Vaginal agenesis involves issues of physical abnormality, body image, sexual identity, and sexual/reproductive functioning that require long-term medical and psychological management. The authors detail the nonsurgical Frank procedure for creating a vagina and discuss counseling techniques for the patient and her family.

Vaginal agenesis, which occurs in approximately 1 in every 5,000 to 7,000 female births, is a significant threat to the mental health and well-being of an otherwise normal, healthy young woman. More than 90% of patients with vaginal agenesis fulfill the criteria for the Rokitansky-Kuster-Hauser syndrome—ie, 46,XX karyotype with normal, functioning ovaries. Although it is typically absent, about 5% of patients have a uterus.

Approximately 7% to 8% of the remaining patients have a more unusual genetic abnormality and fulfill the criteria for testicular feminization (androgen insensitivity syndrome). Genetically, these patients are 46,XY and prove to have only testicular tissue at gonadectomy. A few patients will have yet other chromosomal/gonadal variations; however, the procedure is basically the same whether counseling young women with 46,XX or 46,XY karyotypes.

**Diagnosis**

In all of these patients, the external genitalia are essentially normal. In place of the vagina, there is typically a small pouch or dimple that is 1 to 4 cm in depth. Thorough evaluation at the time of diagnosis is paramount and should include phenotypic studies to determine the exact karyotype. Intravenous py-
Elography is also important since a number of these patients have a congenital urinary tract abnormality. Skeletal abnormalities may also be seen to a lesser degree. Pelvic ultrasonography may be helpful in the differential diagnosis. Vaginal agenesis may be inappropriately diagnosed as an imperforate hymen, and misguided attempts at hymenotomy may result in secondary scar formation. This mismanagement can lead to treatment difficulties in the future.

A further concern is the potential for iatrogenically induced trauma stemming from the physicianpatient discussion. The patient should be told that she was born with an incompletely developed vagina; the expression "born without a vagina" should be avoided. The physician also should reassure the patient that a "more functional" vagina can be created, avoiding references to an "artificial" vagina. This is especially important in the case of the physician who first sees the patient and her family, since inappropriate comments at this point can leave an indelible mark that may require intensive therapy to remove.

**Timing of Treatment**

There is some debate about when these patients should be treated. We believe that medical treatment should be deferred until the patient attains full growth, is psychologically prepared, and is socially and sexually mature. Some pediatric surgeons are much more aggressive in the surgical correction of these abnormalities and choose to treat patients long before they enter their teens. We are strongly opposed to this, because all of the corrective procedures carry a risk of complications and compromised results. Moreover, in the preteen patient, a dilator must be used for a long period before the beginning of sexual activity, and the pediatrician must depend more on parental involvement with vaginal dilatation. If the patient is too young to understand dilatation, then reactions of anxiety, anger, depression, and fear can become associated with the parents' attempt to continue this mechanical therapy. Thus, ironically, procedures designed to promote adjustment and normalcy for these patients can instead result in psychosexual problems.

**Procedures**

**Nonsurgical Approach** Currently, the two most popular methods for creating a vagina in these cases are the nonsurgical Frank procedure and the McIndoe split-thickness skin-graft vaginoplasty. The Frank technique was initially described in 1938. The goal was to increase the depth and caliber of the vagina with the use of graduated dilators, thus avoiding the need for surgical intervention. In the past, patients were advised to sit on a hard stool or firm chair with the tip of the dilator inserted through the hymenal ring into the vaginal dimple. This method was not particularly successful, but the reasons for its overall failure are unclear.

The Ingram “bicycle seat” represented a dramatic improvement in the Frank procedure. The major

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**Case History**

Jane, a 17-year-old girl with vaginal agenesis, was referred by her physician for sexual counseling about interactions with others, self-esteem, and sexual functioning. She had a boyfriend and engaged in other normal adolescent interactions. The disorder was discovered when her mother became concerned that she was not menstruating and took her to the family physician, who referred her to a gynecologist. After appropriate history-taking, physical and pelvic examination, and a thorough discussion of therapy, Jane felt highly motivated to use the Frank nonoperative procedure.

Jane’s mother was perceived by both the physician and the patient as being more upset about the diagnosis than Jane herself and requested several counseling sessions alone. Jane said that her mother’s presence at the initial counseling session was helpful for both of them but requested that they receive subsequent counseling separately. Although the mother was religiously conservative, she expressed no discomfort about discussing sexual functioning and vaginal dilatation with her daughter. They requested any written information that was available.

The counseling sessions took place over 6 months, with weekly visits for the first 6 weeks and then less frequent meetings. At first Jane used the counseling for emotional support and guidance in the Frank procedure. She found that wearing Spandex bicycle pants was effective in keeping the dilator in place while she used a stationary exercise bike; this gave her a sense of control and normalcy.

As Jane’s self-confidence increased, subsequent discussions focused more on her social interactions—especially those with her boyfriend. Although not sexually active at the time, she was particularly interested in rehearsing with the counselor how to explain her disorder if the possibility of sexual intercourse should arise. She decided not to discuss her disorder with her boyfriend unless their sexual relationship progressed beyond kissing and touching and they were considering intercourse. Should this occur, she decided to tell him matter-of-factly that she had been born with a vagina that had not developed completely but that this abnormality had been corrected. She also decided to say that she had normal, functioning ovaries, but was born without a uterus and could not have children. She wanted to reassure him that she was normal in all other aspects of female functioning and that after she had finished school and started a career, she hoped to become a mother through adoption or surrogate motherhood. She went over what she would say with the counselor until she was comfortable discussing these issues.

Follow-up examinations at 6 months indicated that Jane had sustained her motivation and was pleased with the results of the dilator therapy. A subsequent examination when she was 20 indicated continued satisfaction with the nonsurgical approach. The vagina was 12 cm deep and had a satisfactory caliber.
advantage of this racing-type bicycle seat is that it is positioned between the buttocks and is therefore in better contact with the perineum, providing direct pressure from the graduated dilator on the incompletely developed vagina.

First, the patient is instructed to use a mirror to examine her genitalia at home, identifying the external structures and introit dimple. Dilators of graduated sizes are used to create pressure on the vaginal dimple, beginning with a dilator approximately 1.5 cm in diameter and 2 to 3 cm in length. She can use either a stationary bicycle or a multilegged stand with the seat attached and can study, read, or watch television while using it for 20 to 30 minutes three to four times daily. She should be seen monthly, not only for physical evaluation but also for continued encouragement and motivation; she should be made aware that consistent use of this technique will enable her to avoid surgery and will eliminate the risk of postoperative scarring and the need for painful, potentially disfiguring skin grafting. Steady progress should be readily apparent on periodic pelvic examination. Adequate vaginal caliber and depth can usually be demonstrated in 14 to 16 weeks.

On occasion, the position may be difficult to manage because of the shallowness of the vaginal dimple. In these cases, the patient can begin with the lithotomy position, applying manual pressure to hold the dilator firmly in contact with the vaginal dimple until there is sufficient invagination to accommodate the bicycle technique comfortably. Some women can pedal a stationary bicycle during dilatation after several weeks of nonsurgical treatment, but this should be discontinued if there is any chaffing or discomfort. The Ingram technique is successful in approximately 90% of cases, and Ingram himself contended that surgery should not be considered until the patient has undergone a sufficient trial of this approach.

Once satisfactory invagination is accomplished, the patient can begin to use a polyfoam dilator held completely within the vagina by the distal levator muscles. This dilator can consist of any type of soft, cylindrical polyfoam material that can be molded to the shape of the neovagina. This material can be obtained from a local craft store. The dilator is covered with a condom and inserted with the open-ended portion of the condom outside the vagina to permit easy removal. It should be worn for 10 to 12 hours daily, either throughout the day or during the night.

"The Ingram technique is successful in approximately 90% of cases, and surgery should not be considered until the patient has undergone a sufficient trial of this approach.'

Surgical Approach Should the nonsurgical technique fail or prove unsuccessful due to lack of motivation, the most popular surgical approach at present is the McIndoe split-thickness skin-graft vaginoplasty. It can be highly successful but requires an experienced surgeon, close observation, and consistent use of the vaginal insert in nonsexually active patients.

If surgery is necessary, the patient can be reassured that the results are quite satisfactory. After an adequate vaginal vault is surgically prepared, a 6- to 8-cm x 18- to 20-cm split-thickness skin graft is obtained from the anterior or posterior thigh or the buttocks. The graft is applied to an obturator that is placed in the vaginal vault at the recipient site. This dilator is removed on the fifth or sixth postoperative day. The patient is then fitted with a polyfoam dilator, which she uses as described above. Other methods using amnion, peritoneum, or a segment of bowel have been described, but their success has been somewhat limited.

Counseling Professional counseling of the patient and her parents should begin shortly after the abnormality is recognized. However, the physician can help by introducing the subject with appropriate comments at diagnosis; indeed, failing to address the impact of this diagnosis immediately can contribute to subsequent poor psychological adaptation. Early referral for counseling is essential for all of these patients to help them deal with issues of inadequacy, gender/sexual identity and functioning, and motherhood. Low self-esteem and alterations in anatomic development can threaten the patient's self-image and feeling of femininity. Even if the patient seems well-adjusted, she should still be given the opportunity over the long term to discuss any concerns with a professional sex therapist. In both medical treatment and counseling, the concept that sexual identity is more comprehensive than sexual functioning should be emphasized.

Birth to Age 6 If the diagnosis is made during this period, parental guidance is paramount. The parents may experience feelings of guilt or anxiety and anticipate difficulties in their child's future sexual and reproductive functioning. The following guidelines are useful to physicians working with the parents:

- Answer the parents' questions. All questions should be answered as honestly as possible, with re-assurances that medical care and counseling will be available throughout the child's development. The physician should also prepare the parents to answer the child's questions as they arise.
- Provide information. As they would in the case of other congenital or acquired medical disorders, parents need detailed information. When parents are properly educated about their child's condition and ways in which to answer her questions, successful adjustment can occur in even the most complicated cases.
• Address gender issues. Even as early as age 2, a girl begins to understand gender/sexual identity by identifying with her mother and other women. By age 3 or 4, she can recognize anatomic differences. Parents must be prepared to discuss aspects of psychosexual identity stressing early female identification as well as anatomic gender differences. They can benefit from preparation for questions their child might ask during various developmental stages. For example, the child who asks, “Will I be a mommy and have a baby in my tummy when I grow up?” can be told that sometimes mothers cannot have babies in their “tummies” for many reasons and that there are other ways to be a “mommy.” These other ways can be detailed as the child asks. As with other 3- or 4-year olds, the child needs to understand that girls have vulvas and vaginas and boys have penises. At this point, the parent can mention that sometimes a girl’s vagina does not develop completely and that doctors can help it to develop more when she is older. Emphasis should be placed on the female characteristics the child has, on female-associated roles, and on reassurances that parents and doctors will be there to help.

• Assume that parents need support. Parents typically feel guilt, self-blame, anxiety about sexual education, and grief that their child is physically abnormal and will not be able to bear children. If the physician exhibits ease and reassurance in discussing these topics, the parents will be better able to cope with their own feelings and those of their child. Again, it is helpful at diagnosis to recommend that the parents speak with a certified sex counselor. They can also be given appropriate material to read.9,12

Ages 6 to 11 This is when most children develop a sense of competency about their bodies and their mental and physical abilities. The child who has already been diagnosed will continue to need reassurance about her female identity. Questions about female anatomy and physiology can be answered by both the physician and the parents, providing only as much detail as the child desires. If the diagnosis is made at this age, the physician can prepare the parents and child according to the guidelines given previously. Special considerations include:

• Development of secondary sexual characteristics. Hormonal changes resulting in the development of secondary sexual characteristics such as breast buds and pubic hair may begin by age 8 or 9. The physician may want to point out that this development is normal and part of the child’s overall female sexual maturation.

• Development of the clitoris. In discussing anatomy, the physician and counselor can provide a general introduction to the female genitalia and reproductive organs-including the clitoris and its role in sexual arousal and orgasm. This will reassure parents that their child will have normal physiologic responses. The absence of the uterus should also be discussed at this time to explain to the patient her inability to become pregnant.

• Absence of menstruation. The physician can present a simple explanation of the menstrual cycle to the patient as part of a discussion about the reason for the lack of menstruation in her case. It is helpful for the patient to know about the interrelationship between the ovaries and the uterus, but it is more important for her to understand that her ovaries are functioning normally in the absence of a uterus. The physician and counselors should try to anticipate and raise questions that may be on the child’s or parents’ minds rather than waiting to be asked—but care should be taken not to “force-feed” the child overwhelming amounts of information.

• Evaluation of concerns. Often, the young patient and her parents will have questions about physiologic functioning and psychosocial adjustment. They will also have concerns related to treatment options in the development of a more functional vagina. It is important to explain that there are straightforward methods for creating a more completely developed vagina. If this raises further questions, then the specific details must be presented; otherwise, they can be discussed later in the child’s development.

The issue of pregnancy may resurface at this time. The patient can now be told that there are a variety of reasons for a woman’s not being able to become pregnant and that the problem may involve one or both partners; in her case, the reason will be the absence of a uterus. The parents, physician, and counselor can reassure her that there are other ways of becoming a mother, including adoption or surrogate motherhood.

"During arousal, the woman with vaginal agenesis experiences pelvic congestion with resultant lubrication in the genital area and will have an orgasm in response."

Ages 11 to 15 Any adolescent girl has many psychosexual concerns and questions. She wonders if she is like her peers and worries about being different. For the young girl with vaginal agenesis who has grown up aware of her disorder, there may be less trauma associated with puberty. Continued reassurance and additional age-appropriate information about sexual functioning, physiology, and sexrole identification can augment the understanding she already has.

When vaginal agenesis is diagnosed in adolescence—often as the result of an investigation of primary amenorrhea—the girl may react with shock and disbelief. She will fear rejection, imagine herself to be unattractive, and be concerned about sexual relationships and marital potential. Her grief about her
inability to have children must be handled in a very supportive and understanding way. In addition, there will be concern about what to tell others—especially a boyfriend.13,16

The adolescent girl and her parents often ask questions about physiology and anatomy (e.g., absence of menarche, failure of the vagina to lengthen). Underlying these concerns is usually the more basic question of self-esteem—“How normal am I?” The physician and counselor can reassure the patient that other women with vaginal agenesis have had similar concerns and have adapted well and that in all other ways her body is female and is functioning normally. They can promote healthy adaptation and acceptance by treating the patient as a normal young woman with normal questions. The opportunity to speak with someone who has vaginal agenesis and has adjusted successfully is often useful. Special considerations include:

- **Physiology.** The physician should reassure the patient and her parents by citing the normal development of her breasts, ovaries, pubic hair, vulva, and clitoris. The patient can be told that she will be able to experience normal orgasmic responses. During arousal, a woman with vaginal agenesis experiences pelvic congestion with resultant lubrication in the genital area and will have an orgasm in response to the buildup of neuromuscular tension and the spasmocnic contractions of the pelvic muscles.

- **The role of hormones.** Women with vaginal agenesis who have a 46,XX karyotype have normal female levels of the hormones estrogen and progesterone, and they should I), reassuring concerning ovarian function. This means they will ovulate and have the potential for in vitro fertilization. In cases of other karyotypic abnormalities, the patient can be treated with appropriate hormone replacement therapy.

- **Sex roles and fertility.** Both the sex counselor and the physician should anticipate questions about selection of a partner, marriage, and motherhood. When patients ask whether they will be attractive to men, they can be reassured that women with vaginal agenesis find partners, date, and marry just as other women do. The most important predictor of a satisfying relationship with a partner is the quality of the relationships the young woman already has with family and friends. After appropriate counseling, women with this disorder do not perceive vaginal agenesis as a deterrent to dating and having a satisfying relationship with a partner.

In terms of fertility, a number of options are available. Many of these women choose to adopt children, but recent developments have created other options. One mother asked if her own uterus could be transplanted and attached to her daughter's ovaries. This is not medically possible now, but it is possible to harvest the patient's eggs, inseminate them with her partner's sperm, and implant them in another woman's uterus.

- **Treatment.** The adolescent patient and her parents will need a complete understanding of the available treatment options. Unless there are contraindications, we favor the nonsurgical Frank procedure. Again, should this be unsuccessful, a McIndoe split-thickness skin-graft vaginoplasty can be performed.

**Age 15 and Older** If the diagnosis is made at this age, the guidelines outlined previously can be followed. The patient may want to discuss her concerns with the physician or counselor without the parents being present, but it is important to continue to address parental concerns as well. An in-depth discussion should include:

- **Treatment.** The nonsurgical Frank procedure and the McIndoe split-thickness skin-graft vaginoplasty should be discussed as alternatives. The patient should be strongly encouraged to attempt a trial of the nonsurgical approach if at all possible, since it is more anatomic, less painful, requires no healing, and leaves no scarring. The graduated vaginal dilators can be prescribed by the patient's personal physician or can be ordered from a medical supplier (see resource list). Several detailed discussions of the nonsurgical method are available in the literature.1,2,17

- **Results and reactions.** Initially, the patient may express anxiety about the Frank procedure. Common concerns include pain and damage to tissues. The patient can be reassured that the procedure is neither painful nor damaging when the directions are followed. Women report that adjusting to dilator therapy takes some motivation and encouragement and means restructuring one's lifestyle to provide daily periods of privacy.

If active or passive dilatation must be discontinued for a time, causing substantial foreshortening of the neovagina, it will be possible to reestablish the nonsurgical treatment with satisfactory results over a much shorter period of time than originally. If sexual intercourse occurs no more than once every 2 weeks, the condom-covered vaginal insert should be worn for a 10- to 12-hour period three to four times per week. Individual variations on this program can be established by the patient.

- **Sexual functioning and interaction.** Concerns about personal and sexual relationships should be addressed with a review of the patient's normal sex characteristics, hormonal activity, and orgasmic function. Information about human immunodeficiency virus and other sexually transmitted diseases should be included. It may be necessary to reiterate that the patient will not menstruate even after a more functional vagina is created and that she still will have no uterus and cannot become pregnant. The patient may also worry over the possibility of having other physical abnormalities, whether it is "normal" to have a more functional vagina created, and what potential partners may think. Creating time for questions and reassurance is important. Each question should be addressed individually and at the time it is asked.
Sometimes a woman asks whether intercourse will be painful or damage the vagina, and her partner may be concerned about hurting her during intercourse. Some women initially report a feeling of pressure, tightness, or soreness during or after intercourse, but these symptoms abate with repeated activity. The physician and counselor should emphasize sexual responsiveness, the importance of being relaxed and aroused during foreplay, and the possibility of using lubricants during intercourse. Some women report aroused during foreplay, and the possibility of using responsiveness, the importance of being relaxed and periodic use of the vaginal dilator helps to maintain vaginal length and elasticity. The couple can be reassured that post treatment sexual relations can be satisfactorily adjusted to the diagnosis and intervention. In this way, the creation of a vagina is integrated with the patient’s gender identity, self-esteem, and sexual functioning.

Resource List for Vaginal Dilators

Dilators can be purchased from the following companies:
- Syracuse Medical Devices, Inc, 102 White Heron Circle, Fayetteville, NY 13066, (315) 637-9275.
- Ingram Dilators, Faulkner Plastics, Inc, 4504 E Hillsboro Avenue, Tampa, FL 33610, (813) 621-4703.
- F.E. Young Dilators, 1350 Old Skokie Road, Highland Park, IL 60035.
- Milex Products, Inc, 5915 Northwest Highway, Chicago, IL 60631, (708) 831-4080.

References


All patients with müllerian agenesis should be offered counseling and encouraged to connect with peer support groups. Future options for having children should be addressed with patients: options include adoption and gestational surrogacy. Assisted reproductive techniques with use of a gestational carrier (surrogate) have been shown to be successful for women with müllerian agenesis. This information may be useful if the patient requires urgent medical care or emergency surgery by a health care provider unfamiliar with müllerian agenesis. The patient with a low transverse vaginal septum usually will have a normal hymen with more proximal obstruction of the vaginal canal. A rectal examination often will identify a bulging of the proximal vagina. Almost all of the people diagnosed with vaginal agenesis have never heard of it. A congenital disorder of the reproductive system that affects roughly one in 5,000, it occurs when the canal connecting the cervix to the vulva stops developing. The condition typically is diagnosed when a girl enters puberty and fails to begin menstruating. Those patients who retain a uterus may be treated first in an emergency room because of abdominal pain resulting from blocked menstrual blood. Because of the amount of self-care required with this procedure, she tries to delay the operation until patients become sexually active in their late teens or early 20s. Intercourse should eventually help maintain the new vagina. Kolp is also working on a device to facilitate the early stages of vaginal construction. Agenesis of the lower vagina.


Development of the female genital tract is a complex process that is 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. Anomalies of the hymen and vagina may interfere with menstruation, sexual activity, fertility, or childbirth. These typically present after puberty, but are discovered during the neonatal period in some infants. The diagnosis and management of congenital anomalies of the vagina and hymen will be reviewed here. Congenital cervical and uterine anomalies.