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Neovaginoplasty with Isoperistaltic Vascularized of Sygma With Laparotomy

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Abstract

In this paper, we present three patients with Mayer-Rokitansky-Kuster-Hauser syndrome (MRKH), which is a congenital anomaly that clinically presents as aplasia of the vagina and uterus. After the correct diagnosis, these patients were operated on with the open surgical method for the creation of a neovagina with a vascularized and isoperistaltic segment of the sigmoid. Their postoperative follow up has been satisfactory. With the surgical treatment, in all three patients the cosmetic aspect and sexual functionality were achieved, as the final goal of creating the neovagina.

Key words: Mayer-Rokitansky-Kuster-Hauser syndrome, sigmo-neovaginoplasty, laparotomy

Introduction

Mayer-Rokitansky-Küster-Hauser syndrome (MRKH syndrome) is a congenital anomaly that presents clinically as aplasia of the vagina and uterus (Figures 1 and 2). This congenital anomaly results from the dysgenesis of the Müllerian ducts during embryogenesis. Statistically, it is reported in 1: 5000 female births.¹ The syndrome is known based on reports by Mayer (1829), von Rokitansky (1838), Küster (1910) and Hauser (1971).^{1,2} In medical practice, it is commonly known as Rokitansky syndrome.

In most cases, the diagnosis of this syndrome in teenage girls is made during puberty, usually from the first visit to the doctor, due to primary amenorrhea, despite the appearance of secondary sexual physical

characteristics. Examination reveals the complete absence of the vagina or only as a skin deepening of 1-3 cm (Figure 1). In digital rectal examination, an empty pelvis can be found due to the absence of the vagina and uterus. Sometimes, during the digital rectal examination, the presence of a cherry size rudimentary uterus can be ascertained. This condition can be accompanied by aplasia or hypoplasia of the tube of one or both sides. The ovaries may be present, slightly elongated and drawn high towards the linea terminalis (Figure 2). Diagnosis of Rokitansky syndrome is confirmed by ultrasound, MRI and diagnostic laparoscopy. Rarely, this syndrome is accompanied by testicular feminization, which is confirmed by ovarian biopsy and chromosome karyotype (46 XY). Therefore, karyotyping should be done in genital aplasia. In most cases, the hormonal profile of patients is normal and is accompanied by secondary sexual characteristics. It is well known that Rokitansky syndrome, in 40-60% of cases, is also associated with abnormalities

of the urinary system, such as: aplasia of one kidney, horseshoe-shaped kidney, pelvic kidney, pyelonephritis and ureter fissus, while pelvic skeletal abnormalities are associated in 10-12%.¹⁻³

As soon as Rokitansky syndrome is confirmed in the patient, she is notified about the nature of the disease, as a congenital anomaly – i.e. that her vagina and uterus are missing, and therefore she will not menstruate, and cannot get pregnant. This often comes as a shock to her and her partner and to the rest of the family. It usually falls upon the gynecologist to inform the patient with further professional clarifications. The gynecologist also explains it to the patient that the uterus cannot be reconstructed but he also tells her about the possibility of reconstructing the vagina, to complete allow sexual intercourse. This reconstruction of the vagina can be completed with the patient's own surrounding vital structures. She should be informed about a number of possibilities and modalities of this reconstruction, after which she can have a normal sexual life.



Figure 1: External view of the genital region showing the absence of the vagina

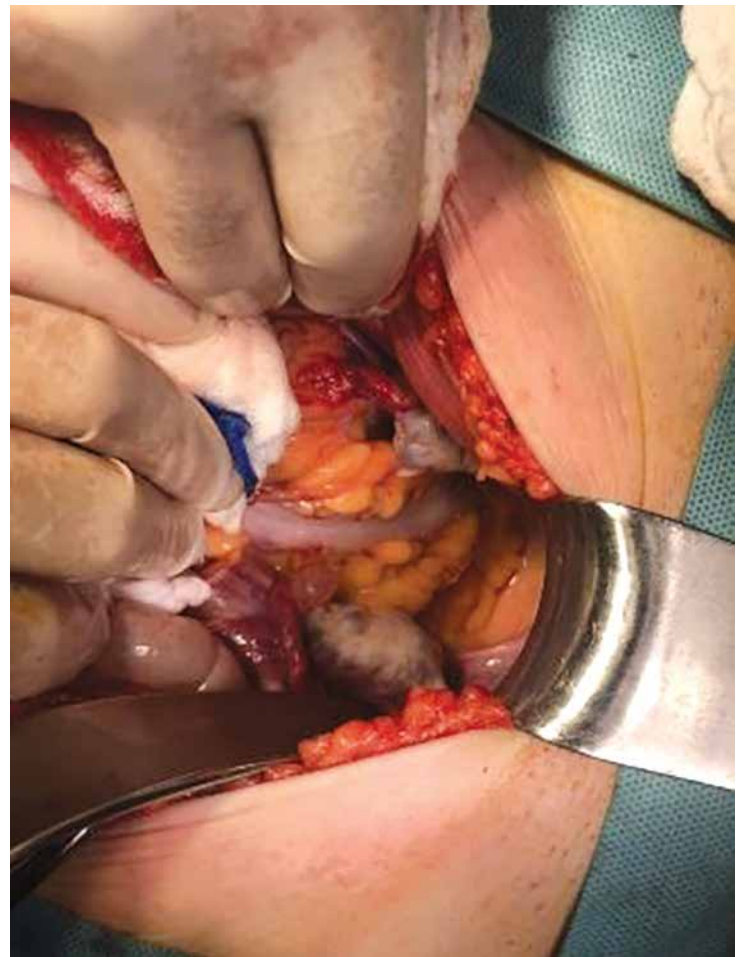


Figure 2: Intraoperative view of Mayer-Rokitansky-Kuster-Hauser

The purpose of neovaginoplasty is the creation of a new vagina (neovagina) from its own structures, to enable intercourse. The personal motivation and emotional maturity of the patient are the main indications for neo-vaginoplasty. Sometimes, it is necessary to consult a psychiatrist and psychologist to assist the patient in managing the mental and emotional stress.²

Cases Presentation

In this paper, we present three patients with Rokitansky syndrome (MRKH), as a congenital anomaly. All three patients were previously diagnosed by a gynecologist outside our clinic. They did not have any concomitant diseases or addictions. Before the surgery, there were gynecological visits, ultrasound examination of the abdomen and pelvis, intravenous urography, a visit to the anesthesiologist and the opinion of a clinical psychologist. All of these various sources of information were taken into account. There was no evidence from a karyogram, but the hormonal profile and secondary sexual characteristics were typically female. They were in a good mental state and they had a strong motivation for surgical correction of this anomaly, in order to regulate their sexual activity.

Methods and possibility of surgical intervention were explained to the patients in detail. Possible complications during and after surgery and general anesthesia were also explained. After the necessary clarifications, the patients willingly signed the consent for reconstructive surgical intervention. During the operation, the ovaries with signs of folliculogenesis and positioned high above the terminal line were found in all three patients (Figure 2). The surgical intervention lasted on average 2 hours and 15 minutes. The postoperative progress in all three patients went without any complications.

Surely, after the intervention, they were treated with infusions, antibiotics, analgesics, antithrombotics, etc. The urinary catheter was kept for six days. The vaginal gauze pad, soaked with betadine and saline solution, was kept for two days. The hospital stay was nine days. During this time, the necessary laboratory analyses were carried out. After discharge from the hospital, the patients came for scheduled check-up visits. One month after the intervention, the patients started local neovagina dilation exercises with adequate silicone prostheses.

Surgery Technique

The operation was carried out by a mixed operating team (surgeon and gynecologist).

The patient was first put under general endotracheal anesthesia, and the urinary bladder was catheterized. The procedure began with a medial infraumbilical laparotomy, allowing further verification of Rokitansky syndrome. The small intestines are packed above and to the right of the abdominal cavity and the sigmoid colon is well exposed. The sigmoid is mobilized, carefully preserving its vascularization. At least 2 sigmoid arteries are carefully followed, identified and preserved with the accompanying veins of the sigmoid segment. After the mobilization of the sigmoid, the surgeons tested whether its mobilization is sufficient in length to create a tension-free neovagina, without its segmental resection. Upon confirming that the length of the sigmoidal segment of 15 cm is sufficient for the creation of the neovagina, the sigmoidal segment is resected, maintaining its vascularization with at least 2 sigmoidal arteries. (Figures 3 and 4). Immediately afterwards, a continuous T-T sigmoid suture is done without tension, with stitches, PDS 3-0, (Figure 5). Then, the isolated and vascularized segment of the sigmoid, 15 cm long, is placed in the isoperistaltic position. The proximal opening of the sigmoid segment is closed (sewn) in two layers, while its distal part penetrates and is placed through the tunnel that was previously created through the muscles of the pelvic diaphragm, without a twist and without tension. At the end, the sigmoidal segment is fixed with special sutures in the vaginal introitus, to create a neovagina.

Since the vagina is missing, as per Rokitansky syndrome, there are no natural holes in the pelvic diaphragm, no window through which the sigmoidal segment can be penetrated to create a neovagina. It is then required to first tunnel the pelvic diaphragm, through which the isolated sigmoid segment will be penetrated and placed. Tunneling of the pelvic diaphragm must be done very carefully, combining actions from the perineal and abdominal side. During this procedure, the urethra, urinary bladder or rectum can be injured very easily, since there is not much maneuvering space during the creation of the window. After the opening and tunneling of the pelvic diaphragm, the distal part of the sigmoid segment is inserted and pulled through it, without twisting, and with special pins without tension, it is fixed in the



Figure 3: Sigmoidal segment

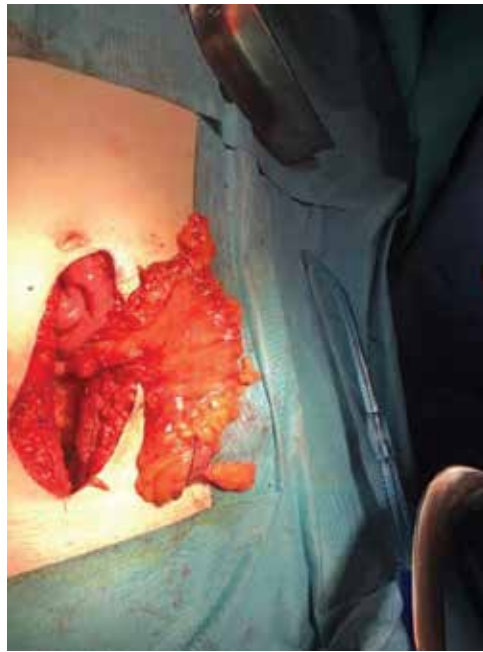


Figure 4: Preservation of vascularization



Figure 5: T-T sigmoid anastomosis

vaginal introitus with special sutures. This completes the creation of the neovagina. Now the neovagina is washed with a warm physiological solution, and vaginal packing is done with gauze soaked with physiological solution and betadine solution. The abdominal cavity is washed out, the Douglas drain is placed and the operative wound is closed in layers.

Discussion

Considerable efforts by surgeons over a long period of time were needed to develop a surgical method to create a neovagina from nearby body structures and to enable a normal sexual life of women with Rokitansky syndrome. The first attempt to create a neovagina was made by Dupuytren, in 1817. After this attempt, many operative methods and techniques have been developed over the 2 centuries, with less invasive methods, such as expanding and pushing back the urogenital sinus with a prosthesis (Frank), to more invasive methods of creating a neovagina, including operative methods very complex such as: skin transplantation and interposition creating a neovagina with vulvo vaginoplasty with the labia of the vulva.^{2,5,6} There were attempts to create a neovagina with amniotic membranes.^{3,5} After these methods, the creation of a neovagina with a rectosigmoid has been tried.² There are authors who created the neovagina with the urinary bladder.²

As in many other surgical branches, the assisted laparoscopic method⁴ is increasingly being used, due to the benefits of a minimally invasive approach. For this purpose, the creation of a neovagina with a sheet of pelvic peritoneum is now very often used.³⁻⁵ Davydov's method dates back to 1969, while the results of this method were followed up and published after 20 years. With the introduction of the endoscopic surgical method, Davydov's method has undergone modifications, in the sense of the assisted laparoscopic method. In Davydov's method, the final length of the neovagina reaches 10-15 cm.³

During a 20-year period, the author Davydov, apart from the results, has also detailed several complications: 4 wound infections, 3 thrombophlebitis, 11 subileus/ileus, 2 pelvioperitonitis, 14 cases of prolonged febrility and 18 cases of cystitis.³ Adamyanova, gives priority to the laparoscopic method over the conventional one, in 223 cases she worked on.⁵ French authors report two complications: migration of the intraperitoneal neovaginal prosthesis and a vesico-vaginal fistula, which was treated laparoscopically.⁶

Various authors also present other complications, such as: rectovaginal fistula, bleeding, pain during intercourse in the first 3 months, which tend to disappear later. Most authors present the neovaginal length as final at 7.6 cm. and that the length tends to decrease on average to 6.2 cm. In spite of this, some authors in the follow-up

of 18 presented cases, did not find a reduction of the neovagina length.⁶ There are other methods as well for creating a neovagina: with the right colon, with the small intestine, with the urinary bladder, the labia majora of the vulva, etc. Recently, the creation of a neovagina with the peritoneal sheet of the pelvis has become common and widely used, with the assisted laparoscopic method according to Davydov, which has its own advantages because it shortens the operative time, the minimally invasive method and the good aesthetic effect.³

In our three cases, the open surgical method was used. This was done for the following reasons:

1. Our experience and greater safety with the open method in the preparation of the vascularized sigmoidal segment.
2. Higher safety of manual closure of sigmoid T-T anastomosis
3. Higher safety in fenestration and tunneling of the pelvic diaphragm, etc.

Early postoperative results in the three operated cases have been satisfactory. The patients had no

wound infection, no urinary infection, no obstructions in defecation. After the hospital treatment, the patients came for check-up visits to monitor the condition which was satisfactory at all times. One month after the surgical intervention, we began with the dilation of the neovagina with a silicone prosthesis, with increasing thickness. Initially, the demonstration of dilation was done in the hospital by the doctor-nurse, and later the patient continued for a few minutes every day at home (Figures 6 and 7).

In the first two operated patients, the sexual active life started after three months. The third patient is waiting for the connection, because the postoperative dilation of the neovagina at the level of the pelvic diaphragm has stalled, as a result of the patient's own irregular and insufficient dilations. In addition to the visual aesthetic effect of the introitus (Figure 8 and 9), and the resultant neovaginal depth and dimensions were satisfactory in all cases (Table 1). The possibility of surrogate pregnancy, with the partners' genetic material, remains a possibility.⁶



Figure 6: Penetration with silicone prosthesis



Figure 7: Depth of penetration

Date of surgery	Age	Profession	Dimensions of the Neovagina	Dimensions after 5 months
28 Sep. 2021	35	housewife	14 cm x 4 cm	15 cm x 5 cm
28 Oct. 2021	26	cashier	15 cm x 4 cm	15 cm x 5 cm
11 Jan.2022	32	housewife	13 cm x 3.5 cm	14 cm x 4.5 cm

Table 1: Neovaginal depth and dimensions



Figure 8: Postoperative abdominal skin scar

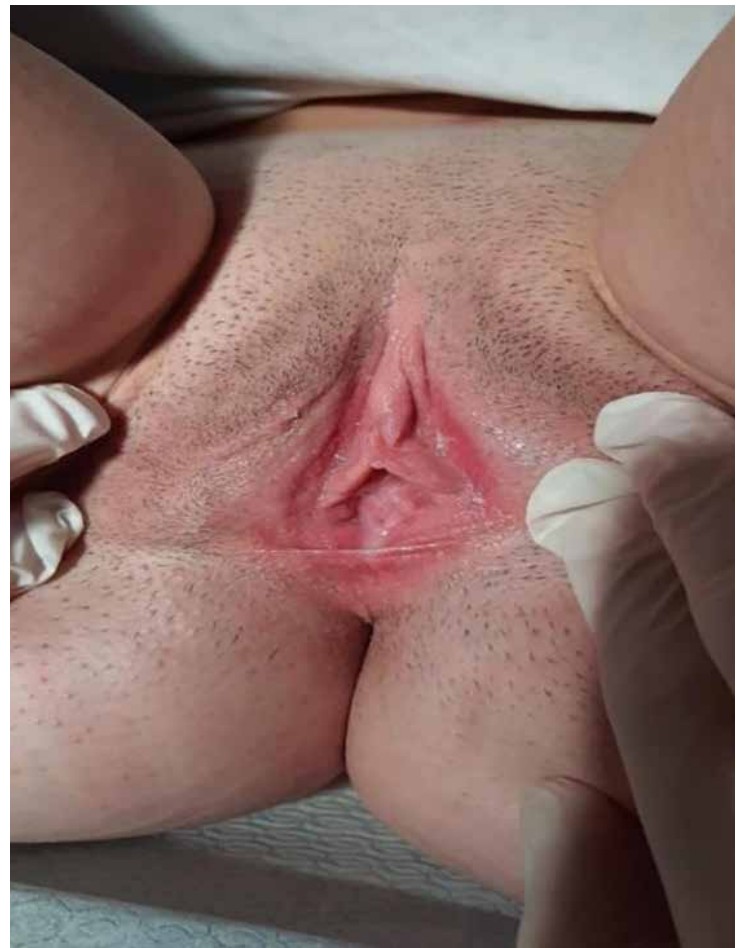


Figure 9: Postoperative view of neovagina

CONCLUSIONS

The benefits for the patient after successfully performing vaginoplasty are, firstly that sexual relationships are enabled; secondly, it can improve the mental and emotional stability of the patient and allow a kind of spiritual and social rehabilitation.

Out of many possibilities, we chose the reconstructive method which can be carried out competently. Future goals are for the reconstruction of the neovagina to be done with the minimally invasive method, according to Davydov, i.e. modification with the assisted laparoscopic method, according to Freidberg.⁴

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