

ORIGINAL ARTICLE

Open Access



# Sigmoid vaginoplasty in Mayer-Rokitansky-Kuster-Hauser syndrome

Ibrahim Amadou Magagi<sup>1\*</sup>, Harissou Adamou<sup>1</sup>, Souleymane Oumarou Garba<sup>2</sup>, Maazou Halidou<sup>1</sup>, Ousseini Adakal<sup>3</sup>, Oumarou Habou<sup>1</sup>, Maman Bachir Abdoulaye<sup>3</sup>, Madi Nayama<sup>4</sup> and Rachid Sani<sup>4</sup>

## Abstract

**Introduction:** Mayer-Rokitansky-Kuster-Hauser syndrome (MRKH) is characterized by congenital aplasia of the uterus and upper 2/3 of the vagina, with normal female secondary sexual characteristics and a normal karyotype (46, XX). The frequency is 1/4500–5000 female births. The aim of this study was to report the management of MRKH syndrome with sigmoid vaginoplasty.

**Patients and method:** This study included 4 patients recruited over a 4-year period from February 2016 to January 2019. MRKH syndrome was retained in the presence of normal secondary sexual characteristics with normal external genitalia associated with vaginal aplasia and uterine agenesis. The approach was a laparotomy and a perineal approach under general anesthesia. The procedure involved the removal of a sigmoidal colonic graft that was anastomosed with the vaginal dimple.

**Results:** The average age was 23 years. All patients had consulted for primary amenorrhea, infertility, and/or difficulties in sexual intercourse. The diagnosis of MRKH type 1 was retained in all patients. The average length of the vagina was 3.25 cm before surgery and 13.63 cm after surgery. The postoperative outcomes were uneventful in 3 patients. One patient developed anastomotic stenosis that was successfully treated with vaginal dilation for 2 weeks. The average postoperative follow-up was 30 months.

**Conclusion:** In the context of a low-resource setting, sigmoid transposition represents a good procedure to treat vaginal aplasia and restore a satisfactory sexual activity to patients with MRKH type 1.

**Keywords:** Mayer-Rokitansky-Kuster-Hauser syndrome, Vaginoplasty, Sigmoid, Zinder

## Introduction

Mayer-Rokitansky-Kuster-Hauser syndrome (MRKH) is characterized by congenital aplasia of the uterus and upper two thirds of the vagina, with normal female secondary sexual characteristics and a normal karyotype (46, XX) [1–4]. The frequency of this anomaly is 1/4500–5000 female births [1–4]. There are 2 types of MRKH syndromes: type 1 represents about 44% of cases and is described as the agenesis of isolated Mullerian derivatives (isolated uterovaginal agenesis). Type 2 is associated with malformations that mainly affect the upper

urinary tract, the skeleton, and the otological and cardiac spheres [1–3, 5]. The etiology of the MRKH syndrome has not yet been clearly elucidated [2, 3, 6]. This pathology results from the interruption of the embryonic development of Muller's canals [2, 4, 7]. It would be an intermediate mesoderm lesion from which the primordia of the genitourinary tract are formed by the end of the 4th week of embryonic life [2, 4]. No precise genetic origin has yet been found, although some deletions in chromosomes 22, 4, and 17 appear to be involved [2, 3]. It would occur sporadically; however, the existence of familial cases has revealed an autosomal dominant mode of inheritance with variable expression (limited to female sex) and low penetrance [2, 6]. Primary amenorrhea remains the main clinical manifestation [2, 5]. The diagnosis of this condition is

\* Correspondence: [imagagi@gmail.com](mailto:imagagi@gmail.com)

<sup>1</sup>Department of Surgery and Surgical Specialties, Zinder National Hospital, Faculty of Health Sciences, University of Zinder, Zinder, Niger  
Full list of author information is available at the end of the article

usually clinical and radiological [2, 3, 8]. Abdominopelvic ultrasonography and magnetic resonance imaging (MRI) can complete the assessment by showing the absence of the uterus and looking for any associated abnormalities [2, 3]. Laparoscopy can be used in cases of diagnostic doubt [2].

In our context, patients with this anomaly are seen after the failure of a first or even a second marriage. The complaints are thus dominated by the non-satisfaction of the spouse during sexual intercourse and the desire for fertility. Many surgical and instrumental techniques have been developed to treat the absence of a vagina and to restore a sexual and social life to women with MRKH syndrome [3, 5, 9]. Currently, surgery is reserved for failures of vaginal dilation procedures that are recommended in the first instance [1, 10]. The aim of this study was to report our experience on sigmoid transposition vaginoplasty in the management of MRKH syndrome.

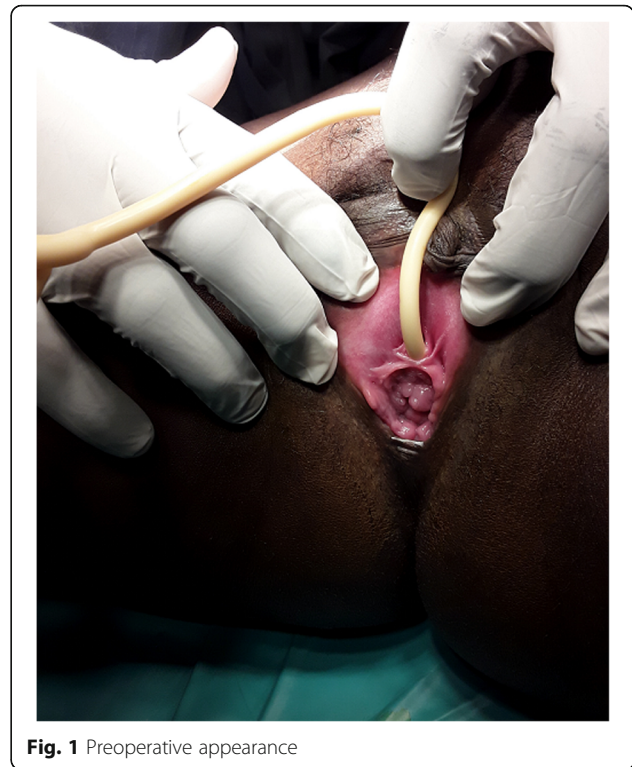
### Patients and methods

This was a retrospective study that enrolled 4 patients over a 4-year period from February 2016 to January 2019. MRKH syndrome was retained in the presence of normal secondary sexual characteristics with normal external genitalia associated with vaginal aplasia and uterine agenesis. A gynecological examination was performed in all patients. Tanner's classification was used to estimate pubertal development. Abdominopelvic ultrasound and an uroscanner confirmed the absence of the uterus and looked for other abnormalities. A vaginal opacification was performed preoperatively to objectify the length of the vaginal dimple and 1 month after surgery to detect possible fistulas. A barium enema was made preoperatively to evaluate the length of the sigmoid. All patients underwent vaginal dilation with appropriate Hégar candles for a sufficient duration without effective results. The surgical technique chosen for all patients was a sigmoid transposition that was described by Baldwin in 1904 [9]. A standard preoperative assessment was performed. Colonic preparation based on a diet without residues, and enemas were instituted 48 h before surgery. Antibiotherapy with ceftriaxone and metronidazole was instituted intraoperatively and continued 7 days after the operation.

The approach was a laparotomy and perineal approach under general anesthesia in the supine position. All patients had a bladder catheter (Fig. 1).

### Abdominal time

A Pfannenstiel incision was made. An exploration of the abdominal cavity was done in search of associated anomalies and also to complete the assessment of the genital anomaly. The sigmoid loop was identified (Fig. 2a), its length and vascularity verified. The vestigial remnants are separated from the bladder base by



**Fig. 1** Preoperative appearance

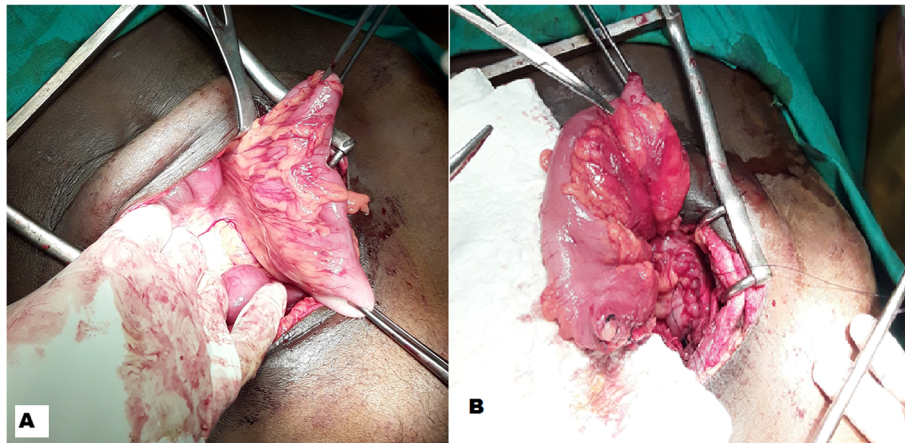
anterior dissection, and the dissection is performed posteriorly between the anterior surface of the rectum and the vestigial remnants. This maneuver allows you to find the vaginal dimple that is sufficiently exposed. This exposure is helped by the intravaginal introduction of a Hégar candle of the appropriate caliber. A segment of the sigmoid colon of about 15 cm was then selected, sectioned at its lower and upper ends, and cleaned with saline. This graft remained pedicled to the inferior sigmoid artery (Fig. 2b). The continuity of the sigmoid colon was restored (Fig. 2b).

### Vaginal time

An assistant was placed between the patient's legs to hold the Hégar candle and push back the vaginal dimple which is incised at the top (Fig. 3a).

The graft was rotated abdominally in the anti-peristaltic direction. The upper end is closed with a tight continuous suture, and the lower end is anastomosed with the edges of the vaginal dimple. To avoid prolapse, the colonic graft was leaned against the anterior surface of the rectum and fixed to the pre-vertebral ligament by its upper end (Fig. 3b).

The bladder catheter is left in place for 5 days. A light diet was allowed when bowel movements resumed. A control X-ray with Hégar candles in the neovagina was performed (Fig. 4). Sexual intercourse was permitted 6 months after the procedure. The variables studied were age, marital status, tanner stages, karyotype, pre- and



**Fig. 2** a Identification of the sigmoid loop. b Preparation of the colonic graft

postoperative vaginal dimple length, duration of vaginal dilation, associated abnormalities, and postoperative complications.

**Results**

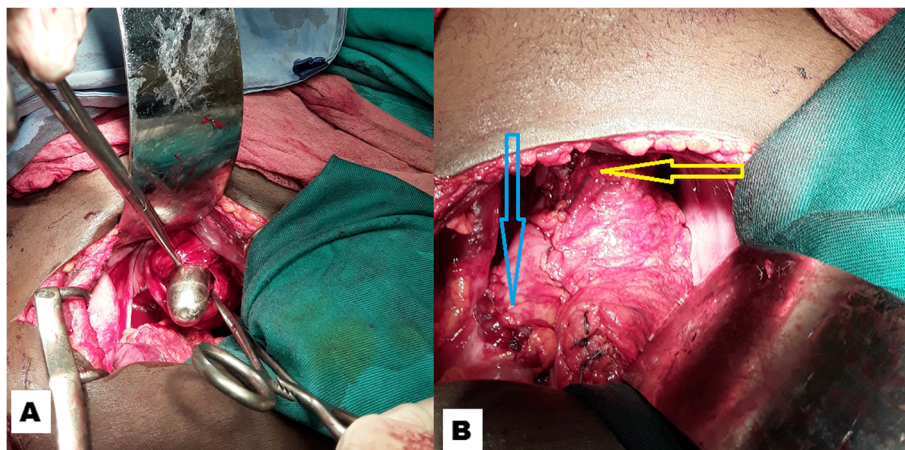
The average age was  $23 \pm 2.16$  years old. All patients had consulted for primary amenorrhoea, infertility, and/or difficulties in sexual intercourse. No medical or family history was noted. No associated anomalies were found. Ovaries were present in all patients. Genotypically, all patients had a normal karyotype 46, XX. The diagnosis of MRKH type 1 was retained in all patients. The average length of the vagina was 3.25 cm before surgery and 13.63 cm postoperatively. The postoperative recovery was uneventful in three patients. One patient developed anastomotic stenosis successfully treated with vaginal dilation for 2 weeks. The median postoperative follow-up time was 30 months (Q1–Q3 = 16.5–41). Two

patients remarried. The clinical and therapeutic characteristics of the patients are summarized in Table 1.

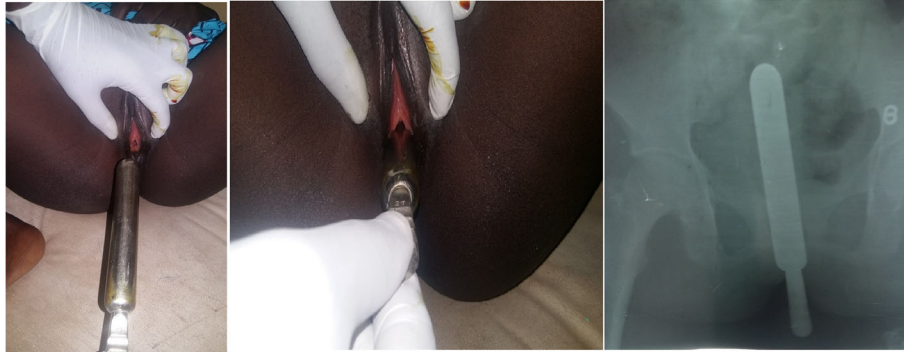
**Discussion**

Once the diagnosis of MRKH syndrome is discovered, it will be necessary to institute appropriate management, which takes into account both the associated abnormalities and the psychosocial component [1, 9]. In fact, this pathology compromises sexual intercourse and fertility and results in intense psychological suffering in these patients who feel diminished in their femininity [1, 9].

The definitive treatment of this condition depends on the age of the patient and is only considered when the patient is emotionally and sexually mature [1, 8, 9, 11]. In this respect, the management of the syndrome must be multidisciplinary involving the surgeon, the gynecologist, the urologist, the plastic surgeon, and the psychologist. Numerous instrumental and surgical therapies have been described treating vaginal aplasia [1, 3, 9]. The main



**Fig. 3** a Vaginal dimple opening. b Final appearance of the assembly (blue arrow: colo-colic anastomosis, black arrow: colo-vaginal anastomosis)



**Fig. 4** Postoperative results: Hégar candle in the neovagina

objective is to restore a sexually active life by restoring a conventional vaginal size, which is usually defined by a length  $\geq 6$ –7 cm and a sufficient diameter between 3–3.5 cm (admitting two fingers) [9–13].

Vaginal dilation is the method recommended on the frontline by expert committees [1, 9]. This method has proven its effectiveness with a success rate ranging from 90 to 96% [1, 10, 13]. All patients may be candidates for vaginal dilation, even with short vaginal dimples less than 2 cm [10]. Two methods are conventionally used, dilation according to Frank or Ingram [3, 9, 14]. Frank was a Czech gynecologist who used waist-increasing candles to obtain a functional vagina; Ingram later improved the process by using dilator candles placed on a bicycle saddle allowing patients to adopt a more accepted and less invasive sitting position [3, 9, 13, 14]. The success of these two methods is underpinned by a frequency of 1 to 3 dilatations of 10 to 30 min per day during 4 to 6 months [3, 10]. Several factors can be sources of failure to vaginal dilatations. These include psychosocial causes (motivation, lack of family support), cognitive (misunderstanding of procedures, young age), logistics (care structure too far away, lack of suitable equipment), and anatomical (associated malformations, absence of vaginal dimples, complications) [10]. In our study, Frank's method was practiced without success. Indeed, most of our patients living in rural areas, far from urban health centers, cannot move easily for regular

intra-hospital dilations. Social and cultural considerations (shame, intimacy, promiscuity, religion) also likely prevented them from performing appropriate dilations at home, following the caregivers' instructions. These vaginal dilations must be performed before considering any surgical procedure [1, 3, 10]. Regular sexual intercourse may be just as effective as vaginal dilatation in some cases [9, 10, 12, 13].

Thus, in case of failure of non-surgical methods, or following the refusal of a patient, surgery should be used. A plethora of surgical techniques exist, either conventionally or laparoscopically. Dupuytren (1817) and Amussat (1835) seem to be the first authors to have proposed a correction of vaginal hypoplasia by surgical methods [9]. None of the techniques has proven to be superior to the others, and there is no consensus on an ideal intervention [3, 8, 9]. These different techniques can be classified into 4 groups according to their principle [9]. The first group includes all the techniques that consist of the creation of a neovagina between the bladder and the rectum (Wharton, McIndoe, Davydov) [3, 9]. Vaginal plasties using intestinal segments (jejunum, ileum, colon, cecum, rectum) constitute the second group [3, 9]. In third place come vulvovaginoplasties (technique of Williams modified by Creatsas) [9]. The last group includes the surgical traction of the vaginal dimple which can be considered as accelerated vaginal dilation (Vechietti, balloon) [9]. Recently, hypoplastic vaginas have been reconstructed from vulvar

**Table 1** Clinical and therapeutic characteristics of operated patients

No.	Age (years)	PMS	Tanner stage	DVD	VDL preop (cm)	VDL postop (cm)	IT (min)	PO	LOS (days)	PFT (months)
1	20	Divorced	4	6	4	14	155	None	7	24
2	25	Divorced	5	5	2.5	13	180	None	8	46
3	24	Divorced	4	6	3.5	13.5	135	Stenosis	15	36
4	23	Single	5	4	3	14	165	None	5	9

PMS preoperative marital status, DVD duration of vaginal dilations (months), VDL vaginal dimple length (preop: preoperative; postop: postoperative) in cm, IT intervention time, PO postoperative complications, LOS length of stay, PFT postoperative follow-up time

biopsies of autologous tissue taken from patients, cultured in vitro, and surgically reimplanted after the necessary cellular maturation [15]. The oral mucosa has also been used to perform vaginoplasty [16].

Vaginal reconstruction using intestinal grafts was reported by Sneguireff as early as 1892 [9]. This method has emerged in recent years due to the many advantages it confers [8, 9]. The ileum was first used as an alternative, then abandoned in favor of the sigmoid colon, which gave fewer complications [9, 17]. Later, authors described the use of the caecum and jejunum [8, 9]. Usually performed by laparotomy, sigmoid transplantation vaginoplasty is also possible by laparoscopy. The main advantage of this technique is that it produces a vaginal cavity that is not very prone to narrowing, requires less dilation, and is naturally lubricated, resulting in less dyspareunia [8, 9, 11, 18]. Bleeding, malodorous discharge, prolapse, colitis, or even malignant degeneration into adenocarcinoma have been reported as possible incidents following colonic vaginoplasty [3, 9]. We have not seen any of these complications in our patients although the main limitation of our study is the small size of our sample, explained by the rarity of the malformation. But more hindsight is needed to assess the real long-term impact, so our patients should have a longer postoperative follow-up period. In 2014, the meta-analysis by Bouman et al. [19] of 21 studies involving 894 patients confirmed that intestinal vaginoplasty was associated with few complications and highly sexual satisfaction among beneficiaries. This study did not find any of the major complications such as cancer or colitis [19]. Vaginoplasty by colonic transposition is a low-cost technique with a simple postoperative follow-up, easily achievable in developing countries [11, 14] such as Niger, where the technical facilities do not allow more complex interventions.

## Conclusion

Mayer-Rokitansky-Kuster-Hauser syndrome is a rare condition that causes primary amenorrhea and infertility while making vaginal intercourse difficult or impossible. This is a hindrance to a proper social life and a psychological frustration for the woman. In the context of a low-resource setting, sigmoid transposition represents a good procedure to treat vaginal aplasia and restore a satisfactory sexual activity to patients with MRKH type 1.

## Abbreviations

PMS: Preoperative marital status; DVD: Duration of vaginal dilations (months); VDL: Vaginal dimple length (preop: preoperative; postop: postoperative) in centimeters; IT: Intervention time; PO: Postoperative complications; LOS: Length of stay; PFT: Postoperative follow-up time

## Acknowledgements

The authors wish to acknowledge all medical staff of ZNH for their contribution to the management of patients and the recording of patient's data.

## Authors' contributions

IAM, HA, and SOG: surgical intervention, study design, bibliographic research, recording data, analysis interpretation of data, and drafting of the article. MH, OA, OH, MBA, MN, and RS: reviewed and completed the writing process. All authors read and approved the final manuscript.

## Funding

None.

## Availability of data and materials

Please contact the corresponding author for data requests.

## Ethics approval and consent to participate

Local ethics approval was obtained from the joint decision of the Scientific Council of the Faculty of Health Sciences of the University of Zinder and the Advisory Technical Advisory Council of Zinder National Hospital. Data was completely anonymized. Written consent was obtained from participants.

## Consent for publication

Consent to publish was obtained from patients. Participants gave written informed consent for their personal or clinical details along with any identifying images to be published in this study.

## Competing interests

The authors declare that they have no competing interests.

## Author details

<sup>1</sup>Department of Surgery and Surgical Specialties, Zinder National Hospital, Faculty of Health Sciences, University of Zinder, Zinder, Niger. <sup>2</sup>Gynecology and Obstetrics Department, Mother and Children Center, Zinder, Niger. <sup>3</sup>Department of Surgery and Surgical Specialties, Faculty of Health Sciences, UDDM University of Maradi, Maradi, Niger. <sup>4</sup>Department of Surgery and Surgical Specialties, Faculty of Health Sciences, Abdou Moumouni University of Niamey, Niamey, Niger.

Received: 24 March 2020 Accepted: 4 August 2020

Published online: 18 August 2020

## References

- Committee Opinion No ACOG (2018) 728 Summary: Müllerian agenesis diagnosis, management, and treatment. *Obstet Gynecol* 131(1):196–197
- Morcel K, Guerrier D, Watrin T, Pellerin I, Levêque J (2008) Le syndrome de Mayer-Rokitansky-Küster-Hauser (MRKH) : clinique et génétique. *J Gynecol Obstet Biol Reprod* 37(6):539–546
- Bombard DS, Mousa SA (2014) Mayer-Rokitansky-Kuster-Hauser syndrome: complications, diagnosis and possible treatment options: a review. *Gynecol Endocrinol Off J Int Soc Gynecol Endocrinol* 30(9):618–623
- Bulfoni A, Motta F, Frontino G, Alberico D, Fedele L (2019) Mayer-Rokitansky-Kuster-Hauser syndrome: associated anomalies in a cohort of 77 patients. *Ital J Gynaecol Obstet* 31(1):43–51
- Yakasai IA, Daneji SM, Natasha A, Yunus EM (2015) Late presentation of Mayer-Rokitansky-Kuster-Hauser syndrome in the tropics-northern Nigeria. *Br J Med Med Res* 5(8):1077–1082
- Fontana L, Gentilin B, Fedele L, Gervasini C, Miozzo M (2017) Genetics of Mayer-Rokitansky-Küster-Hauser (MRKH) syndrome. *Clin Genet* 91(2):233–246
- Kisu I, Ono A, Iijima T, Katayama M, Iura A, Hirao N (2019) Mayer-Rokitansky-Küster-Hauser syndrome with a uterine cervix and normal vagina associated with gonadal dysgenesis in a 46,XX female. *J Obstet Gynaecol Res* 45(7):1386–1390
- Al-Mehaisen L, Amarín Z, Hani OB, Ziad F, Al-Kuran O (2017) Ileum neovaginoplasty for Mayer-Rokitansky-Küster-Hauser: review and case series. *Afr J Urol* 23(2):154–159
- Callens N, De Cuyper G, De Sutter P, Monstrey S, Weyers S, Hoebek P et al (2014) An update on surgical and non-surgical treatments for vaginal hypoplasia. *Hum Reprod Update* 20(5):775–801
- Amies Oelschlager A-M, Debiec K (2019) Vaginal dilator therapy: a guide for providers for assessing readiness and supporting patients through the process successfully. *J Pediatr Adolesc Gynecol*. <https://doi.org/10.1016/j.jpog.2019.05.002>

11. Mungadi LA, Ahmad Y, Yunusa GH, Agwu NP, Ismail S (2010) Mayer-Rokitansky-Kuster-Hauser syndrome: surgical management of two cases. *J Surg Tech Case Rep* 2(1):39–43
12. Cheikhelard A, Bidet M, Baptiste A, Viaud M, Fagot C, Khen-Dunlop N et al (2018) Surgery is not superior to dilation for the management of vaginal agenesis in Mayer-Rokitansky-Küster-Hauser syndrome: a multicenter comparative observational study in 131 patients. *Am J Obstet Gynecol* 219(3):281.e1–281.e9
13. Lee M-H (2006) Non-surgical treatment of vaginal agenesis using a simplified version of Ingram's method. *Yonsei Med J* 47(6):892–895
14. Moudouni S, Koutani A, Attya AI, Hachimi M, Lakrissa A (2004) The use of isolated sigmoid colon segment for vaginal replacement in young adults. *Int Urol Nephrol* 36(4):567–571
15. Raya-Rivera AM, Esquiliano D, Fierro-Pastrana R, López-Bayghen E, Valencia P, Ordorica-Flores R et al (2014) Tissue-engineered autologous vaginal organs in patients: a pilot cohort study. *Lancet Lond Engl* 384(9940):329–336
16. Chan JL, Levin PJ, Ford BP, Stanton DC, Pfeifer SM (2017) Vaginoplasty with an autologous buccal mucosa fenestrated graft in two patients with vaginal agenesis: a multidisciplinary approach and literature review. *J Minim Invasive Gynecol* 24(4):670–676
17. Kwun Kim S, Hoon Park J, Cheol Lee K, Min Park J, Tae Kim J, Chan KM (2003) Long-term results in patients after rectosigmoid vaginoplasty. *Plast Reconstr Surg* 112(1):143–151
18. Labus LD, Djordjevic ML, Stanojevic DS, Bizic MR, Stojanovic BZ, Cavic TM (2011) Rectosigmoid vaginoplasty in patients with vaginal agenesis: sexual and psychosocial outcomes. *Sex Health* 8(3):427–430
19. Bouman M-B, van Zeijl MCT, Buncamper ME, Meijerink WJHJ, van Bodegraven AA, Mullender MG (2014) Intestinal vaginoplasty revisited: a review of surgical techniques, complications, and sexual function. *J Sex Med* 11(7):1835–1847

### Publisher's Note

Springer Nature remains neutral with regard to jurisdictional claims in published maps and institutional affiliations.

Submit your manuscript to a SpringerOpen<sup>®</sup> journal and benefit from:

- Convenient online submission
- Rigorous peer review
- Open access: articles freely available online
- High visibility within the field
- Retaining the copyright to your article

---

Submit your next manuscript at ► [springeropen.com](https://www.springeropen.com)

---