ovacome..
ovarian cancer

Rare ovarian tumours



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Patient Information Forum

Ovacome is the UK's ovarian cancer support charity.
We provide support and information to anyone affected by ovarian cancer. This includes people who have either been diagnosed with the disease or think that they might be at risk, as well as their friends and family, and healthcare professionals.

Our free support services include:

- Telephone, email, text and instant chat support
- A 24-hour online forum
- Regional support and information events
- Online talks, workshops and support groups
- Expert information resources on ovarian cancer, to support with diagnosis, treatment and beyond

We also raise awareness of the signs and symptoms of ovarian cancer, and work alongside clinicians and researchers to ensure that patient voices are heard in the development of new policy and research on ovarian cancer. **For more information, please visit www.ovacome.org.uk.**

Our booklets can be downloaded or you can order printed copies by calling our support line on 0800 008 7054 (Monday - Friday 10am-5pm).



Rare ovarian tumours

Ovarian cancer is not one disease. It includes several different types, less common sub-types and rarer forms of ovarian tumour, and they can behave in different ways. This booklet explains some of the rarer ovarian tumours and the treatments you may be offered.

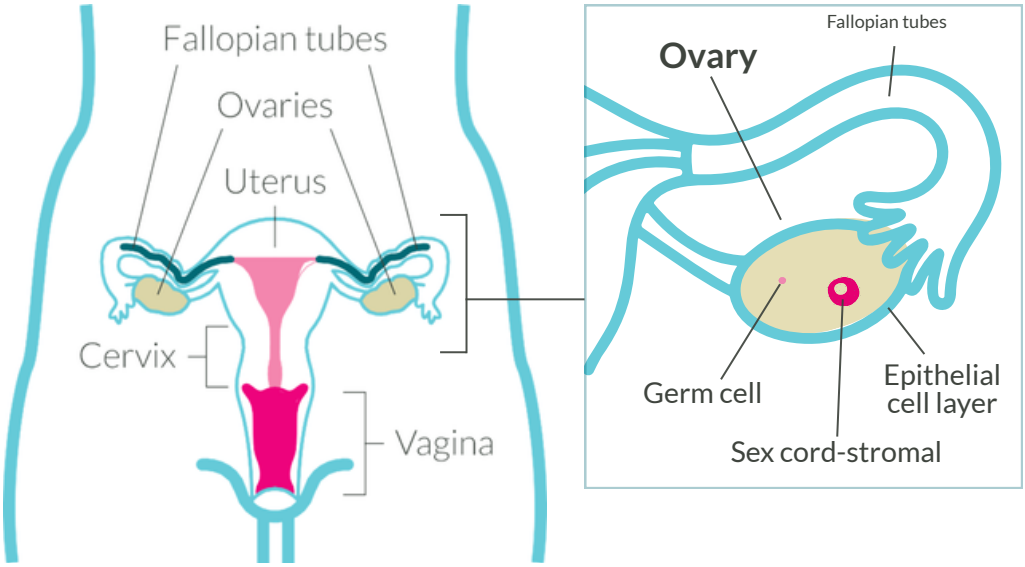
Normal ovaries are made up of:

- epithelial cells, which cover the surface of the ovaries
- germ cells from which ova (eggs) develop
- sex cord-stromal cells inside the ovaries, which provide supportive tissues and are responsible for producing hormones

Tumours can develop from any of these cell types. Because of this, ovarian tumours can vary in how they grow and how they are treated. About 90 per cent (9 in 10) of malignant ovarian tumours develop from the fallopian tubes near the ovaries or from the epithelial cells that cover the surface of the ovaries. These are the most common types of ovarian cancer.

Each year hundreds of people develop one of the rarer ovarian tumours which can be divided into:

- germ cell tumours
- sex cord-stromal tumours
- other rare tumours



Visual of female reproductive organs

The best care and outcome for someone with a rarer tumour depends on careful examination of the ovaries under a microscope and accurate staging of the cancer. Staging means finding out how advanced the cancer is and whether it has spread.

You can find more information on the stages of ovarian cancer here: **ovacome.org.uk/stages-of-ovarian-cancer-booklet**.

If you have a rare ovarian tumour, there may be some uncertainty about the best treatment(s) for you. This is because these tumours affect relatively small numbers of people, so there is often less research available than for more common ovarian cancers.

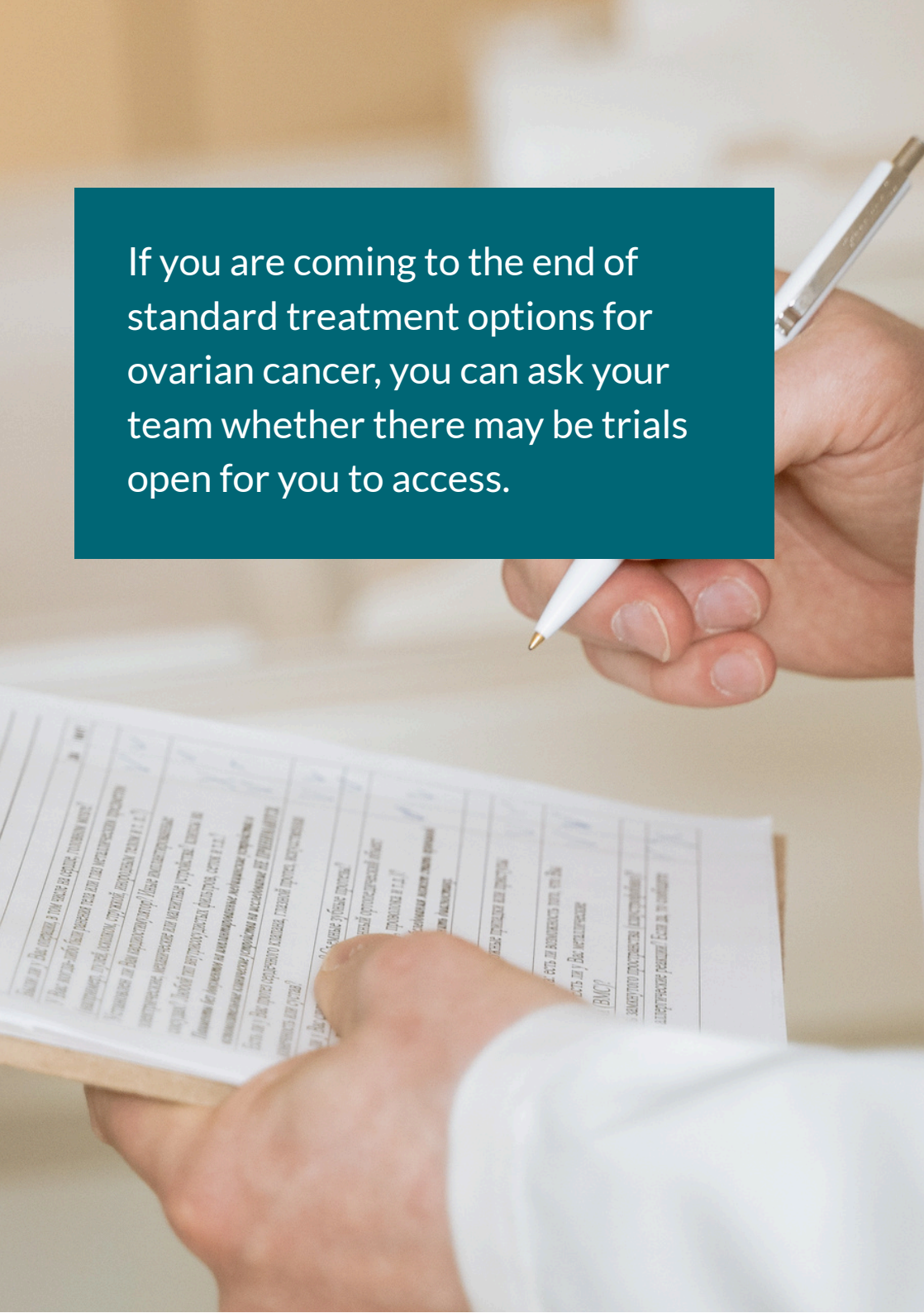
You and your multidisciplinary team (MDT) of specialist surgeons, oncologists and pathologists will make recommendations about your treatment. Sometimes the MDT asks for opinions from other experts. Carefully using tried and tested treatment is often the best option, particularly if your type of tumour is very rare. If you are coming to the end of standard treatment options for ovarian cancer, you can ask your team whether there may be trials open for you to access. There are fewer trials for those diagnosed with rare tumours than for more common types of cancer. However, trials based on molecular or genetic abnormalities are becoming more common and some may be open for people with rare ovarian tumours. You can also contact the Ovacome support team for help in looking for trials.

Ovacome provides more information about some less common types of ovarian cancer in other booklets which can be found here: **ovacome.org.uk/rarer-types-of-ovarian-cancer** and webinars which can be found here: **ovacome.org.uk/rare-ovarian-tumours-webinars**.

Germ cell tumours

These tumours begin in the reproductive cells (egg cells) of the ovary. They tend to appear in teenagers or younger people. The diagnosis is usually made after you develop symptoms similar to all types of ovarian tumour, such as pain in your pelvis and abdomen, a swollen abdomen, difficulty eating and feeling full quickly.

First investigations include a physical examination and an ultrasound or CT scan. If you are younger with a suspected germ cell ovarian tumour, you should have an extra test to measure the hormones beta-hCG and AFP in your blood.



If you are coming to the end of standard treatment options for ovarian cancer, you can ask your team whether there may be trials open for you to access.

If you have very high levels of one or both of these in your blood, this suggests that you might have a germ cell tumour. However, beta hCG can be increased during pregnancy, so please tell your doctor if there is a chance you may be pregnant.

These test results will help your surgeon plan the most appropriate operation. Many ovarian germ cell tumours can be cured, especially when they are found early. Because treatment is often very successful, your surgeon will carefully consider whether one or both ovaries need to be removed. In some cases, ovarian tissue can be preserved, which may help protect your fertility and your chance of having children in the future. After surgery, a pathologist will identify the exact type of germ cell tumour.

These include:

- dysgerminoma
- endodermal sinus tumour
- embryonal carcinoma
- choriocarcinoma
- teratoma

Correct treatment depends on the type and stage of the disease and whether any tumour is left after surgery. Because these tumours are relatively rare, you should be referred to a specialist as soon as possible after diagnosis.

If the tumour was found at an early stage, you may not need chemotherapy. However, if chemotherapy is recommended, a combination of chemotherapy drugs called BEP is commonly used for germ cell tumours (BEP refers to the drugs bleomycin, etoposide, and platinum). This is usually highly effective but requires careful

supervision to reduce side effects. BEP may be given by a specialist in testicular cancer (cancer of the testicle) rather than your ovarian cancer team. This is because testicular cancer and germ cell ovarian cancer are treated with the same chemotherapy, but testicular cancers are more common than ovarian germ cell tumours. Once your treatment is complete, you will usually have regular follow-up appointments arranged with your team.

Ovacome has further information about germ cell tumours which can be found here: **ovacome.org.uk/germ-cell-tumours**.

Sex cord-stromal tumours

The supporting tissues and cells that produce hormones inside the ovary can produce many different tumours. **These include:**

- granulosa cell tumour
- Sertoli-Leydig cell tumour

Granulosa cell tumours

These are the most common kind of sex cord-stromal tumours and tend to appear in middle-age. They sometimes produce the hormone oestrogen, which can cause symptoms such as changes to your periods or bleeding after the menopause. You can find out more about granulosa cell tumours in this booklet: **ovacome.org.uk/granulosa-cell-tumours**.

You will usually have an operation to confirm the diagnosis, find out the stage of the disease and remove as much tumour as possible. If the tumour has spread beyond the ovary, it may also involve removal of the lining of the abdomen, called the peritoneum.

Most patients have a tumour only in the ovary, known as stage 1. In this situation, surgery may be all the treatment you need, and the outlook is usually very good. In some cases, granulosa cell tumours can come back a long time after apparently successful surgery (up to 30 years). If this happens, you may need further surgery to help control the disease. If the disease is more advanced (stages 2, 3 or 4), you may need more treatment after surgery, usually involving drugs. This also applies if your granulosa cell tumour comes back after surgery, and another operation is not likely to help.

If the tumour was found at an advanced stage, the aim will be to try to control the disease. Granulosa cell tumours can grow slowly. In some situations, your team may recommend monitoring the tumour with regular scans before starting treatment. This allows treatment to begin when it is most likely to help, especially if the tumour is not currently causing symptoms.

If you need further treatment, hormonal therapy may be considered before chemotherapy. This can include medicines such as medroxyprogesterone acetate (Provera) or aromatase inhibitors. These treatments reduce the effect of oestrogen in the body, and some granulosa cell tumours may shrink for a time while you are taking them.

Some patients take these drugs for a long time (years) and they may cause side effects. Most commonly, medroxyprogesterone acetate can increase your appetite. Aromatase inhibitors can cause thinning of



Granulosa cell tumours can grow slowly. In some situations, your team may recommend monitoring the tumour with regular scans before starting treatment.

your bones and increase the risk of osteoporosis. Your doctor will monitor your bone density while you are taking the medicine.

If you have an advanced granulosa cell tumour (spread in your abdomen) and need chemotherapy to control it, you may first be offered the standard treatment combination of paclitaxel (Taxol) and carboplatin. Alternative combinations that have been used include BEP, which is used for germ cell tumours (see above), although the bleomycin is often left out.

If you have a granulosa cell tumour, you and your oncologist should discuss the potential benefits and possible side effects of treatment before deciding what to do next. If you have symptoms from specific tumours, radiotherapy can be helpful.

Bevacizumab (also called Avastin) can sometimes be an effective treatment for granulosa cell tumours. Funding options may vary, so your oncologist can explain whether it is available to you. You can also discuss with your oncologist if there are any suitable clinical trials or other treatment options that might be available to you.

Sertoli-Leydig cell tumours

These tumours can produce male hormones like testosterone and may be detected if you develop symptoms such as a lot of body and facial hair, or baldness. Most Sertoli-Leydig cell tumours are not malignant (cancerous) and can be cured by surgery.

However, some are malignant and may need surgery and chemotherapy. The chemotherapy combinations used for granulosa cell tumours are also used for Sertoli- Leydig tumours.

Mucinous tumours

Mucinous tumours are rare and make up one to three per cent (1-3 in 100) of ovarian cancers. You can find more information about mucinous ovarian cancer at **ovacome.org.uk/mucinous-ovarian-cancer**.

Low-grade serous ovarian cancer

Low-grade serous is a form of ovarian cancer that is invasive and grows slowly on surface tissue. You can find more information about low-grade serous ovarian cancer at **ovacome.org.uk/low-grade-serous**.

Clear cell carcinoma

You can find out more information about clear cell ovarian cancer at **ovacome.org.uk/clear-cell-carcinoma**.

Other rare tumours

Some uncommon but important ovarian tumours do not fit into the groups above. As these tumour types are so rare, your doctor and surgeons usually base treatment recommendations on the management guidelines for the more common types of ovarian cancer.

Examples of these rare tumours include:

Small cell carcinoma of the ovary

This tends to appear before the age of 40 and behaves like an aggressive form of the more common epithelial ovarian cancer. If the disease is stage 1 (just in the ovary) some patients will be cured with surgery. But there is a significant risk that the cancer will come back, even at this early stage.

It may be that your team will discuss your treatment with the lung cancer team which treats small cell lung cancer to agree a treatment plan with you.

Brenner tumours

Brenner tumours of the ovary are often discovered by chance. They probably arise from the surface of the ovary. Most are not cancerous, so surgery to remove them is usually the only treatment needed.

Krukenberg tumours

Krukenberg tumours are not primary ovarian cancers. They happen when cancer cells from another organ, usually the stomach but sometimes another part of the bowel, spread to the ovary and grow there. This means that this type of tumour in the ovary is a secondary tumour.

Your doctor can find out where the primary cancer is by reviewing your scans and discussing your case with a pathologist. Under a microscope, Krukenberg tumours look like the tissue they came from. The pathologist can look at the tumour cells and identify where the cancer started.

Occasionally the site of the primary cancer cannot be found. This situation is called carcinoma of unknown primary (CUP). There are specific treatments for CUP and cancer doctors who specialise in this.

Glossary

For help understanding more medical terms, Ovacome has this booklet: **ovacome.org.uk/medical-terms-explained**.

Benign – not cancer - used to refer to tumours which grow slowly in one place and which, once removed by surgery, tend not to come back.

Beta-hCG and AFP – these are hormones. The levels of these hormones can be measured with a blood test.

CT scan – a CT (computerised tomography) scan uses X-rays to produce images of the body.

Malignant – malignant (cancerous) tumours are able to spread to and damage surrounding tissues and other organs in the body.

Pathologist – specialist who examines parts of the body affected by disease.

Radiotherapy – treating cancer using radiation.

Staging – the process for deciding how far the cancer has spread, by using internationally recognised and agreed standards. This can be done using scans, blood tests or surgery.

If you would like to read more about these tumours and how they can be treated, the British Gynaecological Cancer Society Guidelines for treatment (2024) can be found here:
sciencedirect.com/science/article/pii/S0301211524003142

European guidelines for treatment (2018) are available at:
annalsofoncology.org/article/S0923-7534(19)31689-8/fulltext

If you would like to discuss anything about ovarian cancer, please call our support line on 0800 008 7054 (Monday to Friday, 10am - 5pm). You can also visit our website at www.ovacome.org.uk.

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Disclaimer

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Version 3 | Date last reviewed May 2026 | Date for review May 2028

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