Journal of Young Pharmacists, 2024; 16(1):134-136.
https://www.jyoungpharm.org

Case Report

A Rare Scenario: A Female Child with Bladder Exstrophy and Complications

Akshi Prajapati*, Aastha Shah, Chestha Shah, Rahul Gandhi, Neha Suratiya

Department of Pharmacy Practice, Parul Institute of Pharmacy, Parul University, Limda, Vadodara, Gujarat, INDIA.

ABSTRACT

Bladder Exstrophy is an anomaly that is uncommon from birth and influences various aspects of the urinary tract, genitals, pelvis, and anus. The defining characteristic of this condition is the abnormal development of the bladder outside the fetal body. Clinical signs in affected individuals often include the bladder being exposed from the abdomen, a flattened puborectalis sling, separation of the pubic symphysis, and ulcers in the bladder mucosa, urine dribbling, and skin excoriation. The primary treatment for bladder exstrophy typically involves surgery, tailored to the severity of the condition. In one case, a female child was passing urine through her navel instead of the urethra. Following a confirmed diagnosis, a single-stage repair was recommended for the exstrophied bladder. However, before a scheduled follow-up appointment, the patient's stitches ruptured, resulting in urine leakage from the operated site and pus discharge from the urethra. Based on the patient's complaints and laboratory results, a diagnosis of acute kidney injury and urinary tract infection in the context of bladder exstrophy was established, leading to the plan for a resuturing surgery. It's important to note that the risk of bladder exstrophy is increased when the mother is exposed to smoking and radiation during the first trimester of pregnancy. Bladder exstrophy in a female child is a rare and complex congenital condition that necessitates a multidisciplinary approach for diagnosis, management, and long-term care.

Keywords: Epispadias, Cloacal exstrophy (OEIS Syndrome), Hydronephrosis, Urinary Tract Infection (UTI), Acute Renal Failure (ARF), Severe Cystitis.

Correspondence:
Ms. Akshi Prajapati, Pharm D
Department of Pharmacy Practice, Parul Institute of Pharmacy, Parul University, Limda, Vadodara -391760, Gujarat, INDIA.
Email: akshiprajapati858@gmail.com

Received: 29-11-2023;
Revised: 05-12-2023;
Accepted: 31-12-2023.

INTRODUCTION

Bladder Exstrophy is an innate abnormality signified by deformities in various aspects of the urinary tract, genitals, pelvis, and anus. This rare birth defect's defining feature is the bladder's development outside the foetal body. In this condition, the exposed bladder lacks the ability to store urine properly, as the bladder muscles do not develop normally, resulting in a continuous outflow of urine.[1] The exact etiology of bladder exstrophy is not well understood, and it is believed to occur during the 11th week of pregnancy. Inherited and external influences may play a role in its development. While this defect can sometimes be visualized during prenatal ultrasonography, it may not always be detected in all foetuses. Post-birth diagnosis typically involves skiagraphs of the pelvis and abdomen to assess the extent of diastasis symphysis pubis, followed by surgical intervention. Diagnosis confirmation may also involve cystoscopy examination and urodynamic testing.

DOI: 10.5530/jyp.2024.16.18

Publishing Partner : EManuscript Tech. [www.emanuscript.in]

Common clinical signs in affected patients include Exposed bladder from the abdomen, Flattened puborectalis sling, Separation of the pubic symphysis, Ulcers in the bladder mucosa, dribbling of urine, Skin excoriation.[1] To gain a comprehensive understanding of this condition, it's essential to first grasp the bladder's anatomy. Typically, the bladder is a hollow, muscular, pear-shaped organ located in the lower part of the abdomen, supported by pelvic muscles within the pelvic floor. The pelvic floor acts as a muscular diaphragm that provides support to the pelvic organs. Urine is received into the bladder through the ureters, thick tubes connecting each kidney to the upper part of the bladder. The bladder collects urine and ultimately expels it through the urethra. The bladder's base, known as the fundus, is formed by its posterior wall and contains the trigone, which is the structure housing the bladder's outlet, the urethra. Under normal conditions, the bladder is positioned in the lower abdomen and is relaxed when empty.[2] When the kidneys produce urine, nerve signals are sent to the brain, creating an urge to urinate. In response, the brain's nerve signals prompt the bladder muscles to contract, expelling urine through the urethra. Typically, the human bladder can hold a volume ranging from 400 mL to 1000 mL, with an average capacity of 400 to 600 mL, although these figures can vary among individuals.[3] With a clear understanding of bladder anatomy, we can delve into the pathogenesis of

bladder exstrophy. During foetal development, a temporary tissue called the cloacal membrane covers the lower abdomen and is later replaced by maturing abdominal muscles. If the cloacal membrane ruptures before the abdominal muscles fully form, it can result in a condition known as exstrophied bladder. Another tissue, the urorectal septum, separates the developing bladder from the bowels and intestines. Depending on whether the cloacal membrane ruptures and forms an urorectal septum or not, two conditions may develop. Epispadias, is often manifested as a typical bladder exstrophy, is commonly identified either at birth or shortly thereafter. When epispadias is associated with bladder exstrophy, it becomes an uncommon birth defect characterized by the underdevelopment of the urethra into a complete tube and the abnormal expulsion of the urine from the body. Cloacal exstrophy, is otherwise called as OEIS syndrome, is an unusual congenital malformation where babies are born with underdeveloped lower abdominal organs, some of which may be visible outside the body after birth.[4] Epispadias affects 1 in 117,000 newborn males and 1 in 484,000 newborn females. OEIS syndrome, on the other hand, affects 1 in 250,000 births and is more prevalent in males.[5]

Treatment for bladder exstrophy primarily involves surgery, with the approach depending on the severity of the condition. The goals of treatment include closing the bladder to allow proper urine storage without harming renal function and reconstructing the external genitalia for both functional and cosmetic purposes.

There are two primary categories of surgical procedures

1. Complete Repair: In this procedure, the bladder and abdomen are closed, and the urethra and external genitalia are reconstructed in one operation. This surgery is typically performed in a newborn or an infant aged 2-3 months. It may also involve reconstructing the pelvic bones.[6]
2. Staged Repair: Staged repair involves three separate procedures. The first procedure closes the bladder and abdomen within 72 hr of the infant's birth. The second procedure, performed when the infant is 6-12 months old, reconstructs the urethra and external genitalia. The third procedure is done once the patient is mature enough to participate in toilet training.[7]

Each surgical approach has its merits and is chosen based on individual patient circumstances and surgeon preference.

CASE REPORT

A female child was delivered vaginally at a government hospital. Upon birth, it was noticed that the baby was passing urine through the navel region rather than the urethra. Due to the unavailability of appropriate treatment at the government hospital, the infant was transferred to a multispecialty hospital seven days after birth, where she was admitted to the neonatal ICU. The parents expressed concerns about their child's continuous urination through the anterior abdominal wall since birth. Consequently, the physician recommended a series of laboratory tests that encompass a full blood count, urine culture with sensitivity analysis, and blood culture with sensitivity analysis, various biochemical tests, and serological examinations. The results indicated an elevation in neutrophils (45%), the presence of numerous pus cells, gram-negative bacilli, and budding yeast-like cells with pseudo hyphae, suggesting the presence of *E. coli* and *Enterococcus faecium*. Additionally, there was an increased serum creatinine concentration (1.1 mg/dL) and abnormal electrolyte concentrations in the serum, including high sodium (148 mmol/L), potassium (5.6 mmol/L), and chloride (115 mmol/L) levels. Further investigations through ultrasound of the kidney-ureter-bladder revealed an approximate 8 mm-sized defect in the anterior abdominal wall exhibits a protrusion of the anterior bladder wall, suggesting a potential diagnosis of bladder exstrophy. After confirming this diagnosis, the patient was advised to undergo a single-stage repair for the exstrophied bladder. Before the surgery, the patient received six days of infection treatment, including intravenous medications such as INJ. CEFOTAXIME (1.3 mL+5 mL NS), INJ. GENTAMICIN (0.3 mL+5 mL NS) (discontinued after 2 days), INJ. AMIKACIN (0.5 mL+2 mL NS), and INJ. AUGMENTIN (1.5 mL+3 mL NS). On the 5th day, INJ. CEFOTAXIME was replaced by INJ. PIPRACILLIN TAZOBACTAM (1.3 mL+2 mL NS). The surgery was successfully performed on the 7th day of admission. Post-surgery, the patient was closely monitored and received care that included INJ. AMIKACIN, INJ. PIPRACILLIN TAZOBACTAM, AND INJ. LINEZOLID (13.5 mL slowly over 30 min) before being discharged. The parents were advised to maintain proper wound care, including changing dressings and applying betadine solution. However, before a scheduled follow-up appointment, the patient's stitches ruptured, and urine leaked from the operated site, accompanied by pus discharge from the urethra. The physician recommended further laboratory investigations, revealing an elevation in neutrophils (34%), monocytes (8%), eosinophils (10%), and platelet count (5,97,000). Additionally, there was a decline in RBC count and an increase in CRP quantitative levels (30.90mg/L), abnormal electrolyte concentrations in the serum, including high sodium (149 mmol/L), potassium (3.4mmol/L), and chloride (117 mmol/L) levels. Renal function tests indicated a gradual increase in blood urea levels (50 mg/dL). The abdominal and pelvic ultrasound indicated a mildly expanded urinary bladder with irregular thickening of the mucosal wall and debris, as well as significant hydronephrosis and severe cystitis. Considering the patient's complaints and laboratory results, a diagnosis of acute renal failure and urinary tract infection in the case of bladder exstrophy was established. A resuturing surgery was planned for

10 days post-admission. During the period between admission and the second surgery, the patient received treatment for acute renal failure and urinary tract infection, including medications such as Syrup Nitrofurantoin (2.3 mL), Suppository Anomol, Syrup Linezolid (1.5 mL), Inj. Ceftriaxone (1 mL+10 mL Over 20 Min), Inj. Piperacillin-Tazobactam (2.8 mL+5 mL NS), Inj. Calcium Gluconate (5 mL/10 mL NS), Neb. Levolin (0.3 mg/2.5 mL), Inj. Paracetamol (0.2 mL+2 mL NS), Inj. Pantoprazole (0.6 mL+2 mL NS), Vitamin D 3, Vitamin A and multivitamin B Complex. The second surgery was performed successfully, and the patient was discharged after 15 days. Once again, the parents were advised to maintain proper wound care.

DISCUSSION

Being a complex and rare condition, there may be many factors that may lead to bladder exstrophy. The primary consideration is to determine if any problems arose during the pregnancy, childbirth, or in the aftermath of the baby's arrival. Epidemiologically, bladder exstrophy occurs in approximately 1 in every 50,000 births, with a higher prevalence in males than in females. The risk of bladder exstrophy is elevated when the mother is exposed to smoking and radiation during the first trimester of pregnancy. Infant's life can be substantially impacted by this condition, and therefore psychological and emotional support is equally vital as the child matures. In female patient with bladder exstrophy potential reproductive tract anomalies may impact fertility. It is essential to discuss these considerations with the family of the patient. Upon interacting with the infant's parents, it was discovered that her mother had consumed tobacco throughout her pregnancy. This could have been a contributing factor to the infant's current condition. The guardian was mother to three children two sons and a daughter, and this condition did not prevail` in either of the two sons. Following the repair of an exstrophied bladder the relatively shorter female urethra can elevate the likelihood of developing urinary tract infection. The newborns urethra was accustomed to urine excretion subsequently causing gross hydronephrosis along with severe cystitis which led to bursting of stitches. This bursting necessitated resuturing of the wound.

CONCLUSION

Bladder exstrophy in a female child is a rare, challenging congenital condition that requires a multidisciplinary approach for diagnosis, management, and long-term care. Early diagnosis opens a door to timely and tailored treatment plan. Long-term management of bladder exstrophy involves ongoing vigilance for urinary and reproductive health. Equipped with multidisciplinary team of healthcare providers, a strong support network, and a determined spirit help the child achieve the best possible quality of life and functional outcomes.

ACKNOWLEDGEMENT

The authors express their heartfelt gratitude to Dr. Neha Suratiya, Assistant Professor and all the members of the Department of Pharmacy Practice for their invaluable guidance and unwavering support.

CONFLICT OF INTEREST

The authors declare that there is no conflict of interest.

ABBREVIATIONS

BE: Bladder exstrophy; **ICU:** Intensive care unit; **UTI:** Urinary Tract Infection; **ARF:** Acute Renal Failure; **INJ.:** Injection; **CRP:** C-reactive protein; **Neb.:** Nebuliser.

REFERENCES

1. Anand S, Lotfollahzadeh S. Bladder exstrophy. StatPearls. 2023.
2. Stec AA. Embryology and bony and pelvic floor anatomy in the bladder exstrophy-epispadias complex. Semin Pediatr Surg. 2011; 20(2): 66-70. doi: 10.1053/j.sempedsurg.2010.12.011, PMID 21453848.
3. Pierre K, Borer J, Phelps A, Chow JS. Bladder exstrophy: current management and postoperative imaging. Pediatr Radiol. 2014; 44(7): 768-86; quiz 765. doi: 10.1007/s00247-014-2892-5, PMID 24939762.
4. K SK, Mammen A, Varma KK. Pathogenesis of bladder exstrophy: A new hypothesis. J Pediatr Urol. 2015; 11(6): 314-8. doi: 10.1016/j.jpurol.2015.05.030, PMID 26316280.
5. Siffel C, Correa A, Amar E, Bakker MK, Bermejo-Sánchez E, Bianca S, *et al.* Bladder exstrophy: an epidemiologic study from the International Clearinghouse for Birth Defects Surveillance and Research, and an overview of the literature. Am J Med Genet C Semin Med Genet. 2011; 157C(4): 321-32. doi: 10.1002/ajmg.c.30316, PMID 22002949.
6. Grady RW, Carr MC, Mitchell ME. Complete primary closure of bladder exstrophy: epispadias and bladder exstrophy repair. Urol Clin North Am. 1999; 26(1): 95-109. doi: 10.1016/s0094-0143(99)80009-3, PMID 10086053.
7. Baird AD, Nelson CP, Gearhart JP. Modern staged repair of bladder exstrophy: a contemporary series. J Pediatr Urol. 2007; 3(4): 311-5. doi: 10.1016/j.jpurol.2006.09.009, PMID 18947762.

Cite this article: Prajapati A, Shah A, Shah C, Gandhi R, Suratiya N. A Rare Scenario: A Female Child with Bladder Exstrophy and Complication. J Young Pharm. 2024;16(1):134-6.