



## 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 doctors 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 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 do not know the cause of this syndrome but we do know that when a baby grows in the mother's uterus (womb), systems develop. One of the systems is called the reproductive system, which includes the uterus, 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 develop but doesn't completely finish.

**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).

### When is a girl likely to find out she has MRKH?

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 are older.

### What will happen at my doctor's appointment?

Your doctor 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 doctor 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 doctor 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 doctor will have you see a specialist who has experience taking care of young women with MRKH. A **pediatric and adolescent gynecologist** is a doctor 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 doctor can see your female organs in more detail.

### If I have an incomplete vagina, what are my options?

If you have been told that you have MRKH and your vagina is incomplete or absent, you have some options.

- You can create a vagina *without* surgery using dilators
- You can have a surgical procedure
- You can do nothing

## Creating a vagina with dilators

Most of the time girls with MRKH choose to make a vagina by using vaginal dilators. This treatment is very effective and “noninvasive” (*does not have the risks of an operation*). There are different kinds of dilators available and they come in different sizes. The most commonly used dilators look like a thin plastic tube similar to a slender tampon. Starting with the smallest dilator you will learn how to hold the dilator and apply pressure to stretch your vagina. In the beginning, most of the dilator will be on the “outside” and used as a “handle” (*as it can only go in so far*). You will use the dilator for about 15–20 minutes, twice a day. Over time, you will begin to notice that the dilator is able to go into your vagina a bit further. This means you have made progress! As your vagina stretches, your doctor will know when to give you the next size. The next size will be slightly wider.

## Surgery to create a vagina

**MclIndoe procedure** – *This is the most common surgical procedure to make a vagina.* 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 for about a week so that the newly created vagina will heal. A soft dilator must be worn all the time for a while, taking it out only to pass urine or poop. Even though a vagina is created faster with surgery, it is still necessary to use a dilator. Surgery to create a vagina should not be thought of as a “quick fix solution”.

**Bowel vagina** – This is a major operation which involves making a vagina using a section of the bowel. The advantage of this procedure over the MclIndoe operation is that you don't have to stay on bed rest for a week. The disadvantage is that girls who opt for this procedure will likely have chronic vaginal discharge.

### Other surgical procedures to create a vagina include:

- the Frank technique
- Willams vulvovaginoplasty
- the Vecchiotti procedure
- waiting or doing nothing

You are the one who should be in 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 do.

## How often do I need to see my doctor?

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

## How long will it take to create a vagina with dilators?

The average amount of time it takes to create a vagina (*if the dilator is used at least 15–20 minutes, twice a day*) is about 3–14 months. It can take less time for some young women or more time for others. If the dilators are not used every day, it can take a year or more.

A recent study done at Children's Hospital Boston found that 88% of girls who chose to use dilators consistently to make a vagina, were able to do so within a year and a half.

## Do the dilators hurt?

Using dilators to create a vagina shouldn't hurt. Remember, you have control over the amount of pressure that you apply. You should get used to applying enough pressure so the skin will stretch but you should not be in pain.

## When should I start to create a vagina?

**There is no *right* or *wrong* time to create a vagina. The decision to start or not to start should be yours.**

You may be ready to start using dilators when you are in high school or you may decide to wait until you are older. Some questions to ask yourself: “Do I have time in my day to use the dilator?”; “Do I have privacy?”; “Am I comfortable touching myself?”, “When do I plan to become sexually active?”

## What if the dilators don't work or I just can't use them?

Most girls can learn to use a dilator and have success in creating a vagina. Sometimes, it can take a while until you feel comfortable using it. If you have any questions or wonder if you are using the dilator correctly, be sure to ask your doctor or nurse specialist. In some cases, if your doctor feels that little progress has been made, he or she

may suggest that you take a break for a while and try again later. **Dilator treatment is the standard, most efficient and nonsurgical treatment for MRKH;** it is recommended by the American College of Obstetricians and Gynecologists (ACOG) as the first choice of treatment to create a vagina for girls with MRKH. Surgery should only be considered if dilators have been tried (*under the supervision of a gynecologist who specializes in treating young women with MRKH*).

### **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 doctor 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 doctor.

### **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 doctor, 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 deliver 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.*

### **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.

*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.*

## **Frequently Asked Questions**

### **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?**

The quick answer is YES. However, this question requires a more complicated response. The answer to this question is closely tied to what your idea of “normal” means. Every woman, regardless of her 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 partners 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 do not want to share any details of your treatment or diagnosis. *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 doctor, 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 would be most comfortable during the appointment 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.

## **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 doctor 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 would like answered, e-mail us. Our team will review your question. All appropriate questions will be answered and posted here in the FAQ section.*