for the nondilated ureter to prevent advancement of the ureter onto the lateral bladder wall. In addition to this standard approach to bladder neck reconstruction and ureteral reimplant, we have commonly also created a Mitrofanoff channel using the appendix at the time of bladder neck reconstruction, especially in males with exstrophy. Our reasoning for this is that in spite of years of experience, assessing the exact degree of surgical narrowing or determining who will void to completion following this surgery is at best an in-exact science. Our results with this bladder neck reconstruction have frequently led to the need for short and possibly long-term urethral catheterization. We have found this to be difficult for our families to perform after bladder neck surgery and for those sensitive exstrophy patients to tolerate. This group seems to have an almost hypersensitive urethra and bladder neck. We have not found long term suprapubic catheter drainage to be well tolerated during this difficult time of learning to void. We believe appendicovesicostomy to be a much better tolerated option for both parent and child, particularly if long term CIC is required.

The patients will usually require a 7-8 day hospital stay, and are not discharged until the family is comfortable with home care of the postoperative drainage tubes, including a visit from the outpatient nurs-

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Dialogues in Pediatric Urology  

Searching for Continence (continued from previous page)

“In summary, the advances in pediatric urology during our lengthy careers have been nothing short of amazing but reconstructing a functional bladder neck in a child born with such significant anatomic abnormalities as we see in bladder exstrophy remains a quite significant challenge.”

If a small urethral catheter was left in place after surgery it is removed prior to discharge. The Mitrofanoff catheter, or suprapubic tube is left in place for bladder drainage for 3 - 4 weeks, then clamped to check for residual urine as the patient learns to void through their reconstructed bladder neck / urethra. If the patient has difficulty voiding per urethra, they are taken back to the operating room for outpatient cystoscopy and gentle urethral dilation after 6-8 weeks. Renal ultrasound is performed approximately one month after removal of ureteral stents, and is repeated in 2-3 months intervals after the patient begins voiding. We have been fairly liberal in using antimuscarinics for bladder relaxation for the first 3-4 months or longer, as our experience has been that the patients have significant bladder irritability until all the drainage tubes have been removed and the bladder has healed. If one is to undertake this surgery, it is imperative to have outstanding pediatric urologic nursing with expertise in both the care of exstrophy patients and in voiding dynamics to assist the child and parent through the arduous postoperative voiding trials.

Unfortunately, in the long term our personal results with achieving continence that the patient, family and we are satisfied with after isolated bladder neck repair have been disappointing. Additional procedures at the previously surgically altered bladder neck become even more difficult due to significant scarring. We find it unlikely to be able to obtain normal voiding with dryness in re-do bladder neck surgery. Unless the bladder has achieved a functionally normal volume with normal compliance after bladder neck reconstruction, we will offer these families the option of bladder augmentation earlier to achieve urinary continence in an attempt to minimize the overall number of additional procedures.

The management of the child with a very small capacity bladder that does not appear to enlarge as the child ages is also a challenge. The injection of bulking agents to achieve some increased outlet resistance prior to formal bladder neck reconstruction has been tried with variable results. In rare case of the extremely small bladder, we will offer augmentation at the same time as the bladder neck procedure. Patients with very small bladders or multiple prior bladder operations are at much higher risk of eventually needing augmentation, and also at much higher risk of upper tract dilation without it. We will usually wait until the patient is older (>6-7 years), before committing to bladder augmentation, and will exhaust all reasonable medical and less invasive surgical options.

In summary, the advances in pediatric urology during our lengthy careers have been nothing short of amazing but reconstructing a functional bladder neck in a child born with such significant anatomic abnormalities as we see in bladder exstrophy remains a quite significant challenge. Unfortunately this surgery, even in 2016, will frequently result in disappointing results for the child, family and surgeon.

References
Bladder Exstrophy Education: A Survey of Current Pediatric Fellows

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Bladder exstrophy education is hampered by the relative rarity of the disease in the population. In fact, the ACGME does not have a minimum required case number for graduating pediatric urology fellows but instead only tracks the number of cases. We wondered how many exstrophy cases current pediatric fellows were participating in and in what capacity. Additionally, we wondered how fellows perceived multi-institutional exstrophy collaborations and the possible effect on fellow education. We surveyed current pediatric urology fellows about these topics and received 28 responses (25% 1st year clinical fellow, 14% 1st year research fellow, 18% 2nd year research fellow, 43% 2nd year research fellow).

The fellows reported participation in a good number of general exstrophy/epispadias cases (including bladder exstrophy closures, cecal exstrophy closures, epispadias repairs and subsequent reconstruction surgeries on exstrophy patients): 64% 2-5 cases, 29% >5 cases, 4% no cases (but only performed research so far) and 4% no cases (and completed some or all of the clinical training). However, less fellows had experience in primary bladder exstrophy closures: 18% 1 case, 36% 2-5 cases, 11% >5 cases, 21% no cases (but only performed research so far) and 14% no cases (and completed some or all of the clinical training).

Fellows had various levels of participation in initial bladder exstrophy closures (fellows were able to choose more than one choice): 39% as 1st assistant with one attending, 36% as 2nd assistant with two attendings, 4% as 3rd or more assistant with multiple attendings, 25% as observation in person, 11% as observation via video and 11% have not participated in any form. In their future practices, 61% of fellows reported they plan on performing bladder exstrophy closures and 11% report that at the end of fellowship they will feel comfortable performing bladder exstrophy closures alone, 72% will feel comfortable with the assistance of another attending, and 18% will not feel comfortable performing closures.

Current fellows were equally split on whether multi-institutional exstrophy collaborations helped or hurt fellow training; however, there was universal agreement that collaborations would improve on surgical technique and overall patient care. One respondent stated: “I definitely think it hurts fellow training but I think in this rare circumstance, that is OK. There just aren’t enough cases at each individual institution (excluding Hopkins?) for all the trainees in the country to become competent. I think pretending to be competent with limited exposure is not right for our patients. The multi-institutional collaboration is a good idea but I think it proves that even at the attending level, collaboration is needed. Maybe a few programs should be designated for exstrophy closure training.” Another stated: “Overall, I think centralizing management of these complex, rare patients is a good thing secondary to centralizing experience and volume. However, there is no question that this would ultimately hurt fellow training.” Finally: “It would help low volume centers and bolster overall fellowship training. As my program sees many exstrophy patients, I would be excited to have my colleagues come operate with me on exstrophies.”

We then asked fellows their suggestions for improving exstrophy education for fellows. One fellow stated: “The single thing that would improve education is increased number of cases. I personally would not like to have this occur because it would mean more kids born with exstrophy. I’m glad it is rare but the rarity means most fellows don’t get to see or do enough of these cases.” A common theme was increased exposure for fellows. This may be through additional hands-on exposure by fellow rotations through collaborating institutions or international experiences. Alternatively, for fellows that are already observing procedures, they requested increased hands-on participation; for example, the second participating attending could act as a 2nd assistant and direct the course of the surgery in a more hands-off manner. Additionally, fellows suggested the development of educational videos, such as live-feed videos from the multi-institutional collaborations or an organized course sponsored by the multi-institutional collaborative. One respondent stated: “I think the rarity of exstrophy necessitates organized instruction to all fellows from experts with high volume as part of an organized course. Perhaps this could be one goal of the consortium.” Alternatively, some suggested that fellows who want to do exstrophy should train at a center involved in a collaboration or with a large exstrophy population; in the future, additional or specialized training could allow for a designation of “exstrophy trained” pediatric urologist. Universally, fellows requested additional discussion of surgical planning, techniques, and options prior to entering the operating room.

Overall, fellows are excited to participate in bladder exstrophy closures but yearn for more involvement in cases and organized training modules. Hopefully, the multi-institutional collaborations can yield fruitful results in both surgical outcomes and fellow education.