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Short communication

Ileum neovaginoplasty for Mayer–Rokitansky–Küster–Hauser: Review and case series



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Abstract

Objective: To review treatment modalities of Mayer–Rokitansky–Küster–Hauser syndrome, and to present further evidence on the successful use of ileum segment as an additional procedure for the creation of a neovagina.

Methods: Five women presented with primary amenorrhea, normal secondary female sexual characteristics, normal external anatomy, shortening of the vagina, with only vaginal dimples. Abdominal ultrasound scans suggested the absence of uterus. Both ovaries were present with normal kidneys. Karyotyping confirmed XX genotype. Pelvi-abdominal MRI confirmed the diagnoses. All patients were started on nonsurgical treatment, in the form of graduated dilators, as a first-line approach. This was not acceptable to the patients. Decisions were made to resort to ileum vaginoplasty.

Results: The mean surgical time was 5 h. Three patients sustained intra-operative bladder injury. All patients reported watery vaginal discharge. Four patients were followed up for 5 years, and were satisfied with their results. This was confirmed by clinical examination. The fifth patient failed to attend for immediate postoperative care. The vagina was 6–7 cm long with introital stricture.

Conclusion: Women with Mayer–Rokitansky–Küster–Hauser syndrome who need vaginal reconstruction have the surgical option of a vascularized free ileum graft that provides a durable, lubricated neovagina without the scarring of skin grafts.

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Introduction

Mayer–Rokitansky–Küster–Hauser (MRKH) syndrome is a rare syndrome that affects 1 in 4000–5000 women in the general population [1]. It is characterized by congenital aplasia of the uterus and the upper two-thirds portion of the vagina in women who have normal ovarian function and normal external genitalia, with normal secondary sexual characteristics during puberty and primary amenorrhea [1].

Development of the female genital tract is a complex process dependent upon a series of events involving cellular differentiation, migration, fusion, and canalization. Failure of any one of these processes results in a congenital anomaly [1].

The female genital tract is derived from the Müllerian ducts, urogenital sinus, and vaginal plate. The two Müllerian ducts are initially composed of solid tissues and lie side by side. Subsequently, internal canalization of each duct produces two channels divided by a septum that is reabsorbed in a cephalad direction by 20 weeks. The cranial portions develop into the fimbria and fallopian tubes, while the caudal, fused portions form the uterus and upper vagina [1].

The sinovaginal bulbs are two solid evaginations originating in the urogenital sinus at the distal aspect of the Müllerian tubercle. The sinovaginal bulbs proliferate into the caudal end of the uterovaginal canal to become a solid vaginal plate then degeneration of the central cells of this vaginal plate occurs in a cephalad direction. Canalization is complete by 20 weeks [1].

Approximately 50% of MRKH syndrome patients have urologic anomalies, such as unilateral renal agenesis, pelvic or horseshoe kidneys, or irregularities of the collecting system, and 45% have skeletal anomalies involving the spine, ribs, and extremities. Other less common anomalies include congenital heart lesions, abnormalities of the hand, deafness, cleft palate, and inguinal or femoral hernias [2].

The signs and symptoms of MRKH syndrome vary greatly. In most cases, the uterus and/or the vagina are aplastic; in other rare cases, there may be atresia of the upper portion of the vagina and an underdeveloped or rudimentary uterus. In some cases, the Fallopian tubes may be affected. The initial symptom of MRKH syndrome is primary amenorrhea. On physical examination, the external genitalia are normal. A vaginal dimple or small pouch with a hymenal fringe is usually present, as the vaginal pouch and hymen are both derived from the urogenital sinus [2].

The cause of MRKH syndrome remains largely unknown. Initially, the syndrome was thought to occur sporadically due to non-genetic factors such as gestational diabetes or exposure to teratogens, but no link between an environmental cause and MRKH syndrome has ever been established [3].

Some case studies suggest that MRKH syndrome is a genetic disorder that is inherited as an autosomal dominant trait with incomplete penetrance and variable expressivity. Polygenic multifactorial inheritance has also been proposed as a cause. Seven deletions of chromosomal segments have been identified in chromosomes 1, 4, 8, 10, 16, 17 and 22, and 1 duplication on the X chromosome. The candidate genes HNF1B (formerly TCF2),

LHX1, TBX6, ITIH5 and SHOX are currently under investigation [4].

It has been noted that some males may exhibit absence or underdevelopment of the Wolffian duct, thus affecting vas deferens, with oligo- or azoospermia, kidney abnormalities, spinal malformations and hearing impairment. Rare cases in both males and females in the same family suggest a shared genetic origin [5].

Some disorders may be similar to those of MRKH syndrome, where comparisons are useful in the differential diagnosis. A mutation of the WTN4 gene causes a rare genetic syndrome that affects females. It is characterized by the absence of the uterus, short or stenosed vagina, abnormally high levels of androgens, with acne and hirsutism, and normal female secondary sexual characteristics during puberty, but with primary amenorrhea. The gonads may be of ovitesticular types that produce both male and female sex hormones [6].

Other rare syndromes include complete androgen insensitivity syndrome, Winter syndrome, McKusick–Kaufman syndrome, Frasier syndrome, Goldenhar syndrome, VACTERL association, and Turner's syndrome [6].

Other disorders may be associated with MRKH syndrome secondary characteristics, and are not necessary for a differential diagnosis, include Klippel–Feil syndrome, Sprengel deformity and DiGeorge syndrome [6].

The diagnosis of MRKH syndrome is based upon detailed history, clinical evaluation and transabdominal ultrasonography, complemented by magnetic resonance imaging. Karyotyping may be performed to rule out other conditions. Once MRKH syndrome is diagnosed, a search must be undertaken for renal, skeletal, hearing and cardiac abnormalities. Normal levels of follicle stimulating hormone, luteinizing hormone, 17 β -oestradiol and androgens will confirm the integrity of ovarian function.

The treatment of MRKH depends upon the patient's age at diagnosis. Gynecologists, pediatricians, urologists, orthopedic surgeons, plastic surgeons, physiotherapists and psychological support may be needed for a comprehensive approach to treatment.

For the treatment of vaginal aplasia, medical and surgical care is necessary for creating a neovagina when the patient is emotionally and sexually mature.

Nonsurgical techniques are considered. The first-line approach is the Frank and Ingram technique through the application of self-administered progressive pressure to the perineum using vaginal dilators [7].

The surgical modalities include a variety of techniques, with no consensus as to which vaginoplasty technique is best. The most common surgical procedure used for vaginal reconstruction is the McIndoe procedure, where a split-thickness skin graft from the thigh or buttocks is placed in a pocket between the urethra and rectum that is created by blunt dissection. A cylindrical stent is left in situ. The edges of the graft are sutured to the cut edges of the introitus. The labia majora are sutured together to hold in the mold. A Foley catheter is inserted. Both catheters and stent are removed after one week; the neovagina is irrigated [8].



Figure 1 Pre-surgical view.

In Williams vaginoplasty, a vulvar flap is used to make a vaginal tube. Similarly, in rotational flap procedures, pudendal thigh, gracilis myocutaneous, and labia minora, fasciocutaneous flaps are used [9].

Despite reports on many different techniques, the ideal method of vaginoplasty has not been firmly established [10].

The Vecchietti technique is based on stretching the vaginal dimple via continuous progressive controlled traction on an acrylic mold that is passed through the potential neovaginal space and pulled by sub-peritoneal threads, which emerge on the surface of the abdomen and are attached to a traction device, over a period of days to weeks to lengthen the vaginal vault. This technique is now performed laparoscopically [11].

Medical and surgical care are essential for capacity for sexual intercourse, and genital appearance. In rare cases, fertility may be possible. Using assisted reproductive techniques, women with MRKH syndrome can reproduce by having oocytes harvested, fertilized, and implanted in a surrogate. However, because MRKH syndrome appears to be of genetic origin, the risk of passing on the disease to children exists and any decision to conceive should therefore be undertaken after careful consultation.

The use of bowel segments has gained popularity for vaginal reconstruction [12]. For this method, a free intestine is isolated, with its pedicle, transferred to the prepared vaginal pouch. Arterial and venous anastomosis between the flap and the recipient vessels are performed. The main advantages of this operation are the lack of shrinkage, with no need for long-term vaginal dilation, and the natural lubrication provided by the mucous production that obviates the need for artificial lubricants and decreases the risk of dyspareunia [13].

In this case series, the rationale in using ileum over the more commonly used sigmoid colon for vaginoplasty was to avoid the probability of fecal odor. In addition, it was due to the concern surrounding the reported diversion colitis, possibly due to a lack of short-chain fatty acids in colonic contents that are required for mucosal integrity, but being less of a problem the ileum [12] (Fig. 1).

Ethical committee approval for the work, and informed consent from the patients were obtained.

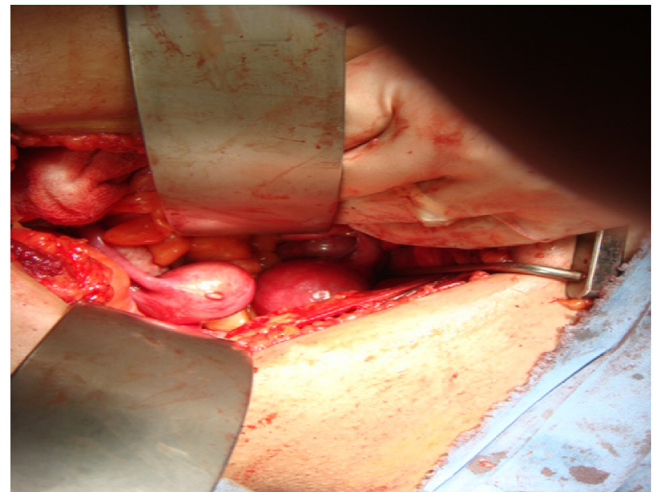


Figure 2 Abdominal view.

Methods

Five women presented with primary amenorrhea. No medical or familial history of note was reported.

On physical examination, all patients had normal secondary female sexual characteristics. Gynecological examination revealed normal external anatomy. Speculum examination was not possible owing to shortening of the vagina, instead only vaginal dimples were seen. Abdominal ultrasound scan suggested the absence of uterus. Both ovaries were present with normal kidneys. Follicle-stimulating hormone and luteinizing hormone levels were within normal. Karyotyping confirmed XX genotype. Pelvi-abdominal MRI confirmed the diagnosis (Fig. 2).

All patients were started on nonsurgical treatment as a first-line approach. Self-administered progressive pressure with vaginal dilators was used for three months. This resulted in lengthening the vagina to 3–5 cm. This was not readily acceptable to the patients. Decisions were made to resort to surgery.

The surgical technique involved admission to hospital 2 days prior to surgery for bowel preparation. Laparotomy was performed through a 10-cm suprapubic transverse incision. All patients had rudimentary uteri and normal looking ovaries.

In general, the surgical procedures were summarized as follows: a 15 cm segment of the ileum, 10 cm proximal to the ilio-cecal junction was isolated and removed, while preserving its vascular pedicle. The ends of the remaining ileum were approximated and anastomosed. Simultaneously, the second surgeon, starting with a transverse lunar vaginal incision, created a pouch for the neovagina from the blind vaginal introitus, through the recto-vesical space, into the abdominal cavity. The resected ileum was transferred into the pouch. The proximal end was closed with sutures and anchored to the retropubic periosteum. The lower end was sutured to the edges of the vaginal introitus and sutures were placed between the colonic wall and introitus. A 3–4 cm segment of prolapse was essential because of the tendency for retraction in the immediate postoperative period (Fig. 3).



Figure 3 View at the end of surgery.

During surgery, three bladder, and one rectal perforation were sustained while dissecting the recto-vesical space. All were repaired straightaway. Cystoscopy examination was performed at the end of the procedure. Urinary catheter was inserted for 4–9 days. Fluid intake was allowed on the third postoperative day, and soft diet was introduced on days 5–7.

A Foley catheter (gauge 24) was left inside the neovagina for 2 weeks. Patients were discharged on oral and local antibiotics, and were followed up twice weekly for the first 2 weeks, then weekly for a month, fortnightly for the following 12 weeks, monthly for the next 6 months, 3 monthly for the next year, then 6 monthly for 5 years.

A sterile vaginal examination was carried out at each visit. Pelvic floor physiotherapy and vaginal dilators were used for 6–8 weeks postoperatively for 6 months to maintain dilation of the neovagina. Sexual intercourse was allowed as soon as the wound had healed and it became comfortable. Generally, patients were allowed to engage in sexual intercourse after 6 weeks.

An Institutional Review Board or Ethics Committee approval was not needed in this case series.

Results

The patients' mean age was 30.8 years (range 26–38 years). All patients had Müllerian agenesis (Table 1).

The mean surgical time was 5 h. Two, of the three patients with bladder injury developed an over active bladder and were treated

with anti-cholinergic drugs. All patients reported watery vaginal discharge. This did not require patients to change undergarments during the day. The only lifestyle modification was to wear a pad that did not require frequent change. There was no need to perform cystogram to ensure there was no vesicovaginal fistula, as the discharge was minimal, and was an acceptable side effect.

Four patients were followed up for 5 years, and were satisfied with the result. This was confirmed by clinical examination. The fifth patient failed to attend for follow-up and immediate postoperative care. On clinical examination, the vagina was 6–7 cm long with introital stricture.

Discussion

Treatment of MRKH syndrome will usually include management of the physical findings and psychological support for the emotional issues associated with the diagnosis.

The treatment of vaginal aplasia involves the creation of a neovagina to women that are emotionally mature and ready to start sexual activity. Nonsurgical techniques, using vaginal dilators, are considered a first-line approach, and have been recommended due to their low morbidity.

In a study by Liao et al. participants using vaginal dilators scored low on sexual esteem and sexual satisfaction and high on sexual anxiety and fear of sexual relationships [14].

In this series, the patient success rate with vaginal dilators was poor. This is not in agreement with retrospective studies that have reported success rates of up to 80% [14]. The reasons for this may have been the lack of counseling by a dedicated women's health nurse, and the time for dilation being prohibitive

In addition, local women feel very uncomfortable with the concept of using graduated dilators because of societal and cultural inhibition that precludes the use of dilators to create a functional vagina as a first-line treatment for vaginal agenesis that may be necessary for several months.

It is important to inform patients who undergo surgery to create a neovagina will most likely need to use vaginal dilators after the surgery to enhance the chance of success.

Of note is the fact that the only unsatisfactory result, in this case series, was in the only patient that failed to attend for postoperative care. This emphasizes the importance of post-surgical regular use of dilators, prolonged postoperative supervision and support by nursing instruction.

Table 1 Summary of patients.

| Patient number | Age | Marital status | Uterus | Result | Notes |
|----------------|-----|----------------|------------------|----------------|---|
| 1 | 38 | Single | Rudimentary | Satisfactory | |
| 2 | 26 | Single | Rudimentary | Satisfactory | |
| 3 | 26 | Divorced | Rudimentary | Unsatisfactory | No post operative care |
| 4 | 29 | Divorced | Rudimentary | Satisfactory | |
| 5 | 35 | Divorced | Uterus didelphys | Satisfactory | Ideal interposition between cervix and perineum was used, preserving the uterus |

There are many different described methods in the literature for neovagina. Traditional techniques include myocutaneous flaps, skin grafts, skin grafts applied to omental cylinders, and combinations thereof, which are associated with skin scarring at the donor graft site.

Every operation has advantages and disadvantages. Surgeon preference and experience influence the choice of technique [15]. In this series, a 3–4 cm segment of prolapse was essential because of the tendency for retraction in the postoperative period. This is cosmetically less optimal than the alternative option of vaginal tissue replacement by autologous buccal mucosal grafting.

Vaginoplasty with autologous buccal micromucosa combined with acellular allogenic dermis has been reported to be an effective and feasible approach for patients with MRKH syndrome, with satisfactory long-term anatomical and functional results, but is limited by the high price and the potential infection [16].

In this series of five procedures, three bladder and one rectal injury were sustained. All four occurred during the perineal pouch creation, and not during the antegrade dissection into the rectovesical pouch. The lessons that were learned to prevent this rate of local injury going forward was that to start with the abdominal rectovesical dissection first, by following the uterosacral ligaments to the rudimentary uterus, then the vaginal dissection.

In this series, fluid intake was allowed on the third postoperative day, and soft diet was introduced on days 5–7. This is probably outside common practice. In recent randomized controlled trials of pelvic surgery, fast track bowel regimens and the enhanced recovery after surgery (ERAS) protocols are increasingly utilized to reduce postoperative convalescence and hospital stays as well as surgical costs [17]. It is suggested that such a prolonged period of fasting due to bowel anastomosis should encourage the implementation of buccal graft neo-vaginal reconstruction without a bowel anastomosis.

The ideal vaginal reconstruction should provide a long-lasting, functional passage that does not need maintenance with dilators or lubrication. The technique of intestinal neovagina uses an isolated segment of bowel for vagina. The advantages of the isolated intestinal neovagina over skin grafts include having its own inherent vascular supply via its intact mesentery and its distensibility. In addition, it requires less regular use of a dilator or stent to maintain vaginal depth and avoid introital constriction.

Regarding technique, Wright and Hanna in their review of 33 years of experience with vaginal reconstruction, including 10 bowel vaginoplasties, suggest that detubularization is needed to put less tension on the mesentery [18]. Although it would seem that the ileum would be more prone to tension on the mesentery than the more regionally located sigmoid colon, in the authors experience there was no difficulty with tension on the ileal mesentery or need to make any modifications for the ileal segments to reach the deep pelvis.

In intestinal neovagina, it is suggested that the normal nonirradiated sigmoid is generally the preferred bowel segment, as it can most easily be mobilized to the perineum in a tension-free manner [19–21]. In contrast, there is anecdotal evidence that small bowel vaginoplasty is associated with less gastrointestinal problems, and less copious vaginal discharge and foul odor [22,23]. Furthermore,

although graft failure is uncommon, if one did occur, replacement material from the ileum is more plentiful than in the case of large bowel transfer.

This report is relatively one of the larger case series of ileum neovaginoplasty in the peer-reviewed literature with low rates of bowel complications and highly functional outcomes in patients compliant with followup. In general, many surgical techniques are described but no consensus exists as to the best one. In addition, outcome data are sparse. The authors concur with the need for well-planned, prospective cohort studies that include assessments of psychological and sexual outcomes of MRKH syndrome [24].

Conflict of interest

The authors state that there is no conflict of interests (financial or non-financial)

Ethical committee approval

There was no funding resource apart from self-funding.

This review involved only data collection about the subject discussed and all were in accordance with the ethical standards of the institutional and/or national research committee and with the 1964 Helsinki declaration and its later amendments or comparable ethical standards.

Authors' contributions

L. Mehaisen: Surgical operations, study conception & design, data acquisition, manuscript writing, editing, and revision.

Z. Amarin: Surgical operations, conception & design, and manuscript writing.

O. Bani Hani: Surgical operations, conception & design, data acquisition, and manuscript writing.

F. Ziad: Conception & design, data acquisition, and manuscript revision.

O. Al Kuran: Surgical operations, conception & design, and manuscript revision

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