Case Report

# Bladder Exstrophy with Exstrophic Rectal Duplication in an Infant: An Extremely Rare Case

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## Abstract

Exstrophic rectal duplication and its association with bladder exstrophy and anorectal malformation is an extremely rare clinical entity. This is a report of the second case of an exstrophic rectal duplication associated with bladder exstrophy in English literature. However, it is the first case, where all these anomalies were accompanied by an anorectal malformation.

**Keywords:** Anorectal malformation, bladder exstrophy, infant, rectal duplication

## Introduction

Bladder exstrophy is a complex spectrum of congenital anomalies characterised by abnormalities of the bladder, bony pelvis, pelvic floor and genitalia.[1] Unlike cloacal exstrophy, classic bladder exstrophy is not typically associated with gastrointestinal malformations.[2] This is a report of an extremely rare variant of bladder exstrophy associated with exstrophic rectal duplication and anorectal malformation.

## Case Report

A 4-month-old male infant was referred for the evaluation of his exstrophy vesica. He had not been surgically treated so far because his family had just immigrated from Somalia. On physical examination, a perineal mucosa-lined lesion looking like a prolapsed rectum and anorectal malformation with perineal fistula were detected in addition to an exstrophic bladder and widening of the pubic symphysis, bifid phallus and scrotum [Figure 1a and b]. The patient was defecating via the perineal fistula and nothing has been done to the anorectal malformation and bladder exstrophy until now. Echocardiographic evaluation of the patient revealed a 4-mm secundum atrial septal defect. However, no additional anomaly was detected radiologically except severe pubic symphysis diastasis of 5 cm. A staged operation was planned. The exstrophic bladder was closed primarily without pelvic osteotomy. There were two exstrophic intestinal tissues in front of the rectum. Although they were in proximity with the native rectum at the midline, they did not communicate with it. They were excised without rectal injury, perineal reconstruction was performed and then a sigmoid colostomy was fashioned [Figure 1c]. Histologically, only colonic mucosa was present in the excised exstrophic intestinal tissues, and these were diagnosed as exstrophic duplications of the rectum. After an uneventful post-operative recovery, the patient was discharged and planned for future anoplasty, pelvic osteotomy with bladder neck and penile and urethral reconstruction.

## Discussion

Alimentary tract duplications are rare congenital anomalies, which can occur anywhere from the oropharynx to the anus. Rectal duplications account for only 5% of all duplications. They are more commonly located in the presacral space behind the rectum as a cystic mass and do not communicate with the rectum in 90% of cases.[3] However, the patient in the present case had an exstrophic anterior rectal duplication. While anterior localisation is rarely seen, it is also far more unlikely

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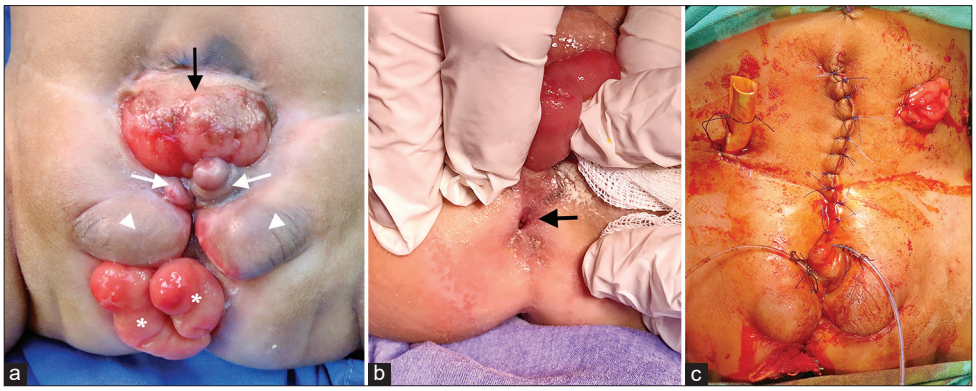


**Figure 1:** Photographs of the patient. (a) The appearance of bladder exstrophy (black arrow), bifid phallus (white arrows) and scrotum (white arrowheads), and two exstrophic rectal duplications (asterisk symbols). (b) The appearance of anorectal malformation with perineal fistula (black arrow). (c) Post-operative photograph

for these to be exstrophied.[4,5] The proposed embryologic mechanism for exstrophic rectal duplication is that the rectal duplication cyst becomes sufficiently large *in utero* to prevent mesodermal migration or increase apoptosis of mesenchyme in the surrounding area, causing weakening of the overlying surface ectoderm, resulting in rupture of the cyst onto the perineum.[5]

Rectal duplications may be associated with other anomalies of the urinary tract and the genitalia.[3] However, exstrophic rectal duplication and its association with bladder exstrophy are an extremely rare entity. Gupta *et al*.[6] have reported only case of exstrophic rectal duplication associated with classical bladder exstrophy. In this respect, this is the second reported case of this association, but this case is unique in two ways: first, two separate exstrophic rectal duplications were observed in the patient and second, all these anomalies were accompanied by a mild form of anorectal malformation.

## Conclusion

An extremely rare variant of bladder exstrophy associated with exstrophic rectal duplication has been presented. This case may be diagnosed as an atypical form of cloacal exstrophy by some clinicians due to its association with an anorectal malformation and exstrophic rectal duplication. However, it is more prudent to leave this case as bladder exstrophy with exstrophic rectal duplication.

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## Declaration of patient consent

The authors certify that they have obtained all appropriate patient consent forms. In the form the patient(s) has/have given his/her/their consent for his/her/their images and other clinical information to be reported in the journal. The patients understand that their names and initials will not be published and due efforts will be made to conceal their identity, but anonymity cannot be guaranteed.

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## Conflicts of interest

There are no conflicts of interest.

## References

1. Ludwig M, Ching B, Reutter H, Boyadjiev SA. Bladder exstrophy-epispadias complex. Birth Defects Res A Clin Mol Teratol 2009;85:509-22.
2. Arlen AM, Smith EA. Disorders of the bladder and cloacal anomaly. Clin Perinatol 2014;41:695-707.
3. Lund DP. Alimentary tract duplication. In: Coran AG, Adzick NS, Krummel TN, Laberge JM, Shamberger RC, Caldamone AA, editors. Pediatric Surgery. 7th ed.. Philadelphia: Elsevier Saunders; 2012. p. 1155-63.
4. Amjadi K, Poenaru D, Soboleski D, Hurlbut D, Kamal I. Anterior rectal duplication: A diagnostic challenge. J Pediatr Surg 2000;35:613-4.
5. Sun J, Vongphet S, Zhang Z, Mo J. Perineal mass protrusion with rectal mucosa: A rectal duplication that underwent exstrophy. J Pediatr Surg 2011;46:e5-8.
6. Gupta RK, Oak S, Parelkar SV, Sanghvi B, Kaltari DK, Prakash A, *et al.* Ruptured rectal duplication cyst with classical bladder exstrophy. J Pediatr Surg 2010;45:1538-41.