

2. ABSTRACT

INTRODUCTION

Exstrophy-epispadias complex is one of the most severe forms of urogenital defects. It includes cloacal exstrophy, bladder exstrophy and epispadias. Cloacal exstrophy affects not only the genitourinary system, but also the gastrointestinal system, and the treatment of this form of defect differs from bladder exstrophy and epispadias. Bladder exstrophy occurs with a frequency of 1:30,000 live births in boys and is three times less frequent in girls (1:100,000). Epispadias occurs in girls with a frequency of 1:650,000 live births, six times less often than in boys.

Treatment in both defects is surgical. In bladder exstrophy, the first stage is the closure of the bladder with the reconstruction of the pubic symphysis and the anterior abdominal wall. An element of this reconstruction in most centers is iliac bone osteotomy, which allows the tissues to be brought closer with minimal tension and significantly reduces the risk of complications in the form of dehiscence of the postoperative wound and re-ruption of the bladder. After the primary reconstruction of the bladder exstrophy, the patient remains with the urethra open, as in the epispadias. The next stage of treatment in girls is common for bladder exstrophy and isolated epispadias and consists of the reconstruction of the bladder neck, urethra and vulva.

The Paediatric Urology Department of the "Children's Memorial Health Institute", developed the method known as the "Warsaw approach". This method was described by dr hab.n.med. Małgorzata Baka-Ostrowska and was dedicated to boys with bladder exstrophy and epispadias but its elements are also used for reconstruction in girls. "Warsaw approach" consists of reconstruction of the bladder neck with bilateral anti-reflux ureters reimplantation, urethral and penile reconstruction in boys, using external rotation of the corpora cavernosa while in girls clitoral and vulvar reconstruction. One of the key distinguishing elements of the "Warsaw approach" is the operative access with the opening of the pubic symphysis. This access allows for the reconstruction of the urethra in a straight line.

AIM OF THE STUDY

The general objective of the study is to evaluate the long-term results of the bladder neck and urethral reconstruction performed using the Young-Dees-Leadbetter method in the modification of the Pediatric Urology Department of the Children's Memorial Health Institute – "Warsaw approach" in female patients with bladder exstrophy and epispadias.

The specific objectives of the study concern, firstly, the assessment of urinary continence after bladder neck reconstruction surgery in the group of patients with bladder exstrophy and isolated epispadias, taking into account the influence of the surgical technique used during the primary reconstruction of the bladder, age at the time of surgery, bladder capacity and the need for reoperation. Secondly, to determine the impact of bladder neck reconstruction on the function of the upper and lower urinary tract and to develop recommendations for surgical treatment of patients with bladder exstrophy and epispadias.

MATERIAL AND METHODOLOGY

The analysis used clinical data of 46 patients with bladder exstrophy and 8 patients with epispadias, operated at the Department of Paediatric Urology of the Children's Memorial Health Institute, born in the years 1976-2010, in whom the follow-up time after bladder neck reconstruction was at least 5 years and the data necessary for the analysis were available. The age of the patients at the end of the observation was 18 years.

Originally, 32 patients with bladder exstrophy were operated on in our institution, while 14 patients were referred from other centers, often after unsuccessful primary reconstruction of the bladder exstrophy. Posterior iliac osteotomy was performed in 19 patients, in 13 as part of the primary reconstruction of the bladder exstrophy, and in 6 patients during reoperation after unsuccessful closure of the bladder exstrophy in other centers.

Urine continence after bladder neck reconstruction, described as periods of dryness between micturition or catheterizations expressed in minutes, was assessed immediately after surgery, followed by evaluation after one year, two, three, five years, and more than five years after surgery.

The condition of the upper urinary tract before surgery and at least five years after bladder neck reconstruction surgery was assessed by ultrasound, voiding cystography, renal isotope examination, and determination of serum creatinine and urea levels.

At the same intervals, the condition of the lower urinary tract was assessed on the basis of bladder diary, voiding cystography, urodynamic examination and measurement of bladder capacity under general anesthesia.

RESULTS

Urine continence with the possibility of micturition or catheterization every 3 hours and remaining dry between micturition periods, at the end of observation, more than 5 years after bladder neck reconstruction surgery, was found in 32 patients, representing 59.3%, another 3 patients (5.7%) kept urine between 2-3 hours and 2 (3.8%) between 1-2 hours. Urine continence during the night was found in 31 (57.4%) patients. The effect of improving urinary continence over five years was statistically significant for patients, $p < 0.001$; both in the group of patients with and without osteotomy. An increase in bladder capacity was observed after bladder neck reconstruction surgery, which met the linear model. The mean bladder capacity at the end of the 18-year-old observation was 231.94 ml, the median was 206.5 ml. This increase was statistically significant, $p < 0.001$. Bladder emptying by spontaneous micturition was found in 21 patients, 3 patients after micturition used additional bladder catheterization, and 22 patients used only clean intermittent catheterization to empty the bladder.

Creatinine and urea levels at the age of 18 were normal at the end of follow-up except in two cases.

CONCLUSION

1. Reconstruction of the bladder neck and urethra performed using the Young-Dees-Leadbetter method in the modification of the Pediatric Urology Department of the Children's Memorial Health Institute – "Warsaw approach", in patients with bladder exstrophy and epispadias, is an effective method, allowing to achieve full urinary continence for more than three hours and urinary continence at night in 59.3% of patients.
2. Lower urinary tract function is preserved in 31% of patients.
3. The most important factor affecting urinary continence after bladder neck and urethral reconstruction is the absence of complications after the primary reconstruction of the bladder exstrophy that would cause the need for reoperation.
4. Upper urinary tract function may deteriorate during observation and requires constant monitoring by performing ultrasound, biochemical and isotope examinations.