Müllerian anomalies occur in 0.1–0.5% of women [1]. Failure of the paired müllerian ducts to fuse laterally can result in uterus didelphys with obstructed unilateral vagina. This congenital disorder is often associated with renal anomalies, such as ipsilateral renal agenesis, renal dysplasia, a double collecting system, and an ectopic ureter [2–4]. Affected girls may present with low abdominal pain, severe dysmenorrhea, a pelvic or vaginal mass, abnormal vaginal discharge, and intermenstrual bleeding. These symptoms usually appear in adolescent girls after menarche.

Because of the small vaginal orifice and relatively high location of the vaginal septum in affected girls with an intact hymen, it is difficult to correct the vaginal anomaly without a hymenectomy. However, young female virgins with this congenital anomaly may have a strong desire to preserve the integrity of their hymenal ring for ethnic or personal reasons. Hysteroscopic resection of the vaginal septum with obstructed hemivagina has been reported to maintain the patient’s virginity [5]. Ultrasound guidance can be used to improve the safety of resectoscopic surgical procedures. [6]. We present the case of a female virgin with this rare syndrome, pyocolpos, and a vertical vaginal septum. She was treated with a minimally invasive resectoscopic operation under transabdominal ultrasound guidance. The integrity of her hymen was preserved after the surgery.

A 15-year-old girl visited our department with a 6-month history of intermittent low abdominal pain and fever. She denied having any sexual experience. Menarche had occurred at 12 years, with subsequent regular menstrual intervals. A pelvic examination revealed low abdominal tenderness and a pus-like discharge from the single vaginal canal. Transabdominal ultrasound and magnetic resonance imaging revealed two separate uterine cavities, normal bilateral ovaries, and a homogeneous cystic lesion in the pelvic cavity, continuing into the right uterus (Figure 1), together with a suspected obstruction and fluid accumulation in the right vagina. The right kidney was absent. Diagnostic sonohysterography with flexible hysteroscopy using transabdominal ultrasound revealed the patency of the left vagina, a normal appearance of the left cervical orifice, and the presence of a vertical vaginal septum with one bulging mass deviating into the left vagina (Figure 2A). No speculum or tenaculum was used during the operation. A 27F resectoscope (PK SuperPulse system; Gyrus ACMI, Southborough, MA, USA) was inserted into the left vagina and a continuous flow of 0.9% normal saline was used as the distending medium. Resectoscopic excision of the bulging vaginal septum was performed under transabdominal ultrasound guidance, and a massive amount of pus-like discharge was drained out (Figure 2B). The patient’s hymenal ring remained intact after surgery. She was diagnosed with uterus didelphys, a vertical vaginal septum, an obstructed right hemivagina with pyocolpos, and right renal agenesis, on the basis of imaging studies and operative findings.

She received postoperative empiric antibiotic management and recovered uneventfully. No recurrence was detected during follow-up.

Uterus didelphys with an obstructed hemivagina and ipsilateral renal agenesis (Herlyn-Werner-Wunderlich syndrome) are characteristic of the rare müllerian anomaly syndrome [7]. Young girls are always diagnosed after menarche with progressive and cyclic pelvic pain and a palpable pelvic mass owing to the unilateral obstructed vagina. An obstructed hemivagina may increase the risk of intraperitoneal endometriosis because of the presence of hematometra and hematosalpinx.
The incidence of endometriosis in patients with this müllerian anomaly is 16% [8]. This congenital defect is often related to renal anomalies, especially ipsilateral renal agenesis, which can easily be detected by prenatal or neonatal ultrasound. Whenever a female fetus or infant is diagnosed with unilateral renal agenesis, potentially associated anomalies such as an obstructed genital tract or congenital müllerian anomaly should be kept in mind. Regular sonographic evaluation of the genital tracts of these asymptomatic patients until the end of puberty is recommended to facilitate early detection of this genital tract anomaly. Accurate and early diagnosis of this syndrome is important to ensure adequate and prompt surgical treatment of the obstructed hemivagina and to prevent further complications such as hematocolpos, pyocolpos and even endometriosis [9].

Adolescent girls with uterus didelphys and an obstructed unilateral vagina often require surgical treatment for symptomatic relief. Traditional methods of transvaginal resection of the vaginal septum using scissors and sutures could easily destroy the integrity of an intact hymenal ring. Minimally invasive surgery without injury to the hymen is preferable. Hysteroscopy has been used in virgins for the clinical diagnosis and treatment of endometrial, cervical and vaginal lesions [10–12]. This procedure can be completed with preservation of the hymenal ring. However, few adolescent virgins with uterus didelphys and an obstructed vaginal septum retained intact hymenal rings after hysteroscopic resection [5,6,13]. The current case presents an additional adolescent virgin with this rare müllerian anomaly, including uterus didelphys, obstructed right hemivagina with vertical vaginal septum, and right renal agenesis. Her vaginal septum was successfully removed by resectoscope using transabdominal ultrasound guidance, without injury to the hymen ring. Young virginal women may prefer the results of resectoscopic excision.
of the vertical vaginal septum with transabdominal ultrasound guidance, compared with conventional transvaginal resection.

References


