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Patient with Mayer-Rokitansky-Kuster-Hauser syndrome (MRKH) who Underwent Laparoscopic Davydov-Moore vaginoplasty

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Abstract

The Mayer-Rokitansky-Kuster-Hauser syndrome (MRKH) constitutes the second most common cause of primary amenorrhea, after gonadal dysgenesis. It is a congenital absence of the vagina with variable uterine development, which is a result of Mullerian duct agenesis or hypoplasia. The incidence for vaginal agenesis is 1 in 5000 (range 1 per 4000 to 10,000 females). We describe two cases of women diagnosed with MRKH syndrome who underwent laparoscopic Davydov-Moore vaginoplasty. Our video demonstrates the succesful operation which is composed of dissection of the rectovesical space, abdominal mobilization of the peritoneum to create the vaginal fornices, and attachment of the peritoneum to the introitus. Treatment of this syndrome should be multidisciplinary and individualized to the patient. It includes a combination of psychosocial support and correction of the anatomic abnormality.

Keywords: MRKH syndrome; Vaginal agenesis; Amenorrhea; Laparoscopic Davydov-Moore vaginoplasty

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Introduction

The Mayer-Rokitansky-Kuster-Hauser syndrome (MRKH) constitutes the second most common cause of primary amenorrhea, after gonadal dysgenesis [1]. It is a congenital absence of the vagina with variable uterine development, which is a result of Mullerian duct agenesis or hypoplasia. Variable uterine development can lead to uterine disorders from lateral hemi-uteri or uterine horns, a midline uterus without a cervix to absence of uterine structures at all. The incidence for vaginal agenesis is 1 in 5000 (range 1 per 4000 to 10,000 females) [2]. Differential diagnosis of vaginal agenesis comprises androgen insensitivity, low-lying transverse vaginal septum, agenesis of the uterus and vagina and imperforate hymen. In terms of ovaries, they may appear normal, extra-pelvic, unilateral or hypoplastic. In addition, these women often present extragenital anomalies, mostly connected with urinary system such as unilateral renal agenesis, pelvic or horseshoe kidneys, as well as irregularities of the collecting system [3]. Currently, underlying etiology remains unknown. The particular genetics is uncertain, but several surrogacy programs show that offspring of women with MRKH do not demonstrate defects. Lack of vagina can be diagnosed during self-examination or through gynecological evaluation. Usually there are no alarming symptoms until puberty when first menstruation (at 15 to 17 years of age) is expected. Amenorrhea, one of the primary presenting signs of Mayer-Rokitansky-Kuster-Hauser syndrome, should be differentiated with vertical fusion defect. Hypomenorrhea may occur with minimal functional endometrium where uterus is obstructed, but otherwise structurally normal, or in a presence of rudimentary uterine horn. Females with MRKH have normal 46XX karyotype with intact ovaries and ovarian function, therefore they develop secondary sexual characteristics (e.g., breast development, axillary hair, and pubic hair) except menarche which is absent in this case. During gynecological examination, the external genitalia appear normal. A vaginal dimple or small pouch with a hymenal fringe is usually present, since the vaginal pouch and hymen both derive from

the urogenital sinus. Rectoabdominal examination determines the presence or absence of midline structures. Ultrasonography demonstrates kidneys, confirms presence of the ovaries and absence of uterus. There are no official guidelines in management of MRKH-syndrome patients. However, treatment should be multidisciplinary to create functional vagina. One of methods is laparoscopic Davydov-Moore vaginoplasty.

Case Report

In this article, we present two cases of women with MRKH syndrome who underwent laparoscopic Davydov-Moore vaginoplasty.

In the first case, an 18-year-old female diagnosed with MRKH syndrome, came to Gynecological Endocrinology and Gynecology Clinic in Krakow. The aim of hospitalization was to create a functional vagina through Moore method in laparoscopy. In terms of extragenital anomalies, the patient reported history of spina bifida. She had atresia of oesophagus and underwent operation during neonatal period. She acquired secondary sexual characteristics at the age of 11. The gynecological examination revealed presence of hymen and vaginal dimple (0,5 centimeter deep). The transanal ultrasonography showed complete lack of uterus and normal, bilateral ovaries.

In second case, a 25-year-old woman with the diagnosed MRKH syndrome, presented to our clinic in order to create a neovagina trough Davydov-Moore method in laparoscopy. She denied any other problems with health and previous hospitalizations. Gynecological examination revealed vulva and perineum typical for nullipara. Also there was aplasia of vagina visualized with tight vaginal dimple (2.0 centimeter).

Davidoff-Moore procedure is a laparoscopic operation which is dedicated for women suffering from MRKH syndrome. This technique is a three stage surgery that includes dissection of the rectovesical space, abdominal mobilization of the peritoneum to create the vaginal fornices, and attachment of the peritoneum to the introitus. During the laparoscopic step, pelvis and abdominal cavity are explored. The peritoneum is incised transversely. Guided by the middle finger inserted in the patient's rectum, the incision is extended in a horseshoe shaped fashion into the connective tissue, which separates the bladder from the rectum. The suture is put on pelvic peritoneum in order to make peritoneal part of vagina. Single sutures strengthen vagina and pelvic wall peritoneum. Purse-string suture is assumed under ureters, to triangle of bladder to make upper wall of neovagina.

The course of operation was without any complications in both cases. Patients were discharged after 7 days of hospitalization. They received gynecological dilators and training guidelines in order to maintain the effect of operation, gynecological follow-up was recommended.

Discussion

The first line therapy is a nonsurgical treatment with self-dilation known as Frank and Ingram procedures – vaginal dilators are highly successful method [4,5]. Unfortunately, there is lack of information, whether our patients have used them. Different dilators from smaller (pediatric) to bigger (adolescent, adult) ones are placed against the vaginal dimple and pressure is applied to invaginate the mucosa. Sexuality of women with MRKH syndrome is another concern. Many of them prefer anal or oral intercourse. A vaginal intercourse is possible after creation of a neovagina. The time needed to create a vagina varies, ranges from four months up to several years, depending upon the patient's compliance. The Ingram modification of the Frank procedure involves the use of a bicycle seat mounted on a stool to create pressure for vaginal dilation. Also, repetitive coitus can also create a functional vagina. The advantage of a non-operative approach is avoidance of surgery with its potential complications. However, some women feel very uncomfortable with a concept of using dilators and therefore they choose surgery methods. It is important to explain that surgery can rapidly create a neovagina, however many of the surgeries require the use of dilators postoperatively to maintain effect. Surgery may be considered if a nonsurgical therapy fails or, if after counseling, patient elects a surgical approach. There are different types and methods of operations which help patients quickly recover. Davydov procedure is a laparoscopically-assisted technique, which has an advantage over traditional approaches. It has shorter operating time, lower intraoperative complications, shorter hospital stay, and no external scars. Postoperatively, sexual function is similar to women without gynecological disorders [6]. Another laparoscopic method is a modified Vecchietti procedure, after that neovaginal mucosae is comparable to normal vaginal mucosa. Active dilation is required until regular sexual activity is initiated like in Davydov procedure [7]. According to Mc Indoe procedure which is one of the most often performed techniques, uses split-thickness skin graft from the buttocks or special artificial skin. Postoperatively, the patient must remain on absolute bed rest and a low-residue diet for seven days. Also vaginal dilator must be used continuously for three months and then at night for six additional months to prevent contraction of the vagina.

Most of patients report satisfactory sexual relationships and vaginal intercourse after this surgery. Graft failure, post-operative hematoma, rectal perforation, and fistula formation are counted as the most common complications. The simpler alternative to the Mc Indoe procedure is Williams' vaginoplasty. Vaginal pouch created during this procedure is only approximately 4 to 5 cm in length and causes an unusual angle for coitus. A vaginal dilator is then used daily for three to four weeks postoperatively [8]. In pediatric surgery, sigmoid vaginoplasty is a common procedure. After it, post-operative dilators are not necessary. The disadvantages of this method are: chronic vaginal discharge, unpleasant odour and risk of developing adenocarcinomas. The best surgical option depends on experience of the attending surgeon and the woman's preference based upon the advantages and disadvantages of each procedure [9]. Both nonsurgical and surgical creation of a vagina should be individualized to the patient. Treatment involves a combination of psychosocial support and correction of the anatomic abnormality. The psychological aspects of a new diagnosis of vaginal agenesis can be difficult for any woman, especially an adolescent. Strong support

and counseling and psychological preparation before treatment plays a critical role. Even after treatment, patient will be unable to carry a pregnancy, they could take into consider surrogacy which is legal in a few countries or adoption. Transplantation of the uterus could be a new solution in the future but nowadays is ineffective.

Conclusion

To conclude, patients with diagnosed MRKH syndrome require interdisciplinary care from childhood through adolescence to adulthood. Presence of different body defects and lack of menarche (primary amenorrhea) should raise doctors, patients and parents' attention. Treatment should be discussed with patient in detail, as well as the moment and necessity of operation. Davidoff-Moore procedure seems to be a good step in gynecology; surgery technique will develop in this direction. Pregnancy and childbearing is a difficult topic, but it needs to find solution in future.

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