NHS
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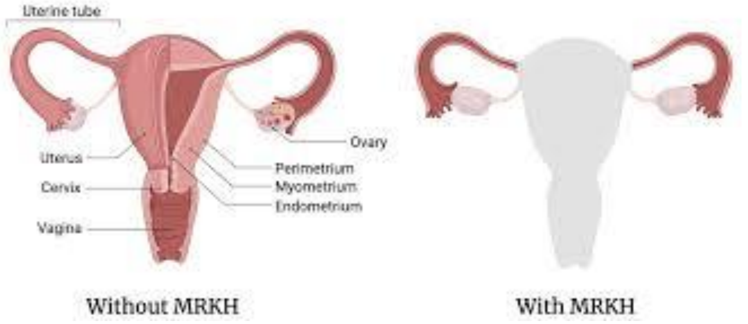
**Saint Mary's Managed Clinical Service**
**Division of Gynaecology**

**Paediatric and Adolescent Gynaecology Service**

**PATIENT INFORMATION LEAFLET**

# ROKITANSKY SYNDROME

Rokitansky syndrome (also known as Mayer-Rokintnasky-Küster-Hauser [MRKH]) is a condition you are born with and affects 1 in every 5,000 female births. In this condition, the womb (uterus) and vagina to not develop normally meaning you have a shortened vagina, no cervix (the neck of the womb) and either no or an undeveloped uterus. This condition is also known as Mullerian agenesis or Mayer-Rokitansky-Kuster-Hauser (MRKH) after the doctors that first learnt about it.



## ARE OTHER FEMALE ORGANS AFFECTED?

In Rokitansky syndrome, the ovaries are normal and make eggs (as they develop from a different place), and the external female organs (vulva) are also normal. The breasts are normal and people with this condition have normal female chromosomes and produce normal amounts of female hormones. No one can tell (not even doctors) by looking at an individual that they have the condition.

## CAN ANY OTHER ORGANS BE AFFECTED?

There are two types of Rokitansky syndrome: Type 1 and 2.

Type 1 affects the vagina, cervix and womb only. About 60% of women with Rokitansky have Type 1.

In Type 2, the vagina, cervix and womb is affected like Type 1 but you can also get other differences in development of the urinary tract and kidneys, hearing difficulties or bone changes. About 40% of women with Rokitansky have Type 2.

## HOW DO I KNOW IF I HAVE ROKITANSKY?

Rokitansky is usually discovered at puberty. Breasts and pubic hair develop because there are normal amounts of female hormones from the ovaries, but periods do not start because there is no womb.

## WHAT TESTS WILL I HAVE?

When you see a doctor, they will ask you questions about your periods and general health and they may examine you. Tests that are performed at the hospital include blood tests to check your hormone levels and your chromosomes along with an ultrasound scan.

In individuals with Rokitansky syndrome, the ultrasound scan shows that there is an absent womb and cervix and a shortened vagina but the ovaries are present. If an ultrasound is not clear, an MRI scan may be needed.

If a doctor is concerned you may have Rokitansky Type 2, other tests may be carried out such as hearing screening, a kidney ultrasound scan or an x-ray.

## CAN IT BE TREATED?

At the moment, there is no treatment to grow or develop the womb.

## DO I NEED TO HAVE SMEAR TESTS?

Smear tests are regular screening tests for cervical cancer. As individuals with Rokitansky syndrome do not have a cervix, they do not require smear tests.
However, it is a good idea to have the HPV1 vaccination (the vaccine that is offered to prevent human papilloma virus – HPV) as it protects against other cancers caused by HPV. These include mouth and throat cancer and some anal and genital cancers.

## WILL I BE ABLE TO HAVE SEX?

Some people with Rokitansky syndrome can have comfortable sex without any treatment. Others need treatment to help them. If treatment is needed, most people with Rokitansky syndrome can create a longer vagina by stretching the vaginal tissue that is already there. This is done by using specially designed smooth cylinders called vaginal dilators that are inserted into the vagina every day and gradually increase in size. This helps people with Rokitansky to have penetrative vaginal sex and is successful in 95% of individuals who use this therapy.
We have a leaflet on vaginal dilators for more information. There are also some surgical procedures available to increase the length of the vagina. All treatment options will be discussed with you as appropriate for your individual case.
Always remember to use protection when having sex, even if you do not have a uterus as you can still be at risk of sexually transmitted diseases and blood born viruses such as HIV.
There are several treatments available to lengthen the vagina to allow sexual intercourse.

## CAN I HAVE CHILDREN?

As people with Rokitansky syndrome are born without a womb, they cannot get pregnant or carry a baby.

An option to have a baby that is genetically yours is via IVF surrogacy. This involves having your eggs removed and fertilised by your partner’s or a donor’s sperm and then placed into another woman's uterus (a surrogate). You can be referred to fertility specialists to discuss this further.

Some women wish to look at other options and consider adoption.

The charity Womb Transplant UK is currently performing research into womb transplants which could be a possibility in the future however at present, this is in the research stage. If this is something that you wish to consider, please review their website where you can self-refer to the charity and we can support you with any other referrals that may be required.

## IS IT NORMAL TO FEEL ANGRY AND SAD?

Yes. Many people diagnosed with Rokitansky can find it overwhelming and hard to accept. Common emotions people can feel include shock, anger, sadness, isolation and rejection in any combination. You may need some time to come to terms with this diagnosis before making any further decisions. Parents of young people diagnosed with this condition can often need support as well. Charities such as MRKH Connect aim to provide support to individuals with this condition.

## WHAT SUPPORT DO YOU OFFER?

At Manchester Foundation Trust, we offer support in our Complex Gynaecology Clinic. In this clinic you can see nurse specialists on vaginal dilation, we can provide advice and guidance regarding fertility and you can seek support from a psychologist.

**If you require any further information or clarification, including clarification of terminology, please do not hesitate to talk to one of the doctors or nurses, who will be happy to discuss your concerns with you.**

# CONTACT DETAILS

Should you require any additional information or help please contact:

☎ **Benign Gynaecology**

0161 701 4455 (Option 1)