



Bladder Exstrophy: A Case Report at Abass Ndao Hospital Center

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ABSTRACT

Bladder exstrophy is a rare and severe congenital malformation belonging to the exstrophy-epispadias complex. It can be diagnosed prenatally using morphological ultrasound, but in resource-limited countries, it is often diagnosed at birth due to insufficient access to specialized obstetric care. We report the case of a 7-year-old girl with bladder exstrophy diagnosed at birth but treated late in our department, in order to highlight the difficulties in accessing specialized care in our setting and to discuss the diagnostic, therapeutic, and prognostic challenges of this rare malformation.

Keywords: Bladder exstrophy; Bladder reconstruction; Urological malformation; Senegal

INTRODUCTION

Bladder exstrophy is a rare and severe congenital malformation belonging to the exstrophy-epispadias complex. It is characterized by a failure of closure of the infraumbilical abdominal wall and the anterior wall of the bladder, associated with anomalies of the urethra, pelvic girdle, external genitalia, and perineum [1]. Its incidence is estimated to be between 1/30,000 and 1/50,000 live births, with a male predominance, the sex ratio varying from 2 to 3 boys to one girl [2,3].

The diagnosis is most often established prenatally using morphological ultrasound. Suggestive signs can be identified as early as the first trimester of pregnancy, around 11 weeks of amenorrhea, primarily the persistent non-visualization of the fetal bladder, observed in nearly 80% of cases [2]. In the second trimester, the absence of bladder visualization, which should normally be identifiable from 16 weeks of

gestation regardless of its state of fullness, constitutes a major warning sign. Other associated anomalies can also guide the diagnosis, including a subumbilical mass, a low-lying umbilical cord, or diastasis pubic bones.

Despite advances in prenatal imaging, some cases are still diagnosed only in the early postnatal period, or even, exceptionally, in adulthood [1]. In resource-limited countries, prenatal diagnosis remains insufficient due to limited access to specialized obstetric care and high-quality morphological ultrasound. Furthermore, some newborns diagnosed at birth are not referred to specialized centers due to a lack of information among families, difficulties in navigating the healthcare system, or socioeconomic and financial constraints [4]. These diagnostic and

therapeutic delays can worsen the functional and psychological prognosis of patients.

The management of bladder exstrophy is primarily surgical and ideally begins in the neonatal period. Early closure of the bladder and abdominal wall promotes bladder development, improves the chances of achieving satisfactory urinary continence, and helps preserve long-term renal function [5,6].

We report the case of a 7-year-old girl diagnosed at birth but treated late in our department with bladder exstrophy, in order to highlight the difficulties in accessing specialized care in our setting and to discuss the diagnostic, therapeutic, and prognostic challenges of this rare malformation.

CLINICAL OBSERVATION

The patient was a 7-year-old girl, not attending school, brought in by her parents for a subcutaneous mass in her lower abdomen, associated with persistent urinary incontinence since birth. The child was born in a

public hospital where a diagnosis of bladder exstrophy was made at birth. However, treatment was delayed due to financial difficulties. Her perinatal history included irregular prenatal visits and no prenatal diagnosis.

On clinical examination, the child was in good general condition with normal growth and psychomotor development. Abdominal examination revealed a reddish, ovoid mass measuring approximately 10 cm along its longest axis and 7 cm along its shortest axis, corresponding to the protruding bladder plaque. The ureteral orifices were visible at the upper part of the plaque, with continuous urinary leakage. The abdominal wall around the exstrophy plaque was supple, with no signs of local infectious complications.

Examination of the external genitalia revealed a malformed vulva with bifid clitoral structure corresponding to two hemiclitorises. The labia majora and minora were difficult to distinguish. A small, median depression corresponding to the vaginal opening was identified (Figure 1).



Figure 1: Preoperative clinical appearance of bladder exstrophy.

The Urinary CT scan revealed bladder exstrophy associated with a 56 mm dehiscence of the anterior abdominal wall muscles below the umbilicus,

complicated by an uncomplicated hernia containing digestive contents. There was also a diastasis of the pubic symphysis measuring 83.7 mm (Figure 2).



Figure 2: Axial section of the uro-scanner.

Blood and urine tests were unremarkable. Initial surgical management consisted of closure of the

bladder plaque combined with abdominal wall reconstruction (Figure 3).

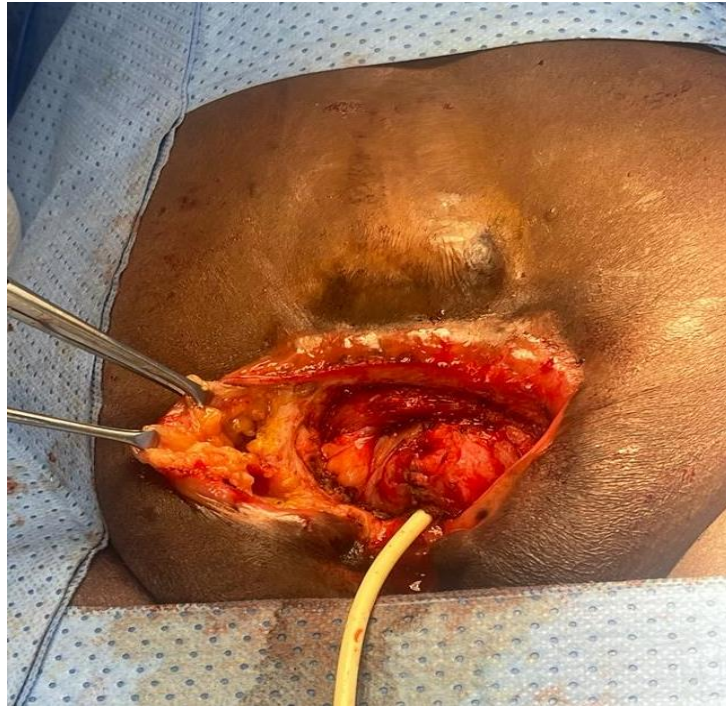


Figure 3: Intraoperative view after dissection of the bladder exstrophy plaque.

The postoperative course was uneventful, marked by good wound healing (Figure 4). However, restoring

urinary continence remains a major therapeutic challenge to this day.



Figure 4: Postoperative appearance after abdominal wall reconstruction.

DISCUSSION

Bladder exstrophy is a rare congenital malformation. Its incidence is estimated to be between 1 in 30,000 and 1 in 50,000 live births, depending on the study, with a male predominance [7].

The diagnosis of bladder exstrophy relies primarily on morphological obstetric ultrasound performed during pregnancy. This examination is now the cornerstone of prenatal screening for this malformation and allows for appropriate neonatal management [2,7].

The diagnostic performance of prenatal ultrasound has improved considerably over the last few decades. Alexander M et al report a sensitivity of up to (82.8%) when characteristic ultrasound signs are systematically sought [8].

Despite these improvements, prenatal diagnosis remains inadequate in many resource-limited countries. Several factors explain this situation, including limited access to specialized morphological ultrasound, insufficient prenatal care, and a lack of awareness of the ultrasound signs of this rare affection. Thus, Siffel et al. report that, although the diagnosis can be made during the fetal period, it is still frequently made at birth [9].

More rarely, very late diagnoses have been described in adolescence or adulthood [10]. Traoré et al., in particular, reported cases diagnosed in patients aged 24, 25, and 28 years [4]. These diagnostic delays are generally observed in disadvantaged socio-economic contexts, characterized by limited access to healthcare facilities, insufficient prenatal monitoring, the absence of obstetric ultrasound and sometimes the occurrence of home births [11].

In our observation, the diagnosis was made at birth in a hospital setting. However, no prenatal diagnosis had been performed due to irregular prenatal care. Furthermore, surgical intervention was delayed until the age of 7 due to financial difficulties. This situation illustrates the persistent challenges faced by health systems in resource-limited countries, where access to specialized prenatal diagnosis and reconstructive care remains insufficient. It also underscores the importance of strengthening prenatal screening and improving access to specialized care to optimize the functional and psychosocial prognosis of children with bladder exstrophy. Fetal Magnetic Resonance Imaging (MRI) plays a complementary role in the prenatal diagnosis of bladder exstrophy. Although obstetric ultrasound remains the gold standard for initial screening, MRI may be indicated when ultrasound data are equivocal or when anatomical analysis is limited by certain technical conditions, including

maternal obesity, oligohydramnios, or an unfavorable fetal position [12,13].

According to Hsieh et al., MRI performed around 32 weeks of gestation is the preferred examination for the precise characterization of severe urogenital malformations [14]. Thanks to its excellent tissue resolution, it allows for better analysis of fetal pelvic anatomy, particularly the ureteral junctions at the level of the protruding bladder plate. It also facilitates the determination of fetal sex, which is sometimes difficult to establish by ultrasound due to associated genital anomalies.

In our case, MRI was not performed. CT urography allowed for the precise evaluation of the bladder wall dehiscence and pubic diastasis, providing the anatomical information necessary for surgical planning.

CT scanning is a useful complementary examination in the anatomical evaluation of bladder exstrophy, particularly in cases diagnosed late. Although not the first-line diagnostic tool for this malformation, it allows for a precise analysis of associated abdominal wall, bone, and pelvic anomalies. Thanks to its multiplanar and three-dimensional reconstructions, the CT scan accurately assesses the extent of the anterior abdominal wall defect, the severity of the pubic diastasis, and the anatomical relationships of the pelvic structures.

The treatment of bladder exstrophy relies primarily on reconstructive surgery. The main objectives are closure of the bladder and abdominal wall, preservation of upper urinary tract function, achievement of satisfactory urinary continence, and reconstruction of the external genitalia to allow for harmonious psychosexual development.

Ideally, treatment is initiated in the neonatal period. Early closure of the bladder plate promotes bladder growth, reduces the risk of infection, and optimizes the chances of achieving urinary continence later in life. Several teams recommend primary closure within the first 48 to 72 hours of life, a period during which the flexibility of the pelvis also facilitates the correction of the pubic diastasis.

Two main surgical strategies are described: Complete Primary Repair of Exstrophy (CPRE), performed in a single surgical procedure, and conventional multi-stage reconstruction involving initial bladder closure, epispadias repair, and then bladder neck surgery when bladder capacity becomes sufficient [15,16]. The choice of technique depends on the patient's age, the

surgical team's experience, and the anatomical characteristics of the malformation [17,18].

Despite advances in reconstructive surgery, achieving complete urinary continence remains a major therapeutic challenge. Some patients require additional interventions such as bladder neck reconstruction, augmentation cystoplasty, or the creation of a continent conduit. Long-term follow-up is essential to monitor renal function, urinary continence, recurrent urinary tract infections, and aesthetic and psychosexual outcomes [19-22].

In our case, closure of the bladder plaque combined with abdominal wall reconstruction resulted in satisfactory anatomical correction with a straightforward postoperative course and good wound healing. However, as in many series in the literature, restoring urinary continence remains the primary therapeutic challenge and may require additional reconstructive procedures during follow-up.

The therapeutic delay observed in our case could compromise optimal bladder development and reduce the chances of achieving satisfactory urinary continence compared to patients who undergo surgical correction in the neonatal period [5]. Indeed, early intervention promotes better bladder growth and optimizes long-term functional outcomes. In the literature, continence rates after bladder exstrophy reconstruction generally vary between 60% and 90%, depending on the surgical strategy adopted, the age at the time of the procedure, the patient's anatomical characteristics, and the possible use of additional reconstructive procedures [23,24].

Beyond the anatomical and functional consequences, bladder exstrophy exposes children to difficulties in social integration, altered body image, and an increased risk of dropping out of school, particularly when treatment is delayed.

CONCLUSION

Bladder exstrophy is a rare and complex congenital malformation usually diagnosed prenatally or at birth. Despite advances in prenatal screening and reconstructive surgery, delays in treatment persist in resource-limited countries due to socioeconomic constraints and insufficient access to specialized care.

Improving the prognosis of bladder exstrophy relies on strengthening prenatal diagnosis, early referral to specialized centers, and multidisciplinary care from the neonatal period onward to optimize functional, aesthetic, and psychosocial outcomes.

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