

Bladder exstrophy: reconstructed female patients achieving normal pregnancy and delivering normal babies

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ABSTRACT

Purpose: Bladder exstrophy (BE) is an anterior midline defect that causes a series of genitourinary and muscular malformations, which demands surgical intervention for correction. Women with BE are fertile and able to have children without this disease. The purpose of this study is to assess the sexual function and quality of life of women treated for BE.

Materials and Methods: All patients in our institution treated for BE from 1987 to 2007 were recruited to answer a questionnaire about their quality of life and pregnancies.

Results: Fourteen women were submitted to surgical treatment for BE and had 22 pregnancies during the studied period. From those, 17 pregnancies (77.2%) resulted in healthy babies, while four patients (18.1%) had a spontaneous abortion due to genital prolapse, and there was one case (4.7%) of death due to a pneumopathy one week after delivery. There was also one case (5.8%) of premature birth without greater repercussions. During pregnancy, three patients (21.4%) had urinary tract infections and one patient (7.14%) presented urinary retention. After delivery, three patients (21.4%) presented temporary urinary incontinence; one patient (7.14%) had a vesicocutaneous fistula and seven patients (50%) had genital prolapsed. All patients confirmed to have achieved urinary continence, a regular sexual life and normal pregnancies. All patients got married and pregnant older than the general population.

Conclusions: BE is a severe condition that demands medical and family assistance. Nevertheless, it is possible for the bearers of this condition to have a satisfactory and productive lifestyle.

Key words: bladder exstrophy; outcomes; female; pregnancy

Int Braz J Urol. 2011; 37: 605-610

INTRODUCTION

Bladder exstrophy (BE) is an anterior midline defect with variable expression involving the infraumbilical abdominal wall including the pelvis, urinary tract, and external genitalia resulting in the exposure of the distal urinary tract to the outer abdominal wall (1). Its incidence varies from 1:30,000-1:50,000 live births, being up to 2.8 times more frequent in the male sex and 1.7 times more frequent in caucasians (2-4). There is a risk

of recurrence of 0.5-3.0% in families with one affected subject. This value represents a small risk of recurrence, but it is as high as 200- to 800-fold when compared with the unaffected population (1). No etiological genetic or non-genetic factor has been identified so far; however some chromosomal candidate regions causally related to bladder exstrophy are starting to be identified (5).

The approach of patients with BE is an enormous challenge in Pediatric Urology. Besides the satisfactory closure of the abdominal wall the

treatment must also aim for kidney preservation, urinary continence, and avoidance of complications (6). The treatment for BE consists of two basic options: staged reconstruction and complete primary repair. The staged reconstruction of BE consists of initially closing the bladder plate and posterior urethra, with subsequent steps of epispadia repair and finally bladder neck reconstruction. The complete primary repair involves the closure of the bladder plate and urinary tract reconstruction through colocolostomy or ureterosigmoidostomy (6). Epispadia repair is also done in the same surgical procedure.

The quality of life of individuals with BE is considerably affected by the disease. Children and adolescents present a high level of school education, social integration and even an active sexual life (7). Several studies demonstrated a good satisfaction level with aesthetical results of surgery, but still many patients are not satisfied with their physical appearance, especially with their genitals, avoiding body exposure situations (7-9). Adult women's quality of life has also been assessed, showing satisfactory results regarding social, professional, and sexual life. Nevertheless, urinary incontinence, genital prolapse and psychological distress are present in many cases (10).

Women treated for BE are capable of having normal children. These pregnancies require a great demand of attention from the physicians since they are complex cases (10). The purpose of this study is to evaluate the sexual function, quality of life and pregnancies from the female patients treated for BE in our institution.

MATERIALS AND METHODS

The patient charts of all patients treated for BE from 1987 to 2007 in our institution were reviewed, totaling 30 charts. Those patients were called by telephone and invited to participate the study. The participating patients were interviewed by phone by the performers of this study about the age they had their first pregnancy; the method of delivery; their children's current ages; whether they had an abortion; and whether any of their children have died and by which cause. They were also asked about any post-delivery complications,

whether they had any kind of urinary leakage, and if they were sexually active. The patients' charts were also reviewed to determine to what kind of reconstruction the patients were submitted.

RESULTS

There were found to be 24 female patients, aged 18 or older, treated for BE in our institution, and 14 of them were successfully contacted and agreed to participate in the study. These 14 patients achieved 22 pregnancies, delivering 17 normal children (thirteen males and four females). The age of delivery of the first child varied from 20 to 32 years old (mean = 25.5 years). All babies were delivered through Cesarean section, with the exception of one that had a premature normal birth. The children's ages in 2007 varied from 1.5 to 11 years old (mean = 4.9 years). One child died one week after delivery due to a pneumopathy. There were also four spontaneous abortions related to genital prolapse.

Regarding urinary tract reconstruction, four patients were submitted to staged reconstructions for urinary tract reconstruction, another five were submitted to colocolostomy for the repairment of BE, and the remaining five patients were submitted to ureterosigmoidostomies. All patients had pelvic posterior osteotomies on the reconstruction.

All the patients referred to urinary continence, which was translated as no need for pads, with mild to moderate eventual stress-related leakages. They were all sexually active and achieved normal natural pregnancies. All of them have stable marital lives and six (42.8%) have formal jobs.

Regarding pregnancy complications (Figure-1), there were the already mentioned four cases of abortions (18.1%), three patients (21.4%) presented urinary infections, one patient (7.14%) presented urinary retention, treated with clean intermittent catheterization, one child (7.14%) had a premature, normal birth and had a good recovery. After delivery (Figure-2) there were three cases (21.4%) of temporary urinary incontinence which had spontaneous improvement within 1 year after delivery, one case (7.14%) of vesicocutaneous fistula that demanded surgical correction, and seven patients (50%) had genital prolapse, including one who had to be submitted to a hysterectomy for this condition. The

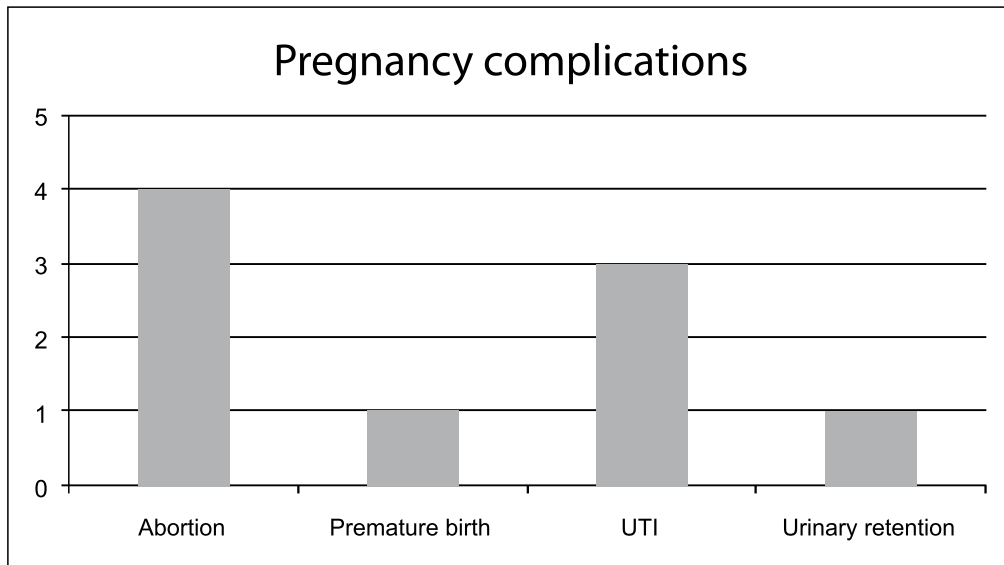


Figure 1 - Pregnancy complications regarding the 22 pregnancies assessed.

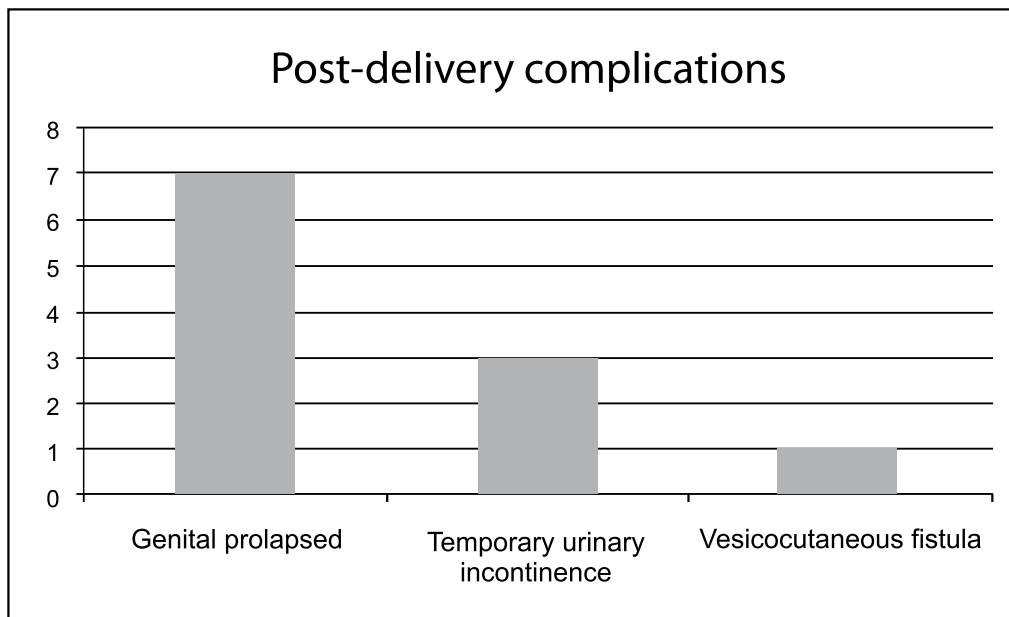


Figure 2 - Post-delivery complications of the 14 patients. One of the genital prolapsed patients had to undergo a hysterectomy.

other six patients had mild to moderate prolapse and were not submitted to surgical correction.

DISCUSSION

Bladder exstrophy has a major impact in the life of patients affected by the disease and their families. Parents of children suffering from BE experience more stressors than the average population (e.g. worrying about the long-term impact of the illness, helping the child with his/her hygiene needs), and when facing increased stress they sometimes cope through avoidance and distancing, harming the child and the familiar structure itself (11). Observing the challenges BE inflicts, it is not surprising that suicidal ideation is also a frequent issue families, patients and physicians have to cope with. A study with 121 patients showed that almost 15% experienced suicidal ideation, with an increase in this number to 38%, when only those aged 14 years and older are considered, including two cases of suicide attempt and one case of suicide (12).

Besides the social and psychological implications of BE there are also several other repercussions of the disease, including the ones from the corrective surgeries and from the associated morbidities of the disease even when treated. Some complications of any method of primary bladder exstrophy closure are complete wound dehiscence, bladder prolapsed, urethral outlet obstruction, bladder calculi, pubic separation, renal calculi and corporal loss; other rarer complications include osteotomy non-union, leg length inequality, persistent joint pain, posterior bladder outlet obstruction, urethrocutaneous fistula, urinary infection, and surgical site infection (13-14). In addition, patients with BE have a 694-fold increase in the risk of bladder cancer by the age of 40, with a higher incidence of cystitis glandularis, which is related to the genesis of adenocarcinomas, demanding intense surveillance of these patients when they achieve adulthood through urine cytology and cystoscopy (15,16). Furthermore, with the optimized treatment options, patients with BE are living longer and the first case of prostate cancer has already been reported, demonstrating the need for screening in these patients, as they get older (17).

Our study presents an abortion rate of 18.1%, against a 12.4% rate of a major pregnancy morbidity study (n = 24.481) (18). Moreover, we present a urinary infection rate of 21.4%, compared with a 5.0% rate of the same mentioned study (18). Regarding temporary urinary incontinence, its incidence can be as high as 80% in some moment of the pregnancy, keeping rates around 20% shortly after delivery, which is comparable to our 21% rate of incontinence after delivery (19). A similar study targeting pregnancy after lower urinary tract reconstructions demonstrated similar findings, with 27 women and 34 pregnancies having the following complications: an injury to the vascular pedicle of the cystoplasty without sequelae and six cases (17.6%) of temporary urinary incontinence (20). Urinary retention and vesicocutaneous fistulas are rare occurrences during pregnancies (21,22).

We found a higher incidence of genital prolapsed in our casuistic when compared with the literature, with 50.0% of our patients with genital prolapsed against a median frequency of 30.0% in the literature, we had one severe case of genital prolapsed which culminated into a hysterectomy, but the other six cases had mild to moderate prolapses, which are being followed since there are no complaints of major discomfort for this condition (10). Female patients with BE are expected to have some degree of vaginal prolapse, especially after a second pregnancy, even without vaginal delivery. Some authors suggest that bed rest for the pregnant mother during the third trimester may diminish this complication (23). Elective Cesarean section is the most recommended delivery for protecting continence and avoiding prolapses (24).

All of our patients were sexually active and had stable marital lives. A study assessing the quality of life of women born with BE presented a stable relationship rate of 64% and sexual activity in 76% (10). A high percentage of women were actively working (59%) against 42.8% of our series, but persistent urinary incontinence was present in 65% of their patients and in none of ours (10). Other studies assessing continent urinary diversion found continence rates of less than 15%, more similar to our findings (25).

CONCLUSIONS

Patients with BE may have a satisfactory and productive lifestyle. They can get married and have children, but they should take extra care during pregnancy and after delivery for they may have more complications than people without this condition.

CONFLICT OF INTEREST

None declared.

ABBREVIATIONS

BE - Bladder Exstrophy

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Submitted for publication:
May 12, 2010

Accepted after revision:
April 15, 2011

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