

MRKH

We understand, we care, we are
here for you.



MRKH support team

Mayer Rokitansky Küster Hauser syndrome (MRKH)

Information for patients, relatives and carers

Introduction

This patient information leaflet talks about Mayer Rokitansky Küster Hauser (MRKH) syndrome. Please use e-link or download our App or match the QR code on the last page for digital interaction.

What is MRKH?

MRKH affects 1 in every 5,000 women.

It is a condition that you are born with. Rarely, MRKH runs in families. There are two classifications of MRKH: Type I or Type II.

- Type I is characterised by a shortened or absent vagina, absent cervix and absent or undeveloped uterus (womb) made up of uterine remnant muscle only. The ovaries are present and function in the same way as any other woman's by producing eggs and female hormones that keep you healthy.
- Type II is characterised by having additional differences such as variations in urinary tract and kidney development, hearing difficulties or bone changes. Around 40% of women with MRKH will have Type II.

Signs suggesting you may have MRKH

Menstrual periods not starting when it should have, is usually the first sign in spite of a normal development of the breasts and pubic hair. The ovaries produce the female hormones that allow puberty to occur but no menstruation due to the absence of the uterus or womb. Girls/women with MRKH are genetically female (46XX karyotype).

What do you do next?

Talk to your GP and express your concerns. The GP will ask you lots of questions and may even advise investigation. Many GPs may refer you to the Gynaecologist at the hospital who will examine you and organize investigations to make diagnosis, including:

- An ultrasound scan to check if the uterus(womb) and its end part called the cervix is absent, the vagina for shortness and presence of both ovaries. Sometimes an MRI maybe necessary but the gynaecologist will explain.
- A special blood test to check chromosomes and to exclude any other genetic abnormalities
- If the gynaecologist is suspecting MRKH II, additional ultrasound of the kidneys or an X-ray maybe performed.

Can you have sex?

Yes.

Will sex be comfortable for you?

Some women with MRKH can have sex without needing to dilate the vagina first.

Others will need to dilate the vagina first to be able to have comfortable vaginal sexual intercourse.

What is vaginal dilation therapy?

Stretching the vagina with a specially designed cylinder-shaped dilator is known as the vaginal dilatation and is therapy of first choice. 95% of patients have been shown to be successful with dilator therapy. Once the vagina has been lengthened, sex should be comfortable. Creating extra length helps patients gain confidence with having sex and helps with their gender and sexual identity.

How long would you need to have vaginal dilator therapy for?

This therapy can take 3 months or sometimes longer. Regular use of these vaginal dilators is important for a good result. These are very effective in making the vagina comfortable for intercourse.

Can surgery help?

Some patients may benefit from surgery. The most commonly recommended operation is the Vecchietti procedure which is a laparoscopic (key-hole) operation done under general anaesthetic designed to create a new vagina by applying continuous pressure to the vaginal area using traction. But patients may experience post-operative pain and risks from the operation.

Who will supervise you during the therapy?

The clinical nurse specialist will teach you how to use your dilators and will support and supervise you throughout the process. If you are having any difficulties we would always encourage you to discuss them with someone in our team.

Treatment

At the moment, there are no treatments to grow or develop the uterus.

Will you need to have cervical smear tests?

No. As you do not have a cervix you will not require a smear test.

Will you need to use protection when I have sex?

Yes. it is always advisable to practice safe sex. If you have male partner, they should use condoms because you are still at risk of sexually transmitted diseases and blood-borne diseases such as HIV or Hepatitis B.

Do you need to get vaccinated against human papilloma virus?

It is beneficial to receive HPV1 vaccinations (the vaccine that is offered to prevent human papilloma virus - HPV), as this protects against other cancers caused by HPV.

Is it normal to feel angry and upset because you have been diagnosed with MRKH?

Yes. It is normal to feel angry and upset. Most women and their parents find this condition difficult to accept and can experience feelings of shock, anger, sadness, isolation and rejection. Many parents also feel very guilty.

Who can help you if you are upset following being told that you have MRKH?

The departmental clinical psychologist who is part of our team is available for appropriate support. Please speak to a member of our team.

Will you be able to get pregnant?

No. Since you were born without a uterus (womb) you will not be able to get pregnant or carry a baby.

Will you be able to have children?

Yes. Your eggs can be removed and fertilised by your partner's or a donor's sperm and then placed in another woman's (surrogate mother's) uterus. This is known as In vitro fertilisation (IVF) surrogacy.

Will the baby be biologically yours?

Yes. The baby will genetically be yours and your partner's/donors.

Does the government help with costs?

It is possible to apply for part-funding so that you can have your fertility and IVF treatment completed in an NHS Centre. Our team can provide you further information.

What if one does not wish to have fertility/IVF treatment?

One can consider other options like adopting children.

Who can you contact for further information or advice?

- You can contact us the MRKH helpline number : 00000000 and the service is available from 09.00 to 17.00, Monday to Friday. Outside of these hours, you can leave a message on the voicemail. You can also contact us at enquiries.mkrh@abc.com or write to MRKH centre, Outpatient department, ABC Hospital, City, Postcode
- We also have a support group which can offer you the opportunity to find out more about MRKH and to meet others who also have MRKH. Some of our own patients who have undergone vaginal dilator therapy, IVF surrogacy and adoption may also attend to share their expertise and experience with everyone. You can email the MKRH centre on support.mrkh@abc.com to become a member of this support group.
- We also produce a twice-yearly newsletter. You can register by emailing registeronline.mrkh@abc.com

- You can find an electronic version of this patient leaflet and links to further information on MRKH by scanning this QR code



or going directly to <https://infoleafletassignment13.com>.

- We love to have your feedback on our services. Kindly spare ten minutes to complete our online feedback form with a chance to win a £10 Amazon voucher. feedback.mrkh@abc.com

AS PER COMMENTS SECTION OF ASSESSMENT MATRIX NEED TO MAKE SURE YOU HAVE INFORMATION IN HERE ON:

- **THE CONDITION IN TERMS OF INHERITANCE AND RISK ASSESSMENT**
- **HOW IS DIAGNOSED**
- **THE DIAGNOSTIC TESTING (DIAGNOSTIC, PRESYMPTOMATIC...)**
- **HOW THE CONDITION CAN BE MANAGED INCLUDING HOW WOULD WORK WITH OTHER SPECIALITIES AND PROFESSIONS**
- **IMPACT ON PATIENT AND FAMILY**