

Open Sigmoid Neovaginoplasty

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Received: 02 May 2025; Accepted: 28 May 2025; Published: 03 June 2025

Citation: Ismet Shaqiri, Rifat Bajrami, Naim Shabani, et al. Open Sigmoid Neovaginoplasty. Surg Res. 2025; 7(1): 1-5.

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, sexual functionality and cohabitation was achieved, as the final goal of creating the neovagina.

Keywords

Mayer- Rokitansky- Kuster- Hauser syndrome, Sigmoid-neovaginoplasty, Laparotomy.

Introduction

Mayer-Rokitansky-Küster-Hauser syndrome (MRKH syndrome) is a congenital anomaly that is clinically presented as aplasia of the vagina and uterus (Figure 1,2). This congenital anomaly results from the dysgenesis of the Müllerian ducts during embryogenesis. Statistically, it is reported in 1: 5000 female births. The syndrome is known based on the authors Mayer (1829), von Rokitansky (1838), Küster (1910) and Hauser (1971). In medical practice, it is commonly known as Rokitanski 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 signs: breast growth, the appearance of female-type hairiness and beautiful modeling of the body with feminine features.

Examination reveals the complete absence of the vagina or only as a skin deepening of 1-3 cm (Figure 1). In the digital rectal examination, an empty pelvis is found due to the absence of the vagina and uterus.



Figure 1: The vagina is missing.

Sometimes, during the digital rectal examination, the presence of a cherry size rudimentary uterus can be ascertained. Sometimes this condition can be accompanied by aplasia or hypoplasia of the tube of one or both sides. The ovaries are present, slightly elongated and drawn high towards the linea terminalis (Figure 2). Confirmation of Rokitanski syndrome is completed 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 Rokitanski syndrome in 40-60% is also associated with abnormalities of the urinary system, such as: aplasia of one kidney, horseshoe-shaped kidney, pelvic kidney, pyelonephritis and ureter fensus, while pelvic skeletal abnormalities are associated in 10-12% [1-3].



Figure 2: Uterus is missing.

As soon as Rokitanski syndrome is confirmed in the patient, she is notified about the nature of the disease, as a congenital anomaly. It is explained to her that her vagina and uterus are missing, and as a result she will not have her periods, cannot get pregnant and that she cannot give birth to a child. This revelation 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 and he also tells her about the possibility of reconstructing the vagina, to complete sexual cohabitation. 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 - cohabitation. The possibility of surrogate pregnancy, with the partners' genetic material, remains a possibility.

The purpose of neovaginoplasty - is the creation of a new vagina - neovagina, from its own structures, to enable cohabitation. The personal motivation and mental maturity of the patient for sexual relationship-cohabitation is the only indication for neovaginoplasty. This surgery is not recommended before the age of 20 because of the psychological maturity of the patient. Sometimes, it

is necessary to consult a psychiatrist and psychologist in order for the patient to cope with mental and emotional stress.

Cases Presentation

In this paper, we present three patients with Rokitanski syndrome (MRKH), as a congenital anomaly. Patients with Rokitanski-MRKH syndrome were diagnosed outside our clinic.

They came to us to perform the surgical intervention.

All three patients operated on with MRKH syndrome 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 was taken into account. There was no evidence of a karyogram, but the hormonal profile and secondary sexual characteristics were typically female. They were in 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 has been explained to the patients in details. Possible complications during and after surgery and general anesthesia were also explained to them. 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, is kept for two days. The hospital stay was nine days. During this time, the necessary laboratory analyzes 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 by a mixed operating team (surgeons and a gynecologist) was done. The patient is put under general endotracheal anesthesia. The urinary bladder is catheterized.

It begins with a medial infraumbilical laparotomy. The case of Rokitanski syndrome is verified once again. 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 two sigmoid arteries are carefully followed, identified and preserved with the accompanying veins of the sigmoid segment. After the mobilization of the sigmoid, it is tested whether its mobilization is sufficient in length to create

a tension-free neovagina, without its segmental resection. Once we find out 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. (Figure 3, 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 vaginal introitus with special sutures. This completes the creation of the neovagina. Now the neovagina is washed with 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.

In Table 1, we present the date of surgery, age, profession and dimensions of the neovagina for the 3 operated patients, at the beginning and five months after the surgery.

Table 1

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

Discussion

It has been a long period of time and numerous efforts by surgeons to find 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, starting with bloodless methods, such as expanding and pushing back the urogenital sinus with a prosthesis (Frank), to bloody methods of creating a neovagina, including operative methods very complex such as: skin transplantation and interposition (Küsner, Grossmann, Kirchner-Wagner, Lange, etc.), creating a neovagina with vulvo vaginoplasty with the labia of the vulva. Then there were attempts to create a neovagina with amniotic membranes, such as (Warton, Brindeau, Bürger, Williams, Davydov, Vecchieti, etc.). After these methods, the creation of a neovagina with a rectosigm has been tried (Schubert-Schmid, Wilfinger). There are authors who created the neovagina with the urinary bladder (D'Argent) [1,4,5].

As in many other surgical branches, the assisted laparoscopic method [4] is being used, where the minimally invasive approach is increasingly being used for the creation of the neovagina.

For this purpose, the creation of a neovagina with a sheet of



Figure 3: Sigmoidal segment.



Figure 4: Preservation of vascularization.



Figure 5: T-T sigmoid anastomosis.

pelvic peritoneum is now very often used (Davydov, Vecchietti). 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.

During a 20-year period, the author Davydov, apart from the results, has published several complications: 4 wound infections, 3 thrombophlebitis, 11 subileus/ileus, 2 pelvioperitonitis, 14 cases of prolonged febrility and 18 cases of cystitis.

A Russian gynecologist, Adamyanova, gives priority to the laparoscopic method over the conventional one, in 223 cases she worked on.

French authors report two complications: migration of the intraperitoneal neovaginal prosthesis and a vesico-vaginal fistula, which was treated laparoscopically.

Various authors also present other complications, such as: rectovaginal fistula, bleeding, pain during intercourse in the first 3 months, but 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 (Brun and colleagues) in the follow-up of 18 presented cases, did not find a reduction of the neovagina [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.

In our three cases, the open surgical method was used to **create neovaginoplasty**. As a justification we had:

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.
4. Good vaginal length and width
5. Natural lubrication
6. No/minimal need of dilators

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.



Figure 6: Penetration with silicone prosthesis.



Figure 7: Depth of penetration.

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, the cause of the patient's own irregular and insufficient dilations.

The visual aesthetic effect of the introitus has been achieved almost to perfection, (Figure 9)

Satisfactory neovaginal depth and dimensions (Table 1).



Figure 8: Postoperative cicatrix.

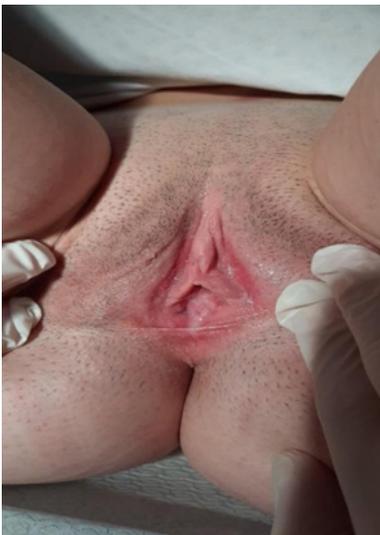


Figure 9: Perfect postoperative aesthetic effect.

Conclusions

The benefits for the patient after successfully performed vaginoplasty are: 1. Sexual cohabitation of partners is enabled; 2. It establishes the mental and emotional stability of the patient and a kind of spiritual and social rehabilitation.

Out of many possibilities, choose 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, respectively modification with the assisted laparoscopic method, according to Freidberg. The possibility of surrogate pregnancy, with the partners' genetic material, remains a possibility.

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