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SEQUENTIAL SURGICAL MANAGEMENT OF VAGINAL ATRESIA AND UTERINE DIDELPHYS PRESENTING AS PRIMARY AMENORRHEA WITH HEMATOMETRA AND HEMATOCOLPOS

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ABSTRACT

Background:Distal vaginal atresia associated with uterus didelphys is an exceptionally rare congenital Müllerian anomaly that may present during adolescence with primary amenorrhea and cyclic pelvic pain due to obstructed menstrual outflow. Delayed diagnosis can result in hematocolpos, hematometra, endometriosis, pelvic infection, and compromised reproductive outcomes. Early recognition and appropriate surgical management are essential to restore normal genital tract function and preserve fertility.**Case Presentation:**A 13-year-old girl presented with severe cyclical lower abdominal pain for six months and primary amenorrhea despite normal secondary sexual characteristics (Tanner stage IV). Local examination revealed normally developed external genitalia with absence of a visible vaginal opening and a small hymenal dimple. Per-rectal examination demonstrated a tense vaginal bulge. Pelvic ultrasonography revealed uterus didelphys associated with hematocolpos and bilateral hematometra secondary to distal vaginal atresia. The patient underwent staged surgical management consisting of examination under anaesthesia with vaginal exploration and drainage of retained menstrual blood, followed by serial vaginal dilatation. Definitive reconstruction was performed using hysteroscopic vaginal septum resection with cannulation of both uterine horns under ultrasonographic guidance. The postoperative course was uneventful, with complete resolution of cyclical pain, restoration of normal menstruation, and satisfactory anatomical and functional outcomes.**Conclusion:**This case emphasizes the importance of considering obstructive Müllerian anomalies in adolescents presenting with primary amenorrhea and cyclic pelvic pain. Careful clinical evaluation supported by pelvic imaging allows timely diagnosis, while a staged, individualized surgical approach can successfully relieve obstruction, restore menstrual function, and preserve future reproductive potential. Reporting such rare cases adds valuable evidence to the limited literature and may help guide the management of similar complex congenital genital tract anomalies.

INTRODUCTION

Distal vaginal atresia is a rare congenital anomaly of the female reproductive tract resulting from defective development and canalization of the distal vagina during embryogenesis. Normal vaginal development occurs between the 11th and 20th weeks of gestation through coordinated interaction of the Müllerian ducts, Müllerian tubercle, and sinovaginal bulbs. Failure of this process leads to partial or complete obstruction of the distal vaginal canal. Although the exact pathogenesis remains incompletely understood, developmental abnormalities involving genes such as TBX3 and AXL have been implicated, and the condition may be associated with renal and vertebral malformations.[1,2]

Unlike complete vaginal agenesis, such as Mayer–Rokitansky–Küster–Hauser (MRKH) syndrome, patients with distal vaginal atresia have a normally developed uterus, cervix, ovaries, and proximal vagina. The defect is confined to the distal vaginal segment, where a fibrous or membranous obstruction prevents communication between the upper genital tract and the vaginal introitus, resulting in obstruction of menstrual outflow despite otherwise normal reproductive anatomy.[2,3]

Distal vaginal atresia represents an exceptionally uncommon subset of Müllerian duct anomalies. Congenital Müllerian anomalies occur in approximately 0.1–0.5% of females, whereas distal vaginal atresia accounts for only a small proportion of these abnormalities, with an estimated incidence of approximately 1 in 10,000–15,000 female births. Because of its rarity, published clinical experience is limited, and standardized management guidelines are lacking.[3,4]

Clinical manifestations typically become evident during adolescence at the expected age of menarche. The most common presentation includes primary amenorrhea associated with cyclical lower abdominal or pelvic pain due to progressive retention of menstrual blood proximal to the obstruction, resulting in hematocolpos, hematometra, and occasionally hematosalpinx. Delayed diagnosis may lead to complications such as endometriosis, pelvic adhesions, infection, infertility, and chronic pelvic pain.[3,4,5]

Diagnosis requires a high index of clinical suspicion and is established through careful physical examination supported by imaging studies. Pelvic ultrasonography is usually the first-line investigation, whereas magnetic resonance imaging (MRI) provides superior delineation of the level of obstruction, associated Müllerian anomalies, and surrounding pelvic anatomy, thereby facilitating surgical planning.[1,5]

Surgical reconstruction remains the definitive treatment and aims to establish a functional vaginal canal while preserving menstrual, reproductive, and future sexual function. Early diagnosis followed by individualized surgical management is essential to relieve symptoms, prevent long-term

complications, and optimize reproductive outcomes. We report a rare case of distal vaginal atresia associated with uterus didelphys presenting as primary amenorrhea with hematocolpos and hematometra, successfully managed by a staged surgical approach.

CASE REPORT

A 13-year-old premenarchal girl presented to the Department of Obstetrics and Gynaecology with complaints of severe cyclical lower abdominal pain for the preceding six months. The pain occurred at approximately monthly intervals, lasted for 3–5 days, progressively increased in intensity, and was associated with abdominal fullness. She had not attained menarche despite normal pubertal development. There was no history of vaginal bleeding, abnormal vaginal discharge, urinary retention, constipation, fever, or weight loss.

Her past medical and surgical history was unremarkable. There was no history of hormonal therapy, chronic systemic illness, previous pelvic surgery, trauma, or congenital anomalies. The family history was non-contributory, with no history of delayed puberty, primary amenorrhea, or congenital genital tract abnormalities.

On general physical examination, the patient was conscious, cooperative, and hemodynamically stable. Her height and weight were appropriate for age. Secondary sexual characteristics were normally developed, corresponding to Tanner stage IV for both breast and pubic hair development, indicating normal ovarian endocrine function. Systemic examination, including cardiovascular, respiratory, abdominal, and neurological examinations, revealed no abnormalities.

Local genital examination demonstrated normally developed external female genitalia with normal labia majora, labia minora, clitoris, and urethral meatus. However, no vaginal opening was identified, and only a small hymenal dimple was present at the expected site of the vaginal introitus. A per-rectal examination revealed a tense, cystic vaginal bulge approximately 2–3 cm proximal to the fourchette, suggestive of distal vaginal obstruction.

Routine laboratory investigations, including complete blood count, renal function tests, liver function tests, serum electrolytes, coagulation profile, and urinalysis, were within normal limits. Hormonal evaluation was not considered necessary because the patient had normal secondary sexual development and imaging suggested an obstructive genital tract anomaly.

Pelvic ultrasonography demonstrated uterus didelphys with two separate uterine cavities, both distended with echogenic fluid consistent with hematometra. A markedly dilated vaginal cavity containing retained blood (hematocolpos) was also identified, confirming obstruction of the distal vaginal outflow tract. Bilateral ovaries appeared normal in size and morphology, with preserved follicular activity. No renal abnormalities were detected on ultrasonographic evaluation. After detailed counselling and obtaining informed written consent from the patient's parents, the patient was taken to the operating theatre for examination under anaesthesia. Intraoperative assessment confirmed distal vaginal atresia with complete obstruction of the lower vaginal segment. Vaginal exploration was performed, and the obstructed segment was carefully opened, allowing evacuation of accumulated altered menstrual blood from the vagina and uterine cavities. Complete drainage of the hematocolpos and hematometra resulted in immediate decompression of the genital tract.

Following adequate healing, serial vaginal dilatation was undertaken to maintain vaginal patency and prevent restenosis. Subsequently, definitive reconstructive surgery was planned. Diagnostic hysteroscopy demonstrated two separate uterine cavities consistent with uterus didelphys. Under continuous ultrasonographic guidance, hysteroscopic vaginal septum resection was performed, and both uterine horns were successfully cannulated to establish unobstructed communication between the uterine cavities and the reconstructed vaginal canal. Hemostasis was achieved, and no intraoperative complications occurred.

The postoperative recovery was uneventful. The patient received prophylactic antibiotics, analgesics, and instructions regarding postoperative vaginal care and scheduled vaginal dilatation. At follow-up visits, the reconstructed vaginal canal remained patent without evidence of restenosis. She subsequently experienced spontaneous menstruation with normal menstrual flow and complete resolution of cyclical pelvic pain. No postoperative complications such as infection, recurrent obstruction, or urinary symptoms were observed.

The patient and her parents received comprehensive psychological counselling regarding the congenital nature of the anomaly, menstrual hygiene, reproductive health, future fertility, and the need for long-term gynecological follow-up because of the associated uterus didelphys. At the latest follow-up, the patient remained asymptomatic with satisfactory menstrual function and an improved quality of life.

DISCUSSION

Distal vaginal atresia is an exceptionally rare obstructive Müllerian anomaly that typically presents during adolescence with primary amenorrhea and cyclical pelvic pain secondary to hematocolpos and hematometra. The association of distal vaginal atresia with uterus didelphys is exceedingly uncommon, with only isolated cases reported in the literature. Owing to its rarity, there are no standardized treatment guidelines, and management is largely individualized according to the patient's anatomy and associated anomalies [1,2].

Our patient presented with the classical manifestations of obstructive genital tract anomalies, including primary amenorrhea, progressively worsening cyclical lower abdominal pain, normal secondary sexual characteristics, and a blind vaginal pouch. Similar clinical findings have been reported by Smorchkova et al., who described a patient with uterus didelphys and vaginal atresia presenting with hematocolpos and cyclic pelvic pain requiring surgical reconstruction [1]. Likewise, Jain and Mutyapwar reported adolescents with distal vaginal obstruction presenting with hematocolpos and hematometra secondary to retained menstrual blood [2]. In agreement with these reports, our patient demonstrated normal pubertal development, indicating preserved ovarian endocrine function and confirming that the underlying pathology was an obstructive outflow tract anomaly rather than an endocrine cause of primary amenorrhea.

Embryologically, distal vaginal atresia results from failure of canalization of the sinovaginal bulbs, whereas uterus didelphys arises from complete failure of fusion of the paired Müllerian ducts during embryogenesis [3,4]. The coexistence of these two anomalies is extremely unusual because they originate from defects occurring at different stages of reproductive tract development. Recognition of this embryological association is important because complex Müllerian anomalies require meticulous preoperative evaluation and individualized surgical planning.

Imaging played a central role in establishing the diagnosis in our patient. Pelvic ultrasonography demonstrated uterus didelphys with bilateral hematometra and hematocolpos,

thereby confirming genital tract obstruction. Previous studies have similarly emphasized that pelvic ultrasonography is the first-line imaging modality, while magnetic resonance imaging (MRI) provides superior anatomical characterization in complex Müllerian anomalies and assists operative planning [3,5]. Although MRI is considered the imaging modality of choice in complicated cases, ultrasonography in conjunction with detailed clinical examination provided sufficient anatomical information to guide successful staged surgical management in our patient.

The differential diagnosis of primary amenorrhea associated with cyclic pelvic pain includes imperforate hymen, transverse vaginal septum, cervical atresia, vaginal agenesis, and Mayer–Rokitansky–Küster–Hauser (MRKH) syndrome. Unlike MRKH syndrome, in which the uterus is absent or rudimentary, our patient had two well-developed uterine cavities with functioning endometrium, resulting in progressive accumulation of menstrual blood. Careful differentiation among these conditions is essential because surgical management, reproductive prognosis, and long-term follow-up differ considerably [3,4].

The principal objective of treatment is restoration of unobstructed menstrual flow while preserving future reproductive and sexual function. Various reconstructive procedures have been described, including vaginal pull-through procedures, vaginal reconstruction using skin or bowel grafts, V-Y advancement flaps, Z-plasty, resection of obstructing vaginal septa, and hysteroscopic canalization [2,6]. In contrast to several previously published reports in which definitive reconstruction was performed as a single-stage procedure, our patient underwent sequential surgical management, beginning with examination under anaesthesia, vaginal exploration, and decompression of hematocolpos and hematometra, followed by serial vaginal dilatation and definitive hysteroscopic vaginal septum resection with cannulation of both uterine horns under ultrasonographic guidance. This staged approach minimized tissue trauma, facilitated accurate identification of both uterine cavities, and reduced the likelihood of restenosis while restoring normal genital tract continuity.

Another important feature of our case was the use of combined hysteroscopic and ultrasonographic guidance during definitive reconstruction. This technique has been described in only a limited number of complex Müllerian anomalies and has been shown to reduce the risk of uterine perforation, false passage formation, and injury to adjacent pelvic structures [6,7]. In our patient, ultrasonographic guidance enabled precise cannulation of both uterine horns and successful restoration of menstrual outflow without intraoperative complications.

The postoperative outcome was excellent, with complete resolution of cyclical pelvic pain, restoration of spontaneous menstruation, and no evidence of restenosis during follow-up. Similar favorable outcomes have been reported when early diagnosis and timely surgical intervention were undertaken before the development of irreversible complications such as endometriosis or dense pelvic adhesions [1,5,7]. Nevertheless, patients with uterus didelphys remain at increased risk of infertility, recurrent miscarriage, preterm delivery, fetal malpresentation, and cesarean section; therefore, long-term gynecological surveillance and reproductive counselling remain essential [4,7].

The present case contributes to the limited literature describing distal vaginal atresia associated with uterus didelphys. Its novelty lies in the successful use of a staged, minimally invasive surgical strategy incorporating hysteroscopic and ultrasonographic guidance to restore menstrual function while preserving reproductive potential. Reporting such rare cases may improve awareness among clinicians and help refine the management of complex obstructive Müllerian anomalies.

CONCLUSION

Distal vaginal atresia associated with uterus didelphys is an exceptionally rare congenital Müllerian anomaly and an important but often overlooked cause of primary amenorrhea in adolescents. This case highlights the importance of maintaining a high index of suspicion in young girls presenting with cyclical pelvic pain despite normal secondary sexual characteristics, as delayed diagnosis may result in hematocolpos, hematometra, endometriosis, infection, and impaired future fertility. Thorough clinical examination complemented by appropriate pelvic imaging enables accurate diagnosis and timely intervention. A staged surgical approach involving initial decompression followed by definitive reconstructive surgery can effectively restore genital tract continuity, establish normal menstrual outflow, relieve symptoms, and preserve reproductive potential. Given the rarity of this anomaly and the absence of standardized management protocols, this case contributes valuable clinical evidence supporting individualized multidisciplinary management and emphasizes the need for long-term gynecological follow-up to optimize reproductive and obstetric outcomes.

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