

A Triplicated Bladder with Diphallia; Rare Variant of Duplicate Exstrophy

Hana Abebe Gebreselassie, Binyam Gebremedihin Godu , Hiwote Girma Assefa, Maru Gama Erge 

St. Paul's Hospital Millennium Medical College, Department of Surgery, Pediatrics Surgery Unit, Addis Ababa, Ethiopia

Correspondence: Hana Abebe Gebreselassie, St. Paul's Hospital Millennium Medical College, Department of Surgery, Pediatrics Surgery Unit, Addis Ababa, Ethiopia, Tel +251 913 766 868, Email hanaabebe23@gmail.com

Background: Exstrophy variants are well described urologic anomalies. They are characterized by atypical anatomical and physical findings than those found in patients with classic bladder exstrophy and epispadias malformation. The combination of these anomalies with duplicated phallus is a rare occurrence. Here we present a neonate with a rare form of exstrophy variant associated with penile duplication.

Case Summary: One day old male neonate who was born at term was admitted to our neonatal intensive care unit. He had lower abdominal wall defect and open bladder plate with no visible ureteric orifices. There were two completely separate phalluses with penopubic epispadias and urethral orifices draining urine. Both testes were descended. Abdominopelvic ultrasound showed normal upper urinary tract. He was prepared and operated with intra operative finding of complete bladder duplication in the sagittal plane and each bladder has its own ureter. The open bladder plate which had no connection with both ureters and urethras was excised. The pubic symphysis was approximated without osteotomy and abdominal wall was closed. He was immobilized with mummy wrap. He had uneventful post-operative course and was discharged on the 7th post-operative day. He was evaluated on the 3rd month post operatively and he was thriving well with no complications.

Conclusion: The occurrence of a triplicated bladder along with diphallia is an exceptionally rare urologic anomaly. As a number of variations are possible in this spectrum, the management of neonates with this anomaly should be individualized.

Keywords: bladder duplication, exstrophy variant, diphallia, duplicate exstrophy

Introduction

Exstrophy variants are rare subsets of an infrequent urologic malformation which is bladder Exstrophy Epispadias complex. Classic bladder Exstrophy has an incidence of 1 in 40,000 to 50,000 live births and exstrophy variants occur 10 times less frequently than this figure making their incidence 2 per one million live births.^{1,2}

Although a spectrum of disorders exists in these variants, they share the musculoskeletal defect of wide pubic diastasis and divergent rectus abdominis muscles. According to Marshal and et al the well-known exstrophy variants include superior vesical fissure, pseudo-exstrophy, duplicate exstrophy and superior vesical fistula.³ Other variants like covered exstrophy and inferior vesical fissure have been described then after showing the complexity of these malformations.⁴

The occurrence of this rare anomaly in combination with another unusual urologic malformation namely diphallia is an exceptional phenomenon. To our knowledge, this is the first report of such combination of complex urologic malformations.

Case Summary

One-day old male neonate who was born at term with a birth weight of 3000 grams was admitted to our neonatal intensive care unit. Upon examination, he was stable and active. He has no dysmorphic features. Chest and cardiac examination was unremarkable. On abdominal examination, he had infra umbilical midline abdominal wall defect with visible open bladder plate with no visible ureteric orifices. Pubic diastasis was 4 centimeters. There were two completely separate phalluses with epispadias and patent urethral orifices draining urine. Both testes were descended. No other

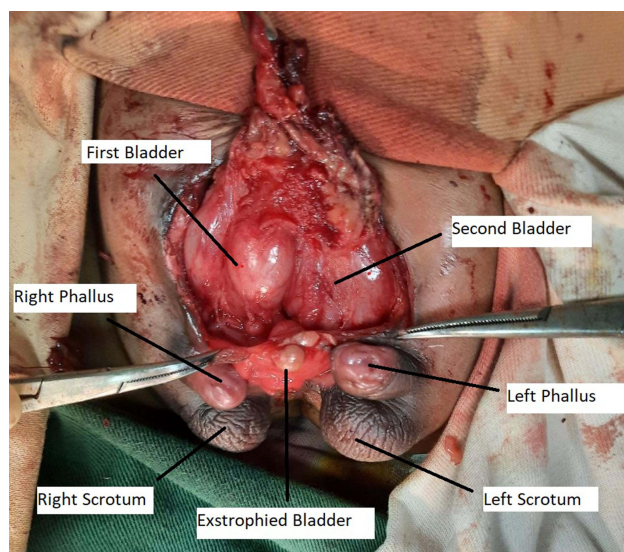


Figure 1 Intraoperative picture revealing a triplicated bladder with diphallia.

skeletal deformity was noted (Figure 1). The anus was normally located and patent. The sacrum was also well noted upon palpation.

Abdominopelvic ultrasound showed normal upper urinary tract. Echocardiography and other blood work ups were unremarkable. After completing investigations, the neonate was taken to the operation theater and explored. As the open bladder plate was being mobilized, it was noted that it had no connection with the ureters and also the urethras. Hence, it was excised. After excising the dry bladder plate, it was found that there was a complete bladder duplication on the sagittal plane. Each bladder was well formed and drain the ipsilateral kidney and was connected to the ipsilateral urethra. The presence of prostatic tissue and the number of corporal bodies in each phallus were not appreciated intra operatively since these structures were located deep in the pelvis. With these findings, no further urologic reconstruction was done.

The pubic symphysis was approximated with a suture without osteotomy. Abdominal wall was closed with mild tension after mobilizing the fascia on both sides. The neonate was immobilized with mummy wrap. He had uneventful post-operative course and was discharged on the 7th post-operative day. Biopsy confirmed that the excised tissue is a bladder. Subsequently, the patient was evaluated 3 months after surgery and he was noted to have smooth post-operative course with no complications.

Discussion

Bladder exstrophy is believed to occur earlier in gestation before the end of the 8th week of gestation as a result of inadequate migration of mesodermal cells in the caudal part of the abdomen. In addition, the timing of rupture of the cloacal membrane is believed to determine the extent of the lower abdominal defect and also the type of the anomaly in this spectrum of malformations. If rupture of the cloacal membrane happens prior to its fusion with the urorectal septum, cloacal exstrophy happens whereas if its rupture occurs after its fusion with this septum, classic bladder exstrophy results.⁵

As to the embryogenesis of exstrophy variants, due to the variability of the defects and the concomitant anomalies whose embryological origin cannot adequately be explained by the mechanisms mentioned above, further discovery is needed to fully understand the embryologic errors that result in the formation of these complex malformations.⁶

Exstrophy variants are characterized by atypical anatomical and physical findings than those found in patients with classic bladder exstrophy and epispadias malformation despite sharing the typical musculoskeletal defects of pubic separation and rectus diastasis. Marshal and his colloquies classified exstrophy variants into the following categories: superior vesical fissure, pseudo-exstrophy, duplicate exstrophy, superior vesical fistula.³ After their publication, other

variants including covered exstrophy and inferior vesical fissure have been described revealing the complex nature of these malformations.⁴

Fewer than 30 cases of duplicate exstrophy have been reported so far.⁷ Benjamin and et al reported their experience on 25 patients with exstrophy variants at their center. They described two types of duplicate exstrophy: anterior-posterior and side by side duplication. Our patient had side by side duplication of the bladder.⁶ We did not find any report describing duplication of the bladder in the sagittal plane along with a third bladder with features of exstrophy making our case unique.

Bladder duplication and diphallia are also rare occurrences by themselves. Approximately 70 cases of bladder duplication have been reported till date while the reported incidence of diphallia is 1 per 5 to 6 million live births.^{8–12} Even though the association of bladder duplication with diphallia has been described in few case reports so far, the occurrence of both of these anomalies in concurrence with a third bladder with features of exstrophy makes our case exceptional.^{13–15}

In our patient, since each duplicated bladder had its own ureters and urethra and the exstrophied bladder was dry, simple excision of the bladder plate and abdominal wall closure was done. This approach is favored as per the recommendations of Benjamin and et al.⁶ The surgical management of the diphallia was left to be done at a later age.

Regarding the future management plan of this infant, we do not expect major concerns on urinary continence as the bladder neck is located deep in the pelvis. In addition, it was noted that the neonate urinates in intervals and stays dry in between. Our plan is to follow his urinary voiding pattern and to send for Magnetic Resonance Urography to have a better characterization of the lower urinary tract and the genitalia. The specific surgical approach for the diphallia will be decided after doing imaging and characterizing the number and level of development of the corpora in each phallus. Either excision or reconstruction of the corporal tissue with or without urethral reconstruction will be done based on imaging findings.

In summary, our case presents a unique constellation of rare urologic malformations namely triplicated bladders with diphallia. This case is another good example showing the complex nature of exstrophy variants in the spectrum of bladder exstrophy and epispadias malformations.

Consent for Publication

Consent for publication was taken from parents of the neonate and can be accessed from the corresponding author upon request. Institutional approval was not required to publish the case details as no personal identifiers were included and there was no other issue requiring ethical clearance.

Disclosure

The authors report no conflicts of interest in this work.

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