Pregnancy in a patient with reconstructed bladder extrophy with osteotomy, continent urinary reservoir with the Mitrofanoff principle, vaginoplasty, and uterus didelphys

Espinosa-Chávez Giordano Bruno,1 Urbina-Bernal Luis Carlos,2 Dávila-Garza Alejandro,3 Carrillo-Treviño Sergio,3 Madrigal-Medina Ricardo,2 García-Rodríguez Miguel Ángel,2 Garza-Rodríguez Ramiro.4

ABSTRACT

Bladder extrophy is a pathology caused by developmental failure of the cloacal membrane, with open bladder exposure and genital malformation. It has been described throughout history and there is a high risk of familial recurrence. Neonates undergo surgery with good results for bladder function and continence. The first procedures of urinary diversion and exeresis of vesical plaque have evolved through the work of different authors that have improved and complemented previously described advances, arriving at bladder closure by stages, and more recently at complete primary closure. The desired urinary continence is difficult to achieve in some cases and in patients that have reached adulthood with no previous treatment, and an effective alternative for these cases is the creation of a continent urinary reservoir.

RESUMEN

La extrofia vesical es una patología ocasionada por el fallo en el desarrollo de la membrana cloacal, con exposición de la vejiga abierta y malformación genital. Descrita desde la antigüedad y con alto riesgo de recurrencia familiar. Los neonatos son intervenidos quirúrgicamente con buenos resultados para la función vesical y continencia, durante el periodo neonatal. Los primeros procedimientos de derivación urinaria y exéresis de la placa vesical fueron evolucionando de la mano de diferentes autores, mejorando o complementando los avances anteriormente descritos, llegando hasta el cierre vesical por etapas y más recientemente, el cierre primario completo. En algunos casos resulta difícil alcanzar la continencia urinaria deseada, y pacientes que llegan a edad adulta sin tratamiento, una alternativa efectiva es la creación de un reservorio urinario continente.
We present the case of a 29-year-old woman born with classic bladder exstrophy, in addition to uterus didelphys, that reached adulthood with no treatment. Cystectomy, anterior osteotomy, and continent urinary reservoir with the Mitrofanoff principle were carried out.

**Keywords**: Bladder exstrophy, urinary reservoir, vaginoplasty, uterus didelphys, Mexico.

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**INTRODUCTION**

Bladder exstrophy is a rare congenital malformation, with an incidence of one case in 30,000-50,000 live births. It is more frequent in males with a 3:1 male/female ratio. Clinically, its anatomical characteristics vary from the simplest to the most complex forms, such as cloacal exstrophy, in which the bladder is exposed through a wedge defect in the infraumbilical abdominal wall with separation of the pubis and the muscles of the urogenital diaphragm and abdomen.

Duplicate or bifid genitals are frequent and are associated with other alterations of the spinal column, esophagus, trachea, heart, and kidneys, among others. Treatment is clearly surgical and should be carried out in the first years of life. Management is a challenge and its aim is to provide adequate bladder function. Upon diagnosis in the neonatal period, parents are counseled and repair is programmed, and therefore it is highly unusual to find this condition uncorrected in adults.

There are only a few reports of bladder exstrophy in the literature and even fewer cases of patients without treatment and associated with uterine malformations such as septate vagina and uterus didelphys.

**CASE PRESENTATION**

A 29-year-old woman sought medical attention because she wanted to become pregnant, but was unable to perform sexual activity.

Physical examination revealed bladder exstrophy with a 5 x 5 cm bladder plate, a trigone with orthotopic ureteral meatuses that ejaculated clear urine under direct vision, whitish bladder mucosa, a triangular defect of the abdominal wall, pubic diastasis, a bifid clitoris, and a vagina with a prominent intravaginal septum (Figure 1). Urologic computed axial tomography (UROCAT) scan showed an intact upper urinary tract (Figure 2). Radiography revealed two normal kidneys and ureters and a 12-cm pubic diastasis (Figures 3 and 4).

Vaginoscopy identified a 12-cm long double vagina ending in a normal cervix and hysterosalpingography revealed uterus didelphys (Figure 4).

In the first surgical stage, the orthopedics service performed anterior osteotomy and the placement of an AO-type frame as the fixation method. In the second surgical stage the urology service carried out simple cystectomy, isolation of 50 cm of the ileum, including Meckel’s diverticulum, with posterior opening of the ileum at its asymmetric edge and reconstruction in U (Figure 5). Ureteral reimplantation was carried out with a submucosal tunnel at the level of the wall of the ileum, tubularization of Meckel’s diverticulum for a continent stoma with the Mitrofanoff principle, reimplanting one end to the ileum and the other to the abdominal wall as a stoma, using the VZQ technique (Figure 6).

The urinary reservoir was closed spherically, later closing the abdominal wall and pelvic ring by applying a tubular frame as the external fixation method (Figures 7 and 8).

Ureteral catheterization was carried out with an 8 F catheter and 16 F cystostomy that were removed 10 days after surgery, with satisfactory outcome. The tubular frame was removed 8 weeks after surgery and clean intermittent catheterization was begun. Physical rehabilitation and ambulation were started on the following day.

Vaginoplasty was performed 3 months after surgery and sexual activity commenced 4 weeks later.

The patient presented with amenorrhea 6 months after the surgical procedure. The immunological test for...
pregnancy was positive and was corroborated with a transvaginal ultrasound study that showed pregnancy to be at 7.1 weeks of gestation (Figure 9).

**DISCUSSION**

Bladder exstrophy was described as long ago as 2000 BCE in Babylon, and then Mowat described it in more detail in 1748. It has a 1:36,000 incidence, a 2.3:1 man/woman ratio, and familiar recurrence risk is 1 in 100.1

It is also associated with other alterations that involve the urinary tract such as epispadias, spinal column defects, intestinal alterations, imperforated anus, and omphalocele. In addition there can also be kidney and ureteral alterations, shortening of penile size, and cryptorchidism. 2

Bladder exstrophy characteristics are absence of the anterior wall of the abdomen, of the anterior wall of the bladder, and of the posterior urethra. There is important diastasis of the symphysis pubis and external rotation of the pelvis.

It represents a great challenge in relation to surgical treatment, due to the anatomical complexity involved, as revealed by the high rate of postoperative complications. Bladder closure, with or without osteotomy, within the time frame of birth to 72-hours of life, is the initial treatment, followed by the creation of a urinary continence mechanism with bladder neck reconstruction. Reimplantation is indicated at approximately two or three years of age, the mean age at which physiological maturity has been reached, enabling social urinary continence.3

Classic treatment is by surgical stages, first, with primary bladder closure with or without osteotomy, and second, with bladder neck restoration with ureteral reimplantation as a urinary continence mechanism, and finally, epispadias repair with elongation of the corpora cavernosa. 4

One of the aspired goals is pelvic ring closure, essential for migrating the separated anatomical structures toward the midline and for facilitating total reconstruction. Osteotomy has been shown to have invaluable and indispensable usefulness for achieving this objective. Many published reports lend support to its usefulness and describe osteotomy as an effective factor in successful reconstruction and in obtaining the
much-desired urinary continence. Addalen emphasizes that symphysis pubis closure, with an interpubic distance as much under 2 cm as possible, is related to a higher rate of urinary continence, allowing the reconstructed bladder neck to be placed intra-abdominally and the urethra to be placed under the symphysis pubis closure. It also enables the urogenital diaphragm musculature to be brought together again, which is a synergic requisite for achieving urinary and fecal continence.

Anterior iliac or unnamed osteotomy was described by Sponseller in 1991. It uses an AO-type tubular frame as a method of external fixation to facilitate highly successful reconstruction. Pelvic ring closure and the reconstruction of deformed organs are principal aims.

In the newborn infant this closure is attained in the first 72 hours due to bone elasticity, without the need for osteotomy. However, in preschool children or older patients, this closure is impossible and osteotomy is required for carrying out total anatomical reconstruction. 5

There are current reports on the use of continent and incontinent urinary diversions. These types of diversions should be chosen exclusively in cases that have been complicated by multiple previous operations and total anatomical urinary reconstruction should be attempted in order to bring the patient as close to normality as possible.

Uncorrected bladder extrophy, together with its devastating psychosocial effect on these patients (social isolation, negative impact in the sexual sphere), also leads to a marked decrease in life expectancy conditioned by recurrent urinary infections and terminal renal failure. 6

**CONCLUSIONS**

Bladder extrophy is a complex congenital malformation involving the urinary tract, skeletal muscle system, and genitals. Treatment is purely surgical and the anatomical complexity involved makes it a great challenge.

The surgical goals include urinary continence and the reconstruction of deformed genitals. Some patients have reached adulthood with no treatment and they require ample reconstruction in order to have a normal life.

Continent urinary diversion is a viable alternative that uses different segments of the digestive tract to achieve urinary continence. Genital reconstruction is required in these unfortunate patients before they reach puberty and adulthood, so that they, too, can aspire to find a mate, have an active sex life, and procreate.
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The present case belongs to the few isolated reports on classic bladder exstrophy in the international literature. Our patient had reached adulthood with no treatment, and underwent urinary, bone, and genital reconstruction, and despite presenting with uterus didelphys, she was able to have a successful pregnancy.

REFERENCES