**Current Management of Bladder Exstrophy**

*Arthur Mourtzinos, MD and Joseph G. Borer, MD*

**Introduction**

In 1987, based on the findings of 10 malformation monitoring systems around the world, the International Clearinghouse for Birth Defects Monitoring Systems reported the prevalence of bladder exstrophy as 3.3 per 100,000 births [1]. Bladder exstrophy affects men an estimated 1.5 to 2.3 times more often than women.

**Etiology**

Perhaps the most prominent theory for the etiology of bladder exstrophy is that proposed by Marshall and Muecke [3] who postulated the basic defect to be an abnormal overdevelopment of the cloacal membrane, which prevents medial migration of the mesenchymal tissue and proper lower abdominal wall development. Subsequent rupture of the cloacal membrane would result in herniation of the lower abdominal contents and the phenotypic appearance of bladder exstrophy or other variants of the exstrophy-epispadias complex.

**Associated Defects**

Exstrophy of the bladder has a constellation of associated anatomical defects involving the abdominal wall, pelvic bones, genitalia, anus, and rectum. Typically, there are no other major organ system anomalies associated with bladder exstrophy compared with cloacal exstrophy, which represents the most severe manifestation of the exstrophy-epispadias complex. In bladder exstrophy, the triangular defect caused by the premature rupture of the abnormal cloacal membrane contains the exstrophied bladder and posterior urethra. The umbilicus lies more caudad than normal, just superior to the cephalad apex of the defect. Inguinal hernias occur in up to 82% of boys and 11% of girls with bladder exstrophy [4]. The perineum is short and broad and the anus is displaced anteriorly, situated directly behind the urogenital diaphragm demarcating the posterior limit of the triangular fascial defect [5].

Patients with bladder exstrophy have characteristic diastasis of the pubic bones from the anterior midline. This is caused by malrotation of the innominate bones in relation to the sagittal plane of the body along both sacroiliac joints. The malrotation of the innominate bones also causes increased distance between the hips and outward rotation of the acetabula and lower limbs [6]. More recently, imaging of the bony pelvis with computerized tomography has detailed the extent of significant external rotation of the iliac wing angle (11.4º greater) and the sacroiliac joint angle (9.9º greater) when compared with gender- and age-matched healthy control subjects [7]. Inferior rotation of the pelvis also was a new finding documented in this report.

The male genital defect is more severe and complex relative to that of the female. The penis is shortened, in part because of the diastasis of the pubic symphysis and because of a marked congenital deficiency of anterior corpora cavernosa tissue. Magnetic resonance imaging (MRI) in adult men found that the anterior corporal length of patients with bladder exstrophy was approximately 50% shorter than that of healthy control subjects [8]. The decreased corporal length relative to normal also has been confirmed on MRI evaluation of anatomical relationships after exstrophy repair in the newborn period [9]. Autonomic nerves course laterally along the corpora cavernosa [10]. Testis function has not been examined in a large group of post-pubertal exstrophy patients, but it is thought that fertility is not impaired by testicular dysfunction.
In women, the clitoris is bifid and the vagina is shorter, but of normal caliber when compared with healthy control subjects. Vaginal dilatation may be required to allow satisfactory intercourse in the mature woman. The fallopian tubes and ovaries generally are normal. Mature women may be predisposed to the development of uterine prolapse, which requires suspension or corrective surgery [11].

Prenatal Diagnosis
Ultrasonography evaluation of the fetus usually allows for an accurate diagnosis of bladder exstrophy. In a review of prenatal ultrasonography examinations with the subsequent birth of a newborn with bladder exstrophy, the following observations were found to be consistent with the diagnosis of bladder exstrophy: absence of bladder filling, a low-set umbilicus, pubic bone diastasis, diminutive genitalia, and a lower abdominal mass that increases in size as the pregnancy progresses and as the intra-abdominal viscera increase in size [12]. At Children’s Hospital in Boston, after the prenatal diagnosis of bladder exstrophy, expectant parents are offered counseling regarding the diagnosis and its implications, advanced obstetrical care, interaction with the urologic team, a tour of the neonatal intensive care unit, and the opportunity to interact with other families who have a child with bladder exstrophy. Obstetrical personnel schedule induction of labor in late gestation with delivery at an institution adjacent to the treating pediatric hospital. Prenatal diagnosis appears to optimize care by affording education and preparation of the expectant parents, scheduled induction of labor, scheduled preoperative evaluation and initial or complete primary repair, and continuity of care with experienced and familiar caregivers. From 1998 through 2003, 10 of 19 newborns with bladder exstrophy cared for at Children’s Hospital in Boston were diagnosed prenatally.

Initial Perinatal Management
After delivery, it is important to limit trauma to the delicate mucosa of the exposed bladder. After the initial plastic clamp occlusion of the umbilical cord at delivery, the clamp should be exchanged for soft cloth umbilical “tape” or silk ligature to limit trauma to the bladder. The bladder then should be covered with a non-adherent film of plastic wrap to prevent irritation from abrasive diapers. The bladder may be irrigated intermittently with sterile saline during diaper changes. At Children’s Hospital in Boston, renal ultrasonography, frontal radiograph of the abdomen, and pelvic MRI are performed in the newborn before complete primary repair is performed within the first 2 days of life.

Surgical Management
Staged approach for repair of bladder exstrophy
The staged approach for functional bladder closure, pioneered and advocated by Jeffs et al. [13] and others [5,14] beginning in the 1970s, has been modified over the past several years. The “modern” staged approach to repair of bladder exstrophy consists of three specific scheduled components. First, bladder, posterior urethra, and abdominal wall closure with bilateral innominate and vertical iliac osteotomy, when indicated, are performed in the newborn period. Second, epispadias repair is performed at 6 months to 1 year of age. Third, bladder neck reconstruction (BNR) and bilateral ureteral reimplantation are performed when patients are between the ages of 4 and 5 years when adequate bladder capacity for BNR and motivation to participate in a postoperative voiding program are documented [5,15]. As part of the staged repair, penile reconstructive techniques strive to fashion a straight and functional penis with a glandular meatus and an acceptable cosmetic appearance.

The goal of the initial stage of the staged approach is successful bladder, posterior urethral, and abdominal wall closure and, in essence, creation of complete (penopubic) epispadias [16]. The need for iliac osteotomy should be rare when closure is performed within the first 72 hours of life. In the newborn, a small, stiff bladder “template” rarely will not be amenable to closure and should be given adequate time to grow to a size that makes closure feasible [17]. Of 19 such patients reported by Dodson et al. [17], primary closure was performed at a mean patient age of 13 months. BNR achieved continence without the need for augmentation or replacement in 47% of these patients. Determining unsuitability for neonatal bladder closure is a judgment only for surgeons with extensive experience. In a review of a 10-year experience of bladder exstrophy management, Oesterling and Jeffs [18] noted the importance of successful initial bladder closure on eventual outcome of the staged approach in these patients. They found that for criteria of bladder capacity at the time of BNR, interval between initial closure and BNR (time to develop sufficient capacity for BNR) and rate of urinary continence, there was a statistically significant difference favoring those patients with versus those without successful initial bladder closure.

In the second stage of the modern staged approach, epispadias is repaired using one of several techniques, typically when the patient is 6 to 12 months of age. Surer et al. [19] have described their experience over a recent 10-year period with the modified Cantwell-Ransley technique. A total of 79 patients with bladder exstrophy were observed (65 repaired primarily and 14 after previous failed repair). At a mean follow-up of 68 months, the penis was horizontal or dangling downward in more than 90% of patients. Urethrocutaneous fistula was noted postoperatively in 11 of 65 (17%) patients who underwent primary repairs, in three of 10 (33%) patients who underwent combined bladder closure with epispadias repair, and four of four who underwent epispadias repair alone after previous failed repair. Urethrocutaneous fistula closed spontaneously in two patients and required repair in 16 [19]. These results are evidence of the diffi-