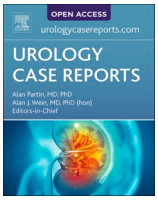
Urology Case Reports 54 (2024) 102736

Contents lists available at ScienceDirect

Urology Case Reports

journal homepage: www.elsevier.com/locate/eucr



Pediatrics

Bladder duplication in a patient with a persistent urogenital sinus: Case report and systematic review of the literature☆

Carla Ramirez-Amoros *, Karla Estefania-Fernandez, Lucas Moratilla-Lapeña, Alejandra Vilanova-Sanchez, Pedro Lopez Pereira, Maria Jose Martinez Urrutia

La Paz Children's University Hospital, Department of Paediatric Surgery, Madrid, Spain

ARTICLE INFO

Keywords:
Bladder duplication
Persistent urogenital sinus
Anorectal malformation
Case report

ABSTRACT

Bladder duplication (BD) is a rare malformation that is often associated to other anomalies. We report a newborn diagnosed with BD in the sagittal plane, associated to persistent urogenital sinus (UGS), given the opening of the vagina immediately below the bladder neck. It is the fourth time this association is reported. Surgical repair was made: both bladders were joined, the common channel was left as urethra and the vagina was descended with a vaginoplasty with an intestinal segment. She also presented an anterior anus, that required posterior mobilization. The patient is currently 3 years old with good sphincter control.

1. Introduction

BD is a rare congenital malformation, with less than 80 reported cases.[1] It is often associated to other congenital anomalies, with an incidence as high as 85 %.[2] Amongst the most common associations we find malformations affecting the gastrointestinal tract,[1,3] genitalia[3–5] or the spine.[6] The association with persistent UGS has only been described in three cases.[3,7] Clinical presentation is variable, ranging from asymptomatic patients in which the finding is incidental to those with vesicoureteral reflux (VUR), recurrent urinary tract infections (UTI) or even renal failure.[7,8] We report a case of BD with UGS who underwent successful surgical correction.

2. Case report

A female patient, diagnosed with megacystic bladder and bilateral hydronephrosis at 16 weeks' gestation and born at 40 + 2 weeks' gestation via euthocic vaginal delivery weighting 2985 g. Clinical examination showed external genitalia consisting of a small vulva with a pubic mound cleft, slight pubic diastasis and an anterior anus. After a neonatal urinary tract infection, ultrasound examination confirmed a complex urogenital malformation with right pelvicalyceal dilatation and left kidney atrophy. Although it was difficult to delineate the complex anatomy, initial cystoscopy revealed what seemed to be a common urogenital sinus with three orifices; towards two hemi-bladders and a vaginal cavity. A suprapubic catheter was placed.

The anterior anus was confirmed with electrostimulation of the muscle complex. Cisto-genito-CT and enema-CT imaging supported the presence of an almost complete bladder duplicity to which the vagina joined 3 mm below the bladder neck to form the common sinus, which measured 44 mm. The patient had a normal uterus and ovaries. Given the future repair and perineal approach, which would include mobilisation of the anus, a laparoscopic colostomy was performed at 11 months of age.

At 18 months the patient's complex urogenital malformation was reconstructed. A cystoscopy and later Phannestiel incision revealed that the anterior-left cavity in which the suprapubic catheter was located corresponded to an hemi-bladder without a clear ureteral meatus or trigone, separated from the other hemi-bladder by an incomplete oblique sagittal septum (Fig. 1A).

The ureteral orifice of the only functioning kidney was in the area of the septum between the two bladders. Posterior to the hemi-bladders, there was a vaginal cavity, with a connection in the area of the bladder neck. The oblique septum between the hemi-bladders was sectioned to create a single cavity (Fig. 1B) and the right ureter reimplanted on the right side of the new bladder cavity (Fig. 1C). The anatomopathological findings were consistent with bladder duplication, showing normal bladder mucosa and detrusor in the resected septum.

☆ The authors declare that they have no known competing financial interests or personal relationships that could have appeared to influence the work reported in this paper.

* Corresponding author. Paseo de la Castellana, 261, 28046, Madrid, Spain.
E-mail address: carlaramirezamoros@gmail.com (C. Ramirez-Amoros).

https://doi.org/10.1016/j.eucr.2024.102736
Received 12 March 2024; Received in revised form 4 April 2024; Accepted 9 April 2024
Available online 10 April 2024

The vaginal cavity was dissected until its complete separation from the posterior aspect of the bladder and the common channel was left as the urethra (Fig. 1D). Likewise, the rectum was released from the posterior aspect of the common channel and a posterior sagittal anorectoplasty was performed (Fig. 1E). For the vagina to reach the perineum, it had to be prolonged by performing a vaginoplasty with the distal colostomy stump (Fig. 1F). The postoperative period was uneventful, with no reported complications. Fig. 2 shows a schematic drawing of the anatomy before and after reconstruction.

The patient proactively underwent dilation of the anoplasty during the first 4 weeks after the surgery, without further need for dilation onwards. At 23 months of age, the colostomy was closed and bowel transit restored. The patient is currently 3 years old with good sphincter control. She has normal voiding volumes every 2 hours with no incontinence and daily bowel movements with senosside laxative treatment and no fecal accidents. She remains asymptomatic with no UTI and no need for continuous antibiotic prophylaxis. Ultrasound follow-up shows a normal right kidney without urinary tract dilation and control voiding cystourethrography shows no VUR. The substitution vaginoplasty appears patent on physical examination, with no need for dilations or other invasive procedures.

3. Discussion

We report the fourth case of BD associated to persistent UGS that was successfully surgically reconstructed.

BD is a rare anomaly with a slight male predominance described in the literature.[1,5] It can be classified according to the axis of the septum in sagittal or coronal BD, but also depending on the extent of the division in complete or partial BD. According to Abrahamson,[9] complete duplications are divided by a muscular wall and have two separate urethras, whereas in incomplete duplications both bladders are separated by a septum and drain into a single urethra. In terms of the axis classification, the sagittal septum is more frequent and more easily explained embryologically.[9] The theories behind the embryological development of the sagittal septum are a maldevelopment of the urorectal septum, an abnormal septum doubling the allantois or partial twinning of the caudal end of the embryo.[10–12] On the other hand, the coronal septum could be explained by a supernumerary cloacal septum that indents the epithelial wall of the bladder causing it to split or by excessive constriction between the urogenital and vesicouretheral portions of the ventral cloaca.[8,9] Persistent UGS could be explained by a failed descent of the urogenital septum.[7]

BD can have a wide range of clinical presentations. Some children are diagnosed early in the neonatal period because of their associated anomalies or the presence of UTI, voiding disorders or urinary tract obstruction and VUR. Other children remain asymptomatic for years, until they are studied because of minor genitourinary symptoms, infertility or simply as an incidental finding in an otherwise healthy child.[3] The diagnosis of BD and some of its associated anomalies can be made with retrograde urethrocystography, lower abdominal CT imaging or pelvic MRI.[13] Ultrasound imaging should be considered to screen for

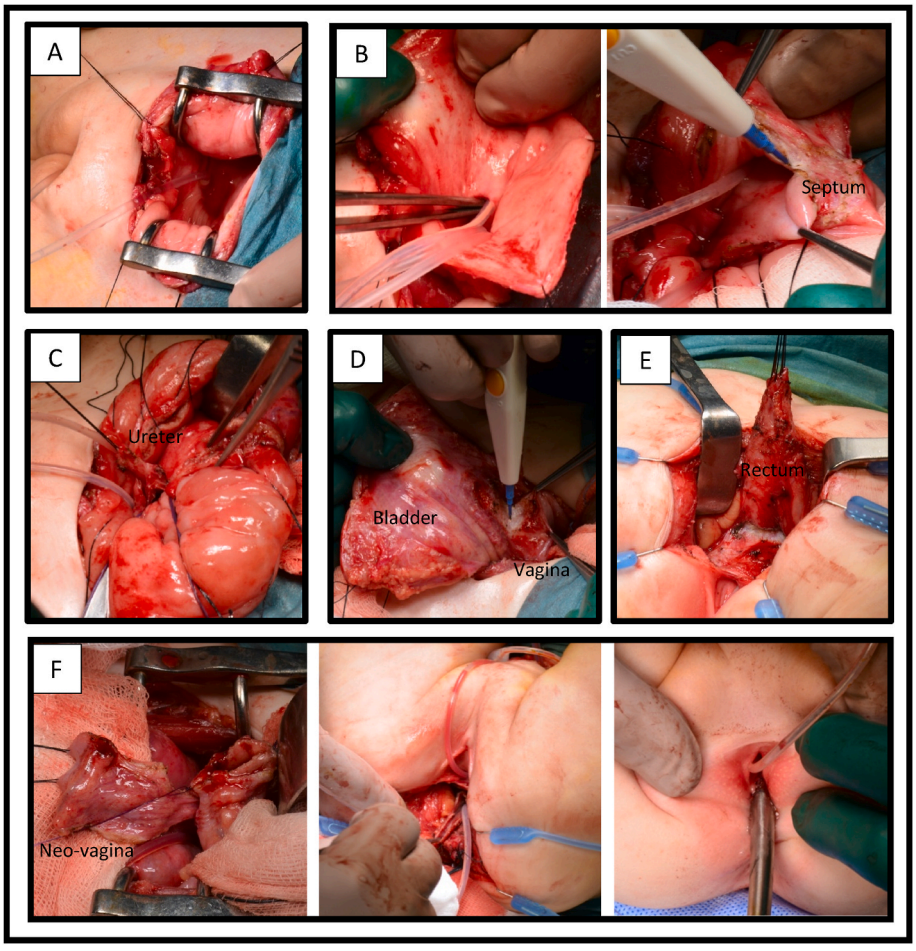


Fig. 1. Surgical reconstruction: A. Anterior cavity with suprapubic catheter. B. Nearly complete sagittal septum separating the hemi-bladders, sectioned to create a single cavity. C. Ureteral orifice of the only functioning kidney in the theoretical area of the septum, reimplanted on the right side of the bladder cavity. D. Dissection and separation of vagina from the posterior aspect of the bladder with the common channel left as urethra. E. Anterior anus. Release of the rectum from the posterior aspect of the common channel and posterior sagittal anorectoplasty. F. Vaginoplasty with the distal colostomy stump.

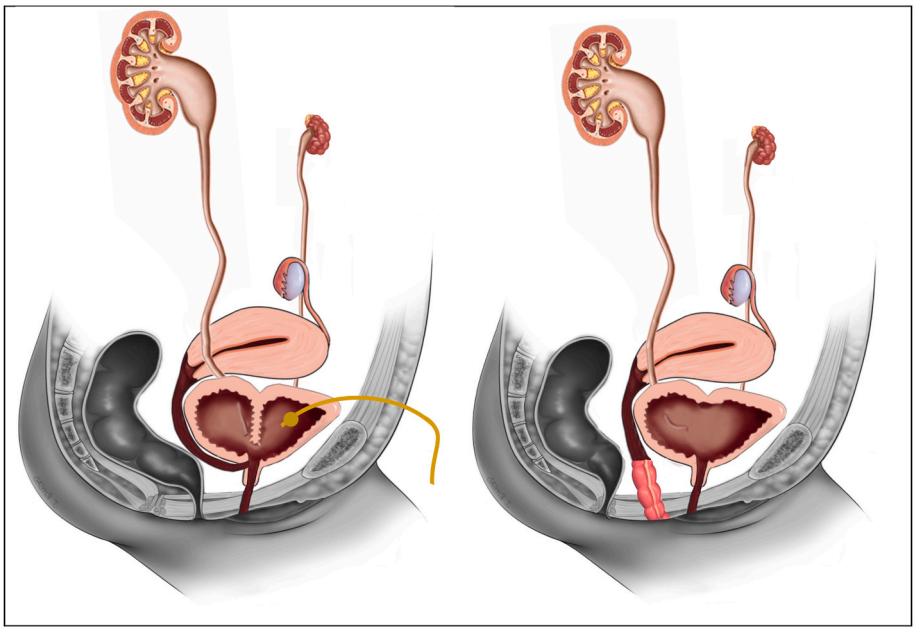


Fig. 2. Before and after reconstruction of bladder malformation with persistent UGS, anterior anus and atrophic kidney.

other upper urinary tract anomalies.[8] However, given the complexity of these malformations, the exact anatomy is often unclear until surgical reconstruction.[7] Lastly, as it was our patients case, antenatal suspicion is also possible[14,15] and can be further confirmed with antenatal MRI.[1]

Although non-urological malformations have more often been linked to sagittal duplications,[1,8] we can also find them in duplications of the coronal plane. The most frequently associated anomalies are caudal gastrointestinal malformations[1,3,6,12,16] (lower gastrointestinal tract duplications, anorectal ectopia, atresia or stenosis), duplication of external and internal genitalia,[3–8] spina bifida or meningocele,[6] pubic symphysis diastasis and vertebral duplications.[3,5] Overall, there seems to be a predominant association to duplication of different systems. For instance, of the 40 patients reviewed by Kossow, 90 % presented duplications of the external genitalia and 42 % duplications of the hind gut.[17] BD has also been described in variants of cloacal exstrophy.[3,18] Anorectal malformations, like the anterior anus our patient had, are described in association to BD in multiple papers.[3,12] As for urinary anomalies, Coker describes 85 % of the reviewed cases of BD to be associated to some genitourinary anomaly.[2] In addition to our patient, there are only 3 other cases reported in the literature in which the BD is associated to persistent UGS[3,7] (Table 1). The first, described by Gastol is a female with cloacal exstrophy with an open bladder and a second closed bladder. She also presented a vaginal duplication with a vagina open to the perineum and the other connected with the urethra of the closed bladder, creating persistent UGS.[3] The other two patients, also female, were described by Shaw and associated two duplicated vaginas and bladders confluent in a single urogenital sinus.[7] All three cases were surgically reconstructed.

Children with BD can remain asymptomatic and be treated conservatively. In cases with two functioning and non-obstructive bladders without a history of UTI or other complications, no treatment is required.[1,3,13] The most common indication for surgery is a symptomatic patient; with recurrent urinary tract infections, urinary incontinence, haematuria, abdominal pain or voiding disorders.[1,13,19] In complete duplications, treatment includes resection of the bladder septum and urethral reconstruction vs. total excision (open or laparoscopically[19]) of the non-functional bladder and urethra.[1,2] For incomplete duplications, both resection of only the septum (open surgery or endourological[20]) or resection of the entire non-functioning duplicated structure have been described.[15] There is no surgical treatment of choice and each case should be individualised to offer the best possible result, prioritising the optimization of the bladder function and drainage, as well as minimising incontinence and risk of infection.[1] Additionally, treatment of associated anomalies can be performed simultaneously or in a different surgical act.

Table 1
Reported cases of BD associated to persistent UGS.

Case 1 (Gastol et.al)[3]: Female, 2 days	
Symptoms: Asymptomatic. Findings: Cloacal exstrophy with BD. Duplicated right kidney and ureters. Vaginal duplication. Persistent UGS. Diagnosis: Physical examination. Intravenous urography and cystogram.	Surgical repair: Staged reconstruction of cloacal exstrophy. Bladders anastomosis. Vaginal anastomosis. UGS as neourethra. Follow-up: Incontinent, pending bladder neck plasty. Cystography revealing a bladder with good capacity and bilateral VUR.
Case 2 (Shaw et al.)[8]: Female, 32 weeks' gestation	
Symptoms: Repeat UTI. Diagnosis: Antenatal ultrasound with left pelvic kidney and right pelvocaliectasis. Post-natal cystoscopy and fluoroscopic examination. Findings: Coronal DB. Sagittal vaginal duplication. Persistent UGS.	Surgical repair: Bladder septum resection. Ureteral reimplantation. UGS as neourethra. Vaginal septum resection and mobilisation to the perineum. Omental flap between neourethra and vagina. Follow-up: Cystoscopy at 6 weeks. Good voiding. Dry intervals.
Case 3 (Shaw et. al)[8]: Female, full gestation	
Symptoms: Perinatal UTI. Diagnosis: Antenatal ultrasound with solitary left kidney and large debris-filled pelvic mass. Post-natal cystoscopy and genitography. Findings: Coronal DB with a dominant bladder. Vaginal duplication with obstructed right vagina. Persistent UGS. Solitary kidney.	Surgical repair: Dominant bladder retained. Young-Dees. Ureteral reimplantation. Vaginal septum resection. UGS as neourethra. Omental flap between neourethra and vagina. Follow-up: Cystourethroscopy at 1 year. Competent bladder neck, with dry intervals.
Case 4 (Ramirez-Amoros et.al): Female, 40 + 2 weeks' gestation.	
Symptoms: Recurrent UTI. Diagnosis: Prenatal ultrasound with megacystic bladder and bilateral hydronephrosis. Postnatal ultrasound, cystoscopy, cisto-genito-CT and enema-CT. Findings: Incomplete sagittal BD. Vagina entering the posterior wall of a bladder. Persistent UGS. Left kidney atrophy. Anterior anus.	Surgical repair: Bladder septum resection. Right ureter reimplantation. UGS left as urethra. Vaginoplasty with an intestinal segment. Posterior mobilisation of the anterior anus. Follow-up: Cystourethrography at 6 weeks and control ultrasounds. Dry intervals between voids and daily bowel movements with laxative treatment.

Our report has certain limitations, as it is a very specific and complex case that is difficult to compare with other patients and whose management cannot be generalised. Nevertheless, given the rarity of these malformations, it is important to report these cases and their management to serve as guides for their management and useful educational information.

4. Conclusion

Bladder duplication is a rare malformation that, to the best of our knowledge, has only been previously described in association with persistent UGS in three other patients. It is a malformation that, when asymptomatic, can be left unreconstructed. However, symptomatic patients should be offered an individualised surgical treatment focusing on optimising bladder function. Given the many associated anomalies, the gastrointestinal and genitourinary systems, as well as the spine of these patients should be studied at diagnosis.

Statements

Informed consent was obtained from the patient's guardians.

All authors attest that they meet the current ICMJE criteria for Authorship.

The authors declare that they have no conflict of interest. We have no funding to disclose.

CRediT authorship contribution statement

Carla Ramirez-Amoros: Writing – original draft, Investigation, Data curation. **Karla Estefania-Fernandez:** Writing – review & editing, Supervision, Conceptualization. **Lucas Moratilla-Lapeña:** Methodology, Investigation, Data curation. **Alejandra Vilanova-Sanchez:** Writing – review & editing, Supervision, Methodology, Conceptualization. **Pedro Lopez Pereira:** Supervision, Funding acquisition, Conceptualization. **Maria Jose Martinez Urrutia:** Writing – review & editing, Supervision, Methodology, Conceptualization.

References

1. Delcont M, Guglielmetti LC, Rajbhandari N, Walker J, Wilcox D, Vuille-dit-Bille RN. Bladder duplication - a case series. *Urology*. 2021;149:199–205. https://doi.org/10.1016/j.urology.2020.11.001.
2. Coker AM, Allshouse MJ, Koyle MA. Complete duplication of bladder and urethra in a sagittal plane in a male infant: case report and literature review. *J Pediatr Urol*. 2008;4(4):255–259. https://doi.org/10.1016/j.jpurol.2008.02.001.
3. Gastoł P, Baka-Jakubiak M, Skobejko-Włodarska L, Szymkiewicz C. Complete duplication of the bladder, urethra, vagina, and uterus in girls. *Urology*. 2000;55(4):578–581. https://doi.org/10.1016/S0090-4295(99)00470-7.
4. Dunn D, Fine RG. Diphallia, double bladder, and two hemiscrotums: a case report. *AORN J*. 2019;109(6):728–740. https://doi.org/10.1002/aorn.12697.
5. Khorramirouz R, Bagheri A, Kajbafzadeh AM. Unusual variant of coronal bladder duplication associated with glans diphallia: a case report and review of the literature. *Case Rep Urol*. 2015;2015:1–4. https://doi.org/10.1155/2015/909102.
6. Azmy AF. Complete duplication of the hindgut and lower urinary tract with diphallus. *J Pediatr Surg*. 1990;25(6):647–649. https://doi.org/10.1016/0022-3468(90)90355-D.
7. Shaw MBK, Cain MP, Rink RC. Two bladders and two vaginas in two planes: one urogenital sinus. *Urology*. 2003;62(1):144. https://doi.org/10.1016/S0090-4295(02)02545-1.
8. Berrocal T, Novak S, Arjonilla A, Gutiérrez J, Prieto C, Urrutia MJ. Complete duplication of bladder and urethra in the coronal plane in a girl: case report and review of the literature. *Pediatr Radiol*. 1999;29(3):171–173. https://doi.org/10.1007/s002470050564.
9. Abrahamson J. Double bladder and related anomalies: clinical and embryological aspects and a case report. *Br J Urol*. 2008;33(2):195–214. https://doi.org/10.1111/j.1464-410X.1961.tb11606.x.
10. Satter EJ, Mossman HW. A case report of a double bladder and double urethra in the female child. *J Urol*. 1958;79(2):274–278. https://doi.org/10.1016/S0022-5347(17)66269-4.
11. Ravitch MM. Hind gut duplication–doubling of colon and genital urinary tracts. *Ann Surg*. 1953;137(5):588–601. https://doi.org/10.1097/00000658-195305000-00002.
12. Veeraraghavan KA, Gonzales ET, Gibbons MD, Wagner ML, Harberg FJ. Cloacal duplication: genitourinary and lower intestinal implications. *J Urol*. 1983;129(2):389–391. https://doi.org/10.1016/S0022-5347(17)52115-1.
13. Pirinççi N, Geçit İ, Güneş M, Tanık S, Ceylan K. Complete duplication of the bladder and urethra in the coronal plane: case report with review of the literature. *Urol Int*. 2013;90(1):118–120. https://doi.org/10.1159/000339922.
14. O'Sullivan GJ, Hunter J, Dubbins PA. Case report: antenatal ultrasonic diagnosis of complete bladder duplication. *Clin Radiol*. 1996;51(11):818–819. https://doi.org/10.1016/S0009-9260(96)80015-8.
15. Evangelidis A, Murphy JP, Gatti JM. Incomplete bladder duplication presenting antenatally. *Urology*. 2004;64(3):589. https://doi.org/10.1016/j.urology.2003.12.011.
16. Esham W, Holt HA. Complete duplication of bladder and urethra: a case report. *J Urol*. 1980;123(5):773–775. https://doi.org/10.1016/S0022-5347(17)56127-3.
17. Kossow JH, Morales PA. Duplication of bladder and urethra and associated anomalies. *Urology*. 1973;1(1):71–73. https://doi.org/10.1016/0090-4295(73)90118-0.
18. Vilanova-Sánchez A, Ching C, Gasior A, Diefenbach K, Wood R, Levitt M. Image of the month: clinical features in a newborn with covered cloacal exstrophy. *Eur J Pediatr Surg Rep*. 2017;5(1):e57–e59. https://doi.org/10.1055/s-0037-1606389.
19. Kord E, Zisman A, Verhovsky G, Neheman A. Laparoscopic excision of complete bladder duplication in a 1-year-old male. *J Pediatr Urol*. 2018;14(4):348–350. https://doi.org/10.1016/j.jpurol.2018.05.012.
20. Parente A, Angulo JM, Romero R, et al. Endourological management in a newborn with incomplete bladder duplication. *J Pediatr Urol*. 2013;9(1):e19–e21. https://doi.org/10.1016/j.jpurol.2011.11.015.