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# Bladder triplication associated with exstrophy variant

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ABSTRACT

A one-day old full-term female neonate presented with a duplicate bladder and exstrophy variant including a patch of exstrophic or ectopic mucosa, duplicate vagina, uterus, and two complete bladders. We report on the surgical management performed in this case and functional urinary results based on a synchronous urodynamic study of the duplicate bladders. To our knowledge, the occurrence of duplicate bladder exstrophy variant with complete urinary continence has not been previously reported.

## 1. Introduction

The exstrophy-epispadias complex comprises significant midline defects including anomalies in the musculoskeletal, gastrointestinal, and genitourinary system. Altogether, approximately 30 cases of bladder triplication have been previously described in the literature.

## 2. Case presentation

A one-day-old female neonate (3.5 kg) was referred to our institution for abnormal appearance of the umbilicus and external genitalia. The pregnancy did not have any complications. There was also no history of maternal disease or medication use during the pregnancy. In addition, although no abnormalities were detected on prenatal ultrasounds, the sex of the fetus could not be determined.

The neonate was delivered at 41 weeks of gestation without any complications. Physical examination at the remote primary care facility revealed a patch of exstrophic mucosa on the infra-umbilical region with a low-set umbilicus. By its appearance, it initially led to the diagnosis of classic bladder exstrophy. Routine laboratory studies were normal and chromosomal analysis indicated a normal female karyotype. Renal ultrasonography showed two normal appearing kidneys without hydronephrosis.

Surgical correction of what seemed like the exstrophic patch was done on day 4 post-delivery. Identification and catheterization of the ureteral orifices was impossible to accomplish. The decision was made to continue dissection as planned even though the orifices weren't found, and that they would be identified later on. After a careful intra-operative examination and dissection (Fig. 1), two complete bladders in the sagittal axis, ureters, urethral orifices, and vaginal openings, as well as a bicornuate uterus and a bifid clitoris were found. In addition, the bladder patch was removed during the surgery since it was nonfunctional. There were two complete and separate bladders. Because of this rare variant, there was hesitation as to reconstruct the two bladders as one because of the possibility that the detrusor function would be irreversibly affected. The decision was made to leave the two underlying bladders divided. The option to operate again later could still be a possibility if ever the two bladders didn't work in a synchronized fashion. Two indwelling urethral catheters were passed into each one of the urethras/bladders and the hemi-clitoris and external genitalia were corrected. The abdominal defect was closed at the midline without tension and without need for osteotomy. Symphyseal diastasis was closed. Lower limb traction was maintained in the zenith position for 19 days after surgery to prevent wound dehiscence. The postoperative period was uneventful and the patient was discharged on day 23, after removal of all catheters. Postoperative renal ultrasonography showed two symmetrical bladders and no hydronephrosis. Signs of bladder cycling were already present, and the patient didn't suffer from total incontinence.

At 17 months of age, two different urodynamic systems were used to perform simultaneous right and left bladder urodynamic testing in the OR. The bladder capacity was similar in both bladders (84 mL each),

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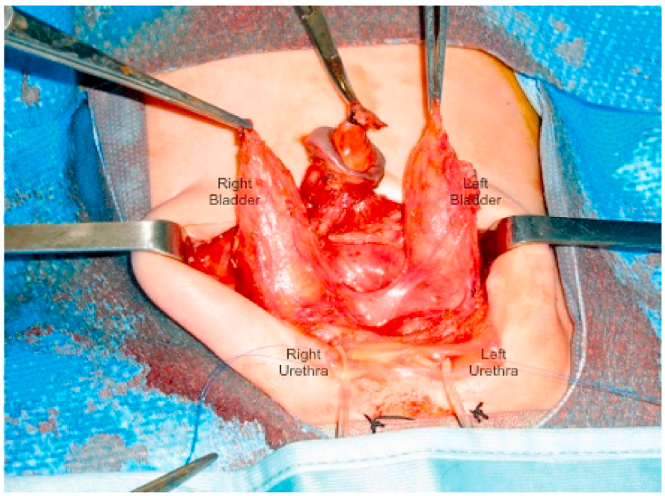


**Fig. 1.** Intraoperative examination revealed two complete bladders, ureters, and urethral orifices in the sagittal axis.

which corresponds to the patient's age. A normal end filling pressure for each bladder was noted: 14 cm $H_2O$ (right) and 22 cm $H_2O$ (left). Bladder contraction pressure were 83 cm $H_2O$ (right) and 82 cm $H_2O$ (left). In addition, no uninhibited detrusor contractions were observed. Postvoid residual urine volume was 6 mL (right) and 71 mL (left). Interestingly, both bladders were functionally synchronized, meaning they would empty at the same time (Figs. 2 and 3). Up until now, ten years later, this patient is still continent and has a normal upper tract on ultrasound.

## 3. Discussion

Exstrophy variants have an incidence of 1 in 400, 000 to 500,000 live births, and seem to be associated with musculoskeletal defects, pubis diastasis, and divergent recti. These variants are classified according to their type of presentation as follows: pseudoexstrophy, superior vesical fissure, superior vesical fistula, duplicate bladder exstrophy, and covered exstrophy. However, these classifications tend to be confused among researchers, because many of these variants do not necessarily conform to a specific type, but rather present on a spectrum.[1]

This bladder triplication variant consists of a non-functional suprapubic exposed patch of bladder mucosa and two underlying intact

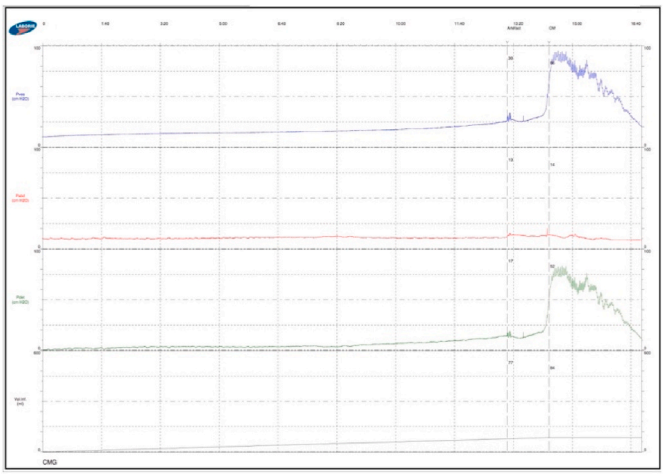


**Fig. 2.** Urodynamic study of the right bladder: bladder capacity 84 mL; normal compliance 14 cm $H_2O$; no uninhibited detrusor contractions; and postvoid residual urine volume was 6 mL.

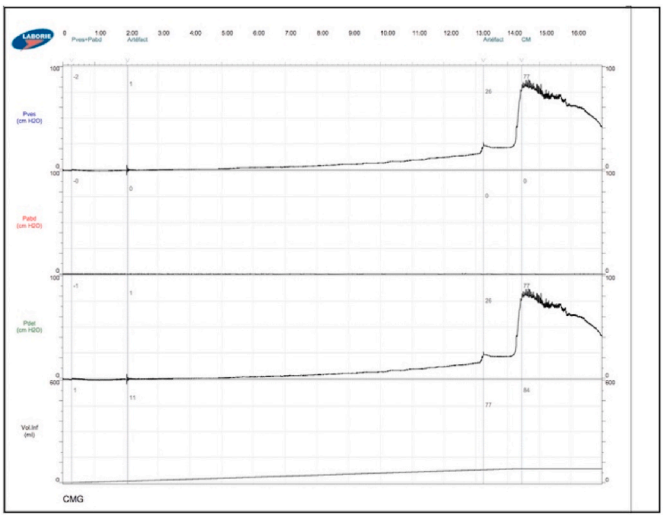


**Fig. 3.** Urodynamic study of the left bladder: bladder capacity 84 mL; normal compliance, 22 cm $H_2O$; no uninhibited detrusor contractions; and postvoid residual urine volume was 71 mL.

bladders, without communication in between any of these structures.[2] Furthermore, as it is a complete duplication, it includes separate urethras originating from each bladder.[3] The etiology of complete duplication of the bladder remains unknown however.

There are no specific signs to diagnose these malformations early, except when external genitalia anomalies are present. In general, careful examination with fluoroscopy and cystoscopy can help to define the exact anatomy and associated malformations. Failure to identify the two ureteral orifices on the exstrophic or ectopic bladder patch is a clue indicating the presence of underlying duplicate bladder. The surgical procedure should be simple in these cases. In our case, the bladder patch was removed, the two bladders left intact, and abdominal wall defect closure performed.

Also, the fact that the two bladders were functioning in a synchronized fashion is a surprising finding, and reminds us that sometimes, less is more. In doubt, it might be beneficial to do only what is needed at the time, and to operate again later if required. The findings of our case, which consisted of an exposed exstrophic or ectopic bladder patch with two complete bladders, diastasis pubis with internal and external genital duplication, makes it unique. Since there are many variants, the management of these pathologies must be individualized.

## 4. Conclusion

The wide variability in the anatomy and presentation of these cases influences its diagnosis and treatment. The management of each case must be individualized with the goal to preserve continence without lower urinary tract obstruction or infection.

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## Author contribution

Monica Salgado-Diaz: Writing - original draft.
Valerie Hogues: Writing - review & editing.
Anne-Marie Houle: Writing - review & editing.
Julie Franc-Guimond: Writing - review & editing.
Diego Barrieras: Writing – review & editing.

## Declaration of competing interest

None.

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