

Transitioning to Adult Gynecology Care

A Guide For Young Women With MRKH



**Boston
Children's
Hospital**

Until every child is well™

Center for Young Women's Health
Division of Gynecology

Transitioning to Adult Gynecology Care

IN APPRECIATION

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DEDICATION

This booklet is dedicated to all the amazing adolescents and adult women who share a special sisterhood.

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Transitioning to Adult Gynecology Care

Congratulations! You're getting ready to transition to routine adult gynecology (GYN) care. It's normal to feel a little nervous about this change at first, especially if you have been followed by your adolescent gynecologist for a while. As a woman, you'll need to work with a gynecologist who is familiar with MRKH or willing to learn about it and who can address your questions about sexual health.

Health systems are different in centers across the United States. Some adolescent GYN providers see patients until 18, 21, or 25 and then recommend a gynecologist for follow-up in another academic adult hospital or community practice. Other gynecologists provide care across a women's lifespan, allowing you to receive GYN care in the same practice. There is a difference though. When you were younger, your parent(s) or guardian most likely accompanied you during your medical or GYN appointments. As an adult, you will make your own appointments and see your provider by yourself. The focus will be on you, not you and your parent(s)/guardian(s).

As you move from adolescence towards adulthood you will likely:

- Take on new challenges and do more things on your own
- Have more choices and options
- Gain a new sense of freedom and independence
- Take on more responsibility for your health care

If your parent(s) or guardian has been making your appointments and communicating with your gynecologist, it might feel awkward at first to advocate for yourself, make appointments and communicate with your medical team directly. The more responsibility you take for these things, the easier it gets, and who else knows you best? An important aspect of becoming an adult is establishing a support system that includes family, friends, and health care providers.

People who can help you transition your care to an adult gynecologist:

- Your pediatrician and your adolescent gynecologist
- Another member of your GYN team (nurse and/or social worker)
- Your parent(s) or guardian(s) – Ask them to help you find someone who is listed on your health insurance plan.
- An older sibling or relative
- A friend with MRKH who has been through the same process

It is not uncommon for a gynecologist to lack prior experience with MRKH. It can be an empowering opportunity to educate and advocate for yourself as an adult woman. For example, you could call the doctor's office and ask the scheduling person if the gynecologist has experience seeing women who have been diagnosed with MRKH. Another way to ask is, *"Does Dr. _ (or anyone in your practice) have experience treating women with "congenital anomalies of the reproductive tract?"* You should also ask if the gynecologist can provide fertility care, meaning they can help you if and when you want to learn more about options to start a family. If you cannot find a gynecologist who knows about MRKH, or prefer to have your primary care doctor or nurse practitioner provide your care, you can ask if she/he is willing to learn about MRKH. You could then give them written information about MRKH and have your current gynecologist write a summary about your course of treatment thus far.

As a young adult moving toward adult care (GYN and general health), you should be able to:

1. **Learn about your health insurance coverage** and the amount of your co-pay and/or deductibles.
2. **Contact the Medical Records Department** where you currently have your GYN care and sign a release to have your records sent to your new gynecologist (and adult health care providers, if applicable) or pick up a copy and bring the documents with you to your first appointment.
3. **Make a list of all the medications** you take and write down any allergies you have (if applicable).
4. **Create a brief timeline of your GYN/medical history:** it's very useful to have the dates of diagnosis, treatment, names of your providers, etc. at your fingertips without having to search for

this information.

5. **Know the names and phone numbers of all your health care providers**, then store the contact information in your phone or someplace else where the information is readily available.
6. **Add your GYN appointments** (and other medical appointments) in your calendar.

If you have to change or cancel a GYN (or medical) appointment make sure you call the office well in advance. Some medical offices will charge you for the visit unless you contact them 24–48 hrs. before your scheduled appointment.

Take this short quiz and find out if you are close to managing your own GYN and other medical care.

- I can explain MRKH (and any other medical conditions I have) to my GYN and medical providers. (If not, it's very helpful to provide your GYN and other medical providers with a copy of the MRKH handout, pages 17–23)
- I know the names and phone numbers of all of my health care providers.
- I ask questions during my GYN and other medical appointments.
- I know what medications I take and I have a list that I bring to my GYN and medical appointments. I tell my providers when I no longer take certain medicine(s) and when I start a new medicine(s).
- I feel comfortable responding to questions my gynecologist (and other medical providers) asks.
- I am able to schedule my GYN and other medical appointments by myself and I have a way of keeping track of them so I don't miss appointments.
- I know about my health insurance coverage and how much my co-pay is for different medical services.
- I know where and how to obtain a copy of my medical records.

I am able to get to my GYN and other medical appointments by myself.

I know where to get my prescriptions filled, and have called the pharmacy when I've had questions or needed refills.

If you checked all or most of the statements:

You're already taking on adult responsibilities and you are prepared to transition your GYN/medical care to adult providers. Talk to your current gynecologist about a transition plan.

If you checked more than half of the statements:

You're on your way! You are actively taking on many new responsibilities regarding your GYN and medical care. Pick a few more responsibilities to do before your next appointment. Begin talking about transitioning with your GYN provider.

If you checked less than half of the statements:

Now is a good time to start taking on more responsibilities from the checklist and practice them at your next appointment. If you need help, ask a parent, friend, social worker, nurse, or doctor.

Commonly Asked Questions

If I don't have a cervix, do I need a yearly Pap test?

It's true that a Pap test is done to detect cervical cancer but the test can also detect vulvar and vaginal cancers. Most gynecologists therefore recommend that you have a yearly Pap test of your vagina starting at age 21.

Do I need a yearly GYN exam?

Yes. You should have a yearly check-up including a pelvic exam or a pelvic ultrasound once a year so that your primary care provider or your adult gynecologist can check your ovaries, vagina, and vulva.

Do I need to get the HPV vaccine?

Yes. At least 1 in every 2 sexually active young women has had a genital HPV infection. The HPV vaccine protects all women (who are immunized) from certain types of the human papilloma virus. There are three different vaccines. Gardasil® protects against the four types of HPV that cause genital warts (types 6 and 11) and cervical, vaginal, vulvar, and anal cancers (types 16 and 18); Gardasil9® protects against types 6, 11, 16 and 18, plus 31, 33, 45, and 52; Cervarix® protects against two types; 16 and 18. If you're sexually active, have had any sexual contact, or are thinking about having sexual contact, your *best* protection is to avoid sexual contact until you are vaccinated. Remember that you are not protected until you have completed the series. Even though you don't have a cervix, (if or when you become sexually active) you are still at risk for genital warts and vulvar or vaginal cancer just like any other woman.

Why might I have pelvic (belly) pain around the same time each month?

In general, some women can tell when they ovulate (make an egg) each month because they feel some discomfort or twinge in their lower belly. Most of the time this mild pain is nothing to worry about as it is caused from normal ovulation.

Some women with MRKH may have a tiny uterus called a "uterine remnant" or "uterine horn". This type of uterus or womb is not big enough to carry a baby but sometimes it can cause pelvic pain if blood leaks into the pelvic cavity. If this is the case, you will need to be on continuous hormone pills (birth control pills) to stop any bleeding until the tiny uterus is removed by a GYN specialist.

Is it harmful if I wait to create a vagina until I'm older?

The choice to have treatment, how, when, etc., is UP TO YOU! Like any other important decision, it's essential that you get all the information before you make the decision to have treatment. Remember, you can create a vagina AT ANY TIME or not at all and still feel complete!

When should I tell my partner about MRKH?

There is no set rule which fits or works for all women, in all circumstances. Sharing anything intimate with a sexual partner should be based on trust, caring and the possibility of a future together but not all sexual encounters have these three components. It is up to you to use your good judgment. Since having MRKH comes with complications about fertility, some women who wish to be parents at some point may choose to have a conversation with their partner sooner rather than later so they can talk about their options. It can be extremely helpful to talk with other women who have MRKH to hear about how they have had similar conversations with their partners.

Will I be able to have a normal sex life?

YES. Most sexual pleasure and orgasm comes from stimulation to the clitoris and intercourse or vaginal penetration can enhance that sensation. Keep in mind that every woman, regardless of her age, health issues or intimate partner preferences, experiences sexual stimulation and sensitivity in different ways. Discovering what you enjoy sexually is an evolving process through your lifetime. Thus, all women are encouraged to explore their sexuality with themselves and with their partner(s) to learn what feels both comfortable and pleasurable. Having MRKH doesn't impact your sexual enjoyment or your ability to have an orgasm.

Will my created vagina feel differently to my partner?

No. After you create a vagina with dilators or surgery, no one will know that you were born with an incomplete vagina. Your partner will not be able to feel any difference.

Will I have pain with intercourse?

If you have completed the dilation process, intercourse should not be painful. Many women do feel some mild discomfort the first time they have intercourse, regardless of whether they have MRKH or not. If you feel any discomfort it may be that you need to use a lubricant such as K-Y jelly before vaginal intercourse.

Can I still get sexually transmitted infections?

YES! Please remember that you should always use barrier protection such as female or male condoms **every time** you have sex to prevent HIV and lower your risk for other sexually transmitted infections (STIs).

Do I need to have intercourse to keep my vagina open?

No. You should not feel pressured to have intercourse until you are ready. You can prevent your vagina from getting smaller by using the largest size dilator or a vibrator just once a week for 15–20 minutes. When you are having intercourse or other kinds of vaginal penetration on a regular basis (1 or more times a week), you don't have to use the dilator anymore.

What might keep me from having an orgasm?

Orgasm is the result of sexual stimulation that is a combination of emotional, psychological and physical pleasure. There are many reasons why women may not experience orgasm at any given time, but none that are directly related to having MRKH. Experiencing orgasm is a very individual process and involves getting to know your body and its responses every time you are stimulated. There are many distractions that prevent women from having an orgasm (climaxing or coming), such as being tired, having negative feelings about your body, ambivalence about your partner, or just not being "into it", etc. Many women find that the process of stimulation is as exciting as the orgasm itself. Your sexuality and responses will evolve throughout your life time.

There are different things that can help:

- Talk about any concerns you might have with your partner: communication is the most powerful form of intimacy!
- Use a lubricant such as K–Y jelly.
- Learn how to tense and relax your pelvic muscles, also called Kegel exercises. These muscles contract during orgasm.
- Learn what kind of stimulation arouses you and how to enjoy your body and share this information with your partner.
- Talk to your GYN team if you have any questions or concerns.

How can I tell if I need to talk with a counselor?

Becoming overwhelmed or distressed by a situation is very common for everyone at some point or another. Sometimes symptoms of depression or anxiety feel unmanageable and in that case counseling may be helpful for addressing these symptoms. There are different kinds of therapists (clinical social workers, psychologists, mental health counselors, etc.) as well as therapies (cognitive behavioral, psychodynamic, mindfulness based, etc.) but the goal is the same: to help you manage your life and challenges with more ease. If you find yourself beginning to be overwhelmed or concerned about something, the sooner you seek help the better. You can ask your primary care doctor or nurse, or your gynecologist for a referral, or seek help through your school counseling center (if you are in college) or a friend, family or clergy. If you ever have thoughts of self-harm, contact a trusted friend or family member immediately and go to the closest emergency room to ensure your safety.

Talking to Your Partner About MRKH

Most young women with MRKH will at some point find themselves faced with the decision of whether, when, and how to tell a romantic partner about their MRKH. The decision to tell your partner about your diagnosis is entirely your own, and there is no set rule which fits for all women, in all circumstances. Sharing anything intimate with a partner should be based on trust, communication, caring, and the possibility of a future together, but not all romantic and sexual encounters will have all of these components. It is up to you to use your good judgment to determine whether this is the right thing to do, and when.

To tell or not to tell?

Whether or not you tell your partner will depend on a number of factors. First and foremost, it is important that your partner can be trusted with your personal information and has demonstrated that he or she is generally supportive of you on an emotional level. Sharing this information may cause you to feel vulnerable and you'll want to know that your partner will be respectful of your feelings. If you are in a relationship with someone with whom you may consider raising a child with in the future, you might choose to have a conversation sooner rather than later about your MRKH so that you can talk about your fertility options. Finally, if you and your partner are considering a sexual relationship, and you have not chosen to create a vagina yet (either with dilation or surgery), it will be important for your partner to be aware of your anatomy so that he or she does not accidentally cause you pain or discomfort. If you have created a vagina, remember that your partner will not feel anything different, so you will have the choice whether or not to discuss your diagnosis.

When is the right time?

While there is no right or wrong time to have this conversation, you may want to consider the level of trust and intimacy that you and your partner have developed before deciding to share your diagnosis. Some women may choose to share this information early on in a relationship

because it feels too difficult to keep it to themselves, while other women may choose to wait months or even years before they feel ready to share this very personal information. Only you can decide when you feel comfortable telling your partner about your MRKH, and you are the best judge of when your partner is ready to hear it. As much as your MRKH impacts you, it will also be important to recognize that it will impact your partner too, and they may need some time to adjust to what you have told them. They also may have a lot of questions about what you have shared, so consider telling them at a time when you feel you are ready to answer any questions that may come up.

How do I do it?

Again, the best way to share this information with your partner depends on you and your level of comfort. Some women may choose to share everything about their MRKH right up front, while others may choose to gradually disclose more information over time. For example, if you haven't created a vagina via dilation or surgery, you may wish to tell your partner that you were born with an incomplete vagina early on but then wait before you discuss the impact of your diagnosis on your future fertility. Alternatively, some women may feel more comfortable sharing that they have an absent uterus but wish to wait before telling their partner that they were born with an incomplete vagina. Keep in mind that if you have not had treatment, a sexual partner may need to know this information to avoid unintentionally causing you pain. If you think it would be helpful, you might consider printing information about MRKH (from our website www.youngwomenshealth.org) for your partner to read.

Finally, remember that you are not alone in this. It can be extremely helpful to talk with other women who have MRKH to hear about when and how they have had similar conversations with their partners. Your health care team is also available to answer any questions you might have or help you talk through your decision. In the end, the most important aspect of this decision is that you feel comfortable and confident having this conversation with your partner. Keep in mind that sexuality is only one part of a well-rounded and healthy relationship, and sharing this information with your partner can lead to a richer, deeper, and more intimate connection where you both can be open to talking about your feelings, desires, and needs. Your partner will likely feel honored that you trusted them enough to share this information, and you will probably breathe a sigh of relief at no longer feeling like this is something you have to keep secret.

What if it doesn't go well?

While these guidelines can help make the process of sharing your diagnosis with a partner go more smoothly, it can be difficult to predict how this conversation will unfold. There may be some situations where, despite your best efforts, the discussion does not go as planned.

Remember that your partner's initial reaction is coming from a place of surprise and maybe even shock, and the feelings and thoughts they express right away may not be the same as those they experience after they've had some time to process the information. After all, you have had time to prepare exactly what you want to say to your partner but they have not had any time to prepare their response, so it's important to give them some time and space to take in what you've shared with them and organize their thoughts about it. It takes a certain level of maturity for an adult to understand and process medical information. Unfortunately you may come in contact with some people who are judgmental or have not had experience dealing with medical issues and therefore their reaction may not be supportive or fair. If you find that your partner makes you feel bad or is not able to be supportive of you and your MRKH, know that you are worthy of far more than what that partner has to offer. You deserve to be with a partner who loves you and accepts you for who you are in every respect and you WILL find that person. In the meantime, you have access to a community of women who share your diagnosis and who will be there for you as you process feelings about MRKH at different points in your life.

Moving Ahead....

There will be times particularly at social events such as baby showers, family reunions, or even casual times with friends in which conversation will naturally come up about fertility issues, children or plans for children in the future. Unfortunately, not everyone is sensitive to the fact that many women, for a variety of reasons, are unable to carry or conceive a child. Try to anticipate the questions you might be asked and practice your responses. Of course you cannot plan for every situation nor can you plan for exactly how you will feel “in the moment”. Remember, you are entitled to how you feel and how you react. After all, it’s your body, you do not owe anyone an explanation, and what you choose to say should be your decision. Just remember, if someone doesn’t know your story they are probably unaware of how their statements may affect you.

BUTTON–PUSHING EXPERIENCES

Most teens and woman with MRKH have had “button–pushing experiences”– times when something will set off their emotions. For example; it is completely normal to feel unsettled or sad when hearing about a friend or relative who is pregnant, or feel vulnerable, unhappy, or even angry when someone innocently asks about your plans to have children. As a teen you may have felt anxious when a classmate asked if you had a tampon or pad to lend, or maybe when your younger sister began her period, you felt upset. Hopefully you were able to learn ways to manage your responses and feelings during those times with the help of your support system.

The reality of having MRKH is accepting the fact that situations will come up in your young adult life and beyond, that will be both “button–pushing” and stressful. It can be very helpful to anticipate which circumstances might be difficult so that you can have a prepared response. It’s also important to validate your feelings and give yourself time to experience the emotions when they come up. Girls and women with MRKH have told us that some of the experiences they find particularly awkward and sometimes stressful include:

- When other women talk about becoming pregnant and when people ask questions such as “*When are you going to start a family?*”

- Medical situations in which you're asked: *'When was your last period?'* or *'How old were you when you got your first period?'*
- Having to explain MRKH to medical providers.
- "Well-meaning" yet insensitive comments from people about the advantages of not having periods: *"At least you don't have to deal with cramps!"*
- Well-meaning but inappropriate comments from people who try to make you feel better by saying something such as: *"At least you'll be able to keep your figure."*
- Receiving baby announcements, invitations to baby showers, christenings, naming, or bris ceremonies.

Even if you choose to have children with a gestational carrier or adoption, some people may "push your buttons" by asking intrusive questions about your childbirth experience. It is therefore very helpful to try to anticipate both the questions and your responses when you know you might be in a situation that could be potentially awkward and/or emotionally stressful for you.

Planning Ahead to Become a Parent, or Not

Planning to start a family is a big decision regardless of whether or not a woman has MRKH. Becoming a parent is right for some people and not for others. Only you can know what the best decision is for you and your family.

As a woman with MRKH it is likely that your uterus is underdeveloped so you will not be able to carry a baby. Since you do have normal ovaries, you are able to have a biological child. This involves taking an egg from your ovary and sperm from a male partner or a sperm donor, and fertilizing them in a lab. When the egg fertilizes it becomes an embryo. The embryo is then placed in the uterus of another woman, who is called a gestational carrier. The woman doesn't have any genetic connection to the baby; she simply is the carrier until birth. There are a lot of steps in between that involve much coordination by a fertility team. Insurance coverage for this procedure can unfortunately be somewhat complicated. Different insurance companies cover different costs and coverage can vary drastically between states and countries. The procedure itself, as well as paying a gestational carrier, can get very expensive. You'll need to do your research to find out if surrogacy is legal in your state and also explore what is financially possible for your family. The websites in the "Resource" section of this booklet will allow you to compare the surrogacy and adoption laws by state. You should also check with your insurance company to find out what they will cover if you decide to have a child via surrogacy. Be aware that different states mandate insurance coverage for fertility treatments such as IVF.

Your options for creating a family include:

1. Adoption: Domestic, International, Private, and via public child and family programs
2. IVF (In-Vitro Fertilization): with a Gestation Carrier

Your Feelings About MRKH

It is likely that your feelings about MRKH will change as you become older and more mature. At first, many young women report that they can think of nothing else much of the time, particularly right after learning about their diagnosis. Other young women have shared that they pushed away all thoughts and feelings about MRKH until much later. This is natural. New information about your body and future has to be assimilated, experienced and understood on many levels.

Often, during the process of dilation, or when it is completed and weekly "maintenance" is practiced, it may feel like a burden or chore. Yet as women move into their 20's and beyond, and their lives fill with college, a career, new relationships, travel and hobbies, women often report a change in how often they think and feel about MRKH or if they think about it at all. With time, most women learn to view MRKH as a smaller part of their life, much like any chronic medical condition for which there is treatment, and life goes on with certain adjustments. This is sometimes referred to as establishing a "new normal." Sometimes this happens naturally and sometimes it can be helpful to talk to a counselor or therapist to help you to get to this point. Regardless, having MRKH should not define who you are nor who you strive to be as an adult, parent, or professional. There is no need to limit your personal expectations or goals.

Resources

Every effort has been made to ensure that the URL's listed in this section are as accurate and up-to-date as possible. We realize that the internet is constantly changing, and we can only guarantee that the links are accurate as of the date this booklet was printed.

State by State Surrogacy Law Practice

<http://www.creativefamilyconnections.com/us-surrogacy-law-map>

This is a private website founded by a lawyer who has expertise with matters involving surrogacy. It is not a website that the CYWH necessarily endorses; however there is a color-coded map of the United States on the website that clearly explains the surrogacy laws that apply to each state, which makes it a very good resource. You can find out how the law works in each state just by clicking on the state image. Keep in mind that the practice of surrogacy is ever changing. An experienced attorney must still determine the prevailing practice in a particular state court at the time surrogacy is initiated.

US Department of Health and Human Services—Administration for Children and Families

<https://www.childwelfare.gov/topics/adoption/laws/>

Adoption laws vary significantly from state to state yet they must comply with general rules set by the United States Federal Government. This resource answers some of the questions you might have about adopting a child from child welfare services; however it is not meant to replace a conversation with an attorney that is knowledgeable about adoption laws, the cost, etc. where you live.

Domestic Adoption—Parent's Magazine

<http://www.parents.com/parenting/adoption/domestic/>

This website is sponsored by “Parents” magazine and does a nice job explaining the different types of adoption offered in the United States. You’ll also find stories from families who talk about their adoption experience.

Intercountry Adoption—Bureau of Consular Affairs—U.S. Department of State

<https://travel.state.gov/content/adoptionsabroad/en.html>

This website offers information for United States citizens who are considering adopting children from abroad. Adoption between the U.S. and other countries is different than domestic adoptions and is governed by both the laws of the country in which a child lives and the country in which the adoptive parents live. According to U.S. law, there are two different intercountry adoption practices. These include the “Hague Convention” process and the “Non–Hague” Convention process. An American citizen who wishes to adopt a child must follow the process of the country for which the child is from. You can learn about the adoption practices in most foreign countries by clicking “Country Information”, then searching for a country in the drop down menu.

Insurance Coverage for Fertility Treatments by State

http://www.resolve.org/family-building-options/insurance_coverage/state-coverage.html

The Resolve.org website provides visitors with a list of the 15 states that are mandated by law to provide infertility treatment. When you access the link you can look to see if your state is listed and then click to find out exactly what your insurance company is legally mandated by law to provide. If you don’t see your state listed, there is no law at this time that requires that insurance companies cover treatments such as IVF. However, your personal health insurance may offer coverage.

MRKH: A Guide for Health Care Providers

What is MRKH?

Mayer–von Rokitsky–Kuster–Hauser syndrome is a congenital anomaly of the female reproductive tract which may also be referred to as mullerian aplasia or mullerian agenesis. It is caused by embryologic growth failure of the mullerian structures. Approximately 1 in 4500 females who are diagnosed with MRKH, have vaginal agenesis, or an incomplete vagina and an absent uterus. Approximately 7%–10% of affected individuals have a normal but obstructed uterus or a rudimentary uterus with functional endometrium. Another variant is the presence of a uterus but with agenesis of the vagina and cervix. Individuals with MRKH have normal external genitalia and a normal female 46, XX karyotype with normal ovarian hormonal/oocyte function. Girls with MRKH have normal female serum testosterone and estrogen levels. MRKH is a syndrome that may or may not be associated with renal (absence of one kidney), skeletal, and hearing problems. The average age of diagnosis is reported to be between 15–18 years of age with the chief complaint being amenorrhea.

Clinical presentation

A young woman typically presents to her pediatrician secondary to primary amenorrhea. If an attempt is made to perform 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. Girls with MRKH generally go through puberty similarly to other adolescents, and have secondary sex characteristics such as pubic hair and breast development. Therefore, it is rare for a health care provider to suspect that anything might be formed differently before this point.

Physical Exam: The external exam reveals normal anatomy with secondary sexual development, normal perineum, and a vaginal dimple or small blind pouch. A rectoabdominal exam is sometimes done to determine whether the midline structures (upper vagina, cervix, and/or uterus) are present. Visualization of the introitus and a gentle exam using a small saline moistened cotton–tip applicator, Calgi swab, or a

gloved finger (if patient is able to tolerate) will help determine the length of the vagina.

A Routine Pelvic Ultrasound: A pelvic ultrasound is done to assess renal structures and can confirm normal ovaries and an absent uterus. Remnants of uterine structures may be present and may cause cyclic or chronic pelvic pain. This is typically due to endometriosis which is caused by retrograde menstruation. An MRI may be necessary to evaluate whether there is functional endometrium present in a normal uterus or rudimentary horn. If endometrial tissue is present in a non-communicating rudimentary horn, a removal of the small horn or other tissue is necessary. In rare cases, an MRI may not be able to capture laterally displaced uterine horns.

Genetic testing: A blood test may be done to confirm normal female karyotype and an FSH level may be evaluated to determine normal ovarian function.

TREATMENT OPTIONS

1. **No treatment– or delayed treatment** until a young woman is mature enough to make her own decision. The decision for treatment or not to have treatment should always be made by the patient, after careful consideration of options.
2. **Non–surgical methods to create a neovagina**–Frank, Ingram, Progressive Perineal Dilation (PPD) and coitus. These methods include using dilators that expand and stretch the vaginal dimple or existing small vagina. Dilators should be prescribed by an adolescent gynecologist who specializes in caring for young women with congenital anomalies of the reproductive tract. Dilator teaching should be done by an experienced provider such as a gynecologist, clinical nurse specialist or nurse educator. Prior to receiving dilator teaching, young women should be evaluated and counseled by a knowledgeable nurse, social worker, therapist, or counselor to evaluate readiness for treatment and more importantly, to allow the patient an opportunity to ask questions about her diagnosis, what the dilation process will entail, fertility options, and how to communicate with family and friends.

3. **Vaginoplasty**– Surgery which involves using a split skin graft from another part of the body such as the buttocks or bowel. (Ex. McIndoe procedure).
4. **Vecchetti procedure**– A combination of surgery and dilation with continuous pressure using a traction device.

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 noninvasive without the risks of surgery or anesthesia and it actively engages the patient in her treatment.

DILATORS

The use of dilators is referred to as the Frank Method. The original dilators were made out of glass. In the early 1980's, Dr. Ingram created different size plastic dilators along with a seat that attaches to a stationary bicycle. This method is rarely recommended because it is difficult and awkward to use.

Today, hard plastic dilators have replaced glass dilators. The most widely prescribed dilators come in a set with different sizes ranging from XS–L. Most young women begin dilation with the XS size dilator. Typically an adolescent gynecologist, or clinical nurse specialist will teach the patient how to insert the dilator, the correct angle and amount of pressure to apply. The patient will take one dilator home with her at a time, and use it 2–3 times a day for about 15–20 minutes. In the beginning, most of the dilator is used as a “handle” because the existing vagina may be just a dimple or 1–2 cm in length. Later, as the vagina stretches, more of the dilator will be able to be inserted. The amount of time that it will take to create a vagina is dependent on how often and how consistently the young woman uses her dilator.

Follow–up: After a young woman begins dilation, she should return monthly for follow–up appointments so that her progress can be monitored. When the patient has made significant progress, the next size dilator is dispensed. It is VERY important that the patient keep her GYN appointments so that her progress can be monitored. Some young women become frustrated if they do not see instant results and may stop

the dilation process and resume treatment later. The patient should not be reprimanded for not using her dilator; on the contrary, she must be reassured that it is her body, and her decision to dilate is hers alone. In general, it takes about 6–12 months but can take as little as 3–6 months if a young woman is motivated. After the desired outcome is achieved, girls who are not sexually active should use the largest size dilator for 15–20 minutes, once a week to maintain patency. Our success rate for helping patients to create a vagina at Boston Children’s Hospital is approximately 85–95%.

SURGERY TO CREATE A VAGINA

McIndoe Procedure: A vaginoplasty involves making a vagina using a segment of the bowel, or skin graft from the buttocks (McIndoe) or another site and a vaginal mold. Young women who have this procedure must stay in the hospital on bed rest for approximately one 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 void or to have a bowel movement. Even though a vagina is created faster with surgery, the patient must be made aware that it is still necessary to use dilators.

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 down side with this procedure is that the angle of the newly created vagina can be awkward and the cosmetic appearance may be less than the patient wishes.

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 the patient will likely have chronic vaginal discharge and may need to wear a pad all the time.

Davydov procedure: The Davydov operation is a type of surgical procedure to create a vagina using a patient’s own peritoneal lining. The peritoneum is a membrane that lines the walls of the abdominal and pelvic cavities. While the patient is under anesthesia, an incision (cut) is made where the vaginal opening should be. Another incision(s) is made

near the belly button to allow a thin instrument to be inserted into the peritoneal cavity. The peritoneal lining is then pulled down and stitched in place at the vaginal opening. The top of the newly created vaginal canal is then sewn closed. The vaginal space is then packed with gauze. The gauze is typically removed about two days after the operation. After the packing is removed, the patient must use various size vaginal dilators a few times a day for several months or until she becomes sexually active. The goal is not to obtain additional length or width but to maintain the new (newly created) vagina and prevent scarring. Most women who have this procedure stay in the hospital overnight for observation and then return for follow-up visits within the 7 to 10 days after surgery.

Laparoscopy–Vecchietti procedure: The Vecchetti procedure combines the use of dilators and a surgical procedure. This technique involves applying a traction device to the abdomen. This is done during a laparoscopy under general anesthesia. 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 a 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. Dilators must be used following the procedure. The Vecchietti procedure is popular in Europe, but it is not commonly done in the United States.

When should a young woman with MRKH begin to create a vagina?

The best time for a woman to either use vaginal dilators or have surgery is **when she is ready**. The timing is different for everyone. The most common age at which girls decide they are ready to create a vagina is approximately between 16 –18, or 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, and some decide not to do anything at all.

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

If your patient is having difficulty or worried that she is using the dilator

incorrectly, she should talk with a member of her GYN team so that they can assess any difficulties she may be experiencing and give her feedback and support regarding her progress. If she has used dilators properly and consistently and has had minimal progress, the GYN team may discuss surgical options.

What if my patient avoids using her vaginal dilator?

There may be one or several reasons why your patient may stop using her dilator. She may say that she doesn't have the time due to a demanding school and extracurricular schedule, is worried that it is painful, or perhaps she is concerned about not having privacy. It is fine to ask your patient how things are going to determine if she is feeling comfortable with her treatment plan but we discourage reminders to use her dilator by her medical team and/or parents.

If your patient continues to avoid using her dilator, it may be that she is simply not ready to create her vagina. Suggest that she speak with a member of her GYN team about delaying treatment until she is ready will likely relieve her anxiety and allow her to have control over the situation.

Will my patient be able to experience sexual pleasure?

Yes. Typically, women do not experience pain with intercourse after treatment and can have a satisfying sex life. We typically suggest the use of a vaginal lubricant such as K-Y jelly since women with MRKH may have less vaginal secretions.

FERTILITY

If your patient 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 will be able to have a biological child with assisted reproductive technology and a surrogate. This, however, is very costly and a financial burden for most families.

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

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

COPING WITH A NEW DIAGNOSIS OF MRKH

It is not uncommon for adolescents 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 patient understands her diagnosis, is able to verbalize her concerns, and becomes involved in her treatment plan. Typical signs that your patient 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 patient may be having a more difficult time adjusting to her diagnosis. If you or her parent(s)/guardian have concerns, you might suggest that your patient talk to a professional. It is highly recommended that the counselor, therapist or social worker have experience with adolescents who have medical issues. It is also very important for the counselor to have accurate information about MRKH. You can direct them to www.youngwomenshealth.org.

Resources at *youngwomenshealth.org*

Annual Conference for Teens with MRKH and their Families:

The Center for Young Women's Health (CYWH) hosts an amazing Annual Conference for Teens diagnosed with MRKH and their Families which is open to interested young women regardless of whether they are patients at Boston Children's Hospital.

For more information and the schedule of events, visit:
youngwomenshealth.org/mrkh-conference/

Monthly Moderated Online Chats:

The CYWH hosts three free monthly MRKH chats that are moderated by a nurse educator and media coordinator; two for teens (one in the afternoon and another in the evening), and a monthly evening chat for parents.

For more information, visit: youngwomenshealth.org/online-health-chats/. This online support group is an opportunity for teens to talk with others with whom they can relate.

