

## SHORT COMMUNICATION

### Management of congenital absence of the cervix: A case report

HUSSEIN L. KIDANTO<sup>1,2</sup>

<sup>1</sup>Department of Obstetrics and Gynaecology, Muhimbili University of Health and Allied Sciences, Dar es Salaam, Tanzania

<sup>2</sup>Department of Obstetrics and Gynaecology, Muhimbili National Hospital, Dar es Salaam, Tanzania

E-mail: [hkidanto@yahoo.co.uk](mailto:hkidanto@yahoo.co.uk)

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**Abstract:** Cervical agenesis or dysgenesis is an extremely rare congenital anomaly. Conservative surgical approach to these patients involves uterovaginal anastomosis, cervical canalization, and cervical reconstruction. In failed conservative surgery, total hysterectomy is the treatment of choice. Success of reconstructive surgery depends on the amount of cervical tissue available. Hence, congenital absence of the cervix is a complex surgical problem and should be dealt with after thorough evaluation. We report an 18 year old girl presented with primary amenorrhoea and cyclic monthly abdominal pain. Initial attempted reconstructive surgery failed and hysterectomy was done. At laparotomy, there was only fibrous tissue and no cervical tissue at all. No findings related to endometriosis were observed. The uterus was removed and sectioning the fibrous tissue level of the blind vaginal cuff. Gross tissue examination showed a non communicating uterine cavity, filled with menstrual blood of about 200mls and a diffusely hypertrophy myometrium. The cervix was absent. Microscopically, there was no cervical tissue in the specimen; the uterine muscles had evidence of adenomyosis. In conclusion, re-canalization and cervical reconstruction procedures may be performed on carefully selected patients, consideration should be directed to the presence of adequate cervical stroma absence of which warrants hysterectomy.

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**Key words:** Cervical atresia; cervical agenesis/dysgenesis, cervical fragmentation/uterine malformations, haematometra. Tanzania

### Introduction

The Müllerian ducts differentiate to form the fallopian tubes, uterus, the uterine cervix, and the superior aspect of the vagina. A wide range of malformations can occur when normal development of the Mullerian ducts is disrupted. Mayer-Rokitansky-Kuster-Hauser syndrome (Golan A *et al*, 1989) is one of the congenital anomaly resulting from abnormal development of the ducts, it is manifested by complete absence of the uterus and vagina. Uterine cervix agenesis is a rare form of lack of development, regarding only a tract of the Müllerian ducts. Patients affected by this rare, "non communicating", abnormality have a functional uterus, but due to lack of cervix they get primary amenorrhoea with cyclic pelvic pain, due to haematometra. Diagnosis can be made in early adolescent period (12–17 years) due to cyclic abdominal pain, but others cases may be diagnosed later with amenorrhoea (Spence *et al*, 2003). Furthermore Müllerian

malformations are frequently associated with abnormalities of the renal and axial skeletal systems like congenital scoliosis of the spine and Klippel-Feil anomaly. Diagnosis of Müllerian duct anomalies may be difficult due to wide variation in clinical presentations as well as anatomical types. Treatment options are available and are usually tailored to the specific Müllerian anomaly. In this report a case of complete cervical atresia is described and the management options are discussed.

## Case presentation

We report the case of an 18 years old girl presented to our outpatient gynaecology clinic in August 2008 complaining of cyclic monthly lower abdominal pain and failure to menstruate. She had normal development of secondary sexual characteristics. The patient's history evidenced primary amenorrhea and cyclic lower abdominal pain; she was initially attended at Mbeya regional hospital and referred to Muhimbili National Hospital (MNH) for further management. The medical and surgical history was normal. Physical examination revealed normal breast development and other sexual characters. The hymen was intact and had normal perforations. The abdominal pelvic ultrasound showed that the uterus was enlarged and ovaries and tubes were normal; however, the cervix was absent. Serum hormonal levels were at normal levels [Estradiol (44.28 pg/ml), FSH(4.5 mIU/ml), LH(8.96 mIU/ml), TSH (2.49 mIU/ml)]. The patient was scheduled for examination under anaesthesia, and the examination evidenced normal vagina, ending in a "cul de sac" where uterine cervix was absent; above this level an enlarged uterus equal to fundus height of 12 weeks was palpable.

After thorough discussion of management options with the parents and the parents including possibilities of reconstruction and the risk of failure, they requested to be referred to India for possible reconstruction of the cervix. In October 2008 she was operated at The Madras Medical Mission where cervicovaginal canalization was performed and the cervical stent fabricated with IUCD (Cu-250) was left to prevent stenosis of neocervix. A plastic mould was introduced in the neovagina with instructions to be changed daily. When she was back in Tanzania the cervical stent was dislodged from the IUCD resulting into retention of the IUCD, stenosis of the opening created and haematometra. She was re-operated again in India and the same procedure repeated at the same hospital in 2009. She was advised to remove the cervical stent and the IUCD after 6 normal menstruation cycles expecting the epithelium to have grown in the newly created cervical canal. However, after removal she managed to have normal menses for 3 months only, the created opening was once again closed. Karyotyping done in India showed that she was a female.

She reported to our emergency medicine department in July 2010 with complain of severe abdominal pain and amenorrhea for 6 months, on examination there was a tender mass extending from the pelvis and the fundus height was equivalent to 18 weeks, Urinary pregnancy test (UPT) was negative and she had no history of sexual intercourse. Abdominal pelvic ultrasound revealed normal liver, kidney, spleen, stomach, bladder and ovaries. There was no paraortic lymphadenopathy; however there

was an enlarged uterus with fluid collection (haematometra), the right tube also had fluid collection (hydrosalpinx). After thorough physical examination there was no skeletal abnormalities, her height was normal for her age, she had no excessive body hair growth, she had feminine look with normal breast development, we discussed with her parents and herself on management options. She was counselled for hysterectomy of which they all agreed.

At laparotomy there were intense anterior abdominal wall adhesions due to previous surgeries; the ovaries and fallopian tubes were normal. Carefully adhesionalysis was done by dissection. After blunt dissection of bladder and rectum, there was just fibrous tissue and no cervical tissue seen. No findings related to endometriosis were observed at laparotomy. The uterus was removed by sectioning the fibrous tissue at the level of the blind vaginal cuff. Both ovaries and tubes were not removed. Gross examination of uterus (Figure 1) evidenced a "non communicating" uterine cavity, filled with an old coffee coloured menstrual blood which measured about 200mls after opening the cavity. The myometrium, was diffusely hypertrophied and the cervix was absent. Microscopically, the specimen showed no cervical tissue, or the characteristic squamous /columnar junction, the uterine muscles had evidence of adenomyosis. After operation this patient was scheduled to see a psychologist for continued counselling on the impact of the procedure on her future reproductive carrier.



**Figure 1: A uterus without cervix after operation**

We report this case to highlight difficulties and challenges encountered in the management of cervical atresia. The uterine cervix provides a conduit for menstrual flow, maintenance of an intrauterine pregnancy, mucus for sperm transfer, and a barrier

to infection from vaginal microflora. Atresia (dysgenesis) of the cervix may result from local segmental atrophy. Although there has been several report of successful reconstruction of cervical atresia (Nguyen *et al*, 2011) management of these cases has remained a challenge in gynaecology. The goals of reconstructive surgery for cervical malformations are to provide a conduit for menstruation, to relieve pain and preserve reproductive potential. The goals are usually achieved when there is substance to the cervix. Moreover, pregnancy has been documented after cervical reconstruction when cervical stroma is substantial (Grimbizis *et al*, 2004).

There is a consensus in the international literature that hysterectomy is the procedure of choice in a patient with cervical agenesis (Golan *et al*, 1989). However, If a long segment of cervix consists of a fibrous cord, a cervical grafting technique may be required. Cervical grafting may speed epithelisation of the newly created cervical canal. If a fragmented cervix is noted, hysterectomy is usually warranted (Rocky *et al*, 1995,). A new technique by prosthetic reconstruction of the cervix using vaginal mucosa-lined polytetrafluoroethylene offers an alternative treatment option for patients with cervical agenesis to preserve their reproductive potential ( Nguyen *et al*, 2011) ;however still more studies in this effect are needed.

Patients with cervical dysgenesis may have one of the four anatomical variations, a) the cervical body is intact with obstruction of the cervical os. Variable portions of the cervical lumen are obliterated. b) The cervical body consists of a fibrous band of variable diameter that may contain endocervical glands, c) Fragmented portions of the cervix with no connection to the uterine body. Hypoplasia of the uterine cavity maybe associated with cervical cords or fragmentation. d) The mid-portion of the cervix is hypoplastic with a bulbous tip.

No cervical lumen or cervical stroma was present in our patient and therefore failure of the primary attempt of reconstruction. The literature has shown that patients with Artesia or cervical fragmentation are not usually candidates for canalization. Patients with either cervical obstruction or a fibrous cord may reasonably be considered for reconstruction. Previous studies have shown that almost all women with either clinical agenesis or fragmentation on whom canalization was attempted failed, requiring reoperation (Edmonds, 2006). Since our patient had two attempted reconstruction that failed and the fact that she had no cervical tissue at all hysterectomy was the management of choice.

Since the reconstruction of the cervix was done in another hospital we cannot specifically tell what the cause of the failure was. However, this case describes the challenges on the management of cases of cervical atresia. In conclusion, this case and the international experience suggest canalization procedures may be performed on carefully selected patients, patients with a fragmented cervix usually warrants hysterectomy. Therefore, consideration should be directed to the presence of adequate cervical stroma to allow a cervico-vaginal anastomosis to be achieved.

### Competing interests

None declared.

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