



# Exstrophy of the bladder

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## ■ ABSTRACT

**Background:** Exstrophy of the bladder is a pathology caused by developmental defect in the cloacal membrane. Described since antiquity, it is a disease with a high degree of family-related recurrence. Surgical intervention in newborns provides good results in bladder function and continence during the neonatal period. Occasionally patients presenting with this pathology reach adulthood without having received the proper medical attention. The case of a female patient whose treatment began in adulthood is presented.

**Clinical case:** A female patient diagnosed with untreated exstrophy of the bladder at 40 years of age while completing a normal pregnancy ending in cesarean section is presented. Due to the changes in the bladder mucous membrane and taking into account the socioeconomic condition of the patient, simple cystectomy and ileal conduit were proposed. The patient was not a candidate for continent reservoir procedure.

**Conclusions:** It is necessary to be familiar with this pathology in which there are still many management challenges as well as a high morbidity.

**Key words:** Exstrophy of the bladder, Primary closure, Secondary closure.

## ■ RESUMEN

**Antecedentes:** La extrofia vesical es una patología ocasionada por el fallo en el desarrollo de la membrana cloacal, descrita desde la antigüedad y con alto riesgo de recurrencia familiar. Los neonatos son intervenidos con buenos resultados para la función vesical y continencia durante el periodo neonatal. Ocasionalmente un paciente con estas características llega a la edad adulta sin haber recibido atención médica adecuada. Se presenta un caso de una femenina que inicia su manejo en la edad adulta.

**Caso clínico:** Femenina con diagnóstico de extrofia vesical clásica, la cual nunca recibe atención adecuada, iniciando su evaluación a los 40 años, cursando además con embarazo normo evolutivo atendido por cesárea. Debido a los cambios en la mucosa vesical y por las condiciones socioeconómicas de la paciente, se propone la cistectomía simple y conducto ileal, no siendo candidata a procedimiento de reservorio continente.

**Conclusiones:** Es necesario conocer esta patología que aun plantea muchos retos en su manejo, presentando una alta morbilidad.

**Palabras clave:** extrofia vesical, cierre primario, cierre secundario.

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## ■ INTRODUCTION

Exstrophy of the bladder is a pathology which still presents many surgical management challenges. Adequate bladder function of emptying and continence as well as reduced morbidity all hinge on the surgery outcome. Diagnosed in the neonatal period, parents receive medical advice and guidance and repair in the neonate is programmed in several stages. This surgery has a high rate of esthetic and functional success.

## ■ CLINICAL CASE

The patient is a 40-year-old married female from Monterrey, Nuevo León, Mexico. She was diagnosed with classic exstrophy of the bladder at birth but due to the low socioeconomic level of her family she never was treated for the condition. She had worn a diaper since infancy and covered the bladder with rough dry cloths that upon removal caused bleeding and ulcerations. She endured this condition for 40 years.

She was married in the year 2000 and months later became pregnant. The pregnancy was normal. During her pregnancy her husband got a job and was able to insure his family in a government health institution (IMSS). The patient was seen medically and scheduled for a cesarean section in the 38th month of her pregnancy.

After her pregnancy and still insured by the IMSS she sought medical attention and was seen by different specialists. She was diagnosed with gastroesophageal reflux and underwent endoscopy that revealed esophagitis, acute antral gastropathy and a 3 cm sliding hiatal hernia. She was treated with proton pump inhibitors and diet.

The patient was referred to the urology service by the family medicine doctor with the diagnosis of incontinency. Bladder exstrophy was corroborated in the first consultation and a department session was called to discuss the case (**Photo 1**).

The patient is obese, weighing 87 kg and measuring 1 m 52 cm in height. General physical examination revealed no other alterations. Genitourinary area examination showed classic exstrophy of the bladder. Bladder diameter was 13 x 17 cm and the walls were ulcerated, bleeding, wrinkled and rough. There were no exophytic or polypoid lesions. Both ureteral meatuses with clear urine ejaculation were observed at the base of the edematous and hyperemic trigone.

The uterine cervix protruded under the trigone presenting a thick whitish exudate. The patient presented with a forked clitoris and no vaginal canal. There was *labia majora* formation and *labia minora* remnants. There were no alterations in the perineum, anus and anal sphincter.



**Photo 1.** Appearance of the bladder at the moment of medical attention. The ulcerated and bloody zone as well as genital alterations can be observed.

Paraclinical tests reported no alterations. Cervix exudate analysis revealed *Cándida sp.* Echosonogram displayed a 10 x 4.7 cm right kidney and a 9.0 x 4.5 cm left kidney. Both were echographically homogeneous and without morphological alterations. Excretory urography was not done.

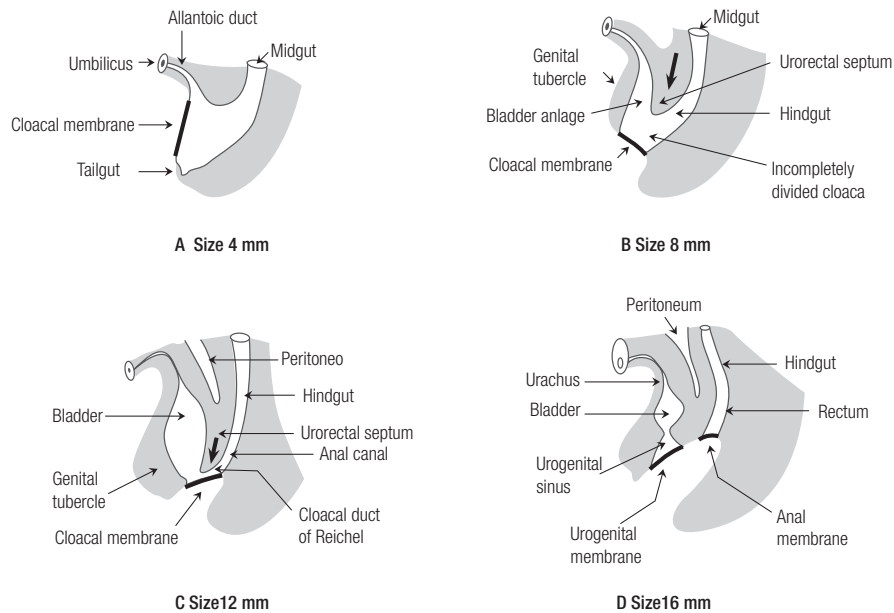
A consensus was reached to define the management of the case. Simple cystectomy and ileal conduit were proposed because exstrophy of the bladder presents a high risk for metaplasia. Socioeconomic conditions of the patient were factors in deciding not to perform continent reservoir procedure.

Simple cystectomy, together with plastic surgery, was carried out and ileal conduit was constructed leaving both ureters free inside it. Prophylactic appendectomy was performed. Plastic surgeons closed the abdominal wall with pediculated flaps and a neovagina with only 3 cm of vaginal canal was formed with flaps from the labia minora. The patient returned 3 weeks later presenting with urine leakage from the Penrose incision. Upon examination dehiscence of the ureteroileal anastomosis was found and repaired. Feeding catheters for temporary external drainage were left in both ureters. After 10 days the patient was released with no urine leakage and an ileal conduit urinary output of 2300 ml per day.

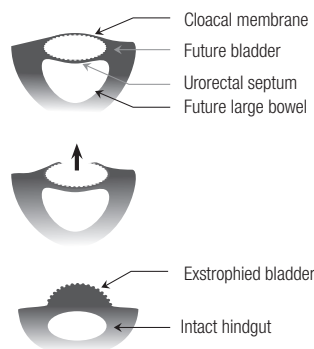
The patient continues to be under surveillance in the hospital out-patient service.

## ■ DISCUSSION

Exstrophy of the bladder was known in Babylon in 2000 B.C.E., von Grafenberg described it in 1597 and a more detailed description by Mowat appeared in 1748. The incidence of the defect is 1 in 36,000 and the man to woman ratio is 2.3:1. The risk of recurrence among relatives is 1 in 100.



**Figure 1.** Development of the caudal region of the embryo.



**Figure 2.** Cloacal membrane development.

In 1968 Marshall and Muecke proposed the theory of embryonal development of exstrophy in which early rupture and an error in the migration of the mesenchyma to the cloacal membrane take place (**Figs. 1 and 2**) (1).

Various risk factors exist such as advanced paternal age, low socioeconomic level, certain types of assisted

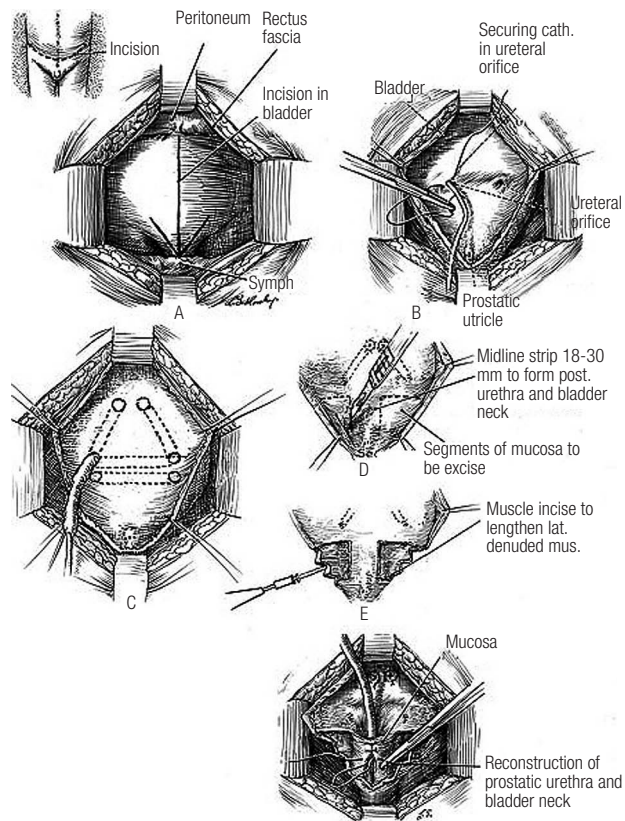
reproduction, previous abortions and even some geographical regions (1,2).

Exstrophy is also associated with other alterations involving the urinary tract such as epispadias, spinal column defects, intestinal alterations, imperforate anus and omphalocele. It can also be associated with kidney and ureteral alterations, reduced size of the penis and cryptorchidism (1,2).

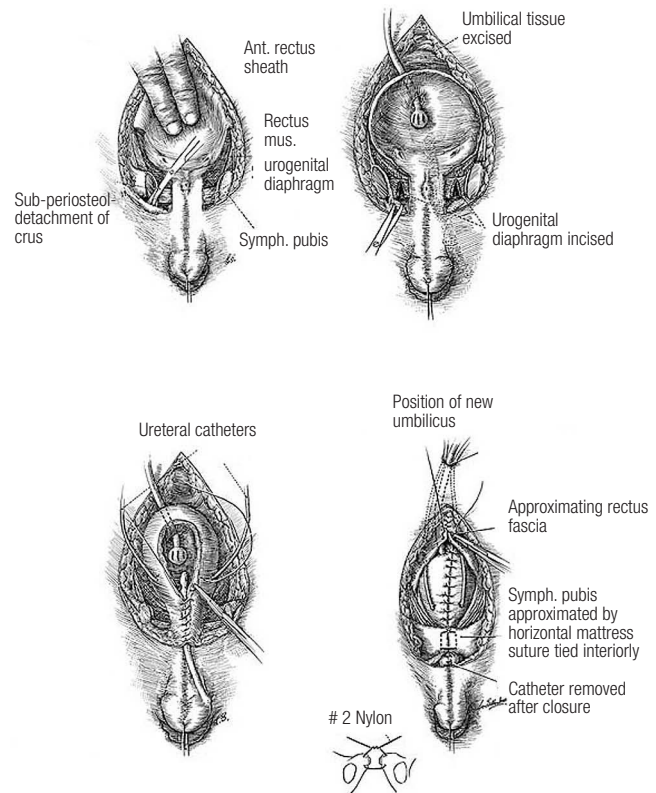
Bladder exstrophy characteristics are absence of the anterior walls of the abdomen and the bladder and absence of the posterior urethra. There is significant diastasis of the pubic symphysis and external pelvic rotation.

Prenatal diagnosis is suspected from ultrasound findings such as absence of bladder filling of the product, low position of the navel, tiny genitals and separation of the pubic branches. When any of these data appear images should be completed with real time high resolution ultrasound, magnetic resonance and sex phenotype determination in addition to routine obstetric evaluation (3).

The following guidelines should be taken into consideration when proceeding to surgical closure: 1) correct placement of the herniated bladder and the urethra within the pelvic ring in relation to muscles



**Figure 3.** Young-Dees-Leadbetter Technique for closing in stages.



**Figura 4.** Technique for closing bladder exstrophy and constructing continent bladder neck.

and bones 2) use of a surgical technique that maintains urinary continence and that preserves kidney function and 3) cosmetic and functional reconstruction of the penis and external female genitalia (4,5,6,7).

There are two schools of surgical repair. One proposes management in stages in which bladder, abdominal wall and posterior urethra closure is the first stage, bladder neck reconstruction and anti-reflux procedure (Young-Dees-Leadbetter technique) is the second stage and epispadias repair (Cantwell-Ransley technique) is the third stage (**Fig. 3**) (6,7,8).

The other proposes primary closure in which the above mentioned stages are carried out in a single surgery. Several authors have reported better results in relation to bladder function (continence and storage capacity) with primary closure because it promotes bladder development and reduces the number of cases requiring amplified cystoplasty. This surgery should be performed within the first 6 to 12 months of the patient's life (**Fig. 4**) (7).

Osteotomies are another important step in hypospadias repair. With this step urinary continence is ensured when the pubic symphysis has been brought closer together and the pelvic ring has been closed, reducing abdominal wall tension. Osteotomy requires a surgeon with experience in pelvic mechanics. If the surgery is contemplated before 72 hours of extrauterine life have passed it only requires the bringing together of the pubic symphysis and its attachment with a suture, with no need of osteotomy (5,8).

The most important complications are uretero-cutaneous fistula, urethral stenosis, prolapsed bladder, tissue loss (penis body and glans) and vesicoureteral reflux and consequent kidney damage. Bladder exstrophy repair should only be carried out by the experienced skilled surgeon.

If surgical techniques do not produce good results or if urinary complications present due to alterations in bladder closure such as fistula or multiple failed interventions, either simple cystectomy with ileal

diversion or continent reservoir is proposed, depending on the patient's sociocultural and socioeconomic situation (5). Psychosocial aspects of the patient should always be taken into account. Being treated as a normal person and having a normal genital appearance and functionality are necessary preventive measures against later psychiatric alterations (9).

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