Vaginal Reconstruction in Adolescent Females with Mayer-Rokitansky-Kuster-Hauser Syndrome

Susanne LeRoy

Throughout time, vaginal and uterine anomalies have had dramatic and disconcerting effects on individuals and families with regard to issues of femininity, reproduction, sexual options, body image, and self-esteem. Mayer-Rokitansky-Kuster-Hauser (MRKH) syndrome is characterized by complete or partial absence of the vagina, uterus, and proximal fallopian tubes. The modified Abbe-McIndoe technique is commonly used to create a neovagina. This technique is considered by many to be the procedure of choice because of its simplicity, low morbidity, and success rate. Females with MRKH syndrome have options for sexual function and the attainment of genetic offspring due to remarkable advances in plastic surgery and the use of assisted reproductive technologies and a surrogate. An improved quality of life may be the impetus for hope, optimism, and a future orientation.

Mayer-Rokitansky-Kuster-Hauser (MRKH) syndrome is characterized by complete or partial absence of the vagina, uterus, and proximal fallopian tubes. The syndrome affects 1 in 4,000-5,000 females (Fedele, Bianchi, Tozzi, Borruto, & Vignali, 1996). Goals of treatment include vaginal reconstruction, education, and psychosocial support of the patient and family.

Etiology

MRKH syndrome occurs as a result of defective mullerian duct development during female embryogenesis. This syndrome represents 90% of all cases of congenital absence of the vagina (Bellemare et al., 1998), and is the second most common cause of primary amenorrhea after gonadal dysgenesis or defective sex organ development (Fedele et al., 1996). The ovaries, distal fallopian tubes, external genitalia, endocrine status (including production of estrogen, progesterone, and ovulatory status), and secondary sexual characteristics are normal (Fedele et al., 1996). Cytogenetic evaluation reveals a 46, XX karyotype. Several familial aggregates with affected siblings have been reported, suggesting that MRKH syndrome is a rare autosomal recessive trait. However, it is unlikely that every case has an easily identifiable genetic cause (Makino et al., 1996).

Associated Anomalies

Various anomalies have been associated with MRKH syndrome (see Table 1), with renal (e.g., pelvic kidney, horseshoe kidney, unilateral renal agenesis, polycystic kidney disease) and skeletal systems (e.g., scoliosis, spine bifida, Klippel-Feil Syndrome manifested by shortness of the neck) being the most commonly affected (Alessandrescu, Petrean, Buhimschi, & Buhimschi, 1996). Other anomalies associated with MRKH syndrome are infrequently reported and include one case of splenosis, the heterotopic autotransplantation of splenic tissue (Griggs, Rudoff, & Coddington, 1990), unilateral and bilateral hearing loss (Strube, Cremer, Dikkers, & Willemsen, 1994), pigmentary skin changes in non-sun exposed areas, disorders of galactose metabolism (Cramer, Goldstein, Fraer, & Reichardt, 1996), congenital heart disease, femoral and inguinal hernias, hypoplastic thanar eminence, cleft palate, sinus inversus - lateral transposition of the organs of the thorax and abdomen (Griffin, Edwards, & Madden, 1976), myomas of the uterine remnants (Farber, Stein, & Adashi, 1978) and endometriosis (Olive & Henderson, 1987). Additionally, the association of certain histocompatibility leukocyte antigens (HLA) has been documented with certain diseases and is important in cross-matching procedures with transplanted tissues. Dabirashrafi, Mohammad, Nibkin, Tabrizi, and Azari (1995) have reported an association with the major histocompatibility complex and MRKH syndrome.

Historical Overview

In ancient times, a young woman was highly valued for her reproductive ability in order to assure "continuation of the tribe and succession of the family name" (Goldwyn, 1977, p. 319). Throughout time, vaginal and uterine anomalies have had dramatic and disconcerting effects on individuals and families with regard to issues of femininity, reproduction, sexual options, body image, and self-esteem. Pain, infection, failure, and death often occurred as physicians attempted to find ingenious ways to correct the "misdemeanors of nature" (Goldwyn, 1977, p. 327).

Reference to various degrees and types of vaginal agenesis and
obstruction have been documented as far back as 300 B.C. Hippocrates advised physicians to spare the urethra during surgery. Efforts to prevent contractures and stenosis of the vagina consisted of repetitive speculum insertions, tin tubes placed in the vagina, or twisted linen tampons soaked with iris oil, saffron, and frankincense. Granulation tissue was treated with applications of copper rust or chips mixed with a lead solution. For centuries there were no advances in treatment. However during the Renaissance, a greater understanding of the genital area occurred secondary to dissection of the human body by anatomists and artists, as well as the gradual growth of scientific inquiry and questioning of the ancient “infallibles” (Goldwyn, 1977, p. 322). In the 16th century, Matteo Realdo Columbo was the first to document the absence of both the vagina and uterus (Goldwyn, 1977). A more scholarly description was written by H.T. Engel in his doctoral thesis (Engel, 1781).

In the 19th century vaginal dilatation was maintained postoperatively with the use of “sponge tents” (Dorsey, 1818) or wax candles known as “bougies” (Abbe, 1898). Septicemia and vaginal stenosis were major problems. In the second half of the 19th century, reconstructive operations were perfected. Robert Abbe (1898) pioneered the use of split-thickness skin grafting using a “thin French rubber pouch,” which was sterilized by boiling, packed with long strips of iodoform gauze, and covered with skin grafts. Once the grafts had taken, a wax “bougie” was used for a few hours daily to prevent contractures (Abbe, 1898). McIndoe (1949-50) reported his experience with this procedure, emphasizing “ease of performance, low morbidity, excellent end result, and absence of mortality.”

During the 20th century satisfactory creation of a vagina has been accomplished using a variety of creative techniques (see Table 2). Although the perfect procedure is a

### Table 1
Anomalies Associated with MRKH Syndrome

<table>
<thead>
<tr>
<th>Renal and Skeletal</th>
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<tr>
<td>Splenosis</td>
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<tr>
<td>Hearing Loss</td>
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<tr>
<td>Pigmentary Skin Changes</td>
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<tr>
<td>(non-sun exposed areas)</td>
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<td>Congenital Heart Disease</td>
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<tr>
<td>Femoral and Inguinal Hernias</td>
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<td>Disorders of Galactose</td>
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<tr>
<td>Metabolism</td>
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<tr>
<td>Hypoplastic Thenar Eminence</td>
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<tr>
<td>Cleft Palate</td>
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<tr>
<td>Situs Inversus</td>
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<tr>
<td>Myomas of the Uterine</td>
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<tr>
<td>Remnants</td>
</tr>
<tr>
<td>Endometriosis</td>
</tr>
<tr>
<td>Presence of Human Leukocyte</td>
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<td>Antigens (HLA)</td>
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</tbody>
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### Table 2
Techniques of Vaginal Reconstruction in Patients with MRKH Syndrome

<table>
<thead>
<tr>
<th>Technique</th>
<th>Reference</th>
</tr>
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<tbody>
<tr>
<td>Intestinal segments (ileum, Sigmoids)</td>
<td>Baldwin (1904); Pratt &amp; Smith (1966)</td>
</tr>
<tr>
<td>Labia minora vascularized flaps</td>
<td>Flack, Barraza, &amp; Stevens (1993)</td>
</tr>
<tr>
<td>Progressive distention of the interlabial space</td>
<td>Vecchiotti (1979)</td>
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<tr>
<td>using pressure or traction (Vecchiotti Technique)</td>
<td></td>
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<tr>
<td>Vaginal transplantation from mother to daughter</td>
<td>Papanicolaou (1973)</td>
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<tr>
<td>Amnion</td>
<td>Jackson &amp; Rosenblatt (1994)</td>
</tr>
<tr>
<td>Interceed (Johnson &amp; Johnson, Somerville, NJ) absorbable adhesion barrier</td>
<td>Frank (1938, 1940, 1941)</td>
</tr>
<tr>
<td>Non-surgical (vaginal mold pressure) dilation of the recto-vaginal septum</td>
<td>Fedele, Busacca, Candiani, &amp; Vignali (1994)</td>
</tr>
<tr>
<td>Laparoscopic approaches</td>
<td>Abbe (1898); McIndoe &amp; Bannister (1938)</td>
</tr>
<tr>
<td>Split-thickness skin grafts using the Abbe-McIndoe technique</td>
<td></td>
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Table 3
Initial Evaluation of a Patient with Vaginal Agenesis

<table>
<thead>
<tr>
<th>Testing</th>
<th>Findings in MRKH syndrome</th>
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<tbody>
<tr>
<td>History and physical examination</td>
<td>Normal external genitalia, normal secondary sexual characteristics, vaginal agenesis, primary amenorrhea, possible associated anomalies (see Table 1)</td>
</tr>
<tr>
<td>Buccal smear</td>
<td>46, XX karyotype</td>
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<tr>
<td>Endocrine evaluation</td>
<td>Normal</td>
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<tr>
<td>Abdominal ultrasound, laparoscopy, and/or MRI (Magnetic Resonance Imaging)</td>
<td>Normal ovaries, variations in vaginal, uterine, and renal anomalies</td>
</tr>
<tr>
<td>Cystoscopy and/or retrograde pyelograms</td>
<td>Possible renal anomalies</td>
</tr>
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matter of continuous debate (Russ & Lee, 1989), the modified Abbe-McIndoe technique is considered by many to be the procedure of choice because of its simplicity, low morbidity and success rate (Alessandrescu et al., 1996). Alessandrescu et al. (1996) have used this modified technique on 201 patients with MRKH syndrome over the past 51 years (see Intraoperative and Postoperative sections).

Assessment
The diagnosis of vaginal agenesis is frequently missed at birth and often is not apparent until the age of puberty and the absence of menses (Sadov & Horton, 1988). Initial evaluation (see Table 3) is necessary to rule out testicular feminization syndrome and other forms of gonadal dysgenesis (Fedele et al., 1996). Assessment includes present perception of body image, attitude toward surgery, need for information (including issues of sexuality and fertility), and evaluation of coping strategies and support systems.

Providing Psychosocial Support
The physical, psychological, and social effects of MRKH syndrome can have dramatic and disconcerting effects on the patient as well as the family. Depression often occurs as a result of altered body image and feelings of helplessness, which are internalized rather than acknowledged or expressed. These feelings can cause a wider deficiency in school, peer relationships, dating, body image, self-esteem, family relationships, and future orientation. Inclusion of the family assists their role as part of a supportive network, as experiences of connectedness within the family are the most important predictors of health-related patterns of problem-solving and goal attainment behaviors (Ford-Gilboe, 1997). Support can also be obtained from other patients with MRKH syndrome who can share their feelings, experiences, and concerns. This can be very effective as the provision of support by those who have experienced similar stressors and situations enhances empathic understanding. Whenever possible, nurses should facilitate such supportive relationships with their former and current patients. The timing of vaginal reconstruction varies depending on the maturity, desire, and circumstances of each patient, however it is recommended between 14 and 16 years of age for psychological assurance and to enhance social integration (Sadov & Horton, 1988). There can be great variability in personal meaning and timing of surgery as a result of religious, social, moral, familial, and cultural values, which all play an important role in decision-making.

Sexuality after Vaginal Reconstruction
Sexuality is a part of human behavior and includes internal and external influences. For the adolescent with MRKH syndrome, altered body image is the result of “invisible disfigurement,” thus affecting feelings of sexuality and femininity. Surgery may be an anticipated event, however, many adolescents describe a simultaneous excitement and fear of their changed body and the responses of others, most notably potential sexual partners. Successful intercourse after vaginal reconstruction is associated with the length of the neovagina (10-12 centimeters is optimal) and the elasticity of the vaginal wall. Vaginal lubricants are highly recommended, especially when first initiating intercourse. Most patients do report adequate lubrication of the neovagina, believed to occur through local plasmatic transudate, and they also report satisfactory orgasms (Alessandrescu et al., 1996). Other than vaginal length, the most common reasons for unsatisfactory results are the absence of sexual activity (thus causing stenosis if adequate dilatation is not maintained) and complications such as infection in the postoperative period. Regrettably, long-term studies of psychosexual adjustment in patients with vaginal reconstruction have not been completed.

Fertility in Patients with MRKH Syndrome
The attainment of genetic offspring is a realistic possibility with the use of assisted reproductive tech-
nologies and the use of a surrogate. This information should be discussed with patients and their families in order to alleviate anxiety if future fertility is desired. The technology of in-vitro fertilization and embryo transfer (IVF-ET) allows for ovarian stimulation with human menopausal gonadotropins, ultrasound guided aspiration of oocytes from the genetic mother (through the lateral fornix of the neovagina or laparoscopic retrieval if there are anatomic difficulties), fertilization with sperm from the genetic father, and embryo transfer to a surrogate or gestational carrier (Wood, Batzer, & Corson, 1999). Patients are informed that the possibility of genetic transmission is unlikely but that it cannot be entirely excluded (Petroska, Gray, Davis, & Reindollar, 1997).

Preoperative Workup

The preoperative workup includes a history and physical, with special attention to pertinent needs of the adolescent with MRKH syndrome. Results of the initial evaluation (see Table 3) and testing, as well as the presence of associated anomalies (see Table 1) or presence of chronic illness need to be documented and addressed with the patient and her family. Allergies and specific reactions to medications, shellfish, iodine, tape, and latex are documented and discussed. Potential skin graft sites are assessed for the presence of tattoos, sun damage, pigmentedary skin changes, moles, scars/keloid, body piercing, and the general condition of the skin.

It is essential to speak privately with the adolescent. Privacy enhances the adolescent’s trust in the nurse and promotes a more honest response to intimate and sensitive questions. It is important to ascertain use of over-the-counter medications, herbal remedies, aspirin, diet pills, caffeine, illegal drugs (mode of use, oral ingestion, smoking, snorting, or injecting), inhalants (toxic fumes such as gasoline or glue), smoking (cigarettes and/or marijuana), and/or alcohol use. Clear explanations for discontinuing these substances should include “graft take,” anesthetic interactions, prolonged bleeding, and the potential for risk behaviors resulting from altered levels of consciousness. Sexual activity and sexually transmitted diseases need to be addressed as some adolescents may have attempted vaginal intercourse without success but may have experimented with oral and/or anal intercourse. A nonjudgmental manner that fosters openness and trust is imperative. Sexual activity and potential sexual activity are essential topics to address preoperatively, including condoms for prevention of sexually transmitted diseases. Additionally, general health promotion behaviors, including seat belt use, access to cigarettes and alcohol, presence of fire alarm in the home, exposure to violence, access to firearms or weapons, nutritional patterns, and social support (family, peers, others who may be important), should be reviewed to assess risk and protective factors for the adolescent during the perioperative process. Adolescents will rarely initiate these discussions, however they will usually respond honestly in a nonthreatening, supportive atmosphere if confidentiality is assured. Education assists in alleviating anxiety and provides a sense of understanding and mastery for the adolescent. The patient is taught about the surgical procedure/skin grafting, NPO status, bedrest during hospitalization, avoidance of thromboembolic phenomena with deep breathing and ankle/ calf movements, and medication for pain control is discussed. Additionally the patient is given the opportunity to see and touch a vaginal mold before surgery.

Intraoperative Care

As for all patients, it is essential that privacy is maintained. The patient is covered with warm blankets during transport and transfer to the operating table, and supportive personnel accompany her through induction of anesthesia. Skin integrity, positioning, prevention of hypothermia, prevention of infection, and care of the graft and donor sites are integral to the safety and outcomes for this patient.

Under general anesthesia, the patient is catheterized to protect the urethra against possible trauma during the dissection of the urethrovaginal space for postoperative urine drainage, and to prevent contamination of the external genitalia by urine. A split-thickness skin graft is taken from the gluteal region or thigh (lateral to the hair-bearing area) and is sutured over a rigid mold with the outer surface of the graft facing outward. A fibrin sponge is applied to the skin graft site and is covered with bandages moistened in antibiotic cream. The patient is then placed in the lithotomy position. Using aseptic technique, the labia minora are separated with traction sutures at the caudal extremity and a transverse incision is made between the traction sutures. An additional incision is made in the midline approximately 1.5 to 2 centimeters below the urethra to broaden the newly created space. The size of the mold is adapted to the urethrovaginal space, which usually reaches 10 to 12 centimeters. The rigid plastic mold is adapted to a length of 8 to 12 centimeters and a diameter of 2.5 to 3 centimeters. The edge of the graft is then sutured to the perineal and hymenal flaps, and the edge of the mold is sutured to the perineal skin to direct the mold ventrally toward the bladder (Alessandrescu et al., 1996).

Postoperative

Postoperative management includes one week of hospitalized bedrest to promote “graft take.” The patient has an indwelling catheter for urine drainage while bedridden, and to prevent contamination of the external genitalia by urine. Skin integrity, infection, thromboembolic phenomena, pain control, anxiety, and altered body image are important implications for nursing care. Discharge planning begins during the week of imposed bedrest. This is a time to illicit discussions of body image and issues regarding self-care and potential future sexual activity. At discharge, the mesh is replaced with a semi-rigid mold covered with a condom. The patient is instructed on how to remove the mold and to self-administer a low-pressure douche with warm water.

After approximately 1 week, the rigid mold is removed, the vagina is irrigated with normal saline, and the mold is replaced with a sterile humidified mesh which is changed daily for the next week.

In cases where the patient can-
not have regular intercourse for reasons other than those related to the surgical procedure, she is instructed to wear a semi-rigid mold for the following 6 months or until she becomes sexually active.

Follow up is 1 week after discharge and the patient is encouraged to call with any questions or concerns. Postoperative follow up continues for 6 months and routine gynecological care, including an annual speculum as well as bimanual exam, is emphasized. This is important as endometriosis and myomas have been reported in some patients with uterine remnants (Beecham & Skinderziewski, 1977; Rosenfeld & Lecher, 1981). Squamous carcinoma of the neovagina has been reported in vaginal reconstruction using skin grafts (Steffanoff, 1973), and two cases of ovarian cancer in patients with MRKII syndrome have been documented (Ghirardini & Magnani, 1995).

**Summary**

Females with MRKII syndrome have options for sexual function as well as fertility due to remarkable advances in plastic surgery and assisted reproductive technologies. An improved quality of life may be the impetus for hope, optimism, and a future orientation.

**References**


trosurgery and conducting a literature search on electrosurgical smoke. The researchers decided that masks were an ineffective barrier to smoke and also provided no protection to the patient. Inline smoke plume evacuation systems were deemed unacceptable because of cost and potentially inadequate wall suction. The researchers concluded that the only safe and effective method was to use dedicated smoke plume evacuation systems with hands-free operation.

The first step undertaken was to determine how many smoke plume evacuation systems were necessary. This was determined by establishing which procedures would require smoke evacuation and then reviewing statistics to determine the number of procedures. After reviewing this information and establishing criteria, a report containing information about the dangers of ESU smoke plume, recommended practices, a discussion of equipment options, and conclusions about what needed to be done was prepared. This report was given to safety officers, the risk management director, the director of materials management, the director of nursing, and other pertinent areas. They then developed the smoke plume evacuation system evaluation tool. After establishing requirements for clinical trial participation, they were ready to trial various systems. Systems were evaluated for effectiveness, ease of use, noise level, versatility, electrosurgical handpieces, on-switches, obstruction of the surgeon’s view, and cost effectiveness. After the period of evaluation, the researchers concluded that there were no perfect models. Since they believed that there were compelling reasons to begin ESU smoke plume evacuation as soon as possible, they chose what they considered to be the best system available.

Before implementing smoke plume evacuation in the institution, the researchers developed a policy and procedure that would be updated annually. Following implementation, they were surprised and pleased with the level of compliance and the expressions of gratitude from physicians and staff members during the period of evaluation.

**Comments.** This article provides valuable information for anyone who is considering changes in the manner in which surgical smoke plume is being handled in their institution. The author and her colleagues seem to have considered all concerned areas as they prepared to undergo this study. The tools that they developed could be used by any institution. They also noted that since this new area of concern has not been adequately researched, there will most likely be additional concerns identified and better evacuation devices being produced.

**References**


Abstract. The author reported on the release of a draft of guidelines for healthcare workers exposed to laser and electrosurgical smoke. The author briefly describes the contents of the draft and OSHA’s recommendations. She also addresses the efforts of other organizations, such as the National Institute of Safety and Health and AORN, in developing awareness of the hazards of surgical smoke. The meetings held as a result of these efforts were attended by representatives from a variety of surgical specialties. The author concludes that it is important to monitor progress of the review of the draft.

The draft, prepared by OSHA, was sent to external reviewers for their comments, which could be incorporated into the document before final publication. The 20-page document covers personnel, types of ESUs, types of lasers, constituents of smoke, pathophysiologic effects of smoke, infectious potential of laser and ESU plume, methods of protection, conclusions, and recommendations. The document is intended to offer guidance for those people exposed to smoke plume. The document recommends that surgical smoke should be removed and properly filtered by a smoke evacuation system as close to the surgical site as possible. An additional recommendation is that staff members exposed to laser or ESU smoke should wear respirators, as traditional surgical masks are inadequate.

**Comments.** This article, written a year later than the above Lanfranchi article, shows that there continues to be a great concern over the hazards of laser and electrosurgical smoke.

In spite of the continued interest in the subject, there have yet to be any specific regulations mandating that institutions be required to have procedures established to effectively protect the patient and employee from the smoke.

**Vaginal Reconstruction continued from page 27**

Obstetrics and Gynecology, 135, 105.


