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**Urology**

# Vesical Extrophy Diagnosed Late: About a Case

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**Abstract** **Case Report**

Vesical exstrophy is a malformation that mainly affects the urinary, genital and musculoskeletal system, whose prevalence is estimated at 2.07/100,000 births. It is usually managed in the first hours of life, but there are still patients who have never consulted for this malformation. We report a case of bladder exstrophy diagnosed late in a 15-year-old patient never treated. Our attitude was the description of the malformation at the anatomical, genital, histological, functional and therapeutic point of view. The patient to benefit from a continental enlargement ileocystoplasty with ostomy.

**Keywords:** Extrophy, Diagnosed.



## INTRODUCTION

Bladder exstrophy is a serious congenital malformation that is characterized by a defect in the closure of the abdominal sub-umbilical wall, the anterior wall of the bladder. Abnormalities of the urethra, pelvic belt, external genitals and perineum are often associated [10]. It is a pathology, whose prevalence is estimated at 2.07 per 100,000 births [1].

At a time when efforts are focused on the pre-natal diagnosis of bladder exstrophy, there are still patients who, despite its impact on their lives, have never consulted for this malformation.

The diagnosis is based on the clinical examination which highlights the bladder plate at the hypogastric region. It presents itself as a red plaque, which bomb under the abdominal thrust and which is entirely part of this wall called the vesical plate associated with epipadias and diastasis of the pubic symphysis [1-4].

The additional examinations, in this case ultrasound and UIV, only allow to appreciate the high urinary tract.

The purpose of the treatment is to obtain a satisfactory urinary continence with preservation of the high urinary tract, based on surgery, which goes from urinary bypass to vessicosphincterian reconstruction. But given the risk of developing cancer on the bladder plate and the frequency of reconstruction failures, many authors opt from the outset for an external urinary bypass continente or ureterrosigmoidosis type.

Neglected bladder exstrophy exposes the two main risks: The impact on the upper device by ascending infection and stenosis due to fibrosis and Cancer [1, 2, 6, 7].

In this article, we report a case of a 15-year-old patient, with a bladder exstrophy never treated before, in order to analyze the particularities of this malformative pathology, epidemiologically, diagnostic and therapeutic.

## PATIENT AND OBSERVATION

Patient 15 years old, single, illiterate, native and resident in Mauritania, who presents a bladder exstrophy since birth never treated, consults for a reddish hypogastric plaque letting flow urine with urinary incontinence. The physical examination had found:

At the hypogastric region, there was a 7 cm large-diameter reddish swelling, roughly triangular at the upper base, with a lower apex leading to the vagina. It was the bladder plate corresponding to the posterior

part of the bladder exposed by the absence of an anterior wall, of pink colour as a whole. At the lower part of this vesical plate, opposite the two hemi-clitoris was visible the vesical abutment of the 2 ureters 3 cm apart (Figure 1).

At the level of the genitals: there was pubic hair distributed in two lateral stripes which continued at the level of the large lips, little marked, with 2 hemi mounts of venus. The large lips were each surmounted by two hemi-clitoris. The little lips were barely visible. The vaginal orifice was narrow.

A Uroscanner had objectified a classic vesical exstrophy complex epipadias and pubic diastasis. Laboratory tests (blood and urine) were within standards. Due to the lack of adequate equipment for her treatment in her home country, the patient was transferred to us for further treatment.

The patient was admitted to the block, we realized a continental enlargement ileocystoplasty with stoma (Figure 2). The surgical follow-up was marked by a good clinical evolution with a good scarring of the abdominal wound, a Redon which stopped bringing back to J3 post operative, a good diuresis which was estimated at 2L per 24h.

## DISCUSSION

The incidence of bladder exstrophy varies across studies: Nelson found 2.5 cases per 100,000 live births [7], Siffel 2.7 cases per 100,000 births [10].

For some authors there is no predominance of one sex over the other. Nelson [7], in his study, showed that the male-female sex ratio is almost equal but that there would be some predominance of this malformation at the level of the white race. Bladder exstrophy can have several aspects. Its complete form is the most common, the partial or enlarged forms, called the cloaca, are much rarer. In its complete form, it concerns the bladder, urethra, anterior abdominal wall, perineum and external genitals.

The diagnosis of this malformation is made especially in antenatal period in the fetus thanks to morphological obstetric ultrasound [4]. Some authors have made the diagnosis in early postnatal or in the early years of the child's life [13]. Others have diagnosed people at least 15 years of age [8]. In his series Traoré found patients aged 24, 25 and 28 [12].

Anatomically, the anterior vesical wall is absent and only the posterior vesical wall, with its two ureteral orifices is called vesical plate. The sphincter apparatus is often hypoplastic and the urethra is constantly malformed in the form of complete epipadias. The abdominal wall is dehiscent at the hypogastric level. The two iliac bones are moved out and the anal sphincter is frequently hypoplastic in its anterior part.

On the genital level, the daughter has clitoral bifidity and the forward displacement of the vaginal orifice which is narrow and sometimes double [5]. In boys, cryptorchidism is common [6] and the penis is wide, short and folded over the abdomen because of the adhesion of the erectile bodies to the intersymphyseal fibrous tissues.

The high urinary tract is usually normal, but there are often abnormalities of the vesico-ureteral junction due to a very short intramural path justifying the need to re-implate the ureters when a vesical reconstruction is performed [7]. Ureteral duplication can be associated with bladder exstrophy, this malformation must be recognized before the operation, to avoid ureteral trauma. Patients not treated in time may develop bilateral hydronephrosis due to the thickness of the bladder wall.

Histologically the bladder plate is abnormal and may be the site of development of vesical adenocarcinomas, especially in patients who live along with their exstrophy [8.

Functionally according to Shapiro [10], the exstrophied bladder has normal neuromuscular activity and should function normally after bladder closure. But studies have shown a significant decrease in detrusory function, even in successful closures [11].

**From a therapeutic point of view**

The treatment of bladder exstrophy in older people as in newborns, remains surgical, it aims to obtain a satisfactory urinary continence, while preserving the high urinary tract. The therapeutic modalities range from urinary bypass to reconstruction surgery [3, 5,12].

According to some authors, urinary bypass is carried out in principle because of the risk of developing cancer and the frequency of reconstruction failures [12]. It is realized of necessity when the reconstruction is unfeasible.

The derivation can be external continental or incontinent skin, it can also be internal, using the anal sphincter as a means of continence.

**External Leads**

Among the external derivations, direct or transintestinal cutaneous ureterostomy is the most widely used. The direct cutaneous ureteromy is indicated when the ureters are dilated because it is possible to perform a transurero-ureteromy that facilitates the apparatus, because there is only one

ostomy. Transintestinal cutaneous ureterostomy using the ileum described by Bricker is the most commonly used.

**Internal Leads**

Internal derivations use the anal sphincter as a means of continence and can therefore only be performed if it is normal, the ureters are not dilated and renal function is within the limits of normal. Several interventions are described.

**Continental external bypasses**

They derive the urine to an intestinal reservoir usually made from ileocecum. Stein [1] uses the Mainz I pocket, but in our training the tank used is the continental ileal pocket with stoma [17].

There is still no consensus on the best surgical technique [13, 18]. Step reconstructive surgery is the most recommended strategy [19]. But even though it is the ideal procedure, it is not without complications. Indeed, besides the frequency of failures even in expert hands, hydronephrosis, vesico-ureteral reflux and deterioration of the high urinary tract are not uncommon [20, 21]. Stein [12], to avoid all these complications and encouraged by the good results of ureterrosigmoidotostomy [22], proposes a primary urinary derivation rather than a vesicular reconstruction.

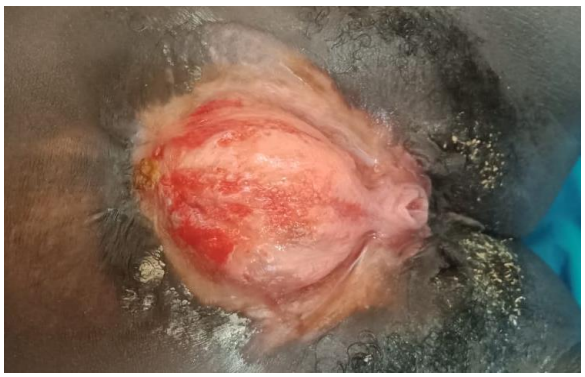

**Fig-1: Image en préopératoire**

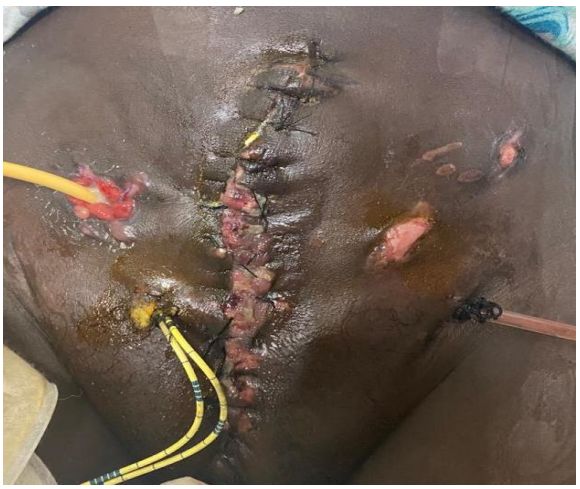

**Fig-2: Image en postopératoire**

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