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Surgical Management of an Obstructive Müllerian Anomaly in a Patient with Anorectal Malformation

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Abstract

Müllerian duct anomalies are rare in the general population, occurring in less than 3% of women, but much more prevalent in female patients with anorectal malformation, occurring in up to 30% of these patients. Unicornuate uterus with a rudimentary non-communicating horn is a congenital anomaly of Mullerian development which can be seen in isolation or in conjunction with other anomalies, with several case reports described in patients with VACTERL association. These anomalies may be asymptomatic until the patient develops dysmenorrhea or devastating obstetrical complications. We describe the successful surgical management of an obstructive Müllerian anomaly in a post-pubertal female patient with anorectal malformation.

Keywords

Müllerian anomaly; anorectal malformation; laparoscopy

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Declaration of interests

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.

1. Introduction

Müllerian duct anomalies (MDAs) are rare in the general population, occurring in less than 3% of women. However, female patients with anorectal malformation have an MDA prevalence of up to 30%. Of these, 15% are a subtype called unicornuate uterus. Unicornuate uterus with a rudimentary non-communicating horn (Figure 1) can be seen in isolation or in conjunction with other anomalies, with several case reports described in patients with VACTERL association. The rudimentary horn can distend with menstrual blood, causing chronic or acute pelvic pain, adnexal torsion or house an ectopic pregnancy and result in rupture and hemodynamic instability.

We present a case of a 14-year-old girl with left renal agenesis and previously repaired anorectal malformation, with chronic left sided abdominal pain since menarche, found to have a right unicornuate uterus with a distended, blood-filled rudimentary left non-communicating horn and fallopian tube. She underwent laparoscopic excision of the left fallopian tube and rudimentary horn.

2. Case Report

A 14-year-old girl with a history of left renal agenesis and anorectal malformation presented to the emergency room with chronic left-sided abdominal pain starting at menarche. At birth, the patient was diagnosed with anorectal malformation with rectovestibular fistula to the posterior introitus. On day-of-life 1 she underwent a diverting sigmoid colostomy. At one year of age, she underwent posterior sagittal anorectoplasty, followed by colostomy reversal at 18 months of age. She had no surgical follow-up for the subsequent decade.

Starting at age 11, she began experiencing intermittent crampy left lower quadrant pain associated with menses each month, worsening in recent years. She had visited various emergency departments more than 15 times over a 2.5-year period with these symptoms and had several ultrasounds as well as magnetic resonance imaging (MRI) which were read as concerning for uterus didelphys with hematocolpos. She had been started on oral contraceptives, which ceased menstruation but did not provide relief of her symptoms. She saw an adult gynecologist who recommended an exam under anesthesia, but the patient was unable to follow-up due to healthcare access issues related to insurance. She continued to experience cyclical left-sided abdominal pain and presented to our emergency department after three years of progressive symptoms.

On initial physical exam, she exhibited significant left-sided abdominal tenderness. Complete blood count (CBC) revealed mild anemia (Hemoglobin 11 g/dL) and no leukocytosis. Beta-hCG was undetectable. Abdominal ultrasound demonstrated a distended left uterine horn measuring 13 × 7.2 × 6.8 cm with an adjacent left adnexal structure measuring 7.0 × 5.1 × 7.2 cm, favored to be a hematosalpinx. A computed tomography (CT) scan was obtained which showed a right unicornuate uterus with an obstructed left non-communicating rudimentary uterine horn and dilated left fallopian tube (Figure 2). Pediatric surgery was consulted for management. Given her long-standing symptoms and lack of gynecological or colorectal follow-up, we proceeded to the operating room to

evaluate her gynecologic and anorectal anatomy, and potentially address her obstructive symptoms.

The patient was taken to the operating room for exam under anesthesia, vaginoscopy, and diagnostic laparoscopy. We began with an exam under anesthesia and vaginoscopy, which showed a normal-appearing, patent vagina. On vaginal speculum exam, there was one normal-appearing cervix oriented towards the patient's right side. We did not identify a vaginal septum or other vaginal anomalies. Importantly, there was no left-sided cervix.

We proceeded with diagnostic laparoscopy (Supplemental Video). Upon entry into the abdomen, we encountered a markedly dilated left-sided uterine horn that was adjacent to the left ovary. The left uterine horn was tracked inferiorly and medially where it met the right uterine horn on the other side. We also identified a dilated left fallopian tube which was attached to the left uterine horn by a thin fibrous ligament. The right ovary appeared normal (Figure 3). There was no communication between the left uterine horn and the vagina, which we confirmed on simultaneous vaginoscopy. As mentioned, the left ovary was adherent to the left rudimentary uterine horn, but otherwise there was minimal inflammation within the pelvis.

After making the diagnosis of an obstructed rudimentary non-communicating left uterine horn, we performed an ovary-sparing laparoscopic excision for definitive treatment. We mobilized the left ovary off the left uterine horn taking care to preserve the gonadal vessels supplying the ovary. The left fallopian tube was then mobilized. The noncommunicating left uterine horn was dissected off the pelvic sidewall down to the confluence of the two uterine horns and separated from the top of the vagina. We verified by repeat vaginoscopy that there was no violation of the vaginal wall as we excised the rudimentary horn, dividing its attachments to the right unicornuate uterus with a bipolar sealing device. The port site at the patient's prior colostomy closure site was enlarged, and the left fallopian tube and uterine horn were removed en bloc after drainage of the accumulated menses. Both ovaries were preserved.

The final diagnosis was a right unicornuate uterus with a dilated, rudimentary, non-communicating left uterine horn and left hematosalpinx. These structures were connected by thin fibrous tissue (Figure 4). The patient recovered uneventfully and was discharged on postoperative day 2. On follow-up at 3 months after surgery she is doing well, and she has regular menstruation without her prior abdominal pain.

3. Discussion

We present a case of a delayed diagnosis of unicornuate uterus with a rudimentary non-communicating left uterine horn in a patient with history of an anorectal malformation and unilateral renal agenesis and describe successful laparoscopic management. A review of the literature suggests that 30% of women with an anorectal malformation have an associated gynecologic anomaly (1). A unicornuate uterus is present in up to 15% of patients with Müllerian anomalies and is most commonly rudimentary and non-communicating, thus placing patients at risk for development of obstructive symptoms after menarche (1).

Several treatment options exist, including oral contraceptives, percutaneous drainage, and surgical excision. We recommend that female patients with an anorectal malformation have an assessment of their Müllerian structures. Often this is a longitudinal assessment – with initial evaluation of the reproductive tract and Müllerian structures at the time of their initial ARM repair to the extent possible and again around the onset of puberty. A screening pelvic ultrasound should be performed after thelarche. A pelvic MRI may be necessary for delineation of the Müllerian anatomy. There should be a high index of suspicion for any signs of menstrual outflow obstruction (2).

There are several different types of Müllerian anomalies, with the most common being the septate or bicornuate uterus variants. One type of anomaly, the unicornuate, results from failed development of one Müllerian duct, with normal contralateral development. Obstructive symptoms commonly develop after menarche, but some patients remain asymptomatic through adolescence and present with obstetrical complications such as ruptured ectopic pregnancies (3,4). In this patient, her clinical history was particularly important for two reasons. First, as many as 30% of patients with anorectal malformation have gynecologic anomalies. Additionally, patients with unilateral renal agenesis have a 50–60% rate of associated genital tract anomaly (5). This is explained by their close embryologic origin, with the Müllerian ducts giving rise to the uterus and upper vagina and the ureteric bud off of the Wolffian or mesonephric ducts giving rise to the kidney. Thus, a high index of suspicion must be maintained for associated anomalies in this patient population.

All patients with anorectal malformation require ongoing surgical follow-up and evaluation, but in particular, female patients require gynecological evaluation at the onset of puberty. In the newborn period, patients with anorectal malformations should undergo a thorough physical exam and abdominal ultrasound should be performed to evaluate the urinary system. Newborn ultrasound may identify markedly abnormal Müllerian structures in newborn girls, but ultrasound at that age is inadequate for thorough evaluation. Thus, females with anorectal malformations should undergo formal evaluation of their internal gynecologic anatomy prior to the onset of menarche to identify and treat any potential sources of obstruction (e.g. vaginal septum, rudimentary horn) (2). We also recommend assessing internal gynecologic anatomy intraoperatively during other indicated surgical procedures prior to puberty, most commonly during colostomy reversal. Importantly, pelvic imaging such as ultrasound or MRI (6,7) should be obtained after the start of thelarche to assess for developing obstructive pathology. Dysmenorrhea in a patient with an anorectal malformation should be investigated as a potential symptom of gynecologic obstruction. It is critical that this condition be recognized and treated prior to child-bearing ages as implantation of an ectopic pregnancy in the rudimentary uterus can result in rupture and catastrophic hemorrhage (3,4). As our case illustrates, it is important to recognize that some women with obstructive anatomy can continue to have regular menses if only one side of their Mullerian structures is affected.

There are several treatment options available for patients with obstructed Müllerian anomalies. Hormonal medication such as oral contraceptives can temporarily pause menses, thus stopping the accumulation of blood in the obstructed structure. However, this

treatment was only partially effective for our patient, and she continued to have significant abdominal pain. The relief of obstructive symptoms may more immediately be obtained by percutaneous drainage of accumulated menses (8). This may be useful in cases of significant inflammation within the pelvis, in cases where the anatomy is ill-defined, or in a patient who is unable to undergo excision of the rudimentary structures at the time of presentation. However, excision of abnormal structures may still be necessary to prevent ongoing obstruction and to avoid future obstetric complications such as implantation of an ectopic pregnancy within the rudimentary horn (3,4). In our patient, due to her long lapse in surgical follow-up and incomplete evaluation of her internal gynecologic anatomy, we elected to definitively assess her gynecological and anorectal anatomy by performing simultaneous examination under anesthesia, vaginoscopy, and diagnostic laparoscopy. Because we encountered minimal inflammation and were able to obtain a thorough assessment of her anatomy, we were able to provide definitive treatment and avoid a later surgical procedure.

Lastly, women should be counseled on the potential effect of Müllerian anomalies on future reproductive capabilities. Among women with unicornuate uterus, the literature suggests that there is 30 to 50% live birth rate (9,10), with 20–30% of pregnancies ending in spontaneous abortion (11). As many as 20–40% of pregnancies end in preterm delivery (10). An ectopic rudimentary horn pregnancy occurs in 3% of cases (10) but can result in life-threatening rupture, underscoring the importance of early diagnosis and management of this condition.

4. Conclusion:

Gynecologic anomalies are far more prevalent in patients with anorectal malformations than the general population. Routine imaging should be obtained after thelarche to assess for obstruction in incompletely developed Müllerian structure as the patient goes through puberty. A multidisciplinary approach, with a pediatric gynecologist if available, is recommended to determine ability for future parturition in complex cases. Excision of rudimentary structures is indicated in select symptomatic women after a complete assessment of Müllerian anatomy.

Supplementary Material

Refer to Web version on PubMed Central for supplementary material.

Acknowledgments:

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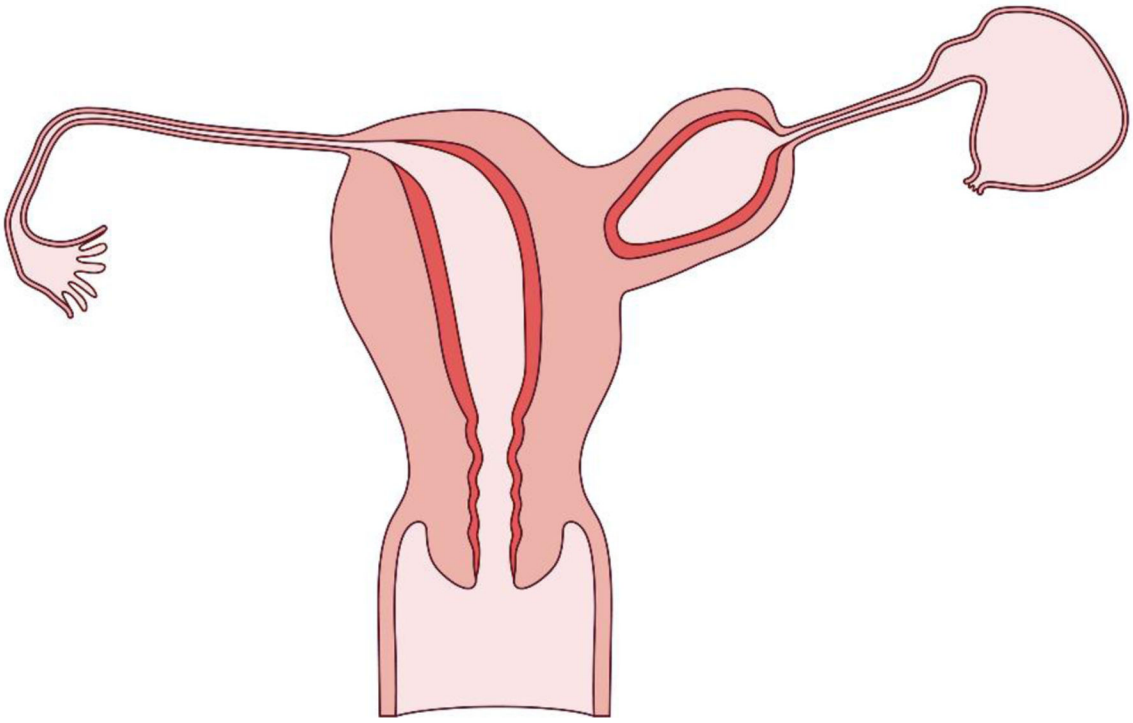

Figure 1:
Anatomy of a dilated rudimentary non-communicating uterine horn and fallopian tube

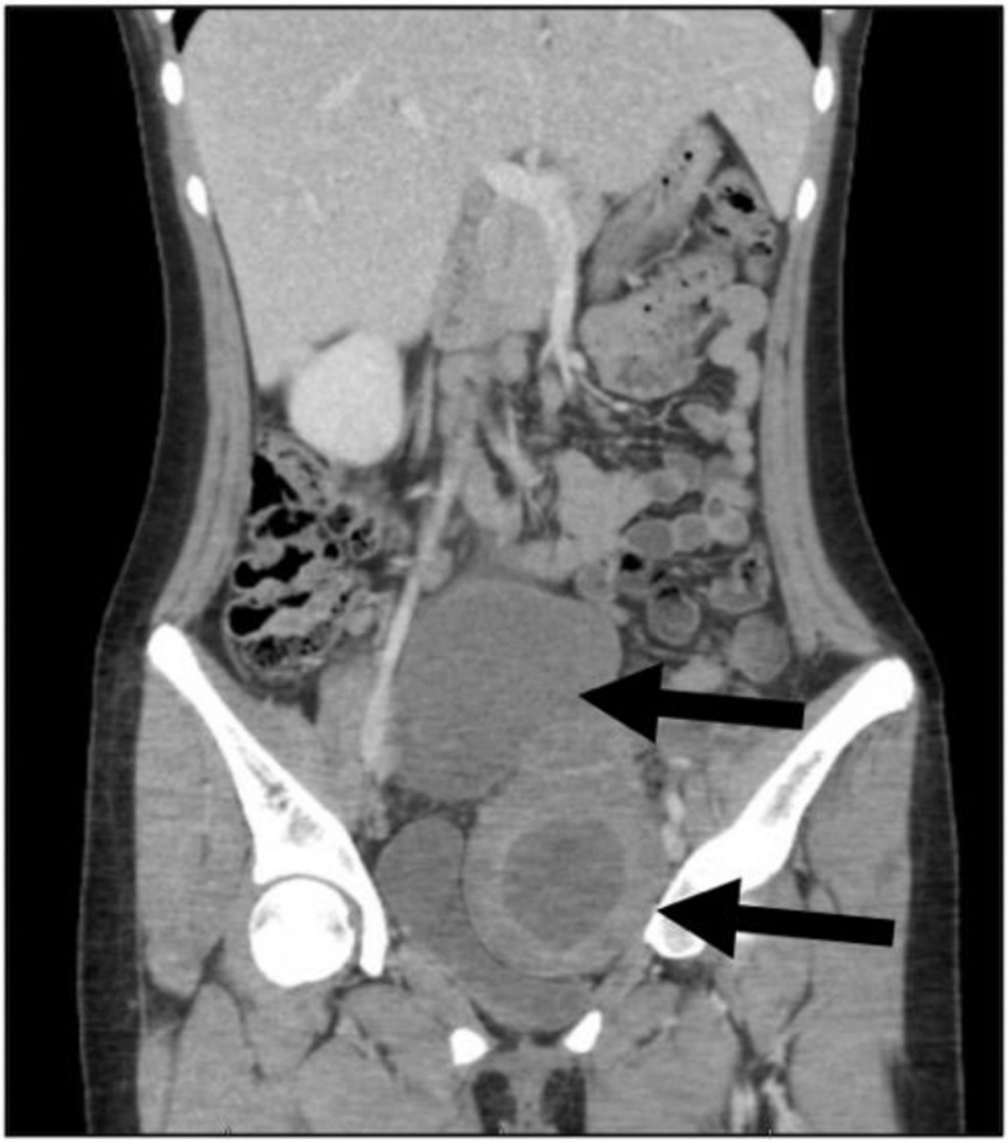

Figure 2:
Computed tomography scan showing dilated Müllerian structures (black arrows).

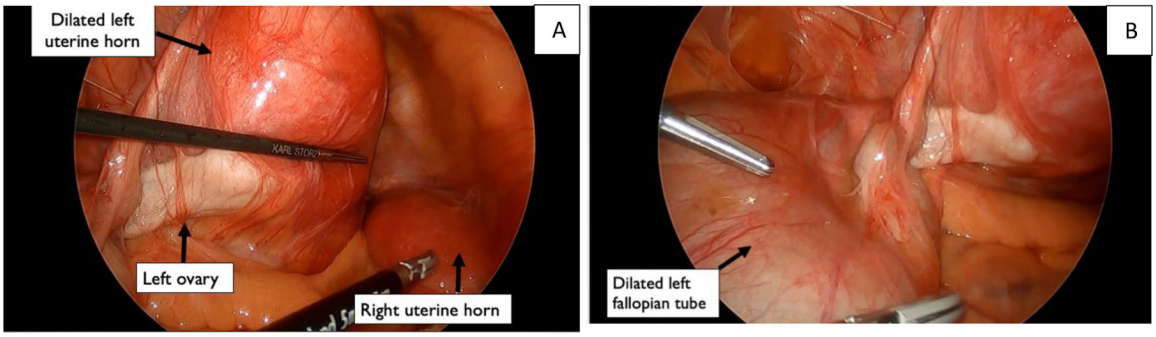

Figure 3: Initial view of the pelvis during laparoscopy.

Figure A shows a dilated left uterine horn with adjacent left ovary. The right uterine horn is visible in the pelvis. Figure B shows the dilated left fallopian tube.

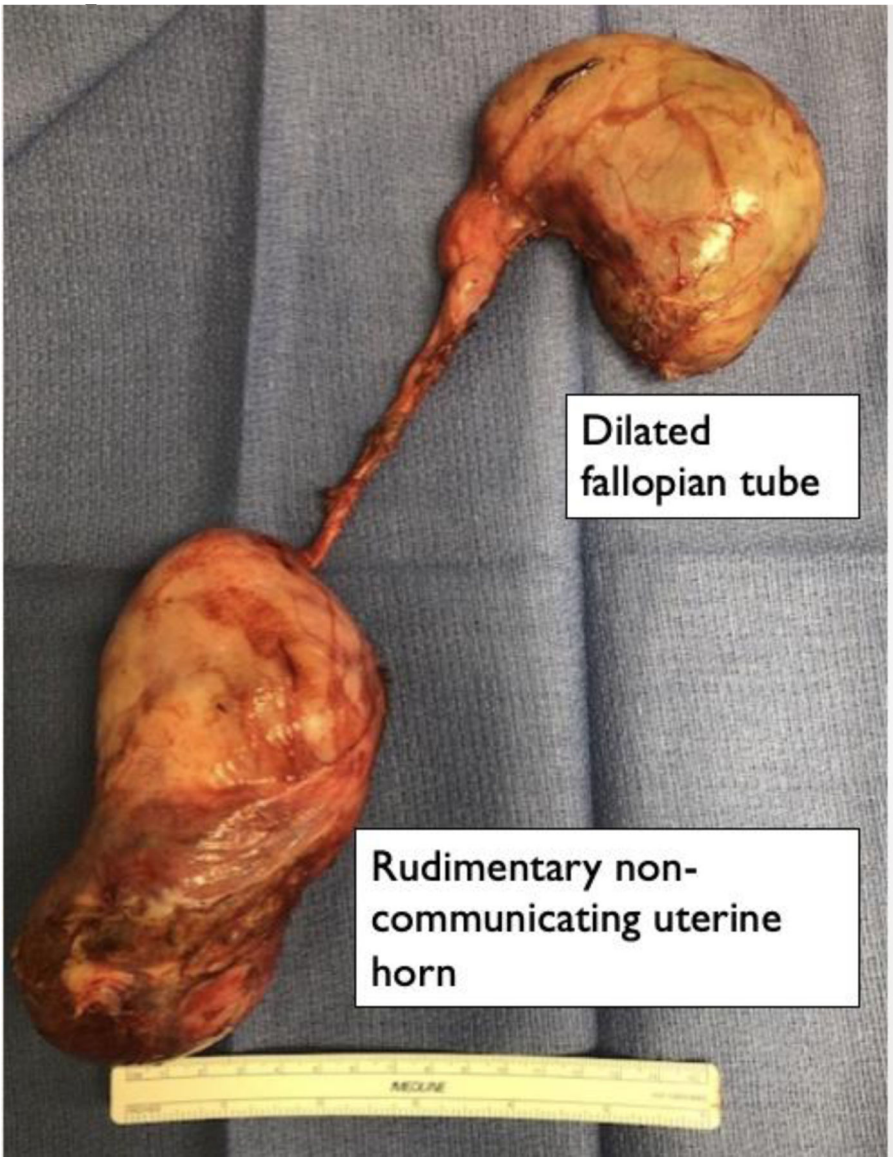

Figure 4:
Excised rudimentary non-communicating left uterine horn and dilated left fallopian tube.