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Orthopaedic management and considerations for classic bladder exstrophy

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Bladder exstrophy results from an embryological aberration that affects the genitourinary and musculoskeletal systems leading to anterior midline defects. The complex urogenital reconstruction procedure is facilitated by the closure of the bony pelvic ring by the orthopaedic team. There are many orthopaedic considerations in the setting of bladder exstrophy. Therefore, this review aims to present a contemporary overview of orthopaedic management in bladder exstrophy, including osteotomy techniques, intraoperative radiology, pin placement, and repeat pelvic osteotomy (RPO) for failed primary bladder closure. The age of the patient and other comorbidities, such as hip dysplasia, are important orthopaedic considerations during the preoperative evaluation. Several types of osteotomy techniques exist to facilitate bladder closure to alleviate tension, enhance genital reconstruction, and improve appearance and possibly continence. Bilateral anterior pelvic osteotomy offers good approximation and improved mobility of the pubic rami at the time of closure. They allow the surgeon to prevent vertical migration of the hemipelvis through direct visual external fixator placement and postoperative adjustments. There is no need for patient repositioning during surgery. In the setting of bladder prolapse or dehiscence after initial bladder repair, RPO augments the success rates of secondary closure with better closure of the abdominal wall and genitalia. Orthopaedic management is essential in treating bladder exstrophy as the approximation of the pubis with pelvic osteotomies is important to optimise primary and secondary bladder closure outcomes.

Keywords: bladder exstrophy, osteotomy, pelvis, orthopaedic management

Introduction

Bladder exstrophy results from an embryologic malformation that affects the genitourinary and musculoskeletal systems, and less commonly affects the intestinal system with cloacal exstrophy. This condition results from a premature anterior rupture of the cloacal membrane during fetal development, therefore, ingrowth of mesenchyme into the abdominal wall is inhibited.[1] The most common form is classic bladder exstrophy (CBE) in which the bladder is open and exposed externally on the abdominal wall along the malformed epispadiac urethra. In cloacal exstrophy, the hindgut is also open with an omphalocele and can be a part of a larger spectrum called OEIS (omphalocele, exstrophy, imperforate anus, and spinal defects) syndrome.

There are many orthopaedic considerations in the setting of bladder exstrophy (Table I). Therefore, this review aims to provide a contemporary perspective of the orthopaedic management of bladder exstrophy, including osteotomy techniques, intraoperative radiology, pin placement, and RPO for failed primary bladder closure. Informed consent was obtained for patient photography contained herein.

Bone abnormalities

Numerous characteristic osseous morphologic changes are observed in bladder exstrophy.[2] Patients with CBE have a mean of 12 degrees of external rotation of the posterior segments of the pelvis on each side, retroversion of the acetabula, a mean additional 18 degrees of external rotation, and 30% foreshortening of the anterior ischiopubic segment (Figure 1).[3]

Table I: Key points regarding pelvic anatomy in bladder exstrophy and pelvic osteotomies when utilised for bladder exstrophy closure

1. The pelvis in bladder exstrophy is different from a normal pelvis in the following ways: • 12 degrees (mean) of external rotation of posterior segments of the pelvis bilaterally • retroversion of the acetabulum • 18 degrees (mean) of external rotation as well as 30% foreshortening of each anterior ischiopubic segment • pubic diastasis is typically 4 cm or greater in bladder exstrophy and much greater in cloacal exstrophy
2. The osseocartilaginous state of the newborn pelvis (up to about 72 hours) means that osteotomy is rarely needed in this group. A large, non-absorbable pubic suture and limb adduction with postoperative traction for four weeks are sufficient for newborn closure.
3. Pelvic osteotomy should always be used after three days of age.
4. Pelvic osteotomy goals are to relieve tension on the bony pelvis, thereby easing closure of and tension on the bladder. Osteotomies do not help urinary continence.
5. Bilateral anterior innominate osteotomy is the preferred method at Johns Hopkins because, relative to posterior osteotomy, they: • do not require prone positioning (i.e. can be accessed anteriorly) • provide easier bone fragment rotation • are more vertically stable
6. The transverse osteotomies are performed midway between the anterior superior iliac spine (ASIS) and anterior inferior iliac spine (AIIS), or higher in younger patients to maintain adequate bone fixation.
7. Vertical osteotomies of the inner wall of the posterior ilium superior to the sciatic notch are necessary in older children or cloacal exstrophy. They are needed because the sacroiliac ligaments in older children have less mobility. Therefore, the vertical iliac osteotomies help mobilise the iliac wings.
8. 120 mm long pins that are 4 mm in diameter are used in toddlers and children (6.5 mm in teenagers and adults) with two placed below and one placed above the horizontal osteotomy.
9. The postoperative immobilisation protocol is as follows: • skin traction • bed rest and immobilisation in Bryant's traction for four weeks (in newborns without osteotomy) • bed rest and immobilisation in Buck's traction for six weeks (if osteotomy is used)

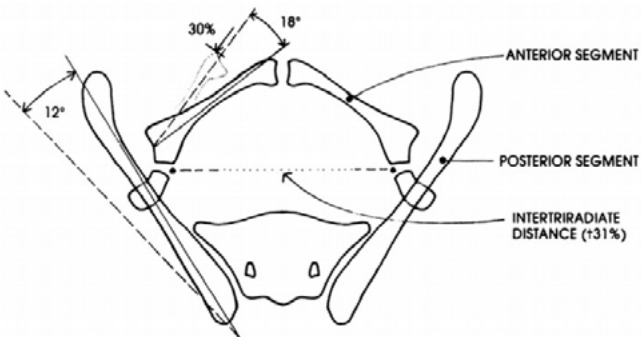

Figure 1: Diagram of osseous changes observed in bladder exstrophy, including external rotation of the posterior iliac segments, acetabular retroversion, and shortening of the pubic rami (reproduced from Sponseller et al.[3])

Although it has no known adverse functional effect, the diastasis is typically at least 4 cm at birth and this proportionately increases with growth to more than 14 cm in adulthood.[2] It is unknown whether this bony abnormality is due to the loss of a tension band function from the symphysis or to a programmed cell growth difference. To investigate the role of pubic symphysis tension in the development of the pelvic ring, Cottrill et al.[4] performed a symphysiotomy in neonatal rabbits, which was maintained with a silicone barrier. As the rabbits aged, they had progressive widening of the symphysis and external rotation of the pelvis. This indicates that the tension band itself could be important in producing this condition.[4] However, in this same small animal model there was no significant difference in anterior segment length, suggesting additional factors may contribute to the growth disturbances in CBE.[4]

Along with the wider diastasis, patients with cloacal exstrophy have more pronounced abnormalities in all forms. The diastasis is greater, deformity is asymmetrical, sacroiliac joints are often unstable, and hips are dislocated in 25% of patients.

Goals of osteotomy

The main goal of osteotomy is to relieve tension on the bony pelvis and thus ease closure of the bladder. It also improves genital appearance, especially in females, and could help promote continence.[5] However, it is rarely needed in newborns if they are closed within a few days of age because the pelvis is malleable in its osseocartilaginous state and can easily be held together with a suture and limb approximation. The exstrophy bladder defect is under a lot of stress, so providing pelvic approximation using external fixator pins facilitates the abdominal wall closure and allows the soft tissue closure to heal under minimal to no tension.

Types of osteotomies

Various approaches have been described, including anterior innominate osteotomy, posterior iliac osteotomy, anterior diagonal mid-iliac osteotomy, superior or anterior pubic ramotomy, and combined osteotomies.[6-11] A posterior osteotomy has several disadvantages, such as the need for prone positioning, increased difficulty in rotating fragments, and a vertically unstable construct in older children. At our institution, bilateral anterior innominate osteotomy is the preferred method for initial bladder exstrophy closure.

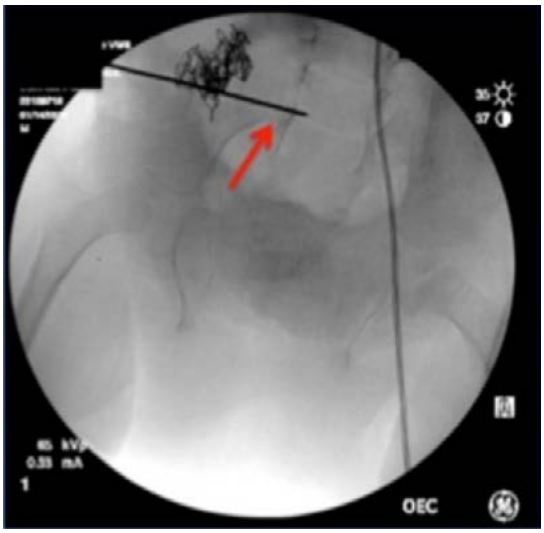

Figure 2: Osteotomy guide pin (red arrow) on fluoroscopic imaging

Surgical technique

The surgical technique involves orthopaedic and urologic correction in sequence with the patient supine. The patient is draped below the costal margin and the bladder is covered in a sterile fashion. Using the ASIS and other pubic anatomic landmarks, an incision is made and the lateral femoral cutaneous nerve and its branches are carefully dissected and retracted medially. After exposing the ilium through a tensor-sartorius interval, a guidewire is used to localise the level of the osteotomy, especially since the bone can be anomalous and small. The guidewire is inserted transversely or with a slight downward angle, aiming for the top of the sciatic notch (Figure 2).

Two pins are placed in the inferior fragment and one in the iliac wing fragment on each side (Figure 3).

A transverse osteotomy is performed using a fine saw through an anterior iliofemoral approach. The osteotomy is performed

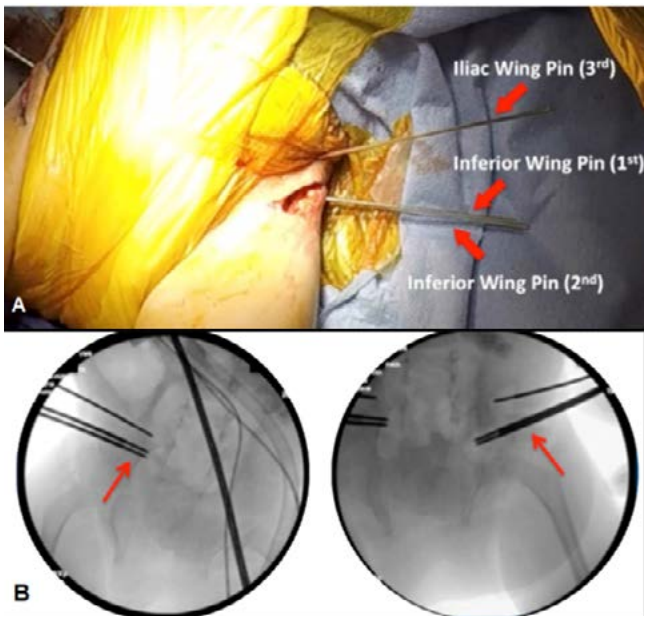

Figure 3: Intraoperative pin placement (A) and fluoroscopic imaging with two pins (red arrows) inserted into the inferior wing (B)

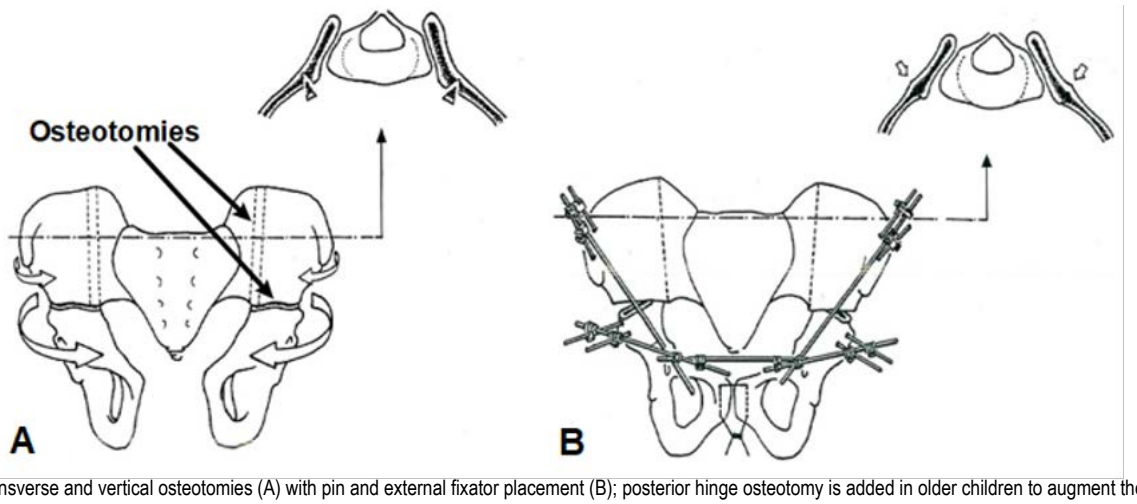

Figure 4: Transverse and vertical osteotomies (A) with pin and external fixator placement (B); posterior hinge osteotomy is added in older children to augment the rotation of the iliac wings

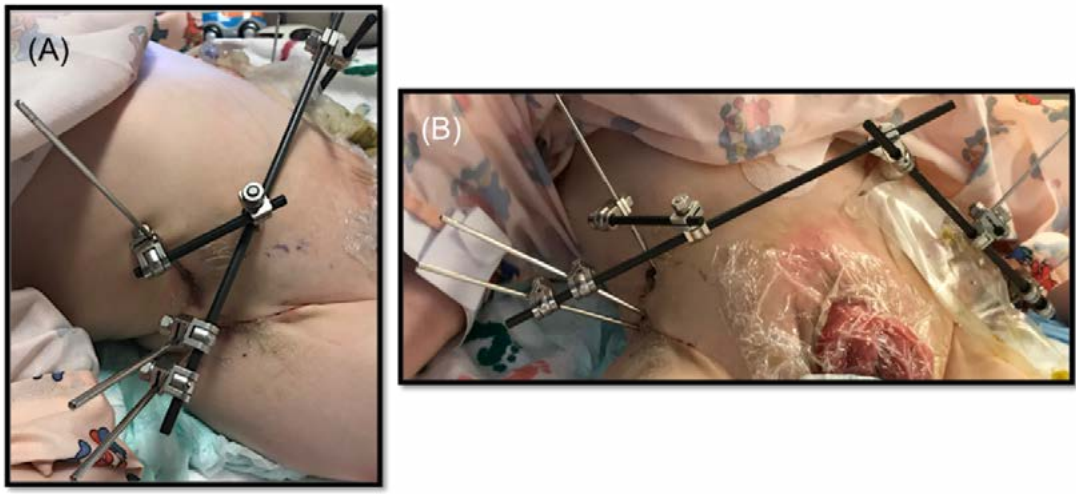

Figure 5: Postoperative right side (A) and bilateral (B) pin placement with external fixator

between the ASIS and AIIS, which is performed slightly higher if the patient is younger to maintain adequate bone fixation.[12] In an older child or a patient with cloacal exstrophy, a second osteotomy can be performed to create a vertical osteotomy of the inner wall of the posterior ilium superior to the sciatic notch, thereby creating a vertical closing wedge of the posterior pelvis in addition to the horizontal anterior pelvic osteotomy (Figure 4).[11]

Vertical osteotomy is recommended to account for the decreased mobility of the sacroiliac ligaments in older children and thus helps to mobilise the iliac wings posteriorly to close this gap.[13] Stabilisation can be achieved by a large non-absorbable suture around the rami, a two-hole plate, or other forms of anterior fixation.

Figure 6: Buck's traction to immobilise postoperatively

After the urologist has finished the bladder and anterior abdominal wall closure, the fixator is assembled (Figure 5) and the patient is maintained in light Buck's traction to immobilise the patient and prevent motion (Figure 6). Aside from aiding the urologic reconstruction and abdominal wall closure, the bony reconstruction helps to keep the tubes draining freely and maintain the bladder repair.

While skeletal traction or casts can be utilised in cloacal patients, skin traction is utilised for CBE repair. Skin traction application involves a single roll of 5–8 cm soft roll wrapped in a single layer without overlap distally from the toes to the proximal thigh. The wrapping should not be applied too tightly during traction to prevent the development of serious complications, such as compartment syndrome or pressure sores.

If no osteotomy is performed, the postoperative protocol involves approximately four weeks of bed rest to remove tension from the closure. If an osteotomy is performed, the postoperative protocol requires six weeks of bed rest, and two more weeks before standing using knee immobilisers while maintaining adduction of the lower extremities during this time. Bones take six weeks to heal and are still not at maximal strength at six weeks. This duration increases with increasing age. Muscle strength also needs to be restored,

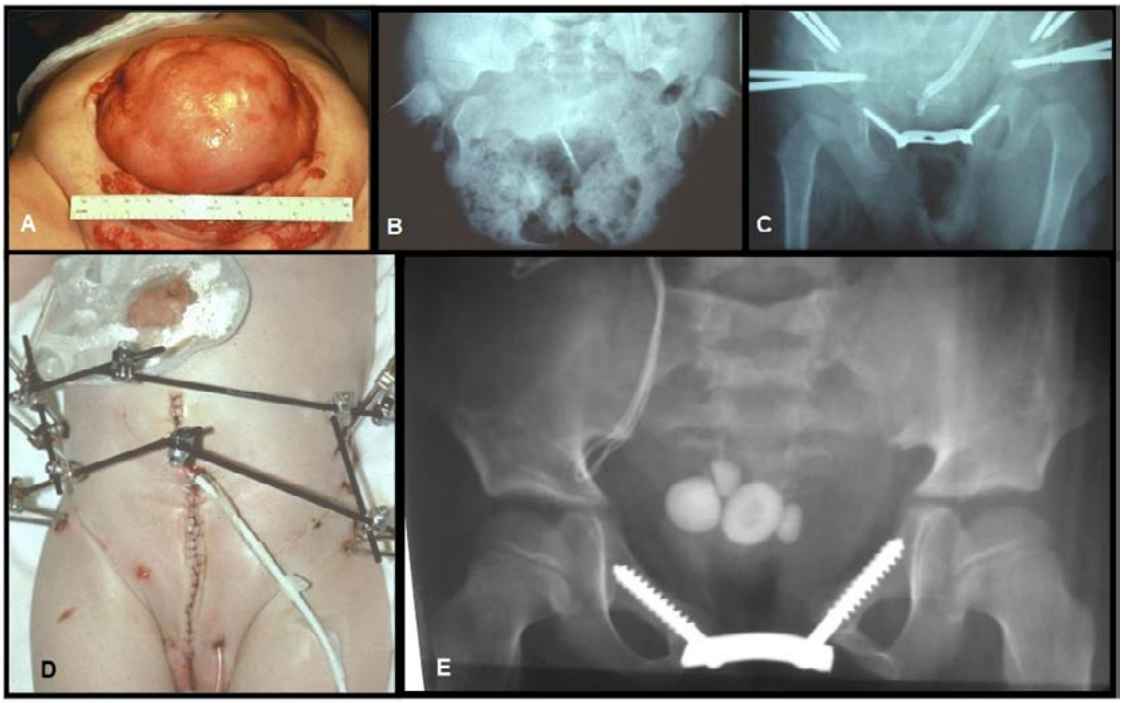

Figure 7: An 18-month-old cloacal exstrophy patient with a large omphalocele (A), significant splay of the pelvis (B), with severe abduction and external rotation, the combination of osteotomies (C), anteriorly and posteriorly, and anterior fixation (D) led to substantial improvement; at age 10, the reconstruction is still well maintained with good hips and a symmetric pelvis (E)

so a painless limp during this period is a sign that the muscle and nerve have not regained normal strength.

The diastasis is immediately reduced after the osteotomy; however, there is some recurrence over time. It will not progress to the preoperative level, but over the ensuing months and years, there is a tendency for the bone to grow back partially to the way it was formed. The osteotomy can minimise this occurrence but it cannot be eliminated.[5] Correction is least maintained in younger patients, likely due to continued undergrowth of rami. Furthermore, correction of diastasis is not correlated with continence.

The older the patient is at the time of osteotomy, the more permanent the closure. Our current recommendation is to close patients with CBE as neonates without osteotomies. Closure is deferred in patients with cloacal exstrophy until they have adequate bone stock, which occurs around 18–24 months (Figure 7).

Complications

From our institution's more than 800 patient series, most complications were transient. Four patients developed pressure sores from traction, of which one patient developed compartment syndrome from the lower extremity wrapping. Femoral nerve palsies occurred in 10 patients, primarily on the left side, which all resolved within a few months. The femoral nerve palsies are likely due to tension on the inguinal ligament as the symphysis is approximated. Placing the pins more transversely also decreased the pressure on the femoral nerve. Additionally, three sciatic nerve palsies occurred; two of which were peroneal nerve-only palsies, but likely occurred at the level of the pelvis. The third was a complete sciatic nerve palsy, which resolved after adjustment to the slide on the inferior fragment. While the incidence of superficial pin infections/inflammation are relatively common, deep infections are rare with only one case of osteomyelitis occurring. From our experience, anterior iliac osteotomies are a safe and effective technique to assist in the closure of difficult exstrophies, especially in older children, after a failed bladder closure and in cloacal exstrophy.

Reclosures

Orthopaedic intervention at repeat closures may be indicated to achieve urologic goals as outcomes worsen with each successive failed closure.[14] Before the reclosure procedure, it is important to perform a hip examination because bladder exstrophy patients are at an increased risk for developmental dysplasia of the hip compared to the general population.[15] Additionally, it is important to ensure the patient is neurologically intact as superior gluteal nerve, femoral nerve, and peroneal nerve injuries are possible. If possible, prior incisions should be used for the reclosure procedure. The initial dissection should be performed carefully as the cutaneous nerve may be challenging to find and the periosteum redevelops. Bony anatomy may be more sclerotic; however, the bone healing is the same. For stabilisation, strong fixation of both superior pubic rami is recommended with the largest screw (4.5 mm in toddlers and 6.5 mm in adults). The connecting plate or rod is contoured anteriorly to avoid urethral impingement in the setting of older CBE or cloacal exstrophies (Figure 8).

From our single institution experience, 56 patients who experienced dehiscence or prolapse after initial bladder repair underwent secondary bladder closure with RPO at a mean age of two years.[16] Of these patients, 15 (27%) had undergone initial bladder closure without osteotomy. The remaining 41 patients (73%) had undergone pelvic osteotomy during the primary bladder closure with posterior iliac osteotomy being the most common initial closure technique (80%).[16] All patients were managed with RPO, of which 38 patients

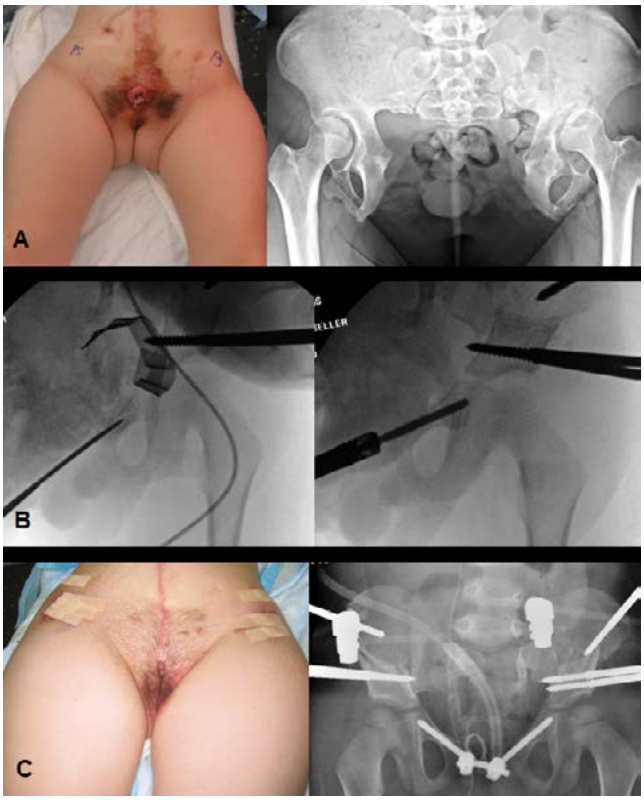

Figure 8: Preoperative (A), intraoperative (B), and postoperative (C) clinical and radiographic imaging of a 19-year-old female with bladder prolapse and hip dysplasia who underwent anterior pelvic reconstruction and bladder reclosure In repeat pelvic osteotomies or when an extreme (>6cm) diastasis exists, fixation of superior pubic rami is accomplished with screws and a connecting plate or rod.

underwent reoperation within one week of failure. Complications included non-union (*n* = 3), repositioning (*n* = 1), and osteomyelitis (*n* = 1).

There is more concern for pelvic floor issues, prolapse, and appearance in extreme or failed cases in older adolescents or adults. Correction in these patients involves a more significant procedure with increased complications and postoperative pain. The surgical time for the orthopaedic component of the procedure is less than 90 minutes. With the use of tranexamic acid, the estimated blood loss of 160 cc is comparable to the index bladder repair. The lower extremities are immobilised for 4 ± 2 weeks and a fixator is in place for 6 ± 2 weeks.

Conclusion

An anterior bilateral pelvic osteotomy allows for better approximation and improved mobility of the pubic rami at the time of closure, prevention of vertical migration of the hemipelvis, direct visual placement of an external fixator, and adjustment postoperatively, with no requirement for turning the patient during the operation. In addition, this procedure allows for adjunctive posterior osteotomy from the anterior approach to provide adequate closure in those with cloacal exstrophy, prior failed closure, or extreme diastasis of > 6 cm. Moreover, RPO is associated with enhanced success rates of secondary closure with better cosmesis of the abdominal wall and genitalia.[17] In particular, repeat anterior iliac osteotomies are a safe and effective technique for failed bladder exstrophy closure with minimal orthopaedic complications and a normal gait postoperatively.

Conflict of interest

The authors have no conflict of interest relevant to this manuscript. This manuscript is not under consideration at any other journal.

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Ethical approval

Not applicable to this review article. Informed consent was obtained for all patient photography, and there is no identifying personal information in the photographs.

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