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CASE REPORT
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MAYER-ROKITANSKY-KUSTER-HAUSER SYNDROME (MRKH) PATIENT TREATED WITH LAPAROSCOPIC DAVYDOV PROCEDURE AT LIAQUAT NATIONAL HOSPITAL, KARACHI

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Abstract
Mayer-Rokitansky-Kuster-Hauser (MRKH) syndrome, characterized by congenital absence of the vagina and uterus, poses significant challenges to affected individuals, particularly in terms of fertility and sexuality. We present a case of a 20-year-old woman diagnosed with MRKH syndrome who underwent laparoscopic Davydov's vaginoplasty at Liaquat National Hospital, Karachi. The patient presented with primary amenorrhea and a blind-ended vagina, with normal external genitalia and well-developed secondary sexual characteristics. Surgical intervention was chosen after thorough counseling, and the Davydov procedure was performed successfully with minimal intraoperative complications. Postoperative care involved self-dilation with Hegar dilators, leading to satisfactory neovaginal length and function. Compared to traditional methods like the McIndoe procedure, the Davydov technique offers advantages such as fewer postoperative complications and a simpler surgical approach. Our case underscores the importance of individualized treatment strategies and multidisciplinary care in managing MRKH syndrome, with laparoscopic Davydov's vaginoplasty emerging as a promising option for vaginal reconstruction. Long-term follow-up is essential to assess the procedure's efficacy and patient satisfaction.

Keywords: Mayer-Rokitansky-Kuster-Hauser Syndrome, Laparoscopic Davydov Procedure, Vaginal Reconstruction, Congenital Vaginal Agenesis, MRKH Syndrome Treatment

Introduction
Congenital absence of vagina and uterus has been eponymously called Mayer-Rokitansky-Kuster-Hauser (MRKH) syndrome. First described in 1829 by August Franz Joseph Karl Mayer, this syndrome affects approximately 1 in 4,500 female births, making it a relatively rare condition [1]. Characterized by the absence or underdevelopment of the müllerian duct structures, including the uterus, cervix, and upper two-thirds of the vagina, MRKH syndrome often manifests clinically with primary amenorrhea in otherwise phenotypically normal females [2].
The vagina may appear as a dimple with the presence of rudimentary uterine primordia and normal functioning ovaries. Patients present with primary amenorrhea, normal external genitalia, and well-developed secondary sexual characteristics. Davydov’s colpopoiesis is one of the methods of vaginoplasty using the patient’s own peritoneum as a graft to line the neovagina. Despite normal

secondary sexual characteristics and functioning ovaries, the absence of a vagina poses significant psychosocial and physiological implications for affected individuals. Infertility and the inability to engage in penetrative sexual intercourse can lead to profound psychological distress and diminished quality of life [3]. Therefore, the management of MRKH syndrome requires a comprehensive approach that addresses both the anatomical defect and the psychological impact on the patient.

Various surgical and non-surgical techniques have been developed for vaginal reconstruction in MRKH syndrome, aiming to restore sexual function and alleviate psychological distress. Among these, the Davydov procedure, first described in 1974 by Dr. Sergei Davydov, has gained attention for its effectiveness and favorable outcomes [4]. By utilizing the patient's own peritoneum as a graft material for neovaginal construction, the Davydov procedure offers advantages such as reduced risk of graft rejection and improved long-term functional outcomes.

This case report presents a patient with MRKH syndrome who underwent laparoscopic Davydov's vaginoplasty at Liaquat National Hospital, Karachi.

Case Presentation

A 20 years old female married for 1.5 years diagnosed case of Mayer-Rokitansky-Kuster-Hauser (MRKH) syndrome presented with complain of unable to consume her marriage. On general physical examination young female of average built and height with normal gait and her vitals were stable. Secondary sexual characteristic was appropriate for age. There was no palpable mass per abdomen and no inguinal swellings. Gynecological examination revealed normal looking vulva and per vaginal examination a blind ended vagina was found with patent external urethral meatus.

Laboratory evaluation revealed a normal hormonal assay. Ultrasound revealed a small hypoechoic tubular area noted in posterior to bladder measures 3.0 x1.4x1.9cm findings are suggestive of hypoplastic uterus. Right ovary measures 2.1x1.7cm and there is right para ovarian cyst measures 2.2x1.8cm, left ovary was normal looking with no associated abnormality with renal system. MRI findings of Figure 1a and 1b shows two rudimentary uterine horns are seen laterally in pelvis adjacent to both ovaries connected by a thin fibrous band.

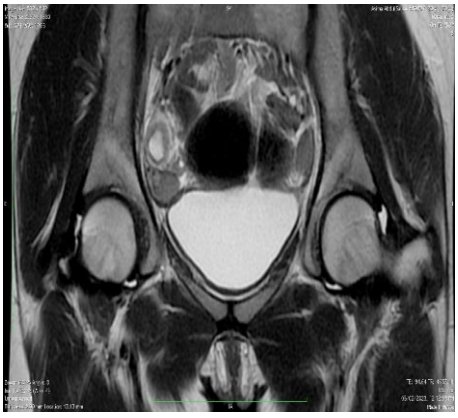

Figure 1a

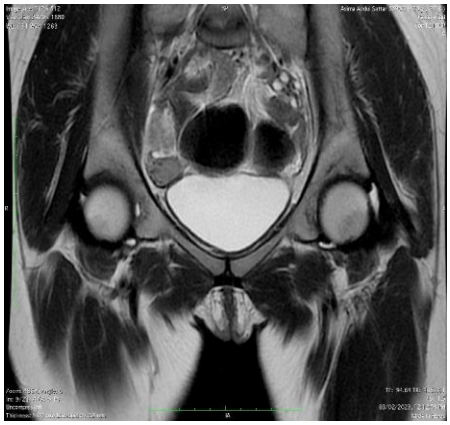

Figure 1b

Figures 1a and 1b both shows normally functioning bilateral ovaries with two rudimentary uterine horn seen laterally in pelvis adjacent to both ovaries

Figure 3 showing cervico vaginal agenesis. Normally functioning bilateral ovaries with multiple follicles. Finding are likely due to Mayer-Rokitansky-Kuster-Hauser (MRKH) syndrome with bilateral rudimentary uterine horns. The patient and her family counselled regarding MRKH syndrome and all the treatment options along with fertility issues were discussed. After counselling and consent, laparoscopic davydov’s vaginoplasty was planned.

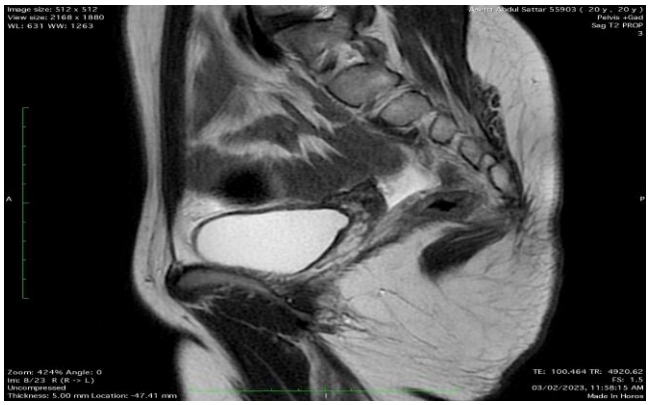

Figure 3: Cervicovaginal Agenesis

Surgical Technique

Preoperative preparation was done which include prophylactic antibiotics. Under General anesthesia she was placed in lithotomy position. Under the laparoscopic view both ovaries and tubes were normal looking. Rudimentary uterus was seen. Supravesical peritoneum along with bilateral rudimentary uterus is dissected from bladder. The peritoneum of pouch of Douglas is incised transversely and mobilized to create the posterior peritoneal flap, Dilator was inserted through vagina Transverse incision below the uterine strand given that serves as opening in Neovaginal apex. Edges of anterio`r and posterior peritoneal flaps pulled through newly created canal under laparoscopy assistance. Posterior flaps are sutured to posterior mucosa and both flaps of peritoneum sutured vaginally by assistant surgeon. The neovaginal apex was supported by suturing supravesical and suprarectal peritoneum along with lateral abdominal wall peritoneum. No bleeding seen at the end. After the final inspection through the laparoscope all ports were removed and the laproscopy incision was closed.

Postoperative Care

On the 3rd postoperative day, examination of neovagina was done. Self-dilation, three times a day (20-30 mins) using lubricated Hegars dilators was advised to patient. Postoperative period was uneventful and the patient was discharged on 4th day with an achieved neovaginal length of 6 cm and 2 finger breadths.

Follow Up

Clinical follow-up planned on 10th postoperative day, revealed patent neovagina of length 6 cm. The next follow-up was scheduled at 3 months' post-surgery**.**

Discussion

Mayer-Rokitansky-Kuster-Hauser (MRKH) syndrome stands as the second most common cause of primary amenorrhea due to vaginal agenesis, presenting a significant challenge in gynecological practice [1]. The absence of a vagina not only impacts fertility but also profoundly affects the sexuality and psychosocial well-being of affected individuals. Psychosocial counseling and support are crucial components of the management of MRKH syndrome, ensuring comprehensive care that addresses both the physical and emotional aspects of the condition [2].

Various surgical and non-surgical methods have been developed for vaginal reconstruction in MRKH syndrome, aiming to restore sexual function and alleviate psychological distress. The choice of procedure must be individualized, taking into account factors such as the patient's needs, surgeon's expertise, previous treatment attempts, and patient compliance [3]. While non-surgical methods like the Frank and Ingram dilation method remain the first-line intervention due to lower complication rates, surgical options offer more definitive solutions [5].

Among surgical techniques, the McIndoe procedure, utilizing split-thickness skin grafts, has been a longstanding method for neovaginal construction. However, its disadvantages, including visible scarring and potential complications such as fistula formation and vaginal stenosis, have led to the exploration of alternative approaches [6]. The Davydov procedure, performed laparoscopically using the patient's peritoneum as a graft material, has emerged as a promising alternative. Compared to the McIndoe technique, Davydov's operation offers advantages such as fewer postoperative complications, quicker recovery, and satisfactory cosmetic outcomes [7].

In our case, the laparoscopic Davydov procedure was chosen due to its simplicity, safety, and effectiveness in creating a neovagina for the patient with MRKH syndrome. The postoperative period was uneventful, with successful self-vaginal dilation leading to the achievement of a good anatomical length. Long-term follow-up is essential to assess the durability and functionality of the neovagina.

Conclusion

Techniques such as the laparoscopic Davydov procedure show promise in neovaginal reconstruction, enhancing sexual function and overall well-being for MRKH syndrome patients. Effective communication and emotional support are pivotal throughout the treatment journey. While non-surgical methods like vaginal dilation remain initial options, laparoscopic techniques like the Davydov procedure offer safe and effective solutions. Success hinges on meticulous surgical technique, postoperative care, and patient compliance. Alternative fertility options such as adoption and surrogacy may be considered. While uterine transplantation holds potential, its feasibility remains under scrutiny. As advancements continue, a holistic approach remains paramount in addressing the complexities of MRKH syndrome.

Human Ethics

Consent was obtained or waived by all participants in this study.

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