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## Challenging Case of Postmenopausal Bleeding and Complete Urogenital Duplication

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**Conflict of interest:** None declared

**Patient:** Female, 58  
**Final Diagnosis:** Congenital duplication of genitourinary system  
**Symptoms:** —  
**Medication:** —  
**Clinical Procedure:** Laparoscopic hysterectomy  
**Specialty:** Obstetrics and Gynecology

**Objective:** Congenital defects/diseases





**Background:** Müllerian duct anomalies represent a wide spectrum of congenital abnormalities ranging from simple uterine anomalies to more complex multisystem derangements. Complete duplication of uterus, cervix, and vagina may be associated with urologic and caudal gastrointestinal malformations.

**Case Report:** We present a case report detailing the management of a morbidly obese patient with postmenopausal bleeding and thickened endometrial stripe who had a very rare condition of pelvic organ duplication, including 2 hemiuteri, 2 vaginas, 2 hemibladders, and 2 each of ovaries, fallopian tubes, kidneys, and ureters. Laparoscopic hysterectomy was complicated by difficulties understanding urinary system anatomy requiring intraoperative urology consultation and imaging.

**Conclusions:** Management of patients with urogenital duplication and abnormal uterine bleeding requires a thorough understanding of possible associated malformations. Thorough preoperative evaluation, careful surgical exploration, and multidisciplinary approach may be necessary to avoid urologic injury in such patients.

**MeSH Keywords:** Congenital Abnormalities • Mullerian Ducts • Urogenital Abnormalities

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## Background

Müllerian duct anomalies represent a wide spectrum of congenital abnormalities and occur in up to 5.5% of females [1]. These anomalies arise as a result of abnormal organogenesis, fusion, or resorption and are frequently associated with congenital renal malformation, ovarian malposition, and anorectal abnormalities. Didelphic uterus is reported to occur in 0.1% to 0.5% of women [2]. Complete duplication of uterus, vagina, urethra, and bladder is even rarer, with very few cases reported in the literature.

Here we present a case report of a patient with a known uterus didelphys associated with a double vagina and 2 urinary bladders (hemibladders) who sought medical attention for postmenopausal bleeding. The complete delineation of her urinary tract anomalies – in particular, the number of ureters and how they drained into her urinary hemibladders – was largely uncertain.

This case report describes an interesting clinical scenario of a common condition, postmenopausal bleeding, in the setting of a rare congenital anomaly and outlines challenges in its evaluation and treatment.

## Case Report

The patient is a 58-year-old Caucasian postmenopausal woman, gravida 2 para 2. The patient sought medical attention at the gynecologic oncology clinic for evaluation of postmenopausal bleeding in the setting of a thickened endometrial stripe. She was known to have a congenital anomaly that included a duplicated genitourinary system (2 hemiuteri and cervixes, 2 vaginas, 2 clitorises, and 2 urinary hemibladders) and also a lower gastrointestinal (GI) tract abnormality surgically corrected in early childhood.

Her past medical history is notable for morbid obesity (body mass index [BMI] 41.5) and hypertension. Her past surgical history included a reconstructive anorectal surgery in childhood, umbilical hernia repair, and 2 low vertical cesarean deliveries. She was a smoker until age 44. Her menarche was at age 12, and she had a history of 2 full-term pregnancies, both in the right uterus, with cesarean delivery. She had no history of hormone replacement therapy. She had no history of abnormal pap smear results and was up to date on pap smear screening. Her last menstrual period was at the age of 50 years.

She was in her usual state of health until 5 months before the current visit, when she had had a 5-day episode of vaginal bleeding intermittently coming from both vaginas. Her initial gynecologic evaluation included a transabdominal ultrasound

examination that revealed a thickened endometrial stripe in her right hemiuterus (9 mm), a normal endometrial stripe in her left hemiuterus (4 mm), a left ovary with a simple-appearing cyst, and a normal right ovary. An in-office Pipelle® endometrial biopsy was attempted but was unsuccessful. A plan was then made to follow up with an ultrasound examination in 3 months.

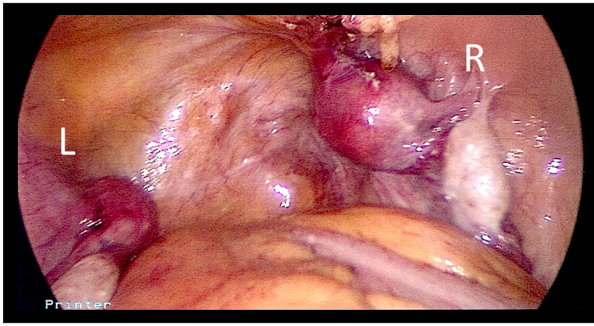
On repeat ultrasound examination, the endometrial stripe measured 10 mm in the right hemiuterus and 4 mm to 5 mm in the left, with both ovaries appearing normal. A subsequent sonohysterogram revealed right uterine endometrial stripe even thicker at 13 mm without evidence of distinct polyps/masses. She was then referred to the gynecologic oncology clinic for further evaluation and management.

During her visit to the gynecologic oncology clinic, the patient was asymptomatic except for minimal postprocedural spotting. Her vital signs were within normal limits. Pertinent physical examination findings included widely separated labia and duplicated vagina and cervixes. Her presentation and ultrasound findings were concerning for possible endometrial neoplasia, and as such endometrial sampling was warranted.

The patient was given an examination in the operating room (OR) under anesthesia, including hysteroscopy with dilatation and curettage (D&C). The OR assessment revealed a larger right vagina with a distinct urethral orifice and a smaller left vagina with an unclearly identified urethral orifice. Through hysteroscopy, left and right endometrial cavities were visualized. Both uteri had an endometrial cavity without obvious evidence of polyps or other gross lesions. The left uterus was notably underdeveloped, with a stenotic cervix; the cornua in the left uterus was difficult to identify definitively. Both uteri were curetted, but only a minimal amount of tissue was obtained from each; the pathologic examination did not reveal enough endometrial tissue to provide a diagnosis.

The endometrial sampling was inconclusive and there was persistent concern for possible endometrial malignancy given her endometrial thickness and BMI. After thorough counseling, the patient made the decision to proceed with removal of both uteri and frozen section evaluation with possible staging depending on these findings. She also decided to have a bilateral salpingo-oophorectomy at the time of the planned surgical procedure regardless of the frozen section findings. She was recommended to have a preoperative magnetic resonance imaging (MRI) scan to better characterize her urologic anomalies; however, she could not afford this.

The surgery was performed laparoscopically. After the abdominal cavity was entered, the survey of the pelvis revealed 2 widely separated uteri with the left uterus in close proximity

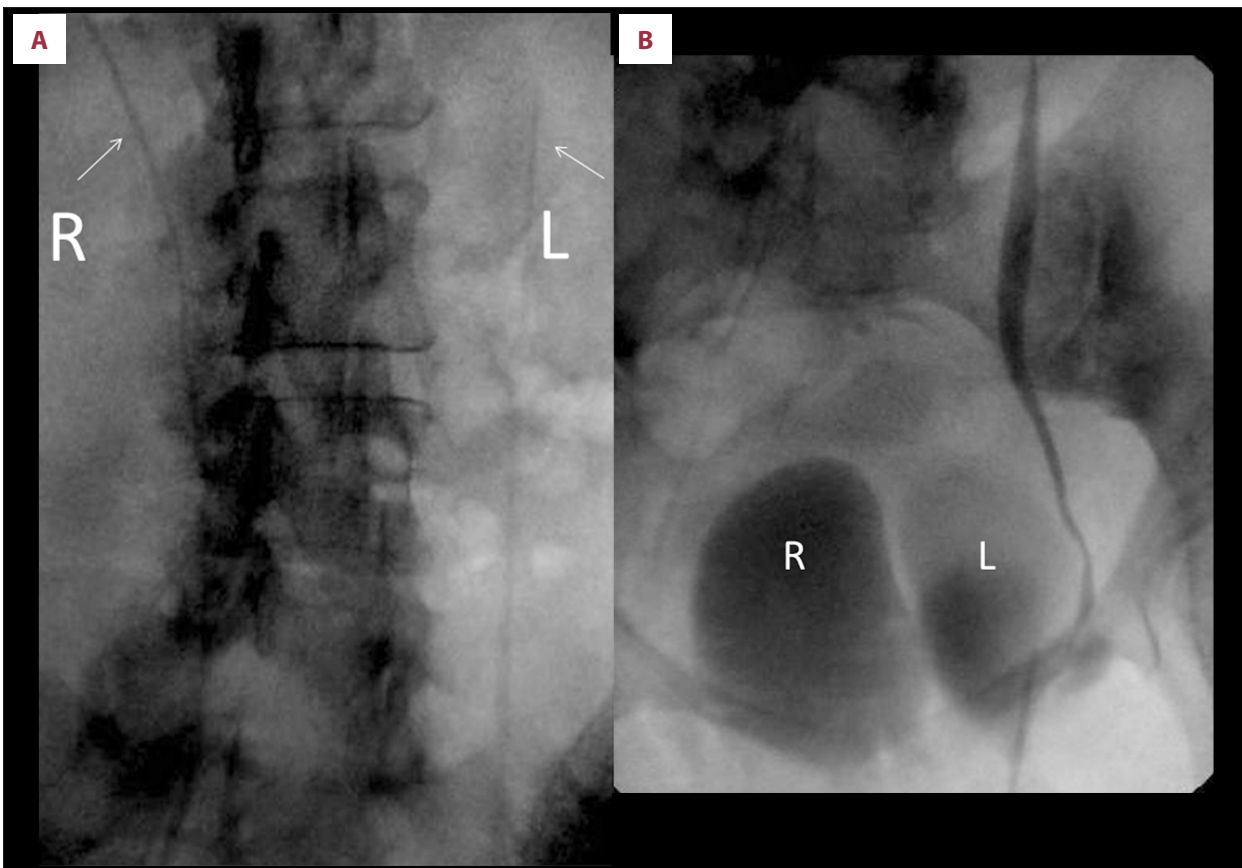


**Figure 1.** Laparoscopic image showing 2 widely separated uteri with the left uterus in close proximity to the left pelvic side wall (L) and right uterus closer to a more central location (R). Each uterus had its own fallopian tube and ovary, with a total of 2 fallopian tubes and 2 ovaries seen.

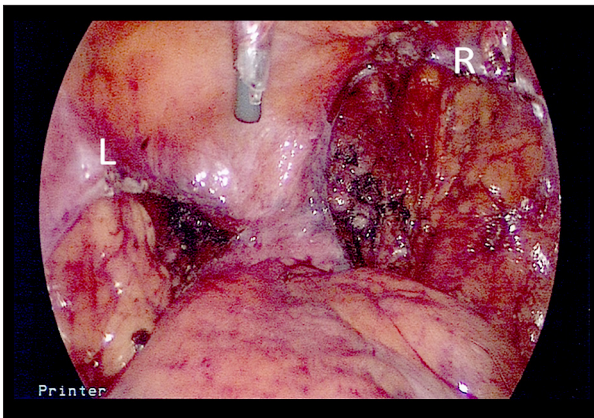
to the left pelvic side wall (Figure 1). Each uterus had its own fallopian tube and ovary, with a total of 2 fallopian tubes and 2 ovaries (Figure 1). Each uterus and cervix communicated to its own separate vagina, as mentioned previously. There were no gross abnormalities otherwise noted in the abdomen or pelvis and no free fluid seen. At the time of laparoscopic evaluation, there appeared to be 1 midline urinary bladder; 2 distinct

urinary bladders could not be identified. Some adhesions of the omentum to the anterior abdominal wall were noted.

Before performing both hysterectomies, the retroperitoneal space was entered bilaterally. Two ureters (1 on either side) were identified, and ureterolysis was done to the level of the corresponding uterine artery. The hysterectomies and bilateral salpingo-oophorectomy were performed thereafter in the usual fashion and the vaginas were sutured laparoscopically. Because of ureteral manipulation during bilateral ureterolysis, the decision was made to perform cystoscopy at the end of the procedure. Before cystoscopy, the patient received methylene blue intravenously to aid in the identification of the ureteral jets. The right urinary bladder was inspected via the right urethra, and only 1 ureteral orifice and jet was noted. Intraoperative intravenous pyelogram (IVP) revealed an obvious right urinary bladder and a less distinct left urinary bladder, with 1 ureter emanating from each renal pelvis (Figure 2A, 2B). Because of distorted anatomy, involvement of the urology team was needed to find the patient's left urethra. A much smaller left urethra was eventually identified slightly retracted above the left vagina. A 17F scope was passed into the left bladder after dilatation. The left ureteral orifice was seen and a blue-tinged jet



**Figure 2.** (A) IVP demonstrating 1 ureter on either side emanating from each renal pelvis. (L) identifies the left ureter and (R) identifies the right ureter. (B) IVP showing an obvious right urinary bladder (R) and a less distinct left urinary bladder (L).



**Figure 3.** Laparoscopic image showing the pelvis after removal of left uterus (L) and right uterus (R) with accompanying ovary and fallopian tube.

was identified from this orifice. The ureter was then instilled with contrast dye and the retrograde pyelogram revealed a normal-caliber left ureter and a nondilated left collecting system. She was therefore noted to have 2 kidneys and interestingly each kidney had a single ureter which drained into a distinct and separate urinary bladder.

At this point, because frozen sections did not reveal malignancy, the procedure was completed (Figure 3). Foley catheters were placed in both urinary bladders. Estimated blood loss during the procedure was 50 mL. The final procedure was total laparoscopic right and left hysterectomy, bilateral salpingo-oophorectomy, lysis of adhesions, bilateral ureterolysis, bilateral cystoscopy, IVP, and retrograde left pyelogram.

The patient's postoperative course was uncomplicated, and she was discharged home on postoperative day 2 in good condition after passing the voiding trials. The final pathology showed both cervixes had no significant abnormality; both uteri contained small endometrial polyps, small (<0.4 cm) leiomyomas, and weakly proliferative endometrium. A benign serous cystadenoma was identified in 1 ovary, and the fallopian tubes were without abnormality.

## Discussion

This case presents a typical clinical scenario of a patient with postmenopausal bleeding and thickened endometrial stripe that warranted further evaluation to rule out an endometrial precancerous condition. Fortunately, the final pathologic evaluation did not reveal malignancy; nevertheless, the evaluation and management steps (in-office endometrial biopsy, followed by hysteroscopy/D&C, then by hysterectomy) were appropriate. However, all steps of this work-up and treatment were complicated by the congenital pelvic organ malformation – urogenital

duplication – that made preoperative evaluation and intraoperative management challenging and required a multiteam approach. A thorough understanding of the embryologic basis for such anomalies and the potential for associated malformations may improve patient care in such complicated cases.

Urogenital duplications are rare congenital anomalies with varying modes of presentation. The exact anatomy depends on the type of anomaly and is related to the error in embryologic development. By the fourth week of embryonic development, the urogenital ridge is formed on both sides of the dorsal aorta, further giving rise to the nephrogenic cord and the gonadal ridge, which subsequently develop into the urinary and genital systems, respectively. By the fifth and sixth weeks, 2 pairs of genital ducts are present in both male and female embryos – mesonephric (wolffian) and paramesonephric (müllerian). In the absence of male developmental stimuli, the paramesonephric ducts continue to develop. By the process of fusion that starts caudally and progresses cranially, müllerian ducts form the uterovaginal canal that later develops into the upper portion of the vagina and uterus, while their nonfused portions become fallopian tubes. Female gubernaculum is an embryonic structure that is thought to play an important role in the development of Müllerian ducts in a female embryo. It is believed to be of muscular origin and by being attached to the müllerian ducts it allows or even induces proper development of the uterus and fallopian tubes. Further into development female gubernaculum persists as the round ligament which penetrates abdominal wall, and ovarian ligament [3]. Reproductive system develops in close proximity in location and time to urinary tract and lower GI systems. During the fourth week of development a blind-ended caudal hindgut is formed. The most distal end of the hindgut is the cloaca, which by the seventh to eighth week becomes separated by urorectal septum into the anorectal canal and the urogenital sinus (subsequently giving rise to urinary bladder and urethra) [4]. The latter contacts the uterovaginal primordium, which results in the formation of paired outgrowths – sinovaginal bulbs. Sinovaginal bulbs eventually fuse to form the vaginal plate, the central part of which breaks down and forms the lower aspect of the vaginal lumen [5].

Three separate classification systems have been proposed to classify female genital anomalies: the American Fertility Society (AFS) classification system; the embryological-clinical classification system; and the Vagina, Cervix, Uterus, Adnexa, and associated Malformations (VCUAM) classification system, with the AFS system being the most widely used [6]. According to the AFS system, the 7 distinct groups of female genital anomalies include (I) segmental or complete agenesis or hypoplasia, (II) unicornuate uterus with or without a rudimentary horn, (III) didelphys uterus, (IV) complete or partial bicornuate uterus, (V) complete or partial septate uterus, (VI) arcuate uterus, and

(VII) diethylstilbestrol (DES) – related anomalies. Embryologically these groups reflect an arrest in 1 of the following processes: initial development (groups I and II), fusion (groups III and IV), and degeneration (groups V and VI). Unfortunately, this classification does not characterize other associated anomalies [7].

Bladder duplication can be partial (1 urethra and 2 bladder halves that are not fully separated) or complete (2 independent bladders with full thickness wall). In cases of complete bladder duplication the septum can be in sagittal or coronal planes, with the former being the most common type [8]. There may be a partial or complete duplication of the genital tract in patients with bladder duplication, ranging from a bicornuate uterus to complete duplication of uterus, vagina, and vulva. Some patients are identified in childhood because of abnormalities of external genitalia, voiding difficulties, urinary tract obstruction, incontinence, anorectal, or skeletal malformations [9]. Other patients can remain asymptomatic for many years and are found during evaluation for infertility, endometriosis, or miscarriage or as incidental findings in adulthood. The prevalence of urologic anomalies accompanying müllerian anomalies is approximately 3% to 50%. Other congenital anomalies that may also be present include skeletal (29%) and cardiac anomalies (14.5%) [10].

A wide spectrum of associated congenital anomalies exists in women with bladder and urethral duplication, including caudal GI malformations, such as anal stenosis, ectopic anus, and partial or complete colon, rectum or anus duplication. Infrequently, the duplication can extend as far proximal as to include the terminal ileum. One of the largest studies to review bladder and urethral duplication reported that of 40 patients of both sexes with bladder and urethral duplication, 42% had duplication of the hindgut and 90% had some form of duplication of external genitalia [11]. Sagittal bladder and urethral duplication (Figure 2A, 2B), as was discovered in our patient, is associated with nonurogenital anomalies, such as duplication of the hindgut, more often than coronal bladder duplication [12]. Our patient had undergone an anorectal procedure in childhood; however, we were unable to retrieve the records for that procedure, and her urologic anomalies were not well characterized before surgery.

Postmenopausal bleeding may be a sign of a variety of conditions, including atrophic endometrium, endometrial polyp, endometrial hyperplasia, and uterine cancer. Endometrial carcinoma is found in approximately 10% of patients with

postmenopausal bleeding and thus warrants thorough work-up, which includes sonographic assessment of pelvic organs and tissue sampling. Not surprisingly, the management of postmenopausal bleeding in this patient was challenging because of her complex anatomy. At the time of the examination under anesthesia, a pediatric speculum was used to visualize the left cervix through the smaller and more lateral left vagina. Hysteroscopy revealed a single cornua in each uterus, which was confirmed at the time of laparoscopic hysterectomy. On laparoscopy, each uterus had a single fallopian tube emanating from a defined cornua (Figure 1). The altered anatomy necessitated retroperitoneal dissection and bilateral ureterolysis at the time of laparoscopic hysterectomy. Careful dissection was especially performed during the left hysterectomy due to the lateral location of the uterus to avoid injury to the ureter and the pelvic side wall vasculature.

In this case, intraoperative IVP was able to substitute for preoperative MRI evaluation of the urologic system. Generally, MRI scan is the imaging method of choice for characterizing complex urogenital abnormalities. T1-weighted delayed images may be used for assessment of the distal ureter if anomalous ureteral insertion is suspected or to identify the ureteral insertion in the case of ureter/bladder duplication [13]. In hindsight, preoperative IVP would have been helpful in the absence of MRI, but it was not performed. In the current case, intraoperative IVP was key in identifying the number of ureters and their drainage into the urinary hemibladders. The IVP assisted with the interpretation of findings on cystoscopy and provided critical information before the left retrograde pyelogram. It would have been difficult to exclude a potential injury to an overlooked double ureter on either side without these intraoperative studies.

## Conclusions

Management of patients with urogenital duplication and abnormal uterine bleeding requires a thorough understanding of possible associated malformations. Careful surgical exploration and intraoperative urologic studies, as well as urology consultation, may be necessary to avoid urologic injury, especially in patients who lack preoperative imaging.

## Statement

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