

Successful Term Pregnancy in a Patient with Vaginal Agenesis, Cervical Atresia and Functional Uterus using a New Method: A Case Report

Marzieh Ghafarnejad^{1*}, Narges Khansari² and Sepideh Nekuie³

¹Associate Professor of Obstetrics and Gynecology, Women's Hospital (Affiliated to Tehran University of Medical Sciences), Tehran, Iran

²Resident of Obstetrics and Gynecology, Women's Hospital (Affiliated to Tehran University of Medical Sciences), Tehran, Iran

³Medical Student, Obstetrics and Gynecology Department, Women's Hospital, Tehran University of Medical Sciences, Tehran, Iran

***Corresponding Author:** Marzieh Ghafarnejad, Associate Professor of Obstetrics and Gynecology, Women's Hospital (Affiliated to Tehran University of Medical Sciences), North Ostad Nejatolahi Avenue, Karim Khan Street, Tehran, Iran.

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Abstract

Vaginal agenesis with functional uterus and cervical atresia is a rare condition with potential threat to the fertility. These patients have amenorrhea and progressive cyclic abdominal pain. Treatment modalities are controversial because of failure in preserving the uterus. We present a successful term pregnancy using a new method in a patient with vaginal agenesis, cervical atresia, and functional uterus.

The patient was a 14-year-old girl with amenorrhea, cyclic abdominal pain, normal secondary sexual development, absent vagina, a large cystic mass on rectal examination, and hematocolpos in ultrasound. She underwent laparoscopy, vaginoplasty, laparotomy, and uterocervical neovaginal cannulation using the Pezzer catheter. 2 years after marriage, she became pregnant and delivered a healthy baby by Cesarean section. Successful pregnancy was achieved in a women with vaginal agenesis after uterocervical cannulation.

Keywords: Vaginal Agenesis; Cervical Atresia, Functional Uterus, Pezzer Catheter, Cannulation, Fertility Preservation

Introduction

Congenital absence of the uterus and vagina is the most common form of vaginal agenesis. This is also called Müllerian agenesis, Müllerian aplasia, or Mayer Rokitansky Kuster Hauser syndrome [1]. Typically, patients with Müllerian agenesis present in adolescence with primary amenorrhea. Puberty and secondary sexual characteristics develop normally, while menarche does not occur. These patients have normal female phenotype, karyotype and ovarian function [1]. With the absence of the vagina, the uterus may be absent or present, normal or abnormal with different anomalies [1]. Vaginal agenesis is a relatively common disorder, whereas vaginal agenesis with a functioning uterus and cervical atresia is rarely encountered in clinical practice. Moreover, because the disorder presents early in adolescence, the clinicians must be familiar with different therapeutic approaches before the patient confronts serious clinical problems [2].

In patients with functional endometrium, cervical atresia, and vaginal agenesis, treatment modalities that preserve the reproductive function are controversial despite successful creation of neovagina. Traditionally, because of the high ascending infection risks and the likelihood of failure in preserving the functioning uterus, hysterectomy with creation of an artificial vagina was the treatment of choice, but nowadays conservative surgery with fertility consideration is becoming more and more frequently recommended. Surgical reconstruction of the vagina, restoration of menses, and maintenance of a patent genital tract are now becoming more challenging for gynecolo-

gists due to improved Assisted Reproductive Technology (ART) and new conservative surgical methods [3]. In this case report, we present a successful term pregnancy after uterine preservation in a girl with vaginal agenesis, cervical atresia, and functional uterus using the Pezzer catheter to cannulate the atretic cervix to the neovagina for menstrual blood drainage and uterus preservation.

Case Report

A 14-year-old girl was referred to our infertility clinic with complaint of primary amenorrhea and progressive cyclic lower abdominal pain. The pain had begun 6 months earlier, with 4-5 days' duration. The patient did not have chills, fever, gastrointestinal, or urinary tract symptoms. She had normal pubarche, adrenarche, and thelarche, appearing at the age of 11. The patient was referred to our hospital by a gynecologist from a city in the west of Iran with the first diagnosis of imperforated hymen or transverse vaginal septum.

On physical examination, the appearance of external genitalia (labia major and minor and clitoris) was normal, but there was no vaginal opening or bulging hymen. On rectal examination, a cystic mass was palpated at the expected level of the cervix with an upward extension. The mass was also palpated on abdominal examination. Physical examinations were otherwise normal. Pelvic ultrasonography reported a large fluid-filled mass, consistent with hematocolpos at the expected level of the cervix. Initial laboratory studies, including complete blood count, urinalysis, and serum chemistries, were within normal limits. Intravenous pyelography (IVP) reported a normal urinary tract. So, before operation our diagnosis was vaginal agenesis, functional uterus with or without cervical atresia. After IRB approval by Ethics Committee of medical faculty, the patient was counseled about this new method versus hysterectomy and she agreed to new procedure would be more accurate. She signed a written consent form. Thereafter, laparoscopy revealed that the uterus, ovaries, and both tubes were normal in the upper part of the pelvic cavity. The atretic cervix was markedly dilated like an enlarged mass, with a mean diameter of 8 × 8 cm. Pelvic endometriosis could not be observed; we, therefore, decided to perform vaginoplasty, followed by laparotomy, in order to create a conduit between the uterus and neovagina and preserve the patient's future childbearing potential. The patient was placed in lithotomic position and a neovagina was created via the McIndoe technique using no grafts. The procedure was commenced with a transverse incision through the mucosa of the vaginal vestibule. The space between the urethra and bladder anteriorly and the rectum posteriorly was opened via a blunt digital and sharp dissection, with the guide of the gloved finger inserted into the rectum and a urinary catheter fixed so as to prevent rectal, uretral, and vesical complications. Thus, a 10-cm deep cavity with a width of 2 cm was made between the rectal and vesical space. Afterward, the patient underwent laparotomy. A small incision was made at the anterior inferior portion of the dilated cervix, and a small biopsy (1 cm) of this part was sent to the pathologist, who reported a "portion of fibromuscular and smooth muscle tissue". After the incision was made, more than 400 cc of a thick, tenacious brownish material was evacuated from the dilated cervix. A connection was made between the upper roof of the neovagina and the lower part of the dilated cervix by opening the atretic portion of the cervix. To keep this orifice open, Pezzer catheter was inserted between the cervix and the upper part of the neovagina using a long clamp. The mushroom-like end of the Pezzer catheter (fenestrated part) was inserted into the cervix and the other part was pulled out from the vagina whilst the extra part was cut.

The site of the incision in the anterior inferior portion of the cervix was sutured. In our technique, no grafts were utilized the catheter was kept in place at least for 6 months. To keep open the newly created vaginal space, we inserted a 20-ml syringe mold into the vaginal cavity and fixed it to the labia with separated silk stitches. The dimensions of the syringe were 10 cm in length and 2 cm in diameter. A 2-cm window was produced on the cap of the tube. Two weeks after the surgery, the mold was removed in order to wash the neovagina and the Pezzer catheter with normal saline and to evaluate the width, length, and surface of the neovagina (2 and 9 cm, respectively). The vaginal surface had no infectious discharge. Again, a new mold was inserted into the cavity.

At the end of the third week, the fixed mold was removed and the patient was trained to use a movable mold regularly until the beginning of sexual intercourse. The patient was discharged and followed up monthly to evaluate the results of surgery. Menses returned and the pain was relieved. There were no signs of stenosis and infection. After 6 months, the Pezzer catheter was removed and the patient was carefully followed up to evaluate the maintenance of menstruation, cervical mucosal discharge, and width and length of neovagina.

She used a dilatator regularly. Menses continued monthly. Two years later, she got married and became pregnant spontaneously. She was advised to have her prenatal care in her hometown and to return at late third trimester to our center for close observation. She was visited at her 36th week of gestation, and ultrasound was performed, which was normal. At 37th week of gestation, as labor pain initiated, she underwent Cesarean section and gave birth to a 2500-gram female infant with good APGAR scores.

Summary and Conclusion

Müllerian agenesis is the second most common cause of primary amenorrhea [2]. Most patients with Müllerian agenesis have rudimentary Müllerian bulbs without any endometrial activity. Rare cases have active endometrium in uterine structures. These patients will present with primary amenorrhea and cyclic and chronic abdominal pain. Initial surgical management of congenital cervical atresia or aplasia with vaginal agenesis remains a subject of debate. In the past, because of postoperative complications of utero-neovaginal canalization, including peritonitis, recurrent obstruction of the utero-neovaginal canal, and persistent infertility, the majority of clinicians viewed hysterectomy as the optimal surgical management in these patients [4]. Nowadays, conservative surgery has become more frequently recommended. Vaginal agenesis with cervical atresia and functional endometrium is a challenging problem in the field of reconstructive surgery. Recent attempts are focused on preserving the uterus instead of hysterectomy and creating and keeping a new tunnel that mimics the normal vagina with fewer complications.

Different techniques have been described in the literature in this regard. Nearly in all of these techniques, some kinds of grafts are used for vaginal reconstruction. William Bates, *et al.* described a surgical technique on an 11-year-old girl with vaginal agenesis and partial cervical atresia, in which a split-thickness skin graft was secured into the newly canalized cervical os in combination with modified McIndoe vaginoplasty. After about 5 years, the authors reported a successful term pregnancy and subsequently cesarean section to protect both the cervix and the new vagina [2].

JV Deffarges, *et al.* reported utero-vaginal anastomosis instead of canalization in 18 cases with cervical atresia. A flap was used to provide more available tissue and avoid stenosis on the utero-neovaginal anastomosis. Ten patients tried to become pregnant during the study period, and there were 6 successful spontaneous pregnancies in 4 of them, while the remaining 6 women were infertile [5].

Recently, Ghafarnejad, *et al.* studied 6 patients with vaginal agenesis and one patient with cervicovaginal agenesis with a functional uterus, performing uterocervico or utero-neovaginal cannulation using the Pezzer catheter between. All the patients had a return of their menses and relief of pain. Three patients developed stenosis, and two cases suffered infection, which was subsequently treated successfully, self-image and quality of life improved in all the cases [3].

Munire Erman, *et al.* reported a 20-year-old woman with primary amenorrhea and cyclic menouria which MRI imaging revealed a remnant upper vagina and unicornuate uterus filled with fluid and renal agenesis in left side, intraoperatively a congenital vesicouterine fistulous tract was observed, after complete resection of fistulous tract and vaginal reconstruction using a sigmoid flap, connection of proximal part of neovagina to remnant cervix carried out. Finally, menses returned and menuria disappeared [7].

In the present study, unlike others [2,5], we did not make use of any grafts for lining the neovagina or flaps for anastomosis [6]. Moreover, we employed the Pezzer catheter to cannulate the atretic cervix to the neovagina. The patient had no signs of infection or stenosis, and the catheter was removed after 6 months. It is clear that usage of any grafts may increase the risk of complications such as infections, scars, and malignant changes in addition to increasing operation time [6]. Using the Pezzer catheter for cannulation is not only less costly but also more convenient and has the added benefit of decreasing operation time and complications. From our opinion, this case is the first report of spontaneous successful pregnancy in these patients using this method.

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