Counseling Pregnant Women with Bladder Exstrophy

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Counseling Pregnant Women with Bladder Exstrophy

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**Abstract**

Background: Bladder exstrophy is a very rare congenital anomaly characterized by a defect in the closure of the lower abdominal wall and eversion of the bladder mucosa, ureteral orifices, bladder neck, and urethra.

Case: A 37-year-old woman arrived at the emergency room with a missed abortion. She was para 5-0-3-1 and had a breech section 7 years earlier.

Conclusion: We describe patients with bladder exstrophy who have not undergone reconstruction surgery and review the literature on pregnancy planning and potential complications.

**Introduction**

A 37-year-old woman arrived at the emergency room with a missed abortion. The patient was para 5-0-3-1 and had a breech section 7 years earlier. She was diagnosed as having bladder exstrophy (BE) without reconstruction. The pelvic examination revealed a uniformly bifid clitoris with an opening in the urethral plate. The vagina and anus were normal. The pubic symphysis was separated widely. BE is a very rare congenital anomaly characterized by a defect in the closure of the lower abdominal wall [1]. Currently, prenatal diagnosis is possible, and BE can be corrected with a staged reconstruction [1]. However, our patient never underwent reconstruction and always wears diapers. In this paper, we describe patients with BE who have not undergone reconstruction surgery and review the literature on pregnancy planning and potential complications.

**Case Report(s)**

A 37-year-old woman arrived at the emergency room with vaginal bleeding. She was para 5-0-3-1 and had a breech section 7 years earlier. She was diagnosed with bladder exstrophy (BE) without reconstruction. Ultrasonography revealed a missed abortion in the right side of a septated uterus. During the pregnancy, she had recurrent urinary tract infections. After her cesarean section, diastasis developed in the surgical scar. The infant was a healthy male with no anomalies. The patient’s abdominal examination revealed no umbilicus, no muscle layer and only a skin layer in the low midline caesarian scar, and divergent rectus muscles to the pubic symphysis. The pelvic examination revealed a uniformly bifid clitoris with an opening in the urethral plate (Fig. 1). The vagina and anus were normal. The pubic symphysis was separated widely. The patient had worn diapers since birth. After dilation and curettage for the missed abortion, she had no complications.

**Discussion**

BE is a very rare congenital anomaly characterized by a defect in the closure of the lower abdominal wall and eversion of the bladder mucosa, ureteral orifices, bladder neck, and urethra [1]. BE is often associated with public bone anomalies. BE is one of eight very rare defects that occur only a few times per 100,000 births; the others are acardia, conjoined twins, amelia, phocomelia, cloacal exstrophy, cyclopia, and sirenomelia [2]. BE has an incidence ranging from 1:30,000 to 1:50,000 live births [1]. Males are affected 2.8 times more frequently than females and Caucasians 1.7 times more often than Blacks, Hispanics, and other races [3]. BE can occur as an isolated syndrome or as multiple congenital anomalies. The isolated form is more prevalent with increasing maternal age [2]. Congenital anomalies associated with BE include epispadias–exstrophy complex, omphalocele, anal defects, neural tube defects, and skeletal defects, including omphalocele/exstrophy/imperforate anus/spinal defects (OEIS) complex [2]. Overall, the isolated form of BE is more prevalent [2]. BE was first described in 1595; however, its pathogenesis remains unknown [2]. It has been hypothesized that the infraumbilical mesenchyme fails to migrate between the ectodermal and endodermal layers of the cloacal membrane [2]. There are no clear associations with environmental factors, drug exposure, or genetic factors [2]. BE is sporadic and the recurrence rate in families is about 1% [2]. We consulted an urologist and orthopedic and plastic surgeons regarding reconstruction surgery after the dilation and curettage, but they did not agree with reconstruction surgery because the patient might...
develop skin defects, skin fistulas, and vesicoureteral reflux. The latter is very common after surgery. In adults, an osteotomy alone may be insufficient for covering the extreme diastasis, because the bony pelvis is rigid with a wider diastasis. In selected cases, a small adult female with BE may undergo a pelvic osteotomy [4]. Without reconstruction, malignant bladder and kidney tumors are more common between the third and fifth decades of life [2]. Pelvic organ prolapse in BE has an incidence rate of 18% [5]. The modified posterior Prolift® procedure is useful for BE patients with symptomatic pelvic organ prolapsed [5]. High-resolution ultrasonographic can diagnose BE at the 15th week of gestation. Non-visualization, a low-set umbilicus, and a lower abdominal mass are the key sonographic findings [6]. However, an antenatal diagnosis is made in only 25% of cases; of these, the mother opts to terminate the pregnancy in 21–80% of cases [6]. Pregnant women with BE reconstruction can develop urinary tract infections, hydronephrosis, and preterm labor [7]. However, a cesarean section may minimize any uterine prolapse and other urinary tract complications or vascular problems involving the augmented ileum [7]. Based on our case and reported reconstruction cases, we encourage pregnancy planning with a team consisting of urologists and gynecologists. The termination rate depends on the team approach and a pediatric urologist can educate the parents regarding the positive results of reconstruction surgery for BE. Before termination is considered, fetal magnetic resonance imaging (MRI) should be performed and the parents should consult a pediatric urologist [6]. Psychosocial problems and the quality of life in BE should be considered when deciding to terminate. Based on long-term follow-up, 5 of 9 females with BE married, 7 of 10 females were active professionals, and their education levels and employment rates matched those of the general population [8]. Six women were sexually active; two women had dyspareunia; and one woman refused sexual intercourse because of genital prolapsed [9]. Two pregnancies were spontaneous, while one resulted from artificial insemination due to partner infertility [8]. Our patient became pregnant five times spontaneously and had four abortions. The greatest inconveniences in the lives of BE patients are the need to wear diapers for urinary incontinence and recurrent urinary infections [9]. Irrespective of gender and surgical procedures, BE patients had a poor quality of life in terms of general health as compared with the normal population [8]. If BE is diagnosed using prenatal sonography, we recommend reconstruction surgery in the newborn period using a team approach and counseling for the parents regarding health concepts and the quality of life of women with BE women based on our experience.

References

Fig. 1 The pelvic examination revealed a uniformly bifid clitoris with an opening in the urethral plate.

Illustrations

Illustration 1

Fig. 1 The pelvic examination revealed a uniformly bifid clitoris with an opening in the urethral plate.
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