ovacome..
ovarian cancer

Clear cell carcinoma of the ovary

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

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 explains what clear cell carcinoma of the ovary is and how it is treated.

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Clear cell carcinoma of the ovary

What is clear cell carcinoma of the ovary?

Clear cell carcinoma of the ovary, also called clear cell ovarian cancer, is an uncommon type of epithelial ovarian cancer. It accounts for five to 10 per cent (five to 10 in a hundred) of patients with ovarian cancer in the western world.

It is likely that the cause of clear cell is different from the most common type of ovarian cancer, high grade serous epithelial ovarian cancer. It may develop from other ovarian tumours called adenofibromas. These are small, fibrous tumours that can occur on the surface of the ovary.

A family history of clear cell ovarian cancer may also increase the risk as there is an association with Lynch syndrome. This is an inherited genetic condition that increases the risk of several cancers.

Clear cell ovarian cancer may also be more common in people with a history of endometriosis.

Diagnosis

The symptoms of clear cell ovarian cancer are often experienced at an earlier stage than other types of ovarian cancer. Those affected may notice a lump in their abdomen or experience pain or discomfort. They may experience some BEAT symptoms such as abdominal bloating, eating difficulties or changes to bowel and urinary habits. You can learn more about BEAT ovarian cancer symptoms here: https://www.ovacome.org.uk/symptoms-of-ovarian-cancer-booklet

There is also an increased risk of developing deep vein thrombosis and pulmonary embolism (a blood clot in the lungs) which can be the first sign of the cancer. Your medical team will be able to help you manage this risk.

The tumour marker CA125, which is measured in a blood test and used in the diagnosis of ovarian cancer, is not always raised in clear cell cancers so it may be less reliable.

How is it treated?

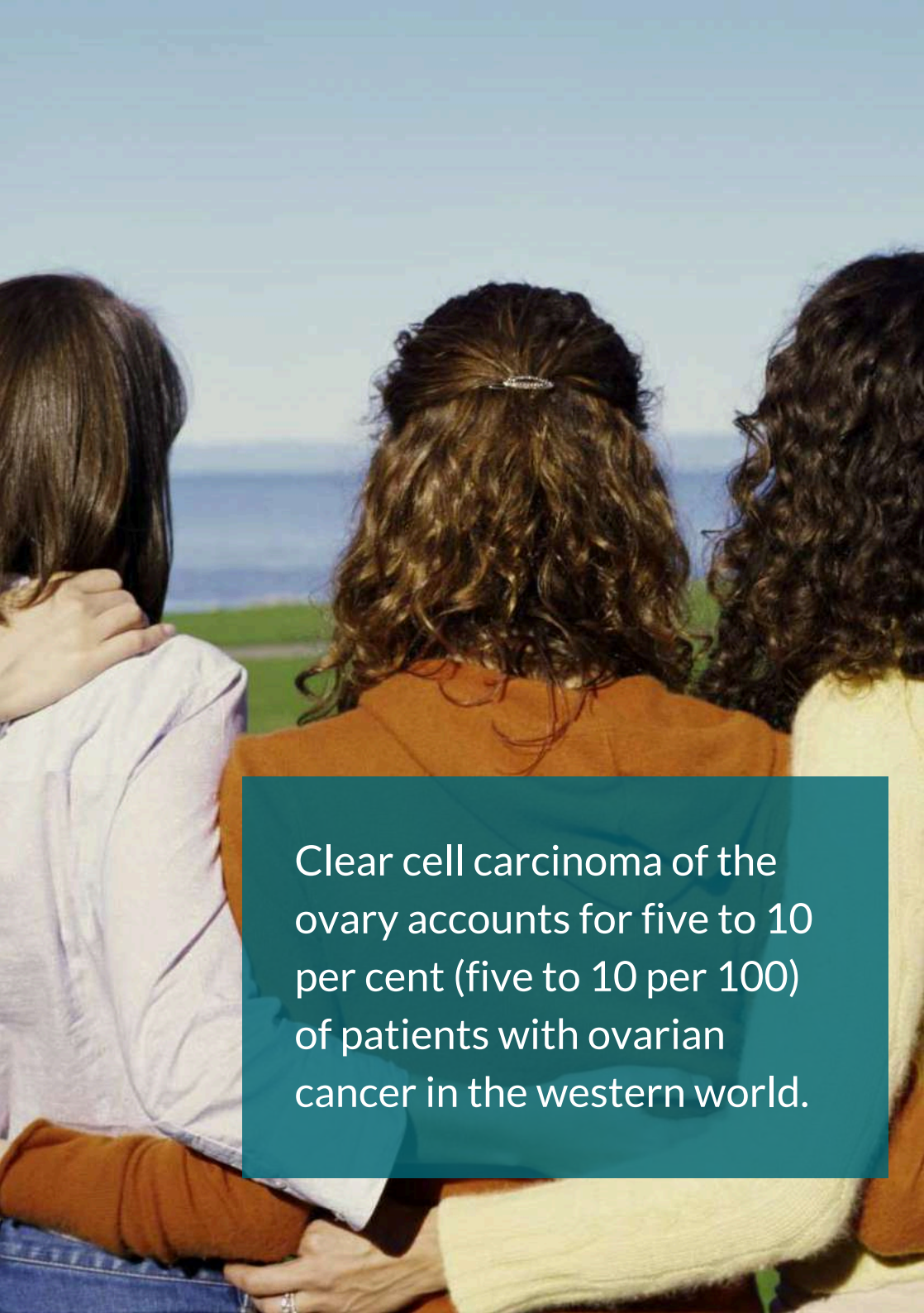
The first treatment is the same as for other more common forms of ovarian cancer.

This means surgery; a total abdominal hysterectomy and bilateral salpingo-oophorectomy and omentectomy (TAHBSO). This is the removal of the womb, cervix, ovaries, fallopian tubes, and omentum, which is a fatty sheet of tissue that covers the abdominal organs. You can learn more about this operation here: https://www.ovacome.org.uk/surgery-for-ovarian-cancer-booklet

The lymph glands in the pelvis and the back of the abdomen may also be removed. These are part of the immune system and carry fluid and white blood cells that help fight infection. They can show the stage of the cancer, meaning how far it has spread.

You can learn more about staging here: https://www.ovacome.org.uk/stages-of-ovarian-cancer-booklet

Fluid from the abdomen will be tested for any cancer cells. A pathologist will examine the tissue and fluid removed to get more information about how advanced the cancer is.



Clear cell carcinoma of the ovary accounts for five to 10 per cent (five to 10 per 100) of patients with ovarian cancer in the western world.

Fertility sparing surgery

If the cancer is at stage 1a, which means it is in one ovary or fallopian tube and has not spread any further, it may be possible to have fertility sparing surgery. This is when the ovary and fallopian tube that are affected are removed, with the womb and other ovary remaining in place.

This means a pregnancy may still be possible if you have not yet experienced menopause.

Chemotherapy

After surgery you may be offered chemotherapy using two drugs, carboplatin and paclitaxel (Taxol).

Where all the visible cancer has been removed, the aim of chemotherapy is to reduce the risk of the cancer coming back. When the tumour is more advanced and cannot be completely removed, the aim is to increase the time until the cancer returns, and further treatment is needed.

Clear cell tumours respond less well to chemotherapy than other ovarian cancers, so the benefit from chemotherapy may be less. Your oncologist will be able to discuss your individual risks and benefits with you, to help you decide about chemotherapy treatment.

What are the side effects of chemotherapy?

If carboplatin and paclitaxel (Taxol) are recommended as your treatment, these will be given as an intravenous infusion (through a drip into a vein) once every three weeks for up to six months.

You may experience side effects from chemotherapy treatment. These might include feeling tired, nausea (feeling sick), hair loss and tingling in your fingertips and toes. Occasionally, you might get pain in your muscles and joints. Your clinical team is there to help with any side effects you may

experience. Once chemotherapy treatment has finished, the side effects will usually lessen over time. You can learn more about chemotherapy here: https://www.ovacome.org.uk/chemotherapy-booklet

What if the cancer comes back?

If your cancer comes back or starts growing again after your initial treatment, the standard treatment for recurrence is to use the same chemotherapy as for other ovarian tumours. This is likely to be carboplatin and paclitaxel (Taxol).

Because clear cell cancers respond less well to chemotherapy, there are research studies looking at different types of drug treatment to see if they are more effective.

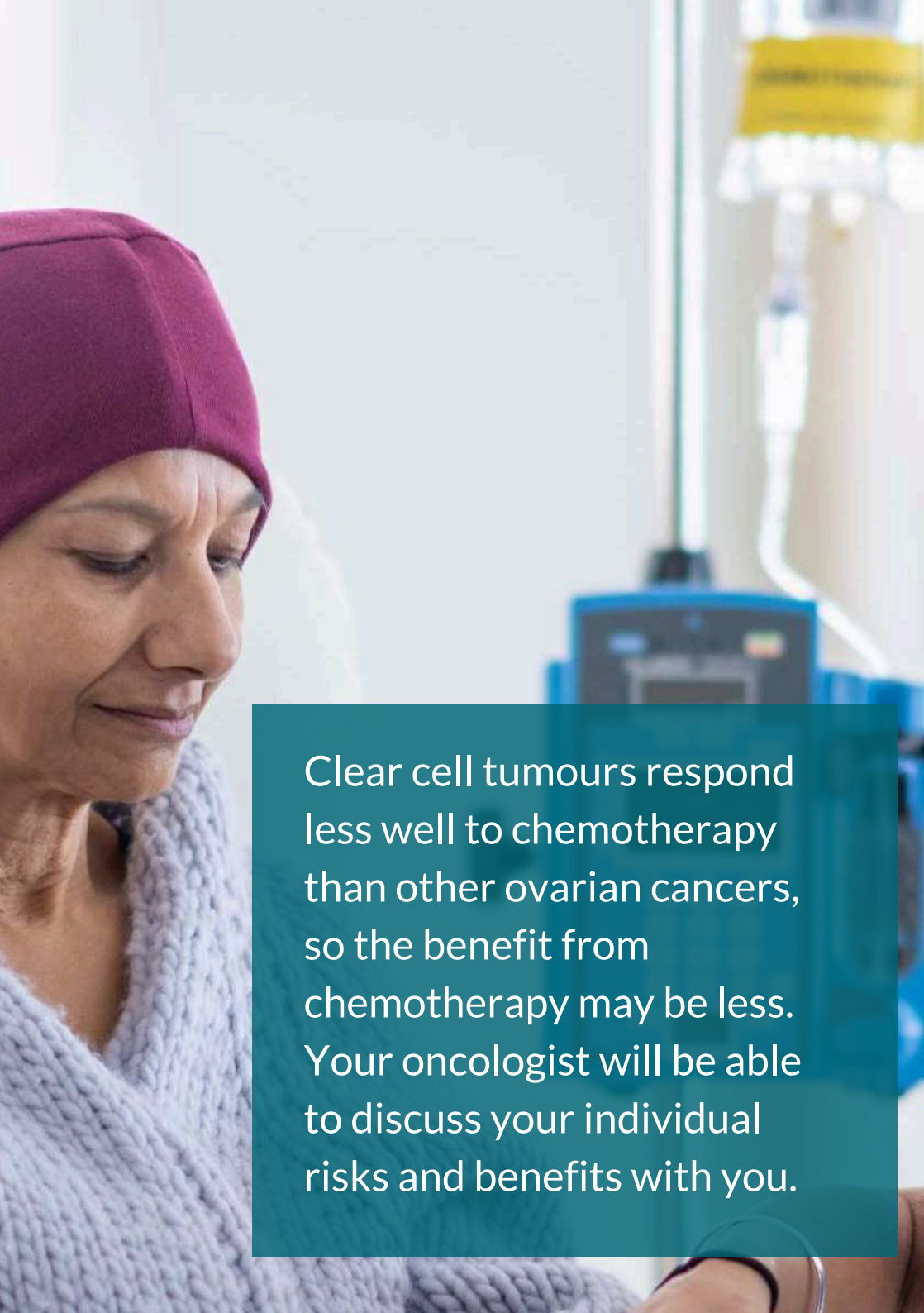
Current research

First line treatment

There is research which suggests that if your clear cell cancer is confined to the ovary (stage IA, IB and IC1) and there has been an assessment of the pelvic and para-aortic lymph nodes to make sure they are not affected, then chemotherapy may not be needed (Management of patients with early stage ovarian clear cell carcinoma 2022).You will need to discuss the risks and benefits for your individual situation with your oncologist.

A trial that studied a different chemotherapy treatment, the combination of cisplatin and irinotecan compared with carboplatin and paclitaxel (Taxol), in the first line setting did not find any advantage. So, carboplatin and paclitaxel (Taxol) remain the standard first line chemotherapy treatment.

The addition of bevacizumab (Avastin) to carboplatin and paclitaxel (Taxol) may prolong the time before the tumour starts to grow again and how long people live in advanced clear cell cancer.



Clear cell tumours respond less well to chemotherapy than other ovarian cancers, so the benefit from chemotherapy may be less. Your oncologist will be able to discuss your individual risks and benefits with you.

Some research has shown that radiotherapy is effective at protecting against recurrence in early stage disease. This has not been accepted into routine clinical practice, but you could discuss it with your consultant.

Treatment of recurrent disease

Unfortunately, chemotherapy does not work well in recurrent clear cell carcinoma, so researchers are looking at other ways to treat clear cell carcinoma if it comes back.

A clinical trial (NiCCC) looked at comparing nintedanib, a drug that targets tumour blood vessels, with chemotherapy when the tumour had recurred or grown within six months of having platinum (carboplatin or cisplatin) chemotherapy. It found that the benefit of chemotherapy was very low. Although nintedanib was a little more effective, the benefit was small and short lasting. The conclusion was not to further investigate nintedanib used alone.

Another study (PEACOCC phase II) trialled immunotherapy drug pembrolizumab. It found that there was some tumour shrinkage in 25 per cent (25 in 100) of patients, and tumours had not grown at 12 weeks in 43 per cent (43 in 100) of patients taking part.

The ATARI study looked at ceralasertib, an ATR inhibitor, and olaparib, a PARP inhibitor, as treatments for rarer gynaecological cancers including clear cell ovarian cancer. In clear cell tumours with a changed ARID1A gene, women received ceralasertib alone. Those without a changed gene received it with olaparib. Researchers saw shrinkage of the tumour in 14 per cent (14 in 100) of women taking part in both groups.

New trials are ongoing, including one looking at the use of an oral immunotherapy with Axitinib, a drug that targets blood vessels. Another trial is studying a new drug that inhibits heat shock transcription factor1 (HSF1) which protects cancer cells from various stresses.

If you are interested in taking part in a trial, discuss this with your oncologist. As clear cell cancers are rare, the trials are only open in a few centres, but you may be able to be referred to one.

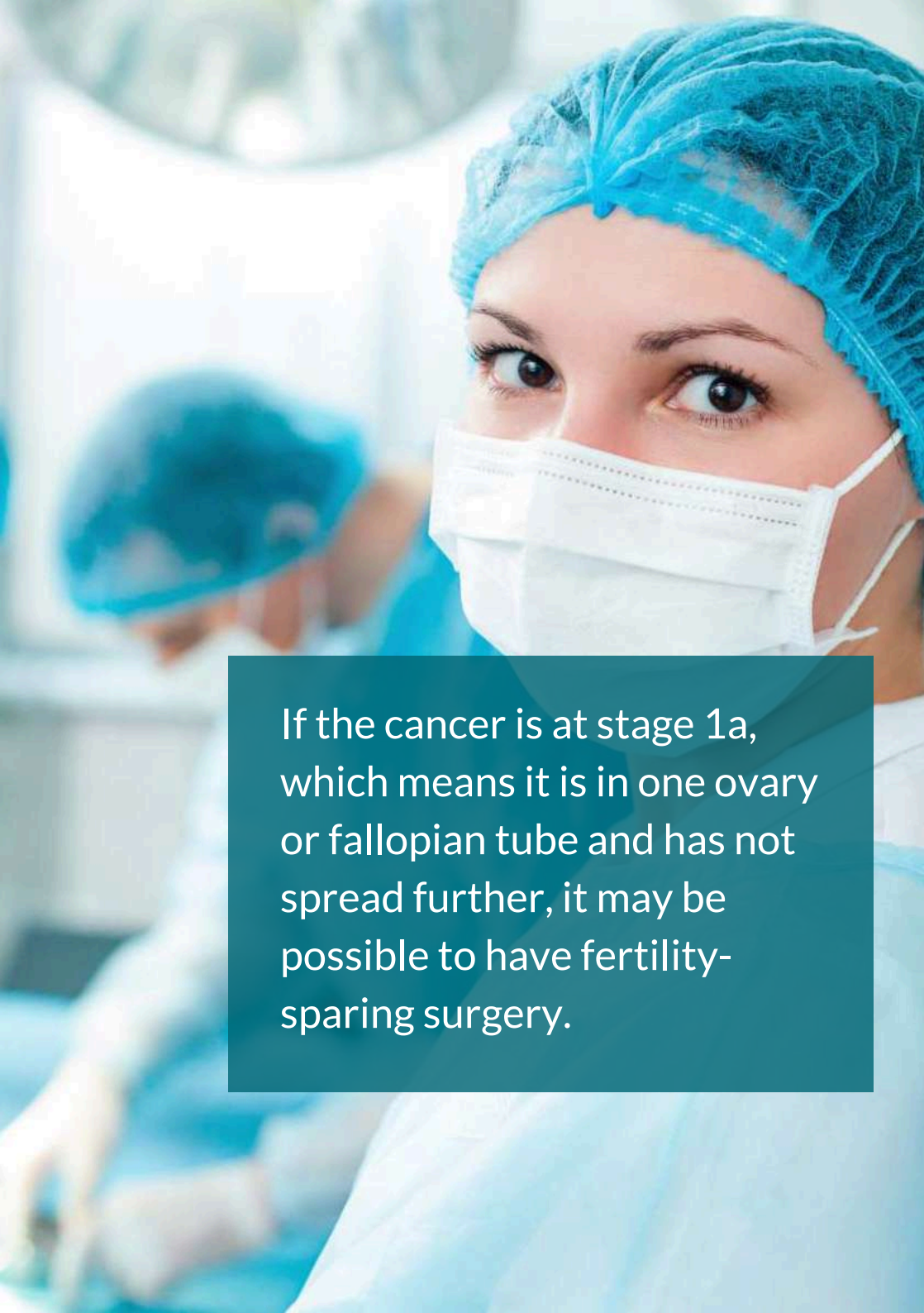
What happens after treatment finishes?

After your treatment, you will need to be carefully followed up by your specialist either in the gynaecology clinic or in the oncology clinic.

Your CA125 level may be checked using a blood test. If it has gone up or if you are experiencing new symptoms, you may be offered a scan. CA125 is not as sensitive to clear cell as it can be to other ovarian cancers, so you may want to discuss further monitoring with your team.

You will usually be seen every three to four months during the first two to three years, and if there are no changes in your health, every six to 12 months after that.

Follow up usually goes on for up to five years after your treatment, but systems vary. Then, if you are feeling well and have no problems it is likely you will have no more follow-up care.



If the cancer is at stage 1a, which means it is in one ovary or fallopian tube and has not spread further, it may be possible to have fertility-sparing 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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