

Rokitansky Syndrome Information Leaflet

Introduction

The full name for Rokitansky syndrome is Mayer-Rokitansky-Kuster-Hauser (MRKH) syndrome, named after the doctors who first recognised it. Girls/women with Rokitansky syndrome are born without a functioning uterus (womb). Sometimes, there may be one/two small uteruses called uterine remnants/horns which normally just consist of muscle. The vagina may be of normal length or it may be short or absent. The ovaries are normal, although they are often sitting slightly higher up in the pelvis. The rest of puberty including breast development occurs normally. Girls/women with Rokitansky syndrome are genetically female (46XX karyotype).

In a small number of women, Rokitansky syndrome can affect development of the kidneys or part of the spine (backbone). This is why all women diagnosed with Rokitansky syndrome are advised to also have a scan of the kidneys.

Rokitansky syndrome affects approximately 1 in 4500 to 1 in 5000 girls/women. The cause of Rokitansky syndrome is not yet known. Rokitansky syndrome very rarely runs in families, so female relatives are usually no more likely to be born with the condition compared to anyone else.

How is it diagnosed?

Rokitansky is usually diagnosed when a girl is referred to a Gynaecologist because her periods haven't started, but the rest of puberty has happened as normal. Several tests have to be done to confirm the diagnosis. These include blood tests (including a genetic test), an ultrasound scan of the pelvis, and possibly an examination of the vagina. Occasionally if the ultrasound is inconclusive or if there are any other problems such as pain, then an MRI scan may be required.

What treatments are available?

At the moment, there are no treatments as yet to grow or develop the uterus. For some girls/women, regular sexual intercourse stretches the vagina so that no other treatment is needed. For other girls/women, the vagina has to be lengthened to allow sexual intercourse to take place. This can be done either by vaginal dilation or vaginal surgery.

Vaginal dilators

Vaginal dilators are plastic moulds that are inserted in the vagina with some pressure. The dilators are inserted and pressure applied for 20 to 30 minutes a day. Gradually wider dilators are used to both lengthen and widen the vagina. Regular use is important for a good result, and treatment can take 3 months or sometimes longer. These are very effective in making the vagina comfortable for intercourse.

We have 2 Consultant Nurses who are available to help you get started on a dilation programme and offer you advice if you encounter any difficulties. You will be given a booklet with top tips for vaginal dilation and this will help you when you are dilating on your own at home.

In the long-term, for most women, regular intercourse helps to maintain vaginal length. For women who are not having regular intercourse, it is usually advisable to continue using dilators once or twice a week. Vaginal dilation is safe and normally used in the first instance.

Vaginal surgery

If vaginal dilators do not work, there are several surgical techniques which can be used. The most commonly recommended operation for girls/women with Rokitansky syndrome is the Vecchietti procedure. The Vecchietti Procedure 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. It works in a similar way to vaginal dilators, but over a shorter period of time. You will usually spend about a week in Hospital. Women will need to use vaginal dilators for some time after having the procedure to widen the vagina and to maintain the vaginal length. Please see our leaflet on Laparoscopic Vecchietti for more information.

Will I be able to have comfortable sex?

Some women with Rokitansky syndrome can have comfortable sex without needing to dilate the vagina first. Others will need to dilate the vagina first to be able to have full penetrative sexual intercourse in the vagina. Some women dilate the vagina with sex, and others use dilators or have surgery as above. Once the vagina has been lengthened you should be able to have comfortable sex.

Will I be able to have children?

Women with Rokitansky syndrome do not have a uterus (womb) that is able to carry a pregnancy. They can have a baby through surrogacy, where another woman with a uterus (womb) can carry the pregnancy for them. They can have their own eggs fertilised with their partner's or donor sperm and have the pregnancy implanted into the womb of another woman (surrogate) by the process of in vitro fertilisation (IVF). We have a Fertility Centre at Liverpool Women's Hospital and we can refer you to have a discussion with a Fertility Specialist if you want more information.

Surrogacy is not the only route to parenthood and many women prefer to adopt and have done so successfully.

There are several uterine transplant trials on going around the world. This may be a possibility in the future, but at the moment it is not an option for women routinely.

Do I have to have smear tests?

No. As you do not have a cervix, you do not have to have a smear test. You might still get the letter inviting you for a smear, but you can let your GP know that this is not required.

Do I still need protection when having sex?

You do not need contraception, but we do still recommend that you use condoms to prevent sexually transmitted infections.

Psychological support

It can be hard to come to terms with various aspects of Rokitansky syndrome, which may not be easy to talk about with other people. Clinical Psychologists can assist girls/women if they experience difficulties in managing the implications of living with Rokitansky syndrome. They provide essential support in making decisions about starting dilation therapy and they can also support you through the dilation treatment programme or surgery, alongside the Consultant Nurses. It can also be very helpful to make contact with other women with Rokitansky syndrome. There are also several support groups on the internet which can be very helpful.

Contact details

Consultant Nurse

Telephone: 0151 708 9988 ext 1053

Support group & web sites:

MRKH support group: www.livingmrkh.org.uk

Surrogacy websites: www.surrogacy.org.uk, www.brilliantbeginnings.co.uk

Adoption and Fostering Academy: www.corambaaf.org.uk

'Top Ten Tips for Dilation': www.dsdfamilies.org

British Society of Paediatric and Adolescent Gynaecologists dilation leaflet: <https://britspag.org/wp-content/uploads/2018/10/Leaflet-Vaginal-dilation.pdf>

This leaflet can be made available in difference formats on request. If you would like to make any suggestions or comments about the content of this leaflet, then please contact the Patient Experience Team on 0151 702 4353 or by email at pals@lwh.nhs.uk

Liverpool Women's NHS Foundation Trust
Crown Street
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Tel: 0151 708 9988
Reference: Gyn/2020-269-v1
Issue Date: 13/02/20
Review Date:13/02/2023

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