The Role of Osteotomy in the Modern Treatment of Bladder Exstrophy

FROM THE GUEST EDITOR

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The role of osteotomy has become more important during the last ten years in the treatment of both primary and secondary closures of classic bladder along with cloacal exstrophy. New imaging modalities have allowed better evaluation of both the soft tissue and bony pelvic structures. While the performance of an osteotomy does not recapitulate the pelvic anatomy completely, it does restore the anatomic relationships to a more normal state. In addition, the performance of an osteotomy helps secure the closure and takes tension from the midline repair. Also, with appropriate immobilization the number of successful closures without prolapse and dehiscence increases significantly following the osteotomy regardless of the exstrophy repair chosen.

As an adjunct to osteotomy, major strides have been made to help relieve pain and movement in the postoperative exstrophy patient. Better medicines for pain and movement and the placement of tunneled catheters which can remain for long periods of time have made the postoperative state smoother for the patients, their parents, and the nursing staff.

An orthopedic consultant with more than a passing interest in pelvic reconstruction is required, along with specialist nurses who will care for the fixator, traction apparatus, and pin sites with great care. With this team assembled the overall care of the exstrophy patient will improve and the routine use of osteotomy will become commonplace in pediatric practice outside of exstrophy centers.

FROM THE EDITOR

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During my fellowship training, which was under John Duckett, osteotomies were not used for bladder exstrophy closure. I can even recall a couple of reclosures in which osteotomies were not thought to be necessary. Data from Johns Hopkins, as well as other institutions, seem to support the fact that good pelvic ring approximation removes the tension on the abdominal wall closure and, therefore, decreases the rate of abdominal wall dehiscence as well as bladder dehiscence. It has been widely accepted that the older child with bladder exstrophy, or the redo bladder exstrophy in particular, certainly benefits from pelvic ring approximation. However, the point at which the pelvis is not as malleable as in the newborn is unclear.

This issue addresses the role of pelvic osteotomy in both initial bladder closure as well as redo bladder closures for exstrophy. What is clear from this issue is that there are many factors that should be considered when deciding whether pelvic osteotomy is helpful for bladder exstrophy closure.

I applaud Dr. Gearhart for his guidance in putting together this issue with an excellent group of contributors. I also applaud the Johns Hopkins exstrophy team, as they have continue to share their experience and data so that we can all learn from their unique patient population.
Modern Pelvic Osteotomy in Treatment of Bladder Exstrophy

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The use of pelvic osteotomies as an auxiliary method to treat patients with exstrophy dates from the mid 50’s, but the modern era of pelvic osteotomy began in the late 1980’s with the widespread use of the anterior innominate osteotomy described by Sponseller.\(^1\) This technique has several advantages over the classic posterior pelvic osteotomies:

1. Easy and safe anterior approximation of the pubic bones and application of an external fixator (which can be readjusted without other surgical procedures).
2. The patient is maintained full time in the same position; it is unnecessary to turn the patient from prone to supine for the urologic procedure.

In patients with extreme diastasis (more than 6cm), or cloacal exstrophy cases, an additional vertical posterior osteotomy of the ilium is performed through the anterior approach, to correct the deformity of the iliac wings.\(^5\)\(^6\) Pelvic osteotomy is indicated not simply because a diastasis exists, but it helps to achieve a urologic goal such as tension-free closure, creation of a symphyseal bar to promote continence, or support of the pelvic floor and reconstruction of the perineum in the older female. It can also treat hip dysplasia which is occasionally associated with exstrophy.

Technique

Here we present a brief technical synopsis of the anterior innominate osteotomy.\(^6\) The osteotomy is usually performed in concert with the genitourinary reconstruction. It starts by positioning the patient supine with the pelvis slightly elevated on folded towels and preparing the back and front of the child’s lower body. A sterile barrier drape is placed over the bladder. An oblique incision measuring about 5cm is made 1 to 2 cm distal to the anterior superior iliac spine (Figure 1). The lateral femoral cutaneous nerve is identified and protected (Figure 2). Each side of the pelvis is exposed subperiosteally from the iliac wings to the sciatic notch, posteriorly to the portion of the ilium near the sacroiliac joint and caudally to just above the triradiate cartilage. The periosteum is dissected free from the sciatic notch.

A small window of subperiosteal exposure is developed on the lateral side of the ilium to visually control the osteotomy and pin insertion sites. A transverse iliac osteotomy is performed at a level halfway between the anterior superior and anterior inferior iliac spines, with an oscillating saw. The inferior segment of the pelvis as a whole should then be able to be moved medially (Figure 3A).

For children older than 1 to 2 years or those who present with a wider diastasis (especially cloacal exstrophy) or a previously failed prior closure a posterior iliac closing-wedge osteotomy is added.\(^5\)\(^7\) This hinge greenstick osteotomy is performed just lateral and parallel to the sacroiliac joints, using a rongeur and osteotomes to create a trough in the ilium (Figure 3B); the posterior cortex of the iliac wing is preserved to allow them to hinge. The osteotomy is tested by rotating the iliac wings internally; the trough should close (Figure 3C).

Two threaded fixator pins are placed in the inferior pelvic segment and one or two pins are placed in the ilium superiorly (Figure 3C). The incisions are closed and the pins are covered, allowing the urologists to complete the genitourinary repair. After the urologic procedure the pubic diastasis may be closed in different ways, sometimes by the urologist. The closure may be obtained applying horizontal nylon sutures around each pubic bone, tied together with the pelvis rotated inward using manual pressure on the pins. In older or cloacal patients, a two-hole plate is often preferred in order to maintain the correction of the diastasis. Older children, patients with cloacal exstrophy and failed exstrophies who present with an extreme diastasis may require staged pelvic closure.\(^7\) (Figure 3D)

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The lower abdominal wall is closed. The external fixator bars are applied between the pins to hold the pelvis in the corrected position. The patient remains supine for 4-6 weeks with each leg individually wrapped in light skin traction. A knee immobilizer may also be useful here.

Radiographs are taken after 7 to 10 days. If a near-complete reduction of the symphyseal diastasis to less than two centimeters has not been obtained, the symphysis can be gradually approximated with use of the external fixator bars. The fixator is left in place for six weeks. After adequate callus is radiologically demonstrated at the osteotomy sites the pins can be removed under sedation.

Orthopedic complications have occurred in less than 5% of the patients according to Okubadejo et al. A quarter of the complications were bony complications on the osteotomy site (non-union, delayed union). The earlier technique, posterior vertical osteotomy, was sometimes followed by unique complications such as sacroiliac joint arthritis as the osteotomy erroneously crossed the joint and leg length inequality after posterior osteotomy. All bony complications were treated and resolved with satisfactory results. It is also important to tell parents preoperatively that some increase in the diastasis with growth is inevitable, but it should still remain more corrected than an unoperated pelvis.

The other complications were neurologic, most commonly transitory femoral nerve injuries (peroneal, sciatic nerve, and superior gluteal nerve have also been registered less commonly). The authors believe that the femoral lesions are secondary to the increased tension on the inguinal ligament by the pelvic repositioning (minimized by addition of a posterior osteotomy, in older children). All patients with femoral nerve palsy recovered completely within 4 to 11 months.

Complications of traction, such as pressure sores and compartment syndrome are uncommon and may be avoided if the patients’ legs are not tied too tightly or against each other (“mummy wrap”).

The rarest complication reported was deep infection and late infections of plate. Infection of plate is attribute to limited skin coverage and proximity to the urethra. The plate should only be removed after the osteotomy is well healed. Physical therapy may be helpful in resuming ambulation after the prolonged interval.

References

Pubic Ramotomy in the Exstrophy Population

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The primary aim of any exstrophy-epispadias surgery is to reconstruct an adequate and functional bladder and urethra. Epispadias repair is normally done as a second stage procedure, however, it has been suggested recently to perform it at primary operation.1

A number of pelvic osteotomy techniques have been described with the aim of reducing the symphyseal pubic diastasis, and, therefore, easing abdominal wall closure. To achieve utmost urinary continence, it is of functional importance to transpose the reconstructed bladder into an intra-abdominal position and to establish a retro-symphysial angle of the posterior urethra. For these reasons the role of pelvic osteotomies is well established, however, from the orthopedic point of view, approximation of the pubic symphysis has only a limited effect on gait, as pubic diastasis recurs with time despite the type of osteotomy.

In an attempt to simplify the procedure, we developed bilateral osteotomy of the superior ramus of the pubic bone alone, which can easily be performed by the pediatric urologist and only slightly increases the overall operative time. Since our limited but encouraging experience with this technique in the late 1980s, we now perform pubic ramotomy in almost all our exstrophy patients.2

Patients from the local population normally undergo surgery between day 3 and 7 after birth, but patients having been sent by a caritative organization from Africa always undergo late surgery due to late referral to our center.

Once general anesthesia is established, the edges of the exstrophic bladder are freed from the abdominal wall, the bladder is closed and the posterior urethra is reconstructed according to the method of Jeffs.3 When urogenital reconstruction is completed, the abdominal skin and its subcutaneous tissues are widely mobilized bilaterally. The rectus muscle and its insertion into the pubic bone are located, and its sheath is opened at the lateral border. The superior ramus of the pubic bone is easily approached through this window. The insertions of the gracilis muscle and its insertion into the pubic bone are located, and its sheath is opened at the lateral border. The superior ramus of the pubic bone is easily approached through this window. The insertions of the gracilis muscle and its sheath are not tied too tightly or against each other (“mummy wrap”).

In the cloacal exstrophy patient, the innominate bones are tilted toward the midline and approximated with wide skin mobilization. The plate should only be removed after the osteotomy is well healed. Physical therapy may be helpful in resuming ambulation after the prolonged interval.

At this stage of the procedure care must be taken not to interpose the reconstructed posterior urethra between the approximated symphysis. After placing 2 wound drains over each osteotomy the rectus muscles and (continued on next page)
their sheaths are easily joined and sutured together using multiple resorbable sutures. The generously mobilized skin flaps are then approached without tension and sutured together with fine non-resorbable sutures.

Prophylactic broad-spectrum antibiotics are administered for 2 weeks. The upper legs are immobilized in adduction with medial rotation of the hips using initially a “Y shaped” plaster cast placed on the inferior legs. Recently plastic orthosis prepared before the intervention has replaced the plaster cast.

Considering the dysmorphic pelvis in the exstrophic bladder patient, anterior osteotomy of the superior ramus of the pubic bone seems to be an easily performed efficient method to facilitate bladder extrophy repair and it can be performed by the pediatric urologist. No further skin incisions or turning of the patient are necessary.

References

Lower Extremity and Pelvic Immobilization in the Exstrophy Patient

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Need for Secure Immobilization

It has long been recognized that regardless of which exstrophy repair is used, a successful initial bladder closure is critical to appropriate bladder development and the achievement of continence in patients with bladder extrophy. A number of factors have been correlated with failed exstrophy closures, including the presence of wound infection, inadequate urinary drainage, and postoperative abdominal distention. A secure and properly stabilized pelvic ring closure is also essential to successful bladder closure, as premature separation of the pubis can place significant tension on the newly closed bladder and result in bladder prolapse or dehiscence. Stabilization of the newly closed pelvis relies on the proper application of post-operative traction and immobilization of the lower extremities and pelvic ring regardless of whether or not osteotomy is performed. The application of traction to the lower extremities effectively stabilizes the closure during this critical initial phase of healing. The need to immobilize a child in traction for up to 8 weeks is a significant undertaking that poses some unique challenges and requires multi-disciplinary cooperation. Traction that is either inadequate or placed improperly can have disastrous results both in terms of surgical outcome and the development of complications.

Types of Traction

We recently analyzed our exstrophy population and compared the various methods of pelvic immobilization and their impact on surgical outcomes. The optimal type and duration of immobilization depend on the age of the child and whether or not osteotomy was used. Of 194 patients identified, 93 underwent primary bladder closure with osteotomy. Twenty-four patients were closed on or before three days of age. Of these 24 patients, 14 closures were successful (58%). Fifteen patients were immobilized with an external fixator and 6 to 8 weeks of modified Buck’s traction. Fourteen of those 15 (93%) had a successful outcome. Two patients were immobilized with a “mummy-wrap” and both failed initial closure. Similarly, all 4 patients immobilized with a spica cast and 3 patients immobilized with 4 weeks of modified Bryant’s traction failed initial closure.

One hundred and one patients were identified who underwent primary bladder closure without osteotomy. Of these patients, 77 underwent primary bladder closure on or before 3 days of age. Thirty of the 77 patients (39%) had a successful primary bladder closure. Forty-six patients were immobilized with 4 to 6 weeks of modified Bryant’s traction and 30 of the 46 (65%) had a successful outcome. Twenty-five patients were immobilized with “mummy wrapping” and all failed initial closure. Similarly, all 5 patients who were immobilized with a spica cast and the one patient who was not immobilized postoperatively failed initial closure.

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Patients who underwent delayed primary bladder closure (greater than 3 days of age) had a greatly improved surgical outcome if osteotomy was performed at the time of surgery (81% vs. 33% without osteotomy). Again, the highest surgical success rates were achieved with an external fixator and 6 to 8 weeks of Buck’s traction, or with 4 to 6 weeks of modified Bryant’s traction depending on the use of osteotomy. The greatest rates of surgical success in those undergoing reclosure were seen in those who had osteotomy and were immobilized with an external fixator and 6-8 weeks of modified Buck’s traction. Universally, if osteotomy was omitted in the reclosure patient the outcome was poor regardless of the method of immobilization.

The results of this study clearly indicate that osteotomy performed at the time of bladder exstrophy closure greatly improves the surgical outcome in each patient group examined. The impact on surgical success, however, was much more marked among patients undergoing delayed or redo bladder closures. This study also demonstrates that among bladder closures performed with osteotomy, the highest surgical success rates are achieved when the patient is immobilized postoperatively with an external fixator and 6 to 8 weeks of modified Buck’s traction. Among initial (early) bladder closures performed without osteotomy, the highest surgical success rates are achieved when the patient is immobilized postoperatively with 4 to 6 weeks of modified Bryant’s traction. We, therefore, advocate routine immobilization of the pelvis and lower extremities with 4 to 6 weeks of modified Bryant’s traction (no osteotomy), or an external fixator and 6 to 8 weeks of modified Buck’s traction (osteotomy) following exstrophy closure. While no form of immobilization will protect a poorly performed exstrophy closure, we do believe that lower extremity traction is the most effective way to protect an otherwise solid closure from the shearing forces exerted by abduction and movement.

**Care of Patient**

The surgical team is responsible for the initial application of the traction apparatus in the operating room. Points of emphasis include the following: 1) The legs should be independently wrapped; 2) The wraps need to be placed judiciously with respect to pressure; and 3) The knees should not be hyperextended. The initial traction should be applied in the operating room and verified following transfers.

The state of traction and immobilization is completely unnatural for the child and resistance can be expected. The child’s reaction is age dependent. While infants will often become quiescent within the first 10-14 days, the acceptance by older children is more variable. Regardless of the age or type of traction, appropriate analgesia and anxiolysis is crucial to success. Routine assessment of pain and anxiety is an important responsibility of the surgical team and nurses. The involvement of a dedicated Pediatric Anesthesia Pain Service is an extremely valuable adjunct.

The immobilized child in traction following exstrophy closure requires the most dedicated and diligent nursing. Wound, pin-site, and perineal care are important in preventing infection and skin breakdown. Neurovascular checks of the lower extremities are important in identifying and avoiding iatrogenic injuries. Parents play an important role in the care of their children even while they are immobilized. Time should be invested in educating the parents on the need for the traction as well as trouble-shooting.

**Complications**

Complications can develop secondary to both immobilization and traction. The prolonged post-operative immobilization makes these children susceptible to skin breakdown, constipation, and atelectasis. Constipation and subsequent ileus can cause serious abdominal distension that could jeopardize the closure. The prophylactic use of stool softeners and fiber supplements is appropriate to prevent this.

Traction on the lower extremities can cause skin breakdown and neurovascular injury. Spica casts and mummy wraps are associated with higher rates of skin breakdown than modified Buck’s or Bryant’s traction. The use of individual non-compressive wraps helps to prevent ischemic skin necrosis. Neurovascular checks of the distal extremity should be performed daily. Compartment syndrome and arterial vascular thrombosis are fortunately uncommon. Okubadejo and Gearhart reviewed the orthopedic complications of exstrophy management in 624 patients from Johns Hopkins. Twenty-six (26) or 4% complications were noted of which 4 were identified as being specifically caused by traction. Three of these complications were pressure necrosis, while one was a bilateral lower extremity compartment syndrome.

**References:**

Reoperative Osteotomy in Exstrophy Surgery
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Despite the improvements in surgical management of bladder exstrophy and cloacal exstrophy in recent decades, exstrophy surgery remains an imperfect science and there remains a subset of patients in whom the initial closure fails. Estimates of success rates vary widely depending on institution, but all patients with unsuccessful initial outcomes face the daunting prospect of reoperation, with the attendant decreases in the chances of long-term continence with the native bladder. Still experience has shown that reoperation can be successful, but the surgeon must recognize that this is aggressive surgery that requires deep dissection of the pelvic structures and full mobility of the bony pelvis, with effective post-operative immobilization. To this end, repeat osteotomy is usually a pre-requisite to successful reoperative exstrophy surgery.

A review of the experience with repeat pelvic osteotomy (RPO) at Johns Hopkins Hospital identified 56 patients who underwent RPO in the course of classic bladder exstrophy care. All of these patients had previously failed at least one primary closure of the bladder, some had failed multiple reconstructive attempts, and all had undergone at least one previous pelvic osteotomy. RPO was performed at a mean age of 23 months (range: 1-72 months). The type of osteotomy used during the RPO was anterior innominate in 24 (44%), combined vertical and transverse innominate in 20 (36%), and posterior iliac in 11 (20%).

Of the 56 patients in the series, only 5 (9%) required additional closure after re-operation combined with RPO. On the other hand, of the 18 patients who failed their second bladder closure procedure and therefore required a third closure, 13 (72%) had not had an osteotomy at the time of the second closure. This suggests that RPO, when combined with re-closure of failed primary repair, improves the outcome of this complex procedure.

Although there would be reasonable concerns over the potential for post-operative complications related to repeated surgical insults to the bony pelvis, it appears that RPO-related complications are relatively uncommon. Ninety-five percent of the patients in the above series had a normal gait; the 3 patients with abnormal gait had non-union of the pelvic bones after osteotomy. This was treated by bone grafting, internal fixation, and repeat osteotomy, respectively. Five patients had superficial fixator pin site infections, which responded to local pin care and oral antibiotics. One patient developed osteomyelitis of the pin site and hip joint 16 months after osteotomy, treated with open incision and drainage, and one patient required a readjustment of the external fixators to correct asymmetry of the pelvic girdle. Both of these patients had normal gait after recovery.

Although the results of RPO in this series were excellent, it should be remembered that these procedures were performed by a highly experienced senior orthopedic surgeon who has spent decades developing these techniques, working in concert with a multi-specialty exstrophy team. RPO remains a daunting surgical challenge. The complications of osteotomy include pelvic asymmetry, proximal migration, bony non-union, and nerve palsy. Techniques to reduce bony complications include placement of the osteotomy lateral to the sacral ala, intraoperative assessment of symmetric iliac mobility, confirming adequate bony apposition at the osteotomy site, and bone grafting along the inner pelvic wall to prevent medial migration. Femoral nerve injury may be related to tension on the inguinal ligament from pubic medialization; combining posterior wedge osteotomy with anterior osteotomy may reduce this tension. Avoidance of sciatic nerve injury requires careful dissection around the sciatic notch, which can also minimize peroneal nerve palsies.

Cloacal exstrophy represents an extreme variant of the exstrophy-epispadias complex, and the bony pelvic defect is more severe in cloacal exstrophy than in classic bladder exstrophy. These patients tend to have a more extreme pubic diastasis, greater pelvic asymmetry, sacroiliac joint anomalies, and hip dislocation. In patients with extreme diastasis, which is often seen in cloacal exstrophy, gradual reduction is performed prior to soft tissue repair using external fixation; in some cases, internal fixation using an intrasymphysial stainless steel plate is required. Given the severity of the bony pelvic lesion, the orthopedic consequences of RPO in this patient population might be expected to be more significant than those of the classic exstrophy population.

Among the 65 patients with cloacal exstrophy in the Johns Hopkins exstrophy database, 15 underwent RPO as part of their treatment for failed primary closure of cloacal exstrophy. Most (80%) of these patients failed their primary closure due to dehiscence. Of the RPO procedures, the osteotomy technique was combined vertical iliac and transverse innominate in 6 (40%), transverse innominate in 6 (33%), and posterior iliac in 3 (20%). Major complications after RPO in this group included infection of the intrasymphysial plate requiring removal in 2 patients. Minor complications included superficial pin site infection in 3 patients, and 1 patient who required replacement of the screws in his intrasymphysial plate. In general, however, orthopedic outcomes after RPO in the cloacal patients were relatively good. Six of the patients now walk with a normal gait, 4 walk with a limp, 1 utilizes a walker, 2 are in leg braces, and 2 are wheelchair bound. None of our cloacal exstrophy patients failed after reoperation combined with RPO as a result of dehiscence or pelvic organ prolapse.

Patients who are older at the time of RPO appear to have less diastasis at follow-up. Five cloacal exstrophy patients underwent pubic re-approximation with a titanium plate and had completely stable diastasis, and in general, more stable approximation does occur when an intrasymphysial plate is used. However, the primary benefit of the RPO lies in facilitating a successful surgical reconstruction, and not in achieving long-term, complete re-approximation of the pubic rami.

Repeat pelvic osteotomy is a critical component of reoperative exstrophy surgery, and in experienced hands can be performed safely and with superb results. The advantages of having a fully mobilized pelvis, which allows the bladder to be adequately dissected and dropped into the deep pelvis, cannot be overstated. Success can be achieved in these difficult patients but close cooperation and surgical coordination between pediatric urologist and orthopedist is essential.

References
Postoperative Analgesia for Bladder Exstrophy

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In the perioperative period, epidural infusions of local anesthetics can provide excellent analgesia with a high margin of safety in all pediatric age groups. Epidural catheters are commonly introduced into the epidural space at the caudal, lumbar, or thoracic level prior to the surgical intervention, while the child is receiving general anesthesia. The tip of the catheter is positioned close to the dermatomes affected by the surgical intervention. Concentration and volume of the administered local anesthetic are higher intraoperatively, resulting in a sensory, motor, and autonomic blockade. For most surgical procedures infusion rates and concentrations of the regional anesthetic can be lower in the postoperative period, as sensory blockade is sufficient for analgesia. For the bladder exstrophy patient the additional maintenance of at least a limited motor block is desirable as it decreases the patient’s ability to place distracting forces on the surgical incision site. A fine balance is required between achieving this goal and avoiding toxic side effects.

Caudal insertion of the catheter for epidural analgesia is most common. The sacral hiatus can easily be palpated in children, and, in children less than 20 kg, the catheter can be advanced cephalad to the lumbar region. As catheters that remain in place for more than a few days, particularly those placed at the caudal site, increase the risk of bacterial colonization and possible infection it is recommended to tunnel the catheter. Epidural infusions have been maintained for up to 240 days without serious local or systemic complications. Clearly, vigilance and excellent sterile techniques during placement and maintenance of the catheter are required.

In many centers the local anesthetic of choice for epidural analgesia is either ropivacaine or bupivacaine. Bupivacaine and ropivacaine infusion rates beyond 0.4 mg/kg/h are usually not recommended as seizures and cardiac arrhythmias have been observed at higher doses. As neonates and infants have an immature liver metabolism and decreased concentrations of serum binding proteins, predisposing them to local anesthetic accumulation and toxicity, continuous infusion of bupivacaine need to be limited to 0.2 mg/kg/h. Pharmacological studies of local anesthetic epidural infusions in infants have not extended beyond the immediate postoperative period with some patients demonstrating increasing total plasma concentrations at the end of the study period. Whereas the measurement of bupivacaine or ropivacaine serum concentrations is essentially unavailable in clinical practice lidocaine can be readily measured in most hospital laboratories permitting daily measurement of plasma levels and adjustment of infusion rates until an equilibrium has been reached. Infusion rates are initiated at 1.5 mg/kg/h for infants and older children and are adjusted to 0.8 mg/kg/h for neonates.

Local anesthetics in the epidural infusion can be supplemented by opioids and clonidine with the goal of increasing efficacy of analgesia while restricting the dose of the individual components to avoid toxicity. A high concentration of opioid receptors in the spinal cord permits administration of smaller doses of the opioid compared to the parenteral route. In the newborn, sole administration of local anesthetics, at least when initiating therapy is advisable, as the risk of opioid-associated respiratory depression is ever-present. The α₂-agonist clonidine can modulate and down-regulate the central nervous system’s response to a painful stimulus. Addition of clonidine has been shown to prolong the duration of analgesia and sedation postoperatively. A mild sedative state is desirable for the bladder exstrophy patient as it will contribute to immobility.

Although epidural infusions may provide adequate analgesia and a mild motor block even for long periods of time, children, particularly of the toddler age, may remain agitated. The intimidating hospital environment, restriction of movement to prevent the dislodgement of various tubes, side effects from therapy like pruritus, underlying temperament seem to contribute to their misery. The need to administer additional sedatives in this situation should not be viewed as a failure of the primary therapy but as another component in the multi-modal postoperative management that these patients require. Benzodiazepines diphenhydramine and butorphanol are commonly used sedative agents. Ideally, the child will have periods of quite wakefulness between sleep. An environment with careful monitoring and an experienced nursing staff are required in these situations as these agents can cause synergistic sedation and respiratory depression with drugs administered epidurally.

References
Osteotomy - Who Can Get by Without One?

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Our best clinical practices, lead us to seek our goals for the patient, with minimal risk and in looking at the daunting challenge of treating the child born with bladder extrophy, it seems impossible to many. Pelvic osteotomy seems to pose an insurmountable risk to many, but I believe it may well be the added complexity in assembling the additional team of surgeons that leads many to avoid the adjunctive surgery. Is this really in the best interest of the patients? Our primary immediate goal is successful closure of the bladder and the abdominal wall, with a minimal risk of dehiscence, and/or prolapse of the bladder. Closure without osteotomy was first supported by Julian Ansell in 1979. He postulated that a substance termed ‘relaxin’ might be responsible for allowing the malleability of the pelvis, and proposed that if the iliac wings could be manually manipulated to allow the pubes to squeeze firmly on the index finger placed in the child’s urethral plate, primary closure could be done without osteotomy. His series used osteotomy in children who were being closed at an earlier age. In spite of the fact that his clinical results showed a substantial initial dehiscence, his hypothesis has been used as a support for the closure of infants without osteotomy. Subsequent molecular studies have failed to show any evidence for a substance of hormonal nature to resemble a ‘relaxin’.

Based on these initial results, many have avoided osteotomy, and although no prospective studies have been done, even in the largest centers with and interest in extrophy, the early results of closure without osteotomy have shown dehiscence rates as high as 10% - this in the era of John Duckett!

An additional objective of osteotomy is as an adjunct to placing the bladder deeply within the pelvis, and allowing the posterior urethra to be more surrounded by the urogenital diaphragm. Recent imaging studies (MRI) reported by Halachmi, have shown that these objectives are achieved when osteotomy is used as a part of complete closure in the newborn period.

When then, should the child be closed without osteotomy? I believe that the circumstances in which this may be accomplished should be carefully selected, and rare. In selecting such a relatively unique population, I would suggest that all potential circumstances be optimized. First, that the child be very young, this to take advantage of any enhanced mobility of the newborn’s skeletal system, and also, to minimize the potential for bacterial colonization of the urinary bladder and surrounding structures. Secondly, that the abdominal wall defect and diastasis of the pubes not be excessively large. Thirdly, that there be no accessory components of the defect, such as duplication of one of the genitourinary organs – a rare but reported event.

These optimal conditions for closure without osteotomy should focus on the known variable aspects of the extrophy defect, each of which plays a part in the achievement of a successful initial closure, as well as leading to the most optimal potential for later success in achieving both urinary continence, and optimal sexual performance. These include: 1. large bladder plate – I use my finger on the bladder plate itself, to try to immerse it into the pelvis, when evaluating the child in the nursery; 2. pubic diastasis that is not excessively wide – there is considerable variation in the degree of pubic diastasis from 1-2 cm., to those children in whom the bony defect is greater than 4 cm. I would not advise closure without osteotomy in those with a plate and prostatic urethra wider than 3cm. This also can be evaluated in the nursery.

In conclusion, I feel that we should plan to have osteotomy available for all our patients with bladder extrophy, and utilize closure without osteotomy as a default, when we judge that the child’s defect does not appear optimal. This approach does demand that we have collaboration in place with our pediatric orthopedic colleague, since the birth of the child with extrophy, although often anticipated on the basis of antenatal diagnosis, is still often a surprise to all. This collaboration may appear difficult, but most pediatric orthopedic surgeons are familiar with the techniques, which have been widely published (Matos and Sponseller, elsewhere in this issue). Our treatment of these children, who are still facing numerous procedures and challenges, even in the best hands, in the most experienced centers, should still aim at the most comprehensive potential outcomes in both short-term, and long-term goals. The benefits far outweigh the risks.

References