

Carcinoid tumours

This information is about a rare type of cancer called a carcinoid tumour. We hope that it will answer any questions that you may have. If you have any further questions you can ask your doctor or nurse at the hospital where you are having your treatment.

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Carcinoid tumours

A carcinoid tumour, sometimes referred to as simply carcinoid, is a tumour of the neuroendocrine system. The neuroendocrine system is a network of glands that produce particular hormones and send them into the bloodstream to affect the function of different organs in the body.

Most carcinoid tumours are found in the appendix or the small bowel (intestine). Less commonly, they may arise in the lung or the pancreas. Rarely, they may arise in other parts of the body.

Carcinoid tumours often grow slowly and it may be several years before any symptoms appear and the tumour is diagnosed.

Causes of a carcinoid tumour

This is a rare type of tumour. Approximately 1200 new carcinoid tumours are diagnosed each year in the UK. Men and women are affected equally and carcinoid is usually found in adults over the age of 30. As with many other forms of cancer, the exact cause is unknown.

Signs and symptoms

The type of symptoms will depend on where the tumour started, where it may have spread to, and whether it produces a hormone known as serotonin. If a carcinoid tumour starts in the appendix it does not usually spread to other parts of the body and does not produce serotonin. As a result there are very few symptoms and it is often discovered by chance when the appendix is removed for another reason.

Carcinoid syndrome

If a carcinoid tumour starts outside the appendix, and particularly if it spreads to the liver, it will often produce serotonin and cause symptoms known as carcinoid syndrome. Symptoms include:

- diarrhoea
- flushing of the skin
- wheezing (similar to asthma)
- loss of appetite
- weight loss.

It may be possible to reduce the symptoms of carcinoid syndrome by avoiding substances or conditions that cause flushing, such as alcohol and stress.

How it is diagnosed

Before your doctor can make a firm diagnosis of a carcinoid tumour a number of tests will have to be done.

Urine test When serotonin is broken down in the liver, it is excreted as 5HIAA in the urine. A 24-hour urine collection is needed to check whether there are raised levels of 5HIAA.

The doctor may also carry out a number of tests to find out whether the cancer has spread.

X-rays and scans These may be taken to show if there has been any spread of the tumour.

Chest x-ray This will show whether there has been any spread to the lungs.

Ultrasound scan A simple, painless scan which uses sound waves to form a picture of the inside of the abdomen.

CT (computerised tomography) scan A CT scan takes a series of x-rays to build up a three-dimensional picture of the inside of the body. The scan is painless but takes longer than an x-ray (about 30 minutes). It may be used to find where the cancer started (the primary tumour) or to check for any spread of the disease (secondaries or metastases).

MRI (magnetic resonance imaging) scan This scan uses magnetism instead of x-rays to form a series of cross-sectional pictures of the inside of the body. During the scan you will be asked to lie very still on a couch inside a metal cylinder. The test can take up to an hour and is completely painless, although the machine is quite noisy. If you don't like enclosed spaces you may find the machine claustrophobic. It often helps to have a friend in the room with you for company.

Octreotide scan An octreotide scan examines how several of your internal organs are working. Octreotide is a substance that is absorbed by carcinoid cells. It can show where a cancer started and any areas that it has spread to. The scan is done in the nuclear medicine department and takes place over three consecutive days.

On the first day you will go to the department twice. In the morning you will have an injection in your arm. You are then usually free to stay in or leave the hospital. In the afternoon you will have a scan taken using a gamma camera. The scan takes about an hour and during this time you will lie still on a bed. On the following two afternoons pictures will again be taken for up to one and a half hours.

123MIBG scan A drug called 123MIBG may be used to show up the site of a carcinoid tumour. 123MIBG is a mildly radioactive drug that is absorbed by carcinoid cells. The scan takes place over two consecutive days in the nuclear medicine department.

On the first day you will go to the department twice. In the morning you will be given an injection in the arm. You are then usually free to stay or leave the hospital. You will go back to the department in the afternoon to have a scan with a gamma camera lasting for about an hour. The following morning you will have further pictures taken for about one and a quarter hours. At this point you may be given a further injection of 123MIBG.

Biopsy A small sample of cells is taken from the tumour to be examined under a microscope. Depending on which part of the body is affected, a local or general anaesthetic is given.

Treatment

The treatment for a carcinoid tumour depends on a number of factors including your general health and the size and position of the tumour. The results of your tests will enable your doctor to discuss the best type of treatment with you.

Surgery

If the tumour is contained in one area (localised), or if there has been only limited spread (regional), surgery is usually the first choice of treatment. If it is possible to remove the tumour completely no other treatment may be necessary.

If the tumour has spread to other parts of the body (metastatic) but there are only one or two areas – for example, in the liver – surgery may still be possible because of the slow growth of the tumour.

Chemotherapy

This is the use of anti-cancer (cytotoxic) drugs to destroy cancer cells. It may sometimes be used to treat carcinoid tumours that have spread, with the intention of reducing the tumour and the secretion of serotonin, and of prolonging a good quality of life.

Interferon

Another type of drug treatment for the symptoms of carcinoid tumours is interferon, which is a biological therapy. Interferon is given as an injection under the skin. At first it can cause flu-like symptoms such as chills, fever, aching joints and tiredness, but these usually disappear.

Radiotherapy

For this treatment, high-energy rays are used to destroy cancer cells. Radiotherapy is usually given to treat symptoms, such as pain, which may occur if the tumour has spread to the bones.

Hepatic artery embolisation

This treatment may be used to slow down the growth of secondary liver tumours (metastases) by reducing the blood supply to the liver. Sometimes it is combined with chemotherapy. The procedure usually requires an anaesthetic, and you will probably have to stay in hospital overnight. You may feel unwell and your temperature may be raised for a few days afterwards.

¹³¹I MIBG

Another drug, ¹³¹I MIBG, may be used to reduce the number of carcinoid cells and the symptoms of carcinoid syndrome. It gives a dose of radiotherapy to the carcinoid cells, and is known as targeted radiotherapy. This treatment can be given as a drink or as an injection into the vein.

Radio-labelled octreotide

Octreotide is absorbed by carcinoid cells. It can be attached to a small dose of radioactivity. This treatment is given as an injection into a vein in the arm. Again, it can reduce the number of carcinoid cells.

Treatments for carcinoid syndrome

Somatostatin analogues

Carcinoid syndrome can be treated with drugs known as somatostatin analogues. These work by reducing the production of hormones by the tumour, and can help to reduce the flushing and diarrhoea.

The somatostatin analogue octreotide (Sandostatin®) is given as a short-acting injection under the skin up to three times a day. Most people are taught how to give the injection themselves.

Other somatostatin analogues are given as longer-acting injections into the muscle (intramuscular) between 7 and 28 days apart, depending on the drug that is used and the response. Commonly used drugs are octreotide (Sandostatin Lar®) and lanreotide (Somatuline® LA, Somatuline Autogel®).

Clinical trials

Research into treatments for carcinoid tumours is ongoing and advances are being made. Cancer doctors use clinical trials to assess new treatments.

You may be asked to take part in a clinical trial. Your doctor must discuss the treatment with you, so that you have a full understanding of the trial and what it means to take part.

Your feelings

During your diagnosis and treatment you are likely to experience a number of different emotions, from shock and disbelief to fear and anger. At times these emotions can be overwhelming and hard to control. It is quite natural, and important, to be able to express them. Everyone has their own ways of coping with difficult situations; some people find it helpful to talk to friends or family, while others prefer to seek help from people outside their situation. Others prefer to keep their feelings to themselves. There is no right or wrong way to cope, but help is available if you need it.

References

This section has been compiled using information from a number of reliable sources, including:

- *Oxford Textbook of Oncology* (2nd edition). Souhami et al. Oxford University Press, 2002.
- *Gastrointestinal Oncology: Principles and Practice*. Kelsen et al. Lippincott Williams and Wilkins, 2002.
- *The Textbook of Uncommon Cancers* (2nd edition). Raghavan et al. Wiley, 1999.