



## Congenital Cervical Agenesis- A Rare Case Report

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### Abstract

*Congenital cervical agenesis is a very uncommon condition. It usually coexists with congenital vaginal agenesis. Years back, many centers considered hysterectomy as the only treatment option. With the advent of newer techniques in reconstruction, most of these cases have achieved normal menstruation and few have had normal pregnancy as well. Herewith, we describe our experience in the management of primary amenorrhea in a 16 years old girl along with highlighting the unconventional uterovaginal anastomosis with V-M technique and thereby, we were able to achieve a successful outcome.*

**Keywords:** *Cervical agenesis/dysgenesis, congenital malformations, MRKH, obstructive Mullerian anomalies, vaginal agenesis.*

### Case Presentation

16 year old girl was evaluated at local hospital for complaints of cyclical lower abdomen pain for 6 months and absence of menstruation. MRI pelvis there showed small sized uterus of 5 x 3 x 3 cm, normal bilateral fallopian tubes and ovaries. Lower part of uterus and cervix was not visualised. Ectocervix of 0.5 cm, Collected menstrual blood in the peritoneal cavity. Vaginal length of 5.2 cm. Findings were confirmed by diagnostic laparoscopy. Peritoneum was cleared off blood. She was on hormonal suppression with oral contraceptive pills. She had consulted several local hospitals where she was advised hysterectomy as final option and hence she self referred to our institution.

On examination, her height was 145 cm and weight 41 kg and had normal secondary sexual characters (tanner stage 4 of breast and tanner 5 of pubic hair). Per rectal examination revealed same findings. Case was discussed with and seen by plastic surgeon and decided to proceed with examination under anaesthesia and plan on further management. She and her parents were counselled regarding the pros, cons and outcomes of surgical repair.

Per speculum examination under anaesthesia showed blind vaginal cavity with dimple at its apex. We proceeded with laparotomy through pfannenstiel incision. Intraoperative findings included: Uterus small in size, distal one third of uterus and cervix was absent. Both ovaries and

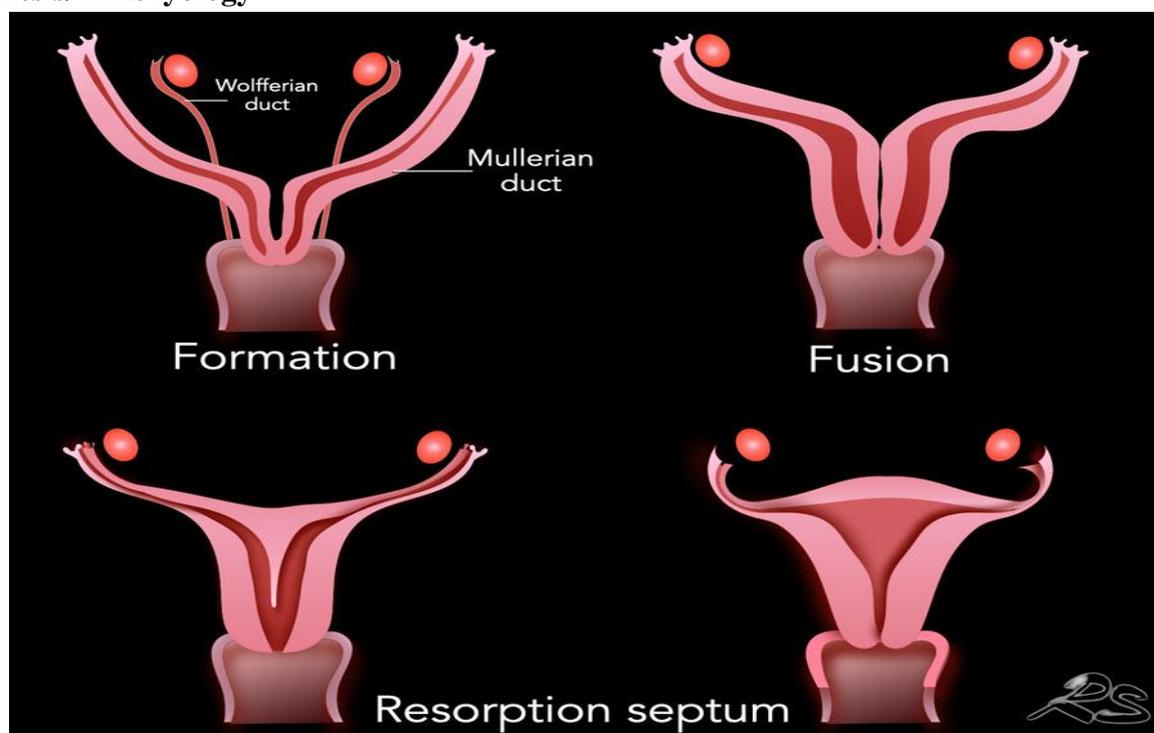
fallopian tubes were normal, we decided to proceed with uterovaginal anastomosis using V-M technique. Distal aspect of uterus was opened transversely till endometrial cavity was seen. Apex of vagina was pushed up with a probe and slit open and anastomosed with lower end of uterus which was further extended upwards in order to compensate for the size difference of uterus and cervix. Full thickness lower uterus to vaginal anastomosis was done with 5-0 pds. No. 8 foleys catheter was used to stent the reconstruction which was fixed to the fourchette to avoid displacement. Vaginal pack was kept. Post operatively, the girl was continued on antibiotics and analgesics, put on complete bed rest with low residue diet and continuous bladder drainage. Vaginal pack was removed after 24 hours and on post operative 6, splinting foleys catheter was removed. Post operative period was uneventful and hence she was discharged on post operative day 7. Hormone suppression was continued for another 3 weeks till complete healing was achieved. After we discontinued

hormonal suppression, her parents were elated to find that their daughter had started menstruating. Thereafter, she has had 5 regular menstrual cycles so far.

### Discussion

Around 200 cases of congenital cervical atresia have been reported in the literature so far. Mullerian anomalies are congenital anomalies with prevalence ranging between 4% to 6% of the general population. Cervical agenesis or dysgenesis has an incidence of about one in 80,000- 100,000 live births and in about 50% cases it coexists with congenital vaginal agenesis. Its prevalence accounts for 3% of all uterine malformations and for 0.1% of the overall population.. Mullerian anomalies appear to have a substantial impact on the reproductive potential and health of the affected women. The earliest reported case of congenital cervical atresia managed by hysterotomy and canalization was described by Ludwig in 1900.

### Pathogenesis/ Embryology



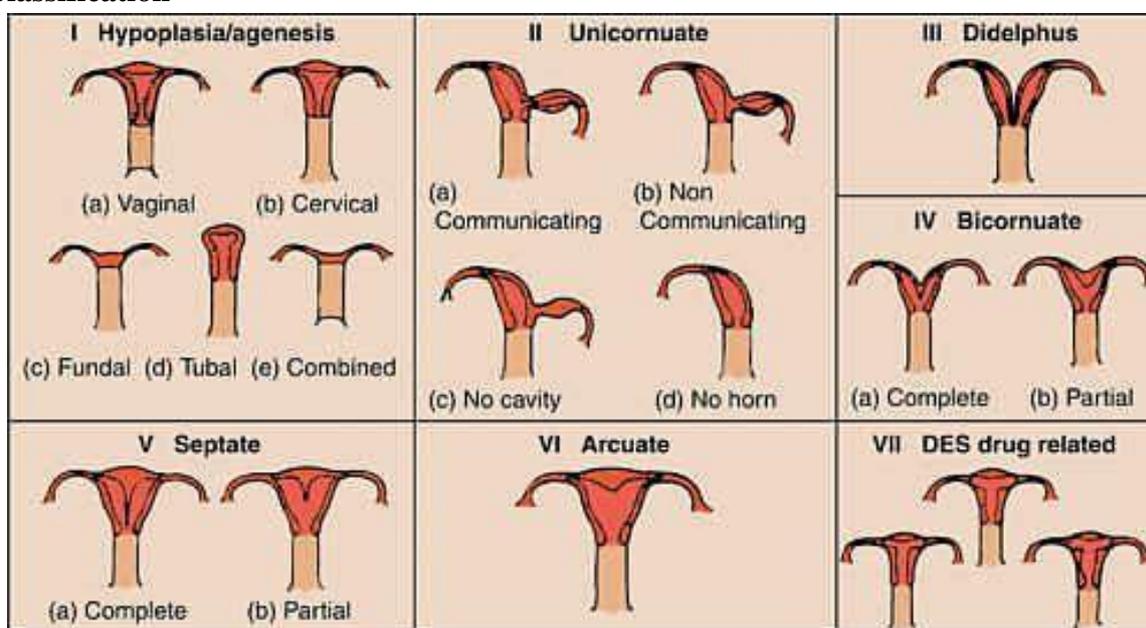
Each paramesonephric duct can be divided into three parts:

- (i) A cranial vertical division that develops into the fallopian tube
- (ii) A median horizontal division that develops into the broad ligament of the uterus
- (iii) A caudal vertical part that fuses to the midline with the contralateral paramesonephric duct constitute the uterus, the cervix, and the upper two-thirds of of the vagina.

Vagina and cervical canal are formed by vacuolization of the paramesonephric tissue. Any event that could upset the vacuolization process could be the cause of various anomalies.

The “atrophy hypothesis” (mechanism of local atrophy at the level of the “primitive) explains isolated cervical dysgenesis with normal vagina. The “ defective elongation hypothesis” (incomplete/partial caudal elongation defect of the merge of the mullerian ducts during the embryonic period) explains cervicovaginal aplasia.

**ASRM Classification**



Our case falls under type Ia according to ASRM classification

**Clinical Manifestations**

Any pubertal female with primary amenorrhoea and a history of cyclic abdominal pain should raise the clinical suspicion of cervical malformations (with or without a normal vagina). This can cause profound physical and psychological agony to the child and her parents. In cases of vaginal agenesis, coital intercourse is physically impossible.

**Diagnosis and Management**

Any pubertal female with history of primary amenorrhoea and cyclical abdominal pain should undergo physical examination as well as radiological imaging modalities like

- (a) Two-dimensional (2D) ultrasound( 67.3% sensitivity and 98.1% specificity)
- (b) Three-dimensional (3D) ultrasound (98.3% sensitivity and 99.4% specificity)
- (c) Transperineal 3D ultrasound
- (d) MRI

Renal anomalies should be looked for as they are commonly associated

The goals of management include:

- (1) Relief from the obstructive symptoms
- (2) Establishment of normal sexual function
- (3) Preservation of the uterus for future fertility

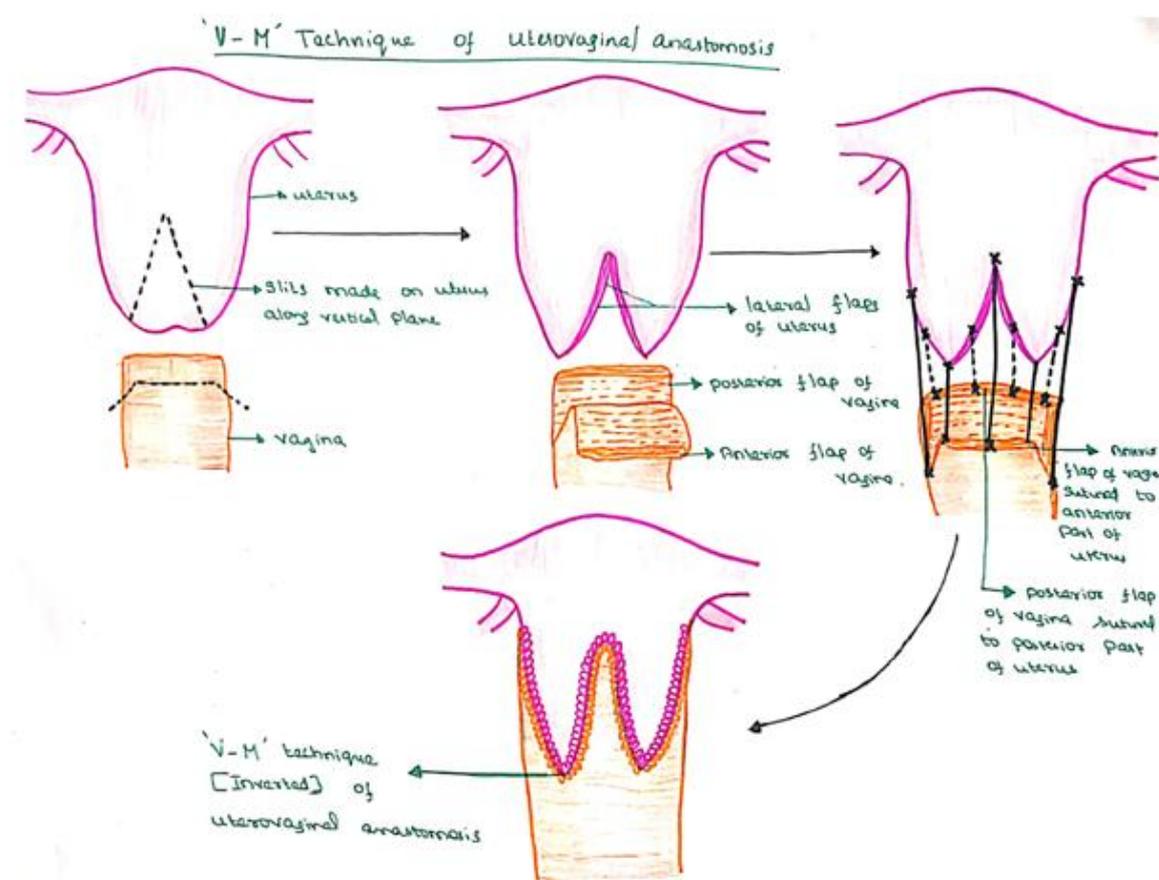
**Medical Management:** Used in the initial stage to alleviate pain and to prevent endometriosis.

Oral contraceptive pills or gonadotropin releasing hormone analogues with add back therapy are generally used.

**Surgical Management:** The aim of the surgery is to restore the continuity of the genital tract. This can be done by the traditional procedure of canalisation which is commonly used. However, the chance of stenosis is more compared to anastomosis procedure. Anastomosis is more time consuming and requires more skill and high learning curve. The degree of pelvic endometriosis and adhesions present needs to be considered in recommending a surgical approach to the problem and can be tackled during the time of surgery. In cases with absent vagina, Vaginoplasty is done with either graft or colon followed by anastomosis as a second step surgical procedure.

Due to extremely small size of the undeveloped distal uterus, it is important, it is important to avoid the risk of future stenosis in the anastomosis. For this, we have introduced an innovative technique to increase the area of anastomosis.

In the V- M technique, in the already transversely slit lower uterus, a longitudinal full thickness slit is made at the lower end. A similarly longitudinal slit is made on the proximal vaginal vault such that the planes of the slits are perpendicular to each other. Thereafter, the flaps are advanced so as to interdigitate the flaps for anastomosis with 5-0 pds in a V-M pattern thereby reducing the cross sectional area and hence reducing chance of stenosis. Along with this it is splinted with no. 8 foleys to stent and fixed to the vaginal fourchette with vicryl.



#### Complications of reconstructive surgeries:

Endometritis, pelvic inflammatory disease, persistent pelvic pain, bowel and bladder injury, reoperations, and even sepsis.

#### Pregnancy and Delivery:

There have been reported six successful pregnancies after restoration of the uterovaginal route of which 3 were spontaneous conception. The advent of

assisted reproductive technologies and gamete-zygote intrafallopian transfer techniques increases the likelihood of pregnancy in patients with congenital cervical atresia who have undergone successful canalization. There is increased risk of second trimester miscarriage and preterm labor. Abdominal encirclement will be necessary during pregnancy and mode of delivery is elective caesarean section.

**Role of Hysterectomy:** Hysterectomy should be avoided as first choice and reserved for cases where canalization attempts fail or in cases with severe endometriosis or pelvic inflammatory disease or if patient prefers to undergo hysterectomy. The patients who desire fertility preservation without reconstructive surgery may have the option of oocyte retrieval with gestational surrogacy.

### Conclusion

- (1) Surgical reconstruction is possible in these cases. Extirpative surgery may be preserved for surgical failures.
- (2) Uterovaginal anastomosis should be considered in difficult cases, especially those prone for stenosis.
- (3) Early surgery is essential for favourable results
- (4) Clinical judgement of the surgeon is most important.

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