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Successful Treatment of Rare Disorder Mayer-Rokitansky-Küster-Hauser Syndrome: 3 Years Follow Up And Reproductive Outcome Case Report

Tamaz Nikolaishvili[1]; David Gagua[2]; Tinatin Gagua[2]*; Beka Metreveli[2]; Aleksandra Gvenetadze[2]

[1]Gagua Clinic: Chiaureli 6a Tbilisi Georgia

[2]David Tvildiani Medical University. Lublian 2/6 Tbilisi Georgia.

***Corresponding Author(s): Tinatin Gagua**

David Tvildiani Medical University. Lublian 2/6 Tbilisi Georgia.

Email: tinatingagua@gmail.com

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Background

Müllerian agenesis, Mayer-Rokitansky-Küster-Hauser Syndrome (MRKH), or vaginal agenesis, has an incidence of 1 per 4,500–5,000 females. It is caused by embryologic underdevelopment of the Müllerian duct, causing agenesis or atresia of the vagina, uterus, or both [1]. The prevalence in Georgia is not known. Although the vast majority of cases of MRKH seem to be sporadic some familial link has been reported with the use of ART [2].

Various forms of Müllerian abnormalities range from minor anatomical variations to total aplasia. Women with MRKH syndrome show 46XX karyotype, normal external genitalia, developed lower two-thirds of the vagina and functional ovaries and fallopian tubes due to different embryogenic origin. MRKH types are typical (type 1 or isolated) and atypical (type 2). Usually, only the caudal part of the Mullerian duct is affected. Type 2 is more frequent and involves renal defects (unilateral agenesis or ectopia of one or both kidneys, horseshoe kidney, in about 40–60% of patients), cervicothoracic (asymmetric, fused or wedged vertebrae, scoliosis, and Klippel-Feil anomaly in about 20% of patients) and, to a minor extend, hearing defects and digital abnormalities of varying severities [2].

Patients with mulerian anomalies are usually present with primary amenorrhea. Differential diagnoses include obstructive anomalies (distal vaginal atresia cervical atresia, imperforate hymen, distal vaginal atresia, transverse vaginal septum, and cervical atresia), androgen insensitivity syndrome, Turner syndrome, Sawyer syndrome, and adrenogenital syndrome [1]. The two main challenges patients face with MRKH syndrome include engaging in adequate and satisfactory sexual intercourse and procreation. We present the MRKH syndrome case man-

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aged at the Department of Obstetrics and Gynecology of Davit Tvildiani Medical University Hospital Tbilisi, Georgia. The successful technique of vaginoplasty from the perspective of sexual life and reproduction.

Case Report

A patient 27-year-old woman presented at the clinic with a chief complaint of inability to perform vaginal intercourse. The patient was previously evaluated at 17 due to primary amenorrhea. Her adrenarche and telarche were normal, and she developed typical secondary sexual characteristics at 14. After investigation, a diagnosis of MRKH type 2 was made. At the time patient chose not to undergo any treatment due to psychological stress and no definite outcomes; when she was referred to our clinic for medical advice at the age of 27, after discussing nonsurgical and surgical options, the Laparoscopic Davydov method was chosen to create the neo vagina. The vulva was normal, with no entrance to the vagina.

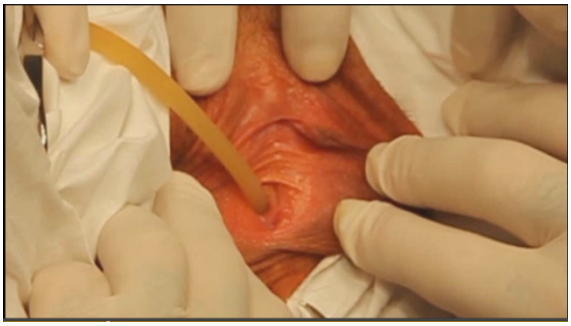

Figure 1: The initial condition.

The operation was carried out under general anesthesia. Four laparoscopic ports were inserted for abdominal access. Four-centimeter incision was made at the vaginal vestibule, and with blunt dissection, the space was created between the rectum and bladder; the damage to both organs was avoided using a Laparoscopic view. Using Laparoscopic accesses corroborating incision was made on the peritoneum behind the bladder (Douglas pouch); three pieces of peritoneum were prepared and sewn into the newly constructed channel between the bladder and rectum, creating walls for neo vagina. Purse stitching was performed from the abdominal side to secure the length of the neo-vagina. Finally, a dilator was inserted inside the newly created vagina. Forty-eight hours after the operation, the dilator was removed.

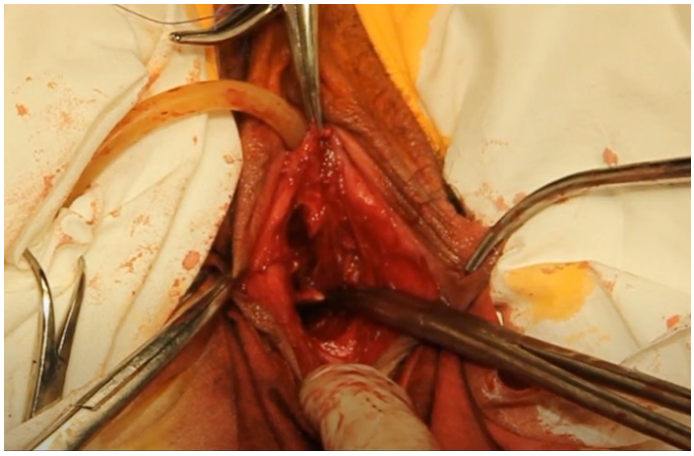

Figure 2: Construction of hanel between bladder and rectum.

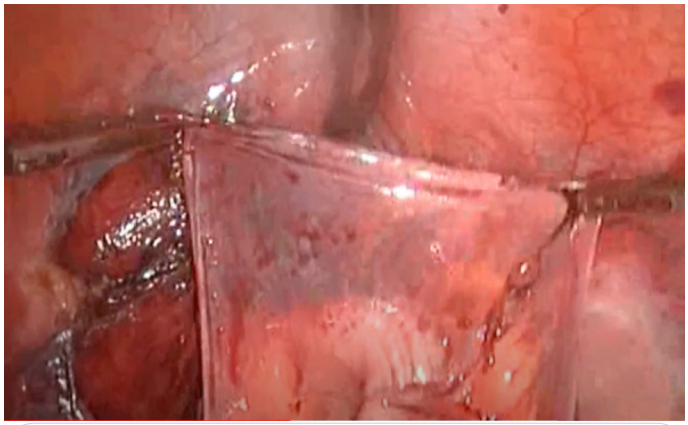

Figure 3: Peritenoum dissection.

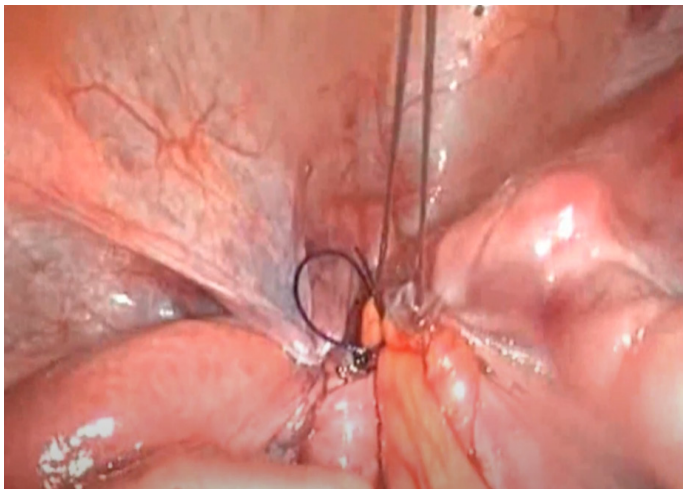

Figure 4: Creation walls for neo vagina.

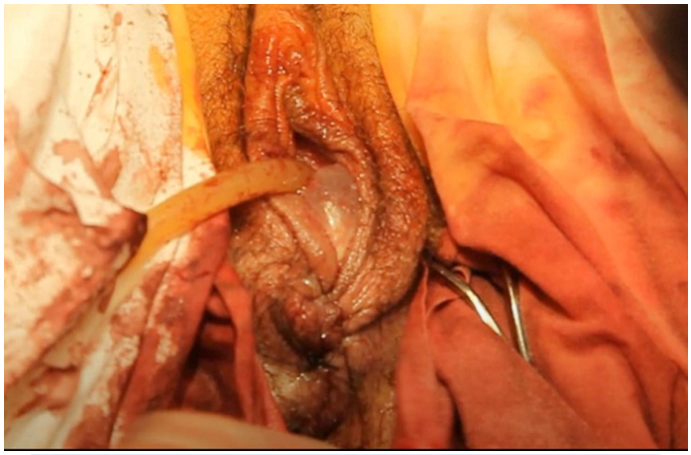

Figure 5: Follow up visit after 1 month.

The patient was discharged from the clinic after three days of observation without complications. She was advised to use a dilator for at least a month, 6 hours a day, and was taught how to insert it correctly. At the evaluation, the neo-vagina was covered with epithelium three months after the operation. The patient reported no discomfort during intercourse.

Patient-reported for evaluation three years after the operation as she was getting ready for Assisted reproductive technology- egg retrieval, fertilization, and surrogacy pregnancy. She reported no discomfort during intercourse and even remarked that the quality of her life had significantly increased after surgery, and she felt fulfilled as a woman.

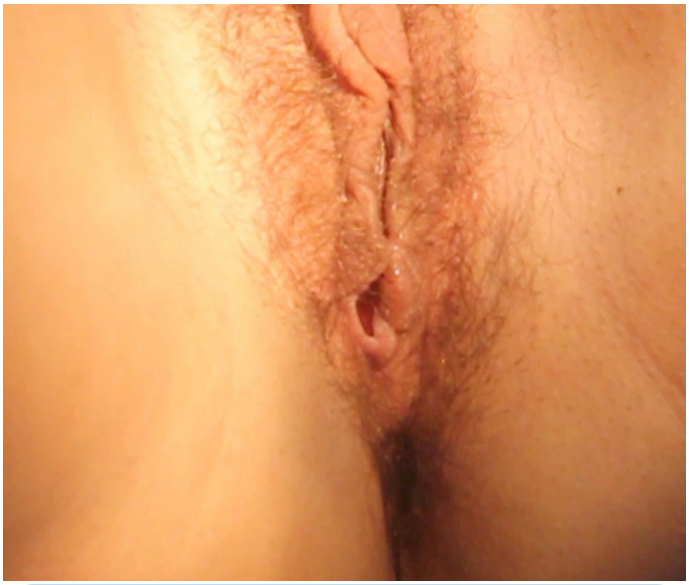

Figure 6: Follow up visit after 1 month.

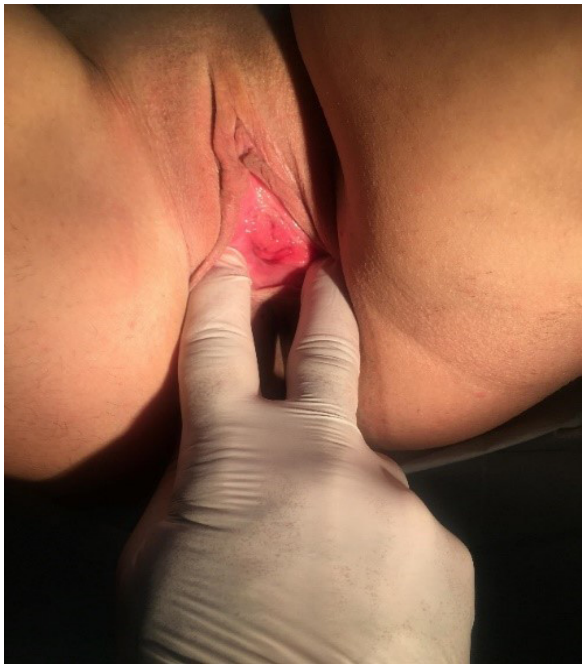

Figure 7: Follow up visit after 3 years.

Discussion

Mayer-Rokitansky-Küster-Hauser syndrome, characterized by vaginal and uterine dysgenesis, can be a psychologically traumatizing diagnosis for a woman. In addition, the inability to perform sexual intercourse and conceive can be stigmatizing since the awareness and information on the syndrome is not readily available outside the medical community.

Since various techniques for creating the Neo vagina exist today, discussing with patients and considering their wishes is especially important. In the case mentioned earlier time limit was crucial. Given the patient's age and unwillingness to perform dilatation, preference was given to surgical treatment [5].

The goal of surgical treatment of MRKH is the creation of a neo vagina with enough depth and lubrication to support regular sexual intercourse. Various methods of surgical techniques exist today. Intestinal vaginoplasty has been previously successfully used, Mc Indoe (split-thickness skin grafts) procedure and conventional or laparoscopic Veicchetti traction procedure. These methods have various advantages and disadvantages; choosing the right one for the patient requires extensive discussion of each of them and giving maximum freedom of choice to the patient. Surgical expertise must be taken into consideration while choosing the best option [7].

The Davydov method of creating neo vagina (The Davydov coreopsis) with Laparoscopic assistance minimizes intraoperative bleeding, availability of abdominal view further assists with the protection of the Urethra, Bladder, and Rectum from iatrogenic trauma, limits discomfort, and hospital stay during recovery time. Neo vaginal walls created by peritoneal flaps can successfully lubricate, do not form painful scars, and support satisfactory intercourse [8, 9].

Conclusion

In the surgical treatment of MRKH – Mullerian agenesis patient's wishes, support system, and psychological state are crucially important. Open discussion and expectation management are essential for successful treatment. The Laparoscopic Davydov method can be a fast, cost-effective, and safe surgical treatment vaginal aplasia.

Written informed consent was obtained from the patient for the publication of this case report and accompanying images. A copy of the written permission is available for review by the Editor-in-Chief of this journal on request.

References

1. ACOG Committee on Adolescent Health Care Number. Müllerian Agenesis: Diagnosis, Management, and Treatment. ACOG. 2018.
2. Guerrier D, Mouchel T, Pasquier L, Pellerin I. The Mayer-Rokitansky-Küster-Hauser syndrome (congenital absence of uterus and vagina) – phenotypic manifestations and genetic approaches. 2006.
3. Fedele L, Frontino G, Restelli E, Ciappina N, Motta F, et al. Creation of a neovagina by Davydov's laparoscopic modified technique in patients with Rokitansky syndrome. Am J Obstet Gynecol. 2010.
4. Robson S, Oliver GD. Management of vaginal agenesis: review of 10 years practice at a tertiary referral centre. Australian and New Zealand Journal of Obstetrics and Gynaecology. 2000; 40: 430-433.
5. Kölle A, Taran F-A, Rall K, Schöller D, Wallwiener D, et al. Neovagina creation methods and their potential impact on subsequent uterus transplantation: a review. BJOG. 2019; 126: 1328-1335.
6. Michala Michala LL, Cutner Cutner A, Creighton SS. Surgical approaches to treating vaginal agenesis. BJOG: Int J Obst Gynaecol. 2007; 114: 1455-1459.
7. Takahashi K, Nakamura E, Suzuki S, Shinoda M, Nishijima Y, et al. Laparoscopic Davydov procedure for the creation of a neovagina in patients with Mayer-Rokitansky-Kuster-Hauser syndrome: analysis of 7 cases Tokai. J Exp Clin Med. 2016; 41: 81-87.
8. Samantray SR, Mohapatra I, Harshini N. Laparoscopic Davydov's Colpopoiesis for a Case of Mayer-Rokitansky-Kuster-Hauser (MRKH) Syndrome. Cureus. 2021; 13: e13974. 2021.