Exstrophy

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Last Updated:

Tuesday, May 2, 2023

1. Introduction

Bladder exstrophy exists on a spectrum between epispadias and cloacal exstrophy.[1] These diagnoses range in severity from involving only one organ to being a part of a larger complex of defects. The spectrum includes:

- Epispadias-the urethra is a partial or completely open “plate” dorsally. (**Figure 1**)
- Classic bladder exstrophy (CBE or BE)-the urinary bladder is an open plate on the lower abdomen. There is also a low set umbilicus, pubic diastasis, and anteriorly displaced anus. Ureters insert into the bladder plate with a short detrusor tunnel. Females have a hemi-clitoris on each side with adjoining labia majora and anteriorly displaced vagina (**Figure 2**). Males almost always are associated with epispadias (**Figure 3**).
- Cloacal exstrophy (CE)-the bladder and the ileocecal junction of the bowel are an open plate on the lower abdomen and the hindgut is truncated and terminates in the perineum. This condition is also known as the omphalocele/exstrophy/imperforate anus/spinal defect (OEIS) complex. (**Figure 4**)
- Exstrophy variants-partial manifestations of the above anomalies are seen.

Since the 19th century, various efforts to manage BE have been described. Because the diagnosis is rare and involves various components, there has been no consensus on the best way to manage the condition. Optimal management remains elusive and surgical reconstruction may require multiple procedures. The goals for initial management are protecting the bladder, closing the bladder to create outlet resistance, protection of the upper tracts of the urinary system, and achievement of continence.

1.1 Keywords

Exstrophy; epispadias; cloacal exstrophy; osteotomy; CPRE; incontinence

2. Definition

In classic bladder exstrophy, the urinary bladder is an open plate on the lower abdomen, and is

associated with epispadias.

Children with BE typically have an anteriorly located anus. Also, the female genital anatomy is altered with a more vertically oriented vaginal opening following repair and a wider and shorter vagina than normal. The anterior component of the penis is also foreshortened in males compared to the general population. Classic BE, however, is rarely associated with other organ system malformation.

3. Epidemiology/Prevalence

Classic BE occurs in approximately one per 50,000 live births and occurs in males more often than females.[2] Male epispadias has an incidence of 1:100,000 live births. CE is rarer with an incidence of 1 in 300,000.[3]

4. Risk Factors and Pathophysiology

Experts disagree on the embryologic explanation for the development of BE. In the prescientific era, the cause of BE was attributed to trauma to the unborn child causing ulceration of the abdominal wall and subsequent bladder herniation. Today, we know that the developing human embryo does not normally pass through a stage that corresponds to exstrophy. This knowledge excludes arrested development and implicates an error in embryogenesis involving the cloacal membrane. [4] The mesodermal membrane folds to form and separate the coelomic cavity from the amniotic space late in the 3rd week of development. The intermediate layer of this mesoderm forms the urogenital system. Disruption in this part of the membrane is thought to lead to exstrophy-epispadias complex (EEC). Normally by the 5 th week, the mesoderm has formed lateral folds that tubularize into the gut tube and cloaca. However, if the mesenchymal cells do not migrate appropriately during the 4th week of development, the exstrophy-epispadias complex (EEC) ensues.[5]

A genetic association for BE has been suggested, but there are no clear studies that indicate a specific genetic pathology. Duplication of 22q11.2 has been found in a small group of unrelated cases of BE [6], while ISL1 (ISL LIM homeobox 1) has been demonstrated as a susceptibility gene.[7]

Some studies have been done on the bladder itself in exstrophy. These have demonstrated an increased in collagen to smooth muscle ratio in BE bladders as well as a 3 fold increase in type III collagen in controls vs. BE.[8] Another study has recently demonstrated reduced smooth muscle to connective tissue ratio, and reduced contractile responses compared to normal pediatric bladders.[9]

5. Diagnosis and Evaluation

Prenatal Diagnosis:

BE can be diagnosed antenatally, although the defect is often not noted until birth.[4] Ultrasound (US) can usually detect BE before the twentieth week of gestation by noting absence of the urinary bladder as a fluid-filled structure within the fetal pelvis. Other ultrasound findings include: [5]

- A semisolid mass protruding from the abdominal wall[10]

- A lower abdominal protrusion
- An anteriorly displaced scrotum with a small phallus in male fetuses
- Normal kidneys in association with a low-set umbilical cord[11]
- An abnormal iliac crest widening[12]

Since kidneys and urine production are typically normal in these fetuses, amniotic fluid levels are usually normal.[5,13] Any of the above abnormalities seen on ultrasound should prompt a fetal MRI when available, which provides better visualization and ability to characterize the exstrophy or variant and distinguish exstrophy from other abdominal wall defects.[14] The most challenging and important distinction to make prenatally, if some diagnosis is suspected, is between classic bladder exstrophy and cloacal exstrophy. A large protuberant bladder wall may have bowel visualized behind it and thus be mistaken for an omphalocele.[15] In this way, BE may be given the diagnosis of isolated omphalocele or being on the OEIS spectrum, which vastly changes the prognosis in counseling.

Prenatal diagnosis helps provide for prenatal counseling, optimal perinatal management, and the chance to deliver near a pediatric center where skilled newborn care is available. While the early care may not be essential, most babies will be transferred to a larger center immediately after birth, so this helps to prevent transfer of baby away from the mother. This counseling should include the expertise of a fellowship trained pediatric urologist experienced in the care of children with exstrophy.

Postnatal Evaluation:

The baby should undergo an ultrasound to evaluate the kidneys and to establish a baseline examination for later ultrasound studies, as well as an anterior-posterior x-ray of the pelvis to assess the degree of pubic diastasis (**Figure 5**). Preoperative spinal ultrasound examination should be considered if sacral dimpling or other signs of rare spina bifida occulta are noted on physical examination. In the setting of a baby with anatomy consistent with cloacal exstrophy, the spine must be evaluated with spinal ultrasound and potentially spinal MRI due to the association of cloacal exstrophy with spinal dysraphisms.

6. Treatment

The goals of bladder exstrophy repair are to close the bladder and urethra, reconstruct the genitalia, and create functional organs for continence, voiding and sexual function. A successful primary repair is one of the key predictors of longer term success, as the bladder can cycle and grow in capacity in order to ultimately provide safe storage under low pressure.[16] Achieving normal bladder storage and emptying minimizes the risk of upper urinary tract deterioration, prevents urinary tract infections (UTI) and vesicoureteral reflux (VUR), and decreases the risk of urinary calculi.

6.1 Preoperative Treatment

After delivery, to reduce trauma to the bladder plate, the umbilical cord should be ligated with silk suture rather than a plastic or metal clamp. A hydrated gel dressing or plastic wrap can be used to protect the exposed bladder from superficial trauma from a diaper (**Figure 6** and **Figure 7**).

If closure is performed beyond the first 72 hours of birth, the baby may be discharged from the hospital with the mother, thus providing time and proximity for bonding with the parents. Pre-operative antibiotic prophylaxis is not required. However, perioperative and post-operative antibiotics are used to decrease the risk for infection following reconstruction.

6.2 Timing of Surgery

Traditionally, primary BE closure was performed in the immediate newborn period, prior to 72 hours of life. This allowed for anatomic closure without the use of osteotomies, and decreased bladder exposure, which has been postulated to decrease the need for bladder augmentation [17], and potentially prevent inflammatory changes to the bladder which might impact future bladder compliance and capacity.[18] There has been more discussion on the benefits of delayed closure, and more data to demonstrate the safety and efficacy of this approach, including equal continence rates[17] and no difference in ultimate capacity of the bladder.[19,20] Other benefits of delayed repair might include safer anesthetic at an older age, more coordinated surgical teams, and a chance for parental bonding before a long recovery period.[21]

6.3 Osteotomy

Infants with bladder exstrophy have a wide and flattened pelvis that is laterally displaced. There is external rotation of the posterior pelvis, shortening and external rotation of the pubic rami, and a wide pubic diastasis. [22,23] Approximation of the externally rotated bony pelvis is critical to decrease tension on the abdominal wall closure, decreases rates of abdominal wall dehiscence, to reapproximate pelvic floor musculature, and to place the bladder and urethra deep within the pelvic diaphragm to aid in continence and to prevent prolapse in females. [24,25]

Iliac osteotomies have been performed to aid closure of the pelvis for any child over 72 hours old, in newborns with exceptionally wide diastasis, and in re-operative BE closure/ repair. However, there is some data supporting osteotomies as valuable even within the first 72 hours.[26,27,28] Osteotomies are performed at the same setting as bladder closure to help secure the closure, except in cases of extremely wide diastasis in cloacal exstrophy, when the osteotomy may be performed in a staged fashion.

Bilateral iliac osteotomy can be performed through either an anterior or posterior approach. Posterior iliac osteotomies are performed with the patient prone, after which the patient is then repositioned for the bladder closure. Anterior iliac osteotomies are performed with the patient supine. This avoids repositioning the patient between the osteotomy and the bladder closure.

6.4 General Surgical Principles

Considerations during general anesthesia for exstrophy repair include minimizing abdominal distention, which can increase intra-abdominal pressures post-operatively and can lead to compartment syndrome which may compromise renal function and increase the risk of wound dehiscence. Awareness of the potential for compartment syndrome is particularly important during

the initial repair of a baby with CE, with requisite omphalocele repair. Nitrous oxide should be avoided as it can cause bowel distension. [29,30] An epidural catheter can decrease the need for narcotics and inhaled anesthetics during the operation and keeps the baby comfortable post-operatively. Tunneling the epidural may reduce the risk of infection if it is to be used for prolonged periods after repair. [31] Post-operatively, maximal urinary drainage with ureteral stents and suprapubic tube are critical to divert urine away from the bladder as it heals.[32] (**Figure 8** and **Figure 9**)

Another component of bladder exstrophy closure includes the repair of inguinal hernias. It has been demonstrated that inguinal hernias are far more common than in the normal population, and more common in boys than in girls with BE. Because of this high incidence, and the risk of incarceration, potentially due to the wide defect at the internal ring that accompanies the patent processus vaginalis, it has been recommended that clinically evident hernias be repaired at the time of initial closure. [33,34] It has also been shown that osteotomy may decrease the risk of recurrence inguinal hernia or prevent primary hernias.[35] Given the demonstrated safety of concomitant inguinal hernia repair, repair at the time of exstrophy repair should be considered, especially in boys. When not performed at initial repair, boys with BE should be carefully examined for hernia at each post-operative visit, as there is an increased incidence of new inguinal hernia in the 6 months following initial surgical repair.[36]

6.5 Staged vs. CPRE

Modern Staged Repair of Bladder Exstrophy (MSRE)

General Principles

The modern staged repair of bladder exstrophy (MSRE) consists of three specific components scheduled with specific goals at approximate ages.[16,37,38,39,40] The benefit of this approach was to convert bladder exstrophy into complete epispadias with an initial bladder closure, to allow time for the bladder to start to cycle and grow, while it is closed and protected.

Stage I - Repair (approximation/ closure) of the bladder [includes repair of the proximal (posterior) urethra in the boy].

Typically performed in the newborn – closure of the bladder, posterior urethra, and abdominal wall defect results in the creation of complete (penopubic) epispadias (in the boy) and is usually performed as a newborn. However, closure may not be advisable in the early newborn period because of a small-sized and/ or stiff bladder "plate" or because the infant is premature. In the latter setting, time allows for adequate growth so that closure is feasible.

Stage II – Epispadias repair

At approximately six months to one year of age, epispadias repair is performed. The goal is to create a straight and functional penis with a glanular meatus in the boy, along with an acceptable cosmetic appearance. Exogenous testosterone supplementation may be given prior to this surgery in the boy based on surgeon preference and penile size. Several techniques exist for this surgery in the boy.

These include well-established techniques: the Complete Penile Disassembly and the Modified Cantwell-Ransley.[40] A controversial component of this surgery is whether to rotate the corpora inward or outward following pubic bone closure.

Stage III - Bladder neck reconstruction and bilateral ureteral reimplantation.

At approximately four to five years of age bladder neck reconstruction (BNR) and, if required, bilateral ureteral reimplantation are performed when there is documented adequate bladder capacity and motivation of the patient to participate in a postoperative voiding program. The goal of this last stage is to provide urinary continence. Bladder neck reconstruction techniques include the Mitchell [41] or the Young-Dees-Leadbetter (YDL).[38] Stage III typically includes bilateral ureteral reimplantation as a means of creating space near the caudal extent of the bladder (bladder trigone), in preparation for BNR surgery. Techniques include the cross-trigonal (trans-ureteral advancement/ Cohen technique) [42] or the cephalotrigonal technique.[43]

Complete Primary Repair for Exstrophy

General Principles

Complete primary repair of bladder exstrophy (CPRE) was introduced by Grady and Mitchell in 1999.[44] This technique includes the combination of bladder closure, anatomic bladder neck narrowing, urethral elongation, and epispadias repair in a single operation in order to provide an environment for bladder cycling. At times, anatomy permitting, bilateral ureteral reimplantation may be performed at CPRE in order to achieve the goals of urinary continence and preservation of renal function. Because CPRE has a risk of the devastating injury of the glans, extreme caution is mandatory during this closure, and alternatives such as staging the closure while still closing the bladder neck and proximal urethra are potential options. [45]

CPRE technique in the boy with bladder exstrophy

Key highlights in the closure of bladder exstrophy in a boy:

- Identification of the verumontanum and ureteral orifices for orientation of important landmarks.
- Polyp resection to aid in closing the bladder plate.
- Complete division of all intersymphyseal band attachments in order to allow appropriate placement of the bladder, bladder neck and posterior urethra deep in the pelvis.[46]
- Ventral dissection of corpora off of the urethral plate.
- Glans is kept in continuity, in order to reduce the risk of venous stasis and glanular ischemia in the first few postoperative hours.
- Urethra may be separated from glans to create a hypospadias urethra in order to gain maximal length on the penis.
- Anatomic bladder neck reconstruction to extend the width of the proximal urethra cranially to the level of the true bladder neck.
- Closure of bladder neck, bladder, and urethra with interrupted monofilament absorbable sutures.

- Placement of suprapubic tube for post-operative bladder drainage and cycling.
- Ureteral stents to maximally divert urine from the bladder
- Pubic bone approximation with large absorbable monofilament suture with knot on outside of bone.
- Epispadias repair with external rotation of corpora for maximal length and minimization of dorsal chordee post-operatively.
- Penile skin shaft coverage
- Umbilicoplasty performed at the level of the anterior superior iliac crest.
- Immobilization after osteotomy and pubic symphysis reapproximation.

CPRE is similar in a girl, with a few adjustments:

- **"Y-V" advancement**
- The vagina is advanced caudally toward the anus along the perineum as a Y-V advancement.
- The labia majora are advanced into the perineum alongside the vaginal orifice to provide an appropriate cosmetic appearance.
- Urethral meatus is matured to the perineum by placing sutures prior to symphysis closure (as it all gets buried after the pubic symphysis is reapproximated. The pre-placed sutures can be tied afterwards.
- The female reconstruction can be completed with a monsplasty which serves to provide extra coverage and support anterior to the clitoral bodies, and makes the bifid nature of the clitori more hidden.

6.6 Post-surgery considerations

After the primary closure of an exstrophy, either with or without osteotomy, the patient must be immobilized to decrease stresses on the closure. There are various types of immobilization used, including: (1) modified Bryant's traction; (2) external fixation; or (3) spica cast. Modified Bryant's traction immobilizes the baby within a bed and the hospital for 4-6 weeks with the legs suspended to a bar over the bed. External fixation, and sometimes internal fixation, often is used for older children or in settings of staged osteotomies. However, these do require daily care and carry a risk of external wound infections along the pin sites.[47] The spica cast allows for more mobility and earlier discharge from the hospital.[48,49] In addition, the spica cast can be hinged, to allow for easier removal if needed. Osteotomies are not a component of the classical Kelly technique.[50] However, there is still no consensus on the optimal mobilization after exstrophy repair as some groups strongly believe that external fixation is the optimal method to maximize successful primary closure.[51]

7. Complications

7.1 Early post-operative complications

A devastating early complication of exstrophy repair can be ischemic penile injury that leads to atrophy of the glans, corpora cavernosa, or urethra. This can occur if the vascular supply to the

corporal bodies is damaged during dissection or during compression of the pudendal vessels or more distal neurovascular bundle from closure of the pubic bones leading to vascular compromise or venous congestion from impaired outflow. [52] There may be a higher risk in babies who are closed without osteotomy.[53]

The most common complication is urethrocutaneous fistula (at the penopubic angle dorsally) in males, occurring in 5-40%. Initial conservative management is recommended along with continued urinary diversion via suprapubic cystostomy tube to promote spontaneous closure in some. If the fistula does not close, redo reconstruction should be delayed for 6-12 months, at which time the bladder and urethra should be examined cystoscopically for the possibility of urethral obstruction distal to the fistula. Major complications of either complete breakdown of the bladder and abdominal wall closure (dehiscence), or bladder prolapse are uncommon but can be devastating as they require a complete redo closure.

Another serious early complication is bladder outlet obstruction. This can be noted after the urethral catheter is removed and bladder cycling begins. If the bladder does not empty, the urethra must be evaluated for a urethral stricture or even complete urethral obliteration in the case of females or a posterior urethral obstruction in males. [54] The suprapubic cystostomy tube should be left in place, and changed as needed, until adequate drainage is confirmed. A rare but dangerous complication resulting from bladder outlet obstruction is bladder perforation. [55,56] Outlet obstruction can occur in a delayed fashion as well, so if a child develops chronic bladder and kidney infections he or she should be evaluated for possible outlet obstruction. Early intervention with CIC for several months will often protect the patient during this period.

Urinary tract infections (UTI) and pyelonephritis are common after BE closure,[57] often exacerbated due to postoperative VUR which present in nearly all patients following closure due to the path the ureters take as they enter the detrusor with very little submucosal tunnel. Adequate bladder emptying under low pressures will help to prevent these infections.

7.2 Long/late term complications

Complications can occur throughout the lifetime of patients with BE. In boys, epididymitis occurs with an incidence of 19-33%. Recurrent episodes should prompt evaluation for high pressure voiding due to urethral stricture.[58] In females, pelvic organ prolapse occurs in up to 20-30% of patients, and osteotomy does not always decrease this risk.[59,60] There is a long term risk of malignancy, most often adenocarcinoma (95%) developing in the bladder plate, especially in bladders that are closed later in life, and those that have been augmented with colon.[61] Decline of renal function is a concern in the setting of recurrent pyelonephritis causing scarring, or from increased bladder outlet resistance leading to progressive increased pressure on the kidneys during storage and emptying. Borer et al reviewed 23 patients who underwent CPRE and found that 5/23 had episodes of pyelonephritis, and 5 had cortical defects visualized on renal scan. [62] In an earlier review of 57 patients who had undergone bladder neck procedures for incontinence, renal scarring was evident in 14/57 patients. From these 57, one developed renal insufficiency in a solitary kidney, and one progressed to renal

transplantation after puberty after undergoing early urinary diversion and bladder closure. [63]

Gait abnormalities due to the inherent boney pelvis abnormalities have been demonstrated later in life,[64] and at least one survey indicated that there may be an increased incidence in pelvic and hip pain in adults with a history of osteotomy. [65]

8. Outcomes

Outcomes after BE repair vary due to the wide range in treatment algorithms regarding type of closure and additional procedures performed and lack of a consistent system to classify voiding and continence. Published rates of continence, using a wide range of definitions, vary from 37% to 90%. [27,55,66,67,68] It is not clear which surgical approach best offers the combination of preservation of normal kidney function, continence, acceptable cosmetic and functional genitalia, and overall durability with the least morbidity. It is not a question, however, that successful initial bladder closure is the most important factor toward achieving ultimate satisfactory urinary continence with voiding. [69,70,71] Some examples of outcomes include a review of 46 patients (32 males, 15 females) who underwent CPRE. 12 underwent bladder neck reconstruction and ultimately of the 12, 6 were dry day and night (dry >3 hours), and 4 of these required clean intermittent catheterization (CIC) via an appendicovesicostomy (APV) and 2 of the 12 ultimately underwent bladder augmentation. [72] In another series that described outcomes of 23 patients managed with CPRE, 17 achieved daytime continence of dry intervals of 2 hours or more. Nine of these 17 had undergone BNR with or without bladder neck injection and 3 had undergone bladder neck injection, 5 children used CIC via an APV to empty to completion, and 1 of the 9 underwent augmentation cystoplasty. [73] In another set of 33 patients who underwent CPRE as newborns without osteotomy, 19 had complications, and 12 males of these 19 underwent a BNR, with only 3 achieving continence. All of the 14 with a successful CPRE closure underwent BNR, 8 were continent day and night, 4 during the day, and 2 were wet. [71] A large single institution review of patients from 1975 through 2017 who underwent MSRE looked at 432 patients. From this group, only 23% void per urethra without CIC or diversion following BNR. The rest required CIC, either after BNR with or without augmentation (and only 61-64% are continent). After BNC with diversion, then continence reaches 93%. [74] From the same institution, a study looked at the effect of BNR after CPRE (which had been performed elsewhere). In a single surgeon series, 42 patients with initial CPRE had achieved a mean volume of 145mL. After BNR, 32/42 (76.2%) achieved day or day and night voided continence. [75] In another multi-institutional long term review of patients who underwent CPRE, 48.1% (26/54) were dry or continent, while only 20.4% (11/54) were truly continent with volitional voiding, 9 of these after a single surgery (CPRE). This study and results lends proof of concept that children with BE can indeed void, even though they may be a minority at this stage in our surgical development. This number is validated in a quality of life survey study that gives insight into outcomes from repairs done historically. In this, 130 female respondents with a median age of 30 years reported a 19.2% rate of volitional voiding. [76] Another multi-institutional retrospective chart review study looked at the probability of bladder augmentation in BE in total, with no indication of the primary surgery. The findings for patients born from 1980 to 2016 demonstrated

that 50.9% of patients underwent augmentation overall, and the probability of augmentation or diversion was 70.1% by 18 years of age. [77] This study included patients repaired by various techniques and at a wide range of times which tempers the applicability, but it does provide sobering data as well. Most of all, this study underscores the precise need for prospective, granular data collection to better understand the surgery as well as the outcomes.

As the surgical approach to exstrophy has evolved, there are new discoveries still being made by careful attention to patients who did not undergo diversion to achieve dryness early in life. While little data exists thus far to prove that over time patients can develop continence, a recent report is just one first report of long term natural changes in children who had undergone a staged repair of bladder exstrophy with bladder closure followed by bladder neck reconstruction. In this cohort, patients who remained incontinent at 10 years of age developed voided continence after the age of 16. [78]

Quality of Life

Health-related quality of life and overall social and emotional well-being in children with exstrophy-epispadias is highly variable with some reports indicating a comparable quality of life to healthy children,[79] while other studies indicating significant impairment consistent with other chronic medical conditions.[80] Adolescent males struggle most with social relationships and adaptive functioning related to phallic appearance and urinary incontinence.[79,81,82,83] Regular assessment of social development in this patient group is important including psychiatric evaluation and parental education. [84] In these patients, anxiety and even suicidal ideation is greater than the general population. [85] A recent survey study looking at HRQOL in women with BE received responses from 130 women with BE treated around the world. In this, the mean ICIQ-SF was 6.2 ± 6.2, which indicated moderate severity, and scores of the Female Sexual Function Index (FSFI) were 20.1 ± 9.0, where sexual dysfunction is defined as a score < 26.55. The scores were low across all domains. [76]

Sexual Function

Erectile function and sensation is intact for most male BE patients and libido is intact, and a majority will marry or live with a partner, however some men may have difficulty or concerns about intercourse due to the size and curvature of the phallus. [86,87] Approximately one-third of adult men choose not to or cannot engage in intercourse.[88] There are increasing numbers of reports of using a radial forearm flap to construct a penis analogue that has shown an improved quality of life.[89]

Antegrade ejaculation has been reported to occur in 63% of men,[58] however seminal emission may be slow and continue several hours after orgasm. Sperm quality and quantity is often diminished which may be due to partial obstruction after surgery, epididymitis, or recurrent urinary infections. Long-term studies in adults with BE found that a minority were able to conceive without assisted reproductive techniques. [90,91,92]

For women with exstrophy, sexual intercourse is possible, and may be normal or may be complicated by dyspareunia.[79,86] Fertility is unimpaired in female patients with BE. However, prolapse

occurs more commonly because of the lack of pelvic floor support. Pregnancy in these patients is considered a high risk.[92,93] Pregnancy complications such as hydronephrosis and bacteriuria are common, and may necessitate antimicrobial prophylaxis.[94] Other risks for pregnant women with BE include a higher risk of spontaneous abortion (22%) and risk of preterm labor, with median age of 36 weeks gestation. Cesarean section delivery using a paramedian incision has been demonstrated as a safe obstetric approach for successful delivery. [95]

9. Figures

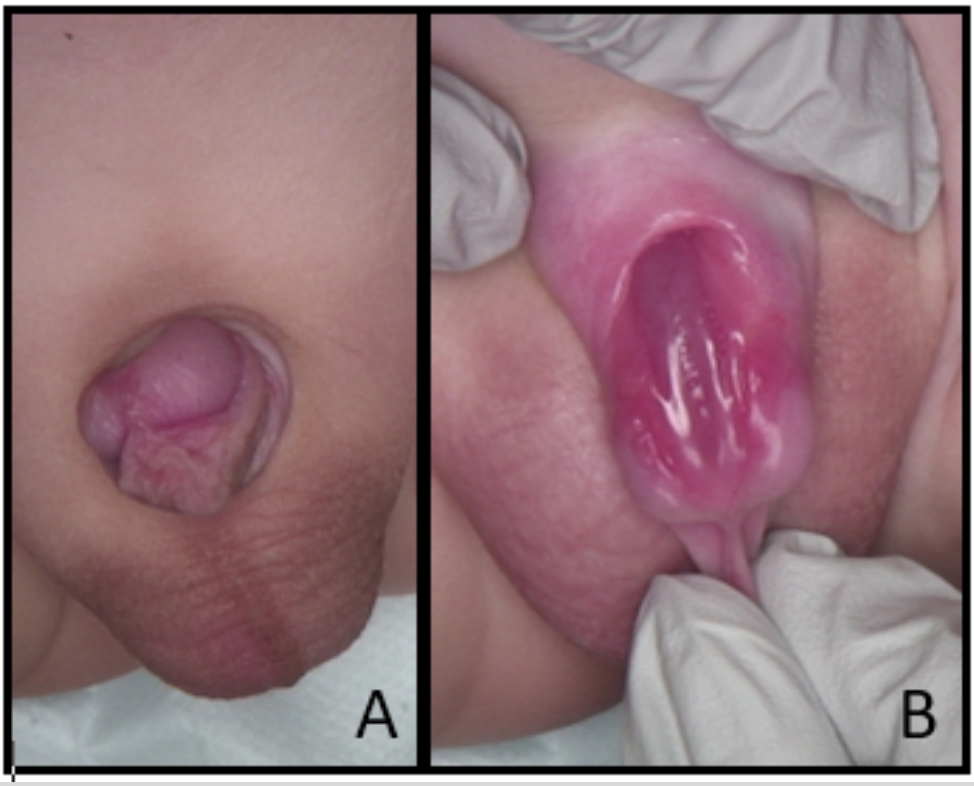

Figure 1: Epispadias. A: Note that the defect can be subtle with the appearance of a circumcised phallus. B: Note the dorsal defect becomes visible with ventral displacement of the glans.

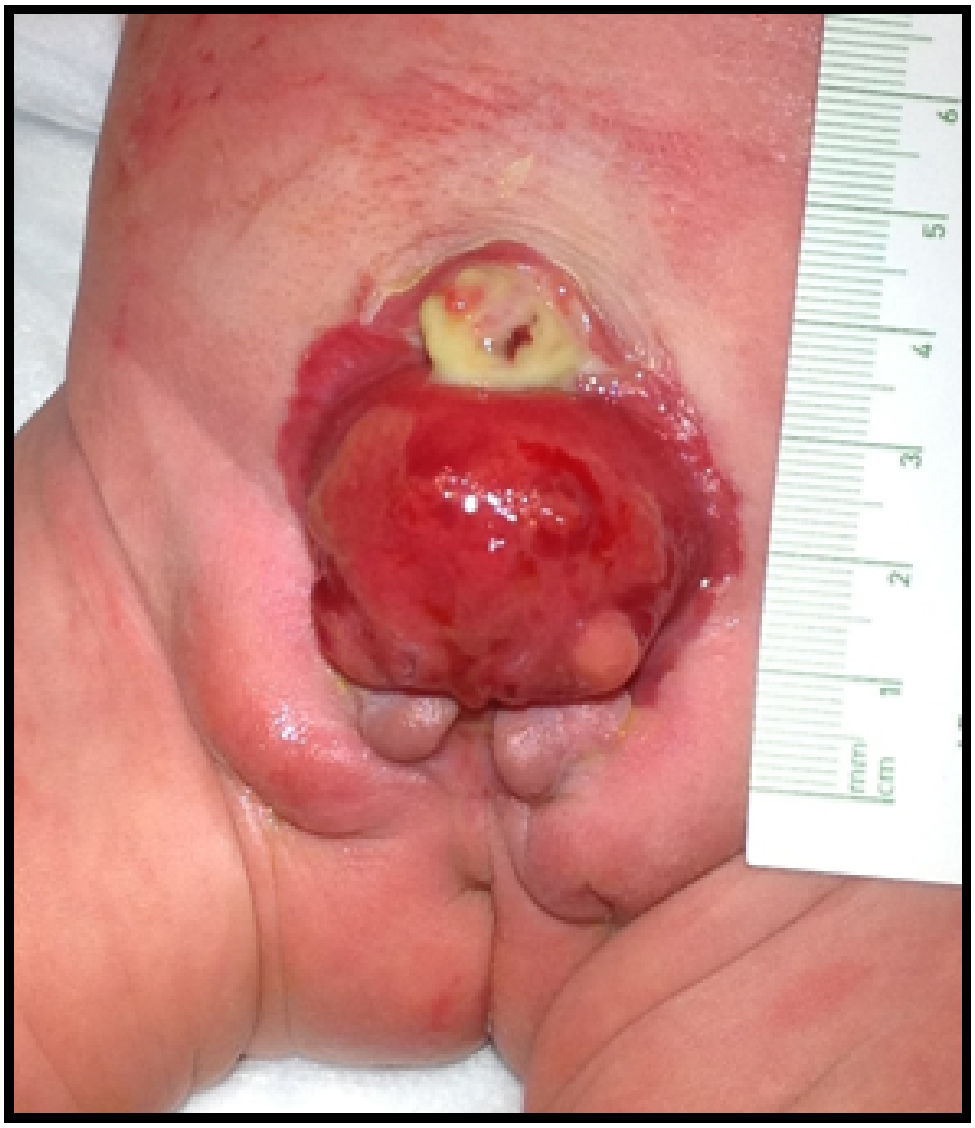

Figure 2: Classic bladder exstrophy – female. Females have hemi-clitoris on each side with adjoining labia majora and anteriorly displaced vagina.

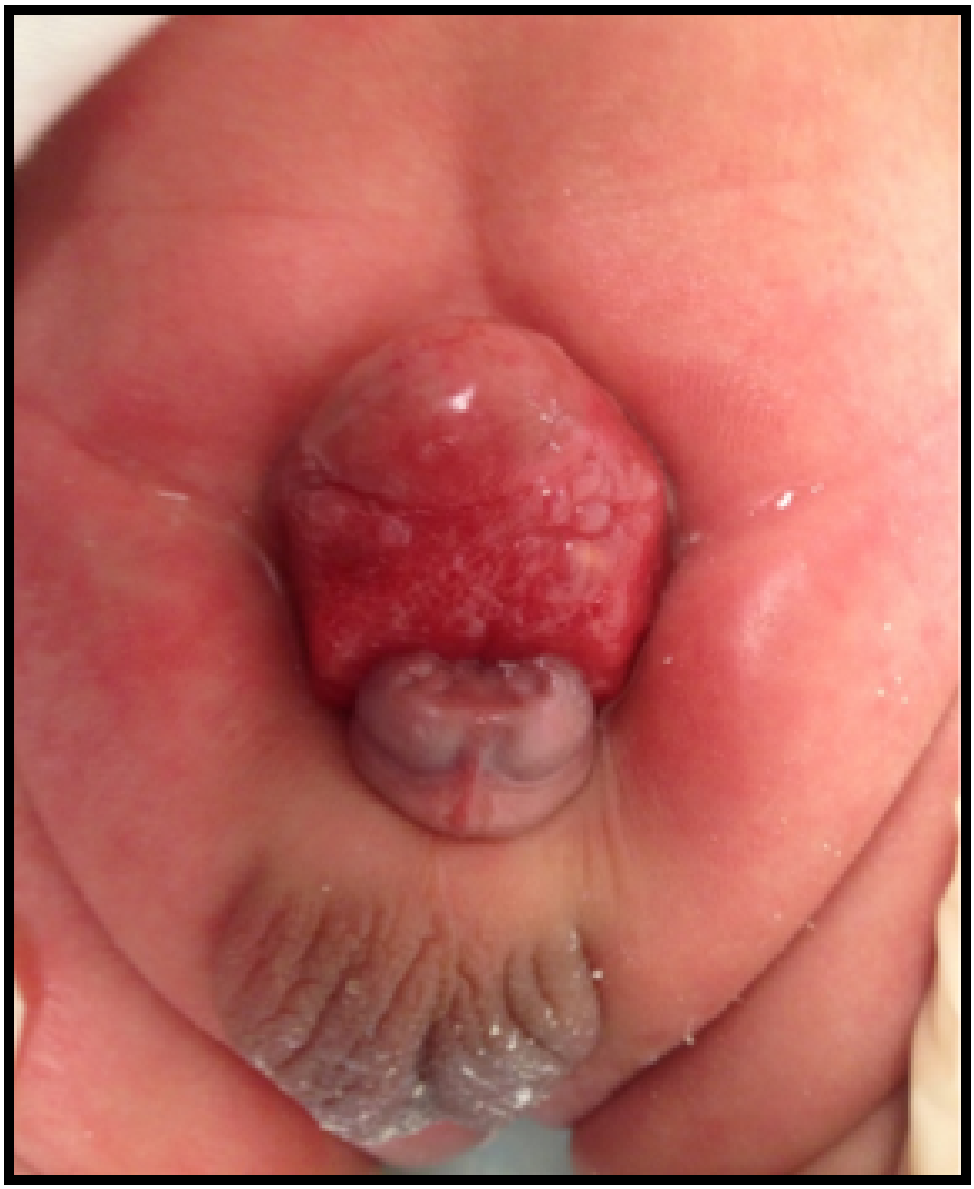

Figure 3: Classic bladder exstrophy – male. Males have epispadias. Ureters insert into the bladder plate with a short detrusor tunnel.

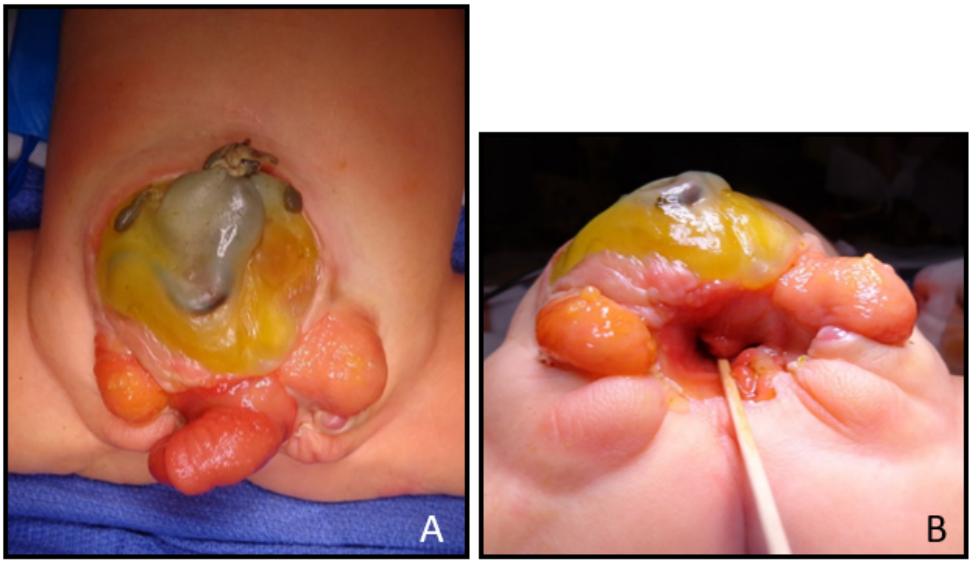

Figure 4: Cloacal exstrophy: The bladder and the ileocecal junction of the bowel are an open plate on the lower abdomen and the hindgut is truncated and terminates in the perineum. A: The small bowel is intussuscepted. B: Small bowel intussusception has been reduced. One can appreciate the 2 bladder halves, scrotal halves, penile halves, and gluteal crease with imperforate anus.

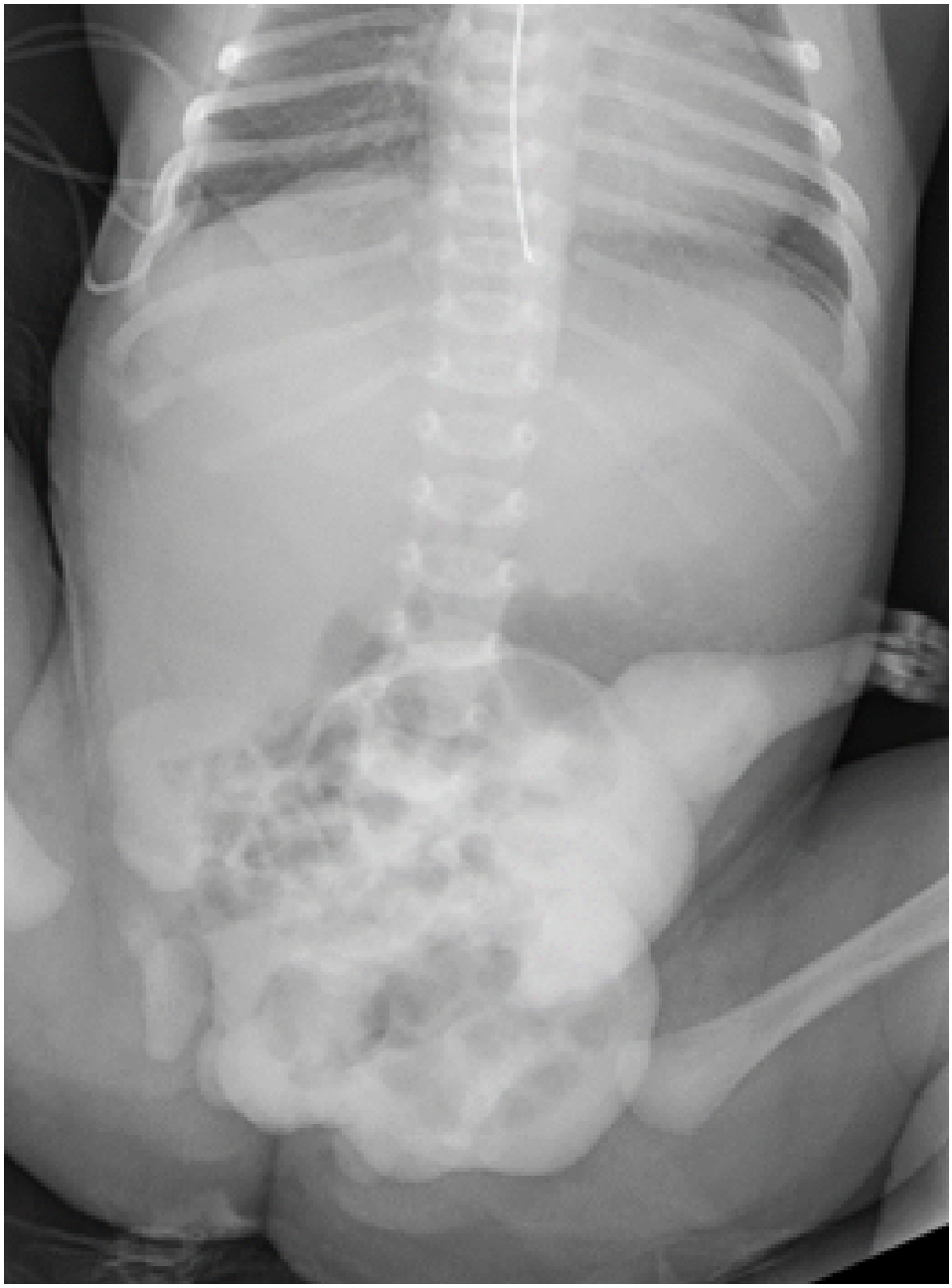

Figure 5: KUB allows assessment of pubic diastasis. Exstrophy will appear as an abnormal soft tissue mass within the pelvis.

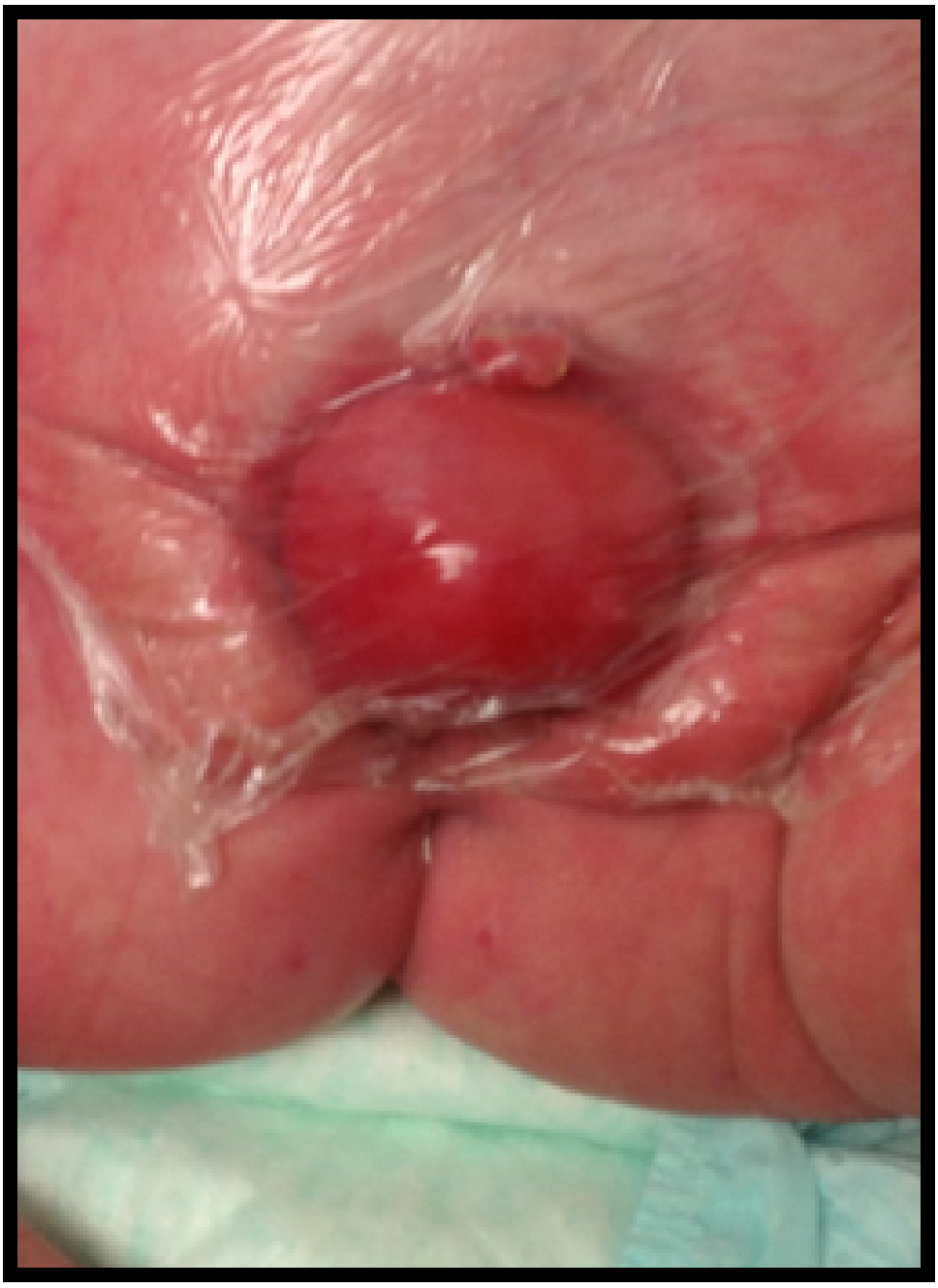

Figure 6: Transparent medical dressing can be placed directly onto the bladder plate to protect the bladder from abrasions from diapers. Pinch a small area at the bottom to allow urine to pass into the diaper.

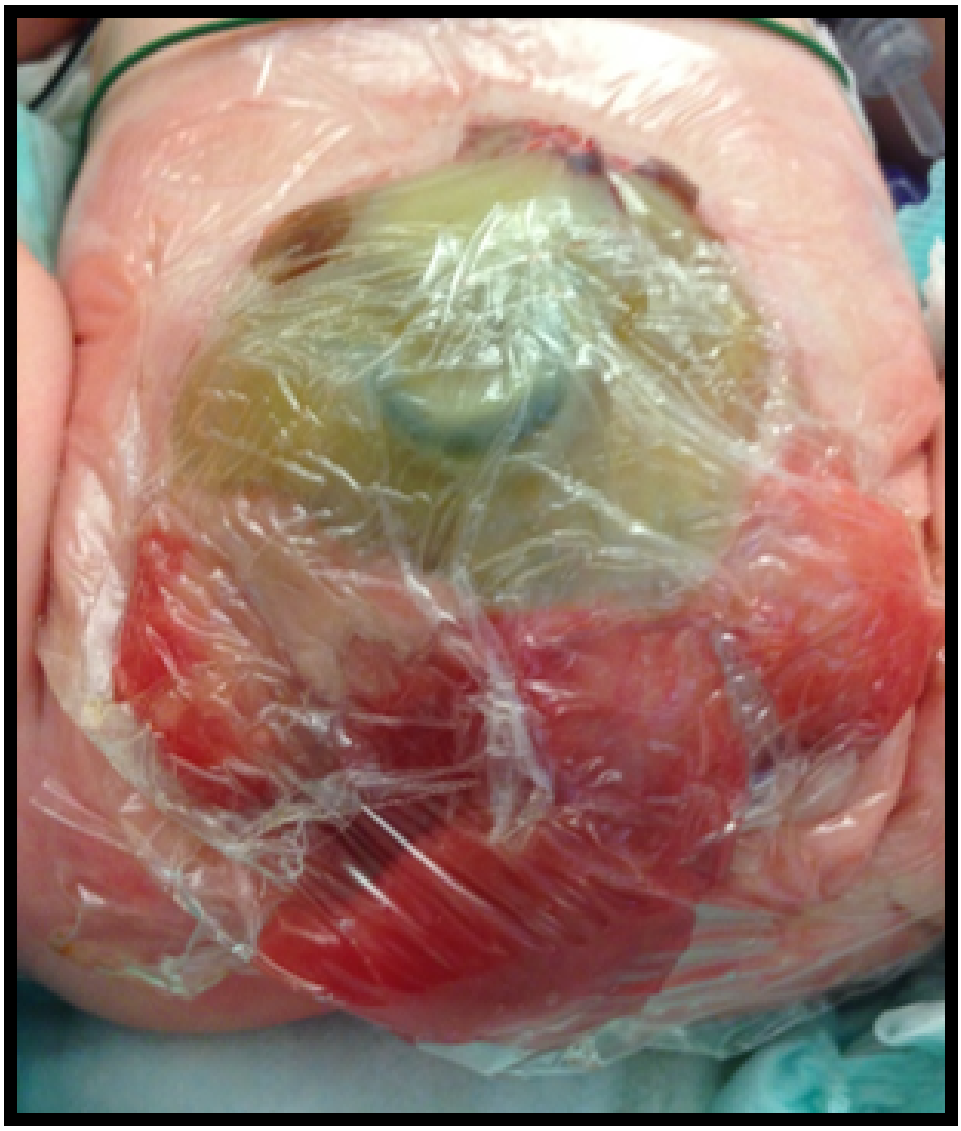

Figure 7: Plastic wrap can be used to protect the cloacal exstrophy defect as transparent medical dressings may not be large enough to cover the entire defect.

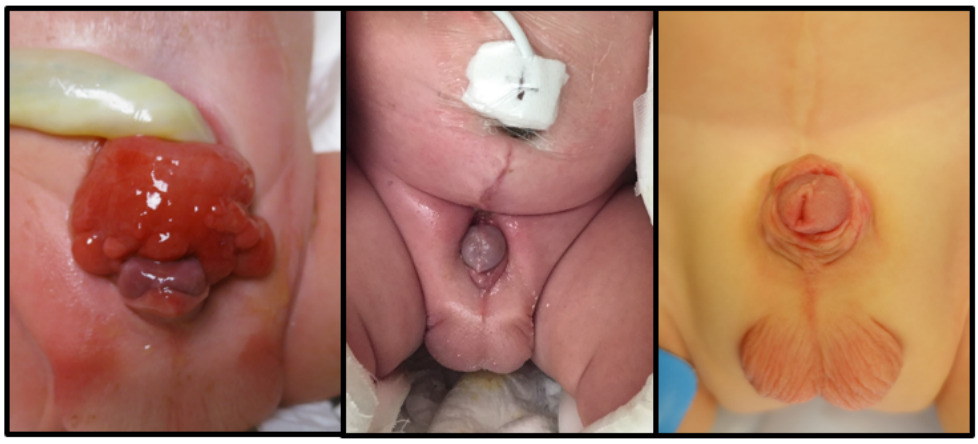

Figure 8: Male classic bladder exstrophy pre- and post-operatively.

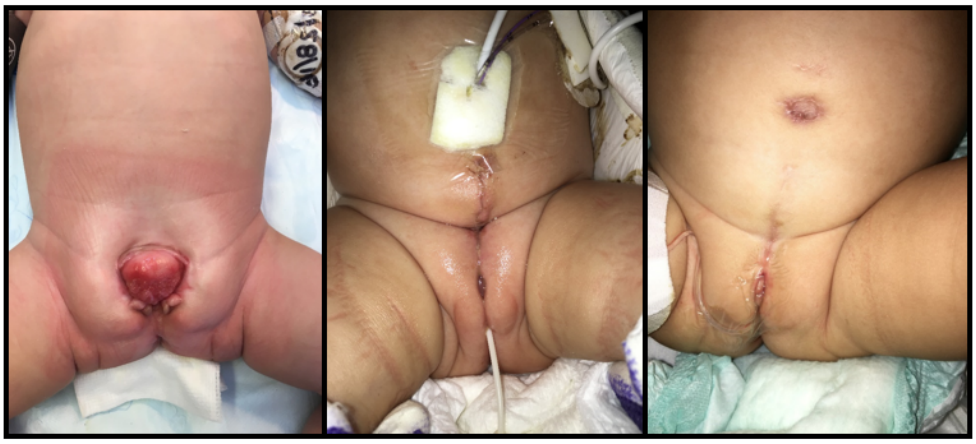

Figure 9: Female classic bladder exstrophy pre- and post-operatively.

Videos

Umbilicoplasty in Bladder Exstrophy Repair

Complete Primary Repair of Bladder Exstrophy in the Girl: Risk Factors for Urinary Retention Lessons Learned and Current Technique

The Richard Grady Monsplasty: A vertical Z-plasty technique

Complete Primary Repair of Bladder Exstrophy in a Male Patient

Presentations

Exstrophy Presentation 1

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