

# Ultrasound-assisted repair of a unique case of distal vaginal agenesis

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**Objective:** To describe a unique vaginal outlet obstruction and its ultrasound-assisted surgical correction.

**Design:** Case report.

**Setting:** An academic medical center.

**Patient(s):** A 12-year-old girl was seen with cyclic abdominal pain, nausea, and an abdominal mass. External genital examination revealed no vaginal opening or dimple. Transabdominal ultrasound revealed a large hematometrocolpos 5 cm proximal to the perineum, with an otherwise normal-appearing uterus and ovaries.

**Intervention(s):** Surgical treatment included perineal incision, creation of a 5-cm passage through connective tissue, drainage of the hematometrocolpos, and mobilization of the proximal vagina, allowing for pull-through vaginoplasty. Ultrasound guidance was used throughout the surgery and allowed for a safe transperineal approach. A vaginal form was placed in the neovagina.

**Main Outcome Measure(s):** Follow-up clinical evaluation and pelvic ultrasonography.

**Result(s):** A well-healed perineum and patent 5-cm-long vagina were observed at 4-month follow-up. The patient reported three regular menses. Postoperative pelvic ultrasound scan was normal.

**Conclusion(s):** We describe a unique case of isolated distal vaginal agenesis of significant length that was successfully treated via a perineal approach with the intra-operative assistance of ultrasound guidance. (Fertil Steril® 2007;87:976.e9–12. ©2007 by American Society for Reproductive Medicine.)

**Key Words:** Vaginal agenesis, hematometrocolpos, ultrasound guidance

Distal vaginal agenesis has been described in association with a variety of anomalies. Mayer-Rokitansky-Küster-Hauser syndrome, defined by vaginal agenesis and an absent or rudimentary uterus, occurs in 1 in 5,000 live female births (1). Distal vaginal agenesis, occurring in association with a rectovestibular fistula or urologic abnormalities, has also been reported in the literature (2, 3). Vaginal outlet obstruction resulting from a transverse vaginal septum or imperforate hymen can resemble vaginal agenesis, but excision of the relatively thin septum or hymen is all that is required (4).

Any deviation from the normal embryology of the reproductive system can result in unique malformations with similar clinical presentations. These variations require detailed knowledge of the specific anatomy to enable optimal surgical correction.

We present a case of a 12-year-old girl with isolated agenesis of a significant length of distal vagina with resultant hematometrocolpos 5 cm proximal from the perineum and describe the sonographically assisted surgical creation of a neovagina via a perineal approach.

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## CASE REPORT

A 12-year-old girl was seen locally for evaluation of cyclic abdominal pain, nausea, and vomiting. She was found to have an abdominal mass thought to be consistent with hematocolpos. She was taken to the operating room locally for planned repair of imperforate hymen, but examination under anesthesia was inconsistent with this diagnosis, and the surgery was abandoned.

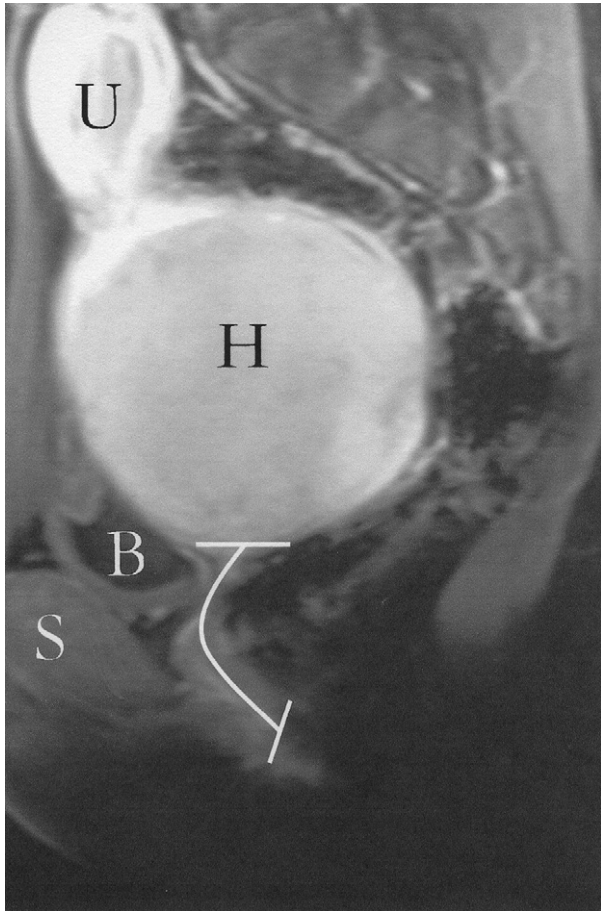
Further workup, including magnetic resonance imaging and computed tomographic scan, revealed a large heterogeneous fluid collection in the cervix and possibly the upper vagina. Magnetic resonance imaging also suggested a distance of 5 cm from the fluid collection to the perineum (Fig. 1). Intravenous pyelogram and voiding cystourethrogram were normal.

## Preoperative Evaluation

The patient was referred to our center for evaluation. She reported breast development for 1 year and a recent increase in growth rate, but no menses. Her symptoms of abdominal pain and emesis had persisted for several months, and she now complained of dysuria and dyschezia as well. Ultrasound examination at our center showed normal ovaries and a 5 × 6 cm hematometrocolpos, with an estimated length of 5 cm between the fluid collection and the perineum (Fig. 2).

## FIGURE 1

Magnetic resonance image showing a sagittal view of the patient's blood-filled uterus pushed cephalad to the level of the sacral promontory. The large hematometocolpos is demonstrated compressing bladder and bowel. The superimposed curved line delineates the significant distance from the hematocolpos to the perineum. U = uterus, H = hematometocolpos, B = bladder, S = pubic symphysis.



Kresowik. A unique case of distal vaginal agenesis. *Fertil Steril* 2007.

Possible diagnoses considered included a transverse vaginal septum, although there was no visible or palpable distal vagina, distal vaginal agenesis, or complete vaginal agenesis. Surgical intervention under ultrasound guidance was planned, and preparations were made for a possible vaginoplasty with skin graft to bridge the distance from the perineum to the mass if there was insufficient proximal vagina for a pull-through vaginoplasty.

### Operative Methods

First, cystoscopy was performed in the operating room to confirm that there were no concurrent urologic abnormal-

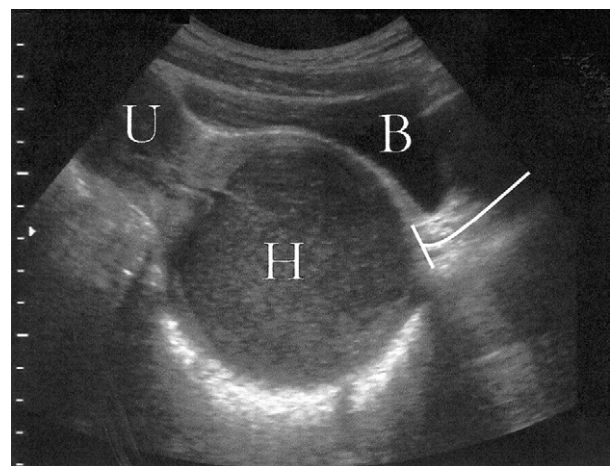
ities, with normal results. A three-way Foley catheter was placed, and the bladder was filled to assist in transabdominal ultrasound imaging. A 5-cm inverted U-shaped incision was made in the perineum below the urethral opening, and the tissue was undermined, thus creating a functional flap in case this was needed for the neovagina.

Under continuous transabdominal ultrasound guidance and concurrent rectal examination, a space was created bluntly through the incision and cephalad toward the hematometocolpos. When complete, the created space was approximately 5 cm in length and 2 finger-widths across and allowed direct visualization of the bulging fluid collection. Before incising the hematometocolpos, the proximal vaginal mucosa surrounding the blood collection was undermined bluntly with a finger to improve mobilization of this tissue (Fig. 3). This was greatly facilitated by ultrasound guidance, which reassured us that we had reached the hematometocolpos and could undermine without injury to the bladder or rectum.

The vaginal mucosa was anchored with sutures, and the hematometocolpos incised in the midline, draining approximately 500 mL of old, chocolate-colored blood. There appeared to be normal vaginal tissue lining the hematometocolpos, and multiple circumferential sutures were placed at the distal limits of this tissue. This vaginal tissue was then pulled down and circumferentially attached to the mucosa at the introitus to create a neovagina.

## FIGURE 2

Preoperative transabdominal ultrasound image showing a sagittal view of the patient's hematometocolpos. The superimposed line delineates the distance to be bridged during surgical repair. This image suggests a normal cervix, with most of the blood collected in an expanded proximal portion of the vagina. U = uterus, H = hematometocolpos, B = bladder.



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