

Information for family members

ROKITANSKY SYNDROME



INSTITUTO ROKI

Apoio a mulheres com
Síndrome de Rokitansky

To parents,

When I found out that my daughter had Rokitansky Syndrome, the news not only brought a whirlwind of emotions, but also many doubts. I sought information from the best doctors, psychologists, and psychiatrists, but their answers were too basic and without any explanation that gave me confidence to help her through this disruptive moment, in a phase where doubts are already an intrinsic part of adolescence.

For us, parents, love never changes, but the need to see less pain in my daughter made me search for more answers. That's how I came across the organization Beautiful You and the MRKH Conference in Boston. I found medical answers and emotional support for girls, parents, and family members.

When I returned to Brazil, loneliness, anxiety, and doubts no longer took over my life, and the urgent need to make my daughter feel this peace - which I felt when I received that support - brought the idea of helping other families in the same situation, to deal with their emotions and reactions to this new reality.

Thus, together with Isabella and Claudia, my partners, we have made it possible to create the Roki Institute in order to answer all your questions, concerns, and, most importantly, to welcome you with great affection!

To you, my daughter Isabella, your father and I dedicate this institute, very proud of the way you have faced this obstacle in your life, of the woman you have become, and of the courage you have had by turning pain into love for others!

With love,

Luciana Leite, co-founder of the Roki Institute

Rokitansky Syndrome

INFORMATION FOR FAMILY MEMBERS



THE SYNDROME

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

DISCOVERING THE SYNDROME

The characteristics include normal ovaries and fallopian tubes, an absent or incomplete vaginal canal, and underdeveloped uterus or complete absence of the uterus. [Read here](#)

TREATMENT

Treatment options include dilation of the vaginal canal, surgery, or a combination of both. [Read here](#)

SEXUAL LIFE AND MOTHERHOOD

Women with the Syndrome will have alternatives for motherhood. [Read here](#)

TALKING ABOUT THE SYNDROME

Talking is important, respecting your daughter's time. [Read here](#)

Rokitansky Syndrome

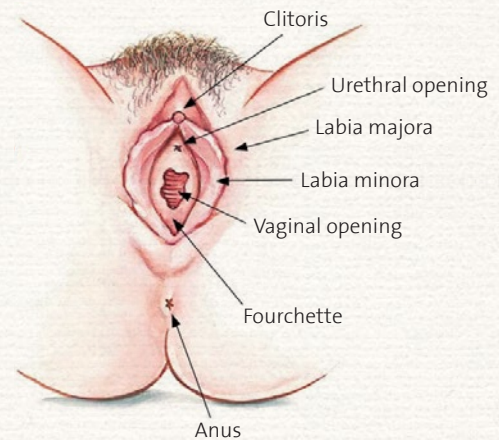
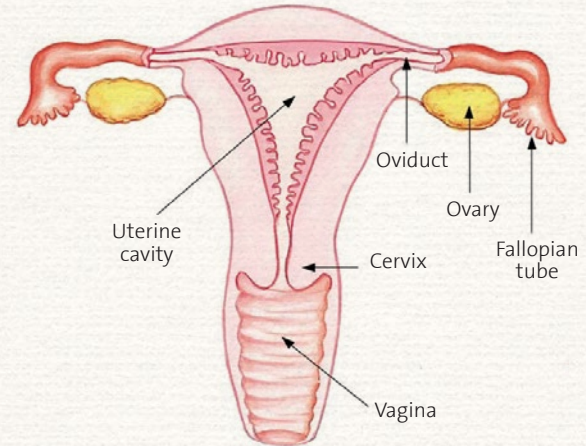
Rokitansky Syndrome is the name used in Brazil and is named after one of the doctors who described it. Worldwide, the term MRKH Syndrome is used, which corresponds to the initials of all the doctors who described the syndrome (Mayer, Rokitansky, Kuster, and Hauser). Perhaps one day we can simply call it Roki Syndrome.

It is a congenital syndrome that affects the female reproductive system. Syndrome refers to a set of symptoms and signs, and congenital means that the condition is acquired during fetal development and present at birth. It affects about 1 in every 5,000 women.

The cause of Rokitansky Syndrome, in which the reproductive system does not develop completely, is still unknown. The external genitalia, called the vulva, which includes the clitoris, urethra (urinary canal), labia minora, labia majora, hymen, and anus, develop normally. Ovaries and Fallopian tubes also develop normally, so your daughter will produce eggs and hormones naturally.

The altered formation occurs in the uterus and vaginal canal. In most cases, the uterus is absent or underdeveloped. The vaginal canal is shorter and narrower than usual or may even be absent.

In a rarer form of this syndrome, type II, which affects 1 in 10,000-15,000 women, there may be alterations in kidney formation, slight hearing loss, spinal problems, and heart abnormalities.



Discovering the Syndrome

Why are we only discovering this condition now and what are the diagnostic tests?

The most common age for diagnosis of Rokitansky Syndrome is between 13 and 18 years old. The most frequent reports are those in which the family seeks medical attention because the girl does not menstruate or experiences pain and difficulty during sexual intercourse. Some may discover the condition earlier due to other reasons, such as accidents, urinary incontinence, or lower abdominal pain.

Pelvic exams are not done at birth or during childhood, so it is rare for the diagnosis to be made before adolescence. Girls with the syndrome go through puberty like other young adults, with normal sexual characteristics such as the development of pubic hair and the typical increase in breast and hip size. Therefore, it is rare for a doctor to have any suspicion before adolescence.

For diagnosis, the pediatrician or gynecologist may perform an adapted vaginal touch exam, if your daughter has not had sexual initiation, or may request imaging tests to confirm the diagnosis.

Pelvic ultrasound is usually the first test to check if the girl's uterus is present and what its size is. A very small uterus is called "infantile uterus or

remnant uterus." This test can also confirm if the patient has normal ovaries and kidneys. Magnetic Resonance Imaging can assist in viewing the female internal organs with more detail.

Blood tests may be requested to confirm if ovarian hormones and female karyotype (46, XX) are normal.

Could I somehow have caused this syndrome in my daughter?

The cause of the syndrome is still unknown. It is acquired during fetal development, so it is common for many mothers to feel responsible, as with other congenital conditions. No maternal behavior or cause is associated with this syndrome. Do not blame yourself, nor worry, you were not the cause of the alteration in your daughter.



Treatment

Treatment options

If your daughter has been diagnosed with incomplete or absent vagina, she has the following options:

- She can wait and do nothing at the moment until she is ready for treatment;
- She can create a vagina. The first option is the use of dilators that expand and stretch existing vaginal tissue with intermittent pressure. This procedure should be taught by a gynecologist and/or specialist nurse. Surgeries are recommended only in case of failure of clinical treatments.

Waiting or doing nothing

Your daughter has control over her own body. Deciding whether to create a vagina or not should be her decision, after conversations with specialist doctors and parents. If your daughter plans to have vaginal intercourse at some point, creating a vagina will be necessary.

If your daughter is thinking about treatment, she should definitely consider using dilators initially. All surgical options have risks, require general anesthesia, and the use of dilators after the procedure. Therefore, surgery is not a quick or easy solution, nor a way to avoid the need of dilators.

Specialized medical groups in Brazil and worldwide recommend vaginal dilators for the creation of the vagina without surgery as the first treatment option for Rokitansky Syndrome. Surgical options should be discussed if dilation is unsuccessful.

Dilators

The progressive dilation technique, known as the Frank Method, was described in 1938. The original dilators were made of glass. Today, dilators are made of plastic and have different progressive sizes. The most recommended kits have 5 or 6 dilator sizes, ranging from 6 to 15 cm in length.

The first dilator recommended, at about 6 cm, is similar to a thin vaginal tampon. Your daughter should try to practice dilation daily, with sessions between 20 and 30 minutes. As the vagina stretches, the immediately larger size dilator is used until treatment is complete with the use of larger dilators, which can reach lengths between 14 and 15 cm.



The time period to create a vagina will depend on how consistently your daughter uses the dilators. The treatment typically ranges from 6 to 12 months, with some reports of shorter processes lasting 3 to 6 months when dilation exercises are done 2 to 3 times per day. There are also reports of longer processes lasting over 12 months, when the girl frequently interrupts dilations for days or weeks.

Is treatment with the dilator successful?

Yes, it is very successful. However, there is a wide variation in the success rate of dilation, mainly related to the teaching and expertise of healthcare professionals who educate their patients on this method. Make sure your daughter is assisted by an experienced gynecology program with high success rate in training patients to create a functional vagina.

How often does my daughter need to see a gynecologist?

Typically, your daughter will be asked to return for monthly or bi-monthly evaluations. A consistent follow-up during treatment is very important to assist and monitor progress. Your gynecologist will need to check if she is applying pressure in the right place and with the correct angle, so that the success rate of treatment increases and the dropout rate decreases.

When should my daughter start vagina construction?

The best time for your daughter to start treatment is when SHE is ready. The most common age for girls to decide to create a vagina is around 16 or 17 years old. Some young women are encouraged to start treatment when they encounter a sexual partner. As a parent or guardian, you may want your daughter to solve the problem as quickly as possible, but remember that treatment should only be started when she is ready.

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

If your daughter is having difficulty or you are concerned that she is using the dilator incorrectly, she should be encouraged to talk to her doctor. If she is not satisfied with her progress after using dilators for about 6 months, she and her gynecologist should discuss appropriate ways to carry out the treatment. Perhaps the solution is to wait and use the dilators in the future, or to start discussing surgical options.

What if my daughter is not using the dilator?

There may be one or several reasons why your daughter is avoiding the use of the dilator. She may feel she does not have the time, or may be concerned about having pain; or perhaps she is uncomfortable with lack of privacy. It is helpful to ask her how the process is progressing, and also if she is feeling comfortable with her treatment plan.

Meanwhile, it is not advisable to constantly remind her to use her dilator.

It's important for your daughter to maintain regular visits to her gynecologist, so that she can be evaluated, she can identify any difficulties that might come up, and she can receive guidance and support regarding her progress.

If your daughter continues to avoid using the dilator, it may be that she simply isn't ready to create her vagina. Having a discussion about delaying treatment until she is ready can alleviate her anxiety and allow her to have control over the situation. This will allow her to focus on other activities that are more important to her at the moment.

Surgeries

When a woman is not successful with dilator maneuvers, several surgical techniques are described and indicated in the minority of cases. Vaginoplasty or neovaginoplasty are the names of surgeries to create or enlarge a vagina.

We often create fantasies, believing that a surgery will form a vagina easily, with no problems. This is not true. Every surgical procedure has risks and possible complications during surgery and in our daily life. At the appropriate time, the medical team can discuss with both patient and family, surgical possibilities and the team's experience with each available technique.

Surgeries that used parts of the intestine to create the vagina were the first to be performed in the late 19th century. Due to mucus secretion in the region, bleeding, pain, and other side effects, this technique became less used nowadays. Naturally, technology has evolved a lot in the last two centuries.

Surgery with the *Davydove* technique uses part of the peritoneum for vaginal construction. However, cases of discomfort and pain due to traction are reported.

Another technique, initially described by *McIndoe*, used skin grafts on a rubber mold. The procedure, now called *McIndoe-Banister*, has evolved in relation to molds and grafts. After surgery, daily use of dilators is necessary for approximately 3 months.

The *Williams* technique uses the skin of the labia majora to form a "vaginal pouch." Women undergoing this technique mention discomfort during sexual activity, due to the axis of the new vagina, and also the occurrence of hair growth.

The *Vecchietti* surgery fixes threads by laparoscopy, which are attached to a traction system in the abdomen. Time, costs, care with the equipment, and a second surgery to remove the device are the complaints of patients during treatment.

Above all, make sure your daughter is being accompanied by a team of experienced doctors who are sensitive to the feelings, concerns, and doubts that everyone may have.

Sexual life and motherhood

Will my daughter experience “sexual pleasure”?

Most of the pleasure and sexual orgasm comes from clitoral stimulation. Usually, women do not feel pain during sexual intercourse and develop comfortable and pleasurable movements and positions after treatment.

When the time is right, talk to your daughter about sexual activity. A good suggestion is to recommend the use of lubricants. Before attempting vaginal intercourse, your daughter should speak with her gynecologist.

Can my daughter have children?

If your daughter was born with an incomplete vagina but has a normal-sized uterus, it is likely that she can conceive and have a baby. If she was born without a uterus or with a small one, she cannot have a “natural” pregnancy.

Since her ovaries should produce eggs normally, she can have a baby carried by another person, called a surrogate mother. Adoption can also be a choice for her.

Surrogacy

A family member, friend, or another person can “lend” their uterus for 9 months. Beautiful surrogacy stories are narrated and available for those who would like to know them. Your daughter would use her own egg, her partner’s sperm, or a donor’s sperm and be the biological mother.

Uterine transplant

The process is complex, with many steps, and is performed in large medical centers. The first step is in vitro fertilization, in which eggs and sperm are harvested, and embryos are created and frozen for later use (up to this point, the same process as surrogacy).

Next, a compatible uterus donor must be found, and then the transplant surgery is performed. It is necessary to wait for several months or up to a year while taking immunosuppressive medications so that the embryo can be placed in the transplanted uterus. Immunosuppressive drugs must be taken to prevent rejection of the transplanted uterus.

Some complications are reported, but if everything goes well, a cesarean section is performed at the time of birth. After the baby is born, there is an option to keep the received uterus for a second pregnancy or remove it immediately after delivery.

Currently, the procedure is performed on an experimental basis and is only recommended for women who have not undergone previous surgical neovaginoplasty. There are already some babies born from transplanted uteruses in the world, but there are also cases of failure with complications during the process. Science is constantly seeking fertility options. When she is ready to have children, other options should be available.

Talking about the Syndrome

How can I talk to my daughter if I feel uncomfortable discussing any topics related to her sexuality?

Teenagers often sense when their parents are stressed. It's okay to tell your daughter that you're feeling uncomfortable or having difficulty talking. At the same time, let her know that you want to talk to her and offer support. Most likely, she feels uncomfortable too. Although both of you may be feeling shy about talking, she will know that you're there to help her deal with the situation.

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

Your daughter may feel uncomfortable discussing her diagnosis because it often raises questions about her sexuality. A diagnosis of vaginal alteration can prematurely force a discussion about her as a sexual being, a conversation that can leave both daughters and parents in an uncomfortable or embarrassing situation. Trying to have a conversation when she's not willing to talk will be frustrating for both of you, so it's best to wait until your daughter is ready.

You can suggest that she read about Rokitansky Syndrome, participate in online support groups with other girls who have the same diagnosis, or talk to a trusted friend or sibling about her feelings. You should also let her know that you're available to listen if she wants to talk.

Remember, it's very important to start communicating by asking her what's the best way to help her! Your daughter should be completely involved in planning her best health care and, ultimately, in deciding her treatment.



How can I respect my daughter's right to make decisions about her body?

If your daughter is under 18, your consent for medical treatment will be necessary. It is crucial that all options are discussed in clear language with her, so she can participate in these important decisions about her body. Ultimately, the medical team will respect her wishes regarding treatment.

Should I tell someone about my daughter's diagnosis?

Sharing information about your daughter's diagnosis is a personal decision. It is always best to ask for her permission first. Typically, parents are selective about who they share this personal information with, as they find comfort in talking to a close relative or friend. Additionally, participating in online support groups for parents of girls with Rokitansky can also be very enriching.

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

It is up to you to decide if you want to share information and with whom. However, the more information you know about your daughter's diagnosis, the easier it will be to answer specific questions. Being able to share information will make these discussions easier. Do not hesitate, however, to say "I don't know". It is good for parents to set boundaries when people ask inappropriate questions. Remember, your daughter's privacy must be respected.

How do I know if my daughter needs professional help?

It is not uncommon for young women with the diagnosis of Rokitansky to undergo temporary mood swings, while they are adjusting to new comprehension of their body development. These concerns, however, should decrease over time, as they understand the diagnosis, are able to verbalize their concerns, and engage in appropriate medical care.

Typical signs that your daughter is upset may include changes of her sleep or eating patterns, comments on her body, concerns about treatments and medical appointments, and also changes in her behavior, like being too quiet, withdrawn, or irritable.

If you, your daughter, or the medical team have concerns, do not wait too long. You should talk to the gynecologist about referring her to a therapist or social worker. It would be helpful for the chosen professional to be familiar with the syndrome. You can present the Guide for Health Professionals or share information with them before your daughter's first meeting.

As parents, you may also have difficulty controlling your emotions regarding your daughter's diagnosis. If you are experiencing significant mood swings or difficulty dealing with your work or at home, you should also seek professional help. By doing so, you will be showing your daughter that there is nothing wrong with seeking help.



INSTITUTO ROKI

Apoio a mulheres com
Síndrome de Rokitansky

If your daughter has 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. 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.