ovacome..
ovarian cancer

Rare ovarian tumours

Ovacome is a national charity providing support and information to anyone affected by ovarian cancer.

We give information about symptoms, diagnosis, treatments and research.

We run a free telephone and email support line and work to raise awareness and give a voice to all those affected by ovarian cancer.

This booklet is part of a series giving clinical information about ovarian cancer. It describes rare kinds of ovarian cancer and their treatments.

There is a glossary to explain words you may be not be familiar with.

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Patient Information Forum

Rare ovarian tumours

Ovarian cancer is not one disease. There are many different kinds of ovarian cancer.

Normal ovaries are made up of:
- epithelial cells, covering the surface
- germ cells from which ova (eggs) develop
- sex-cord stromal cells which provide supportive tissues and are responsible for producing hormones.

A range of tumours, which behave differently and have different outcomes, can develop from each of these cell types.

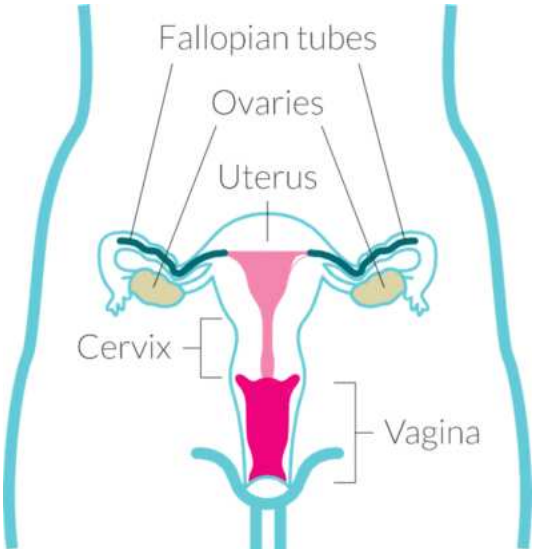
About 90 per cent (90 in 100) of malignant (cancerous) ovarian tumours develop from the fallopian tubes near the ovaries or from the epithelial cells that cover the surface of the ovaries. These are the most common types of ovarian cancer affecting about 7,500 patients each year in the UK. Their behaviour is well understood, and there are reliable guidelines about treatment and results.

There are other subtypes; you can find out more about clear cell ovarian cancer at **ovacome.org.uk/clear-cell-carcinoma-booklet**
Mucinous tumours are rare and make up one to three per cent (one in 100 to three in 100). You can find more information about these at **ovacome.org.uk/mucinous-ovarian-cancer-booklet**

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

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.



A diagram of the female reproductive system

Basic principles

The best care and outcome for someone with a rarer tumour depends on careful microscope examination of the ovaries and accurate staging of the cancer. Staging means seeing how advanced the cancer is and if 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 for you. This is because research into different treatments cannot be carried out on the small numbers of patients who are affected.

You and your multidisciplinary team (MDT) of specialist surgeons, oncologists and pathologists will make decisions 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.

Germ-cell tumours

These tumours begin in the reproductive cells or 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 ovarian tumour, you should have an extra test to measure the hormones beta-hCG and AFP in your blood.

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 tests will help your surgeon choose the most appropriate operation. As most ovarian germ-cell tumours are curable, the surgeon will consider whether both of your ovaries need to be removed. As treatment of germ-cell tumours is so successful, some ovarian tissue can be retained so that you may still be able to have children in the future.

After surgery the pathologist can identify the 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. Sometimes chemotherapy is the preferred option rather than immediate surgery.

A special 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 because side effects are common. Once your treatment is complete, regular follow-up will be organised.

Sex-cord stromal tumours

The supporting tissues and cells that produce hormones in 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 if you have had your menopause.

First you will have an operation to make the diagnosis, confirm the stage of the disease and to remove as much tumour as possible if it has spread beyond the ovary to the lining of the abdomen, called the peritoneum.

Most patients have a tumour just in the ovary (stage 1). In this case you will not need any more treatment after surgery and the outlook is usually very good.

However, in rare cases, granulosa cell tumours come back a long time after apparently successful surgery (up to 30 years). When granulosa cell tumours come back you may need more surgery to control the disease for longer.

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.

The aim is to try and control the disease. As granulosa cell tumours can grow slowly, it is sometimes appropriate to keep an eye on the tumour by having repeated scans.

This means you can wait until the condition is causing you problems before having treatment.

If you need further treatment you may want to try using hormonal therapy with drugs such as medroxyprogesterone acetate (provera) or drugs called aromatase inhibitors, before starting chemotherapy. These stop your body producing oestrogen. Some granulosa cell tumours will shrink for a while if you take these medicines.

Some patients take these drugs for a long time (years) and they can have side effects. Most commonly medroxyprogesterone acetate can increase your appetite. Aromatase inhibitors can cause thinning of your bones (osteoporosis) and therefore 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 decide what treatment you are going to have. You should think about the benefits and the possible side effects. If you have symptoms from specific tumours, radiotherapy can be helpful.

Avastin (bevacizumab) can be an effective treatment for granulosa cell tumour but is not funded by the NHS for this so you may have to obtain it privately. You can discuss with your oncologist if there are any other treatments or trials that would suit 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 the main treatments include surgery and chemotherapy. The chemotherapy combinations used for granulosa cell tumours are also used for sertoli-leydig tumours.

Other rare tumours

There are some uncommon but important ovarian tumours that do not fit into the categories 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 is not clear whether giving chemotherapy immediately after surgery reduces this risk. If you have small cell carcinoma which is advanced or comes back, the benefits of chemotherapy are limited. However, you can have times when you do not have cancer (remission).

You can see more information about ovarian cancer and younger patients here: **www.ovacome.org.uk/younger-people**

- **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 no other treatment is needed except surgery to remove them.

- **Krukenberg tumours**

Krukenberg tumours are not ovarian cancers. They are formed when seedlings of cancer from another organ (usually the stomach, but sometimes other parts of the gut) grow on the ovary. This means the tumour that was found on your ovary is a secondary tumour.

Your doctor can find out where the primary cancer is by looking at your scans and discussing your case with a pathologist.

Each primary cancer causes changes to show in the secondary cancer's cells. So, the pathologist can study the changes and work out where the tumour has come from. Identifying the primary cancer site is important because the best treatment is the standard treatment of the primary cancer, not the secondary one in your ovary.

So, after surgery to remove the affected ovary, people with Krukenburg tumours should receive treatment for the area of the body the cancer first affected.

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

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 tumours are able to spread to and damage surrounding tissues and other organs in the body.

Pathologist
A 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.

We welcome your feedback on this booklet. Please email ovacome@ovacome.org.uk or call 0800 008 7054. If you would like to discuss anything about ovarian cancer, please phone our support line on 0800 008 7054 Monday to Friday between 10am and 5pm. You can also visit our website at www.ovacome.org.uk. This is one of a series of information booklets produced by Ovacome. You can see them here: ovacome.org.uk/information.

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Disclaimer

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