

THE FEMALE PATIENT

Care and Counseling of the Patient With Vaginal Agenesis

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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 physician/patient 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 1938.¹ 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.² The major

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 maintained 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.

- 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

Agnes 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."



Agnes 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 (eg, 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 spasmodic 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 be reassured 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.

Originally published as: Foley, Sallie, and George W. Morley. 1992. Care and Counseling of the Patient with Vaginal Agenesis. *The Female Patient* 17 (October):73-80.

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