Carcinoid Lung Tumour

A patient’s guide
What is a carcinoid lung tumour?
Lung carcinoid tumours are unusual tumours that start in the lung. They are rare, accounting for fewer than 5% of all lung cancers. They tend to grow slowly and may not cause symptoms for many years.

Carcinoid tumours are a type of tumour of the neuroendocrine system. This system is made up of special types of nerve and gland cells called neuroendocrine cells. It is responsible for making hormones that are released into the bloodstream.

Neuroendocrine cells are found in the:
- gut, including the food pipe (also called the gullet or oesophagus);
- lungs;
- pancreas.

Neuroendocrine cells in the lung release hormones that control the flow of air and blood in the lungs. Carcinoid tumours develop when a change has occurred in the neuroendocrine cells that cause them to grow too much.

Around 25 in 100 (25%) carcinoid tumours develop in the lung.

Types of carcinoid tumour
There are two types of lung carcinoid:
- Typical carcinoids grow slowly and only rarely spread to other parts of the body;
- Atypical carcinoids are much rarer, tend to grow faster and are slightly more likely to spread to other parts of the body than typical lung carcinoids.

Doctors can only tell the difference by looking at the cells under the microscope. 9 out of 10 (90%) lung carcinoids are typical in nature.

A carcinoid tumour may form in the walls of the large airways (bronchi) near the centre of the lungs, or develop in the smaller airways (bronchioles) towards the outer edges of the lungs and is called peripheral. The tumour’s location may affect which symptoms a patient has and how the carcinoid can be treated.
What are the symptoms of lung carcinoid tumours?
Because carcinoid lung tumours tend to grow slowly they may not cause any symptoms for many years. Sometimes they are found by medical tests carried out for other reasons.

If a carcinoid is present in the large airways symptoms may include:

- cough and sometimes blood in the phlegm or sputum;
- wheezing;
- shortness of breath;
- chest pain.

Lung carcinoid tumours can cause partial or complete blockage of an air passage that can lead to a chest infection.

Carcinoid syndrome is a condition related to over-production of certain hormones by the neuroendocrine cells of the tumour. It is very uncommon to have carcinoid syndrome from a lung tumour.

How are lung carcinoid tumours staged?
The stage of a cancer is a term used to describe its size and whether it has spread beyond its original site. Knowing the stage and position of a cancer helps the doctors decide on the most appropriate treatment.

**Stage 1:** the carcinoid is smaller than 3 cm, limited to one area and there is no sign of cancer in the main airways or evidence of any spread.

**Stage 2A:** the carcinoid is small but has spread to the lymph nodes closest to the affected area.

**Stage 2B:** the carcinoid is bigger than 3cm with carcinoid cells in the lymph nodes closest to the affected area, OR cells have grown into the chest wall, outer covering of the lung (pleura), the diaphragm (muscle at bottom of chest cavity) or outer covering of the heart (pericardium).

**Stage 3A:** either carcinoid cells are present in lymph nodes further away from the affected lung but on the same side, OR there are carcinoid cells in the nearest lymph nodes but there is also spread of the tumour.
into the pleura and chest wall or the middle of the chest.

**Stage 3B:** either carcinoid cells have spread to the lymph nodes on the other side of the chest or above the collarbone OR the tumour has grown into another major structure in the chest (such as the heart, windpipe or blood vessels).

**Stage 4:** the carcinoid has spread to the other lung or another organ of the body.

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**What are the main treatments for lung carcinoid tumours?**

The method of treatment will depend on the type of carcinoid (typical or atypical) and its staging. It may also depend on your overall health. You may not have any treatment straight away if your carcinoid is not causing symptoms or growing. Your doctor will monitor you with regular check-ups and you will be offered treatment if you develop symptoms. This approach is sometimes called “watchful waiting”.

**Surgery for carcinoid tumours**

If you have a growing carcinoid or symptoms, you may be offered surgery. Many lung carcinoid tumours can be treated with surgery alone.

You may have surgery if you:

• have early stage or localised carcinoid in the lung;
• are generally well.

When the aim of this surgery is to remove the carcinoid completely, it is known as curative or radical surgery. If the tumour is peripheral a lobectomy or segmentectomy would be required to remove it completely. For central
carcinoids there are specialised procedures that aim to remove the entire tumour called bronchial sleeve resection or sleeve lobectomy.

Atypical carcinoid tumours may require more extensive surgery during which nearby lymph nodes are removed as well. After surgery to remove an atypical carcinoid tumour, the doctors may recommend further treatment with chemotherapy or radiotherapy.

**Chemotherapy**
Chemotherapy uses drugs to destroy cancer cells. This works by disrupting their growth. Chemotherapy isn’t generally the first choice of treatment for typical carcinoid tumours.

If the carcinoid tumour has grown too much and is atypical in nature or has spread too far to be removed completely, then treatment with chemotherapy or radiotherapy might be considered more appropriate.

**Radiotherapy**
Radiotherapy is treatment with high energy x-rays that are aimed at the tumour to destroy cancer cells, or sometimes to the place from which the tumour was removed surgically.

**What are the other treatments for carcinoid tumours?**
When a carcinoid tumour is present in the airways it is called endobronchial. There are various forms of endobronchial treatment using a bronchoscope. This is a special scope with either a laser or freezing probe fitted to it. This can be used to destroy the tumour cells. It can open up an airway, and can result in excellent local control of the tumour. In some cases this can be curative. The use of a freezing method is known as cryotherapy.
What happens after initial treatment?
Lung carcinoid tumours are often cured by the initial treatment but sometimes they can come back (recur) many years later. If you have completed your treatment, your doctor will still want to monitor you closely. Your doctor will want to see you for follow-up, for example 3-monthly to begin with then 6-monthly and after that yearly up to 5 years. The time between visits may be extended if there are no problems.

It is very important to go to all of your follow-up appointments. During these visits, your doctor will ask about any problems you are having and may do imaging tests such as chest x-rays or CT scans as required.

Recurrent carcinoid tumours
Generally the prognosis of lung carcinoids is very good.

Carcinoid tumours can sometimes come back, even years after initial treatment. If this happens, further treatment options will depend on where the cancer is and what treatment has been used already.

Further information can be obtained on carcinoid tumours from the following sources

NET Patient Foundation
www.netpatientfoundation.org

Cancer Research UK
www.cancerresearchuk.org

Macmillan Cancer Support
www.macmillan.org.uk