INTRODUCTION
Alterations in the normal sequence of development of Mullerian ducts lead to a wide spectrum of reproductive anomalies. Uterine malformations are very common, minor malformations can be observed in 7-10% of all women and major malformations are seen in 2-3% of fertile women, 3% of infertile women and 5-10% of those with repeated miscarriages. A rare form of abnormality involving only a part of the Mullerian duct leading to functioning uterus and cervical agenesis is also seen. It occurs in 1 in 80,000 to 100,000 births. It is known to be associated with both partial and complete vaginal aplasia and renal anomalies. According to the American Fertility Society, cervical agenesis should be classified as type Ib Mullerian anomaly. Presentation in these cases is usually with primary amenorrhea and cyclical lower abdominal pain.

CASE REPORT
A 25-year-old nulliparous woman presented with the history of primary amenorrhea, infertility, lower abdominal pain and dyspareunia underwent vaginoplasty, total abdominal hysterectomy with right adnexal clearance for congenital absent cervix, vaginal septum with functioning uterus and right sided endometrioma. Mould was kept in vagina for 2 weeks followed by intermittent vaginal dilatation for one week. Couple was advised normal coital function and intermittent vaginal dilatation at home. Patient was followed regularly in OPD for 4 months and there was no complaint regarding sexual life.

ABSTRACT
A 25-year-old married lady with primary amenorrhea, infertility, lower abdominal pain and dyspareunia underwent vaginoplasty, total abdominal hysterectomy with right adnexal clearance for congenital absent cervix, vaginal septum with functioning uterus and right sided endometrioma. Mould was kept in vagina for 2 weeks followed by intermittent vaginal dilatation for one week. Couple was advised normal coital function and intermittent vaginal dilatation at home. Patient was followed regularly in OPD for 4 months and there was no complaint regarding sexual life.

Functioning uterus with absent cervix, short vagina and transverse septum

DISCUSSION

Complex malformations of the female genital tract are often incorrectly identified, treated and reported, probably due to not considering the malformation as a cause of the clinical symptoms and neither the embryological origin of the different elements of the genitourinary tract. Atresia of the uterine cervix is an uncommon Müllerian malformation which may be associated with vaginal aplasia. Its exact incidence is unknown and the management of women with this malformation remains confusing. Total hysterectomy is recommended when canalization procedures fail or are impossible.

Any abdominal or pelvic, acute or chronic pain in a pubescent girl must evoke an obstructive genital syndrome. As in this case, the patient had primary amenorrhea, chronic lower abdominal pain and mass in hypogastrium, which indicated an obstruction in genital tract. The presence of a mass inside the vagina, discovered on rectal examination, suggests blood retention above an obstruction. Clinical examination easily eliminates imperforate hymen or blind hemivagina, but might not differentiate cervical atresia from high vagina transverse septum. Transabdominal or transperineal ultrasonography may specify the level of obstruction, but it is not very reliable for the diagnosis of cervix atresia. Magnetic Resonance Imaging (MRI) currently appears to be the most reliable morphological examination for the diagnosis of utero-vaginal malformations with a surgical correlation >80%. Laparoscopic exploration has the ability to assess the type of uterine malformation, and reveals other complications of the upper genital tract that may require appropriate surgery.

When both the vagina and cervix are absent and a functioning uterine corpus is present, it is difficult to obtain a satisfactory fistulous tract through which menstruation can occur. Conservative surgical treatment of uterine cervical atresia, mainly canalization techniques, are at high risk of secondary stenosis of the cervix, up to 40-60%. Moreover, some authors conclude that chance of subsequent pregnancies is unlikely, particularly in association with vaginal aplasia. Consequently, hysterectomy was recommended as first-line treatment by many authors.

Many authors have recommended hysterectomy as an initial procedure as it will eliminate much needless suffering from associated problems such as cryptomenorrhoea, sepsis, endometriosis and multiple operations. At the same time, plastic surgeon should be prepared to perform a vaginoplasty with the use of a split-thickness graft. If the neovaginal space is allowed to close and scar, future operations to develop an adequate neovagina are associated with increased risk of graft failure and fistula formation.

Vaginally, dissection was done for neovagina for approximately 10 cm in length. Unfolded septum was used to cover the raw neovagina. Upper extent was stitched to the top most raw of neovagina. Mould of 20 cc syringe was covered with gauze and condom, fixed in place with labial suturing. She was kept catheterized for 7 days. Laxative was also prescribed.

After 2 weeks, mould was removed under general anaesthesia to keep the procedure painfree, proper evaluation and wash down. In the ward, she was daily applied with triple sulpha cream and trained for insertion of mould thrice-a-day. After one further week, after the satisfactory result of neovagina, she was allowed for coitus with proper lubrication and twice daily mould insertion.

Figure 1: Unfolded upper vagina.

Figure 2: Peroperative abdominal finding showing uterus with right adnexa and absent left adnexa and exploration of uterine cavity with sound.
However, recent advances in reproductive technology and laparoscopic surgical techniques have made conservative surgery possible and is perhaps going to be the first-line treatment option in highly specialized unit with the expertise in laparoscopic surgery.

REFERENCES


