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Classic Bladder Exstrophy Lately Treated: A Case Report

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Abstract: The exstrophy of the bladder is a serious congenital malformation characterized by a defect in the closure of the inferior abdominal wall, the anterior wall of the bladder, often associated with an abnormality of the urethra, pelvic girdle, external genitalia and of the perineum. While the diagnosis of this malformation is generally made in the antenatal period and is based on a failure to visualize the bladder on morphological ultrasound in the second trimester, in some patients the diagnosis is either made in the early postnatal period or in adulthood. In African societies, the birth of a malformed child is experienced as a real tragedy, taking into account the mystical-religious considerations that surround it on the one hand and the weight it constitutes for families on the other. We report our experience about a case of the bladder exstrophy diagnosed and treated late in a 12-year-old child at university clinics of Lubumbashi in Democratic Republic of the Congo.

Keywords: Case Report**,** Congenital Malformations, Bladder Exstrophy, Epispadias, Urology

1. Introduction

The Classical exstrophy of the bladder is part of the exstrophy-epispadias complex (EEC), a group of embryological abnormalities characterized by partial or total exposure of the mucosa of the lower urinary tract to the external environment through a defect in the anterior wall of the abdomen [1, 2]. In classical bladder exstrophy, the bladder is presented as an open plaque in the hypogastrium, always associated with epispadias [3].

In boys, genital anomalies are represented by a separation of the corpora cavernosa, which is shorter than normal and has a prominent dorsal cord. The urethral plaque is also shorter and the glans is bifid, everted, covered with the foreskin only on its ventral surface [9]. In girls, the clitoris is bifid, the pubic mount and labia minora are divergent, the urethra and vagina are shortened, the vaginal opening is displaced anteriorly and is often stenotic [12].

It is a rare condition, occurring globally in 2.1 to 4.0 OUT of 100,000 births, but its incidence in Africa is not known [4]. It mainly affects boys and its diagnosis can be made during the prenatal period. The management is done either early (48 to 72 hours of life) or late (6 to 12 weeks) according to various techniques, each with its advantages and disadvantages [5]. In Africa, affected patients present late, around 6 to 9 years of age, which considerably delays management and functional prognosis [1, 6]. We present to you a case of a 12-year-old boy seen late in order to provide our therapeutic approach and the results of which seemed encouraging to us.

2. Patient and Observation

The patient is a 12-year-old boy, whose parents came to consult for a congenital hypogastric mass letting urine flow. Since birth, the parents have not consulted any medical facility and no management of the anomaly has been made.

Her mother is 25 years old and had not had any prenatal consultation during the pregnancy, which reportedly went smoothly. The parents are of low socio-economic status and illiterate. No notion of consanguinity has been identified.

General physical examination of the patient did not reveal any particulars. On examination of the abdomen, the bladder appears as a conoid hypogastric mass with a height of about 3.5 cm, whose oval base has a large horizontal axis, about 7 cm and a small vertical axis, 5 centimeters. Its anterior top is light brown in appearance. The rest of the mass is pink, its base continues with the anterior wall of the abdomen. The adjacent skin is erythematous. Presence of 2 lateral ostia, one on the left and the other on the right, low-located on the hypogastric mass, continuously ejecting urine (ureters) (Figure 1). Note also the presence of a pubic symphysis disjunction on palpation. Examination of the external genitalia showed dorsal separation of the cavernous bodies, with exposure of the urethral plaque, following the herniated bladder mucosa in the hypogastric region. The hemi-glans are everted, united at their ventral surfaces by hemi-foreskins merging on the median line and absent on the dorsal surface. The scrotum has longitudinal folds, the testes are present in the hemi-bursas and retain their physiological sensitivity. The anus is in the urogenital triangle, 1 centimeter from the bi-ischial line. Its sphincter is tonic.

Abdominopelvic ultrasound revealed no upper urinary tract abnormality and pelvic x-ray revealed a gap at the site of the pubic symphysis of 5 centimeters. (Figure 2).

The patient underwent repair using the two-stage exstrophy reconstruction method while taking care to place the ureteral catheters (Figure 3). Taking into account the patient's age, the first and second steps were carried out during the same intervention, thus resulting in the closure of the bladder (Figure 4) and the repair of the epispadias according to the Ransley technique, without osteotomy. The postoperative consequences were complicated by a urinary tract infection with the release of the threads from the epispadias reconstruction (Figure 5). The child was taken back three months later for the penis plasty (Figure 6). The post operative course was simple.

3. Discussion

The exstrophy of the bladder is part of the exstrophy-epispadias complex, a set of developmental abnormalities, also called lower coelosomy, resulting in various defects affecting the urinary and musculoskeletal systems, the pelvis, the pelvic floor, the abdominal wall, the external genitalia and sometimes the spine and anus. Lower coelosomy can be categorized into typical (Epispadias, Classic Bladder Exstrophy and Cloacal Exstrophy) and atypical (Duplicated Exstrophy, Covered Exstrophy and Pseudo-exstrophy) [4]. The generally accepted mechanism is that of an early rupture of the cloacal membrane due to lack of development of the underlying mesoderm, which contributes to the formation of the pelvis and muscles of the abdominal wall. Regarding the lack of mesoderm development, experts have yet to find a compromise. Muecke's hypothesis suggests that an abnormally larger cloacal membrane has a blocking effect in the development of the mesoderm [9]. Another hypothesis, based on clinical observations which concluded in a cephalic displacement of the base of the penis (or of the clitoris) suggested that the abnormally more cephalic origin of the genital tubercle interferes with the development of the mesoderm, the genital tubercle behaving like a wedge [10]. In this case, it is a classic form combining an exstrophy and an epispadias (figure 1).

Classical bladder exstrophy is characterized by the herniation of the posterior wall of the bladder through the hypogastrium and is always associated with epispadias. It can occur in isolation, as in the case of our patient, or in a polymalformative context [7].

It is a rare disease, but its incidence in African countries is unknown. Several factors contribute to this situation, including the fact that newborns with severe forms are neglected and die early [1]. Although some studies report an equal frequency in both sexes, it is generally accepted that there is a clear predominance of men. Our patient is male and no epidemiological study in our possession can specify the frequency.

Due to the rarity of the disease, the exact cause is not known. Some risk factors have been identified. These are white race, young age and maternal multiparity as well as low socioeconomic status. The risk is higher in members of the same family, which has raised suspicion of a genetic mechanism. Recently, a probable candidate gene was identified, ISL1, and studies are underway to clarify its implication [1, 8]. In this case, let us note the maternal multiparity, the low socio-economic level, the black race, no notion of consanguinity. We believe that the literature blames the white race for the study poverty in the black race.

The diagnosis of bladder exstrophy can be suspected as early as the first trimester. Indeed, since renal function establishes between the 9^{th} and 11^{th} week of amenorrhea, the bladder can be recognized as early as the 11^{th} week. Its non-visualization during 2 ultrasound examinations spaced 30 minutes apart should raise the suspicion of bladder exstrophy, which is confirmed from the 16^{th} week, during the second trimester. Checking for associated abnormalities completes the remainder of the examination [7, 11]. In this case, no maternal follow-up during gestation, no antenatal consultation and no ultrasound examination was performed. The diagnosis was made in a remote setting after childbirth and no treatment was established.

In late presentations, failure to close the bladder constitutes a persistent trauma, leading to inflammation, fibrosis, metaplasia and carcinoma later in life [1].

Pelvic abnormalities are bone and muscle. Pubic diastasis is constant, causing eversion of the pubic branches at the iliac and ischial junctions. The levator ani are more posterior and in external rotation, while the puborectal muscle is depressed. This exposes this type of patient to pelvic organ prolapse [12]. In this case, we noted a pubic diastasis of about 5 cm with a duck gait.

The initial management aims to protect the bladder plaque from any trauma, which can come from a compress, clothes or the umbilical cord clamp. It is for this reason that the cord must be clamped using wire and the bladder plaque covered with plastic material, sucking irrigated with physiological saline each time the diapers are changed. Then, the newborn is transferred to a tertiary hospital center. In poor countries, where health coverage is non-existent, parents most often have to prepare financially before any transfer, this delays the care, as in our case where the newborn was treated at 20 months old [1]. Early definitive management, no later than the 72nd hour of life, is recommended in settings with large neonatal intensive care units. Otherwise, late management, between 6 and 12 weeks is recommended. In addition, late management gives the possibility of stimulating the penis with testosterone, which reduces damage to the glans during the procedure [5]. In our case, we did not have the opportunity to choose between these two schools given the delay in presenting our patient.

Since 1950, several techniques have emerged for the treatment of exstrophy of the bladder. The principle is to close the bladder and the abdominal wall, to repair the urethra. The most popular are: the Modern Staged Repair of exstrophy (modern reconstruction of exstrophy in different operating stages) proposed by Jeff and Gerhart, the Complete Primary Repair of Bladder Exstrophy (Complete Repair of bladder exstrophy in one stage), proposed by Grady and Mitchell and the Radical Soft Tissue Mobilization proposed by Kelly. The technical choice depends on the surgeon's habits. So, in our case, the technique used for the repair of the bladder was the SERM and the epispadias was repaired according to the Ransley technique which consists of an incision then a Dorsomedial anastomosis of the corpora cavernosa above the urethra [2].

Nowadays, this technique has been modified by allowing better reimplantation of the urethra between and below the corpora cavernosa, this is the Cantwell-Ransley technique [5]. Other epispadias repair techniques, such as Mitchell's (complete penile disassembly) and Kelly's have in common that they require careful dissection of the anatomical layers in order to maintain the vasculonervous structures of each structure. The lesion of which will lead to erectile dysfunction and atrophy of the cavernous bodies. Everything is done under magnifying glasses, which we do not have in our environment.

Regardless of the technique used for bladder reconstitution, osteotomies have long been considered as an obligatory adjuvant. Their roles are to allow easy symphysal approximation, to secure the suture of the abdominal wall by reducing the tension and the positioning of the entire bladder in the pelvis. However, recent studies have shown that there is no statistically significant difference between patients who have had an osteotomy and those who have not [13]. Other studies report good results apart from the use of osteotomy (95% success) and complete pelvic girdle closures without osteotomy [14-16]. Based on these recent reports, we did not prefer to perform an osteotomy.

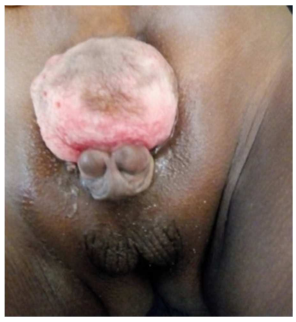

Figure 1. *Image showing the posterior bladder wall with a bifid penis, typical of classic bladder exstrophy.*

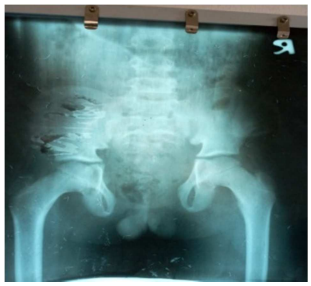

Figure 2. *Radiograph showing the pubic diastasis on an anteroposterior incidence.*

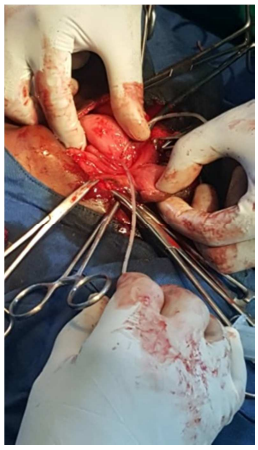

Figure 3. *Image showing ureteral ostium after placement of ureteral catheters.*

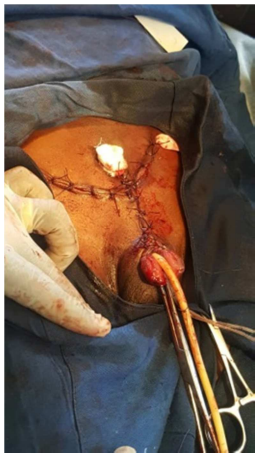

Figure 4. *Image showing the aspect of the anterior wall of the abdomen after vesical repair and coeliorraphy.*

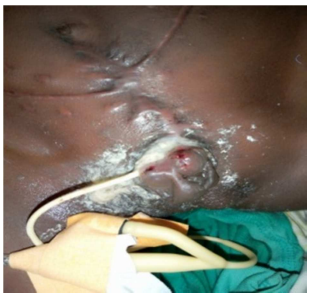

Figure 5. *Image showing urinary infection with release of the plasty threads.*

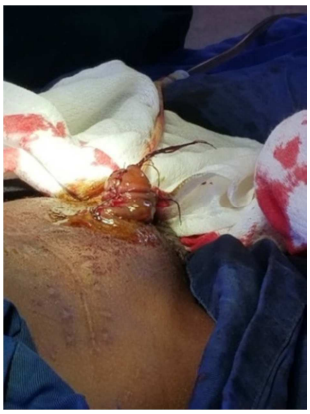

Figure 6. *Image showing the aspect of the penis after repair of the epispadias.*

4. Conclusion

Bladder exstrophy is a very rare congenital malformation whose lack of direct life-threatening prognosis contributes greatly to delayed diagnosis in Africa. The management is a real challenge for the surgeon, so that there are several techniques trying to provide solutions as the lack of consensus in the final management is glaring. Most of the treatment is aimed at anatomical reconstruction while preserving as much as possible the chances of normal bladder-genital function. The diagnosis is made antenatally, hence the need for prenatal consultations which unfortunately are not often followed by pregnant women in rural areas. Delay in care is a factor of poor prognosis

Conflicts of Interests

The authors declare no conflict of interest.

Contributions of the Authors

Pitchou Mbey Mukaz initiated the drafting of this article. Dimitri Kanyanda Nafatalewa and Augustin kibonge Mukakala, wrote and arranged according to the recommendations of the review. Florent Tshibwiz a Zing made the review of the literature. Igor Mujinga wa Mujinga, Serge Ngoie Yumba, Eric Mbuya Musapudi, Vincent de Paul Kaoma, Prince Muteba katambwa, and Jeff Bukasa Misenga read and corrected the article.

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