

MRKH: A GUIDE FOR PARENTS AND GUARDIANS

Learning that your daughter has MRKH may bring up many questions and emotions for parents. It's normal to be worried about how your daughter is coping and it's important to be aware of your own reactions to this news. Discussing issues such as puberty, reproduction, and sexuality is often challenging for parents. When your daughter is diagnosed with vaginal agenesis, you may feel overwhelmed with your own concerns, your lack of knowledge about the condition and how to discuss it with your daughter, your family, and her primary health care provider or pediatrician. This information guide is intended to help you to understand MRKH syndrome, answer your concerns, and help you support your daughter. Commonly asked questions and concerns are included at the end of this guide.

What is MRKH?

MRKH is a congenital condition of the female reproductive system that affects approximately 1 out of every 5000 females. Girls diagnosed with MRKH have **vaginal agenesis**, which refers to an absent or incomplete vagina. The uterus is also very small or absent. It's important to understand that young women with this syndrome are genetic females. They have normal ovaries and will experience puberty without having periods. MRKH is a syndrome that may or may not be associated with renal (kidney), skeletal, and hearing problems.

TREATMENT OPTIONS

If your daughter has been told that she has MRKH and her vagina is incomplete or absent, she has the following options:

1. **She can do nothing.**
2. **She can create a vagina *without* surgery, using dilators** that expand and stretch her existing vagina over time, using intermittent pressure. *This is a nonsurgical procedure.*
3. **She can have a surgical procedure, or "vaginoplasty", which involves surgery** (using a skin graft from another part of her body).

4. **She can have a procedure that involves a combination of an operation and dilation** with continuous pressure: **Vecchetti procedure.**

WAITING OR DOING NOTHING

Your daughter ultimately has control of her own body. *Deciding to make or not make a vagina should be **her decision**.* If she decides she would like treatment, "when" should be her decision too! If your daughter plans on having vaginal intercourse at some point in the future, making a vagina is something she may wish to consider.

Aside from doing nothing, there are three basic approaches to expand or create a vagina. If your daughter is thinking about treatment, she should definitely consider using dilators first. **All of the surgical options require general anesthesia and the use of dilators after each procedure, therefore surgery is not a quick fix, or a way to avoid the need for using dilators.**

ACCEPTED STANDARDS OF CARE

The American College of Obstetricians and Gynecologists recommend vaginal dilators without surgery as the optimal treatment for creating a vagina. Health care providers recognize that this treatment is very effective and is noninvasive (does not have the risks of surgery or anesthesia).

DILATORS

Sometimes the use of dilators is referred to as the **Frank Method**, named after the first health care provider who became famous for introducing a treatment that a young woman could do on her own, in the privacy of her own home. The original dilators were made out of glass. In the early 1980's, Dr. Ingram created different size plastic dilators along with a bicycle seat that attached to them. Young women were then taught how to dilate using this special seat on a stationary bicycle. The Gynecology Program at Children's Hospital Boston does not recommend the Ingram method because it's difficult and awkward to use.



Today, hard *plastic dilators* have replaced glass dilators. There are different kinds of dilators available, but the most widely prescribed dilators come in a set with different sizes ranging from XS–L. The dilators look very similar to tampons. Starting with the smallest dilator, a young woman learns how to hold it and apply pressure to stretch her vagina. In the beginning, most of the dilator is used as a “handle” (as it can only go in so far). Later, as the vagina stretches, more of the dilator can be inserted into the vagina. If your daughter is a patient at Children's Hospital Boston and selects this method of treatment, Dr. Laufer's nurse specialist will teach her how to use the dilators. She will take one home with her and use it twice a day for about 20 minutes. She will likely return for follow-up appointments once a month. As her vagina stretches, Dr. Laufer will give her the next size dilator, which will be slightly wider. The length of time it will take to create a vagina will depend on how consistent your daughter is with using the dilator. Typically it takes about 6–12 months but can take as little as 4–6 months if dilation is done consistently, 2–3 times a day.

Is dilator treatment successful?

Dilator treatment is very successful. However, there is great variation in the success of dilator treatment, mainly related to the teaching and expertise of the health care providers who teach their patients this method. **Make sure that your daughter is followed by a gynecology program that has at least an 85–95% success rate for helping their patients create a functional vagina. Both you and she should ask the gynecologist how many patients he/she treats monthly/yearly who have vaginal agenesis (incomplete or absent vagina) and ask about their success rate.** If the program or health care provider that you are working with has a low success rate, your daughter will likely get frustrated during treatment, have a poor chance of success using dilators, and thus be at a higher risk of needing a surgical procedure.

How often will my daughter need to see her gynecologist?

Generally, your daughter will be asked to return for monthly follow-up appointments so that her progress can be monitored. It's VERY important that your daughter keeps her appointments with her gynecologist so he/she can check her progress

(whether she has decided to use dilators or have surgery). If she decides to use dilators, her gynecologist will need to check to see if she's applying pressure in the right place and at the correct angle. As her vagina stretches, her gynecologist will give her the next (slightly larger) dilator.

SURGERY TO CREATE A VAGINA

A **vaginoplasty** involves using tissue from another area of the body (such as the bowel, or buttocks) to create a vagina, surgically.

- **McIndoe procedure:** A vagina is created with a skin graft, usually from the buttocks (bottom) or with a special skin-like material and a vaginal mold. Young women who have this procedure must stay in bed in the hospital after the procedure for about a week so that the newly created vagina will heal. After the operation, a soft dilator must be worn all the time for about 3 months, taking it out only to use the bathroom. **Even though a vagina is created faster with surgery, it's still necessary to use dilators afterwards.**
- **Williams procedure:** This procedure involves the creation of a vaginal “pouch”. It's sometimes used when other surgical procedures have failed. This procedure is more popular in Europe than in the United States. Dilators are necessary following the procedure, but for only about 3–4 weeks, instead of up to 6 months (which is more typical with the McIndoe procedure). The downside with this procedure is that the angle of the newly created vagina can be awkward and the cosmetic appearance may be poor.
- **Bowel vagina:** This is a major operation which involves making a vagina using a section of the bowel. The recovery involves 4–6 weeks of healing from major surgery, and dilators will need to be worn afterwards. Another disadvantage of this procedure is that your daughter will likely have chronic vaginal discharge and need to wear a pad all the time.

LAPAROSCOPY–VECCHIETTI PROCEDURE

The **Vecchetti** procedure combines the use of dilators and a surgical procedure. This technique (named after the health care provider who developed it) involves applying a traction device to the outside abdomen (stomach) of a young woman.

This is done during a laparoscopy while the patient is under general anesthesia. At the same time, a plastic bead (about the size of an olive) is placed in the vaginal space, and is held with string that is threaded up through the vagina, into the abdominal cavity, and out to the traction device (which is secured to the skin outside of the abdomen). The vagina is formed using continuous pressure, 24 hours a day. The tension is set by turning the “crank” that is on the outside of the abdominal wall. The tension pulls the plastic bead upwards, creating vaginal length. The vagina can be created in about 7–10 days, but requires a long hospital stay to complete the process. Afterwards, a second procedure with anesthesia is necessary to remove the equipment.

It's very important to understand that this procedure also requires the need to use vaginal dilators. Additionally, the traction device is awkward and often uncomfortable. The Vecchietti procedure is popular in Europe, but it's not commonly done in the United States.

Make sure your family is working with a team of experienced medical professionals who are sensitive to you and your daughter's feelings and concerns. When deciding about what treatment is best for her, it's important to remember that dilators are the standard of care and should be tried before surgery is considered. With that being said, your daughter should never be under any pressure. The choice to have treatment or not should always be hers!

ACCEPTING YOUR DAUGHTER'S DIAGNOSIS

It's common to experience a range of emotions when your daughter is diagnosed with MRKH, such as confusion, guilt, sadness and helplessness. Some parents/guardians may feel overwhelmed, or may move into “high gear,” seeking all the information they can access over the internet or from other health care providers. You may feel the need to discuss your daughter's new diagnosis frequently with your family and friends or you may feel the need to withdraw from friends and family for a while. It's also common for parents/guardians to want to “check-in” with their daughters more often than usual about their thoughts or feelings. Typically parents/guardians compare these feelings to riding a “roller coaster” of emotions.

As you learn more about your daughter's diagnosis and treatment options, it's unlikely that you will feel the same intense feelings that you initially experienced. Over time you will begin to accept your daughter's diagnosis and be able to support her emotionally and with any treatment she chooses.

TAKING CARE OF YOURSELF

Taking care of yourself is important for you and your daughter under any circumstances and particularly essential when dealing with a diagnosis related to your daughter's sexual and reproductive health. As you become more comfortable with your own reactions to her diagnosis, you will feel stronger, enabling you to support her. Parents/guardians need time to separate what they feel from what their daughter might be feeling.

- **Find reliable sources of information** about MRKH and the surgical and non-surgical treatment options as well as the option to delay treatment.
- **Keep a notebook or journal** and write down questions as you think of them. Bring it with you to your daughter's medical appointments.
- **Talk about your worries and concerns** with your daughter's medical team, spouse, significant other or a close friend. Many parents have expressed that it's much easier to be supportive to their daughter when they accept ongoing support for themselves.
- **Learn about helpful ways to cope with stress** such as deep breathing, relaxation exercises, and yoga.
- **Join a support group** or find out about networking opportunities with other parents through your local hospital, clinic, or online chats.
- **Talk with a mental health counselor or social worker** if you are having difficulty coping.

HELPING YOUR DAUGHTER

It's natural for you as a parent/guardian to want to help your daughter. Knowing when and how to be helpful may be difficult in the beginning because you may not know exactly how she is coping. Your daughter may be quiet and withdrawn, retreating to her room, making it difficult to know if she wants to talk or be left alone.

She may feel isolated or embarrassed because she is not menstruating and can't take part in conversations about "periods" with her friends. It's important to remember that most teenagers are naturally struggling for independence from their parents/guardians while at the same time seeking their support. The diagnosis of vaginal agenesis, treatment options, sexuality, and future fertility issues, make this struggle more complex than usual!

The following suggestions may help you support your daughter as you and she become more comfortable with new knowledge about her body.

- **Encourage your daughter to ask questions and talk about her worries with you.** There's no right or wrong time to start a conversation with your daughter. Some parents and daughters find it easier to talk spontaneously while others prefer to plan a special time to chat such as during a walk together.
- **Encourage your daughter to ask questions and talk about her concerns with her health care providers.** Suggest that your daughter keep a notebook of any questions and concerns she may have for her health care team: health care provider, nurse, social worker. Understand that she may want to talk with her members of her team alone, without you present.
- **Give your daughter as much privacy as possible.** Privacy is important to the healthy development of all adolescents. Your daughter will need time alone in her bedroom to use the dilators. If she shares a room with a sibling, be sure to arrange time when she can have the room to herself.
- **Reassure your daughter that you will ask her permission before talking about her diagnosis with anyone.** Ask her if you may talk with one person who gives you support; for example, a favorite aunt.
- **Don't worry if you aren't sure how to answer some of your daughter's questions.** It's fine to say "I don't know" and offer to help find the answers. If your daughter is interested in learning more, it may be a good idea to share research tasks. This can provide a safe way to talk about vaginal agenesis by shifting the focus from your daughter to the condition. Your daughter's team of medical experts can help answer questions, such as future fertility options.

TALKING WITH SIBLINGS

It's essential to decide with your daughter what she wants her brothers or sisters to know, if anything, about her diagnosis. You can help guide her about what should be said to whom and by whom. She may not want them to know anything initially and may choose to discuss it after she has become used to her diagnosis. However, if your daughter will be using dilators, she is going to need some private time. Then, it may be helpful to say something to her siblings such as, "Your sister is healthy and she has some special medical issues she needs to take care of. She will need to have some private time for herself every day." The information that you and your daughter share with her siblings will depend on the age, language development, the quality of their relationship and the personality of the child. If siblings are adults, your daughter may choose to talk more openly with them.

COMMONLY ASKED QUESTIONS

How is the reproductive system affected?

The reproductive system is formed during the first few months of fetal life. It includes all the female organs: uterus, vagina, fallopian tubes, and ovaries. Vaginal agenesis (incomplete or absence of the vagina) occurs when the reproductive system doesn't finish developing. When this happens, the vagina can be shorter than usual or absent. The uterus is also typically absent or smaller than usual. The ovaries (the organs that make hormones) and fallopian tubes are normal.

Why is it that we are only finding out about this now?

The average age that MRKH is diagnosed is between 15–18 years old. A young woman typically presents to her pediatrician at this time because she has not yet had her first period. If a pediatrician does an internal exam a diagnosis may be made right away. In other cases, a young woman is referred to a specialist such as an adolescent gynecologist. Typically further testing such as a pelvic ultrasound or MRI (magnetic resonance imaging) is done to confirm whether or not a uterus can be visualized and if there are 1 or 2 kidneys. **Since a pelvic exam is not done at birth or during childhood check-ups, it's uncommon for this diagnosis to be made before the teen years.**

Girls with MRKH generally go through puberty like other young adults, and have secondary sex characteristics such as pubic hair and breast development. Therefore, it's rare for a health care provider to suspect that anything might be wrong before this time.

Are there other tests that are done to confirm this diagnosis?

In addition to an ultrasound or MRI, sometimes a blood test may be done to confirm normal karyotype (female genes) and ovarian function.

Is there any way I could have caused my daughter's diagnosis?

No. MRKH is not heredity or genetic. It's a congenital anomaly or birth defect that occurs during fetal development in about 1 out of 5,000 female births. There is no known cause for this condition and there is nothing an expectant mother can do to prevent this from happening to her unborn daughter.

When should my daughter start to create a vagina?

The best time for your daughter to either use vaginal dilators or have surgery is when she is ready. Timing is different for everyone. The most common age at which girls decide they are ready to create a vagina is around 16 or 17, before college. Sometimes girls wait until they are in college, and start treatment over summer break. Some young women prefer to wait until they have a sexual partner before beginning treatment and some decide not to do anything at all. Please remember that as a parent/guardian you may wish for her to address the issue as soon as possible to help resolve her vaginal agenesis, but this is your daughter's vagina. A vagina should be created when your daughter is ready and wants to create a vagina.

What if the vaginal dilators don't work or my daughter can't use them?

If your daughter is having difficulty or worried that she's using it incorrectly, she should talk with her health care team. If she is not pleased with her progress after she has used dilators consistently for about 6 months, she and her health care provider should discuss the options of surgery or waiting until later.

What if my daughter isn't using her dilator?

There may be one or several reasons why your daughter is not using her dilator. She may feel like she doesn't have the time, is worried that it's painful, or perhaps she is concerned about not having privacy. *It's fine to ask your daughter how things are going, or if she is feeling comfortable with her treatment plan but it's **not** okay to constantly remind her to use her dilator.* It's important for your daughter to keep regular check-ups with her gynecologist so that the medical team can assess any difficulties she may be experiencing and to give her feedback and support regarding her progress. If your daughter continues to avoid using her dilator, it may be that she is simply not ready to create her vagina. *Having a discussion about delaying treatment until **she's ready** may relieve your daughter's anxiety and allow her to have control over the situation.*

Will my daughter be able to experience sexual pleasure?

Most sexual pleasure and orgasm comes from stimulation to the clitoris and intercourse can enhance that pleasure. Typically, women do not experience pain with intercourse after treatment. If your daughter feels discomfort with sexual intercourse it may be that she needs to use a lubricant such as K-Y jelly. Before attempting vaginal intercourse, your daughter should talk with her health care provider.

Will my daughter be able to have children?

If your daughter was born without a uterus or if her uterus is tiny, she will not be able to "carry" a pregnancy. Since her ovaries are normal and make eggs, she may be able to fertilize an egg with the assistance of fertility experts. A surrogate mother could then carry the pregnancy to term. Your daughter and baby's father would be the biological parents of the child. Adoption is another choice for some couples. Keep in mind that fertility options are improving every day. By the time your daughter is ready to have children, she may have more options available to her.

In rare cases, if your daughter was born without a vagina but has a normal size and normally-located uterus, it's likely that she will be able to become pregnant with the assistance of fertility interventions (in-vitro fertilization) and carry a baby.

How can I talk with my daughter when I'm uncomfortable discussing anything related to her sexuality?

Teens can usually sense when their parents are stressed. Be honest. It's fine to tell your daughter that you are feeling a little awkward. At the same time tell her that you want to talk to her and offer her support. Most likely she's uncomfortable too. Although you may both be feeling shy about talking, she will know that you are there to help her cope.

My daughter doesn't want to talk with me about her vaginal agenesis. What should I do?

Your daughter may be uncomfortable about discussing her diagnosis because this often raises issues about her sexuality. The diagnosis of vaginal agenesis may prematurely force a discussion about her sexuality that some daughters and parents find uncomfortable or embarrassing. Attempting to have a conversation when she is unwilling to talk will be frustrating for both of you, therefore it's better to wait until your daughter is ready to have an open conversation with you. You can suggest that she read about MRKH, join a medically moderated chat room discussion with other girls who have this diagnosis, or talk with another trusted friend, sibling or adult about her feelings. You can also let her know that you are available to listen if she would like to talk. There are also online support groups that can offer support and guidance before, during, and after treatment.

Remember, it's ALWAYS important to begin communicating by asking your daughter how best you can help her! Your daughter should be as involved as possible in the planning of her care and ultimately deciding her treatment plan.

How can I be respectful of my daughter's right to make decisions about her body?

If your daughter is under 18 years old, your consent for medical and surgical treatment is necessary. It's critical that all options be discussed in clear language with your daughter so that she can participate in these important decisions about her body. Ultimately her medical team will respect her wishes regarding treatment.

Should I tell anyone about my daughter's diagnosis?

Telling someone about your daughter's diagnosis is a personal decision. It's always best to ask your daughter for her permission first. Most parents of girls with vaginal agenesis tell us that they are selective with whom they share their daughter's personal information with but at the same time they find comfort and support by talking with at least one close relative or friend.

How do I respond to people's questions about my daughter's diagnosis, the treatment and future implications?

It's up to you to decide if you want to share information and with whom. However, the more you are comfortable understanding your daughter's diagnosis, the easier it will be to respond to specific questions. Being able to share information factually will make these discussions easier. Additionally, listen carefully to what the other person is asking. What is it that they really want to know? Is it medical information or reassurance that your daughter will be physically and emotionally healthy? Answer the questions as well as you can and do not hesitate to say "I don't know." It's fine for parents to set limits when people ask questions they feel are inappropriate. Remember, your daughter's privacy must be respected.

How can I tell if my daughter needs professional help?

It's not uncommon for young women with a new diagnosis of MRKH to have temporary changes in their emotions and behaviors as they adjust to a new understanding of how their body has developed. These concerns should decrease over time as your daughter understands her diagnosis, is able to verbalize her concerns, and becomes involved in her medical care. Typical signs that your daughter is upset may include changes in her sleep or eating patterns, worried comments about her body, concerns about treatments and medical appointments, and changes in behavior, such as becoming quiet and withdrawn, or aggressive and angry. If the changes in her emotions and behaviors lasts longer than a few weeks or seem very concerning to you, your daughter may be having a more difficult time adjusting to her

diagnosis. If you, your daughter, or her medical team has concerns, don't wait too long. You should talk with her gynecologist about having her meet with a counselor, therapist, or social worker. This can usually be arranged by obtaining a referral from her primary health care provider. It's essential that the counselor, therapist, or social worker have experience with adolescents who have medical issues. It's important for the counselor (that she is referred to) have accurate information about MRKH. You can direct them to our MRKH guide online at: youngwomenshealth.org/mrkh_teen.html, or share written information with them prior to your daughter's first meeting.