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Case Report

# Conservative Treatment of a Herlyn-Werner-Wunderlich Müllerian Anomaly Variant, Noncommunicating Hemiuterus with Gartner Duct Pseudocyst

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**ABSTRACT** Asymmetric obstructed uterus didelphys (Herlyn-Werner-Wunderlich syndrome) is a rare congenital müllerian anomaly consisting of uterus didelphys, hemivaginal septum, and ipsilateral renal agenesis. Herein is reported a case of incomplete Herlyn-Werner-Wunderlich syndrome diagnosed using 3-dimensional transvaginal ultrasound in a 14-year-old patient with absence of the hemivaginal septum. The most contributive diagnostic factors and appropriate therapeutic management in such cases are discussed. Journal of Minimally Invasive Gynecology (2011) 18, 262–266 © 2011 AAGL. All rights reserved.

*Keywords:* Atypical; Conservative; Hemihysterectomy; Herlyn-Werner-Wunderlich syndrome; Uterine anomaly

Congenital anomalies of the female reproductive tract occur in 3% to 4% of fertile and infertile women [1]. Classification of müllerian anomalies according to the American Society for Reproductive Medicine (ASRM) includes hypoplastic defects, lateral fusion defects, vertical fusion defects, and defects caused from in utero exposure to diethylstilbestrol [2]. According to the ASRM, the anomaly described herein most closely represents a class III variant of Herlyn-Werner-Wunderlich syndrome with a noncommunicating uterine horn, ipsilateral renal agenesis, and Gartner pseudocyst (atretic blind hemivagina). Classification 2b of Acién [1] describes this anomaly both embryologically and clinically.

More commonly, a noncommunicating horn is categorized as an ASRM class IIB defect, and can be observed in 35% to 40% of women with a unicornuate uterus [3]. In the ASRM classification, the noncommunicating horn of a class IIB müllerian anomaly is described as rudimentary. Traditionally, treatment of unicornuate uterus with a noncommunicating rudimentary horn consists of laparoscopic resection of the noncommunicating rudimentary horn [4]. Laparoscopic removal of the noncommunicating horn was first reported in 1990, and since then 32 case reports have been published that discuss the technique and complications of this surgical practice [2,5,6]. Reasons to consider removal of the noncommunicating horn include improvement in dysmenorrhea, prevention of endometriosis, and prevention of pregnancy complications due to implantation in the rudimentary horn [5].

For class IIB müllerian anomalies, Falcone et al [7] described anatomic variations in the composition of the attachment between the rudimentary horn and the unicornuate uterus, and highlighted that the muscular or fibrous composition of this attachment should be a key consideration when choosing a treatment and surgical approach. The present case describes a unique unification procedure of a functional noncommunicating uterine horn in this rare anomaly (Fig. 1). This approach was chosen based on symmetric horn dimensions, muscular contiguity, endometrial adequacy, and the presence of only a thin, avascular, fibrous separation between the obstructed horn and the lower uterine segment in a patient who strongly desired a conservative approach.

## Case Report

All University of Utah Institutional Review Board requirements for this case report were met. The patient was

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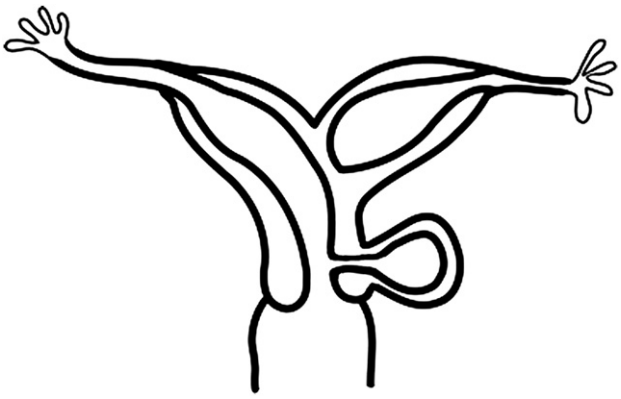

Fig. 1. Depiction of present case uterus.

a 14-year-old (gravida 0, para 0) young woman who underwent menarche at age 13 years. She reported to the referring physician regular 28-day cycles with bleeding for 7 days, and a 1-year history of increasing pelvic pain. Initial abdominal ultrasound examination documented a bicornuate-appearing uterus with a masslike effect distending into the left uterine horn filled with bloodlike products. Follow-up magnetic resonance imaging (Fig. 2) revealed an absent left kidney consistent with unilateral renal agenesis, uterine bicornis unicollis with a normal right uterine horn, and a thin (1–2 mm) fibrous band separating the obstructed left horn from the lower uterine segment. There was no vaginal septum, diverticulum, pseudocyst, or fluid collection in the vaginal or cervical region except the protruding proximal portion of the noncommunicating left uterine horn. The patient underwent laparoscopy, left salpingectomy, and drainage of hematometra. The initial operative report described 1 cervix, absence of a vaginal septum, a dilated left uterine horn, and a proximal tube with no communication between the left uterine horn and the cervix.

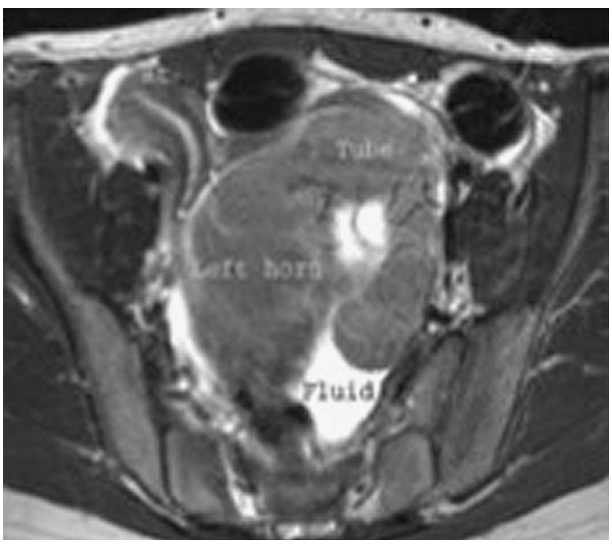


Fig. 2. Axial T2-weighted magnetic resonance image of pelvis before outside surgery. Magnetic resonance image shows a normal right uterine horn (RT) separated from the left horn by a thin low-signal fibrous band.

Two months after the initial surgery, the patient had increasing pelvic pain, and was referred to our tertiary care center for evaluation. During the initial assessment, the patient reported increasing daily pelvic pain, which worsened with menses. Abdominal and perineal ultrasound examination revealed a uterus measuring 6.9 cm transverse and 4.5 cm anteroposterior. The left horn of the uterus was distended with fluid consistent with blood. The patient was given norethindrone suppression therapy for 1 month, and a pelvic examination and vaginal ultrasonography were performed with the patient under conscious sedation to differentiate between an obstructed vaginal septum vs a noncommunicating left uterine horn. Ultrasonography revealed a smaller right uterine horn with functioning endometrium; a large left uterine horn measuring 38 × 38 mm with a fluid collection consistent with retained blood; no evidence of an obstructed hemivagina, vaginal septum, diverticulum, or pseudocyst; and a thin (2–3 mm) tissue plane separating the left uterine horn from the lower uterine segment. The patient and her family earnestly requested conservative therapy for management of the pain, with an attempt to avert hemihysterectomy, if warranted. The family and patient were aware of the absence of the left fallopian tube and potentially reduced utility of the left horn, but were also aware of transuterine migration, a common occurrence in animal species with bicornuate uterine configurations [8]. The patient and her family were also aware of the potential for recurrent obstruction. The pain was well controlled using norethindrone, and scheduling of surgery was for patient and family convenience, not because of pain.

Examination with the patient under anesthesia revealed no vaginal septum, diverticulum, or pseudocyst. Hysteroscopic and laparoscopic evaluation of the right uterine horn revealed a patent right tube and normal endometrial cavity. No communication was observed between the left uterine horn and the lower uterine segment, cervix, or right uterine horn. Laparoscopic evaluation revealed a normal-appearing right uterine horn, tube, and ovary; a dilated left uterine horn; a surgically absent left tube; and a normal left ovary. There was no evidence of any pelvic or abdominal endometriosis. The muscular connections between the lower uterine segment and the patent right and noncommunicating left uterine horns were firm, circumferential, and symmetric in appearance. Because of this muscular contiguity and symmetry and the presence of only a thin, nonvascular, nonmuscular fibrous tissue band between the left horn and the lower uterine segment, the decision was made to proceed with an attempt to create a communication with the lower uterine segment rather than hemihysterectomy. At laparoscopy (Fig. 3), a fundal incision was made in the left uterine horn to enable insertion of a 5-mm suction irrigation device, and the hematometra was suctioned and irrigated. A laparoscopic Kittner sponge was inserted into the fundal incision and directed toward the lower uterine segment of the