EDITORIAL COMMENT

Congenital absence of the vagina: in search of the perfect solution. When, and by what technique, should a vagina be created?
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Purpose of review
To review the Mayer–von Rokitansky–Küster–Hauser syndrome and to address means of diagnosis, patient education and counselling. The timing of, and vast options for, creation of a functional vaginal are also discussed.

Recent findings
The diagnosis of Mayer–von Rokitansky–Küster–Hauser usually occurs during an evaluation of primary amenorrhea. Counselling and support are of great importance for affected young women and their families. Educational materials have increased with the availability of Internet web sites and there is a vast number of options for creation of a functional vagina; most international centers promote the utilization of vaginal dilators.

Summary
Young girls, adolescents and women with Mayer–von Rokitansky–Küster–Hauser should be offered a comprehensive evaluation, and presented with information regarding all options for management and support. Ongoing psycho-social and educational support is extremely important. International centers that focus on congenital anomalies of the reproductive tract should be developed. These centers of excellence will facilitate long-term follow up studies to improve patient care and evidence based medical options.


Keywords
vaginal agenesis, MRKH, reproductive tract anomalies


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Abbreviation

MRKH Mayer–von Rokitansky–Küster–Hauser

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Introduction
Mayer–von Rokitansky–Küster–Hauser (MRKH) Syndrome [1–4] describes vaginal agenesis with variable müllerian duct and possible associated renal, skeletal and auditory abnormalities. Vaginal agenesis is usually accompanied by cervical and uterine agenesis, although affected girls and women may have a normal, but obstructed, uterus, or rudimentary uterine horns with functional endometrium. It is generally thought that MRKH affects 1/5000 live-born females and results from a cessation of müllerian duct development around the fifth gestational week. MRKH Syndrome is the second most common cause of primary amenorrhea, gonadal dysgenesis being the most common cause of primary amenorrhea.

Individuals with MRKH Syndrome have a normal female 46 XX karyotype and normal ovarian hormonal/oocyte function. Since ovarian sex-steroid production is normal, the development of secondary sexual characteristics and puberty progress normally. The diagnosis is generally made because of an evaluation of primary amenorrhea; the average age at the time of diagnosis has been reported to be between 15 and 18 years of age. The presence of normal breast and pubic-hair development and normal female serum testosterone and pubertal estradiol levels makes this syndrome the likely diagnosis.

This diagnosis can be quite unsettling for a girl, young adolescent, or adult woman, and for her family. Counseling by experienced nurses, social workers, psychologists and/or psychiatrists is strongly recommended, and some people feel that it should be mandated by the treating health-care provider. It is important to stress to the young woman and her family that she has normal ovarian function, normal production of sex steroids, that a functional vagina can be created, and that fertility is possible with assisted reproductive technologies and a gestational carrier [5,6].

Treatment of MRKH Syndrome should be preceded by counseling of the young woman and her parents. Attention must be given to the psychosocial issues as well as to the correction of the anatomical abnormality. The patient’s cooperation and mental status are vital to the ultimate success of the creation of a functional vagina [7,8]. The timing for correction, non-surgical or surgical,
of this anomaly is purely elective, and thus pediatric surgeons, pediatric urologists and gynecologists should refrain from creating a vagina for girls with MRKH Syndrome during childhood. It is challenging for a child and her parents to be asked to utilize a vaginal dilator after surgical creation of a vagina during childhood. Parents commonly complain that they are not comfortable about inserting a dilator into their daughter’s vagina. Long-term follow-up has also shown that vaginas created during childhood have high failure rates and require ‘re-do’ vaginoplasties for the creation of a functional vagina [9]. Parents of girls with MRKH Syndrome may seek consultation for surgical correction during childhood to ‘resolve’ the anomaly. It is recommended that any technique for creation of a functional vagina be delayed until the mid to late teens, when the young woman (and not her parents) opts for treatment and is comfortable, and willing to be compliant, with her role in the process. Some women may elect never to create a vagina. It is important to provide patients and their families with resource information (e.g. http://www.youngwomenshealth.org or http://www.MRKH.org) presenting all options.

The creation of a vagina may be accomplished both non-operatively and operatively. The goal of any method is to create a vaginal canal of adequate diameter and length and appropriate axis to accommodate sexual intercourse. In addition, the vagina should have a ‘normal’ amount of secretion and lubrication and should require minimal care for maintenance. No procedure achieves all of these goals, and thus there are many options (each of which has associated advantages and disadvantages).

The non-operative approaches attempt to use progressive invagination of the vaginal dimple to create a vagina of adequate diameter and length. It has been noted that a functional vagina can be created with repetitive coitus [10], but care must be taken not to dilate the urethra, which can lead to urinary incontinence.

On the basis of the observed success of creation of a vagina by repetitive coitus, Frank developed the concept of the use of graduated hard glass dilators [11]. The use of vaginal dilators is the first line of therapy offered to our patients with MRKH Syndrome. The rationale for this approach is based on the fact that this technique requires no surgical intervention or anesthesia, leaves no scars, results in ‘normal vaginal lubrication’, and is completely under the control of the young women themselves. Recent studies have shown that the utilization of dilators is more than 85–90% successful in creating a functional vagina [12,13], and we have had a similar experience with our patient population. The young woman is instructed on how to use a hard dilator for constant pressure in order to create a vagina. This technique works best if she has a vaginal dimple, but can be utilized in all cases of MRKH Syndrome. It may take 2 months to 2 years to create a functional vagina, depending on patient motivation and the frequency of dilation. We have found that the greatest success can be achieved through education, nursing support, mental health counseling, and a ‘big sister’ program. Patient instructions for the utilization of vaginal dilators are available at the http://www.youngwomenshealth.org website.

To facilitate the process, Ingram modified the Frank method by using a dilator mounted on a bicycle seat upon the base of a stool [14]. Although this modification is an interesting concept, many young women initially find this technique very awkward; however, as the vagina gains length, this adaptation of the Frank method can be helpful.

Surgical procedures should be considered only if the young woman has been unable to create a functional vagina with the utilization of dilators. Surgery should not be used as a first option; standard guidelines addressing when to proceed to a surgical procedure need to be developed. Some young women are interested in a ‘quick fix’, and think that surgery will be able to achieve this goal. It is important that the health-care provider and the mental health provider help these young women to understand the short- and long-term issues related to surgery, and to appreciate the fact that most surgical interventions require the postoperative utilization of dilators.

The surgical techniques are manifold, and there is great controversy as to the best technique. Unfortunately, there is a lack of data comparing differing surgical procedures and on long-term follow-up of sexual function. A summary of the surgical procedure options is presented below.

The method most commonly used by gynecologists has been the Abbé–McIndoe vaginoplasty [15,16]. A vaginal canal is created by dissecting the potential neovaginal space. A split-thickness skin graft is taken from the buttocks, mounted on a mold, and left in place for 7 days. The graft epithelializes. Surgery is usually done in the summer or during a school vacation so that the patient has adequate time for recovery without having to miss school or answer embarrassing questions from her peers. The young woman must be counseled regarding the requirement to wear a vaginal dilator continually for 3 months postoperatively. After the initial 3 months, the patient is instructed to wear the dilator at night for 6 months (unless she is having regular intercourse). Many young women are under the false impression that if they have a surgical procedure they will be able to avoid the
The use of full-thickness skin grafts for the creation of a vagina has been described [21,22]. The proponents of the use of a full-thickness graft report that there is a decrease in the graft stricture or stenosis seen with the split-thickness technique.

The concept of dissecting the potential neovaginal space and lining the cavity with a material to facilitate epithelialization has brought forth variations on the Abbe–McIndoe vaginoplasty. Wharton created a potential vaginal space and placed a stent without a covering [23]; this technique was not popularized because of reported prolonged vaginal bleeding and discharge from granulation tissue. Modifications of this technique were made with utilization of peritoneum [24–26], amnion [27,28], and Interceed (Johnson & Johnson Patient Care, Inc., New Brunswick, New Jersey, USA) [29,30].

The William vulvovaginoplasty technique [31] involves the creation of a vaginal pouch by making a ‘U-shaped’ incision and using full-thickness skin flaps from the labia majora to create a ‘kangaroo-like’ pouch, horizontal to the perineum. Coitus and the use of a dilator assist in the creation of a functional vagina. It is particularly useful for patients with a previously failed vaginoplasty, and for those who have had radical pelvic surgery or irradiation. This procedure has not been popular in the USA as it is felt to create an awkward angle for sexual intercourse and a poor cosmetic appearance. This is not the feeling in other parts of the world: Creatsas et al. have reported greater than 90% success with their modified technique [32].

Some pediatric surgeons and urologists utilize a segment of bowel to create a vagina; this requires a laparotomy and bowel reanastomosis [33–37]. This procedure has the advantage over a McIndoe procedure in that vaginal dilators are not required to maintain a patent vagina postoperatively. Also, the patient is not required to remain subject to bedrest for 7 days (in contrast to the McIndoe procedure). Disadvantages have been reported: patients complain of the need to wear a pad daily because of the chronic vaginal discharge from the bowel mucosa; also there may be stenosis at the introitus, and the need for additional surgery. In addition, some patients find that they need to douche daily to avoid a foul-smelling odor. Because of the increasing risk of transmission of human immunodeficiency virus during adolescence and young adulthood, together with the poor barrier effect of gastrointestinal mucosa relative to skin, we prefer a procedure that does not utilize bowel. That technique is best preserved for cases in which there is no adequate space between the urethra and the rectum allowing alternative options for vaginal creation.

The Vecchetti procedure has also been used for the creation of a functional vagina [38]. A sphere, attached to two wires, is placed in the vagina dimple. The wires are guided through the potential neovaginal space to exit on the anterior abdominal wall. An apparatus is attached to exert continuous pressure, increasing daily, stretching the blind vaginal pouch. This technique was originally performed by laparotomy, but modifications have succeeded laparoscopically [39–41]. Once the vagina has been initiated with the dilation apparatus, the patient continues with the use of hard vaginal dilators to create a functional length for the vagina. The apparatus for the Vecchetti procedure is not approved by the Food and Drug Administration and thus it is not widely performed in the USA.

As one reviews the vast number of different procedures (and their modifications) for the creation of a functional vagina, one is provoked into asking the question ‘Why?’ Why does the literature supply so many different procedures designed to address this fairly uncommon congenital abnormality? One explanation may be that, because this condition is rare, the ability to perform long-term follow-up studies comparing the various options is limited. The creation of regional, national and/or international centers of excellence (as have been developed in the United Kingdom) for the care of women with complex congenital anomalies, and the development of associated databases, may help to facilitate our ability to compare differing techniques offered in various parts of the world. With the accumulation of such data, we may be able to offer evidence-based medical options when counseling our clients with MRKH Syndrome.

References


