

## True Duplicate Bladder Extrophy: A Case Report and Review of the Literature

Rossella Angotti\*, Doski Kader, Giulia Giannotti, Maria Pavone, Giovanni Di Maggio, Francesco Ferrara and Mario Messina

Division of Paediatric Surgery, Dept of Paediatrics, Obstetrics and Reproductive Medicine, University of Siena, Policlinico "Le Scotte" Viale Bracci 16, 53100 Siena, Italy

### Introduction

Bladder exstrophy is a rare malformation occurring predominantly in males, with an incidence of 1:50,000 live births. Variants of bladder exstrophy including "split symphysis variants", are very rare malformations with an incidence of 1 in 400,000-500,000 live births, making up 8% of all exstrophy patients [1, 2]. We describe a child with duplicate exstrophy, that is one of the rarest variants, and add a further case to the scarce published literature.

### Case Report

A 2-year old female, born by normal vaginal delivery at 37 weeks of gestation with a birth weight of 3180 g, was admitted to our clinic for assessment of a congenital infra umbilical midline lesion. Antenatal history and family history were negative, there were no other congenital anomalies, and the child's postnatal development had met normal milestones. Examination revealed a low-set umbilicus, below which was a soft, dry, pigmented 5 x 6 cm mucosa lined plaque. There was no urinary discharge and no ureteric orifices could be identified (Figure 1). The external genitalia were normal female and the child was dry, passing urine with an intermittent forceful stream. There was a minimal diastasis at the pubic symphysis that measured 1.5 cm on X-ray. All routine laboratory studies including chromosomal analysis, urinalysis and urine culture were normal. Renal anomalies were excluded on ultrasound scan, and a micturating cystourethrogram showed a normal bladder, bladder neck and urethra, without UV reflux or urinary fistula. Urodynamic studies showed a bladder capacity of 100 cc that was appropriate for her age. At cystourethroscopy the urethra and bladder neck were normal leading up to a normal trigone with bilateral competent single ureteric orifices. The rest of the vulva and the vagina were normal. Surgical reconstruction consisted of a total excision of the mucosal lesion, with primary closure of the abdominal defect allowing the umbilicus to rise to a normal midline position. There was no communication between the abdominal lesion and the urinary tract or other viscera. Histology identified the mucosal plaque as 'urothelium without atypia'. The child was discharged on the 7th postoperative day with a well healing wound. At 1 year follow-up the scar had stretched and was pigmented reducing the overall aesthetic appearance, but was otherwise satisfactory.



Figure 1: Preoperative appearance duplicate bladder extrophy.

### Discussion

Duplicate exstrophy is one of the rarest variants within the spectrum of bladder exstrophy. It is characterized by the presence of a non-functional exstrophic mucosal plate on the abdominal wall, with a normal bladder and no exposed ureteric orifices. The aetiology of this malformation is unknown, and the embryologic theories are many [3]. Sheldon et al. [4] divided the duplicate exstrophy into those with a true duplication with the classic findings of the exstrophy complex, and another group, to which our case conforms, characterized by a suprapubic exstrophic mucosal plaque with no communication to a covered normal functional bladder. These children usually have well formed external genitalia (as defined by Marshall and Muecke) [5] and are continent of urine with a normal voiding pattern [1, 6-7]. Surgical reconstruction for this latter group is relatively straightforward consisting of excision of the exstrophic plaque with primary aesthetic closure of the abdominal wall defect. In our case immediate healing was uncomplicated with good positioning of the umbilicus. However at 1yr follow-up the scar had stretched and was pigmented, detracting from the overall aesthetic appearance. Long term follow-up with and possible scar revision, are relevant to avoid psychological concerns in adolescence and young adulthood.

### Conclusions

A literature review confirmed the rarity of the disease [3, 7, 8] revealing only 24 reported cases of duplicate bladder exstrophy, most of which are isolated case reports. This paper adds a further case to the scarce literature, emphasizing the need for a full evaluation to exclude any connection of the exstrophic plaque to the urinary tract, and the need for a long-term follow-up with possible scar revision in adolescence or young adulthood.

### References

1. Bouhafs A, El Azzouzi D, Halim Y, Arifi M, Belkacem R, et al. (2006) Duplicate bladder exstrophy: a unique variant. *J Pediatr Urol* 2: 483-485.
2. Corroppo M, Zampieri N, Pietrobelli A, Giacomello L, Camoglio FS (2007) Bladder exstrophy variants. *Minerva Urol Nefrol* 59: 109-113.
3. Andiran F, Tanyel FC (1999) Duplicate exstrophy of the bladder. *J Pediatr Surg* 34: 626-627.

\*Corresponding author: Dr. Rossella Angotti, Division of Paediatric Surgery, Dept of Paediatrics, Obstetrics and Reproductive Medicine, University of Siena, Policlinico "Le Scotte" Viale Bracci 16, 53100 Siena, Italy, Telephone: +39 0577/586502; Fax: +39 0577/586174; E-mail: [rossellaangotti@libero.it](mailto:rossellaangotti@libero.it)

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4. Sheldon CA, McLorie GA, Khoury A, Churchill BM (1990) Duplicate Bladder Exstrophy: a new variant clinical and embryological significance. J Urol 144: 334-336.
5. Marshall VF, Muecke EC (1962) Variations in exstrophy of the bladder. J Urol 88: 766-796.
6. Cerrah Celayir A, Celayir S, Oner N, Vural G, Danişmend N (2000) Duplicated exstrophy in one of identical twins. Urology 56: 330.
7. Gupta DK, Charles AR, Srinivas M (2003) Exstrophy variants: should they be considered malformation complexes separate from classical exstrophy? Eur J Pediatr Surg 13: 377-382.
8. Kumar Basant, Sharma Chetan, D Shinha Devendra (2009) True Duplicate Bladder exstrophy: a rare lesion. Indian J pediatr 76: 852-853.

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