

Vaginoplasty in vaginal agenesis associated with MRKH syndrome with tabularized peritoneal pull-through

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Abstract

In this paper, a 16 year old girl who was diagnosed as a case of Mayer-Rokitansky-Küster-Hauser (MRKH) syndrome, underwent a combined laparotomy-peritoneal approach to create a neovagina by using tubularized peritoneal graft with uneventful postoperative period. Vaginal dilation was maintained with a vaginal mould daily for six months and three to four times weekly thereafter. She was followed-up after 2 and 4 weeks in the first month and three monthly for a duration of six months. On second follow-up, adequate vaginal length of 6-7 cm and width of 2.5-3 cm were achieved with healthy vaginal tissue. Hence, the laparotomy-peritoneal approach of using a peritoneal graft for creations of a neovagina can be an effective approach with minimal surgical morbidity to create a passageway for satisfactory intercourse.

Introduction

Mullerian agenesis, or Mayer-Rokitansky-Küster-Hauser syndrome, has an incidence of 1 in every 4000-10,000 females. It results from embryologic failure of development of the müllerian duct, which leads to agenesis or hypoplasia of the uterus and vagina. Patients normally present with primary amenorrhea but have normal secondary sexual characteristics and are genetically female. They have an absent or short blind-ending vagina. Most affected individuals have small, rudimentary uterine bulbs without functional endometrium. The ovaries are normal in structure and function because they have a different embryologic source. There is an association with other congenital anomalies; 30% have renal anomalies and 12% have skeletal, mainly vertebral anomalies.^{1,2}

Treatment aims to create a neovagina that can provide a passageway for satisfactory intercourse with the least morbidity. Options include nonsurgical and surgical methods. The nonsurgical Frank and Ingram methods of progressive self-vaginal dilation with hand-held dilators or dilators mounted on a bicycle seat stool carries the least morbidity but requires a prolonged period of treatment and may cause discomfort.^{3,4} Surgical options for the creation of a neovagina include split-thickness skin graft (McIndoe procedure) or full-thickness skin graft, sigmoid vaginoplasty, peritoneal graft (Davydov procedure), Vecchiotti

procedure which uses an external traction device placed on the abdomen to exert continuous pressure on the vaginal dimple via a 2 cm olive bead, and amnion grafts.² However, there is still no clear consensus on the best method. Davydov procedure is an effective approach with minimal surgical morbidity that can create a passageway which is adequate for normal sexual function.⁵

In this paper, we have described the use of peritoneum in vaginoplasty (Davydov procedure) with slight modification, that is, the use of tubularized peritoneal flap. Combined laparotomy and vaginal approach was used to create a neovagina. Though laparoscopic modification of the original Davydov procedure is common now-a-days but laparotomy can still be done.

Case Report

A 16 year old girl presented with primary amenorrhea with well development of secondary sexual characteristics. Height, weight, and external genitalia were normal. On examination, the vagina was absent with a presence of small dimple. Ultrasound and contrast enhanced CT scan of lower abdomen showed normal ovaries and hypoplastic uterus. Karyotype studies showed the normal female chromosomes 46XX. Hormonal analysis including follicle stimulating hormone, luteinizing hormone and



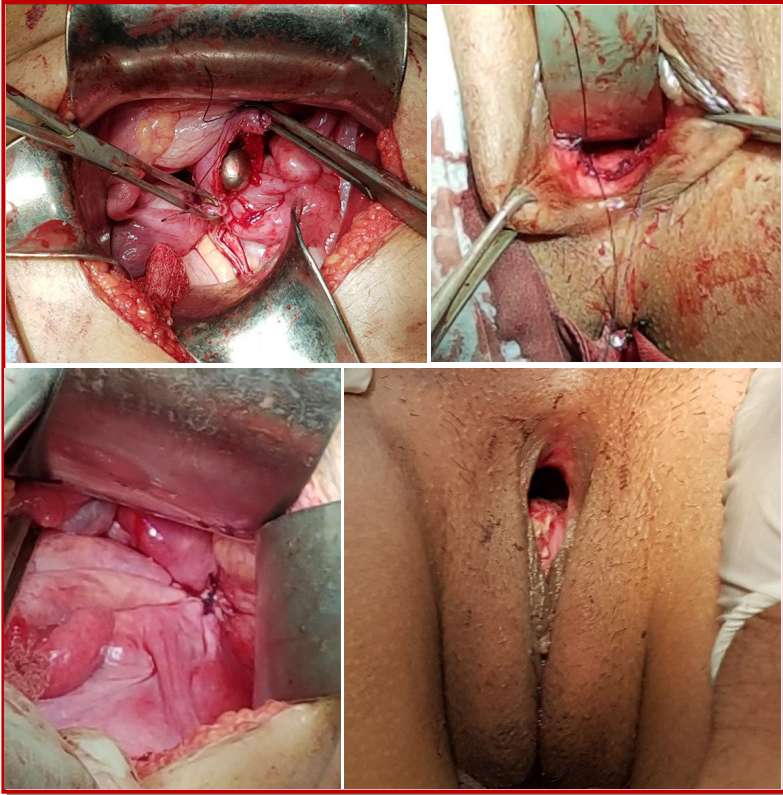


Figure 1: Tubularized peritoneal flap held by 2 tissue forceps at the margin of anterior and posterior flap. Hegar's dilator is at middle of flap (upper left); Tubularized flap was pulled through and fixed with margin of neovaginal introitus using 2-0 polyglycolic acid-interrupted sutures (upper right); Neovagina was closed at peritoneal side using 1-0 polyglycolic acid with peritoneum of bladder anteriorly and peritoneum of rectum posteriorly by a purse-string suture (lower left); Second follow-up; Near normal vaginal opening was present with vaginal length of 7-8 cm allowing index finger easily without discomfort (lower right)

prolactin were all in the normal range.

Operative technique

Keeping patient in the lithotomy position, the surgical painting, draping and catheterization were done. A transverse incision was made midway on the blind vaginal pouch. Blunt and sharp dissections were done till apex of the vaginal pouch to create an adequate vaginal space (8-9 cm), taking care of not to injure rectum and urethra. A pack was placed in the neovaginal space and the patient was repositioned in supine position. Lower midline incision was made and peritoneal cavity reached. Intraoperative findings showed rudimentary uterine bulb with normal bilateral ovaries. The anterior and posterior leaves of the broad ligament were opened from lateral to medial (5-6 cm). Bilateral clamps were applied lateral to the flap and cut downwards to create two anterior and posterior flaps and rudimentary uterine tissue was removed leaving serosal covering. These flaps were tubularized by closing it laterally (3-0 polyglycolic acid). The top of the neovaginal space was made prominent by a

Hegar's dilator and kept in between tubularized flap and the incision was given over Hegar's dilator (Figure 1). The tubularized flap was pulled through the opening so as to reach the neovaginal introitus and fixed with margin using 2-0 polyglycolic acid interrupted sutures. The top of the neovagina was closed using 1-0 polyglycolic acid with peritoneum of bladder anteriorly and peritoneum of rectum posteriorly by a purse string suture to isolate from rest of the general peritoneal cavity. Vaginal mould was placed and abdomen closed in layers keeping drain *in situ*.

Postoperatively, patients were covered with intravenous ceftriaxone and amikacin for 3 days followed by 7 days of oral cefexime. She was started on graduated feedings to a soft diet on the 2nd day and encouraged to ambulate. The cylindrical vaginal mold and bladder catheter were kept *in situ* and removed on the 3rd postoperative day and then vaginal mould was replaced every alternate day till discharge. The patients were monitored up to 10th postoperative day to assess wound healing and any complication. She was discharged on the 10th postoperative day with a vaginal length of 6-7 cm and a width of 2.5 cm. She was advised to follow-up after 2 weeks and use of iodine ointment at vaginal margin twice a day.

Follow-up

On first follow-up (2 weeks after discharge), near normal vaginal opening was present with vaginal length of 7-8 cm allowing index finger tightly. A band of constriction was felt near vault and hence patient was taught to use a vaginal mould daily.

On second follow-up (4 weeks after discharge), near normal vaginal opening was present with vaginal length of 7-8 cm allowing index finger easily without discomfort. Mild constriction was felt near vault.

She was advised to follow-up three monthly for six months and use dilator daily for six months and 3-4 times a week thereafter.

Discussion

Timing for nonsurgical or surgical creation of a neovagina is critical and is best planned when the patient is emotionally mature and expresses the desire for correction. This is important for success of the procedure because it requires a period of daily postoperative vaginal dilation for 3 months followed by maintenance with sexual intercourse or vaginal mold use a few times a week.

Nonsurgical creation of the vagina with self-vaginal dilators has the lowest morbidity but may require a long duration of vaginal mold use, high degree of self-motivation, and may be uncomfortable.⁴ In a study by Gargallo³ of 64 patients, 88% achieved

functional success at a median of 18.7 months. Success rates depend on the frequency of vaginal mold use, sexual activity, and initial vaginal depth. Manual dilation is less likely to be successful in patients with a completely absent vagina (as in our case). Surgery is an option for patient for whom dilator is unsuccessful or for patient who prefers surgery. Many surgical methods are available but the best option for functional outcome and sexual satisfaction with the least morbidity is still unknown.

Using peritoneum to create a neovagina is ideal because the peritoneum is naturally moist and can provide lubrication for intercourse.^{4,6} The thick peritoneal flap gives added strength to the tissue, making it less friable and less likely to break down during intraoperative handling and during the postoperative healing phase. It avoids the morbidity from the use of other grafts such as skin grafts and bowel grafts.

Skin grafts are associated with skin scarring and risks of infection at the donor site. Split thickness skin grafts (McIndoe technique) have a high-risk of postoperative contracture and require long-term regular use of a vaginal mold for the prevention of complication.⁷ Full-thickness skin grafts have a much lower risk of neovagina skin contracture but still have risks of skin scarring from the donor site.⁸ Bowel grafts are associated with surgical risks from major bowel surgery, for example, intestinal stenosis and obstruction, intestinal wound dehiscence, and fistula formation, and may also have neovagina complications of introital stenosis and mucosa prolapsed. Bowel grafts also tend to have excessive mucoid discharge.⁹⁻¹¹ An amnion graft does not have a blood supply and disintegrates as re-epithelialization takes place. The amnion graft is associated with foul smell and risk of infection.⁴

Postoperatively, it is important to use a vaginal mould that is large enough to allow apposition of the uterine serosa-peritoneal graft to the vaginal walls yet not large enough to cause pressure necrosis.⁷

In this case, we used a 4-5 cm vaginal mould in the initial postoperatively which is removed before the patient is sent home. On first follow-up, patient was taught to use vaginal dilator and advised for further follow-up.

Conclusion

Tubularized peritoneal neovaginoplasty is a simple and effective method with good outcome and minimal morbidity to the patient. Though laparoscopic modification of original Davydov procedure is common now-a-days but laparotomy can still be done. It avoids the problems associated with graft use from other sites and has good form and function to enable satisfactory intercourse.

Ethical Issue

Written and signed informed consent from the patient was taken for publishing this case report.

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