DOI: https://doi.org/10.54393/pbmj.v9i1.1335

PAKISTAN BIOMEDICAL JOURNAL
https://www.pakistanbmj.com/journal/index.php/pbmj/index
ISSN (E): 2709-2798, (P): 2709-278X
Volume 9, Issue 01 (January 2026)

Case Report

OPEN ACCESS

Cervicovaginal Agenesis: A Case Report of a Rare Congenital Anomaly with Delayed Clinical Recognition

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ARTICLE INFO

Keywords:
Premenarchial, Hematometra, Cervical Agenesis, Uterovaginal Anastomosis, Case Report

How to Cite:
Ali, N., Osama, M., Attique, M., Asghar, R., & Aman, A. (2026). Cervicovaginal Agenesis: A Case Report of a Rare Congenital Anomaly with Delayed Clinical Recognition : A Case Report of Cervicovaginal Agenesis . Pakistan BioMedical Journal, 9(1), 52-56. https://doi.org/10.54393/pbmj.v9i1.1335

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Received Date: 19th January, 2025
Revised Date: 4th July, 2025
Acceptance Date: 12th July, 2025
Published Date: 31st January, 2026

ABSTRACT

Rare congenital Mullerian anomalies include cervical agenesis. It has been found in 39% of cervical agenesis instances, which makes its association with vaginal agenesis which occurs even less frequently much rarer. For this ailment, a hysterectomy was the standard of care. However, due to improvements in assisted reproductive techniques and surgical advancements, conservative surgery can now be considered the first-line therapy Patients with primary amenorrhea should be actively evaluated for underlying causes. cervicovaginal agenesis is uncommon and has a variety of presentations. In a successful case of cervicovaginal agenesis treated with cervicovaginoplasty, a 14-year-old adolescent girl who had been experiencing cyclical pelvic discomfort and primary amenorrhea for five months came to the Women and Children Hospital Dera Ismail Khan's outpatient department. The patient's menstrual periods returned after surgery, and her cyclical abdominal pain decreased. Patients with primary amenorrhea should be actively evaluated for underlying causes. cervicovaginal agenesis is uncommon and has a variety of presentations. This might cause misdiagnosis and treatment delays. it is important to establish early diagnosis and proper management of these surgically amenable lesions to preserve normal physiology and fertility and prevent complications.

INTRODUCTION

Cervical agenesis is a rare congenital Mullerian anomaly occurs in one in every 80,000 to 100,000 births. It has been documented in 39% cases of cervical agenesis, making its relationship with vaginal agenesis, which is even more uncommon in occurrence [1]. Cervical agenesis is categorized as Ib by the American Fertility Society (AFS), which has historically been frequently utilized due to its simplicity and convenience of usage [2]. Due to the blockage of menstrual flow from the uterus, patients with cervical agenesis typically appear with amenorrhea and cyclic pelvic discomfort in early adolescence, around the time of menarche. Preventing significant endometriosis requires early identification and treatment since it can cause irreparable harm to reproductive potential and requiring major surgery like a hysterectomy or adnexectomy [3]. Two cases of cervicovaginal agenesis reported with the normal uterus, which managed them with a conservative surgical approach. They created neovagina and connected it to the uterine cavity, but after six months, there was no passage between the uterus and neovagina. Despite recurrent reconstructive surgeries, the patient presented with pelvic infection and sepsis, for which the hysterectomy was done [4]. Considering these harmful consequences of uterine conserving surgery, in the case of cervicovaginal agenesis, there are some important issues for both the patient and surgeon before choosing the best

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surgical decision; first, the patient and her parents should be completely informed about regular follow-up visits, appropriate hygiene, regular use of vaginal dilators after surgery. Second, in conservative surgery after the creation of the neovagina in the case of cervical agenesis or cervical malformation, insertion of the stent between the lower uterine segment and neovagina may prevent cervical stenosis [5]. This work has been reported with respect to the SCARE 2023 criteria [6]. Ethical approval was obtained under reference number 134/GJMS/JC.

Cervicovaginal agenesis is a rare Müllerian anomaly that often presents with primary amenorrhea and cyclic pelvic pain, leading to delayed diagnosis and mismanagement, particularly in resource-limited settings. Due to its low incidence and variable clinical presentation, there is limited standardized guidance regarding optimal timing, surgical technique, and long-term outcomes of conservative management. Most available literature consists of isolated case reports, with insufficient long-term follow-up data on fertility preservation and postoperative complications. Therefore, this case report aims to highlight early diagnostic considerations, describe a successful conservative surgical approach, and contribute to the existing evidence on functional and anatomical outcomes in cervicovaginal agenesis.

Case Presentation

A 14-year-old young girl with primary amenorrhea and severe cyclical pelvic pain for 5 months presented to the Outpatient department at Women and Children hospital Dera ismail khan, she took multiple treatments for abdominal pain at private setup but was not resolved. she was referred for gynaecological examination. On physical examination, her breast development was tanner stage 2, axillary and pubic hairs stage 3. She had no known history of medications, allergies, adverse reactions or her family disease. Transabdominal ultrasonography revealed a fluid collection of 172ccin intrauterine cavity. These findings were consistent with hematometra. The patient was admitted to hospital for examination under anaesthesia, external genitalia was normal, a blind-ending vagina 1.5 cm from the introitus. Dissection was given in loose areolar tissue between urethra and rectum, there was no connection between uterus and vagina, the cervical opening was not visualized. So vaginoplasty with abdominopelvic approach was planned. laparotomy followed by hysterotomy was done. Hegar dilator 7 was introduced from above to create a space followed by the insertion of foley's catheter to guide the cervical opening, inflated and secured from below. A Mold was kept in the space created to prevent from stenosis. Which was changed after 72 hrs. Her post-operative period went unremarkable. She had no pain or discomfort. She was stable and discharged home on postoperative day five. The patient successfully underwent cervicovaginoplasty through a combined abdominopelvic approach leading to the creation of a neovagina and continuity was established between the uterine cavity and the neovaginal tract using a Foley catheter as a temporary stent to maintain patency. The immediate postoperative period was uneventful. The patient remained stable, experienced no pain or complications, and was discharged on the fifth postoperative day. Vaginal molds were introduced postoperatively to maintain the tract and prevent restenosis. At her first follow-up one month after surgery, the Foley catheter had been removed, and she reported spontaneous menstruation 15 days' post-discharge. Clinical examination revealed a neovaginal length of 5–6 cm, with a palpable cervical dimple and adequate epithelialization at the neocervical junction. Ultrasound confirmed absence of hematometra and free flow of menstrual blood. The patient was followed up for six months. She continued mold use consistently and was monitored regularly over a six-month follow-up period. Menstrual cycles remained regular with no recurrence of pelvic pain or signs of vaginal stenosis. Vaginal patency and cervical continuity were maintained, and no further intervention was required. No complications such as restenosis, infection, or sexual dysfunction (although the patient had not yet initiated sexual activity) were reported. The patient and her guardians were counselled on the importance of continued annual follow-up to monitor vaginal and cervical status and future fertility potential. The experience and recommendation while dealing such case are, the patient should continue using vaginal molds for at least 3–6 months postoperatively to maintain the vaginal tract and prevent stenosis. She should be advised to monitor her menstrual flow and report any symptoms of dysmenorrhea, reduced flow, or amenorrhea, which may indicate re-stenosis. Every 3–6 months follow ups for the first year and then annually to assess: Vaginal Caliber and epithelialization status. Cervical patency using ultrasound (to ensure no hematometra formation). Hormonal Evaluation: Breast and pubic hair development at Tanner stage 2-3 suggests possible incomplete puberty or hormonal imbalance. Serum FSH, LH, estradiol, and AMH may be checked to assess ovarian function.

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DISCUSSION

Because of the variation in presentation, the therapy strategy for cases of hematometra caused by cervical atresia or agenesis is debatable Compared to patients with full agenesis, those with fibrous cord/fragmentation may have a greater chance of repair [1]. This condition often manifests in adolescent girls with a 46, XX karyotype, normal ovarian function, normal developed secondary sexual characteristics, and primary amenorrhea [2]. It is crucial to comprehend and define the precise anatomy of the Mullerian abnormality before considering genital tract reconstruction or surgical repair. Examining the current instance revealed an outflow restriction at the cervix level conservative surgical methods [4]. Since a stent was unavailable, a self-made Mold was used to put between the lower uterine segment and the neovagina after the neovagina was created in cases of cervical agenesis or cervical malformation. This may help avoid cervical stenosis. It's unclear how long the stent should be left in place, although some research suggests leaving it in place for at least six months to allow the catheter's surrounding tissue to fully epithelialize [5]. To keep the newly formed vaginal canal open and stop the tissues from settling inward, vaginal molds are helpful. They also guarantee that the vaginal canal is the proper size and form and aid in its shaping. According to studies, individuals who utilize vaginal molds following surgery had a greater success rate for vaginal dilatation and a lower risk of restenosis. It is crucial to remember that vaginal molds are not always required, and the choice to use them should be evaluated individually. The extent of the cervical agenesis, the size of the newly formed vaginal canal, and the patient's adherence to the mold-wearing protocol are all factors the surgeon should consider while doing surgery on this patient [7]. One surgical strategy that presents several difficulties is the restoration of utero-cervix/neocervix-vagina/neovagina continuity in individuals with cervical deformity. After menarche, and preferably after starting a sexual relationship, is the optimal age. Making a neovagina is a rare procedure that calls for surgical expertise and a multidisciplinary team. Rebuilding a neocervix is an even more difficult and uncommon procedure [8]. It is clear that using two approaches at the same time comes with challenges and possible consequences. Some writers contend that in order to prevent problems such ascending infection and the necessity for reintervention because of stenosis, the first line of treatment should be traditional care (hysterectomy) [9]. However, taking into account the preservation of fertility, the new surgical methods might be employed as the first resort [10]. There are significant risks of recurrence and complications associated with early uterovaginal anastomosis operations that involve cervical drilling or catheterization. Numerous papers report that uterovaginal anastomosis by laparotomy or laparoscopy has been effective in cases with cervix agenesis [11]. Regretfully, if conservative measures are unsuccessful, a hysterectomy may be required. Even in the ill-reputed types of congenital cervical agenesis, cervicoplasty with mucosal lining allows for the development of a patent cervical canal [8]. When doing vaginoplasty in situations of congenital cervical and vaginal agenesis, the abdominopelvic approach necessitates meticulous planning and risk assessment. The creation of a vaginal canal that is sufficiently long and wide, the preservation of the urethral and rectal sphincters, the maintenance of bladder and bowel function, the reduction of infection and hematoma formation risk, and the achievement of a cosmetically acceptable result are all important surgical factors. Hematoma development, vaginal stenosis, infection, persistent pelvic discomfort, urethral and rectal fistulas, and sexual dysfunction are possible side effects. Before beginning, the surgeon should thoroughly examine the patient's anatomy and medical history and have expertise doing this kind of surgery [12]. Congenital anomalies of the female reproductive tract, particularly cervicovaginal agenesis, pose a significant diagnostic and surgical challenge due to their rarity and variable clinical presentation. The complexity of such malformations often leads to delayed recognition, particularly in settings where adolescent menstrual irregularities are overlooked or misinterpreted. Acién and Acién provide a comprehensive overview of complex genital tract anomalies, emphasizing that early diagnosis is critical for planning optimal surgical intervention and for psychological well-being [13]. However, as seen in many cases, including the one discussed in this report, diagnosis is frequently delayed until adolescence or early adulthood when symptoms such as primary amenorrhea and cyclic pelvic pain manifest. Carranza-Mamane *et al.* stress the importance of a systematic approach to the diagnosis of congenital reproductive anomalies, recommending the use of imaging modalities such as MRI for precise anatomical delineation [14]. This is particularly relevant in cervicovaginal agenesis, where detailed imaging aids in differentiating among types of Müllerian anomalies and informs surgical planning. Leitao *et al.* describe a successful uterovaginal anastomosis for cervicovaginal agenesis, reinforcing that timely intervention using appropriate surgical techniques can result in restoration of menstrual function and preservation of reproductive potential [15]. Similar approaches have been supported by Folch *et al.* who categorize Müllerian agenesis and its subtypes, emphasizing individualized treatment strategies depending on anatomical and functional considerations [16]. In resource-limited settings, Adeyemi *et al.* highlight the utility of non-surgical approaches such as vaginal

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dilation in selected cases, particularly for vaginal agenesis. While this may not be applicable in cases with a functional uterus and cervical agenesis, it underscores the need for context-specific treatment modalities and counselling [17]. Liu *et al.* document the use of modified laparoscopic uterovaginal anastomosis techniques in cervical agenesis, demonstrating the advancements in minimally invasive procedures that improve patient recovery and reduce long-term complications [18]. Their report supports the growing body of literature advocating for laparoscopy as the preferred approach in experienced centers. Dreisler *et al.* explore long-term outcomes of vaginal reconstruction using bowel segments in cases where conventional techniques are unfeasible. While this technique is more commonly used in vaginal agenesis without a functional uterus, it serves as an alternative in complex cases requiring neovaginal construction, with satisfactory functional outcomes [19]. Mungan *et al.* emphasize the importance of long-term follow-up in patients undergoing surgical correction of vaginal agenesis. Complications such as stenosis, fistula formation, and sexual dysfunction may arise if postoperative care and patient compliance with dilation protocols are inadequate [20].

This study is limited by its single-case design and relatively short follow-up duration of six months, which restricts the assessment of long-term reproductive outcomes, fertility potential, and risk of restenosis. Additionally, advanced imaging modalities such as MRI were not utilized for detailed preoperative anatomical mapping. Future studies should focus on multicenter case series with longer follow-up periods to evaluate reproductive outcomes, quality of life, and surgical success rates. The development of standardized management protocols and long-term registries for congenital cervical anomalies would further strengthen clinical decision-making and improve patient outcomes.

CONCLUSIONS

The underlying causes of primary amenorrhea should be carefully assessed in patients. Cervicovaginal agenesis is a rare condition with a range of manifestations, despite vaginal anomalies like TVS. Misdiagnosis and treatment delays might result from this. For these surgically treatable lesions to maintain normal physiology and fertility and avoid consequences, early identification and appropriate care are crucial. For teenagers in particular, psychosocial support is a critical component of treatment. Two well-known side effects of the illness include infertility and restenosis.

Authors' Contribution

Conceptualization: NA, MA, RA
Methodology: NA, MA, RA
Formal analysis: MO, MA, RA
Writing, and Drafting: NA, MO, AA
Review and Editing: NA, MO, MA, RA, AA

All authors approved the final manuscript and take responsibilities the integrity of the work.

Conflicts of Interest

The authors declare no conflict of interest.

Source of Funding

The author received no financial support for the research, authorship and/or publication of this article.

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