

MRKH: A GUIDE FOR TEENS

You may have just learned that you have **MRKH (Mayer Rokitansky Kuster Hauser Syndrome)**. You're probably thinking, "Why is the name so long?" It's extra long because this condition is named after all of the health care providers who discovered it. Aside from being overwhelmed with the name of this condition, it is also normal to feel confused, scared, and sad about having MRKH. Most likely you and your parents have a lot of questions. We hope that this guide will help answer your concerns. We also have a special guide for your parents.

What is MRKH?

MRKH is a congenital disorder (birth defect) that affects the female reproductive tract. Congenital means that it is there at birth. About 1 in every 5,000–10,000 female babies has this condition. *MRKH is a syndrome (group of symptoms)*. We don't know the cause of this syndrome but we do know that when a baby grows in their mother's uterus (womb), organs and systems develop. One of the systems is called the reproductive system, which includes the uterus, cervix, vagina, fallopian tubes, and ovaries. The reproductive system is formed during the first few months of "fetal" life (while a baby is still in her mother's womb). With MRKH, the reproductive system starts to grow but doesn't completely develop.

Girls with MRKH have normal ovaries and fallopian tubes. Most often the uterus is absent or tiny. The vagina is typically shorter and narrower than usual or it may be absent. Sometimes, there may be one kidney instead of two. About 3% of girls will have a minor hearing loss and some may have spinal problems such as scoliosis (curvature of the spine).

DIAGNOSIS

When is MRKH diagnosed?

The most common age for MRKH to be diagnosed is when a young woman is between 15 and 18 years old. That's when a young woman is likely to see her health care provider because she hasn't started her

period. Some girls may find out at an earlier age or when they're older.

What will happen at my health care provider's appointment?

Your health care provider will probably ask you questions such as: "When did you notice that your body was changing...going through puberty?" Next, he/she may want to take a look at your outer female organs and also check to see how long your vagina is. Your health care provider will gently put a Q-tip or gloved pinky finger at the opening of your vagina and then very slowly and carefully put it into the vagina to see how deep your vagina is. If your health care provider thinks you might have MRKH, he/she will probably order a test called an ultrasound or an MRI (magnetic resonance imaging). These tests do not hurt and are similar to having an x-ray. Usually your health care provider will refer you to a specialist who has experience taking care of young women with MRKH. A **pediatric and adolescent gynecologist** is a health care provider with special training in young women's reproductive health.

What can a pelvic ultrasound or MRI show?

A pelvic ultrasound is usually the first test to check to see if a uterus or womb is present. This test can also confirm that you have two ovaries and two kidneys. Sometimes a very tiny uterus can be seen. A tiny uterus is called a "uterine horn or remnant". You may need to have an MRI so that your health care provider can see your female organs in more detail.

TREATMENT OPTIONS

If you've been told that you have MRKH and your vagina is incomplete or absent, you have the following options:

1. **You can do nothing.**
2. **You can create a vagina *without* surgery, using dilators** that expand and stretch your existing vagina over time, using intermittent pressure. *This is a nonsurgical procedure.*



3. **You can have a surgical procedure, or “vaginoplasty”, which involves surgery** (using a skin graft from another part of your body).
4. **You can have a procedure that involves a combination of an operation and dilation with continuous pressure: The Vecchetti procedure.**

WAITING OR DOING NOTHING

YOU have control of your body. *Deciding to make or not make a vagina should be **your decision** and if you decide you want to, when should be your decision too!* If you are not planning to have vaginal intercourse, it is not something you need to do. If you plan to have vaginal intercourse now or in the future, making a vagina is something you might want to consider.

Aside from doing nothing, there are three basic approaches to help you expand or create a vagina. If you're thinking about treatment, you should definitely consider using dilators first. **All of the surgical options require general anesthesia and the use of dilators after the operation, 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 is 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 you are a patient at Children's Hospital Boston and select this method of treatment, Dr. Laufer's nurse specialist will teach you how to use the dilators. You will take one home with you and use it twice a day for about 20 minutes. You will likely be asked to return for follow-up appointments once a month. As your vagina stretches, Dr. Laufer will give you the next size dilator, which will be slightly wider.

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 you work with a gynecology program that has at least an 85–95% success rate for helping their patients create a functional vagina. You 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, you 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 do I need to see my health care provider?

It is VERY important to keep appointments with your health care provider so he/she can check your progress (whether you have decided to use dilators or have surgery). If you decide to use dilators, your health care provider will need to check that you are applying pressure in the right place and at the correct angle. As your vagina stretches, your health care provider will give you the next (slightly bigger) dilator.

SURGERY TO CREATE A VAGINA

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

- **McIndoe procedure:** A vagina is created with a skin graft, usually from your 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 is still necessary to use a dilator afterwards.**
- **Williams procedure:** This procedure involves the creation of a vaginal “pouch”. It is 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 you 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 is 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 is not commonly done in the United States.

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

FREQUENTLY ASKED QUESTIONS

Can anyone tell that I have MRKH?

Some young women wonder if anyone can tell if they have MRKH. *The answer is no.* No one, except you and your health care provider can tell that you were born with an incomplete vagina and following treatment, with dilators or surgery, your sexual partner will not be able to feel any difference.

Will I be able to have children?

If you were born *with* an incomplete vagina but have a normal size uterus, it is likely that you will be able to become pregnant and carry a baby.

If you were born *without* a uterus or if your uterus is tiny, you will not be able to carry a pregnancy. Since your ovaries are normal and make eggs, an egg can be fertilized with your partners' sperm.

Someone else such as; your sister, friend, or another person you choose, could be the surrogate mother. Surrogate mothers are women who agree to carry a pregnancy for a couple. You and the baby's father would be the biological parents of this child. Adoption is another choice for some couples. Fertility options are improving every day. *By the time you are ready to have children, there may be more options available to you.*

Why might I have pelvic (belly) pain each month?

Some women with MRKH may have a tiny uterus called a "uteri or uterine remnant". The uterus or womb is not big enough to carry a baby but it can cause pelvic pain if blood from this small uterus goes into the belly. Your health care provider will be able to tell if you have a small uterus and if it needs to be taken out. If you have belly pain, it is important to tell your health care provider.

If I don't make a vagina but I do decide to have sex, will anything bad happen?

If you have vaginal intercourse before your vagina is created using dilators or surgery, sex will likely be very painful. It could cause a tear in your vagina and bleeding. Making a vagina with sex can be done but comes with risks and is usually very uncomfortable.

Will I ever be able to have a "normal" sex life?

YES. Keep in mind that every woman, regardless of her age or health issues experiences sexual stimulation and sensitivity in different ways. *Discovering what you enjoy sexually is an evolving process throughout your life.* Thus, women with MRKH who are sexually active are encouraged to explore their sexuality with themselves and with their partner(s) to learn what feels both comfortable and pleasurable.

I'm in boarding school/college and have a roommate – How do I get some privacy to use the dilators?

Most young women find that they need privacy when using the dilators. When you are sharing a room, either with a sibling or roommate, it can be

uncomfortable asking for time alone, especially if you don't want to share any details of your diagnosis or treatment.

There are many reasons why people need to have some time alone: meditating, studying, napping, praying, etc. It is always best to plan ahead, so check with your roommate to find out when she will be in class or out of your dorm room so you will know when you will have private time to use your dilator.

I'm really embarrassed with all the medical visits – will this ever get easier?

Many young women diagnosed with MRKH feel pushed into a world of new information and new experiences. It is perfectly normal to have a range of emotions: sadness and anger, hope and worry, fear and embarrassment can all be part of the experience. Your medical team: the health care provider, nurses and social worker, are aware of this and they are trained to be sensitive to your unique situation. You may be the kind of person who wants a lot of interaction and information at all the visits or you may be someone who just wants the facts and prefers to keep the visits as brief as possible. One thing that you can do to make this process easier is to tell your medical team what approach would be most comfortable for you during the appointments and exams. The team will do their best to make sure your appointments are as stress-free as possible.

My parents want to keep talking to me about MRKH but I already feel like my privacy has been taken away – how do I keep some boundaries?

An essential part of growing up is becoming more independent as well as setting some boundaries between you and your parents. When there is a medical issue that requires many appointments and exams, it can be hard to feel a sense of privacy. Additionally, since MRKH by definition involves both you and your parents thinking of you as a sexual person, the stress level in families may be very high at first. For many young women and their parents, the conversations you have been having about your body may just feel too private. Just as you have had to get used to this diagnosis and what it all means, so do your parents.

Sometimes this may feel like an invasion of your privacy. Your parents may find it helpful to read the Parent's Guide to MRKH which answers the most frequently asked questions parents have.

You can also talk with members of your medical team for help with communicating with your parents.

Is there anyone else I can talk to about having MRKH?

Some young women find it helpful to talk with a parent(s), while other girls prefer to talk with a counselor or a close friend. *We know that it can be very helpful to talk with someone your own age that has MRKH.* The Center for Young Women's Health at Children's Hospital Boston offers free monthly chats for young women with MRKH.

Am I still a biological woman?

It is not uncommon for young woman to wonder if they are "really" a girl when they first learn they are born with an incomplete vagina and uterus. If you have asked yourself this question, you are not alone. *But it is very important for you to understand that **you are a female**.* Your health care provider may order a special blood test that can confirm that you are a genetic female and have 46XX chromosomes.

Will my vagina ever close up?

Once you have created a vagina your vagina shouldn't change or shrink if you are having vaginal intercourse or using the dilator about once a week. If you are not sexually active, you should use the largest size dilator once a week for about 15–20 minutes (only after you are finished making your vagina).

I don't want to use the dilators now – can I wait until I feel ready? What happens if I never create a vagina?

The choice to have treatment – how and when – IS UP TO YOU! Like any important decision, it is essential that you get all the information before you make the decision to have treatment. Talking with other women who have MRKH is valuable too. *You control your body. You should never be forced or pressured into using dilators by your parents, partner or medical team.*

Rather, they should support you during the treatment process only when you decide the time is right. *Your medical team* has the responsibility of giving you information and resources to help you understand your reproductive health issues so YOU can make informed decisions. Your parents have the responsibility of helping you get medical care and helping you obtain privacy at home if/when you begin using dilators. You have the responsibility to learn more about MRKH and to talk with a trusted adult if you are feeling overwhelmed.

If you have a question about MRKH that you'd like answered, e-mail us at: cywh@childrens.harvard.edu. Our team will review your question. All appropriate questions will be answered and posted in the FAQ section.

If you have been diagnosed with MRKH or you would like an evaluation or second opinion, you may contact the Gynecology Program at Children's Hospital Boston at: (617) 355-7648.