

# Cervical Agenesis: Is Hysterectomy the Only Answer?

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**Abstract:** Cervical agenesis is an uncommon Mullerian anomaly with occurrence of 1 in 80, 000-1, 00, 000 births. Presentation is typically cyclic abdominal pain and primary amenorrhoea. Patients can be considered for canalisation surgeries but most of them eventually end up in hysterectomy due to recanalisation. We report here a case of cervical agenesis and the dilemmas in its management.

**Case Report:** 17 years old unmarried girl presented to our OPD with primary amenorrhoea and acute abdominal pain. MRI was suggestive of lower vaginal atresia, gross hematometra, hematocolpos and hematosalpinx. Examination revealed normal secondary sexual characters and a blind ending vagina. Dilemmas in diagnosis, options of management and comparison with other national and international studies will be discussed.

**Keywords:** hematometra, hematocolpos, agenesis

## 1. Introduction

Cervical agenesis is type 1B Mullerian anomaly according to American Fertility society. It results from abnormal fusion of Mullerian ducts with the urogenital sinus or atrophy segment of normally formed Mullerian system. It is an extremely rare congenital anomaly with an occurrence of about 1 in 80, 000 to 1, 00, 000. (1). It represents 3% of all uterine anomalies. (2) Patients can present with primary amenorrhoea, hematometra, hematocolpos, infertility, abortions, recurrent pregnancy loss and endometriosis. (3) The main objectives of treatment are symptom relief, regular menstruation and restoration of fertility.

## 2. Case Description

A 17 years old unmarried girl presented in the outpatient clinic at R. N. Cooper Hospital with primary amenorrhoea and acute abdominal pain. She had cyclic abdominal pain. Her bowel and bladder habits were normal. She had undergone diagnostic laparoscopy in the past. However, reports were unavailable. On examination, secondary sexual characters were present. On per abdominal examination, a firm, non-tender, globular swelling of 20 weeks size was palpable. On per rectal examination, a mass was felt in midline which was 20 weeks in size. Pelvic ultrasound revealed a grossly bulky uterus with echogenic fluid extending into cervical and vaginal canal. MRI showed lower vaginal atresia and stenosis of external os, gross hematometra, hematocolpos and hematosalpinx with leak and collection along lateral wall of cervix.

The patient was admitted and IV analgesics were given. The patient was counselled since it was a repeat surgery and acute abdomen made it a semi-emergency state. Examination under anaesthesia revealed a normal perineum. Vaginal dimple was present. Hymen was not visualised. There was no swelling or discoloration. On per rectal examination, a mass was felt in midline and rectal mucosa was intact.

After labial separation lower ¼ of vagina was patent. Some amount of vagina with epithelium was present. (Figure 1)



Figure 1

Vaginal dimple was caught with Allis forceps and dilated with Hegar's dilators upto no 8. Transverse incision was taken over vaginal dimple. A neovaginal space was created by blunt dissection. A tense collection identified between bladder and urethra above and rectum below. It was suctioned with a needle and syringe and minimal chocolate coloured fluid was aspirated.



Figure 2

However, communication with uterine cavity could not be identified. Hence, decision of exploratory laparotomy was taken. A Pfannenstiel incision was taken and In situ: a uterus of 20 weeks size and flimsy adhesions were found. There were endometriotic nodules on fallopian tubes. An incision was taken on isthmus and chocolate coloured fluid (150 cc) was drained. A cervico-vaginal tract was created by railroad technique. A thick red rubber catheter with multiple holes was kept in the uterine cavity for drainage. (Figure 3)



Figure 3

An Amnion graft and vaginal mould were used to keep the neo-vagina patent. A vaginal mould was prepared by expandable foam over red rubber catheter with 2 condoms and amnion graft was applied over it. This mould was then kept in the space created between bladder and rectum. (Figure 4)



Figure 4

Postoperatively, she received antibiotics, analgesics and intravenous fluids. She was kept nil by mouth for 24 hours. The drain output on day 1, 2, 3 was 50, 45 and 220cc respectively. The Foley's catheter was kept for 7 days. Uterovaginal catheter expelled spontaneously on day 6. The graft was taken up after 7 days.

The patient was counselled about the usage of glass dilators. She recovered well and was discharged on Day 8. On her follow up visit on day 20, the uterus could not be felt. 1

finger could be inserted and cervix was not felt. She is now menstruating monthly. After 3 months, 1 finger per vaginal examination was possible. Vaginal depth was about 2 and half inches.

### 3. Discussion

Obstructive uterine anomalies occlude the normal menstrual flow resulting in primary amenorrhoea. The main objectives of treatment are symptom relief, regular menstruation and restoration of fertility. Ultrasonography is the modality of choice to define internal genital anomaly and classify the level of obstruction or aplasia. (4, 5) Three dimensional ultrasound further improves the ability to accurately diagnose cervical anatomy though it requires operator expertise and adequate knowledge about the anatomical types. (6) MRI has been considered as a superior method for evaluating the vaginal and cervical anatomy but recently three dimensional ultrasound has gained importance. (7)

Reconstructive surgeries are used to create epithelialised uterovaginal canal. Controversies exist in the treatment options and uterovaginal anastomosis is considered as fine line treatment by some authors. (8, 9) To prevent closure of surgically formed canal, uterovaginal catheter stent is recommended. (10, 11, 12) Re-operations may be required due to restenosis of canal and eventually patients may end up with hysterectomy. Hysterectomy was the eventual treatment for cervical agenesis due to common complications of recanalization of the cervix and the unlikelihood of a viable pregnancy. Conservative surgical approach aiming at the restoration of anatomical integrity of female genital tract presents a permanent challenge.

In this patient, this conservative surgical approach was tried considering the simplicity of the technique and possibility of restoration of normal menstrual outflow tract and fertility preservation.

### 4. Conclusion

The procedure is a simple and effective method for creation of a menstrual outflow tract for cases of cervical agenesis with functioning uterus. However, the procedure needs further evaluation. Restenosis of the canal may occur eventually resulting in a hysterectomy.

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