



Pelvic Osteotomies in Bladder Exstrophy: Surgical Technique and Case Follow-Up

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DOI: 10.31080/ASOR.2022.05.0620

Received: July 15, 2022

Published: November 07, 2022

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Abstract

The Classic Bladder Exstrophy is part of the Exstrophy - Epispadias Complex; it is a rare condition in which there are anomalies in the formation of the genitourinary system, and in its more severe cases compromises the digestive system and the pelvic ring as well. Orthopedic treatment remains challenging and involves improving the pelvic ring continent and reducing the diastasis of the pubic symphysis through pelvic osteotomies.

We present the case of an 08-month-old male karyotype patient with bladder exstrophy, omphalocele, imperforate anus, sacral anomaly and pelvic alteration, who underwent surgical management for genitourinary and pelvic ring reconstruction; We describe the surgical technique used, the evolution and post-surgical follow-up at 3 years.

Keywords: Bladder Exstrophy; Pelvic Osteotomy; Congenital Abnormalities

Introduction

The exstrophy-epispadias complex groups together a wide spectrum of malformations ranging from classical bladder exstrophy to more severe forms such as cloacal exstrophy. They result from the disruption of the cavitation processes of the pelvic organs and the closure of the pelvic enclosure and the lower abdominal wall during embryogenesis [1]. Classic Bladder Exstrophy has an estimated incidence of 1:30,000 live births [2] and a slightly higher predominance among males, with a boy/girl ratio of 2.3:1 [3].

Although several hypotheses have been proposed for the occurrence of the malformations, the underlying cause remains unknown [4].

Patients with Classic Bladder Exstrophy have widening of the anterior bony pelvis caused by external rotation of the iliac bones along each sacroiliac joint and rotation or eversion of the pubic

rami at their junction with the ischium and ilium [5]. In addition, there is a pubic diastasis which, if untreated, increases from an average value at birth of 4 cm to 8 cm at age 10 years, compared to the normal average of 6 mm at all ages [6,7].

Specific indications for osteotomy as part of a reconstruction include wide diastasis, re-closure after an initial failed repair, patients with persistent abnormal perineal appearance, and uterine prolapse due to a wide pelvic floor [8].

Case report and surgical technique

We present a patient with 46XY male karyotype, who is evaluated for the first time in our service at 08 months of age; he had a history of classic bladder exstrophy, omphalocele, imperforate anus, sacral anomaly and pelvic ring alterations since birth. He had undergone previous surgery for the creation of a left ileostoma due

to the alteration of the digestive tract. At the time of evaluation, he presented external rotation of the lower limbs, significant pubic diastasis and genitourinary exstrophy without definition of the external genitalia (Figure 1).



Figure 1: Pre-surgical clinical presentation.

The evaluation by tomography with reconstruction showed external rotation of the posterior pelvis and both iliac wings, acetabular retroversion and pubic diastasis of 6.5 cm (Figure 2).

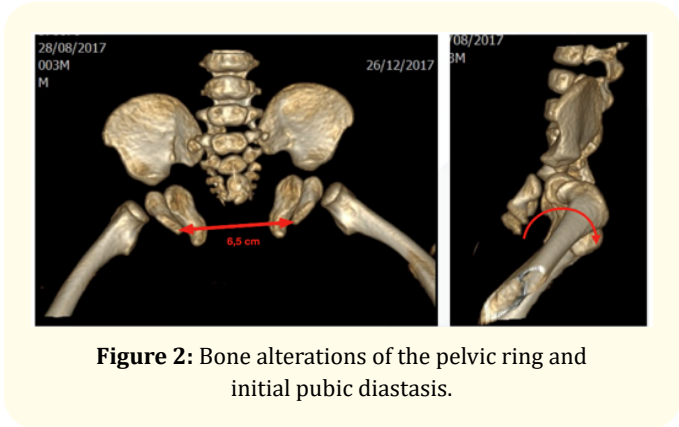


Figure 2: Bone alterations of the pelvic ring and initial pubic diastasis.

In a team meeting between the orthopedics, pediatric urology and pediatric surgery services, it was decided to perform the surgical management for genitourinary and pelvic ring reconstruction in a single surgical act, leaving the resolution of the imperforate anus for a later surgery.

The surgical procedure was performed in 3 stages

In the first orthopedic surgical time, we used the modified Spon-seller [9] technique; through a 5 cm long ilio-inguinal approach, the femoral cutaneous nerve was dissected and protected and iliac wings were sub-periosteally exposed both in the external and internal table up to the greater sciatic notch. Subsequently, we per-

form two osteotomies: a horizontal osteotomy (transversal - Salter type) with a Gigli saw and an incomplete vertical osteotomy (only in the internal iliac table), located 1 cm from the sacroiliac joint, which we do with an arthroscopic drill to create a groove and allow a hinge, in order to close the pelvic ring (Figure 3). Then, under fluoroscopic guidance, we inserted 4 Schantz pins per side: 2 oblique and 2 horizontal and closed the approaches by layers (Figure 4).

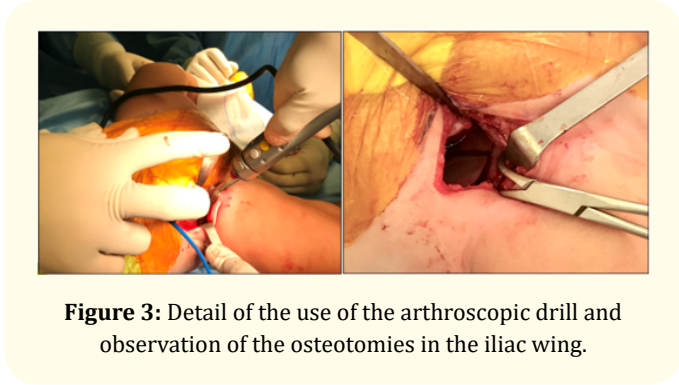


Figure 3: Detail of the use of the arthroscopic drill and observation of the osteotomies in the iliac wing.

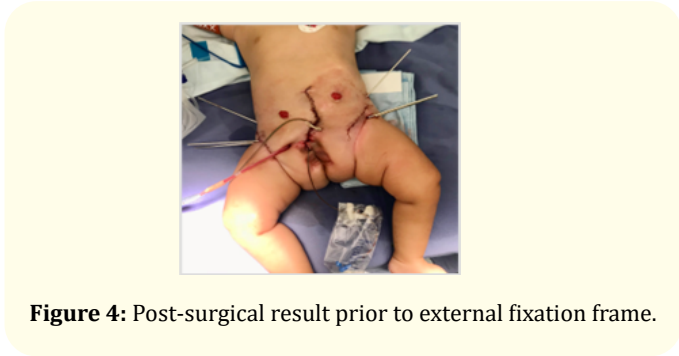


Figure 4: Post-surgical result prior to external fixation frame.

After that, urological reconstruction with cystostomy and urethroplasty was performed.

In the second surgical orthopedic stage, a double external fixation frame was placed, closing the pelvic ring, reducing the pubic diastasis and avoiding abdominal tension. Finally, the lower limbs were bandaged to maintain a skin traction of 800 grams (Figure 5).

Results

The patient was admitted to the pediatric intensive care unit for better management of analgesia and homeostasis, he remained in that unit for two weeks with no major complications except for local irritation in the area of entry of the upper Schantz pins. The



Figure 5: Final post-surgical clinical results.



Figure 7: Post surgical clinical presentation.

traction and external fixation frame was maintained for 8 weeks, after which they were removed under sedation. A tomographic control was performed at 12 weeks, showing a better configuration of the pelvic ring and a pubic diastasis of 3.2 cm (Figure 6).

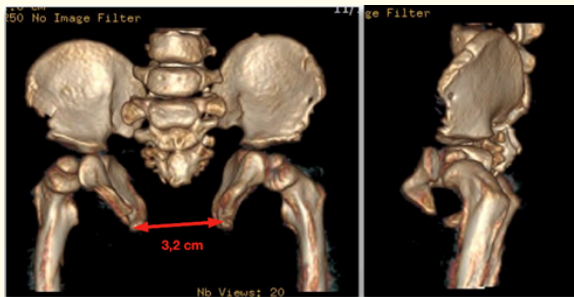


Figure 6: Tomographic control with better configuration of the pelvic ring and decrease of the pubic diastasis.

After confirmation of the favorable urological and orthopedic evolution, an intensive physical therapy and rehabilitation program was started, achieving an independent gait at one year and six months. The last clinical and radiological control was performed when the patient was 3 years and 10 months old; the pelvic correction was maintained, the alignment and rotation of the lower limbs was within normal limits and the patient had no limitations for physical activities (Figure 7).

The complete correction of the genitourinary and digestive alterations in this patient has not yet been completed and we are maintaining clinical and radiological follow-up every 6 months.

Discussion

The main objective of orthopedic reconstruction through pelvic osteotomies is to reduce pelvic diastasis and improve the pelvic ring continent, thus allowing closure of the bladder, the abdominal wall and decrease the previous tension due to anatomical compromise.

Different surgical techniques have been published over the years [10], from the classic posterior approach and 2-stage correction described by O'Phelan in 1963 [11], to the oblique pelvic osteotomies in the most recent years [12]. All surgical techniques have shown favorable results; however, the anterior innominate osteotomy with posterior vertical iliac osteotomy described and modified by Sponseller [9], presents advantages over other conventional techniques that include: less intraoperative blood loss, better apposition and mobility of the pubic branches at the time of closure, tolerance for placement of an external fixator under direct vision, tolerance for safe external fixation in children older than 6 months, and it is not necessary to turn the patient during the operation.

We decided to use this surgical technique, making a modification and changing the rongeur for a 3.5 mm diameter arthroscopic drill for the vertical osteotomy, which allowed us a less invasive dissection of the iliac wing and a more controlled management of the internal iliac table.

Although there were no major complications, those related to external fixation such as infection of the pins entry sites, transient paralysis of the lateral cutaneous nerve of the thigh and delays in

the consolidation process have been described. Other less common complications include pseudarthrosis, lower limb dysmetria, and injuries to the sciatic nerve, femoral nerve, and superior gluteal nerve [13].

Conclusions

Classic bladder exstrophy is a rare pathology with comorbidities that significantly affect organs and systems, especially the genitourinary and pelvic anatomy.

Orthopedic treatment is indicated to improve the pelvic continent as a complement to urological surgeries, to reduce post-surgical stress and avoid urological complications such as prolapse, as well as to improve the patient's function, especially ambulation without excessive external progression.

A multidisciplinary and coordinated approach between specialties is essential; surgical planning and execution are essential to obtain a result that contributes to improving the patient's quality of life.

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