

Case Report

Laparoscopic neo-vagina creation with peritoneal pull through vaginoplasty technique: Minimal access surgery on a patient with MRKH syndrome in Sri Lanka

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Introduction

Mullerian dysgenesis has an incidence of about 1/5000 females [1] and is caused by embryological underdevelopment of the Mullerian duct. This multifaceted condition represents a defect in mesodermal organization and is often associated with renal or skeletal abnormalities (53%). In most individuals, karyotype evaluation reveals a normal 46, XX but there have been a few karyotype rearrangement abnormalities reported including duplications or deletions [2,3]. The resulting inability to reproduce is a highly emotionally detrimental condition and psychological support has been the mainstay in patient management. Establishing a functional vagina with approximately 7cm length is important for comfortable sexual intercourse and this can be successfully done with vaginal dilators (90-96%). If this fails, there are a vast variety of surgical options available for neo-vagina creation [4,5].

Surgical interventions can be divided into 2 main types, traction based (Vehietti procedure-open/laparoscopic) and graft based (Abbe-McIndoe technique-vaginal route with skin graft, sigmoid vaginoplasty-combined vaginal and open/laparoscopic approach, Davydov technique -combined vaginal and laparoscopic approach with peritoneal graft, William's vaginoplasty and Wharton-Sheares-George method) [4,6]. The literature described procedures give varying success rates and failure/complication rates. Though most surgical options are classical open interventions, advanced laparoscopy techniques have led to surgical neo-vagina creation in unique ways [7].

Case History

A 27-year-old woman, married for 2 years, was assessed for subfertility. During focused history taking, it was found that the couple's sexual intercourse was abnormal as they could not perform penetrative vaginal intercourse. On examination, she was of average build with a female phenotype. However, genital examination showed no vaginal orifice and the vaginal dimple was less than a centimetre (Figure 1). On further assessment, she confirmed that she had primary amenorrhea but denied symptoms such as cyclical pelvic pain, galactorrhoea or visual disturbances. There was no history of difficulty in gender assignment in the past.

Her routine biochemical workup, that included FSH, LH, TSH, prolactin and estradiol levels, was normal. However, ultrasound scan failed to detect a uterus although both ovaries were visualized. These findings were later supported by MRI pelvis (Figure 2) where bilateral uterine horns were noted with no other malformations seen in the abdomino-pelvic viscera. Patient's karyotype was 46, XX. A diagnosis of Mayer-Rokitansky-Küster-Hauser syndrome (MRKH) was made. (Figure 3)



Figure 1: View of vulva at surgery-

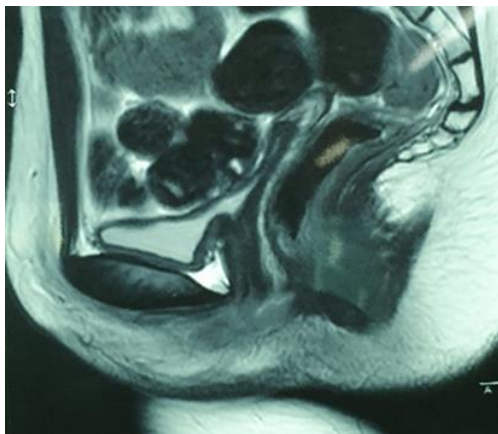


Figure 2: Sagittal plane view of MRI pelvis which show bilateral adnexa with no proper uterus formation

Figure 3: Karyotype result of patient- 46, XX)

Following counselling, the couple requested for surgical neo-vagina formation over the use of vaginal dilators and were referred for specialized care to the Colombo South Teaching Hospital.

Management

The patient and her partner were counselled and primary surgical neovagina formation over dilation was arranged as there was a technical difficulty due to a very short end vaginal pouch.

The surgical technique that was followed was a modification of the Laparoscopic Davydov Procedure combined with the Wharton-Sheares-George method. Surgery was performed in two stages. First, as a perineal open dissection (Figures 4-11) and secondly, as intraperitoneal dissections to reach the final step where the mobilized peritoneum was anchored to the new vaginal opening (Figure 12).



Figure 4: Laparoscopic view of patient's pelvis. uterine horns on either side can be seen along with B/L tubes and ovaries. Connecting each side, there is a thick peritoneal band



Figure 5: Image shows development of an anterior peritoneal flap which requires identification of vesical peritoneal reflection and division of both round ligaments - derivative of Wolffian duct.

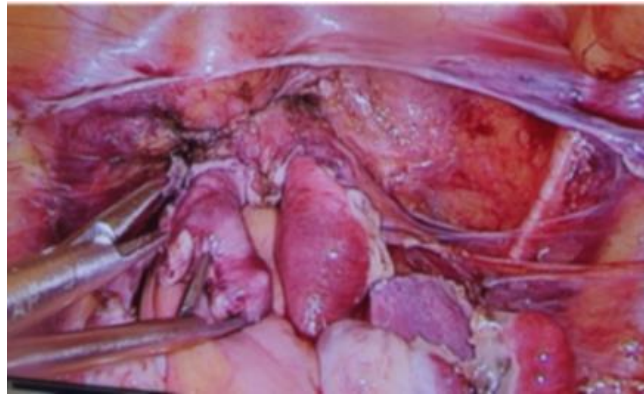


Figure 6: Image shows intact uterine horns as one unit.

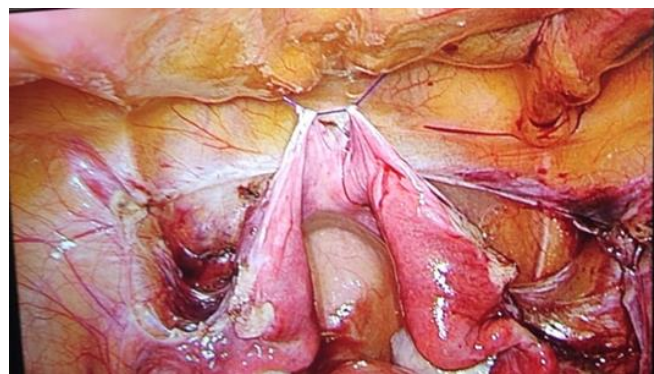


Figure 7: Anterior peritoneal fold is suspended to abdominal wall to get access to remaining pelvic peritoneum with minimal interference to the surgical field



Figure 8: Peritoneum over cul de sac is mobilized and rectum is displaced to reach the potential space for new vagina



Figure 9: The rudimentary Mullerian ducts are dilated by pushing Hegar dilators in the direction of the pelvic axis, and the resulting median raphe was then intersected

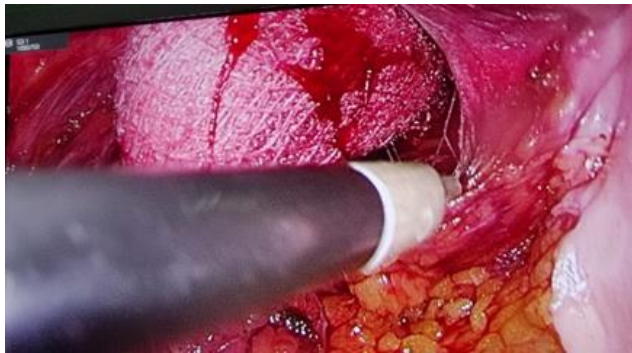


Figure 10: Newly developed tissue space for vagina. A swab was placed for identification purpose

Figure 11: Approximation of peritoneal fold using absorbable 1-0 polyglactin and incorporation of rudimentary uterine horns to apex of neo-vagina

Figure 12: Mobilized peritoneum brought to vaginal orifice to anchor using absorbable sutures

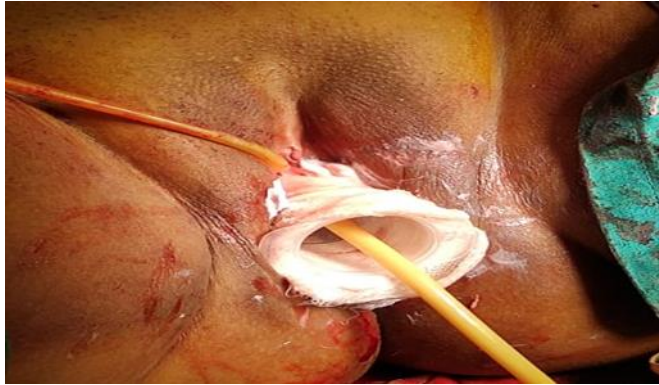


Figure 13: Local oestrogen containing mould is applied inside neo-vagina)



Figure 14: Self vaginal dilation in post-operative period)

Post operatively, the vaginal mould (Figure 13) was kept for 6 weeks and afterwards, self-dilation (Figure 14) was started till post-operative 12 weeks was reached. Thereafter, the couple was advised for penetrative sexual intercourse involving the neovagina.

Discussion

Vaginoplasty can be done by various non-surgical and surgical techniques. Non-surgical management includes serial dilatation with vaginal dilators [8,9,10] (Table-1). Dilators are held in place for 20–30 minutes, two or three times a day, in order to stretch the vaginal epithelium into the potential space between the bladder and rectum.

Table 1: Non-surgical techniques of vaginoplasty

Frank's method (1938)	Ingram's method (1981)
Self vaginal dilation	Progressive dilators are attached on a bicycle seat
Gradual process	Use own perineal pressure
Takes about 6 months to reach desired length	More successful than former method [11] (88% vs. 91%)

Disadvantages associated with these methods are urinary tract infection, vesico-vaginal fistula formation or secondary prolapse. Being non-invasive methods with high success rates, it is recommended as a first line therapy by gynecologists [12,13]. However, when the vaginal dimple is very short, this is practically challenging.

Surgical methods possess distinct advantages and disadvantages (Table 2).

Table 2: Studies comparing advantages and disadvantages of surgical methods

Surgical method	Advantages	Disadvantages
Vecchietti procedure [4,14] (Traction method), 98% success rate	Laparoscopically possible & 7-9 days is sufficient for adequate length of neo-vagina formation. Traction can be adjusted as an outpatient procedure	Intra-operative viscus injuries, Urinary incontinence
McIndoe procedure [4,15] Open surgery where split thickness skin graft is used as neo-vaginal epithelium following potential space development between bladder and rectum	Traditionally popular method, Only perineal approach is required	Potential vaginal stenosis, perforation of the bladder and rectum, graft failure and unsightly scarring at the graft site. Has to live with a mould for 3 months after surgery
Sigmoid Vaginoplasty [4,16] Complex surgery where one end of a resected sigmoid segment is pulled down with its vascular pedicle to the introitus to form a new vagina and the other end is closed to create a blind pouch	Laparoscopy is possible. Lubrication is naturally present as intestinal epithelia is used	Chronic vaginal discharge, foul odor, stenosis at the anastomotic site and the risk of developing adenocarcinoma in the graft. Operative morbidity and mortality is high. Infection risk is more.
Williams vulvovaginoplasty [4,15] Skin flaps from the labia majora are used to create a pouch horizontal to the perineum	Perineal procedure. Simple procedure compared to others	Short length and unusual angle for coitus of the resulting pouch
Davydov procedure [4,17] An autologous peritoneal graft is used for vaginoplasty. Peritoneum graft from pouch of Douglas is dissected and mobilized. After creating vaginal space, peritoneum is reached and then mobilization is started by dissection of recto-vesical space. abdominal mobilization of the peritoneum is done to create vaginal fornices and attachment of the peritoneum to the introitus is then performed.	Laparoscopically possible Good anatomical and functional success is reported with this procedure Laparoscopy has better visualization, shorter hospital stays and lesser post-operative pain	A vaginal mould is inserted for 6 weeks. No major complications are reported other than growth of granulation tissue at the vaginal vault

Wharton-Sheares-George method The basic Müllerian ducts are gradually expanded by advancing Hegar dilators along the pelvic axis, followed by intersecting the resulting median raphe with diathermy. A vaginal mold is inserted into the newly created cavity and held in position by two sutures	No major Intraoperative and post-operative complications or prolapse were reported Minimally invasive, quick and safe surgical option that does not require allogenic or autologous transplants	
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There are wide variations in the use of grafts for neovagina formation starting from skin graft, amnion, intestines and peritoneum. Peritoneal grafts were first used by Russian clinicians and gained wider popularity with Davydov method. However, there were many subsequent modifications including the use of thin peritoneal graft, thick peritoneal graft with substratum and the combined use of peritoneum with amnion grafts [7]. The uniqueness of our surgery is related to utilization of the rudimentary uterine horns to create an apex to the neo-vagina with provision of an additional support and to mimic the cervix which has a role in intercourse [18,19].

Laparoscopic peritoneal pull-through neovagina formation offers a relatively easy surgical procedure with excellent results on long term follow up [7]. This procedure is practically devoid of morbidity associated with other traditional techniques. Some clinicians avoid use of postoperative mould as vaginal shrinkage and adhesion formation was minimal in their experience. Peritoneal flaps must be kept in continuation with the parent site through the attachment for continuation of blood supply. As the pelvic peritoneum lining the neo-vagina is responsive to estrogen, it is safe to use local estrogen. Early phase of sexual intercourse require lubricants and metaplasia of pelvic peritoneum to normal vaginal epithelium occurs after about 6-12 months in almost all patients. As the ovary became accessible per vagina, ovum retrieval and pregnancy using surrogate mother is possible, thus making this a fertility enhancing procedure for MRKH patients. This reduces patient's mental stress of being not able to have intercourse and reproduce offspring with own genetic material.

Conclusion

Rokitansky syndrome is a rare congenital disorder characterized by uterine and vaginal aplasia. Classical presentation is primary amenorrhea with normal secondary sexual characteristics, blind vagina and absence of uterus and cervix on ultrasonography with normal 46 XX karyotype. MRI abdomen/pelvis gives more information about additional congenital abnormalities. Management includes extensive counselling on reproductive issues and provision of psychological support. This is followed by neovagina formation at an appropriate age. At present, minimal access surgeries are becoming more popular though most academic bodies recommend non-surgical dilation at first.

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Staff of Theatre Complex B of Colombo South Teaching Hospital

Informed consent

Informed written consent was taken from the patient for photographs and use of clinical records as appropriate.

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