

Mayer-Rokitansky-Küster-Hauser syndrome: a review of 245 consecutive cases managed by a multidisciplinary approach with vaginal dilators

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Objective: To understand the efficacy of vaginal dilators in the management of Mayer-Rokitansky-Küster-Hauser (MRKH) syndrome.

Design: Retrospective sequential study.

Setting: Hospital.

Patient(s): 245 women.

Intervention(s): Vaginal dilators.

Main Outcome Measure(s): Functional vaginal length and sexual satisfaction.

Result(s): Of the patients who completed the program, 232 (94.9%) achieved a successful vaginal length (defined as greater than 6 cm in length and maximum width throughout the vagina and especially at the apex) and sexual function. When the program was completed by all patients, 100% of patients were successful.

Conclusion(s): Vaginal dilator therapy is the treatment of first choice for creation of the vagina in MRKH syndrome, and the success rates suggest that surgery is rarely, if ever, required. (Fertil Steril® 2012;97:686–90. ©2012 by American Society for Reproductive Medicine.)

Key Words: MRKH syndrome, neovagina, vaginal dilators

The National Centre for Congenital Abnormalities of the Genital Tract was established in London in 1998. The remit was to coordinate the treatment of congenital abnormalities for England and Scotland. Mayer-Rokitansky-Küster-Hauser (MRKH) syndrome has been one of the commonest referrals, a syndrome of uterine and vaginal agenesis with an incidence of 1:5,000 female births (1). Most patients present with primary amenorrhea in their early teens with the presence of normal secondary sexual characteristics other than menstruation. Ovarian function in these patients is entirely

normal, hence their normal breasts, pubic hair, and axillary hair growth (2). The external phenotype is normal female, and the philosophy of managing this condition for many years has simply been the creation of a neovagina for sexual function. This has challenged gynecologists to create both surgical and nonsurgical approaches, and reports of extensive and innovative techniques have been submitted to the literature over many years (3).

However, it has become apparent that a holistic approach to the management of these patients is much more important, involving more than simply

the anatomic correction of their vagina. The introduction of routine psychological and expert clinical nurse involvement is essential to the management of patients with this condition at the National Centre. The aim of all treatment for this condition is not only the creation of a vagina that is functional both in terms of its length and in sexual satisfaction for the couple, but also improved quality of life and psychological well-being of the patients. The risk of psychological morbidity in this group of patients cannot be overestimated (4), and the dilator treatment in particular can give rise to feelings of shame and embarrassment (5). The use of vaginal dilators in these patients has been our first-line treatment approach for the last 12 years, and we review our experience in this paper.

MATERIALS AND METHODS

Between 1998 and 2010, the total number of consecutive referrals of women

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with MRKH was 360, of whom 245 were primary referrals who were entered into our treatment program. All patients were initially seen and assessed by one of two gynecologists with extensive expertise in this condition (D.K.E. and G.L.R.). All patients received a clinical examination and an assessment of their condition, including an ultrasound scan or magnetic resonance imaging (MRI) when clinically indicated. This was followed by a discussion of the etiology of the condition, its causes, and an explanation both to the patient and, if appropriate, her parents or partner of the treatment that would be offered and the support that would be given. It was explained to all patients that dilator therapy was the initial treatment and that only if this failed would a surgical solution be offered.

The patients were then seen by a clinical nurse specialist with expertise in the creation of the vagina using vaginal dilators, and an explanation was provided again as to why we were adopting this approach as opposed to surgery. The main reasons given to the patients are that avoidance of surgery has obvious risk advantages and that the creation of the neovagina from existing vaginal tissue means that it is more likely to have better sexual function and sexual sensation. Further, a surgical approach may lead to scarring, contraction, and the need for more long-term dilation, with eventual failure in some cases with no further options. The patients were shown the dilators and introduced to the treatment program. They were also given information about the helpline telephone number, regular support group meetings, a newsletter, patient contacts, and the Web site (<http://www.mrkh.org.uk> and <http://www.imperial.nhs.uk/femgenab>), which has a password-protected chat-room.

Before they started the vaginal dilator program, all patients were then briefly screened by a psychologist who has expertise in the management of patients with this type of disorder. Patients who were considered to be experiencing psychological problems, including adjustment difficulties or other significant mental health problems, were invited for a full psychological assessment to collaboratively ascertain whether it was an appropriate time to engage in the dilator treatment. This is important to establish, as psychological difficulties may act as a barrier to dilator treatment, or, conversely, the dilator treatment may serve to exacerbate preexisting psychological difficulties.

Once the clinical and psychological assessments were completed and the treatment was deemed appropriate, the timing of the dilator therapy was then agreed on, and the patients were admitted to hospital into a single room for direct supervision. The dilator therapy included guidance on understanding the anatomy, the use of the dilators, and support. The average length of stay was 3 days, during which time dilator treatment occurred two to three times daily, always in the presence of the clinical nurse specialist. This ensured that the patients were performing the vaginal dilator therapy correctly in terms of direction, pressure, and duration of treatment. The psychologist visited the patients on the ward to monitor their well-being during the process, and any potential difficulties were identified with the view of stopping the treatment if required or offering adjunctive psychological therapy after the inpatient stay.

Repeated dilatation of the vaginal dimple, avoiding the urethra and the anus, is essential. Because they have no periods, nearly all the teenagers and young women have little knowledge of their own anatomy at the beginning of therapy. Thus, supervision is imperative for successful results. The patients adopt a semirecumbent position, and the dilator is placed on the vaginal dimple using firm pressure for 10 minutes three times a day. A lubricant in the form of KY jelly (Johnson & Johnson) or Aquagel (Ecolab) is used to facilitate easy application. After successful instruction and confidence, patients were discharged from hospital, and they then were seen at two weekly intervals by the clinical nurse specialist and if necessary by the physician to ensure that progress was supervised.

The dilators come in a set of eight different sizes (QCH dilator modified pattern; Phoenix Surgical Instruments), which are graduated. On leaving hospital, most patients have already progressed to the no. 4 and no. 5 dilators, which they use in graduated order each time they dilate. At each 2-week assessment, progress is assessed by the clinical nurse specialist, who observes a dilator session and ensures that progress in terms of depth and direction are proceeding effectively. If the therapy is proceeding successfully, the next size of dilator is added to the regime, and the smaller size is withdrawn. Progress takes on average 5.5 months (range: 2 to 19 months) (6), with most women achieving a no. 7 (14.5 × 4.5 cm) dilator by the end of this time. Only for women with partners whose penises are above average in size is it necessary to progress to no. 8 dilators.

Treatment with dilators was ceased when it was successful, satisfactory intercourse was achieved, or by size criteria defined as greater than 6 cm in length and uniform width. Throughout this time, patients were given psychological support if deemed necessary, such as helping them overcome motivational difficulties through goal setting and increased social support, or addressing any underlying diagnosis-related adjustment difficulties that arose while undergoing the dilator treatment. In those patients who were in relationships, sexual intercourse was encouraged any time that the patient felt ready. Sexual satisfaction in terms of functional length and sexual pleasure was recorded for all patients at the end of the treatment time, based on the patient's reporting. A subgroup of 60 were assessed using sexual function questionnaires, and their answers were compared to a normal population, who demonstrated equally enjoyable sexual satisfaction (6).

RESULTS

Between 1998 and 2010, 360 patients with MRKH were referred to the national center. Of these, 245 had primary referrals and no previous attempts at creating a vagina. The remaining 115 patients were referred seeking advice and psychological support, information about MRKH syndrome, and advice on fertility/surrogacy. The mean age at referral was 22.7 years (range: 14 to 56 years). Patients received a 30-minute psychological screening before embarking on treatment. Of the 245 patients with primary referrals who entered the dilator program, the mean age of commencing dilators

was 18.6 years (range: 16 to 22 years). A successful length (defined as greater than 6 cm in length and maximum width throughout the vagina and especially at the apex) and sexual function was achieved by 232 patients during this time (94.9%), with one patient lost to follow up and 13 patients not completing the treatment. The reasons for not completing treatment, as seen in Table 1, were related to social, cultural, and psychological factors. No patients since 1998 have required a surgical approach to create a vagina.

The MRKH Web site was established in 2003, and in the last 4 years has received between 86,000 and 94,000 hits. In the last year, the helpline has been used by almost 1,000 patients and health care professionals, two-thirds of whom used e-mail contact and one-third the telephone. The reported complications have been one patient with urethral irritation and inflammation, one patient with urinary leakage, and one with vaginismus.

DISCUSSION

Some patients with MRKH syndrome can create an adequate vagina using repeated natural intercourse, and the use of dilators is a modification of this technique. The ability to create a neovagina without surgery was first reported by Frank in 1938 (7), but this report received scant attention over the ensuing 40 years. Rock et al. (8) in 1983 reported a 66% success rate in 21 patients; Broadbent et al. (9) in 1984 and Roberts et al. (10) in 2001 reported results of 95% and 91% success, respectively. Ingram (11) in 1981 modified the technique used by Frank by inserting dilators into the saddle of a bicycle stool and having the patients sit astride this to gently create perineal pressure. Although many patients complained that this was an uncomfortable technique, the results were 92% success.

However, in spite of this, gynecologists have continued to seek a surgical solution rather than a nonsurgical one. Vecchietti invented an operation, again on the principle of the vaginal pressure techniques, which in his own series showed 100% success (12). Although this technique can now be performed laparoscopically with similar success, this is a surgical variant of the use of vaginal dilators. Other surgical techniques have flourished, including McIndoe-Reed graft, skin graft procedures, amnion vaginoplasty, bowel neovaginas, and peritoneal lining of the vaginoplasty. All of these techniques have success rates that range

between 80% and 90%, but none exceed the success rate of the nonsurgical approach (3).

Many patients view the creation of a vagina as a solution to attaining "normality." The adoption of a surgical approach would seem to fulfill this wish, hence the development of numerous surgical procedures. However, normality is not achieved through anatomic surgical correction alone. Some centers will still advocate surgery, but our data suggest that a nonsurgical approach offers better results. In 2006, the American Congress of Obstetricians and Gynecologists Committee (13) stated that nonsurgical techniques were the technique of first choice in all patients with an absent vagina. However, since then, there has only been one publication (14) on nonsurgical technique success and 16 publications on surgical techniques (15–30). None of these surgical techniques match the results of the nonsurgical. The definition of successful length has been variously reported from 6 to 13 cm, but the reports of vaginas 10 to 12 cm after the laparoscopic Vecchietti procedure also have described needing long-term dilator use in many patients (31, 32).

With the nonsurgical approach, we have not found it necessary to continue dilator use once satisfactory length/stretch or intercourse has been achieved. Although psychological support after completing the treatment is beneficial for patients, continued use of dilators may serve as a constant reminder of being "different." When embarking in a relationship after a substantial absence of dilators or intercourse, some women may need to use the dilators for a short time, but they very quickly and easily regain their original length. The ability to cease using dilators completely is a major psychological advantage with this treatment regime. Our own series shows that success rates of 95% can be achieved; the failures in our study could be attributed to the presence of multiple congenital abnormalities together with social, psychological, and cultural factors as opposed to a failure of the technique. When the program was completed by all patients, 100% of patients were successful.

All the surgical techniques have reported complication rates, which include vesicovaginal fistula, rectovaginal fistula, bladder perforations, graft contracture, keloid scar formation, and skin graft site disfigurement. The use of bowel in the surgery has complication rates reported as high as 20% (33). There have also been cases of malignancy reported in surgical neovaginoplasty (34), which has never been reported in a case of the nonsurgical approach (Table 2). The nonsurgical approach has none of these risks, and thus the nonsurgical approach is unequivocally safer.

It is indisputable that a multidisciplinary approach is required for all patients with MRKH syndrome. Investigators who argue that this is only required for the nonsurgical approach miss the fact that patients with MRKH have a high level of psychological distress and may require psychological support alongside the dilator therapy to help them complete the treatment. Whether they have a surgical or nonsurgical approach, the input of psychology is required and a holistic approach will determine the success of outcome. In the last 11 years, feedback from clinical experience suggests that the input of our psychologists has made a significant

TABLE 1

Reasons for patients not completing treatment.

Reason	No. of patients
Learning difficulties	1
Interpersonal conflict related to the treatment	1
Multiple congenital abnormalities	3
Significant mental health problems	2
Difficulties coping with treatment and subsequent nonattendance	2
Cultural beliefs related to medical treatment	4

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TABLE 2

Comparison of complications of nonsurgical versus surgical techniques.

Complications of nonsurgical management	Complications of neovaginoplasty
Urinary tract infection	Vaginal stenosis
Prolapse	Vesicovaginal fistula
Urinary incontinence	Rectovaginal fistula
Bleeding	Adenocarcinoma in bowel graft
	Prolapse
	Urinary incontinence; surgical challenge due to previous surgery

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contribution to changing the success rates that we have achieved. After incorporating an intervention based on support around dilator treatment, Heller-Boersma et al. (35) showed that group therapy improved psychological outcomes in MRKH patients. Our own failures and the complications we report in terms of psychological distress in this group of patients as well as the frequent use of the helpline and Web site further supports the need for psychological input in the management of dilator therapy in the context of MRKH syndrome.

One of the major limitations of the outcome of treatment for the absent vagina is the lack of data on sexual satisfaction as opposed to vaginal length. Our published data by Nadarajah et al. (6) demonstrates there was no difference in sexual desire, arousal, or orgasmic satisfaction between patients treated with vaginal dilators and controls. We did, however, find a small but statistically significant difference in the ability of the study group to vaginally lubricate during sexual arousal, and some patients do require lubricants for sexual satisfaction.

This study, the largest ever reported, shows that vaginal dilators to create a vagina in women with MRKH by a nonsurgical technique is superior to any reported surgical series and carries no physical risk to the patient. A large proportion of patients with MRKH require psychological input, and this should be an essential part of any treatment approach. We believe we have demonstrated that with the correct multidisciplinary input, surgery can become unnecessary in the management of these women. The needs of MRKH patients exceed their physical desire for a vagina, and long-term quality-of-life outcomes will only be achieved if units adopt this type of holistic approach.

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