Current Urology Reports (2019) 20: 48
https://doi.org/10.1007/s11934-019-0916-2

NEW IMAGING TECHNIQUES (S RAIS-BAHRAMI AND K PORTER, SECTION EDITORS)

Anatomy of Classic Bladder Exstrophy: MRI Findings and Surgical Correlation

Emily A. Dunn[1] · Matthew Kasprenski[2] · James Facciola[3] · Karl Benz[2] · Mahir Maruf[2] · Mohammad H. Zaman[2] · John Gearhart[2] · Heather Di Carlo[2] · Aylin Tekes[1]

Published online: 8 July 2019

Abstract

Purpose of Review The exstrophy-epispadias complex (EEC) represents a group of congenitally acquired malformations involving the musculoskeletal, gastrointestinal, and genitourinary systems. Classic bladder exstrophy (CBE) is the most common and best studied entity within the EEC. In this review, imaging features of CBE anatomy will be presented with surgical correlation.

Recent Findings Magnetic resonance imaging (MRI) has emerged as a useful modality for pre- and postnatal assessment of the abdominal wall, pelvic floor, and gastrointestinal and genitourinary systems of children with CBE. The authors' experience supports use of preoperative MRI, in conjunction with navigational software, as a method for identifying complex CBE anatomy.

Summary Imaging facilitates surgical approach and improves visualization of complex anatomy, potentially helping to avoid complications. Continued investigation of imaging guidance in CBE repair is needed as surgical techniques improve.

Keywords Classic bladder exstrophy · Exstrophy-epispadias complex · Magnetic resonance imaging · Anatomy · Surgical repair · Imaging guidance

Introduction

Classic bladder exstrophy (CBE) falls on the spectrum of the exstrophy-epispadias complex (EEC), a group of congenitally acquired malformations involving the musculoskeletal, gastrointestinal, and genitourinary systems. CBE is reported in approximately 3.3 per 100,000 live births. CBE is the best studied and most common entity within the EEC, comprising more than half of all cases, and it will be the focus of this review [1].

Deformities of the genitourinary system as seen in CBE are severe and, without proper treatment, can lead to urinary incontinence, renal failure, and sexual dysfunction. Surgical reconstruction of the bony pelvis, bladder, and urethra is necessary to achieve urinary continence and satisfactory cosmesis. Genital reconstruction may enhance sexual and reproductive function and is typically performed after repair of bladder and musculoskeletal defects. An understanding of genitourinary, hindgut, and musculoskeletal anatomy and development is important for diagnosis of CBE and operative planning. Plain film radiography (XR), computed tomography (CT), and magnetic resonance imaging (MRI) are modalities most commonly utilized for imaging CBE before and after surgical repair. At the Johns Hopkins Children's Center, MRI is the cross-sectional modality of choice in the initial assessment of CBE defects. MRI is preferred over CT, given its superior contrast resolution for soft tissues and use of non-ionizing radiation. In this review, we present imaging features of CBE anatomy, with an emphasis on MRI findings, and their surgical correlation.

This article is part of the Topical Collection on *New Imaging Techniques*

✉ Emily A. Dunn
edunn18@jhmi.edu

[1] Division of Pediatric Radiology, Russell H. Morgan Department of Radiology and Radiological Science, Johns Hopkins University School of Medicine, Baltimore, MD, USA

[2] Robert D. Jeffs Division of Pediatric Urology, James Buchanan Brady Urological Institutions, Charlotte Bloomberg Children's Hospital, Johns Hopkins Hospital, Baltimore, MD, USA

[3] Russell H. Morgan Department of Radiology and Radiological Science, Johns Hopkins University School of Medicine, Baltimore, MD, USA

Brief Embryology

Exstrophy-Epispadias Complex

EEC is thought to arise from disturbances in development occurring as early as the 4th week of gestation, during which the lower abdominal wall and pelvic bones form and the cloaca is divided. In normal development, mesodermal mesenchyme migrates toward and is thought to reinforce the cloacal membrane, which ultimately perforates giving rise to a separate urogenital system and hindgut. Additionally, migration of mesenchyme results in formation of the medial portions of the anterior abdominal wall musculature, the anterior bladder wall, the pubic symphysis, and precursors of the external genitalia [2, 3].

While many theories seek to explain the developmental origins of EEC and a related defect, cloacal exstrophy (CE), they are still incompletely understood; failure of normal mesenchymal migration as well as rupture of the malformed cloacal membrane is likely involved. The timing of the abnormal cloacal membrane's rupture is thought to determine the malformation's severity, with epispadias and CE representing the least and most severe manifestations, respectively. CBE, which is of intermediate severity, results from rupture of the cloacal membrane after the urogenital system and hindgut have separated [2–4].

After the 4th week of gestation, mesenchymal migration and folding gives rise to the genital tubercle, urogenital folds, and labioscrotal folds which are precursors for the external genitalia in both sexes. Sex differentiation begins at week 7 and is complete by week 17. Prior to this stage, developing male and female external genitalia are morphologically indistinct [5].

In males, the SRY gene, the sex-determining region of the Y chromosome, initiates development of the testicles and stimulates production of androgens [6]. Fetal androgens induce development of the male sexual ducts, the prostate, urethra, and penis. The urogenital folds fuse and the genital tubercle elongates forming the corporal tissue and glans. The labioscrotal folds fuse to form the scrotum [7, 8].

In the absence of a Y chromosome and SRY gene, default female genitalia are formed. The genital tubercle does not elongate and instead deviates inferiorly, giving rise to the clitoris. The urogenital folds and labioscrotal folds remain unfused and give rise to the labia minora and labia majora, respectively [7, 8]. Genital defects observed in EEC and CE range from epispadias to bifid penile/clitoral and scrotal/labial halves.

Imaging Assessment of CBE Anomalies

In EEC, disruption of normal genitourinary and musculoskeletal development at certain stages is thought to produce anomalies with varying severity. CBE is characterized by deformities of the urinary system, anterior abdominal wall, pelvic bones, pelvic floor musculature, rectum, anus, and genitalia, which result in an "open book" configuration of the pelvis.

Imaging non-invasively identifies deformities not readily apparent on physical exam and may help to distinguish CBE from CE and exstrophy variants. Plain film XR and CT are typically used to assess deformities of the bony pelvis. Some centers utilize low-dose CT with field of view limited to the pelvis in order to limit radiation exposure. MRI is well-suited to evaluate CBE anomalies due to its multiplanar capability and, in the authors' institution, is preferred over CT given its superior contrast resolution, especially for soft tissues. Due to increasing awareness of effects of ionizing radiation from medical imaging, MRI has emerged as a useful modality to assess the anatomy of children with CBE. Hence, in the authors' institution, children with CBE are evaluated with MRI of the abdomen and pelvis prior to closure and reconstruction.

MRI Protocol

In the authors' institution, the examination is performed on a 1.5-T system (Avanto, Siemens, Erlangen, Germany). The field of view extends from the superior aspect of the kidneys through the distal-most portion of the genitalia and from anterior to posterior skin surfaces. The standard imaging protocol for evaluation of CBE includes axial, sagittal, and coronal T1- and T2-weighted 2D turbo spin echo sequences, obtained without fat saturation. The addition of a 3D T2-weighted turbo spin echo sequence provides high-resolution, isotropic images which may be reconstructed in multiple planes. Together, these sequences provide detailed information regarding complex CBE anatomy. Typically, exams are performed under general anesthesia in order to reduce motion and ensure patient safety. Moderate sedation with rectal midazolam and swaddling are also employed when appropriate.

Musculoskeletal Defects

Plain film XR demonstrates characteristic bony pelvis external rotation and pubic symphyseal diastasis, and therefore is among the first examinations performed in the early assessment of children with CBE and CE. Early studies utilizing CT further defined musculoskeletal anatomy in CBE. Sponseller and colleagues described the configuration and dimensions of the CBE bony pelvis compared with age-matched controls, including pubic bone diastasis, shortening of the pubic rami, external rotation of pelvic bones, and convergence of the iliac wings. Coronal rotation of the sacroiliac joints and retroversion of the acetabula and femurs were also reported [9]. Stec and colleagues studied 3D models of the pelvic bones derived from CT in CBE children prior to closure. Compared with age- and sex-matched controls, CBE children had greater

coronal rotation of the sacroiliac joints, greater outward rotation of the iliac wings, and more inferior pelvic rotation [10].

Stec and colleagues similarly assessed 3D models of the pelvic floor musculature constructed from CT of children with CBE. External rotation of the pelvic bones alters the configuration of the pelvic floor. In CBE, the levator ani was found to be more externally rotated, wider, more posteriorly distributed (relative to the rectum), and consequently more flattened compared with age- and sex-matched controls [11, 12].

High-resolution T1- and T2-weighted MRI sequences accurately depict musculature, soft tissues, and the ossifying pelvis (Fig. 1) of growing children. Consequently, use of MRI in the assessment of CBE has increased in recent years. Several studies have used MRI to describe the pelvic floor of CBE patients. Williams and colleagues studied the pelvic floor of male CBE infants with MRI. They generated a 3D model of the levator ani musculature and demonstrated its irregular contour with a central kink and loss of the typical "dome" configuration [13]. Tekes and colleagues analyzed the CBE pelvic floor with MRI and depicted individual muscular components of the levator ani using a 3D visualization system [14].

MRI has been used to assess functional and anatomical changes in CBE patients after surgery: Stec and colleagues demonstrated reshaping and internal rotation of the bony pelvis and anterior redistribution of a portion of pelvic floor musculature following primary closure [15]. Gargollo and colleagues observed that pelvic anatomy of children following complete primary repair of exstrophy (CPRE) closely resembled that of age- and gender-matched controls. They also showed that children older than 3 years who had continent intervals lasting more than 3 h had pelvic anatomy which appeared even more similar to controls [16]. Halachmi and colleagues assessed CBE male children after neonatal single-stage repair with osteotomies and found that levator ani and obturator internus configurations were overall similar to those in age-matched controls [17].

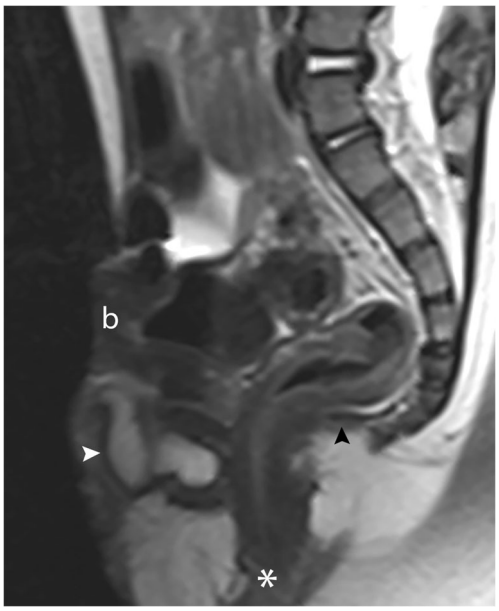

Fig. 1 MRI assessment of the CBE male. Sagittal T2-weighted MRI at the level of the pelvic midline. The bladder template (b) is exposed. Pelvic floor musculature (black arrowhead), which is depicted as a thin layer of soft tissue with low T2 signal intensity, is flattened. There is anterior displacement of the rectum and anus (asterisk). Paired corpora cavernosa are identified by their high T2 signal. The phallus in males is shortened due to diastasis of the pubic symphysis, congenital deficiency of the anterior corpora, and dorsal chordee (white arrowhead)

Urinary Defects

In CBE, the urinary bladder is everted and externalized. The urethral plate is open and extends along the dorsum of the penis from the bladder to the glanular groove [18]. The bladder and posterior urethra occupy an infraumbilical abdominal wall defect and are usually visualized on physical exam without difficulty. Verification of bladder template size may determine the timing of bladder closure [19]. In the authors' experience, cross-sectional imaging is often helpful for identification of portions of the bladder and urethra templates which may be obstructed by adjacent soft tissue or herniated intestines. Fluid-sensitive MRI sequences may be used to identify abnormal ureteral course and ureterovesical junctions, which contribute to vesicoureteral reflux in CBE.

Isolated renal anomalies, unrelated to the EEC, occur in 2.8% of children born with CBE and include duplication of the collecting system, absent or hypoplastic kidney, pelvic kidney, multicystic dysplastic kidney, and ureteropelvic junction obstruction. Identification of an abnormally located kidney or ureter could result in a different surgical approach [20]. Imaging may be used to identify associated malformations of the whole urinary tract prior to surgery.

Gastrointestinal Defects

In CBE, the anus is typically displaced anteriorly. As patients age, divergence of the levator ani and puborectalis muscles contributes to anal incontinence and rectal prolapse. Prenatal MRI can distinguish CBE from CE, by assessing for the presence or absence of severe anorectal, spinal, and urogenital malformations [21]. Isolated colorectal malformations, unrelated to CE and exstrophy variants, occur in 1.8% of children born with CBE and include imperforate anus, rectal stenosis, and congenital rectal prolapse [12].

Genital Defects

Silver et al. described MRI features of genital defects in adult men with CBE following reconstruction. In these males, there is an overall decreased length of the phallus which is, in part, secondary to the deficiency of the anterior corpora cavernosa segment. The paired corpora cavernosa are broad in

appearance and divergent. Dorsal chordee and diastasis of the pubic symphysis also contribute to diminished visualization of the external genitalia. Diastasis of the pubic symphysis increases the transverse distance between corporal bodies [22]. The urethral plate is open and shortened. In males with CBE, the neurovascular bundles are deviated more lateral than usual as they course along the penile shaft [23, 24].

Gearhart and colleagues described the prostate configuration in adult men with CBE. Prostate volume, weight, and maximum cross-sectional area in this group appeared similar to those of normal controls. In all 13 men studied, the prostate gland was located posterior to the urethra [25].

The addition of 3D MRI sequences provides a detailed spatial representation of the genitalia, which are typically very small and often situated deep within the pelvis. The authors have demonstrated value in MRI identification of CBE genital defects prior to reconstruction. Compared with normal infants, preclosure CBE infants have prostates with smaller craniocaudal dimension, but overall greater volume due to greater transverse and anterior-posterior dimensions. In CBE infants, the periprostatic vasculature and pudendal neurovascular bundle courses are externally rotated compared with normal infants. Identification of these structures with preoperative MRI may help to avoid their injury during genital reconstruction [26•].

Genital abnormalities in females with CBE are less precisely described when compared with those in males. Woodhouse and colleagues described genital anomalies in adult females with CBE. The clitoral bodies are bifid, with the labia and mons pubis divergent anteriorly. The urethral plate is shortened and opens anteriorly. The vaginal canal is shortened but has normal caliber. The vaginal introitus is displaced anteriorly and often stenotic. The cervix is closely related to the introitus and lies in the anterior vaginal wall. Additionally, uterine prolapse is common during pregnancy and after childbirth due to weakening of the pelvic floor musculature. The ovaries and fallopian tubes are usually normal [27].

The authors described MRI anatomy of genitalia in CBE female infants before reconstruction. In CBE, the majority of the clitoral bodies were identified anterior to the pelvic attachment. The mean CBE vaginal length was shorter than that of normal infants, a similar finding in earlier studies of adult CBE females [28•].

Imaging-Guided Surgical Navigation

In the authors' institution, preoperative MRI used in conjunction with BrainLab® navigational software (Munich, Germany) has been investigated for intraoperative assessment of CBE anatomy. Surgical landmarks are identified on high-resolution MR images in axial, sagittal, and coronal planes. This data can be reconstructed into a 3D volumetric rendering, which serves as an anatomical roadmap and guides dissection of the pelvic floor [29••].

Surgical Approaches to Repair

Advances in surgical technique have increased the success of exstrophy closure. The modern staged repair of exstrophy (MSRE) for primary closure is gaining significant popularity, although alternative methods exist. Other approaches include the Kelly Repair, the CPRE, the Warsaw Approach, the Mainz Repair, and the Erlangen Approach [30, 31]. The Kelly Repair involves radical soft tissue mobilization to avoid use of osteotomy in a multistaged procedure. The CPRE combines bladder closure with the "penile disassembly" technique for epispadias repair to potentially reduce the number of reconstructive procedures and provide continence without the need for bladder neck reconstruction. The MSRE is the method used at the authors' institution and will be the focus of the surgical anatomy provided below.

Pelvic Osteotomy

To achieve the urologic goals of exstrophy closure, bilateral pelvic osteotomies may be required. Pelvic osteotomy is a proven adjunct to a successful exstrophy closure in select patients [32, 33]. The osteotomy reduces tension on the suture lines of the abdominal wall, posterior urethra, and bladder in an effort to minimize shearing forces that may lead to dehiscence and bladder prolapse. It also eliminates the need for abdominal wall fascial flaps. A combined anterior innominate and vertical iliac osteotomy is used and has several advantages over prior types of osteotomies. The advantages of this approach include (1) both urologic and orthopedic portions of the case are performed in the supine position without the need to place the patient prone; (2) cosmesis is better than the posterior approach; (3) the anterior approach allows for more secure external fixation; (4) the combined approach allows for large cancellous surfaces to maximize chances of healing; and (5) the combined approach also results in superior bone malleability and easier apposition of the pubic bone due to less tension [34].

At the authors' institution, osteotomy is not routinely performed on newborns unless the pelvic diastasis measured on plain film XR is greater than 4 cm in diameter or there is lack of malleability of the pelvis during exam under anesthesia. An osteotomy is also indicated if the patient is greater than 72 h of age and rotation of the greater trochanters does not allow for easy approximation of pubic bones to the midline.

Modern Staged Repair of Exstrophy

The goal of the MSRE is to convert the bladder exstrophy into a complete male epispadias with the urethra relocated to the proximal or midshaft of the penis. The bladder is completely mobilized off the rectus fascia and dissection continues cephalad until the urogenital diaphragm fibers are encountered bilaterally. It is crucial that these fibers are taken down completely on both sides. Disruption of these fibers ensures successful placement of the posterior urethra and the bladder deep within the pelvis, which avoids prolapse and optimizes continence. Patients who have undergone osteotomy are subsequently immobilized in external fixation with modified Buck's traction for 4 to 6 weeks postoperatively. External fixation hardware is removed once XR confirms adequate bone healing.

Surgical Correction of CBE Male Genitalia

During functional bladder closure, it is important to recognize the male genital defects inherent in the exstrophy condition. With deficient corporal length, it is imperative that dissection acts to optimize penile and urethral length as well as preserve the neurovascular bundles that are laterally located. If the urethral plate is left intact, urethral and penile length can be enhanced by mobilizing up to the level of the prostate. Penile length is also achieved by exposing the corpora cavernosa bilaterally and freeing them from the attachments to the suspensory ligaments on the pubic rami. Additional penile length can be obtained at the time of modified Cantwell-Ransley epispadias repair which is typically performed between 6 and 10 months after MSRE.

Complications such as urethrocutaneous fistula, persistent chordee, and wound dehiscence can occur after both modified Cantwell-Ransley and complete penile disassembly for male genitalia repair [19]. A complication that occurs exclusively after the penile disassembly technique is ischemic damage to the glans or corpora [35]. There have been no reported cases of penile loss following the MSRE to date. Nonetheless, imaging-guided surgical navigation could have future applications for genital reconstruction. Identification of complex genital anatomy with navigation software may help surgeons develop penile disassembly techniques which avoid dissection of collateral vasculature, thus avoiding ischemic penile injury.

Surgical Correction of CBE Female Genitalia

The surgical techniques in the female exstrophy patient are similar to those in the male, but there are several important differences to provide for a successful functional and cosmetic outcome. Similar to males, osteotomy is used in cases where the diastasis would not allow for a tension-free closure. The vagina is prepped for the procedure so it may be repaired primarily if it is inadvertently entered. The dissection proceeds along the plane of the rectus and bladder, and continues along the medial aspect of the clitoral/corporal bodies moving deep into the pelvis. The vagina is dissected laterally and posteriorly taking care to leave the urethrovaginal septum intact. The dissection continues until the levator hiatus is encountered. These maneuvers allow for the placement of the bladder and urethra deep within the pelvis and prevent it from being displaced anteriorly when the pelvic bones are approximated. Surgical complications can also occur after female genital reconstruction. Following MSRE, all females universally require vaginoplasty as a teenager for both satisfactory cosmesis and intercourse. At the time of exstrophy closure, there is also potential for damage to the clitoris and erectile bodies in females which are much smaller and less defined than their anatomic counterparts in males. Preoperative imaging used in conjunction with navigation software can potentially avoid clitoral injury, thereby preserving functional and esthetic outcomes.

Future Directions

Continued investigation of imaging guidance in CBE repair is needed as surgical techniques improve. Conventional and 3D ultrasound, most often used for imaging the fetus, do not utilize ionizing radiation, are portable for use in many clinical settings, and may have novel applications for pre-, post-, and intraoperative assessment of CBE. Optimization of MR protocols, for instance by performing a single volumetric imaging sequence which may be reconstructed into multiple planes, may allow for faster scans, potential for intraoperative use, and diminished need for sedation.

Conclusion

Current reconstructive techniques involve closure of CBE defects and aim to improve long-term functional and cosmetic outcomes. The authors' review of the literature describing MRI and surgical anatomy in CBE emphasizes the role of imaging in diagnosis and operative planning. While useful during pre- and intraoperative assessment at the authors' institution, MRI is not routinely performed in all centers, due to high cost, frequent need for sedation, and limited availability. Compared with other available imaging modalities, MRI provides superior anatomic detail of CBE defects without use of ionizing radiation. At present, MRI is best used for identification of defects associated with CBE which may alter surgical approach, for instance abnormally coursing vessels and ureters as well as ectopic kidneys. At the authors' institution, preoperative MRI used in conjunction with surgical navigation software has been investigated as a method for identifying

complex CBE anatomy. With imaging guidance, the surgeon may precisely dissect delicate supporting pelvic structures during repair and repositioning of the bladder and urethra, potentially minimizing complications. Development of imaging-guided surgical techniques could be applied to other stages of CBE repair which require precise dissection, for instance during genital reconstruction.

Compliance with Ethical Standards

Conflict of Interest Emily A. Dunn, Matthew Kasprenski, James Facciola, Karl Benz, Mahir Maruf, Mohammad H. Zaman, John Gearhart, Heather Di Carlo, and Aylin Tekes each declare no potential conflicts of interest.

Human and Animal Rights and Informed Consent This article does not contain any studies with human or animal subjects performed by any of the authors.

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- •• Of major importance

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