

For those who have a sarcoma of the genital tract



Causes,
symptoms,
treatment
and research

Each year, 1700 Norwegian women are diagnosed with a form of gynaecological cancer. Today, around 22,000 women in Norway are living with a form of gynaecological cancer or have undergone treatment for this. Sarcoma of the genital tract is a rare form of gynaecological cancer. What are the symptoms and how are they treated?

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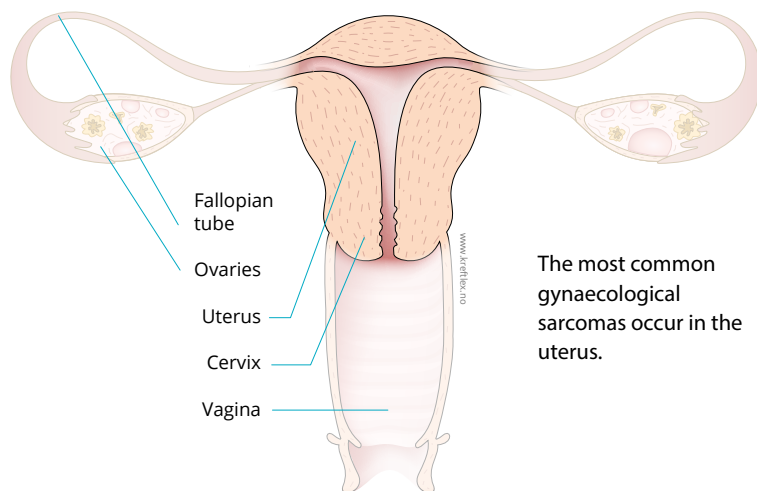


30

women in Norway are
diagnosed with sarcoma
of the genital tract each year

Sarcomas of the female genital tract comprise approximately one percent of all gynaecological cancers. The cancer may have started in the labia and ovaries, but in the vast majority of cases, the cancer starts in the vagina. The cancer usually arises in the wall of the uterus, which consists of muscle, but it can also occur along the inner surface of the uterus (endometrium).

Sarcoma of the female genital tract is more likely to occur after menopause. Around 30 new patients are diagnosed each year.



Vulval cancer (cancer of the female external genitals), rare ovarian tumours and placental cancer are also rare forms of gynaecological cancers. The Norwegian Gynaecological Cancer Society has created separate brochures for these forms of cancer.

Symptoms

- Pain in the lower abdomen
- Pressure sensation in the lower abdomen
- Irregular bleeding

If symptoms persist, you should not wait to contact your doctor. You can also ask to be referred to a gynaecologist. Women who experience bleeding after menopause must always contact a doctor and should always be examined for cancer.

Causes and prevention

Little is known about the cause of this type of cancer, and there is no evidence that it is hereditary. Nor is there any evidence that common fibroids develop into sarcomas.

Assessment

Harmless fibroids may be difficult to distinguish from sarcoma. In general, fibroids do not grow after menopause. If this happens, if this happens sarcoma should be suspected unless the patient is taking hormones. General practitioners should always keep this diagnosis in mind. It is common to take a pelvic MRI and CT scan of the chest and abdomen upon suspicion of sarcoma in the genital tract. Biopsies of the tumour and samples from the uterine lining is a standard part of the assessment, in addition to an ultrasound, if possible.

Treatment

If a sarcoma in the genital tract is localised and considered resectable (meaning that the cancer has not infiltrated surrounding structure and can be surgically removed), surgery is always the primary treatment. In this procedure, both the uterus and ovaries are removed. This type of cancer rarely spreads to the lymph nodes, so these are not routinely removed. There are different types of sarcomas that originate in the uterus. In women who have not reached menopause, surgeons will assess whether the ovaries should be removed. This will always be discussed with the patient.

If all the cancerous tissue is removed during surgery, neither chemotherapy nor anti-oestrogen therapy will be necessary. Radiation therapy is rarely needed.

If surgery is not possible, or the patient has a relapse, they will be considered for chemotherapy or anti-oestrogen therapy, depending on the type of sarcoma that has been diagnosed. Should there be a recurrence of the sarcoma, doctors will always consider a new surgical procedure. Other forms of treatment for recurrence may be considered, such as radiation therapy or some type of drug therapy.

Treatment of sarcomas of the genital tract is centralised to the centres for gynaecological cancer at one of the four university hospitals in Norway, and managed by multidisciplinary teams.

Check-ups

Follow-up after completion of treatment must be adapted to the individual, depending on the risk of recurrence, age and general condition. It is the doctor who is responsible for treatment at the hospital and who will outline a plan for follow-up and check-ups afterwards.

Patients who are treated for sarcoma of the female genital tract have check-ups every 6 months for 5 years, and then every year for the next 5 years. A CT scan will be taken before each check-up.

Late effects of treatment

Both the body's cells and organ functions may be affected by cancer treatment. Side effects and late effects will vary from person to person, depending on the type of treatment, your age and general condition, and on any other illnesses you may have. Below is an overview of late effects that may arise from treatment for gynaecological cancer.

Early menopause

Surgery that involves the removal of the ovaries will trigger menopause in women who have not yet reached menopause. This is a process that normally occurs over a long period, where the woman gradually loses hormones produced in the ovaries.

- Less oestrogen results in less moisture in the mucous membranes, which may make sexual intercourse painful or uncomfortable.
- It is common to experience hot flashes, dry and sore mucous membranes in the vagina, and mood swings. You may also experience fatigue, sadness, depression, poor appetite and hair loss.
- If your body is no longer producing testosterone, this may affect your sex drive. Many will notice reduced libido and general interest in being sexually active. You may experience fewer sexual dreams and lack of interest in sex, but will still feel the need for intimacy.
- It is important to get the right hormonal treatment, adapted to your individual needs.

Nerve damage (polyneuropathy)

Chemotherapy may in some cases cause minor damage to the nerves, especially in the fingers and feet. These symptoms may appear gradually during treatment, but will often go away once treatment is concluded. Sometimes these symptoms persist or become chronic. Such nerve damage is often described as numbness in the fingers and under the feet, or a stinging, burning sensation.

Fatigue

Fatigue is a frequent side effect of radiation therapy or chemotherapy. Around 10 to 35 percent of cancer patients experience fatigue. This is a feeling of exhaustion that does not improve with sleep or rest. There is no quick and effective cure for this. Many patients find that this gradually subsides after a shorter or longer period.

Fatigue that lasts more than six months after the end of treatment, when there are no longer signs of active disease, is called chronic fatigue. Those who experience fatigue may also feel depressed, have trouble concentrating, have trouble with short-term memory, and will generally have little energy.

Fatigue that arises within a limited period of time and subsides when treatment is finished is called acute fatigue.

Rehabilitation

Municipal health services are responsible for providing rehabilitation where you live. Most municipalities offer multidisciplinary services, with an occupational therapist, physiotherapist, nurse and social worker. Should you need assistive devices or adaptations to your home, the municipality can help you with this.

Although municipalities and health trusts are still developing local and regional rehabilitation services, many patients have found it helpful to participate in a rehabilitation programme with other cancer patients spanning several weeks. Such programmes provide a good atmosphere and group dynamics.

By participating in a rehabilitation programme after cancer treatment, you can meet others who are in a similar situation. These programmes also offer better insight and tools to help you adjust to your “new life”. Most patients find that life is not quite the same as before. Rehabilitation programmes are also for patients who had cancer many years ago, and who are still struggling with the long-term effects of treatment.

At www.helsenorge.no, you can find more information about rehabilitation services and how to apply. Search for “rehabilitering kreft” (“rehabilitation cancer”) and your region or health trust.

Many municipalities have local cancer coordinators who have an overview of the services and opportunities in your local area. Here you can find an overview of municipal cancer coordinators:

www.kreftforeningen.no/tilbud/kreftkoordinator-i-kommunen/

See our website for an overview of rehabilitation services
www.gynkreftforeningen.no/2022/01/rehabiliteringstilbud/

Patient care pathway

A standard patient care pathway describes how assessment, treatment, communication and dialogue with the patient and family members, distribution of responsibilities, and specific trajectory schedules are all organised. The purpose of a patient care pathway is to ensure that cancer patients receive a well-organised, comprehensive and predictable trajectory without unnecessary delays in assessment, diagnostics, treatment and rehabilitation. There is no specific patient care pathway for rare gynaecological cancers, but patients with suspected cancer will be referred for an assessment by specialist health services.

Learn more about patient care pathways at www.helsedirektoratet.no





CURRENT RESEARCH

There is unfortunately little to report on the research front for this diagnosis. Senior consultant Tone Skeie-Jensen, at the Dept. of Gynaecological Oncology, Oslo University Hospital, says that many patients ask for immunotherapy. She explains, however, that there are currently no research results to indicate that this is effective. Skeie-Jensen and her colleagues would like to include patients in studies, but there are no relevant studies in Norway at this time.

CLINICAL TRIALS AND APPROVAL

Clinical trials

Clinical trials must always be conducted before a new drug or treatment method can be approved for use. In these trials, drugs are tested on patients with the disease in question. Participation in a clinical trial is not a right, and it is always voluntary. Participants in clinical trials are given the opportunity to test new medicines, which will contribute to better knowledge and research progress. Generally, the physician responsible for the patient's treatment will have an overview of relevant clinical trials, and can therefore request that the patient is considered for participation in the trial at the hospital conducting the study.

Occasionally, a patient will come across a clinical trial. Patients can also contact the physician in charge of the study directly. Patients participating in a clinical trial must always fit the criteria set by the researchers for the study, such as the appropriate age, diagnosis and prior treatment.

An updated overview of current clinical trials in Norway can be found at helsenorge.no, or on the websites of various university hospitals.

You can also check the website of the Norwegian national centre of competence for gynaecological oncology:

www.oslo-universitetssykehus.no/fag-og-forskning/nasjonale-og-regionale-tjenester/nasjonal-kompetansetjeneste-for-gynekologisk-onkologi

Impress-Norway

IMPRESS-Norway is a large Norwegian study open to all patients with advanced cancer who have undergone standard treatment and have no remaining treatment options. IMPRESS was initiated in early 2021. Its aim is to offer extended molecular diagnostics and potential targeted treatment for several Norwegian cancer patients. This is done by taking drugs that are already approved for certain cancer diagnoses and applying these to other types of cancer, based on genetic changes in the cancer cells (molecular profile). Patients who are referred to these clinical trials undergo a screening process, where their cancer cells are examined for more than 500 genes to determine molecular or genetic alterations. If genetic alterations are identified that would have a consequence for treatment recommendations, this is discussed at a national meeting for the research group, held weekly. If the patient is eligible for another ongoing clinical trial in Norway, they will be referred to this study. If a molecular profile is identified that is suitable for a drug through the IMPRESS study, the patient may be assessed for inclusion in an IMPRESS clinical trial. A separate treatment arm will then be created for this specific combination of diagnosis, genetic alteration and drug therapy.

Expert Panel

In 2018, the Norwegian regional health authorities established an Expert Panel scheme. The aim of the Expert Panel scheme is to provide patients who have a life-shortening disease with a new and thorough assessment of treatment options, after established treatment has been attempted and is no longer effective. One important aspect of the Expert Panel is to help patients and their family members feel secure in knowing that all relevant treatment has been considered. The physician in charge of the patient's treatment can request a new assessment by the Expert Panel.

The Expert Panel will assess and advise on the following:

1. Assess whether adequate established treatment has been provided, or if further established treatment is appropriate, either in Norway or abroad.
2. Assess and advise on whether there are relevant clinical trials or experimental treatment in Norway or abroad, preferably in the Nordic region. Experimental treatment must be within approved protocols with criteria for participation and documented effect.
3. Assess and possibly advise on off-label treatment with drugs that have a documented effect. Off-label refers to marketed drugs that are used to treat diseases for which the drugs have not been approved.
4. Assess and possibly advise on undocumented treatment that the patient has obtained information about and wishes to have assessed.

Approval of new medicines

In Europe, a medicine is first approved by the European Medicines Agency (EMA), which grants European marketing authorisation for the medicine. The medicine must then be granted Norwegian marketing authorisation (MT) by the Norwegian Medicines Agency (SLV). In order for an approved drug to receive public funding as a "blue prescription", or for use in hospitals, it must be value assessed. This process can take time, and it is not always easy to gain a good overview of the process. Medicines that are to be financed by hospitals must be sent for assessment by the Norwegian Decision Forum for new health technologies with specialist health services in Norway. This is governed by the four regional health authorities. This Decision Forum is comprised of the directors of the four regional health authorities. It is these four individuals who decide which methods specialist health services can or cannot use. Once the Decision Forum has approved a drug, it can be used by the hospitals. This process takes time. Figures from 2018 show that from the time a drug has been granted marketing authorisation in Norway, it takes an average of 333 days until it can be introduced for use. The National System for Managed Introduction of New Health Technologies is currently under evaluation, partly due to criticism of its use of time.

Ragnhild was diagnosed with a uterine sarcoma

Ragnhild Tombre Bjørnebekk (age 76) was diagnosed with a uterine sarcoma 30 years ago. At that time, she was told that only 10 percent of patients lived longer than a year. Luckily, she met a doctor who had a different opinion, and who believed this was a disease with far more individual outcomes.

“I remember being told I had a sarcoma over the phone one Saturday. My husband was away, but my children were home, and my son said “Just remember that you have me!”. Hearing something like that means so much in a situation like this. I was able to mobilise energy quickly. Understanding the seriousness of the situation, while also motivating yourself is crucial,” says Ragnhild.

She underwent surgery already on the following Tuesday.

Trust yourself!

Prior to the diagnosis, Ragnhild had experienced minor bleeding and felt that something was just not right in her body. She had seen her doctor several times, with no results.

“It is essential that we trust our own bodies and take our symptoms seriously. I couldn't accept that my doctor found nothing wrong. I was concerned and decided to see another



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doctor. The doctor I saw at the medical centre of the University of Oslo realised immediately that something was wrong and sent a message to Ullevål

Hospital. I was quickly admitted and they performed a dilation and curettage (D&C).

After the D&C, everything happened quickly. Ragnhild underwent surgery at Aker Hospital the following week. During this procedure, both her uterus and ovaries were removed.

Individual trajectories

After the surgery, she was transferred for further treatment with chemotherapy at Radiumhospitalet. There, she was given information that was difficult take in.

“At Radiumhospitalet, I was told that I had to expect to be paranoid the rest of my life, because this would return. I feel that this was a destructive message, as it takes away all hope!

Ragnhild is thankful that the doctor at Aker Hospital had a different attitude, which was that this type of disease was difficult to predict, as there are individual trajectories.

Ragnhild received chemotherapy for a long time after her surgery. During this time, the doctor at Aker Hospital was supportive.

“When I was given the diagnosis, I was told by my doctor that “what you will be going through now will be hell, but you will and must get through it.” That became my motivation throughout the treatment. This was something I would and had to get through.

Breast cancer and relapse

There was such little research on this type of cancer that Ragnhild was viewed as an interesting case. She was put on experimental oestrogen therapy for a few years, which resulted in breast cancer, as it turned out that she was sensitive to oestrogen. Breast cancer surgery went fine, and she underwent radiation therapy afterwards. She was unable to receive more chemotherapy, as she had already been given a large dose of chemotherapy earlier in her treatment.

What followed was anti-oestrogen therapy, which triggered new metastases in the first type of cancer a year later. And the recurrence of sarcoma became a reality. The surgery she underwent due to the recurrence was comprehensive and difficult. She was also put on a long period of drug therapy for a total of seven years, which eventually led to osteoporosis. Since then, she has also developed diabetes and polymyalgia rheumatica (a rheumatic disorder that causes significant pain), and both are due to the treatment she underwent, or a consequence of the sarcoma diagnosis, although no one knows for certain.

The four responses

She has accepted the fact that she must struggle with late effects.

“Basically, I have been fortunate with the treatment I received and my response to it. When you have cancer, you just have to acknowledge that this is how it is – and what can I do about it? I have

established four responses for dealing with the cancer: Face it, cope with it, mobilise strength, and motivate myself. This has worked very well for me. I have always set short-term goals that can be carried out and that give me a sense of accomplishment. Finding out what brings joy to me and my family has always been important. For instance, we have made sure to experience art, which gives me energy in my daily life.

Finding energy at work

Ragnhild is considered one of the foremost researchers on violence in Norway. Despite a long and difficult disease trajectory, she has managed to remain a significant resource at work. Although the doctors told her she should take sick leave at times, she was never interested in taking 100 percent sick leave. Her job and the energy she gets from working has been crucial during her illness.

“When you're working, you're in a situation where there is no illness. It feels like a free space. You don't have to talk about illness. But it's also vital because you're doing completely normal and everyday things – just like before. Working is also about having a significant role. That what you do has value. This has been key for me,” says Ragnhild.

At the same time, the disease has given her the opportunity to read more, as she hasn't been “out in the field” as much as before.

“So I have had time to become more informed of current international research, which is very interesting to me. I try to view cancer as a turning point. It can also contribute to something positive,” she says.

For Ragnhild, learning more about professional matters related to cancer, such as what type of food is wise to eat, has been important for keeping a forward-looking and constructive perspective.

Looks for the positive in most things

In 2018, Ragnhild developed a form of skin cancer that she views as a minor issue. The previous tough therapies she underwent helped her put her disease into perspective.

“I don't really have a tendency towards depression. I'm able to see the positive in most things. Being in the present is easier when you're ill. You don't think as far ahead as you normally do. Creating meaning in life and having an interest you can continue to develop is of enormous importance.”

She believes that even simple steps can help keep the dark thoughts at bay.

“Focusing on the aesthetic and things that spark joy has great value. At Radiumhospitalet, for instance, I notice that some people focus on being nicely dressed. It gives them a sense of self-esteem, which is important. Participating in pleasant things prevents stress. Learn what is important for you in life and try to facilitate that,” is her advice.

Peer support services

Through our peer support services, those who have or have had cancer, and their family members, have the chance to talk to someone in the same life situation. The principle of peer support is that people who have been ill themselves can share their experiences with others, yet at the same time be a person who understands and provides support outside the healthcare system. We have certified peer support persons all over the country. Most are patients themselves, but some are also family members. All of our peer support persons have a duty of confidentiality.

You can contact our peer support persons directly. See the overview of all peer support persons on our website:

gynkreftforeningen.no/likepersonstjenesten



The content of this brochure has been quality assured by Tone Skeie-Jensen at the Department of Gynaecological Oncology, Oslo University Hospital.

Sources:

www.kreftlex.no/Blotvevsarkom-i-buk-og-bekken

About the Norwegian Gynaecological Cancer Society

The Norwegian Gynaecological Cancer Society is a patient association for women who have or have had gynaecological cancer, women who have been treated for gynaecological precancerous conditions, and women who have been diagnosed with a genetic risk of gynaecological cancer, and their family members. The Norwegian Gynaecological Cancer Society has over 1000 members. We have local chapters and peer support persons all over the country, and our association is run by volunteers – women who have or have had gynaecological cancer. Our main focus is the patient and generating knowledge about what should be improved in healthcare services with regard to treatment, rehabilitation and follow-up.



Join our community – become a member of
the Norwegian Gynaecological Cancer Society:
gynkreftforeningen.no

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