Posterior Sagittal Approach for Uterovaginal Anastomosis in a Case of Congenital Cervical Atresia with Anorectal Malformation

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INTRODUCTION

Congenital cervical atresia is a relatively rare Mullerian duct anomaly found in 50% of cases of vaginal atresia.[1] It presents usually at puberty with acute or chronic pelvic pain and primary amenorrhea. The management is controversial ranging from hysterectomy to recanalization or conservative management in the form of uterine catheterization. Hysterectomy eliminates the symptoms related to hematometra, but loss of reproductive function is irreversible. Thus, it should be avoided as the first choice and reserved only for cases where canalization attempts fail or are impossible. However, canalization procedures may lead to recurrent obstruction of the uterovaginal neocanal, and persistent infertility. Hence, hysterectomy had conventionally been the treatment of choice. Recently, some series have reported an improved reproductive performance after uterovaginal anastomosis.[2] Endoscopic canalization, laparotomy with uterovaginal anastomosis, and laparoscopic techniques have been mentioned in literature.[3,4] We report posterior sagittal approach for this anomaly which was associated with anorectal malformation (ARM).

Cervical atresia is a rare association with anorectal malformation (ARM) which can be missed till puberty in the presence of normal vaginal orifice. A 12-year-old girl operated for ARM in neonatal age presented with primary amenorrhea. She had a normal vaginal opening, short perineal body, and prolapsed anteposed anus and was diagnosed with cervical agenesis. As the posterior sagittal approach is standard to place the rectum in correct anatomical position, reconstruction of the anus along with adequate perineal body and uterovaginal anastomosis was performed through this approach. This report highlights the utility and versatility of this approach for the management of such complex cases.

KEYWORDS: Cervical atresia, hematometra, hematosalpinx, posterior sagittal approach

Case Report

A 12-year-old girl, who had undergone abdominoperineal pull through for ARM in neonatal age, presented with cyclical lower abdominal pain, primary amenorrhea, perineal excoriation, and rectal prolapse. On examination, she had normal external genitalia with two orifices in introitus representing urethra and vagina, an anteposed anus with prolapsed mucosa, perineal excoriation, and no perineal body. On abdominal examination, she had a long longitudinal midline scar with lumps palpable in bilateral iliac fossae and hypogastrium. On examination under general anesthesia, the possibility of imperforate hymen or vaginal septum was ruled out. She had a normal vagina with an adequate length of 6 cm, but on genitoscopy, no cervical impression was seen. The diagnostic laparoscopy confirmed the presence of Mullerian structures with bilateral hematosalpinx and hematometra. Contrast-enhanced computed tomography (CT) abdomen and pelvis also

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showed cervical agenesis and rectum and vagina lying close to each other for significant length [Figure 1]. An magnetic resonance imaging (MRI) pelvis was done for better delineation of anatomy preoperatively, which confirmed the CT findings with rectum lying anterior to sphincter muscle complex with no perineal body. The posterior sagittal approach was used to place rectum in correct anatomical position and uterovaginal anastomosis [Figure 2]. After the rectum was mobilized and retracted away, the upper end of the vagina was identified by palpating the vaginal stent. The lower end of the uterus identified by the bulging hematometra was confirmed by aspirating. Transverse incision was made over lower limit of hematometra and was decompressed completely. A similar incision was given over upper blind end of the vagina and uterovaginal anastomosis was done over 18 Fr Foley kept as stent per vaginally, which was guided into uterine cavity under vision [Figure 3]. Neoanus was created within sphincter muscle complex and perineal body was reconstructed. Residual hematosalpinx were decompressed laparoscopically. She required a diverting colostomy which was closed after 6 weeks. The vaginal stent was left in situ for 3 months and she menstruated through the stent twice before removal. Check genitoscopy was done 6 weeks after removal of stent which confirmed patent uterovaginal canal and the girl is having regular menstruation on the follow-up of 2 years with satisfactory rectal continence.

**Discussion**

Congenital cervical atresia is a rare Mullerian duct anomaly with an incidence of 0.01% in general population.\(^{[1]}\) It is estimated that only 4.8% of women with cervical atresia have a functioning uterus.\(^{[5]}\) These cases present with primary amenorrhea, well developed sexual characters, and cyclic abdominal pain. Clinical examination eliminates vaginal atresia or an imperforate hymen, but it may not always be possible to differentiate cervical atresia from a high vaginal septum.

Transabdominal or transperineal sonography might be helpful to identify the level of obstruction but is not reliable. The MRI is a more reliable imaging technique and may diagnose associated upper genital tract anomalies or problems such as hematosalpinx and endometriosis. Laparoscopy allows an exploration of the pelvis with the assessment of the internal genital organs.

Unlike most of the other Mullerian anomalies, the management of congenital cervical atresia is challenging. The earliest reported case of congenital cervical atresia managed by hysterotomy and cervical canalization was described in 1900 by Ludwig.\(^{[6]}\) The most successful surgical methods employed involved a transvaginal or transabdominal approach to create a neo-ostium through the dense fibrous cervix and
communicate it to the endometrial cavity and vagina. Patency was maintained by application of stents, with or without a surrounding full or split-thickness skin graft. However, re-stenosis of fibrous tissue, postoperative severe infection, or septicemia in occasional cases resulted in the recommendation of hysterectomy. In recent years, there is resurgence toward conservative surgery which is now being frequently attempted in congenital cervical atresia, with an aim to preserve the reproductive capability and relieve the menstrual symptoms. The goal is to sustain cyclic menses and avoid re-stenosis. Canalization techniques such as drilling, though easier, are associated with re-stenosis with complication rate of nearly 40%–60%. Hence, the technique of uterovaginal anastomosis described by Deffarges et al., is now preferred as it has lower risk of re-stenosis and can be performed even in the presence of associated vaginal atresia. Subsequently, novel techniques have been described by El Saman using laparoscopic assistance (i.e., endoscopic monitored canalization under vaginoscopic monitoring and retropubic balloon vaginoplasty). However, the long-term outcome and the reproductive performances after the laparoscopic techniques are yet to be reported.

Due to the risk of retrograde menstruation and subsequent development of endometriosis, creating an outflow tract should be done as early as possible (12–16 years). A successful uterovaginal anastomosis will depend on the size of the created ostium, the length of the canal, the presence of vaginal mucosa adjacent to the end of the neo-ostium, and the duration of stenting. The duration of the stenting ranges from 3 weeks to 3 months. We chose uterovaginal anastomosis through posterior sagittal approach and kept the stent for 3 months with no restenosis. We think keeping stent for a longer duration has reduced chances of stenosis.

**Conclusion**

In cases of cervical atresia, every attempt should be made to preserve the uterus. The posterior sagittal approach can be used for cases associated with ARM for uterovaginal anastomosis with good outcome.

**Declaration of patient consent**

The authors certify that they have obtained appropriate patient consent forms. In the form, the patient has given her consent for her images and other clinical information to be reported in the journal. The patient understands that name and initials will not be published and due efforts will be made to conceal identity, but anonymity cannot be guaranteed.

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**Conflicts of interest**

There are no conflicts of interest.

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