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REVIEW ARTICLE

Bony abnormalities in classic bladder exstrophy: The urologist's perspective

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Abstract *Introduction:* As the primary practitioner managing patients with classic bladder exstrophy (CBE), it is incumbent upon the pediatric urologist to understand the associated orthopedic anomalies and their management.
Methods: A Pubmed search was performed with the keyword exstrophy. Resulting literature pertaining to orthopedics and published references were reviewed.
Results: Anatomic changes to the bony pelvis include outward rotation, acetabular retroversion with compensatory femoral anteversion, anterior pubic shortening, and pubic diastasis. Imaging options have improved, which impacts surgical planning. Surgical approach, including type of osteotomy and method of pubic approximation, is evolving. Most centers employ immobilization after surgery, with external fixation, Bryant's traction, Buck's traction, and spica casting being the most common methods. Orthopedic complications range from minor pin-site infections to neurologic and vascular compromise. Most experts agree osteotomy aids bladder closure beyond 72 h of life, but effect on continence remains controversial. Although no significant orthopedic benefit has been expounded, it may be too early to appreciate improvement in frequency or severity of osteoarthritis or hip dysplasia.
Conclusion: While orthopedic surgeons remain vital to managing exstrophy patients, knowledge of the anatomy, imaging, surgical approaches, and immobilization enable effective communication with parents and other physicians, improving care for these complicated patients.

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Introduction

Since osteotomies were first described, our understanding of the pelvic anatomy has improved, new radiographic technologies have emerged, novel procedures have been developed, and outcomes have been measured. As the pediatric urologist assumes the central role in caring for the patient with bladder exstrophy, it is important to appreciate the bony anomalies accompanying the condition, including the appropriate work-up, surgical options and resulting complications, and impact on the patient's outcome.

Historical perspective

Prior to the advent of performing osteotomy with urologic reconstruction, abdominal closure relied on anterior flaps and the urinary tract was typically managed with diversion [1,2]. The use of osteotomies to correct pelvic bony anatomy in conjunction with bladder closure was first published in 1958. Dr. Shultz planned to close the bladder of a 2.5-year-old exstrophy patient, and Dr. Schwartzmann wanted to perform osteotomies for gait correction during the same admission. On discussion, they recognized the advantage of bringing the pubic bones together before bladder closure, forever changing management of the exstrophy—epispadias complex [3]. In response to the poor outcomes of bladder closure attributed to undue tension from a widened symphysis pubic and outwardly rotated iliac bones, O'Phelan began routinely performing posterior osteotomies in 1957. Posterior osteotomies were performed as a first stage, followed by closure of the bladder and pubic symphysis reapproximation, typically with wire suture, as a second stage. The child was then immobilized in a spica cast for 10—12 weeks until radiologically proven to have mature bone growth at the osteotomy site. Shortly after regaining mobility, the wire suture was removed. While there was little impairment of growth at the osteotomy sites, the pubic diastasis typically recurred. He noted a low urologic complication rate [1]. By 1970, the UCLA group reported its experience in 15 patients. They did not reapproximate the pubic symphysis, decreased the period of immobility to 4—6 weeks, and used a molded body cast for immobilization. While no objective measures were reported, they noted in their comparison of nine patients with osteotomies and six without, that 'urologic procedures were greatly facilitated' [2].

Anatomy

Bony pelvic anomalies can be classified as changes in orientation or dimension. Over time, some of these anomalies evolve. Additionally, changes may occur in the spine and knee.

With improvements in imaging technology, multiple groups have delineated pelvic anatomy (Table 1). While there is no debate as to presence of pubic diastasis or external rotation, hip configuration is somewhat controversial. Some suggest persistent acetabular retroversion [4,7], while others found lesser degrees of retroversion with age [6,9]. There is also an increased angle of femoral anteversion and tibial torsion [6,7,9]. One may conclude that there is probably acetabular retroversion, but it potentially improves over time and does not affect gait as there is compensatory femoral anteversion. External rotation of the posterior and anterior pelvic segments appears to improve with age. Pubic diastasis, however, tends to worsen over time [4].

As different groups have found conflicting data concerning acetabular retroversion, the presence of hip pathology is also unclear. Some report hips that function well over the long-term [9] and no evidence of hip dysplasia [6], while others found radiographic evidence of hip pathology in 35% of their population. Additional radiographic abnormalities were noted in 54% of patients, including: hip subluxation, coxa breva, congenital coxa vara, pistol grip deformities, asymmetric hemipelves, and acetabular retroversion [10].

Bones outside of the pelvis also sometimes form abnormally. Patients with exstrophy may be born with congenital spinal anomalies or develop spinal curvature over time; while this is often clinically insignificant, some patients exhibit functional deficits [10—12]. Spinal abnormalities seen with exstrophy include: dysmorphic sacrum, lumbar sacralization, spina bifida occulta, spondylolisthesis, scoliosis, and kyphosis [10]. Knee pathology has also been described. In a series of 14 patients, 50% had signs of patellofemoral instability, including two with subjective complaints. Radiographically, 71% of patients had pathologic patellofemoral congruency [6].

Various clinical measurements are used to decipher the anatomy of exstrophy children. In Sponseller et al.'s study of children who had not undergone osteotomy prior to evaluation, clinical measurements included angle of foot progression, internal rotation of the hip in extension, external rotation of the hip in extension, and thigh—foot angle. These clinical measures were largely within the normal range, except the angle of foot progression relative

Table 1 Changes to the bony pelvis in classic bladder exstrophy.

Rotational anomalies	Dimensional anomalies
External rotation of posterior pelvis/iliac wings [4,5]	Increasing pubic diastasis [4,5]
External rotation of anterior segment [4]	Shorter anterior pubic segment [4,8]
Coronal rotation of sacroiliac joint [5]	Increased inter-triradiate cartilage distance [4,5]
Acetabular retroversion [4,6,7]	
Convergence of iliac wings [8]	
Femoral retroversion [6,7]	