



Bilateral iliac osteotomy in symphysis pubis diastasis together with extrophia vesicalis: A case report and literature review

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ABSTRACT

Classical extrophia vesicalis is a rare congenital defect requiring multidisciplinary surgical reconstruction. It is observed with symphysis pubis diastasis in which orthopedic intervention is required. Reducing pubic distance by applying pelvic osteotomy reduces tension in the organs in the lower abdomen. In the presented study, a 13-month-old male patient, who underwent iliac osteotomy with the diagnosis of symphysis pubis diastasis with extrophia vesicalis, and who was operated together by pediatric surgery and orthopedic departments, and who underwent a second operation due to complications on the 15th day of the post-operation has been discussed as case report along with the literature review.

Keywords: extrophia vesicalis; iliac osteotomy; symphysis pubis diastasis.

INTRODUCTION

Classical extrophia vesicalis is a rare congenital defect requiring multidisciplinary surgical reconstruction. It is seen in one of every 30 thousand childbirths and is 4 times more in baby boys than baby girls.¹ It is formed as a result of the developmental defect of the lower anterior abdominal wall and pubic bones, there is no anterior vesical wall and the posterior bladder opens directly to the middle-lower abdomen. The bladder wall merges with the skin from both sides and urine goes directly into the abdominal wall. Urethral anterior wall defect and epispadias also accompany this anomaly.

In this anomaly, there is also symphysis pubis diastasis.² This space, which is around 0.6 cm in a healthy person of all ages, reaches an average of 8 cm at about age 10 in children with extrophia vesicalis, while it is about 4 cm at birth. In these patients, the width of the sacrum and the length of the posterior (iliac) segment are

normal, while the length of the anterior (ischio-pubic) segment is 30% shorter than normal. In addition, these two segments have undergone external rotation.³

The formation process of the defect begins with the hypertrophy of the cloacal membrane during the development stages of the urogenital sinus during the sixth and tenth weeks of intrauterine, preventing the migration of the mesoderm between the ectoderm and the endoderm. This malformation not only inhibits the development of the lower abdominal muscle structure and pelvic bones, but also makes the cloacal membrane unstable and prone to early rupture. The time and location of the prolapse determine the condition of the case in the extrophy-epispadias spectrum.¹

The treatment is complicated and mostly in several stages. In babies born with Congenital Extrophia Vesicalis (CEV), a series of multidisciplinary reconstructive surgery is required, including closure of the

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anterior abdominal wall, closure of urethra and bladder with the aim of controlling urine so that renal functions are preserved, reconstruction of genital organs for cosmetic and functional purposes and finally reduction of pubic gap. Closing the bladder is essential in the early stages to maintain the patient's kidney function and to ensure spontaneous urination. It is also recommended that pelvic osteotomy be performed in the first 72 hours, when the pelvic bones are flexible. Other surgeries can be left for the next sessions.^{3,4,5}

In this study, the application of bilateral iliac osteotomy for the symphysis pubis diastasis in the second operation of a 13-month-old male patient, who was born with the diagnosis of exstrophy vesicalis and whose first operation was performed on the 5th postnatal day, and the short-term results of this application have been documented as a case report by reviewing the literature information.

CASE REPORT

A 13-month-old male patient was brought to the pediatric surgery department of our hospital for the purpose of operation due to CEV. The patient was consulted to our orthopedic clinic for the closure of anterior pelvic defect. The information about the patient was that the patient was born prematurely at the 32nd gestational week, was operated due to chloacalectomy + high-type anal atresia + bilateral inguinal hernia on the 5th day, and that there were no other additional diseases. In the pelvis and lower extremity physical examination, it was found that the symphysis pubis anterior had a soft tissue defect of approximately 50 x 50 mm. It was observed that both lower extremities were in external rotation and joint range of motion was normal in all joints. No length difference was observed in the extremities. Neuromotor and vascular examination was natural in both lower extremities. In x-ray and CT examinations, it was found that there was 51.5 mm pubic diastasis and no additional bone problem (figure 1a, 1b).

Surgical treatment was planned in the same session by us in order to close the anterior pelvic diastasis in addition to the surgical operation to be performed in exstrophy vesicalis by the pediatric surgery department.

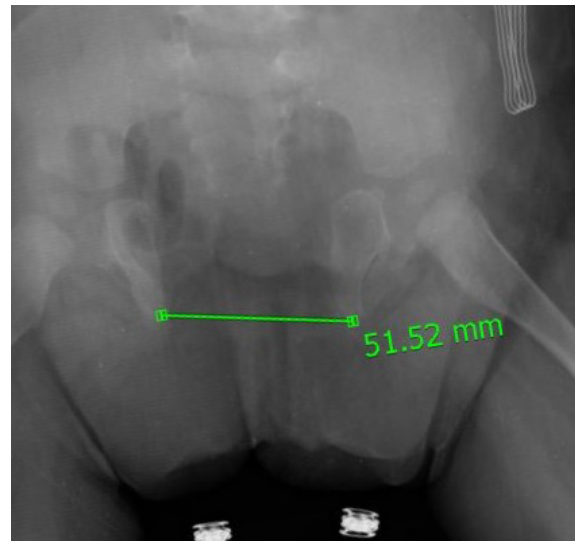


Figure 1a: Preoperative anteroposterior X-ray graphy.



Figure 1b: Preoperative CT imaging.

Right general jugular vein and right femoral artery catheterization were performed after general anesthesia. After general sterilization applications, the operation was started by the pediatric surgery department. Bilateral undescended testis, bilateral inguinal hernia repair, exstrophai vesicalis epispadias complex repair, bladder augmentation (ileocystoplasty), bilateral uretero neocystostomy, bridectomy, right ureterostomy (Mitrafanoff Procedure), colostomy closure, abdomino perinal colon pullthrough (anesthesia), and penile reconstruction were performed. Then, approximately 8 cm incision was made on both iliac bones by us. The sensitive nerves of the iliac apophysis were preserved and divided into two. With the help of the periosteal elevator, the sciatic notch was reached by subperiosteal controlling. Transverse Salter osteotomy was performed on both iliac bones under scopy.

Then, the anterior pubic diastasis was closed and fixed with a cerclage wire. Then, in order to fix the pelvic osteotomy line and increase the stability of the pubic diastasis, a tubular external fixator was applied to 2 supraacetabular areas and 2 iliac bones using Schanz screws. Osteotomy line and symphysis pubis were evaluated with scopy. It was found to be a suitable alignment (figure 2).

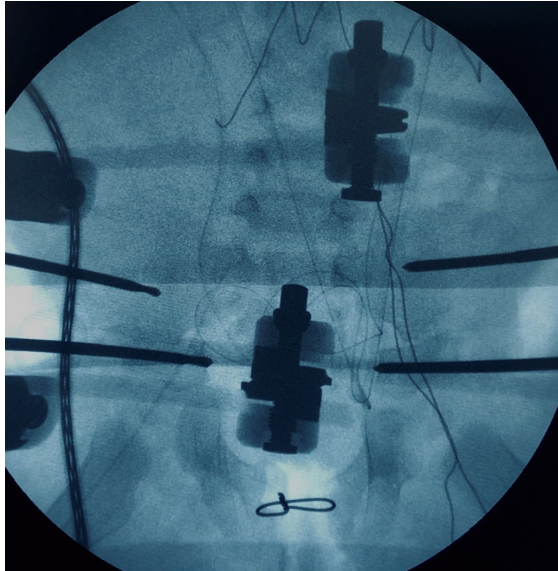


Figure 2: Intraoperative flourescopic imaging at the first surgery.

It was observed that there was a perfusion disorder in the right leg after the operation. It was observed that the distal capillary filling was impaired and the right lower extremity was cyanosed. Although the right femoral artery catheter was removed, vascular circulation did not improve and cardiovascular surgery was included in the operation. Embolectomy was performed with 3 French embolectomy catheters in the right femoral artery and vascular circulation was observed to improve.

The patient was evaluated at the visit made at the 12th hour postoperatively. Neurological examination could not be performed because the patient was followed up as intubated. It was observed that the right femoral and popliteal pulses were taken, the soles of the feet, the back of the feet and the 2nd, 3rd, 4th and 5th fingertips of the right feet were cyanotic. It was deemed appropriate to follow up the patient with heparin infusion. In addition, it was found that the circulation of the glans penis was impaired. The patient was re-operated together with pediatric surgery and 1 vicryl suture was used to bring the cerclage wire in the pubis and the anterior abdominal wall closer. The anatomically combined symphysis pubis was loosened and separated partially.

As a result of the return of the glans penile circulation to normal within the postoperative 15th day, a reconjoin operation was performed using 1.0 pds suture for pubic diastasis. 1 Schanz screw in the right iliac bone was removed due to loosening. It was observed with scopy that the position was appropriate. Dressed follow-up was performed for ischemic pulp necrosis in the necrotic area in the 5th finger pulp was followed by debriding (figure 3).

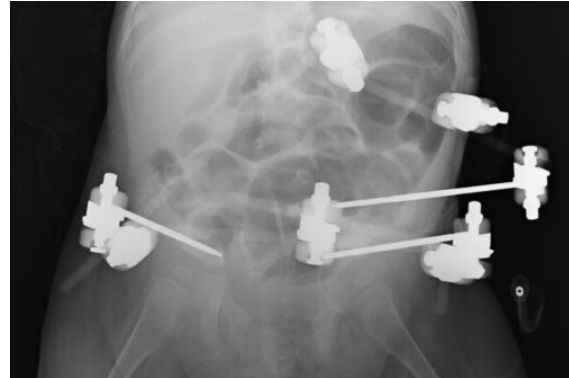


Figure 3: Anteroposterior X-ray graphy on the 15th day.

The external fixator was removed after seeing in the x-ray graphy taken on postoperative day 63 that the pelvic osteotomy line was setting (figure 4).



Figure 4: Anteroposterior X-ray graphy on the 63th day.

It was observed that there was no diastasis and the pelvis was stable. The symphysis pubis gap was measured to be 2.1 cm. It was observed in the patient follow-up that the patient could actively perform hip and knee flexion, but not ankle dorsi-flexion. It was observed that the pulp necrosis in the 2nd, 3rd, 4th and 5th fingers completely healed with granulated tissue.

DISCUSSION

Bladder extrophy is a complex defect involving the skin tissue, muscle tissue, pelvic bone and soft tissues, bladder and external genital organs of the abdominal wall. In the method to be planned in the correction of this anomaly, it is aimed to improve all these different structures functionally and cosmetically as much as possible.^{1,6}

In a study evaluating bone pelvis using three-dimensional CT in patients with classical bladder extrophy, it was found that the iliac wing angle was 11.4 degrees wider than the control group, the sacroiliac joint showed 9.9 degrees external rotation and the pelvis showed 14.7 degrees rotation in the supero-inferior plane. It was seen that the mean pubic diastasis of 0.6 cm in the control group was 4.2 cm and the intertriradiate distance of 4.2 cm in the control group was 6 cm.⁶ In another study in which pelvic bone and soft tissues were evaluated by CT, it was found that the elevator ani muscle was wider in this anomaly and it supported the bladder and posterior urethra less. Again, the pelvic structure has been reported to be substantially flattened in sagittal and coronal sections.⁷ External rotation of the pelvic wings causes lateral rotation of the elevator ani and obturator muscles. In this case report, the patient's 3D tomography was taken and pelvic measurements were evaluated. Accordingly, pelvic rotation in the antero-posterior plane was 41.4 degrees, pelvic rotation in the supero-inferior plane was 142.6 degrees, and the iliac wing angle was 150 degrees on the left and right. These values are not similar to the literature. For this reason, 3-dimensional tomography should be performed in the preoperative period, considering that each patient may have unique anomalies and bone deformities. With these measurements, preoperative planning can be made more effective.

Various single or multi-stage surgical procedures are available for reconstruction of the anomaly, including pelvic osteotomies. Studies do not identify an important orthopedic problem in the long term, whether or not there is osteotomy in children with bladder extrophilia, even with symphysis pubis diastasis. Kantor et al reported long-term orthopedic outcomes in children of different ages operated with different techniques with and without osteotomy, and reported no significant differences in hip joint structure and spine structure.⁸ Posterior pelvic osteotomy provides better continence in older ages. It creates a suitable area for the bladder and urethra to settle in the abdomen. It is also included in the procedure to support the closure of the abdominal wall. Halachmi et al showed a significant improvement in the size and angle of the elevator ani and obturator muscles in MR imaging

after single-stage reconstruction surgery involving anterior iliac osteotomy. They also reported that the support of the posterior urethra and bladder neck increased by increasing the length of the elevator ani.⁹ Performing osteotomy can prolong surgery time and partially increase the risk of post-operative complications. However, it significantly increases the success rate of the primary closure of the anomaly.¹⁰ In the presented case, posterior iliac wing osteotomy was applied to both iliac wings. The fixation of the osteotomy line was provided with an external pelvic fixator. After ensuring the rigidity of the fixation, the cerclage wire passed through the holes drilled through the symphysis pubis strengthened the pubic diastase closure.

A fixator or a pin can be placed in pelvic parts to ensure immobilization during osteotomy. These keep the pelvis stable for 4-6 weeks after the operation together with the external fixator. When the specified time has elapsed, the fixator and pin can be removed after seeing callus formation on the radiograph. The fixation of the pelvis can also be achieved by immobilization of the lower extremities through longitudinal traction (modified Buck's traction). Another method that does not require fixation or traction is pelvipedal plastering with similar success and complication rates. In this method, if the child is to be discharged, it is easier to provide immobilization, while the control of the pelvic tension is not as easy as in the external fixator.¹¹ In our case, an external pelvic fixator was applied for fixation of the osteotomy line in accordance with the literature. Since the strength of the fixation is considered to be sufficient, pelvipedal plaster was not applied in order not to hinder the detection of possible wound problems.

Early operation of the cases of exstrophis vesicalis is important in terms of easier approximation of the pelvic bones, which are still flexible due to maternal hormones.¹² For this reason, in most cases, bilateral pelvic osteotomies are attempted to be applied before the child has completed 72 hours after delivery. Also, early application of primary bladder closure is reported to cause less inflammation and fibrosis in the bladder and less need for urinary correction.¹³ In our patient, the first diagnosis was made in an outer center and the first intervention was made in this center. In the first operation of the

patient, a colostomy was opened in order to ensure stool passage. However, due to the large defect in the anterior abdominal wall and the length of the surgical procedure, the osteotomy operation was delayed to be performed after age 1.

Some early and late complications associated with osteotomy have been described. Okubadejo et al studied complications associated with orthopedic approaches in the follow-up of 624 patients with bladder extrophy, which they carried out in their clinics for 14 years. Complications related to orthopedic surgery were observed in 26 patients (4%). They reported bone structure complications in osteotomy region such as bone non-union, delayed bone union, joint pain or inequality in the lengths of the extremities in 5 patients, neurological complications in 13 patients (femoral nerve damage-possibly traction-induced), early deep tissue infections in 2 patients, and late deep tissue infections in 2 patients.¹⁴

In the retrospective study of 100 patients born with bladder extrophy, Ostrowska et al showed that osteotomy application significantly reduced the rate of op wound site opening after primary closure in addition to the closure of the extrophy.¹⁵ In the presented case, no complication such as non-union of the osteotomy line was encountered. The external fixator was removed on the 63th day of the controls due to callus formation in the osteotomy lines. However, during operation, femoral artery thrombus developed in the patient due to catheterization. Revascularization was achieved as a result of intervention by cardiovascular surgery. Accordingly, ischemic lesions in the 2nd-3rd-4th toes were followed with dressings and healed after granulated tissue. In addition, we think that the neurological deficit that develops in the lower right extremity of the patient is due to thrombus-induced ischemia that occurs more during the operation than osteotomy.

The most common late complication in women born with bladder extrophy is pelvic organ prolapse. In another study of 67 female patients with an average age of 23 who were born with bladder extrophy and 25 of whom underwent osteotomy surgery, it was reported that the size of the initial diastasis was related to pelvic organ prolapse, but osteotomy did not reduce

the risk of prolapse.¹⁶ The most important late complications in men are sexual dysfunction, cosmetic appearance and fertility problems. Reducing symphysis pubis gap can improve the cosmetic appearance of the genital organs but has no effect on sexual function and fertility.¹⁷

Another complication seen in the presented case was the deterioration in penile blood supply that occurred at the postoperative 12th hour. Partial opening of the sutures in the pubic wings was required to improve perfusion. There are examples of penile complications in extrophy closure surgery in the literature, and the pathogenesis of extrophy or epispadias during the repair of the terminal arteries of the penis or increased pelvic pressure after pubic approximation, compression of the pudental vessels and venous congestion has been held responsible.¹⁸ In a study, a 50% reduction in perfusion was detected by approximating the pubis in tissue perfusion measurement performed during laser angiography closure of extrophy.¹⁹ If congestion develops after pubic closure, it is recommended to remove the intrapubic suture and bring it forward to the pubic arm.²⁰

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