

## Surgical Approach to a Case of Complete Cervical Atresia and Partial Vaginal Agenesis

**Umber Jalil Bakhtiar\***

Consultant OB\GYN, Pakistan

\*Corresponding Author: Umber Jalil Bakhtiar, Consultant OB\GYN, Pakistan.

Received: April 12, 2019; Published: July 09, 2019

### Abstract

**Objective:** To construct a utero-vaginal canal in a young 15 year old girl with complete cervical atresia, vaginal agenesis and the recto-vaginal fistula.

**Design:** Case Report.

**Setting:** Pakistan Railway Teaching Hospital.

**Patient:** A 15 Years old girl presented with lower abdominal pain and rectovaginal fistula from a previous vaginal surgery. Examination showed 2cm blind vaginal canal and ultrasound showed isolated haematometra.

**Procedure:** First stage surgery done to repair rectovaginal fistula. Second stage abdominoperineal surgery included drainage of haematometra and attaching cervical area to vaginal canal. Foly's (22Fr) catheter was used as a stent.

**Result:** Menstruation started after 2 months of removing the catheter.

**Keywords:** Cervical Atresia; Partial Vaginal Agenesis

### Introduction

Congenital atresia of cervix is a relatively rare mullerian duct development disorder which has been primarily reported as individual cases. In most cases the nature of uterine outflow obstruction in congenital cervical atresia has been confirmed intraoperatively in the past [1,2].

### Case Report

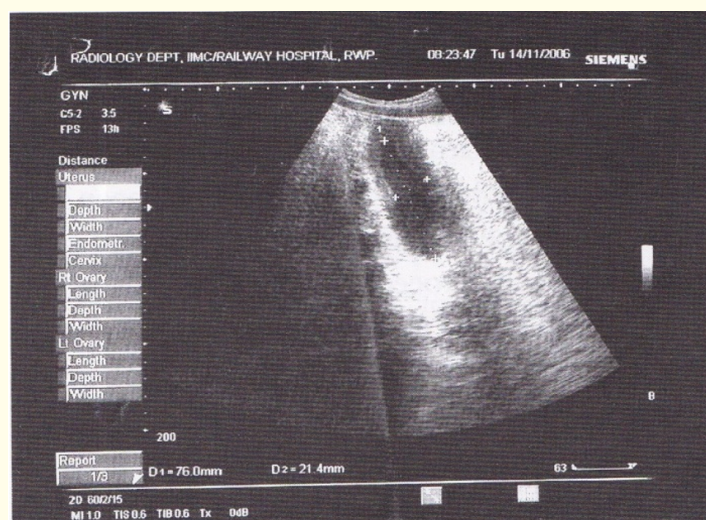
A 15 Year old girl presented in outpatient gynae With 2 years history of severe cyclic pelvic pain and primary amenorrhea. She was referred from a rural health centre with the history of pelvic surgery done by a general practitioner in order to treat the problem considering it hematocolpos.

The surgery did not result in drainage of any collected amount of blood rather the patient developed the problem of fecal incontinence after the surgery. After 1 week of this surgery she presented to us. On examination a 2 cm blind vaginal canal with thick vaginal bands was seen. On rectal examination a rectovaginal fistula 2 cm above the anus was seen and a mid-line 10 week size mass was palpated anteriorly which was mobile. Intravenous pyelography was done that showed normal kidneys and ureters. Pelvic ultrasound demonstrated normal ovaries, a haematometra measuring 9.0 x 8.0 cm and absence of vagina. All her baseline laboratory investigations were done and she was prepared for surgery. In the first stage the recto-vaginal fistula was repaired and blunt dissection of vaginal canal was done up to about 3 - 4 cm. Detailed counseling of her parents was done.

## Surgical Approach to a Case of Complete Cervical Atresia and Partial Vaginal Aggenesis

After one month of initial surgery a second surgery was done. This time the approach was abdomino-perineal. The blunt dissection of a vagina was done between rectum and urethra and the length of vaginal canal increased up to 4 - 5 cm. On opening the abdomen retrovesical pouch was opened, uterine mass measuring 8.0 x 8.0 cm was seen with blind lower bulb like end and right sided tube also was dilated. Both ovaries and left sided tube were all healthy. There were no endometriotic spots in the cavity and cavity was clean and healthy. Uterus was opened by making 1.5 cm midline vertical hysterotomy incision; 100cc of thick chocolate color fluid was drained. The fibrous edge of the incision was trimmed to make an ostium. This ostium was attached to the proximal end of vagina by the abdominal route. A foley catheter of 22 Fr was placed in the uterine bagged was attached with it. During the dissection of vagina small portion of posterior urethral wall was injured. It was repaired and another foley catheter was retained in the bladder. Patient was put on board Spectrum antibiotics. Her recovery was uneventful and she was discharged on the 5<sup>th</sup> day. After 2 weeks the bladder catheter was removed. The foley catheter from the uterine cavity was removed after four weeks. Patient was trained along with her mother to do vaginal dilatation with Hegars dilators starting with size 8. She was assessed after 15 days and she could pass hegars dilators no. 10. She was trained to use anal dilators later on. One month after surgery she had regular normal menses without lower abdominal pain. She was assessed after periods. She had normal flow. It has been 4 months now that she is having regular menses and comes every month for follow up.

She had decreased flow. It has been more than 10 years now she is having irregular menses and come every year for follow up.



## Discussion

Congenital atresia of cervix is a relatively rare mullerian duct development disorder which has been primarily reported as individual cases. In most cases the nature of uterine outflow obstruction in congenital cervical atresia has been confirmed intraoperatively in the past [1,2]. However modern imaging procedures such as magnetic resonance imaging and ultrasonography have recently been shown to be successful in accurate preoperative diagnosis of congenital cervical atresia [3-6]. In a large number of patients, in the literature endometriotic implants have been described at the time of laparoscopy or laparotomy.

Lower reproductive duct tract development in these patients is variable. Of the 50 cases in the literature in which vaginal anatomy has been described, 24 cases (48%) were associated with a normal vagina [3,4]. Twenty one cases of complete vaginal aggenesis and 5 cases of "shortened blind vaginal pouches" (partial vaginal aggenesis) have been described. The incident of renal anomalies associated with congenital cervical atresia is difficult to determine because many of the reported cases did not describe the result of intravenous pyelography.

Nonetheless, because four renal anomalies were described in 20 patients including left ureteral ectopia, it is recommended to have renal collecting duct system imaging (i.e. intravenous pyelography or renal ultrasonography) in all these patients.

Cervical atresia primarily results from defect in elongation of the mullerian ducts. Mullerian duct development would suggest that a partial (incomplete) elongation defect occurs in patients with congenital absence of the both the cervix and vagina. Congenital cervical atresia with normal vagina indicates normal mullerian elongation. In these cases, neither differentiation nor canalization of the cervix occurred. The differentiation of the uterine cervix is a complex, dynamic process. After elongation and fusion, cervical tissue differentiation ensues around the fifteenth week of gestation with thickening of the caudal aspect of the mullerian duct tissue, which is in contact with the urogenital sinus (mesodermal origin) and portions of the lateral woffian ducts [1,3]. Thus it is possible that the developmental aberrations that concern in congenital atresia of the cervix may involve both early (elongation) and late (canalization) stages of differentiation [4,5].

Majority of these patents have evidence of pelvic endometriosis or pelvic adhesive disease or both. The degree of pelvic endometriosis and adhesions present needs to be considered in recommending a surgical approach to the problem. To reduce cost and morbidity pelvic magnetic resonance imaging and diagnostic/laparoscopy to define pelvic disease may be considered as an alternative to exploratory laparotomy in providing diagnosis, as well as to give information that is pertinent to future management. The idea candidate for canalization would appear to be patient with minimal pelvic endometriosis and adhesions at the time of laparoscopy or laparotomy. It must be emphasized that patients whose chances of fertility would be markedly compromised by the presence of endometriosis, pelvic adhesions, salpingitis or advanced maternal age should be advised to consider primary hysterectomy rather than conservative management.

Many cases of congenital cervical atresia are associated with normal vaginal development (48%). When a canalization procedure is performed with our vaginoplasty, the chance of success as measured by cyclic menstrual function is about 70%. When a combined approach is necessary because of vaginal atresia the chance of surgical success resulting in normal cyclic menses is lower (about 40%). Furthermore, combined vaginoplasty canalization procedures appear to be associated with a higher risk of infection, including life threatening sepsis [2,5]. As such, the degree of vaginal atresia present should be considered in the management recommendations given to patients with congenital cervical atresia.

Medical suppression of ovarian function would allow additional time management decision making. Consideration should also be given to medical therapy as a means to retard the development of severe and progressive endometriosis, particularly for patients who are considering conservative management.

The likelihood of spontaneous pregnancy occurring as a result of canalization appears to be low. The lack of normal endocervical canal glandular function in patients who have undergone successful canalization may be a contributing factor to infertility. There are only 3 cases in literature that report spontaneous pregnancies occurring after canalization of completely or partially atretic cervix [3,7,8]. However, the advent of assisted reproductive technologies and gamete- zygote intrafallopian transfer techniques makes possible the greater likelihood of pregnancy in patients with congenital cervical atresia who have undergone successful canalization [5,9]. It is recommended that physicians should carefully document the anatomy of the patient and attempt canalization only in those cases in which it is likely to be successful.

In short, surgical canalization can be considered in selected patients with congenital cervical Atresia to achieve normal menstrual bleeding with resolution of symptoms. Clinical judgment on part of physician (considering severity of endometriosis and pelvic adhesions at the time of laparoscopy, degree of vaginal atresia and responsiveness to gonadotropic releasing hormone analogs) must be used in appropriate management of the patients with be used in appropriate management of the patients with this unique mullerian anomaly. Hysterectomy should always be offered as an option, given the risk of morbidity associated with conservative management. Finally, recent advancements in assisted reproductive terminologies may afford patients who successfully undergo conservation management a better opportunity to achieve pregnancy in future.

### Conclusion

Cervicovaginoplasty, attaching the endometrial mucosa to vaginal mucosa can help in creating a patent cervical canal with the help of a stent.

### Bibliography

1. Dillon W, *et al.* "Congenital atresia of the cervix". *Obstetrics and Gynaecology* 54.1 (1979): 126-129.
2. Geary W and Weed J. "Congenital atresia of the uterine cervix". *Obstetrics and Gynaecology* 42.2 (1973): 213-217.
3. Hampton B, *et al.* "Pregnancy after successful vaginoplasty and cervical stenting for partial atresia of the cervix". *Obstetrics and Gynaecology* 76 (1990): 900-901.
4. Markham S, *et al.* "Cervical agenesis combined with vaginal agenesis diagnosed by magnetic resonance imaging". *Fertility and Sterility* 48.1 (1987): 143-145.
5. Niver D, *et al.* "Congenital atresia of uterine cervix and vagina: three cases". *Fertility and Sterility* 33.1 (1980): 25-29.
6. Valdes C, *et al.* "Sonography in the surgical management of vaginal and cervical atresia". *Fertility and Sterility* 40.2 (1983): 263-265.
7. Zarou G, *et al.* "Pregnancy following the surgical correction of congenital atresia of the cervix". *International Journal of Gynecology and Obstetrics* 11.4 (1973): 143-146.
8. Fraser I. "Successful pregnancy in a patient with congenital partial cervical atresia". *Obstetrics and Gynaecology* 74 (1989): 443-445.
9. Solomons E. "Conception and delivery following construction of an artificial vagina: report of case". *Obstetrics and Gynaecology* 7.3 (1956): 329-331.

**Volume 8 Issue 8 August 2019**

**© All rights reserved by Umber Jalil Bakhtiar.**