Introduction

During the last decade, the role of pelvic osteotomy has come to the forefront in the modern treatment of bladder and cloacal extrophy. New basic science and clinical discoveries analyzing the deficiencies and deformity in the bony pelvis and the muscular pelvic floor in the patient born with extrophy has led to a greater understanding of how reconfiguring the pelvic ring facilitates the safe and reliable placement of the bladder and posterior urethra deep within the pelvis. We have current experience and follow-up with >900 patients with this condition at our institution. The use of pelvic osteotomy in both primary extrophy closures and reoperative closures has led to the authors’ opinion that the...
judicious use of osteotomy in the closure of the newborn and its obligatory use in failed previous closures significantly enhances the surgical success of lower tract reconstruction along with effective postoperative lower extremity immobilization. The aim of this treatise is to demonstrate to the practicing pediatric urologist and pediatric surgeon new discoveries in this field using osteotomy and its application in their practice of both primary and failed exstrophy closures.

2. **Musculoskeletal anatomy of exstrophy**

The spectrum of genitourinary and musculoskeletal anatomic anomalies encountered with exstrophy suggests an aberrant migration of progenitor cells during embryologic development. Along with the obvious defects in the bladder and urethra, a wide pubic diastasis has been realized for decades. Modern radiologic imaging, including computed tomography (CT) scans and magnetic resonance imaging (MRI), have recently contributed to a far greater description of pelvic bone and muscular abnormalities that manifest in the patient with exstrophy.

In 1995, Sponseller and colleagues [1] obtained CT images of the exstrophied pelvis and compared them with age-matched controls. This demonstrated that there is an approximate 12° external rotation of the posterior aspect of the pelvis (Fig. 1). Follow-up by Stec [2], using three-dimensional CT imaging suggested that this is mostly due to the fact that the sacroiliac joint was tilted 10° more toward the coronal plane than the sagittal plane. The anterior pelvis is affected even more, with an external rotation of 18°. Along with these defects, an overall 15° inferior rotation of the entire pelvis and a retroversion of the acetabulum account for the waddling gait of patients with uncorrected exstrophy. Additional findings from the aforementioned studies included a 30% foreshortening of the pubic rami and a sacrum that is 43% larger by volume and 24% larger in surface area than in normal children.

With regard to pelvic musculature, a second paper by Stec [3] in 2001 demonstrated that the levator ani muscles, which normally assume a conical configuration, are 32° flatter and rotated outward 15° in the exstrophied pelvis. The result is a wider than normal levator hiatus. In the normal pelvis, 52% of the levator group is posterior to the rectum, as opposed to the 68% in affected patients (Fig. 2). Essentially, then, urinary incontinence results from the flattening of the “pelvic sling” and a wider levator hiatus that positions the muscles of continence further away from the urethra. The posterior positioning of the supporting pelvic tissues requires that they support two times

![Fig. 1 – Abnormal bony pelvic anatomy of classic bladder exstrophy. The posterior segments are externally rotated a mean of 12° each; the anterior segments are rotated by a mean of 18° and are 30% shorter than normal.](image)

![Fig. 2 – (A) Displacement of the levator (Lev) ani to more posterior (Post) position in exstrophy, that is, 68% posterior to anus versus normal musculature. Note shortened anterior (Ant) segment of levator ani in exstrophy, 32% anterior to anus versus 48% in controls. Obt int = obturator internus. (B) Greater outward rotation of 15° of obturator internus in exstrophy versus normal. Note that area encompassed by puborectalis is twice that of normal and more flattened.](image)
more body weight than normal and suggests the mechanism by which prolapse occurs.

Multidimensional MRI provides a better tool for investigating anatomy, particularly the soft tissues. Williams and coworkers [4] confirmed the CT studies that showed a flattening of the levator ani muscle. Interestingly, they did not see a correlation between the amount of pubic diastasis and the extent of disproportionate curvature of the levator ani group. MRI has been helpful in examinations of the postoperative pelvis. Haalchi and coworkers [5] found that continence after closure was associated with a sharper angle of levator ani divergence, deeper position of the bladder neck within the pelvis, and a decrease in the intrasymphyseal distance. A Boston Children's study [6] looked at preoperative and postoperative MRIs of children who underwent complete primary closure without osteotomy. They compared the images with normal age and gender controls, specifically focusing on iliac wing angle, ischial angle, obturator internus angle, puborectalis angle, posterior bladder-neck distance, and symphyseal diastasis. They found that after closure, the exstrophy values were more similar to, but not within the limits of, normal. Most importantly, patients whose anatomic parameters shifted closer to normal after surgery were those who were more likely to become continent. These results supported an earlier study designed to evaluate the prostate in exstrophy and furthermore showed that patients who achieved dryness had the tendency of a more narrow pelvis [7].

3. Osteotomy: who needs one?

The goal of an initial exstrophy closure or even reclosure is a secure approximation of the pubic bones and soft tissues in the midline without tension. In some patients with exstrophy who are diagnosed prenatally we have been able to perform closure in the first few hours of life when the pelvis is malleable due to maternal production of the hormone relaxin. If closure is performed after 48–72 h of age, this hormone has decreased and an osteotomy will need to be performed because pelvic bone malleability is markedly decreased. Even before 72 h in the newborn a paradox exists in that while the pelvis may be malleable enough to close without tension, if a significant diastasis exists (>4 cm) this means a good sized bladder template exists and the need for a primary closure to be successful becomes even more important. Clinical clues to the need for osteotomy include taking the child to the operating room and examining the infant under anesthesia. With the surgeon’s hand under the child’s buttocks, pressure is placed on the greater trochanters and they are rotated medially. If the pubic rami can be brought together easily without tension and the diastasis is <4 cm, an osteotomy does not need to be performed. This, however, does not obviate proper immobilization and traction, relief of pain, and movement in the postoperative state if osteotomy is not performed. If during the examination under anesthesia the pubic bones do not come together easily, if the diastasis is >4 cm, or if there has been a prior closure with or without an osteotomy, the performance of a pelvic osteotomy is mandatory. Thus, if the surgeon has any doubt about the need for osteotomy then it should be performed because the morbidity of the modern combined pelvic osteotomy is minimal compared to that of a failed exstrophy closure. Lastly, with easy bony pelvic approximation the stress on the midline structures and incision is lessened and lower rates of wound dehiscence have been reported when osteotomy is performed.

4. Modern pelvic osteotomy

The use of osteotomy in the treatment of the exstrophy complex originated in Europe with the work of Trendelenburg in 1906 who first described the importance of bringing the pubic bones into apposition for closing bladder exstrophy. It was not until 1958 that Schultz described a bilateral posterior osteotomy followed 2 wk later by closure of bladder exstrophy in a 2-yr-old boy. The success of this early effort led authors to recommend the posterior iliac osteotomy as part of the routine surgery of exstrophy closure. At our institution this approach was used successfully for a number of years but has been abandoned because it was associated with increased blood loss and postoperative pain and did not provide a sufficiently secure pelvic closure. In patients with cloacal exstrophy this approach can also be very close to the spinal defect and can cause complications. In addition, the posterior approach requires the patient to be turned from prone to supine at the end of the osteotomy to continue with bladder mobilization.

Several alternative osteotomies have been offered including the anterior innominate osteotomy alone for primary or failed closures with good results [8]. However, in 1995 basic science information demonstrated that combining the anterior innominate osteotomy with a posterior iliac osteotomy done from an anterior approach recapitulated the pelvic anatomy in a more precise manner; thus, the single
anterior innominate approach was abandoned for the combined approach [9]. Other approaches including an oblique iliac wing osteotomy have been described and some promising clinical results have been reported [10]. In 1996, Frey [11] described the pubic ramotomy as a useful adjunct as a method of pubic bone approximation and suggested that this approach would preclude the need for a pediatric orthopedic surgeon. However, although the pubic bones can be brought together with this approach, the pelvic bony rotation provided is inadequate and provides little advantage except in female newborns when manual rotation of the pelvis leaves a small residual diastasis and a ramotomy would allow closure without tension. The modern treatment of the bladder exstrophy complex requires a pediatric orthopedic surgeon with experience in pelvic osteotomy, immobilization, and traction as part of the “exstrophy team.”

The modern era of pelvic osteotomy began in the late 1980s with the widespread application of the anterior innominate osteotomy by Sponseller et al [12]. This technique allows ease of approximation of the pubic bones and the application of an external fixator under direct vision without turning the patient. In all patients as newborn or others who have an extreme diastasis (>6 cm) or cloacal exstrophy or in those in whom the anterior approach alone is used and mobility seems impaired, an additional osteotomy of the posterior part of the ileum is performed through the anterior approach to facilitate correction of the deformity. This step is important because anatomic studies have shown that the posterior portion of the pelvis is externally rotated in exstrophy patients and older patients lose the elasticity of the sacroiliac ligaments.

Although the combined “Sponseller” osteotomy has been well described, a brief synopsis follows (Fig. 3). The infant or child is prepped front and back from the nipples to the knees and from the tip of the scapula to the popliteal fossa. A sterile barrier drape is placed over the bladder. An oblique incision is made inferior to the anterior superior iliac spine as described by Salter [13]. The femoral nerve is exposed and protected. Each side of the pelvis is exposed subperiosteally from the iliac wings inferiorly to the pectineal tubercle and posteriorly to the sacroiliac joint. The periosteum of the sciatic notch is elevated carefully, and a Gigli saw is used to create a transverse iliac osteotomy exiting anteriorly at a point halfway between the anterior superior and anterior inferior spines. This osteotomy is created at a slightly more cephalad level than that described for a classic Salter osteotomy to allow placement of external fixator pins in the distal segments.

For patients with a wider diastasis, poor mobility, a failed prior closure, or cloacal exstrophy the posterior iliac osteotomy is added. This region is approached by continuing the subperiosteal dissection of the medial surface of the iliac wings posteriorly to the sacroiliac joint. A bone rongeur or burr is used to create a closing wedge osteotomy vertically by removing cortical and cancellous bone from the anterior portion of the ilium at least a centimeter from the sacroiliac joint. The more pliable proximal part of the posterior iliac cortex is left intact and used as a hinge. This combination of osteotomies corrects the abnormalities in both the anterior and posterior parts of the pelvis.

Two threaded fixator pins are placed in the inferior pelvic segment and two pins are placed in the wing of the ileum superiorly. An x-ray is taken of the pelvis to confirm accurate pin placement. The soft tissues are closed and the bladder closure begun. At the end of the urologic procedure the pubic bones are closed with a horizontal mattress suture of no. 2 nylon. Patients with cloacal exstrophy and failed exstrophies who have had an extreme diastasis may require staged pelvic closure with the fixator screws placed in the superior pubic ramus and a two-hole plate used to bring the bones into close apposition. The lower abdominal wall is then closed and the external fixator bars are applied between the pins to hold the pelvis in the corrected position.

Postoperatively, patients remain supine with the lower extremities in light skin traction for approximately 4-6 wk. If postoperative radiographs taken at 10 d postoperatively do not show complete reduction of the symphyseal diastasis, it can be gradually approximated with the use of the fixator bars over several days. The external fixator is left in place for 6 wk until adequate callus is seen at the osteotomy sites. Often, the fixator is removed at the bedside under sedation.

Sponseller et al [12] published long-term outcomes of a very large series using the combined

![Fig. 3 – Combined transverse anterior innominate and anterior vertical iliac osteotomy with pin placement and preservation of the posterior periosteum and cortex.](image-url)
osteotomy with very satisfactory results. The most serious complication was that of transient femoral nerve palsy in 7 of 92 patients. This occurred after pure anterior innominate osteotomy in five and after combined anterior and posterior osteotomy in two patients. All resolved completely within 12 wk postoperatively. They were managed with bed rest for 8 wk and then with a knee immobilizer for the remaining 6–8 weeks until quadriceps weakness resolved. Other complications included delayed union on the anterior iliac osteotomy site (n = 3), superficial wound infection (n = 1), transient adductor weakness in the right thigh (n = 1), a pin site infection that required irrigation and debridement (n = 1), and transient motor palsy involving the right peroneal nerve (n = 1); one child had a persistent Trendelenburg gait of unknown cause. Postoperative skin inflammation occurred commonly around the fixator pins and was usually controlled with oral antibiotics. There were two instances of closure failures, one with bladder prolapse and the other with complete wound dehiscence after respiratory syncytial virus infection caught in the hospital during healing. Both were classic exstrophies.

The mean symphyseal diastasis always recurs over time even with osteotomy because there is undergrowth of the pubic bones in exstrophy. Basic science studies from our laboratory shows normal microscopic bony architecture of the cells involved in bone formation so this growth deficit is hard to explain [14]. However, in osteotomies performed in older children any recurrence of the diastasis is much less compared to those younger than 6 mo of age [12]. The relationship of continence and diastasis is somewhat unclear. Some children with a wider diastasis at the time of bladder-neck reconstruction become continent quite easily, whereas others with a narrower diastasis at the time of bladder-neck repair remain incontinent. Thus, other factors such as bladder capacity at the time of bladder-neck repair, surgeon’s experience with bladder-neck repair, and urodynamic parameters all come into play in the attainment of urinary continence. Nonetheless, studies have clearly shown that the chance of a child attaining urinary continence with a failed primary closure are minimal compared to a successful one and that osteotomy certainly enhances the chance for success by allowing healing of the soft tissue without movement or tension as shown by several series [15].

Long-term evaluation of gait outcomes and walking has been performed in this group [16]. Other than the child with a Trendelenburg gait who had an injury to the superior gluteal nerve all have normal gaits on examination. The patients with resolved nerve palsies all have normal gaits and restoration of strength. All patients can run and participate in sports to the same degree preoperatively.

Lastly, a recent long-term study found that exstrophy patients without osteotomy have higher relative joint force and joint stress [16]. In addition, there was more radiologic evidence of degenerative changes in the hip in these relatively young middle-age patients. Long-term follow-up studies are underway because this new finding may add to the algorithm for the use of osteotomy in more patients if the above findings continue to be seen over time.

5. **Staged pelvic reconstruction in exstrophy with extreme diastasis**

Ideally, a complete osteotomy should be performed at the time of the primary closure. However, when the anatomy is unfavorable due to severe pubic diastasis or a paucity of soft tissue, this may not always be possible. Patients with cloacal exstrophy present a particular problem due to the frequent presentation of pelvic asymmetry, malformation of the sacroiliac joints, and hip dislocation. A solution to this problem is to perform the osteotomy in a staged fashion prior to attempting closure [17]. This approach involves performing the initial osteotomy and then placing an external fixator that is gradually tightened over a period of several weeks, followed by an attempt at closure.

In the most recent update of a series of patients undergoing staged closure at our institution, Matthews and colleagues [18] reported on 15 patients, all but one of whom had cloacal exstrophy. All patients were found to have a pubic diastasis of at least 8 cm and all were over the age of 12 mo. The latter point must be emphasized because children younger than 1 yr have inadequate bone strength to permit stable pin placement. All patients underwent combined anterior innominate and vertical iliac osteotomies with placement of interfragmentary pins, followed by placement of an external fixator and positioned in modified Buck’s traction. Pin size was dependent on the size of the child, with infants receiving 2.5-mm pins, young children receiving 4.0-mm pins, and adolescents receiving 5.0-mm pins. Gradual reduction of the pelvis was performed at the bedside under appropriate analgesia and sedation, 48–72 h postoperatively for infants and 1 wk after osteotomy in older children. Final closure was performed when the diastasis was reduced to <1 cm. An interpubic stainless steel plate was used (Fig. 4) to maintain the
closure in nine of the patients, using 4.0-mm screws for the infants and 6.5-mm screws for the older patients. Buck’s traction with the external fixator was used for 6–8 wk following closure, until bone healing was identified on radiographs.

None of the patients described above experienced dehiscence, organ prolapse, or urethral stricture. Immediate complications included one case of the pins loosening that required replacement, one case of ureteral obstruction from a hematoma that required a temporary nephrostomy tube, and one case of temporary femoral nerve palsy. The only long-term complication was a case in which the interpubic plate fractured 4 yr postoperatively and required replacement. Otherwise, patients maintained good pelvic stability with no negative orthopedic consequences.

Although the preceding work primarily included patients with cloacal exstrophy, gradual pelvic reduction has also been investigated in classic bladder exstrophy by Kandemir and coworkers [19]. Their technique of distraction osteogenesis differed in terms of actual approach and technique; however, the principles were similar. Of their 14 patients, only one suffered dehiscence and this was the result of an uncontrollable pin site infection. Their overall success supports the use of gradual reduction in classic exstrophy patients but also highlights the risks of pin site infection and the requirement of meticulous postoperative care.

The use of gradual pelvic reduction should not be advocated as a replacement for a single-stage osteotomy for all patients. However, in extreme cases as described above, its use may significantly reduce the risk of a failed closure.

6. Reoperative osteotomy in bladder and cloacal exstrophy

Although pelvic osteotomy has been common in the contemporary repair of bladder exstrophy, little has been published about reoperative osteotomy in this population. At our institution extensive experience has been accumulated in the reoperation of patients with exstrophy after failure of primary closure caused by dehiscence, prolapse, or other problems. In these patients repeat pelvic osteotomy (RPO) has become a key component of these complex reconstructions. Formerly, little was known about repeat disruption of the pelvic ring. In a recent review of 56 classic exstrophy patients [20] and 15 cloacal exstrophy patients [21], most had undergone posterior iliac osteotomy as their primary closure technique. Additionally, most had either a “mummy wrap” or spica cast as a means of postoperative immobilization. Most of the classic exstrophy patients had a complete dehiscence (>70%; n = 40), whereas the remainder had a bladder prolapse (n = 16). Of the cloacal exstrophy patients, 80% (n = 12) had complete dehiscence and three had bladder prolapse. At the time of reoperative osteotomy, most patients underwent an anterior innominate approach or in the recent past a combined anterior innominate and posterior iliac osteotomy. Of the 56 failed classic exstrophy closures only 5 (9%) required a third closure after reclosure combined with RPO. Of the 15 cloacal patients, none required a reclosure after RPO.

Complications of RPO in classic exstrophy were mainly those of local pin site infections (n = 5), one case of bony nonunion, and septic arthritis requiring incision and drainage. In the cloacal patients the intrasymphyseal plate became infected and was removed in two. Superficial pin site infection occurred in three and one required replacement of screws in the intrasymphyseal plate. Gaits were normal in all patients except those with cloacal exstrophy who were already walking with short leg braces, walkers, etc.

7. Immobilization and traction with pelvic osteotomy

After a successful osteotomy and soft-tissue closure one must not overlook the importance of postoperative pelvic and lower extremity immobilization. Meldrum and associates [22] compared the different immobilization techniques in primary closures for children younger and older than 72 h of age, as well as for secondary closures after
patients had suffered prior, failed attempts. Successful closure was defined as cases in which dehiscence and prolapse were avoided. Children closed primarily with osteotomy before 3 d of age had a success rate of 93% when immobilized 6–8 wk with an external fixator and modified Buck’s traction (Fig. 5). Success rates using 4–6 wk of modified Bryant’s traction (Fig. 6), spica casting, or mummy wrapping were all 0%. For children closed primarily with osteotomy after 3 d of age, the success rates for Buck’s traction and Bryant’s traction were 98% and 50%, respectively, whereas spica casting, mummy wrapping, and no immobilization yielded 0% success rates. Finally, closures after prior failed attempts were investigated and found to be 97% successful when the modified Buck’s was used and slightly less so, 87%, when modified Bryant’s was used. Spica casting and mummy wrapping were significantly less effective with success rates at 33% and 0%, respectively. It is worth noting that all three subgroups had much higher success rates when osteotomy was used rather than when it was omitted.

At our institution, all patients undergoing closure with osteotomy have their pelvis and lower extremities immobilized with an external fixator and 6–8 wk of modified Buck’s traction. Sufficient evidence supports the idea that leg traction effectively controls leg movement and positions the buttocks and pelvis flat on the bed. The fixator and traction devices are removed when the radiographs, as reviewed by the orthopedic surgeon, demonstrate sufficient callous formation at the osteotomy site. Once the hardware is removed, the child is allowed to begin mobilization. We believe that regardless of the immobilization method chosen, adequate pain control, including prevention of bladder spasms, constitutes a critical component in stabilizing the repair.

For children who do not require osteotomy at the time of closure, as outlined previously, our standard postoperative immobilization strategy uses 4–6 wk of modified Buck’s traction. Due to frequent complications and poor success rates, spica casting and mummy wrapping have been abandoned in all cases.

8. Conclusion

Although it may seem that the role of the pelvic bony structures and that of pediatric orthopedists has little to do with the urologic reconstruction of bladder exstrophy, nothing could be further from the truth. The treatment for this major birth defect requires a team of multidisciplinary experts all operating in concert to ensure success. Because the single greatest contribution to eventual continence is a successful primary closure regardless of technique, the role of osteotomy becomes paramount. Osteotomy has proven benefit in the field of exstrophy reconstruction and with the current level of clinical, anatomic, and scientific knowledge it is appreciated why this procedure has many advantages. Although it is not currently possible to replace the shortage of bone that exists in the exstrophy pelvis, the pelvic osteotomy with low surgical morbidity, excellent cosmetic results, and where the effect on success of closure is clearly advantageous, pelvic osteotomy remains a vitally important part of the surgical armamentarium in the modern treatment of the bladder exstrophy-epispadias complex.

Conflicts of interest

None declared.
References


CME questions

Please visit www.eu-acme.org/europeanuroplogy to answer these CME questions on-line. The CME credits will then be attributed automatically.

1. Of the three major common exstrophy closure strategies, which could benefit from concurrent performance of pelvic osteotomies?
   A. Complete primary repair of exstrophy (CPRE).
   B. Modern staged repair of exstrophy (MSRE).
   C. Radical soft tissue mobilization (Kelly repair).
   D. All of the above.

2. Which musculoskeletal defect is characteristic of classic bladder exstrophy?
   A. Internal rotation of the posterior pelvis.
   B. More narrow levator hiatus than normal.
   C. Flattening of the levator ani muscles.
   D. Decreased volume and surface area of the sacrum.

3. Which patient would be a reasonable candidate for undergoing exstrophy closure without an osteotomy?
   A. Infant who is 24 h old, has a 2-mm pubic diastasis, and whose pubic rami can be brought together easily without tension.
   B. Infant who is 24 h old, has a 6-mm pubic diastasis, and whose pubic rami can be brought together easily without tension.
   C. Six-year-old child who requires reclosure after a prior failed attempt that included an osteotomy.
   D. Six-year-old child who requires reclosure after a prior failed attempt that did not include an osteotomy.

4. Which is true regarding recurrence of pubic diastasis after a successful primary closure with an osteotomy?
   A. Rarely occurs but, when it does, can lead to long-term bladder dehiscence and prolapse.
   B. Almost invariably occurs but does not appear to affect continence rates.
   C. Occurs more frequently in children who undergo closure after 6 mo of age.
   D. It is best prevented by placing a no. 2 nylon suture to secure the apposition of the pubic rami.

5. At the time of reclosure for a failed prior attempt that initially incorporated pelvic osteotomies, repeat pelvic osteotomies
   A. Do not need to be performed because the bony abnormalities should have been corrected at the first procedure.
   B. Are more likely to result in significant blood loss than one should expect during an initial attempt.
   C. Should always be performed along with a plating of the symphyseal diastasis to prevent a second failure.
   D. May develop superficial skin infections at the pin sites, which commonly lead to osteomyelitis and septic arthritis.

6. Which form of postoperative immobilization has been shown to be the most effective in all patients who undergo pelvic osteotomies at the time of closure?
   A. Mummy wrapping.
   B. Spica casting.
   C. Modified Bryant’s traction for 4–6 wk.
   D. An external fixator with modified Buck’s traction for 6–8 wk.