SURGICAL MANAGEMENT OF BLADDER EXSTROPHY IN A COHORT OF PATIENTS

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Abstract

This retrospective study evaluated patients treated in the Pediatric Surgery Department of the Clinical Emergency Hospital for Children „Grigore Alexandrescu”, Bucharest, Romania, between 2006 and 2016. The inclusion criteria were the diagnosis of a classic form of bladder exstrophy. All the patients underwent primary bladder closure with or without bilateral anterior innominate and vertical iliac osteotomies. Regarding the pelvic osteotomies, we analyzed their use depending on the age the primary bladder closure was performed at. Complications after primary bladder closure, like dehiscence followed by bladder prolapse were also analyzed. Epispadias repair was performed in the male patients who underwent modern staged reconstruction of exstrophy (MSRE) and the bladder neck repair and other additional procedures were performed in order to achieve continence. The study included 35 patients from which 24 were male and 11 were female. All patients underwent primary bladder closure at a mean age of 2,62 months. Pelvic osteotomies were performed in 28,6% of the patients and one patient died after primary bladder closure. The major complication after primary bladder closure was wound dehiscence followed by bladder prolapse and occurred in 22,85% of the patients. Epispadias repair was performed in 18 male patients who underwent MSRE. Mean age for epispadias repair was 1,21 years. Bladder neck repair was performed in 25 patients at a mean age of 5,13 years. Some of these patients needed additional interventions in order to achieve continence like bladder augmentation and urinary diversion. Continence was analyzed in 26 patients and the overall rate of continence was 84,64% (continent patients – 73,08%, partially continent – 11,54% and incontinent patients – 15,38%). The purpose of the treatment of bladder exstrophy is to achieve continence, a good esthetic result and to preserve renal function. One should use all the methods to achieve continence in patients, as bladder neck reconstruction alone is not enough sometimes.

Keywords: bladder exstrophy, continence, primary bladder closure, epispadias repair, bladder neck reconstruction, complications

Introduction

Bladder exstrophy is a rare disease and part of the exstrophy-epispadias complex (EEC). When having a bladder exstrophy, the patient presents a lower abdominal wall defect through which the open bladder and an epispadic urethra are exposed. In addition, the patients present wide
pubic diastasis and an anterior placed anus [1].

The incidence is estimated between 1:30,000 to 1:50,000 live births [2]. According to EUROCAT, the prevalence of the extrophy-epispadias complex in Europe, between 2006 and 2015, was 5.7:100,000 births [3].

Surgical management of the patients with bladder exstrophy consists of primary bladder closure, epispadias repair and reconstruction of the bladder neck, in order to create a continence mechanism [1]. Contemporary approach of these patients includes a modern staged reconstruction (MSRE) or a complete primary repair (CPRE). The modern staged reconstruction of bladder exstrophy is a 3-staged approach in which the first stage is performed early after birth and consists of closure of the bladder, posterior urethra and abdominal wall. During the first stage, the pelvic osteotomies can or cannot be used. The second stage consists of epispadias repair and is performed in infants, while the third stage consists of bladder neck reconstruction and is best performed at the age of 4 [1]. During the complete primary repair of the bladder, the closure of the bladder, posterior urethra, abdominal wall and the repair of the epispadias are being made simultaneous.

The purpose of this study was to analyze the pattern of surgical management of the patients with classic form of bladder exstrophy.

Materials and method

This retrospective study evaluated patients with classic form of bladder exstrophy treated in the Pediatric Surgery Department of the Clinical Emergency Hospital for Children „Grigore Alexandrescu”, Bucharest, Romania, between 2006 and 2016.

All the patients underwent primary bladder closure with or without bilateral anterior innominate and vertical iliac osteotomies. To analyze the usage of the pelvic osteotomies the patients were divided in 4 groups depending on the age of primary bladder closure: patients who underwent primary bladder closure in the first 72 hours of life, patients who underwent primary bladder closure between 72 hours of life and 30 days of age, patients who underwent primary bladder closure between 30 days of age and 12 months of age and patients who underwent primary bladder closure at over 1 year old.

The complications after primary bladder closure consist of dehiscence followed by bladder prolapse, wound infections and urinary tract infections (UTIs).

Epispadias repair in male patients who underwent MSRE was performed using a modified Cantwell-Ransley repair.

Regarding the continence, there were 3 groups of patients: continent patients (permanent continence), partially continent patients (dryness for over 2 hours and incontinent intervals) and incontinent patients. In the first two groups the continence was either spontaneous or through clean intermittent catheterization.

The continence was also analyzed in association with the procedures performed: bladder neck reconstruction, bladder augmentation and urinary diversion.

All the data were analyzed using SPSS.

Results

The study includes 35 patients from which 24 were male and 11 were female. All the patients underwent primary bladder closure, 18 of the male patients underwent epispadias repair, while 25 patients underwent bladder neck reconstruction. Only one patient died during hospitalization after primary bladder closure due to prematurity and not as a post-operative complication.

The mean age for primary bladder closure was 2.62 months with a minimum of 0 days and a maximum of 3.36 years. The MSRE was used in 20 males and the CPRE was used in 4 male patients. Pelvic osteotomies were used in 28.6% of all the patients and were performed only when the primary bladder closure was performed at over 72 hours of age. The number of patients who underwent primary closure, with or without pelvic osteotomies depending on the age the interventions were performed at are detailed in Table 1.
Mean length of stay after primary bladder closure was 18 days with a minimum of 8 days and a maximum of 1.7 months.

<table>
<thead>
<tr>
<th>Age at the moment of primary bladder closure</th>
<th>Primary bladder closure without pelvic osteotomies</th>
<th>Primary bladder closure with pelvic osteotomies</th>
<th>Total</th>
</tr>
</thead>
<tbody>
<tr>
<td>≤ 72 hours</td>
<td>20</td>
<td>0</td>
<td>20</td>
</tr>
<tr>
<td>&gt;72 h - 30 days</td>
<td>4</td>
<td>4</td>
<td>8</td>
</tr>
<tr>
<td>1 month - 12 months</td>
<td>1</td>
<td>4</td>
<td>5</td>
</tr>
<tr>
<td>&gt; 1 year</td>
<td>0</td>
<td>2</td>
<td>2</td>
</tr>
<tr>
<td>Total</td>
<td>25</td>
<td>10</td>
<td>35</td>
</tr>
</tbody>
</table>

Table 1 - Usage of pelvic osteotomies depending on the age of primary bladder closure

Epispadias repair was performed in 18 male patients using a modified Cantwell-Ransley repair. Mean age of epispadias repair was 1.21 years with a minimum of 5.59 months and a maximum of 3.47 years. Mean length of stay after epispadias repair was 10.72 days.

Bladder neck repair was performed in 25 patients. Out of the 35 patients, one patient didn’t need bladder neck repair, while in the other 8 the bladder neck repair was to be performed after the end of the study. Mean age of BNR was 5.13 years with a minimum of 3.14 years and a maximum of 9.46 years. Mean length of hospitalization after BNR was 15.88 days.

Following the primary bladder closure, we analyzed the post-operative complications. The major complication was wound dehiscence, followed by bladder prolapse (Table 2). This complication was present in 8 patients (22.85%). 6 of them were present in the patients who did not undergo pelvic osteotomies during primary bladder closure and 2 underwent pelvic osteotomies during primary bladder closure. After this, those 8 patients underwent a new bladder closure after 6 months and pelvic osteotomies were used.

<table>
<thead>
<tr>
<th>Age at the moment of primary bladder closure</th>
<th>Bladder closure without pelvic osteotomies</th>
<th>Dehiscence followed by bladder prolapse after bladder closure without pelvic osteotomies</th>
<th>Bladder closure with pelvic osteotomies</th>
<th>Dehiscence followed by bladder prolapse after bladder closure with pelvic osteotomies</th>
</tr>
</thead>
<tbody>
<tr>
<td>≤ 72 hours</td>
<td>20</td>
<td>4</td>
<td>0</td>
<td>0</td>
</tr>
<tr>
<td>&gt;72 hours</td>
<td>5</td>
<td>2</td>
<td>10</td>
<td>2</td>
</tr>
<tr>
<td>Total</td>
<td>25</td>
<td>6</td>
<td>10</td>
<td>2</td>
</tr>
</tbody>
</table>

Table 2 - Wound dehiscence followed by bladder prolapse depending on the age of primary bladder closure and usage of pelvic osteotomies.

The minor complications after primary bladder closure were present in 4 patients, 2 of them presenting wound infections and the other 2 UTIs. All the patients were treated using antibiotics according to antibiogram.

Following the epispadias repair complications were present in 3 patients out of 18, 2 of them presenting urethral fistulas and 1 of them urethral stricture.
We evaluated the continence in 26 patients out of 35 (1 patient died after the primary bladder closure and the other 8 patients were planned to undergo a continence procedure after the end of the study). The total rate of continence in the 26 patients was 84.62%. 19 patients (73.08%) were continent and 3 patients (11.54%) were partially continent. The others were incontinent.

As a reminder, 4 patients underwent CPRE. One of those patients achieved continence after the surgery and didn’t need further continence procedures. The other 25 patients whom continence we evaluated underwent BNR. In the patients whose bladder either was too small, or the bladder neck repair failed, in order to achieve continence bladder augmentation (ileocystoplasty) and urinary diversion (Mitrofanoff procedure or ureterosigmoidostomy) were used. In this 25-patient group, 18 were continent. 9 out of the continent patients underwent only BNR, 4 of them underwent BNR and bladder augmentation, 2 of them underwent urinary diversion (Mitrofanoff procedure – continence using clean intermittent catheterization) in addition to bladder augmentation and BNR and 3 of them underwent urinary diversion (ureterosigmoidostomy) after the BNR (Table 3).

<table>
<thead>
<tr>
<th>Procedure</th>
<th>Continent</th>
<th>Partially continent</th>
<th>Incontinent</th>
<th>Total</th>
</tr>
</thead>
<tbody>
<tr>
<td>BNR</td>
<td>9</td>
<td>2</td>
<td>2</td>
<td>13</td>
</tr>
<tr>
<td>BNR + bladder augmentation</td>
<td>4*</td>
<td>1</td>
<td>2</td>
<td>7</td>
</tr>
<tr>
<td>BNR + bladder augmentation + urinary diversion</td>
<td>2*</td>
<td>0</td>
<td>0</td>
<td>2</td>
</tr>
<tr>
<td>BNR + urinary diversion</td>
<td>3</td>
<td>0</td>
<td>0</td>
<td>3</td>
</tr>
<tr>
<td><strong>Total</strong></td>
<td><strong>18</strong></td>
<td><strong>3</strong></td>
<td><strong>4</strong></td>
<td><strong>25</strong></td>
</tr>
</tbody>
</table>

Table 3 - Continence rates depending on the procedure used.

Continence rates in the group of patients who underwent only BNR was 69.23%. In the group of patients who underwent bladder augmentation after BNR the continence rate was 57.14%. For the patients who underwent urinary diversion (Mitrofanoff procedure) in addition to bladder augmentation and BNR and for the patients who underwent urinary diversion (ureterosigmoidostomy) after BNR the rate of continence was 100%.

Using a Pearson test, we determined that there was a weak to moderate association between continence rates and the age of primary bladder closure (p=0.033; p<0.05).

In addition to the basic procedures for the bladder extrophy some patients needed other interventions. One patient who needed bladder augmentation underwent cystolithotomy and 13 patients underwent inguinal hernia repair. 12 patients out of these were males and one was female. 50% of the male patients and only 9% of the female patients developed inguinal hernia.

**Discussions**

The management of the bladder extrophy is a complex process requiring multiple surgical procedures in order to achieve continence, a good esthetic and functional result and to preserve renal function. Unfortunately, sometimes, despite the efforts made by the medical team some patients do not achieve continence and a few of them die. In our case only one patient died (2.85% of total) after primary bladder closure, resulting a low mortality rate. Nelson et al. also reported a low mortality rate of only 1.5% [4]. Similar to Nelson et al. our patient died of prematurity and not as a post-operative complication. Usually, patients with bladder extrophy have associated anomalies. In our study was reported one patient having coloboma of the iris, microcornea and sclerocornea and another patient having annular pancreas.

As we should expect bladder extrophy is more frequent in males, male to female ratio in
our study being 2.18:1. Literature reports this ratio between 1.5:1 to 6:1 [5-9].

The primary bladder closure is the first step to be done in a patient with this condition. This step should be performed as soon as possible after birth in order to avoid the possible complications, which would be determined by exposing the bladder mucosa. In our study all the patients underwent primary bladder closure at a mean age of 2.62 months, with a minimum of 0 days and a maximum of 3.36 years. Contemporary guidelines for bladder exstrophy management suggest that primary bladder closure should be done in the first 72 hours of life. A literature review shows a mean age of primary bladder closure of 1.97 days [10] to 6.57 days [11]. In some cases, the mean age of primary bladder closure is even smaller, of [12].

The age of primary bladder closure depends not only on the medical team, but also on the patients’ access to medical services, educational and financial status of the parents and other aspects.

Our study shows that 57.14% of the primary bladder closure were performed at an age less than 72 hour. This rate is 12% greater than other studies in the literature [13]. The number of primary bladder closure performed between the age of 72 hours and 30 days was 12% greater than the number of Baka-Ostrowska et al. reported and for the age of over 12 months, our study shows 32% smaller than Baka-Ostrowska et al. reported and for the age of over 12 months, our study shows 90% more cases than Baka-Ostrowska et al. and 5.71% in our study).

We performed bilateral anterior transverse and posterior iliac osteotomies in 28.57% of the patients. The rate of performing pelvic osteotomies is low probably because most of the primary bladder closures were performed in the first 72 hours of life, when this procedure is not needed. The rate is similar with the ones reported in other studies – between 30% and 36% [4,10,13].

The mean length of hospital stay after the primary bladder closure was 18 days, with a minimum of 8 days and a maximum of 1.7 months. One study regarding CPRE reported a mean length of stay of 13.2 days [14] after the procedure, while another one reported 24.6 days [4].

The epispadias repair is the second step in the male patients who underwent MSRE as a first step. The contemporary approach suggests the epispadias repair should be performed at the age of 6 months to 12 months [1, 15-18]. 18 male patients from our study underwent epispadias repair at a mean age of 1.21 years using modified Cantwell-Ransley procedure. Mean length of stay after the procedure was 10.72 days.

Regarding the bladder neck repair, 25 incontinent patients, males and females, underwent Yound-Dees-Leadbetter procedure at a mean age of 5.14 years. The smallest age for BNR was 3.41 years and the greatest was 9.46 years. According to literature, the recommended age for BNR is 4 to 5 years old [1,15,19], but some studies report the mean age for BNR at 7 years [11], even at 9 years old [20].

Post-operative complications are very important because they can affect the patient evolution and can increase the costs in healthcare. The most important complication after primary bladder closure is wound dehiscence followed by bladder prolapse. In our study wound dehiscence and bladder prolapse occurred in 8 patients (22.85%). 75% of them occurred in patients in which primary bladder closure was performed without pelvic osteotomies and the other 25% in patients in which primary bladder closure was performed with pelvic osteotomies. In our study the rate of wound dehiscence and bladder prolapse is lower than the one Baka-Ostrowska et al. reported - 31% [13]. Further comparison between our study and the one of Baka-Ostrowska et al. showed lower rates of wound dehiscence and bladder prolapse in our study for the patients who underwent primary bladder closure without osteotomies at less than 72 hours - 20% compared with 27.65%. Wound dehiscence and bladder prolapse in patients who underwent primary bladder closure without osteotomies at a age of over 72 hours was better in our study 40%, compared with 75% reported in literature. The difference between our study and the one published by Baka-Ostrowska et al. was not
important regarding the patients who underwent primary bladder closure with pelvic osteotomies at an age of over 72 hours – 20% compared with 16% [13]. The complications occurring after epispadias repair (urethral fistula) were present in 11.1% of the male patients. For this kind of complications, the literature reports a frequency of 2 to 26% [1].

We evaluated the continence of 26 patients. The patients were split into 3 groups depending on the continence: continent, partially continent and incontinent. 73.08% of the patients were continent, 11.54% partially continent and 15.38% were incontinent. Overall continence rate was 84.62%, being almost the same as the literature reports. The overall rates of continence reported in other studies are between 79% [21] and 90% [22]. Shaw et al. reports a proportion of 8% of partially continent patients and 2% incontinent patients. 25 patients out of the 26 evaluated underwent BNR and other additional procedure in order to achieve continence. 13 patients underwent only BNR and 69.23% of them achieved continence. Literature reviews show a rate of continence for the patients who underwent only BNR of 34% [22], 56% [21] and even 70% [19]. 57.14% out of the 7 patients who underwent bladder neck repair and bladder augmentation are continent. This is a better rate than Shaw et al. reported for a similar group of patients. Another group of 2 patients underwent bladder neck repair, bladder augmentation and urinary diversion – Mitrofanoff procedure, both patients being continent using clean intermittent catheterization. In this group of patients, we saw a better rate of continence than Capolicchio et al. reported – 67% - for the same group of patients. One last group of 3 patients underwent bladder neck repair and urinary diversion – ureterosigmoidostomy – achieving 100% rate of continence.

Regarding the procedure used for bladder augmentation literature reviews show that ileocystoplasty was used in 64% [22]. However, we used this procedure in all our patients. Due to abdominal wall defects, the patients having bladder extrophy are more likely to develop inguinal hernia [18-20]. In our study 50% of the male patients and 9% of the female patients developed inguinal hernia.

Conclusions

Bladder extrophy is a rare anomaly which requires multiple surgical interventions to achieve continence, a good esthetic result and preserve renal function. Surgical management of the patient should begin immediately after birth and is it very important that primary bladder closure to be performed in the first 72 hours of life in order to achieve a greater continence rate.

In order to achieve continence, the most wished outcome of the treatment, sometimes bladder neck reconstruction alone is not enough, and patients need additional interventions like bladder augmentation and urinary diversion. Using all the methods we achieved a total rate of 84.62% continence among our patients. In order to succeed in achieving continence medical team’s efforts have to be supported also by parents who have to understand what the treatment involves.

References