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Successful Pregnancy Outcome Following Cervico-Vaginal Dysgenesis –A Case Report

Banu PJ[1*], Nasreen K[2], Ansary SA[3], Ishrat S[4], Deeba F[4], Begum N[4], Anwary S[5]

[1]Professor, Chairman, Department of Reproductive Endocrinology and Infertility BSMMU, Dhaka, Bangladesh
[2]Junior Consultant, OSD (DGH, Mohakhali), Attachment-BSMMU, Dhaka, Bangladesh
[3]Junior Consultant, OSD (DGH, Mhakhali) Attachment: BSMMU, Dhaka, Bangladesh
[4]Associate Professor, Department of Reproductive Endocrinology and Infertility BSMMU, Dhaka, Bangladesh
[5]Assistant Professor, Department of Reproductive Endocrinology and Infertility BSMMU, Dhaka, Bangladesh

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***Corresponding author:** Banu PJ

Abstract **Case Report**

Background: Congenital cervical dysgenesis with or without vaginal agenesis is a rare Müllerian developmental disorder that occurs in one in every 80,000 to 100,000 births. Cervical abnormalities two basic anatomical types. First, the cervix is absent with narrow lower uterine segment and absent vagina cervico vaginal agenesis. The second type is cervical dysgenesis. Management options for such conditions include total abdominal hysterectomy or conservative surgery with uterovaginal anastomosis, cervical canalization, or cervical reconstruction. In our case report, the patient had cervico vaginal dysgenesis. ***Objective:*** Aim is to successful reproductive out come after reconstructive surgery of cervicovaginal dysgenesis. ***Case report:*** Mrs. Rabeya came to BSMMU on 08/11/08 with primary amenorrhea with hematometra with vaginal agenesis at 15 yrs old. Reconstruction of cervix & vaginoplasty done & menstruation started. Upto 7 years after 1st operation her menstrual cycle was regular but scanty flows with severe dysmenorrhea. Above these complaint's she again came to BSMMU at 2015 and reconstruction surgery was done due to narrowness of cervico vaginal space at that time. After 2nd operation her menstrual flow was average & dysmenorrhea decreased. She got married at 2017. Four months after her marriage she developed severe dyspareunia & came to BSMMU & admitted on 10/08/17. Re-anastomosis of cervico vaginal stenosis and resection of septum done for 3rd time and cervico vaginal canal restored. Her dyspareunia was improved. Now after her 3rd time operation she enjoying her new life & spontaneous pregnancy occur. Finally LUCS was done at her 36wks pregnancy on 6th May 2019**.** ***Discussion:*** A total of approximately 200 cases of congenital cervical atresia have been reported in the literature. The experience from reconstructive uterovaginal anastomosis was not satisfactory. Rock *et al*., recently reviewed 30 cases of cervical agenesis and dysgenesis that underwent reconstructive surgery. Reobstruction was observed in all patients, of which 25 patients had hysterectomy and only 1 patient achieved spontaneous pregnancy. ***Results:*** In REI dept. of BSMMU we handled 14 cases of cervicovaginal dysgenesis. After successful reconstructive surgery 12 cases started regular menstrual cycle, one case needed hysterectomy and one patient achieved spontaneous pregnancy. ***Conclusion:*** Our case suggests that successful pregnancy can be achieved by reconstructive surgery for patients who have congenital cervico vaginal dysgenesis with functioning uterus.

Keywords: Meniscus abnormality, medial meniscus, double-layered meniscus.

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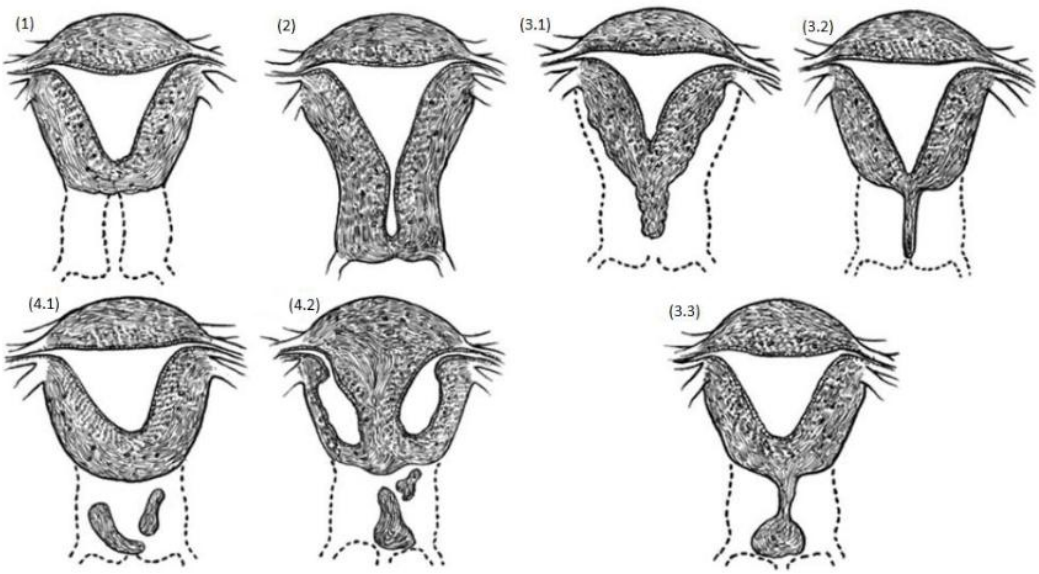
INTRODUCTION

Congenital cervical dysgenesis with or without vaginal agenesis is a rare Mullerian developmental disorder that occurs in one in every 80,000 to 100,000 births [1]. Cervical anomalies are among the rarest Mullerian duct anomalies which can be observed in the absence of the cervical canal or concomitantly with its obstruction. Although the prevalence counts only accounts for 3% of all uterine malformations and for 0.1% of the overall population [1], it is relevant because it generally affects women during their childbearing age, impairing fertility [2]. In the new ESHRE/ESGE classification, cervical anomaly is classified into 5 groups or classes: C0 (normal cervix), C1 (septate cervix) C2 (double normal cervix) C3 (unilateral cervical(aplasia) and C4 (cervical aplasia) [3]. In another type of classification [4],this condition is divided into 4 categories according to both the anatomical variants and the type of surgical treatment: type 1 cervical agenesis (CA)(characterized by the complete absence of the cervix and no endo cervical

canal); type 2 cervical dysgenesis (CD) 2(a)(characterized by cervical obstruction with the cervix being well formed but lacking an endocervical canal) type 2(b) cervical dysgenesis (characterized by cervical fibrous cord whose diameter as well as the stroma nature may vary) type 2(c) cervical dysgenesis (characterized by cervical fragmentation with separation of the segments) (Figure-1) [5]

Cervical agenesis has two basic anatomical types (1) Cervical agenesis type 1 and Cervical dysgenesis type 2 a,b,c .clinical presentation Primary amenorrhea with cyclical abdominal pain. Management options for such conditions include total abdominal hysterectomy or conservative surgery with utero vaginal anastomosis, cervical canalization, or cervical reconstruction to relieve symptoms and restore regular menstruation with fertility [4, 5]. However, the reconstructive surgery may be complicated by pelvic infection or restenosis of the neocanal, resulting in reoperation and hysterectomy [4, 5]. Further, the chance of a spontaneous pregnancy is small. An alternative option to restore fertility is by the use of assisted reproductive technology (ART). In our case report, the patient had cervical dysgenesis, associated with obliterated cervix to the uterine body by a fibrous (types 2 a) band. The patient achieved pregnancy after reconstruction of cervix with repeated surgery and vaginoplasty.

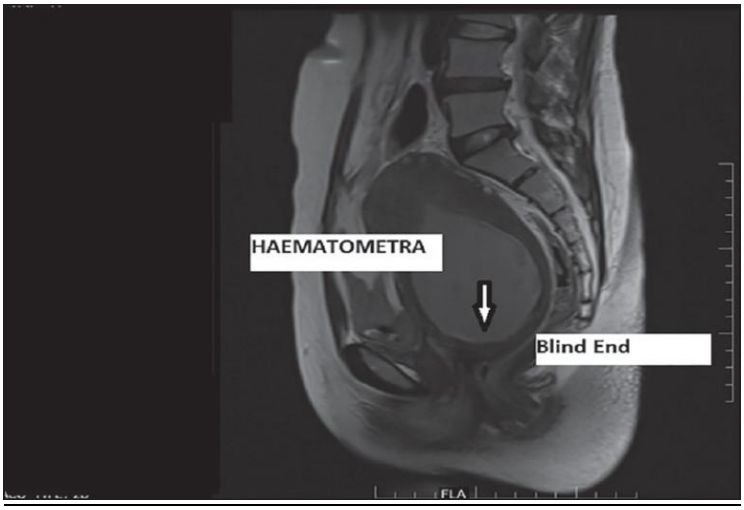


Legend: (1) Cervical agenesis type 1; (2) Cervical dysgenesis type 2; (3) Cervical dysgenesis type 3; (4) Cervical dysgenesis type

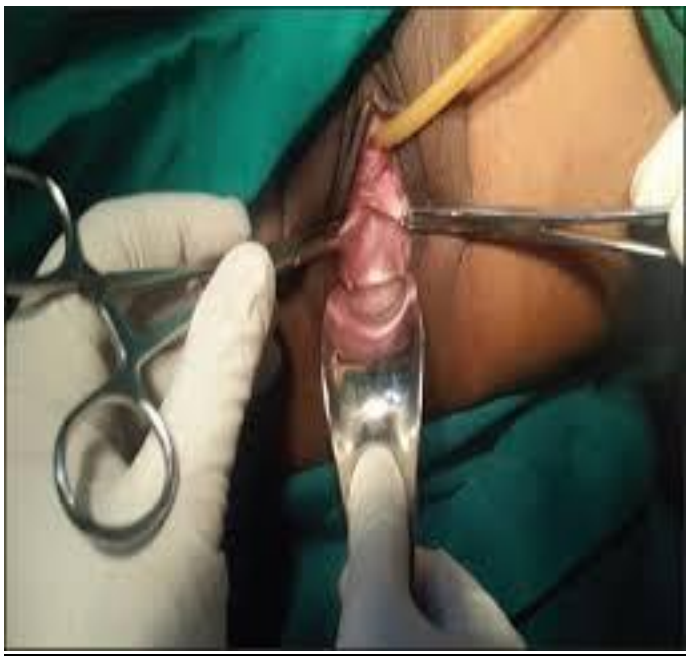
PATIENT AND OBSERVATION

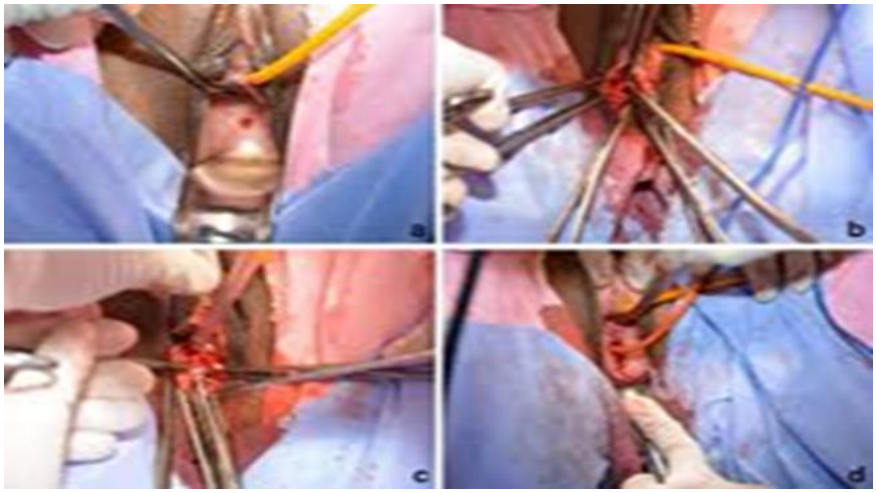
A 22 year old female presented to the infertility outpatient depertment BSMMU complaining primary amenorrhea with cyclic, monthly, lower abdominal pain on October 2015and. There was no history of uterus exposure to diethylstilbestrol or radiation. She had a past history that at 15 year of her age presented gynae outpatient department BSMMU with same complain on 2008. At that time 1st time reconstruction of cervix & vaginoplasty done & menstruation started. After operation her menstrual

cycle was regular but scanty flows with severe dysmenorrhoea with these complaint's she again came to BSMMU on October 2015 .On examination development of secondary sex characteristics was normal (Tanner staging 4 and 5 for breast and pubis).... As the patient was virgin cotton tip examinanation of vagina was done. Lower part of vagina present. A bimanual rectal examination was performed found bulky uterus. An ultrasound examination showed that uteri corpus with haematometra and both ovaries were normal.



A MRI scan sustained the presence of a uterus with haematometra. Serum hormonal levels were within normal limits. She diagnosed a case of primary amenorrheacervicovaginaldysgenesis.2nd time On 2015 again cervical dilatation with pull through operation (Folys catheter kept in situ for 2 weeks) with vaginoplasty followed by gradual vaginal dilatation for 3 month , dysmenorrhoea decreased.





surgical end-to-end anastomosis was decided upon and performed in the following steps: (i) surgical anatomical preparation of both cervical edges, (ii) sharp opening, patency check and dilation of the cervical canal distally, (iii) insertion of a 16F Foley catheter transvaginally through the distal cervical part and forward of the catheter into the endometrial cavity where it was anchored with 5 ml of fluid pumped into the catheter balloon, (iv) suturing of cervical edges to each other with the use of Vicryl absorbable suture No. 2.0, (v) suturing of anterior and posterior uterus vaginal vault. After completion of treatment her menstrual flow was average & dysmenorrhoea decreased. She got married 2yrs after her 2nd operation at her 24yrs age on 2017. Just after her marriage she developed severe dysperunia & again she came to BSMMU on 4 months of her marriage & admitted on 10/08/17 and diagnosed a case of cervical stenosis and iatrogenic transverse vaginal septum. On 3rd time resection of septum done and re-anastomosis of cervico vaginal stenosis and cervico vaginal canal resumed followed by silicone cather was kept in situ for 6 weeks. The length of the remaining cervix was 1.5cm dysperunia was improved & also dysmenorrhea decreased. The patient was discharged on the pill. After 6 weeks silicon Cather was removed. A month later she had normal menses. After her 3rd time operation she enjoing her new life & spontaneous pregnancy occur. These methods have included creation of neo vagina and reconstruction of cervix around catheter, which is both challenging and controversial. 1yr after 3rd operation spontaneous pregnancy occur on 05/11/18 at her 8 weeks pregnancy diagnosed by USG.

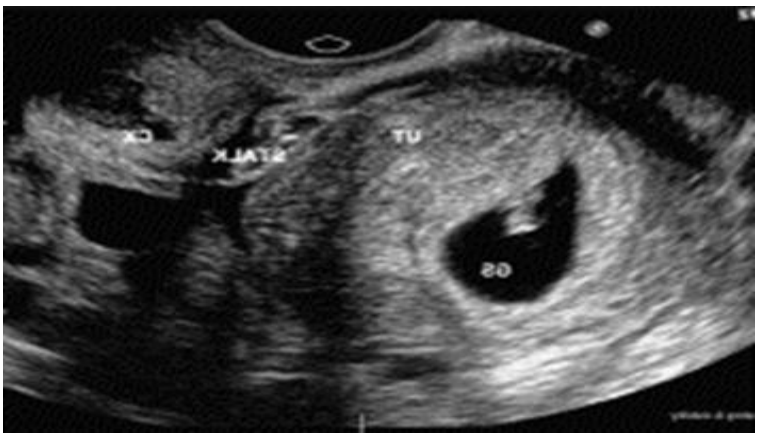


Fig-1: Ultra sonogram showing intrauterine pregnancy

Now she is continuing 30 weeks pregnancy. Her LMP was 01/09/2018, accordingly her EDD was 08/06/19.Her antenatal period was not uneventful. She was admitted in REI Department of BSMMU on 05/11/18 at her 8wks of pregnancy for abdominal pain and again admitted 03/02/2019 at her 22 wks of pregnancy due to fear of abortion. Then she was treated with inj. Gestone, microgest, with other medication and discharged at her 31 wks pregnancy. She went Barisal again with our medication. She again admitted with abdominal pain at her 33 wks pregnancy. As problem in vaginal part of cervix was less. Finally LUCS was done at her 36wks pregnancy at 6.5.19 and female baby was born, A Successful journey ended.

DISCUSSION

Pregnancy in patients with cervical dysgenesis is unattainable without either a reconstructive uterovaginal canalization or ART. A total of

approximately 200 cases of congenital cervical atresia have been reported in the literature [9]. However, there has been a lack of uniformity with regard to the ideal managements. The experience from reconstructive uterovaginal anastomosis was not satisfactory. Rock *et al*., [10] recently reviewed 30 cases of cervical agenesis and dysgenesis that underwent reconstructive surgery. Reobstruction was observed in all patients, of which 25 patients had hysterectomy and only 1 patient achieved spontaneous pregnancy [10] prematurely at 27 th weeks. The demolitive treatment consists in hysterectomy or hemi-hysterectomy. This possibility is generally reserved to patients with bicorporeal uterus [11] or to patients without pregnancy desire [11]. It is also employed in case of repeated failures of the conservative treatment or in case of complications after surgery (infections or cervical restenosis) [12]. The risk of recurrence is associated with fertility-sparing surgery. Hysterectomy should be recommended if canalization procedures fail or in the absence of pregnancy desire [12]. While waiting for larger case studies to have comparable results about surgical outcomes and fertility, In summary, our case suggests that successful pregnancy can be achieved by reconstructive surgery of cervix and Vaginoplasty for patients who have congenital cervicovaginal dysgenesis with functioning uterus.

To reduce the risk of stenosis it is possible to use [13] a biological mesh or a partial vaginal epithelium. Other surgeons have explored the use of a cellular porcine small intestinal submucosa graft for cervicovaginal reconstruction [15]. The demolitive treatment consists in hysterectomy or hemi-hysterectomy. This possibility is generally reserved to patients with bicorporeal uterus [7] or to patients without pregnancy desire [13]. It is also employed in case of repeated failures of the conservative treatment or in case of complications after surgery (infections or cervical restenosis) [14]. The risk of recurrence is associated with fertility-sparing surgery. In our series, none of the patients treated with UVA had a restenosis but median follow-up compared with the CDT group's follow-up was too short. No conclusion can be drawn and any impact on recurrence due to different conservative treatment would require a rather large series with a longer follow-up. Pregnancies are reported only in the CDT group. In this case, too, the series is too short to carry out a methodological analysis based on the UVA (uterus vaginal anastomosis, CDT (cervical dysgenesis treatment) results. Strengths of the Study .This paper is the first systematic review specifically focused on the surgical treatment of type 2(a)cervical dysgenesis.

CONCLUSION

Standard treatment of type 2(a) CD has not yet been established due to its rarity. The conservative technique Cervico vaginal anastomosis should be recommended, especially for young women. There is no evidence that one technique is better than the others in terms of recurrence and fertility outcome. RCT with larger sample size are needed. Hysterectomy should be recommended if cana lization procedures fail or in the absence of pregnancy desire [14]. While waiting for larger case studies to have comparable results about surgical outcomes and fertility, e proposed intraoperative laparoscopic ultrasound guidance as an innovative approach.

Conflicts of Interest: The authors have nothing to disclose.

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