

The National Centre for Adolescent and Adult Females with Congenital Abnormalities of the Genital Tract

MRKH

Information for patients, relatives and carers

Introduction

MRKH (Mayer Rokitansky Küster Hauser) syndrome is a congenital (born with) abnormality, characterised by the absence of the vagina, cervix and the uterus (womb), which affects one in every 5,000 women. It is also associated with kidney, bone and hearing difficulties. The ovaries are usually present and function in the same way as any other woman's by producing eggs and female hormones that keep you healthy.

How will I know if I have this condition?

Women usually discover that they have no vagina 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 vagina and uterus mean that there is no period. Most women seek advice about this when they are 15 or 16 years old. However, some women may have found out because they had difficulties with sexual intercourse.

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 have referred you for this. The doctor may refer you for further investigations, which may include:

- 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 vagina, uterus (womb) and cervix and the presence of ovaries

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

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

Will I need to have cervical smear tests?

No. As you do not have a uterus or cervix, you will not need to have any cervical smear tests or HPV1 vaccinations (the vaccine that may be offered to prevent human papillomavirus).

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 – 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. About 95 percent of our patients are successful in creating a normal-sized vagina using this technique alone. If you are struggling to complete the dilator treatment, it may be advisable to discuss this with the specialist team. Please note, that when you have fully stretched your vagina to a normal functional length, you would have completed the treatment. You can then stop using your dilators altogether. A small minority of women might be offered surgery if they had vaginal surgery before and their vaginas have closed up due to adhesions (scarring). Our specialist consultant(s) will be able to discuss with you whether this is appropriate.

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 vagina, cervix and 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. However, 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 their 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 a diagnosis of 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 percent 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 support and encouragement for you, your parents and your partner. The group meets twice a year, in 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 9.00am to 5.00pm, 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 nuala.dixon@imperial.nhs.uk 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 treatment?

We aim to provide the best possible service and staff will be happy to answer any questions you may have. However, if your experience of our services does not meet your expectations and you would like to speak to someone other than staff caring for you, please contact the **patient advice and liaison service (PALS)** on **020 3313 3322** or **020 3312 7777** (St Mary's Hospital). You can also email PALS at pals@imperial.nhs.uk.

The PALS team will listen to your concerns, suggestions or queries and is often able to help sort out problems on behalf of patients.

Alternatively, you may wish to express your concerns in writing to:

The chief executive
Imperial College Healthcare NHS Trust
Trust Headquarters
The Bays, South Wharf Road
London, W2 1NY

Alternative formats

This information can be provided on request in large print, as a sound recording, in Braille, or in alternative languages. Please contact the communications team on **020 3312 5592**.