

Centre for disorders of reproductive development and adolescent gynaecology

Mayer Rokitansky Küster Hauser syndrome (MRKH)

Information for patients, relatives and carers

Introduction

This leaflet talks about Mayer Rokitansky Küster Hauser syndrome (MRKH). Please feel free to ask our team any questions you have about the information below.

What is MRKH?

MRKH is a congenital abnormality (an abnormality you were born with), characterised as a shortened vagina, absent cervix and absent uterus (womb). 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.

MRKH affects 1 in every 5,000 women.

How will I know if I have this condition?

Women usually discover that they have MRKH during puberty. Although they develop breasts and pubic hair, they do not start having periods. This is because the ovaries produce the female hormones that make normal development occur but the absence of the uterus means that there is no period.

What tests will I have?

Your GP will normally ask you some questions and may examine you. Some GPs feel that this examination is best carried out at a hospital by a gynaecologist and may refer you for this. The doctor may also refer you for further investigations, including:

- a blood test to check your chromosomes and exclude any other genetic abnormalities
- an ultrasound scan or MRI scan to confirm the absence of the uterus (womb) and cervix and a shortened vagina, and the presence of ovaries

Women with this condition have a 40 per cent chance of having differences in the development of their kidney and urinary tract (for example, 15 per cent have only one kidney), therefore a renal scan or x-ray of this area may also be recommended.

Ten per cent of women with MRKH may experience some hearing difficulties and another ten per cent may have bone changes. If required, your doctor will arrange for some tests.

Will I need to have cervical smear tests?

No. As you do not have a cervix you will not require a smear test. However, it is still beneficial to receive HPV1 vaccinations (the vaccine that is offered to prevent human papilloma virus - HPV).

Will I be able to have comfortable sex?

Yes. The majority of women with MRKH are able to create a vagina by stretching the small amount of vaginal tissue already present. This is usually done by using specially designed smooth cylinder-shaped objects created for this purpose called vaginal dilators. Following this, you will be able to have sex.

You will be taught dilator therapy and supervised by the clinical nurse specialist throughout your treatment. This is the preferred and first line of treatment for women seen at our centre. Most patients (95 per cent) are successful in creating a normal-sized vagina using this technique alone. If you are struggling to complete the dilator treatment, discuss this with the specialist team. Please note that when you have fully stretched your vagina to a normal functional length, you will have completed the treatment. You can then stop using your dilators altogether.

Having sex with a partner is a personal and intimate affair. Once your vagina has been lengthened you should be able to enjoy sex normally. However, men's penises are different sizes and a man who has a large penis may cause you some discomfort as he would any woman. For this reason, you may need to stretch your vagina further. We will be happy to advise you on this.

Will I be able to have children?

As you were born without a uterus (womb) you will not be able to get pregnant or carry a baby. However, your eggs can be removed and fertilised by your partner's sperm and then placed in another woman's (surrogate mother's) uterus. This is known as IVF surrogacy and the baby will genetically be yours and your partner's.

This service is commonly available privately in recognised centres or clinics. It is possible to apply for part-funding from your GP so that you can have your fertility and IVF treatment completed on the NHS. Therefore, we advise that you and your partner discuss this further with your GP.

Your alternative fertility option is adoption and many of our couples have been successful in adopting children.

Is it normal to feel angry and depressed?

Yes. Most women and their parents find this condition very difficult to accept at first and express feelings of shock, anger, depression, isolation and rejection. Many parents also feel very guilty. There is a psychologist available to all women with MRKH who will offer appropriate support, which is a vital aspect of your care.

Are there any alternatives to this treatment?

The first line of treatment and the preferred choice for creating a vagina is dilator therapy. This is because our results show that 95 per cent of women will be successful with dilator therapy and surgery has risks, is painful and requires long-term dilator use.

Will I need to use protection when I have sex?

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

Who do I contact for further information or advice?

At this centre we have a support group which offers encouragement for you, your parents and your partner. The group meets twice a year, in the spring and autumn. The meeting in the spring is usually informal and offers the opportunity to meet other girls with the same condition. The autumn meeting tends to be more formal and informative, with specialist speakers and some of our own patients who have undergone IVF surrogacy and adoption coming to share their expertise and experience with everyone.

As well as the support group, we also have a helpline and a list of patient contacts, so please feel free to call us whenever you have any questions or if you would like to be put in touch with other women with the same condition. The helpline number is 020 3313 5363 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 for the clinical nurse specialist.

Alternatively, you can visit our MRKH support website at www.mrkh.org.uk

For details of how to register online, please telephone 020 3313 5363, email enquiries.drd@nhs.net or write to us at the address below:

Gynaecology outpatient department
Queen Charlotte's & Chelsea Hospital Du Cane Road
London W12 0HS

How do I make a comment about my visit?

We aim to provide the best possible service and staff will be happy to answer any of the questions you may have. If you have any **suggestions** or **comments** about your visit, please either speak to a member of staff or contact the patient advice and liaison service (**PALS**) on **020 3313 0088** (Charing Cross, Hammersmith and Queen Charlotte's & Chelsea hospitals), or **020 3312 7777** (St Mary's and Western Eye hospitals). You can also email PALS at imperial.pals@nhs.net The PALS team will listen to your concerns, suggestions or queries and is often able to help solve problems on your behalf.

Alternatively, you may wish to complain by contacting our complaints department:

Complaints department, fourth floor, Salton House, St Mary's Hospital, Praed Street
London W2 1NY

Email: ICHC-tr.Complaints@nhs.net

Telephone: **020 3312 1337 / 1349**

Alternative formats

This leaflet can be provided on request in large print or easy read, as a sound recording, in Braille or in alternative languages. Please email the communications team:
imperial.communications@nhs.net

Wi-fi

Wi-fi is available at our Trust. For more information visit our website: www.imperial.nhs.uk