Absent vagina a description of cases with varied ages, presentations and reconstructions


1Department of surgery, medical college Kirkuk university, Iraq,
2Department of Gynecology, medical college Kirkuk university, Iraq
3Department of Obstetrics and Gynecology, Azadi teaching hospital, Iraq
4Department of Gynecology, medical college Kirkuk university, Iraq

ABSTRACT

The aim is to describe cases of vaginal agenesis of varied ages presentations and managements and report the outcome of interventions regarding restoration of continuity of reproductive tract and normal sexual activity. A prospective descriptive case series study. Five female patients with vaginal agenesis were recruited from November 2005 to November 2016. Three were adolescents and had a functioning uterus. Three patients underwent vaginoplasty using McIndoe method, one had vagino-vaginal anastomosis, and the other had hysterectomy to remove source of menstruation. Age ranged from 12 to 24 years. The outcome of vaginoplasties to the 2 young females and to one of the adolescent girls was adequate vagina, whereas the other 2 adolescent girls had failed redo vaginoplasty and ended with a patent narrowed vagina in one and a closed vagina in the other which required a hysterectomy. On follow up (range 5 - 42 months) 2 of the adolescent’s girls had regular menstruation with no pain and pelvic collection, while the third one had amenorrhoea following a hysterectomy. The 3 patients who had successful vaginoplasty got married and enjoyed satisfactory intercourse. Patients with absent vagina presents at variable ages depending on whether a functioning uterus is present or not. The success of vaginoplasty is less when it has to be done in adolescence than when it is done in adulthood when patient is grown-up. Best outcomes require accumulative experience, ideally with the aid of doctors of other specialties.

INTRODUCTION

Complete vaginal agenesis is a rare anomaly that may result from a variety of different underlying diagnoses. The reported incidence is 1 in 4,000 to 10,000 female births. The condition most frequently associated with agenesis of the upper two-thirds of the vagina is Mullerian aplasia or Mayer-Rokitansky-Kuster-Hauser (MRK) syndrome. Vaginal agenesis may be accompanied by a functioning uterus, which requires special management. It may be found in 2 to 7 percent of women with (MRK) syndrome and may present with cyclic abdominal pain, primary amenorrhoea, and a palpable mass due to distension of upper vagina with menstrual blood. Endometriosis is a common finding in such cases and may be a cause of infertility (1 Fedele et al, 2006) (Sanfilippo et al, 1986). These women have a normal female genotype, phenotype, and normal ovarian function.
The congenital anomalies of the female reproductive tract can be evaluated by several radiological techniques. The different modalities employed include hysterosalpingogram, ultrasonography, sonohysterography, and MRI. Although MRI is considered the gold standard technique for diagnosing Mullerian anomalies (Pellerito et al., 1992), each imaging technique has its inherent strengths and limitations; therefore, a combination of several techniques may best evaluate a particular abnormality.

The most important aspects of the surgical management of this condition are correct diagnosis of both the underlying abnormality and its anatomy, documentation of any associated renal or skeletal anomalies, and proper psychological preparation of the young woman for any anticipated corrective surgery. The ideal surgical procedure would be low-risk and to create a vaginal canal in the correct axis, of adequate size and secretory capacity, to allow intercourse to occur without the need for continued postoperative dilatation. An important influence on the perioperative complication rate is previous vaginal/perineal surgery because this makes dissection of the recto-vesical space and the attainment of hemostasis more difficult (BuSS and Lee, 1989a).

This study aimed to describe many cases of vaginal agenesis of variable ages presentations and management and report the outcome of the interventions regarding restoration of continuity of the reproductive tract and enjoyment of normal sexual activity.

**MATERIALS AND METHODS**

In this prospective descriptive case series study, five cases with absent vagina of variable ages presentations and management attending gynecologic clinic of Azadi General Hospital from November 2009 to November 2016 are described.

Case 1: The first case is a 14-year-old adolescent girl presented with primary amenorrhea and periodic lower abdominal pain of a few months' duration. On examination, she had normal female physical characteristics with a lower abdominal mass and an absent vagina. Abdominal ultrasonography revealed a distended uterus and a pelvic cystic mass. The kidneys were normal. The patient and her family were concerned about her amenorrhea and future fertility. A provisional diagnosis of the absent vagina with a functional uterus was made.

The first surgical procedure was to relieve the obstruction of retained menses by constructing a neovagina in order to reach the wall of the pelvic cyst which was incised and over 200 mL of chocolate-colored retained blood was released. The use of on a split-thickness skin graft fabricated mold placed into the dissected space was delayed until the vagina was clear from discharge.

Four weeks later, a thick split-thickness skin graft was taken from the left thigh and wrapped over a solid, hollow perforated vaginal mold. This was inserted into the previously created vaginal space. The patient was then placed on strict hospital bed rest for 7 days. Four weeks later, the mold was removed. The patient was instructed to wear a vaginal dilator daily before going to bed.

A few weeks later, the patient developed lower abdominal pain and mass, which was confirmed on ultrasonography. Vaginal drainage of the collection was performed. Later the patient was instructed to wear a vaginal dilator regularly.

Again, several weeks later the patient had a recurrence of the pain and mass. This time abdominal approach was decided to relieve the obstruction. The distended upper vagina was opened and it was found to be separated from the neovagina by a thin membrane. It was excised and vagino-vaginal anastomosis performed. A hollow vaginal mold was left in and was removed 2 weeks later.

Case 2: A 24-year-old woman presented, asking for vaginoplasty prior to marriage. She has a known history of primary amenorrhea due to absent vagina and uterus with a single right pelvic kidney. Vaginoplasty using McIndoe method was performed as a one stage operation. The vaginal mold was removed 25 days later and was instructed to wear a vaginal dilator daily before going to bed. Four months later she got married.

Case 3: A 12-year-old adolescent girl presented similar to case 1. She had appendicectomy 1 month before. Surgical relief of obstruction was performed by constructing a neovagina similar to case 1. The vaginal mucosa lining the hematocpos was anastomosed to the mucosa of introitus ending with a patent vagina. Three weeks later, after the vagina was clear from menstrual discharge, a graft covered mold was planned to be placed into the previously created neovagina. Unfortunately, this space was almost closed, and while separating adhesions, the rectal wall was injured, which halted continuation of the planned procedure. The injury was repaired, and later, the patient was instructed to dilate the already patent narrow vagina by using a dilator daily.

Case 4: A 13-year-old adolescent girl presented similar to cases 1 and 3. Hymenotomy failed to relieve obstruction because it was due to absent vagina with a functioning uterus. Relief of menstrual obstruction...
tion was performed similar to cases 1 and 3. The patient did not show up for skin grafting of the created vaginal space until 2 months later when she developed abdominal pain and mass with a closed vagina. During redo vaginoplasty, the rectal wall was injured even with vigilant dissection. The injury was managed similar to case 4, and the patient remained to have a closed vagina. Abdominal Hysterectomy and salpingectomy was performed after 2 months due to inability to tolerate pain arising from cryptomenorrhoea.

Case 5: A 23-year-old woman presented similar to case 2. She requested vaginoplasty prior to a planned marriage. The operative and postoperative management was similar to case 2. A few months later she got married.

RESULTS AND DISCUSSION

The age of patients ranged from 12 to 24 years (mean 17.2 years). Three of them were adolescent’s girls, and the other 2 were young females. The 3 adolescents girls had a functioning uterus in addition to an absent vagina, and all presented acutely as severe abdominal pain. The 2 young female patients presented requesting vaginoplasty in order to get married. The 3 adolescent’s girls received erroneous treatment; exploratory laparotomy (1) and appendectomy (1) and all 3 had undergone hystereotomy.

Three successful McIndoe vaginoplasties were done to the 2 young females (cases 2 and 5) and to one of the adolescent girls (case 1). The average operating time was 90 minutes, and the average blood loss was 50 mls. There were no intra-operative and post-operative complications in these 3 cases. The mean vaginal depth was recorded to be 8 cm, and the ultimate outcome of these vaginoplasties was an adequate vagina with skin graft fully taken. There was no granulation tissue or adhesions. No patient reported dyspareunia, exceptional dryness, or disturbing vaginal discharge. Unfortunately, in the adolescent girl (case 1) the McIndoe vaginoplasty was not straightforward as in the other 2 cases since there was failure to restore utero-vaginal continuity vaginally which was important in her case since she had a functional uterus. Hence, an abdominal approach had to be contemplated in order to restore this continuity which was ultimately successful. Endometriosis complicated this case, and right salpingectomy was performed during laparotomy. Great anxiety, and worry to the patient and her family resulted from these staged surgical procedures. Nevertheless, the patient and her family were pleased with the restoration of menstruation and retain fertility.

In the adolescent girl (case 3) a durable patent but narrow vagina ended after anastomosing the vaginal mucosa lining the hematocolpos to the mucosa of introitus. During redo vaginoplasty and while attempting to place a graft covered mold in the previously surgically created neovagina rectal injury occurred which halted continuation of the procedure. Subsequently, she had regular menstruation with no abdominal pain, and a number of ultrasonographic examinations revealed no pelvic collection. The patient was instructed to dilate the narrow mid-vaginal segment, below which the vagina was adequate and stretchable.

The last adolescent girl (case 4) had pain relief and disappearance of the pelvic mass after vaginoplasty similar to cases 1 and 3. Unfortunately, she did not show up for skin grafting of the created vaginal space until 2 months later when she developed abdominal pain and a pelvic mass. On examination, the vagina was closed again, and during redo vaginoplasty, the rectal wall was injured even with vigilant dissection. The injury was healed, but the patient remained to have a closed vagina with pain and pelvic mass. She disappeared to show up again, being unable to bear pain arising from cryptomenorrhoea. Hysterectomy was performed to relief her pain and pelvic collection.

The average hospital stay for creation of a neovagina to relief obstruction was 5 days, while for the implantation of skin graft was 18 days. On Follow up (range 5 - 42 months; mean 22 months) the 2 adolescents patients (cases 1 and 3) had regular menstruation with no pain and no pelvic collection, while case 4 had amenorrhoea following hysterectomy and relief from pain and collection. The vagina was adequate in cases 1, 2, and 5, while it was patent with mid zone narrowing in case 3 and closed in case 4. All 3 patients (cases 1, 2, and 5) got married and enjoyed satisfactory intercourse. Case 1 failed to conceive even after 3 years of marriage. Due to the rarity of complete vaginal agensis and particularly when it is accompanied by a functioning uterus, the general gynecologist is likely to encounter this condition only once or twice in his/her professional career. This inexperience is accompanied by the fact that there are so many operations for the correction of this anomaly, which suggests that no one procedure is ideal and satisfactory and at present, there is no consensus in the literature regarding the best option (Templeman et al, 1999). The most commonly performed operation is the Abbe-McIndoe vaginoplasty, which involves the use of thick split-thickness skin graft on a fab-
ricated mold placed into a space dissected between the bladder and rectum and diligent use of vaginal dilatation postoperatively (McIndoe and Banister, 1938). Multiple studies have demonstrated long-term patient satisfaction after such a technique (BUSS and LEE, 1989b) (Martinez-Mora et al., 1992) (Strickland et al., 1993) (Goerzen et al., 1994). It is felt to have a greater than 80% functional success rate (Klingele et al., 2003) (Frost-Arner et al., 2004) (A et al., 2005) (Rock et al., 1983). However, it leaves the patient with a potentially disfiguring scar from the donor site. Other complications include graft failure, wound infection, hematoma, and fistula formation (Sparac et al., 2004). The most important considerations for the long-term success of this operation include: the motivation and mental status of the woman and her willingness to comply with the postoperative program of regular vaginal dilatation that continues for at least 6 months or until regular sexual activity commences. In adolescent girls with vaginal agenesis accompanied by functioning uterus, the usual practice of postponing McIndoe vaginoplasty until the patient is wanting to have regular sexual intercourse (Templeman et al., 1999), has to be broken because of the urgent demand to relief the obstruction of menstrual blood.

In this study it was found that the 2 young women with absent vagina and uterus had better results in terms of functional vagina than the 3 adolescent girls with accompanying functioning uterus, since vaginoplasty in the 2 young women was delayed until they were in their twenties when they were mature, psychologically stable and kept her vagina dilated postoperatively through regular marital sexual intercourse. Any surgical approach that requires postoperative dilatation, the patient must be developmentally and emotionally mature enough to accept this responsibility (Templeman et al., 1999). On the other hand, relief surgery had to be done rather urgently in the 3 adolescent girls at the time when they had their clinical presentation (Sparac et al., 2004). Such girls were not fully-grown and psychologically not ready for sexual activity yet. That is why technical difficulties were faced. They were not cooperative in spite of a thorough explanation of the procedure and the importance to keep all physician’s instructions. Their families were confused and bewildered with what was going on and in cases 3 and 4 they couldn’t afford to attend scheduled dates which led to closure of the primary constructed neovagina which made redo vaginoplasty difficult and risky. There was failure to open the closed vagina and the procedure ended in rectal injury which was dealt with successfully. This was expected since the first procedure is the most likely to succeed, and any subsequent surgery at the vaginal area may be more technically difficult and will put the patient at significantly greater risk of operative injury to surrounding tissues and a poor functional result for these adolescents (Templeman et al., 1999). I feel that in order to minimize risks of redo vaginoplasty one should not delay grafting of the created neovagina for more than few days before its closure and occurrence of false feeling of cure by the patient and her family which will make them fail to show up for further surgery. Some authors consider hysterectomy plus creation of neovagina an accepted treatment for this kind of anomaly, especially when the patient and her family do not comply with follow up (Sparac et al., 2004) (Rana et al., 2008).

Plastic surgery Gynecologists with experience in reconstruction, ideally with the aid of general and plastic surgeons, may provide the best outcomes for the patient undergoing major reconstruction. A psychologist familiar with female reproductive anomalies is invaluable for helping the patient and surgeon determine impact of the diagnosis and readiness for correction (Miller and Breech, 2008). In 3 of our cases, the assistance of a plastic surgeon contributed to the successful use of split-thickness skin graft covered mold in vaginoplasty.

**CONCLUSION**

Patients with absent vagina presents at variable ages depending on whether a functioning uterus is present or not. If it is there, the presentation will be in adolescence when vaginoplasty would have to be done to relief obstruction of menstrual flow. Its success rate is less than when it is done at adulthood when the patient is psychologically and emotionally stable and ready for postoperative vaginal dilatation usually through marital sexual intercourse. Best outcomes of such cases require accumulative experience in vaginoplasty, ideally with the aid of doctors or other specialties including a psychiatrist.

**REFERENCES**


