

Information for Women

ROKITANSKY SYNDROME



INSTITUTO ROKI

Apoio a mulheres com
Síndrome de Rokitansky



For You.

I am a teenager with Rokitansky Syndrome and one of the creators of the Roki Institute. When I received my diagnosis at 13 years old, I felt a pain like I had never felt before, as if that news had ripped a part of myself away. My subconscious believed that my life path was going to be like the norm: losing my virginity, meeting someone special, getting married, and having children. After all, that is what is expected of us nowadays. The strangest part was discovering something so significant about myself that had always been there, and I just didn't know it, as if I had lived my entire life with a lie about who I believed I was. I went through a period of anger right after my diagnosis. Why me? What did I do to deserve that? What will people think of me if they find out? No one could tell me how to deal with it, and even worse, no one knew WHAT it was. For a long time, I felt completely lost, didn't know who to turn to, and didn't know who I could trust to tell about this newfound discovery. The years went by, and as I matured, I gradually began to accept myself. One thing that went through my mind was, will anyone ever be able to love me because of that? It has been 5 years since my diagnosis, and nowadays, I can finally answer that question. The first person who has to love me is myself, and only this way, I will be able to be loved. For a long time, all I wanted was not to have been born like this, but nowadays, I can say with conviction that this is a part of me, it is a part of my history, it is a part of who I am, and most importantly, it has given me the opportunity to help you, who might be as lost and destroyed as I was. You are not alone, we are a very large group of warriors, and we will go through all of this together!

With love,

Isabella Barros, co-founder of the Roki Institute.



Rokitansky Syndrome

INFORMATION FOR WOMEN

THE SYNDROME

Rokitansky Syndrome is a congenital disease of the female reproductive system. [Read more here](#)

DISCOVERING THE SYNDROME

How the medical diagnosis is made. [Read more here](#)

TREATMENTS

Treatment options. [Read more here](#)

SEXUAL LIFE AND MOTHERHOOD

Alternatives for motherhood. [Read more here](#)

TALKING ABOUT THE SYNDROME

Talking is important. [Read more here](#)

The Rokitansky Syndrome

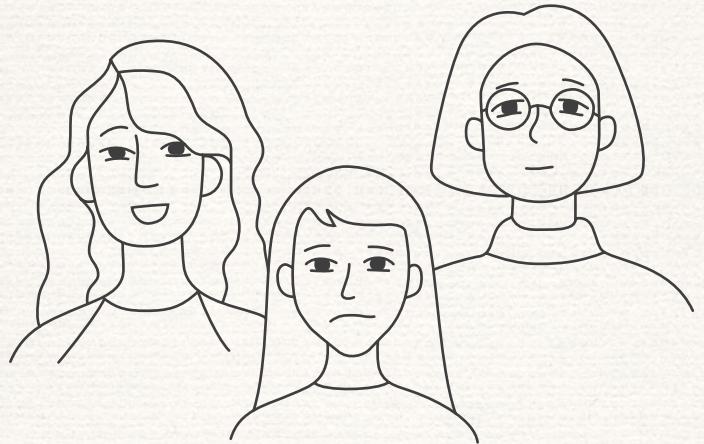
Rokitansky Syndrome is the name used in Brazil and is named after one of the doctors who described it. Worldwide, the term MRKH Syndrome is used, which corresponds to the initials of all the doctors who described the syndrome (Mayer, Rokitansky, Kuster, and Hauser). Maybe one day we will be able to simply call it Roki Syndrome. It is a congenital syndrome that affects the female reproductive system. Syndrome is a set of symptoms and signs. Congenital means that it is acquired during fetal development, inside the mother's womb, and present at birth. It affects about 1 in every 5,000 women. We do not know the cause of this syndrome, but we know that the reproductive system (vagina and uterus) is formed during the first months of fetal life (while a baby is still in its mother's womb). With the syndrome, the reproductive system begins to grow, but does not develop completely.

Girls with Rokitansky have normal external genitalia. This means that the part we see, called the vulva, which includes the clitoris, urethra (urinary canal), small and large labia, hymen, and anus, have normal development. The ovaries (where eggs are produced) and fallopian tubes (the conduit that carries the egg from the ovary to the uterus) also have normal development. The altered formation occurs in the uterus and vaginal canal.

Most of the time, the uterus is absent or tiny. The vaginal canal (the path between the uterus and the vulva) may be shorter and narrower than usual or be absent.

When is Rokitansky Syndrome diagnosed?

The most common age for diagnosis is between 13 and 18 years old. The most frequent history is seeking medical care because the girl does not menstruate or because she has pain and difficulty in sexual intercourse. Some may discover it earlier or later for other reasons, such as accidents, urinary loss, or lower abdominal pain.



WITH
Rokitansky Syndrome

WITHOUT
Rokitansky Syndrome

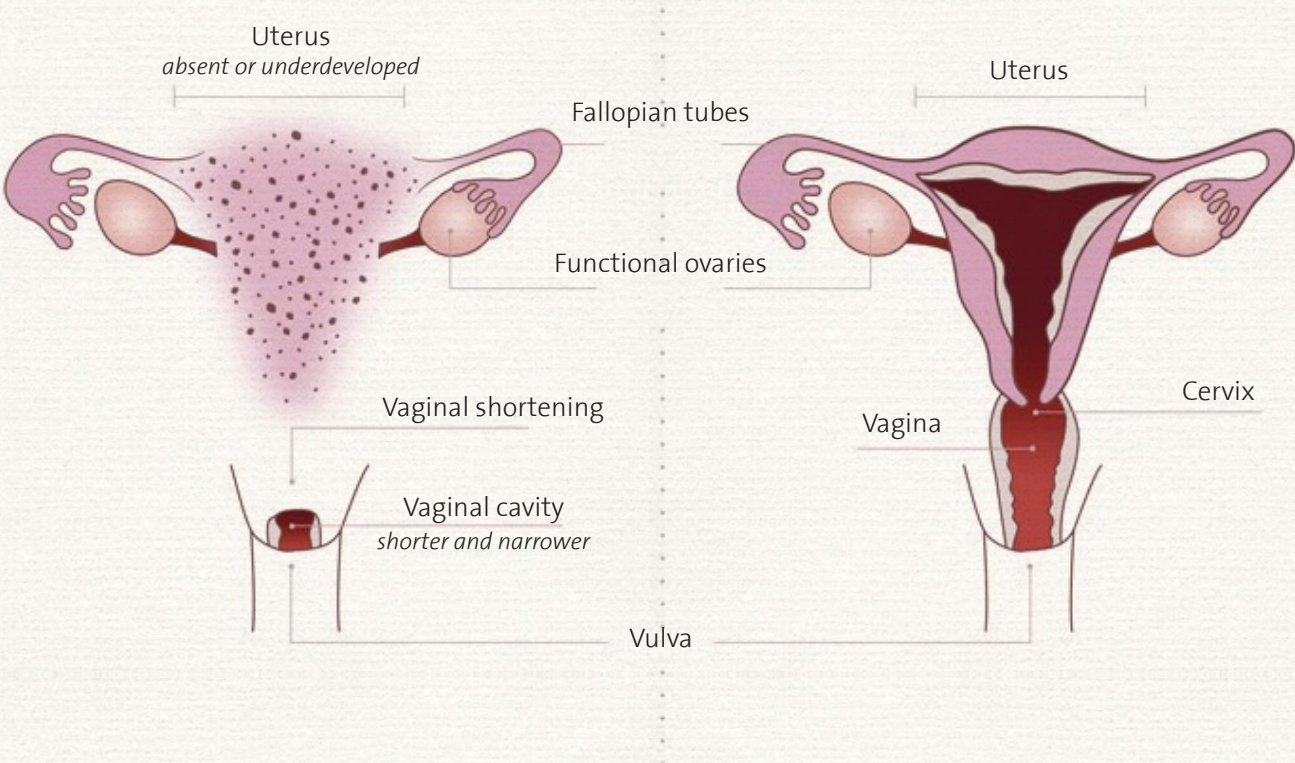


IMAGE: FLORAVI.COM

Discovering the Syndrome

Can anyone diagnose me with Rokitansky Syndrome (besides my doctor)?

The answer is no. Nobody except your doctor can say that you were born with an incomplete vagina. After treatment, with dilators or surgery, your sexual partner will not be able to feel any difference.

What will happen at the doctor's appointment?

Your doctor should ask questions like: "When did your body change (breasts, hips, hair growth)?" "Do you feel pain or any other changes?". It will be important for you to tell him everything that bothers you! Next, you should be examined: your doctor will gently evaluate your external genitalia and check your vagina through a manual exam or with the use of a small speculum. It is normal to feel embarrassed and nervous, but the exam is quick, and discomfort will be less if you can relax. Your doctor will probably request an imaging test called an ultrasound or magnetic resonance imaging. These tests do not hurt and are similar to an X-ray. Normally, your doctor will refer you to a specialist (gynecologist or pediatrician) who has experience with Rokitansky Syndrome.



What can a pelvic ultrasound or magnetic resonance imaging show?

A pelvic ultrasound is usually the first test to check if the uterus is present and what its size is. A very small uterus is called a "childish uterus" or "uterine remnant". This exam can also confirm that you have normal ovaries and kidneys. You may need a Magnetic Resonance Imaging so that your doctor can see your internal female organs in more detail.

Treatments

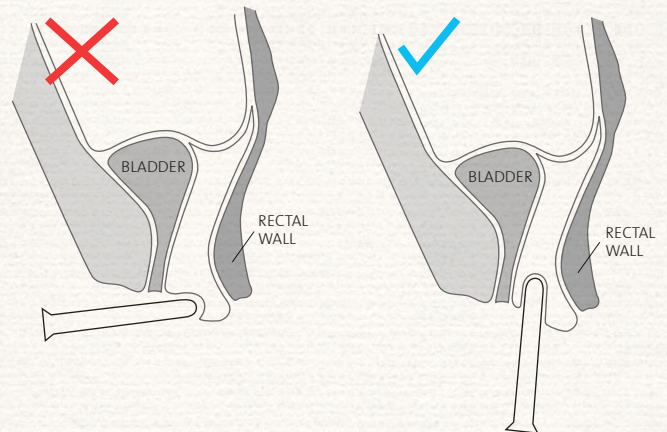
What is the difference between treatment with dilators and surgery?

Dilation is the most commonly used and recommended method by specialists. You will be the one performing the treatment, daily or a few times a week, for a few minutes up to half an hour per session. Progressive size dilators are used. The complete duration of treatment can last for months up to 1-2 years, depending on your choice. The beginning is more painful, so you may use vaginal creams to facilitate it, and as the sessions progress, the discomfort decreases.

Surgery may seem easier and faster, but complications can occur, as in any surgical procedure. Your doctor will provide more details.



CAREFUL PLACEMENT OF DILATORS



Sexual life and motherhood

If I don't create a vagina and decide to have sex, will something bad happen?

If you have vaginal intercourse before your vagina is created, either by using dilators or surgery, the sex might be very painful. You must be careful not to cause trauma or bleeding. Creating a vagina through sexual intercourse can be done, but it's important to be aware and gentle to avoid risks and discomfort. However, there are other ways that couples can be sexually intimate that don't involve vaginal penetration. When your vagina is ready, you can have normal sexual activity and pleasure, just like any other woman.

Will I ever be able to have a satisfying sexual life?

Keep in mind that every woman, regardless of her age or anatomical characteristics, experiences sexual stimulation and pleasure in different ways. Discovering what pleases you sexually is a process of evolution for all women throughout their lives. Exploring your sexuality with yourself and your partner is important in order to find comfortable and pleasurable movements and positions.

Can I have children?

If you were born with an incomplete vagina but have a normal-sized uterus, you will probably be able to get pregnant and have a baby. If you were born without a uterus or with a small uterus, you will not be able to have a natural pregnancy. Since your ovaries should produce eggs normally, you could have a baby carried by someone else's womb, called a surrogate. Adoption may also be a choice for you.

Surrogacy

Someone like your sister, friend, or another person can have a pregnancy for you. You will hear beautiful stories of surrogacy. This child will be generated from your egg and your partner's or a donor's sperm, so you will be the biological mother. Your baby will have your genetics and will grow for 9 months in a "borrowed" womb.

Uterine transplant

This is a surgery that was first performed in Sweden in 2014 with a living donor and in Brazil in 2016 with a deceased donor. The process is complex, with many steps, and performed in large medical centers. The first step is the in vitro fertilization procedure, in which eggs and sperm are collected, embryos are created and frozen for later use in the transplanted uterus (up to here the same process for surrogacy).

Next, it is necessary to find a living or deceased uterus donor. The donor must be compatible, and the transplant surgery is then performed. It is necessary to wait for several months or up to a year using immunosuppressive medications before

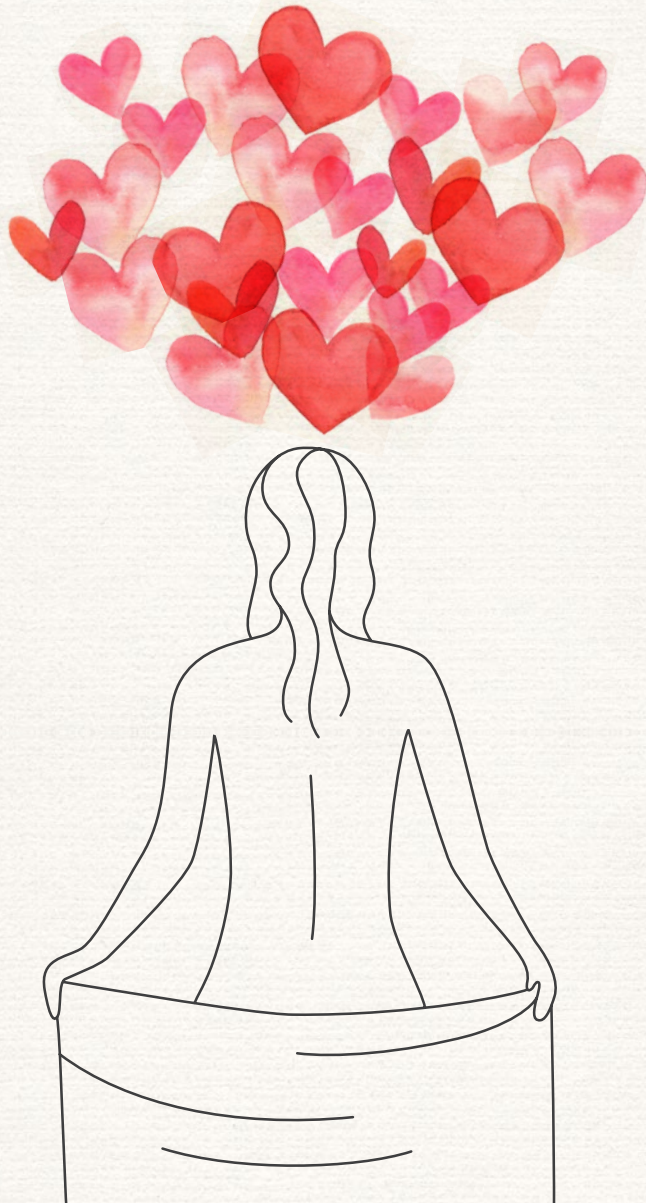
placing the embryo in the transplanted uterus. The immunosuppressive drugs must be taken to prevent the body from rejecting the transplanted uterus while it is in place.

Some complications may occur, but if everything goes well, a cesarean section is performed during childbirth. After the baby is born, you have the option to keep the donated uterus for a second pregnancy or remove it immediately after delivery. Some babies have already been born from women who underwent uterine transplant, but there have also been cases of unsuccessful transplants with complications during the process. Currently, the procedure is performed on an experimental basis and is only recommended for women who have not undergone previous surgical neovaginoplasty.

Science and the search for fertility options are evolving, and other options should be available when you are ready to have children.

Why do I have pelvic pain (cramps) every month?

Generally, some women may feel discomfort or a twinge in their lower belly when they ovulate each month. Most of the time, this pain is mild, and there is nothing to worry about as it is caused by normal ovulation. Some women with Rokitansky syndrome may have a small uterus that is not enough to carry a baby, but sometimes it can cause cramps if there is minor bleeding in the pelvic cavity. Your gynecologist will be able to tell if you have a small uterus and if it needs to be treated. If you have belly pain, it is important to tell the medical team.



Talking about the Syndrome

In my room, more people sleep. How do I get privacy to use the dilators?

Most young women need privacy when using dilators. When you're sharing a room, whether with siblings or roommates, it can be uncomfortable to ask for some private time, especially if you don't want to share details about your diagnosis and treatment. There are many reasons why people need some time alone: to meditate, study, nap, pray, shower, etc. It's always best to plan your routine and talk to whoever shares your room to know when you can have more privacy to use your dilator.

I'm really embarrassed about all the medical visits - will it ever get easier?

Many young women diagnosed with Rokitansky feel pushed into a world of new information and experiences. It's perfectly normal to have a range of emotions: sadness and anger, hope and worry, fear and embarrassment, all of which can be part of the experience. Talk to your doctor, tell them what bothers you, as they may refer you to other healthcare professionals who can help. You may want interaction and information in appointments, or just be as objective and brief as possible. Tell your doctor which way you feel most comfortable.

My parents want to keep talking to me about the syndrome, but I feel like my privacy has been invaded. How do I maintain 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 problem that requires many appointments and tests, it can be difficult to have privacy. The syndrome involves you and your parents, thinking of you as a sexual person, so the level of distress in the family can be very high. For many young people and their parents, conversations about their body can be uncomfortable or even felt as an invasion of privacy. But remember, they are all together, and want only the best for you.

At the same time, just as you had to get used to this diagnosis and everything that it means, your parents did too. They are probably worried about you and how you're coping with this new information. Many parents show this by asking a lot of questions.

It is important for you to be honest with your parents about what you need, including respectfully letting them know when you don't feel like talking. Your parents may find it necessary to talk to other family members, such as an aunt or grandparent,

but you should be included in the decision about who will be informed about your diagnosis.

Your parents may also find it helpful to read the Information Guide for Families, where we answer some frequently asked questions and help clarify doubts. You can also talk to your doctor to help communicate with your parents.

Is there anyone else I can talk to about having Rokitansky Syndrome?

Some women find it helpful to talk to other family members, while others prefer to talk to a close friend. Sharing your experiences and emotions with other women with the syndrome can be empowering. The Roki Institute is here to talk and connect you with other young women.

Can my vagina close up?

Once you have created your vagina, it should not shrink if you have vaginal intercourse or use the dilator once a week for 15-20 minutes.

Am I still biologically a woman?

Yes. It is common for a girl to question whether she is “really a woman” when she is told she was born with an incomplete vagina and/or no uterus. If you

have asked yourself this question, you are not alone. However, it is very important to understand that you are a woman. Your doctor can order a test that can confirm that you are a genetic woman and have 46,XX chromosomes.

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

The choice of when and how to undergo treatment is yours to decide! Like any important decision in life, it is essential that you obtain all the information to decide on the best possible way. Talking to other women who have the syndrome will help you mature your decision. You are in control of your body. You should never be forced or pressured by your parents, partner or medical team to undergo treatment. Instead, they should support you during the treatment process, only when you decide it is the right time.

The medical team should inform and clarify all your doubts so that you can make safe decisions. Your parents should offer support, seek medical care, and ensure privacy if you opt for treatment with dilators. And we will be here to help and support you whenever you need it. You are not alone on this journey! Everything will be alright.



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Síndrome de Rokitansky

If you have been diagnosed with Rokitansky Syndrome,
please contact the **Roki Institute**.

Our commitment is to welcome and assist you!

www.institutoroki.org.br



All information is educational in nature. Created by the Roki Institute, based on material provided by the Center for Young Women's Health and Boston Children's Hospital.

For diagnosis and treatment, please consult your doctor.