LONG TERM FOLLOW UP OF COMPLETE BLADDER EXSTROPHY REPAIR.
A CASE REPORT

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ДОЛГОСРОЧНОЕ ПРОСЛЕЖИВАНИЕ ПРИ ПОЛНОЙ РЕКОНСТРУКЦИИ ЭКСТРОФИИ МОЧЕВОГО ПУЗЫРЯ. СООБЩЕНИЕ О КЛИНИЧЕСКОМ СЛУЧАЕ

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ABSTRACT
OBJECTIVE: Bladder exstrophy is a congenital anomaly which is not always successfully managed by surgery. Major goals of surgical intervention in such cases are preservation of normal renal function, development of adequate bladder function and urinary continence and avoidance of future urinary tract infections. We present 5-year data on a patient who underwent complete repair of the bladder exstrophy. CASE REPORT: We describe a full term female infant who presented at birth with complete bladder exstrophy. Complete repair of the condition was performed 3 days after birth (Ransley technique). During hospitalization the patient had a positive urinary culture with Candida lusitaniae, enterococcus and septicemia with Klebsiella pneumoniae ESBL. The patient had no complications until the age of 20 months when she developed an episode of pyelonephritis and five further episodes of cystitis with E. coli. Radiographic testing showed small bladder capacity (23 ml at the age of 3 years), bilateral vesicoureteral reflux, a long stenotic urethra and no loss of renal function. Because of the recurrent urinary tract infections, dilatations of the stenotic urethra (Scheldinger technique) were successfully performed at the age of 3. Twenty-two months later the child had negative urinary cultures, a normal renal function and had also gained partial control of bladder function and urinary continence and avoidance of future urinary tract infections. We present 5-year data on a patient who underwent complete repair of the bladder exstrophy.

CONCLUSIONS: Surgical repair of bladder exstrophy remains a challenging surgery for the pediatric urologist. Following surgical correction both early and long-term post-operative complications may be present. Longitudinal follow up is required by an experienced team of health care professionals.

Key words: bladder exstrophy, neonate, complications

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Резюме
Цель: Экстrophy мочевого пузыря представляет собой врожденную аномалию, которую не всегда возможно успешно лечить оперативным методом. В таких случаях основной целью хирургической интервенции является сохранение нормальной почечной функции, достижение адекватного функционирования мочевого пузыря и нормальной континенции, а также и избежание будущих инфекций мочевых путей. В работе авторы представляют пятилетнее прослеживание пациента, перенесшего полное восстановление экстракорпорального мочевого пузыря. Клинический случай: Авторы сообщают о малчике, родившемся в сроке с полной экстrophy мочевого пузыря. Проведена полная реконструкция пузыря на третий день после рождения по методу Ransley. Лабораторные тесты пациента во время госпитализации показали положительную культуру мочи - Candida lusitaniae с энтерококками и септициемией с Klebsiella pneumoniae ESBL. Пациент без осложнений до 20-имесячного возраста, когда развился пиелонефрит и 5 раз заболевал циститом, вызванным E. coli. Рентгенологическое исследование показывает мочевой пузырь небольшого объема (23 мл, в возрасте 3 лет), двусторонний везикоуретеральный рефлюкс, стеноз уретры и сохраненную почечную функцию. За 3 года проведена дилатация уретры (по Scheldinger) из-за постоянных инфекций мочевых путей. 22 мес. позже тесты пациента показывают отрицательные уринарные культуры, налици нормальная почечная функция, а также и контроль за сфингером мочевого пузыря.

Выводы: Оперативное восстановление при экстрофии мочевого пузыря для детского уролога представляет очень трудное хирургическое вмешательство. После хирургической коррекции возможно как раньше, так и поздние постоперационные осложнения. В таких случаях необходимо осуществлять долгосрочное прослеживание пациента со стороны опытного состава в этой области.

Ключевые слова: эксттрофия мочевого пузыря, новорожденные, осложнения
INTRODUCTION

Bladder extrophy is a rare congenital anomaly that occurs in 1 in 30,000-50,000 live births.\(^1\) It occurs more frequently in males (male/female ratio: 3-4/1), in firstborns and in Caucasian neonates.\(^2-3\) It is a complex congenital abnormality which affects the musculoskeletal, urogenital and gastrointestinal systems.\(^4\) The abnormality can be diagnosed prenatally.

The treatment for bladder extrophy is surgical and it presents a challenge both in terms of the surgical repair itself and in maintaining normal renal function and development of adequate bladder storage. Early and long term complications following surgical correction are not rare and include bladder dehiscence, urinary tract infections (UTI), bladder stones and perforations as well as an increased risk of malignancy, vaginal and rectal prolapsed in female patients and epididymitis in adult males.

A case of a patient with complete bladder extrophy is described due to the rareness of this anomaly, the complications and the treatment followed.

CASE REPORT

We report a case of a female neonate, first child of phenotypically healthy immigrant parents, born in a public maternity hospital by cesarean section at 41 weeks gestation. Birth weight was 3320 gr (50th centile curve). During the prenatal scanning which included repeated ultrasound examination, the anomaly was not detected. It was also not possible to identify the gender of the baby. Immediately after birth, the baby was transferred to a Neonatal Intensive Care Unit, as the bladder extrophy with visible outer ureteral orifices became evident. Three days after birth, complete surgical reconstruction was carried out, with reconstruction of the bladder neck and neourethra, reconstruction of the outer sphincter, narrowing of the pubic bones and placement of the bladder within the pelvis (Fig. 1). The neonate was positioned at a right angle (modified Bryant’s traction, Fig. 2) for 21 days and received a triple antibiotic regimen for 14 days. Throughout her stay in the hospital, the patient had positive urine cultures for Candida lusitaniae, Klebsiella enterococcus as well as clinical and laboratory evidence of septicaemia-urinary tract infection with Klebsiella pneumoniae ESBL. She was treated with amphotericin B liposome, imipenem and amikacin. Multiple ultrasound scans revealed an increasing bilateral distention of the pyelocaliceal system while the DMSA scan showed a slight reduction in renal function of the left kidney. It was difficult to perform a cystography due to the increased stenosis of the urethra. The neonate was discharged at 51 days, with instructions for chemoprophylaxis with cefprozil and regular monitoring by a paediatric nephrologist. The patient progressed without complications until the age of 20 months when, due to episodes of pylonephritis, initially with E. coli ESBL and later with Pseudomonas aeruginosa, she was given iv treatment. Despite chemoprophylaxis with nitrofurantoin 3 episodes of asymptomatic and 2 episodes of febrile urinary tract infections occurred. The cystography that followed showed a 3rd to 4th degree vesicoureteral reflux (VUR) bilaterally and low bladder capacity (29 ml at the age of 3 years). Due to the recurrent UTIs and stenosis of the urethra, intermittent sterile dilatations of the urethral stricture were carried out (urethral width 12 Fr) up to 16 Fr at the age of 3. Twenty-two months later, the patient received chemoprophylaxis...
with nitrofurantoin. She had normal renal function, no significant variations in the DMSA scan and in terms of urinary continence, she had gained partial control of the bladder sphincters (she had learned to void her bladder voluntarily every two hours during the day, although there was a small amount of urine leakage to her underwear).

DISCUSSION

Bladder extrophy is a congenital abnormality which can be diagnosed prenatally. The commonest ultrasound findings are identified before the 20th week of gestation and include absence of bladder filling, low set umbilicus, pubic bone diastasis, diminutive genitalia and lower abdominal mass that increases in size as the pregnancy progresses.\(^4\)\(^6\) In our patient, the only finding at the prenatal checkup was the difficulty in determining the gender.

Typical findings include the bladder opening onto the lower abdominal wall and the eversion of the posterior wall of the bladder exterior to the body. The external genital organs of male neonates appear to have more serious deformities due to the reduced length of the corpora cavernosa of the penis and the coexistence of epispadias.\(^7\) Coexistence of an inguinal hernia has been reported in both genders, but more frequently in males (82%) than in females (11%). Radiographic study is necessary in order to rule out other abnormalities of the kidneys such as hydronephrosis and aplasia as well as to determine the anatomy of the internal genital organs.

The treatment is only surgical and primarily aims to provide a satisfactory capacity and function of the bladder, maintaining the renal function but also giving an acceptable appearance and functionality of the genital organs. Currently there are two surgical procedures that are used for the repair of bladder extrophy, the modern staged repair of extrophy (MSRE) and the complete primary repair of extrophy, with or without the performance of osteotomy after one year (CPRE).\(^8\) Our patient was treated with CPRE, which had to be performed within the first 72 hours of the neonate’s life and aims at the gradual increase of bladder capacity and achievement of continence, early bladder neck resistance and bladder cycling, reducing the number of surgical interventions.\(^9\)\(^10\)

Early post-operative complications include incidents of UTIs (especially in patients with a concurrent VUR) which can result in scarring and eventual renal failure as well as urolithiasis, which is related to relapsing UTIs. The occurrence rate for pyelonephritis and renal scarring is 20%-28% and is more common in patients with bladder augmentation.\(^11\)\(^-\)\(^12\) More specifically, Gargollo reports that 28% of the patients who underwent CPRE had 1-4 episodes of pyelonephritis and 19% had cortical defects visible with a renal scan. After initial bladder closure, fistulas (urethrococutaneous, vesicocutaneous) are the most commonly reported complications in a rate between 4-16%, depending on the surgical procedure. Hypospadias, appears to be a more common result at the initial closure especially in patients with CPRE. Bladder perforations rarely occur following procedures that increase bladder outlet resistance (such as bladder neck reconstruction and bladder augmentation). Bladder dehiscence is also not very frequent but is associated with an increased risk of incontinence.\(^13\)

As far as the ability to control the bladder for a period of at least 2-3 hours and achieving continence in patients, which is one of the key goals of surgery, the rates vary between 37% and 90%. Shnorhavonan et al. report that 74% of their patients, who are 4 years of age or older, have achieved daytime continence with voluntary voiding - 20% of boys and 43% of girls have achieved primary urinary continence without the need for bladder neck reconstruction.\(^14\) Scafeer et al. report lower percentages: 41% underwent bladder neck repair, with 56% subsequently continent, 19% daytime continent with nocturnal incontinence and 26% continuously incontinent. Similarly, Gargollo reports that 6 months after CPRE, all patients had urinary continence periods of 2 to 3 hours or greater, with females performing better than males. As for fecal incontinence or soiling, none were reported in children older than 4 years.\(^10\)\(^,\)\(^11\)\(^,\)\(^15\)\(^,\)\(^16\)

In this particular patient, the coexistence of VUR, the relapsing UTIs as well as the low bladder capacity led to the decision to perform dilatations of the urethral stricture in order to increase the capacity of the bladder, while particular emphasis was given to training the patient to void her bladder regularly (every two hours) and avoidance of constipation. The good progress she made and the partial continence that she achieved prove the efficacy as well as the importance of the conservative approach.

In older ages, the complications that are mentioned include epididymitis (19-33%), prolapse of the uterus and the rectum in female patients at an average age of 16 years old (20%) as well as an increased risk for malignancies of the bladder and the colon, particularly in patients whose bladder was increased in size with the use of intestinal
components. As far as the reproductive ability of these patients is concerned, there is an insufficient number of studies. However, it appears that male patients have reduced fertility due to oligospermia/azoospermia as well as reduced sperm motility. On the other hand, there do not seem to be any particular problems in female patients where a satisfactory number of normal pregnancies have been reported.

Although the major goals are surgical outcome and bladder function, follow-up studies demonstrate that adolescents and young adults are at risk for developing a poor body and genitalia image, anxiety and adjustment disorder. Results also suggest that incontinence was associated with increased psychological dysfunction and need for psychological support.

CONCLUSIONS

The occurrence of complications in patients with bladder exstrophy is not rare. For this reason, regular and longitudinal follow up of such patients is necessary, with particular attention to the evaluation of renal function and prevention of UTIs. In order to improve quality of life and given the complexity of the condition, such patients must be life-long monitored by a team of specialists (urologists, nephrologists, gynecologists, psychologists).

REFERENCES