

Laparoscopic Morcellation of Didelphic Uterus With Cervical and Renal Aplasia

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ABSTRACT

This is a case report (and review of the literature) of a 12-year and 10-month-old girl with a rare congenital anomaly of uterus didelphys, unilateral cervix aplasia, and ipsilateral renal aplasia. She had severe dysmenorrhea from the first menses. In an effort to preserve fertility, a cervical fistula was made that closed over. A laparoscopic hemi-hysterectomy was done successfully and rapidly with laparoscopic morcellation. Because no ureter was present, it was not necessary to trace it. For this congenital anomaly, laparoscopic morcellation of the obstructed hemiuterus is the preferred treatment either as a primary procedure or as a secondary procedure following failure of a surgical cervical fistula for the young patient.

Key Words: Laparoscopic morcellation, Uterus didelphys, Cervix aplasia.

INTRODUCTION

A congenital anomaly syndrome was reported in 1922 of uterus didelphys, unilateral hematocolpos with distal vaginal aplasia, and ipsilateral renal agenesis.¹ Since then, over 180 such cases have been reported.² It has no eponym, and we refer to it as “no eponym” syndrome.³ The patients present with unilateral upper hematocolpos and painful menses that start many months after menarche, because initially the upper vagina can slowly distend without pain (**Figures 1 and 2**). The diagnosis is suggested by a paravaginal mass (hematocolpos) readily detected by vaginal or rectal examination. The appropriate surgical management for the usual no eponym syndrome is conservative vaginal marsupialization of the upper ipsilateral hematocolpos and leaving both hemiuteri in place.

Our patient had an unusual variation. Instead of obstruction by the unilateral distal vaginal aplasia, the obstruction was higher up due to cervix aplasia (**Figure 3**). In view of the young age and the desire to preserve fertility, we created a uterine fistula, which closed over. Rather than repeat the fistula with the possibility of infection, the hemiuterus was removed by laparoscopic hemi-hysterectomy morcellation.

CASE REPORT AND RESULTS

The patient was first seen at age 12 years 10 months because of severe left lower abdominal pain with the first menstrual period, which began 1 year and 9 months earlier. For the previous 6 months, the pain had become continuous.

The enlarged left hemiuterus was palpated arising from the pelvis to 6 cm above the symphysis pubis. On rectal examination, the lower pole was tense, tender, cystic, and high (6 cm from the pelvic floor). Abdominal sonogram indicated left renal agenesis. Pelvic sonogram showed a 13-cm x 10-cm hematometra. MRI depicted a uterus didelphys. The left hemiuterus wall was dilated by old blood and elevated the left adnexa (**Figure 3**). The right uterus was normal. The impression was uterus didelphys with obstruction of the left uterus and hematometra. There was no hematocolpos. The MRI did not detect the left cervical

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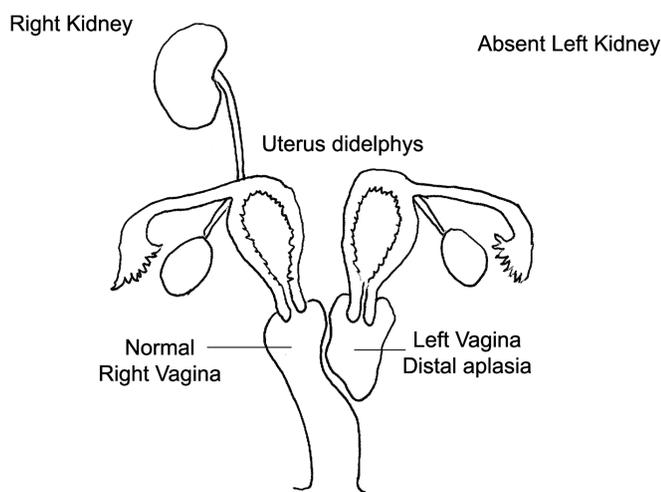


Figure 1. The usual “no eponym” anomaly syndrome before menstruation begins.

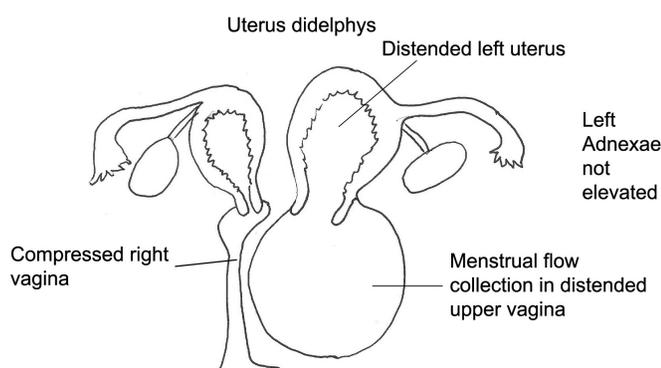


Figure 2. The usual “no eponym” anomaly syndrome after the start of menstruation. The hematocolpos is retroperitoneal and deep in the pelvis, requiring a vaginal surgical approach.

aplasia. The combination of uterus didelphys, cervical aplasia, and elevated adnexa has not been described in the literature to the best of our knowledge.

Examination with the patient under general anesthesia revealed one normal vagina and one right normal cervix. There was no left cervix. The bulging left vaginal fornix was punctured and dilated with the release of a large volume of old, pasty menstrual blood from the left hematometra. A wide number 24 Foley bladder catheter with a large 10-mL balloon was left indwelling for drainage. The catheter passed 2 weeks later. After surgery, the first menses were painless. Eight weeks later, a 1-finger vaginal examination palpated a distended 7-cm left hemiuterus fundus without any fistula opening. An MRI showed marked distention of the left hemiuterus with old and new blood and a new left hydro-hemato-salpinx. Two months

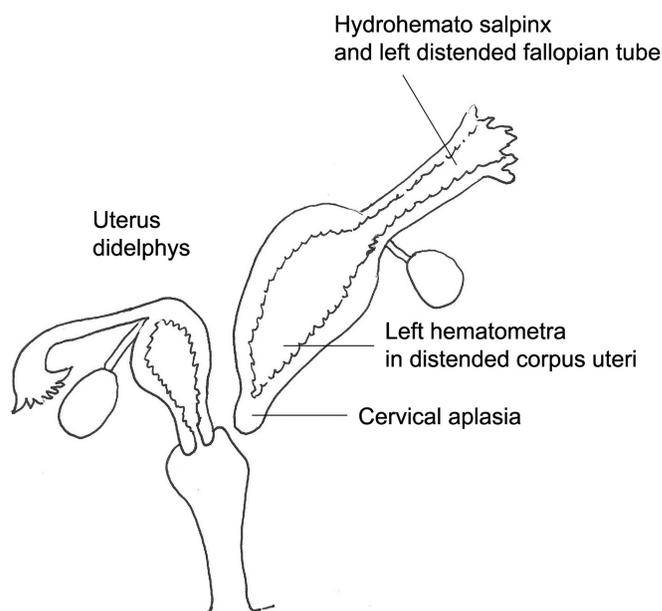


Figure 3. Our case report. The obstruction was left cervix aplasia. The hematometra is intraperitoneal and surgery for it requires a laparotomy or laparoscopy. The hematometra has elevated the adnexa.

later, the patient had painful menses. A 1-finger examination palpated a 10-cm left posterior cystic mass. A repeat MRI showed no left cervix.

At the second surgery, there was no left hemiuterus fistula visible or palpable in the left vaginal fornix. Laparoscopy was done for the first time. There were extensive adhesions in the left pelvis. The anterior cul-de-sac had superficial endometriosis. A left salpingectomy was done because of the severe hematosalpinx. After dissection and hemostasis by bipolar and harmonic wave ultrasound instruments, the left hemiuterus was completely removed by a relatively simple and rapid laparoscopic morcellation (**Figure 4**) down to the left vaginal vault but not penetrating it. There was no unusual blood loss or need of suturing. Because there was no left kidney and therefore no left ureter, there was no need to identify it. There was no sign of the fistula. Six months later, an MRI showed an absent left hemiuterus and a normal right hemiuterus with a patent cervix, and the patient’s menses were painless.

DISCUSSION

Our case was similar to no eponym syndrome because of the uterus didelphys and unilateral renal agenesis. Our case differs from no eponym syndrome because of the unilateral cervical agenesis and the lack of hematocolpos.



Figure 4. Laparoscopic morcellation of enlarged left uterus. Normal right uterus with ovary and tube. Right sacro-uterine ligament and cul-de-sac visible. Diluted blood in deep pelvis.

The surgical approach is different for each, vaginal for the no eponym syndrome (**Figures 1 and 2**) versus abdominal for cervix aplasia (our case) (**Figure 3**).

Morcellation of a congenital hemi-uterus is an ideal surgical option especially for the young patient in whom there is a volume of solid benign tissue to be removed. In our patient and in the more common no eponym anomaly, no kidney and no ureter are present on the side being operated upon.

No unusual blood loss occurred. The morcellation itself only took a few minutes. The main lateral uterine blood vessels and the smaller vessels in the base and medially were readily controlled.

Whereas with the usual no eponym syndrome, dysmenorrhea begins many months or even a year after menarche; in our case, there was severe pain with the first menses because of cervix aplasia, hematometra, retrograde menstruation, hematosalpinx, and eventually pelvic endometriosis.

Although MRI imaging is more precise than pelvic ultrasound, the first MRI missed the left cervix aplasia.

Prior to the first surgery, it was assumed that the patient had the usual no eponym syndrome with a unilateral distal vaginal agenesis, which is treated by vaginal medial marsupialization of the upper obstructed vagina. Thus, the surgeon should be prepared for variations of the basic anomaly.

In retrospect, other clues were the lack of left hematocol-

pos, the left adnexa being elevated by the left hematometra on imaging, and a palpable lower abdominal pelvic mass.

The fistula rather than hemihysterectomy was created because of the desire to preserve all possible fertility and the young age of the patient. The fistula failed and rather than make another fistula to evacuate the hematometra with the danger of infection, the left hemiuterus was removed by laparoscopic morcellation.

Vaginal surgery in a teenager may be difficult because of a deep, narrow vagina and difficult exposure access. There are inherent problems in surgical management. The precise details of the anomaly may not be discovered until the surgery is in progress. The surgeon wishes to preserve possible fertility, and the informed consent usually comes from the parents based on the expected findings.

LITERATURE REVIEW

In 1999,⁴ there was a report of a 17-year-old with a uterus didelphys with a right hematometra, hypoplastic cervix, a right obstructed upper hemivagina, and an ipsilateral renal agenesis. The authors indicated that the “. . . majority of clinicians view hysterectomy as the optimal primary surgical management.” The right hemiuterus was removed by laparoscopy rather than attempting to make a fistula for fear of infection. They wrote, “laparoscopic hemihysterectomy is a rather sophisticated technique. However, the automatic endoscopic stapler and suturing technique make these procedures possible.”

Our experience with laparoscopic morcellation was the opposite. We found it relatively easy, fast, and without the need for suturing. In addition, there was no need to search for a ureter.

In 2001,⁵ the same group wrote, “although treatment of didelphic uterus with a hypoplastic cervix is controversial, we believe that creation of the neocervix with prophylactic endometrial ablation may be a conservative effective modality.” Their article favored the latter. They reported 2 similar patients age 18 and 16 with a hypoplastic cervix, didelphic uterus, and ipsilateral renal agenesis. In the first, laparoscopic salpingectomy for hematosalpinx was done. Then they opened the hematometra, suctioned it out, and stitched it closed. Leaving the uterus in place, they did a uterovaginal canalization and endometrial ablation. Their second case was similar with a right cervix aplasia. A laparoscopic salpingectomy was done followed by a vaginal creation of a neocervix and an endometrial ablation.

They recommended a laparoscopic salpingectomy to prevent an ectopic pregnancy, leaving the obstructed hemiuterus, followed by a vaginal creation of a neocervix and endometrial ablation. Apparently they developed their procedure because of the technical difficulty of removing the obstructed hemiuterus by laparoscopy using traditional surgery. Their procedure is lengthy and leaves the problems of endometrial ablation in the young patient. There have not been any other reports of their procedure in the United States literature.

A 14-year-old⁶ had severe menstrual backache and urinary retention due to a large right hematometra of a didelphic uterus in the Pouch of Douglas with cervical aplasia, and ipsilateral renal aplasia. An abdominal right hemihysterectomy was done.

A 48-year-old had bilateral cervical agenesis of uterus didelphys that was treated by an abdominal bilateral hemihysterectomy.⁷

Despite the title of a paper in 1979, the operative note favors the no eponym syndrome. When the upper hematocolpos septum was incised without any other surgery, the hematocolpos and hematometra drained old menstrual blood.⁸

CONCLUSION

Our patient had a rare congenital anomaly that was a variation of an unusual syndrome (no eponym syndrome) anomaly. The latter is characterized by uterus didelphys, unilateral aplasia of one distal vagina, and almost 100% ipsilateral renal aplasia.

The initial MRI did not discover the single cervix aplasia, and the preoperative diagnosis was assumed to be the no eponym syndrome, which is treated by the vaginal route of a large marsupialization of the obstructed hematocolpos. Thus, the surgeon has to have experience with anomalies and be prepared for unexpected situations.

In retrospect, the clues to our case were immediate severe dysmenorrhea starting with the first menses; the image showed elevated adnexa on the affected side, and a tense tender cystic hematometra whose lower pole was relatively high.

In general, the usual approach to complete cervix agenesis is abdominal hysterectomy, because attempts to make a fistula usually are unsuccessful, and there is a danger from severe infection.

Nevertheless, after informed consent because of the

young age and the desire to preserve all possible fertility, an attempt was made to create a fistula cervix. After it failed, we did a laparoscopic morcellation of the obstruction hemiuterus.

We found laparoscopic morcellation to be relatively fast, simple, and with essentially no blood loss. An additional factor was the ipsilateral absent kidney and the ureter.

Two recent papers^{9,10} report a long-term rare complication of small pieces of tissue left after morcellation of the uterus forming benign painful tumors.

Donnez et al⁹ reported on a woman age 48 who had an uneventful laparoscopic morcellation subtotal hysterectomy because of menorrhagia and myomas. The histology was myomas and adenomyosis. She was given estrogen hormone replacement therapy. Five years later, she had pain and a 4-cm mass in the pararectal fossa that was an adenomyoma due to a piece of tissue left after morcellation.

Paul and Koshy¹⁰ reported multiple parasitic myomas after laparoscopic myomectomy with morcellation.

The surgeon should endeavor to remove any small pieces of morcellated tissue in the abdomen, perhaps by extensive washing. Patients should have long-term follow-up.

The usual morcellator is for single patient use. It is contraindicated for use on vascularized tissue or as a dissecting tool. It is not used for possible malignancy.

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