About Lung Carcinoid Tumors

Overview and Types

If you have been diagnosed with a lung carcinoid tumor or are worried about it, you likely have a lot of questions. Learning some basics is a good place to start.

- What Are Lung Carcinoid Tumors?

Research and Statistics

See the latest estimates for new cases of lung carcinoid tumors and deaths in the US and what research is currently being done.

- What Are the Key Statistics About Lung Carcinoid Tumors?
- What's New in Lung Carcinoid Tumor Research and Treatment?

What Are Lung Carcinoid Tumors?

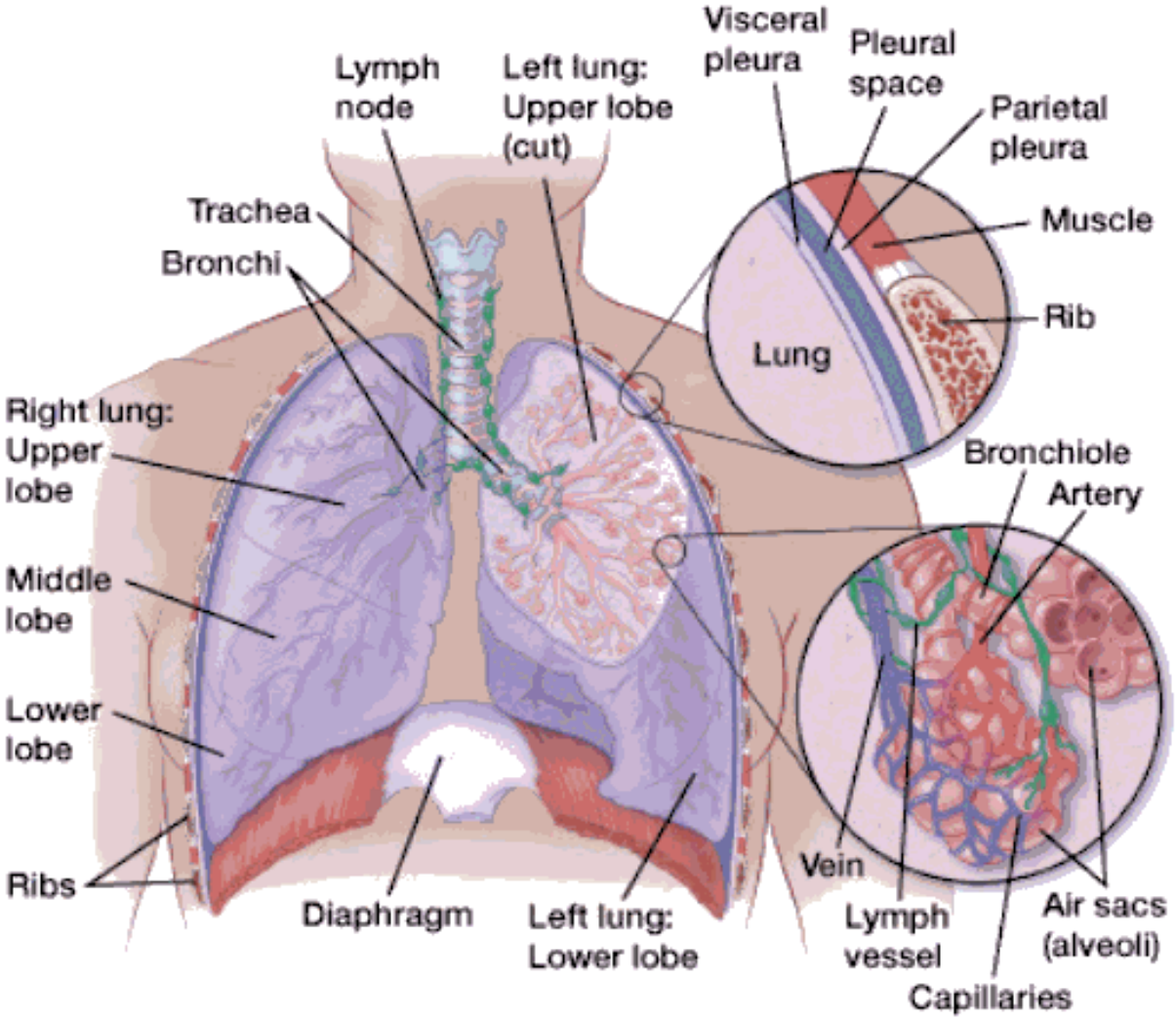
Lung carcinoid tumors (also known as *lung carcinoids*) are a type of lung cancer, which is a cancer that starts in the lungs. Cancer starts when cells begin to grow out of control. Cells in nearly any part of the body can become cancer, and can spread to other areas of the body. To learn more about how cancers start and spread, see What Is Cancer?

Lung carcinoid tumors are uncommon and tend to grow slower than other types of lung cancers. They are made up of special kinds of cells called *neuroendocrine cells*.

To understand lung carcinoid tumors, it helps to know something about the normal structure and function of the lungs, as well as the neuroendocrine system.

The lungs

The lungs are 2 sponge-like organs in your chest. Your right lung has 3 sections, called *lobes*. The left lung has 2 lobes. It is smaller because the heart takes up room on that side of the body.



When you breathe in, air enters through your mouth and nose and goes into your lungs through the *trachea* (windpipe). The trachea divides into tubes called the *bronchi* (singular, *bronchus*), which enter the lungs and divide into smaller branches called the *bronchioles.* At the end of the bronchioles are tiny air sacs known as *alveoli.*

Many tiny blood vessels run through the alveoli. They absorb oxygen from the inhaled air into your bloodstream and pass carbon dioxide (a waste product from the body) into the alveoli. This is expelled from the body when you exhale. Taking in oxygen and getting rid of carbon dioxide are your lungs' main functions.

A thin lining called the *pleura* surrounds the lungs. The pleura protects your lungs and helps them slide back and forth as they expand and contract during breathing. The space inside the chest that contains the lungs is called the *pleural space* (or *pleural cavity*).

Below the lungs, a thin, dome-shaped muscle called the *diaphragm* separates the chest from the abdomen. When you breathe, the diaphragm moves up and down, forcing air in and out of the lungs.

The diffuse neuroendocrine system

Carcinoid tumors start from cells of the diffuse neuroendocrine system. This system is made up of cells that are like nerve cells in certain ways and like hormone-making endocrine cells in other ways. These cells do not form an actual organ like the adrenal or thyroid glands. Instead, they are scattered throughout the body in organs like the lungs, stomach, and intestines.

Neuroendocrine cells make hormones like adrenaline and similar substances. In the lungs, this may help control air flow and blood flow and may help control the growth of other types of lung cells. Neuroendocrine cells may detect the levels of oxygen and carbon dioxide in the air we breathe and then release chemical messages to help the lungs adjust to these changes. People who live at higher altitudes have more lung neuroendocrine cells, probably because there is less oxygen in the air they breathe.

Types of lung neuroendocrine tumors

Like most cells in your body, lung neuroendocrine cells sometimes go through certain changes that cause them to grow too much and form tumors. These are known as *neuroendocrine tumors* or *neuroendocrine cancers*. Carcinoid tumors are one type of neuroendocrine tumor.

Neuroendocrine tumors can develop anywhere in the body. Neuroendocrine tumors that begin in the digestive system, another common site for these tumors, are discussed in Gastrointestinal Carcinoid Tumorsand Pancreatic Cancer.

This document focuses only on carcinoid tumors that start in the lungs.

There are 4 types of neuroendocrine lung tumors. Starting with the fastest growing, they are:

- Small cell lung cancer
- Large cell neuroendocrine carcinoma
- Atypical carcinoid tumor
- Typical carcinoid tumor

Small cell lung cancer

Small cell lung cancer (SCLC) is one of the fastest growing and spreading of all cancers.

Large cell neuroendocrine carcinoma

Large cell neuroendocrine carcinoma (LCNEC) is a rare cancer. It is a subtype of non-small cell lung cancer (NSCLC). Although it shares some features with SCLC (including a tendency to grow quickly), it is typically treated as a type of NSCLC.

Typical and atypical carcinoid tumors

The other 2 types of lung neuroendocrine tumors are carcinoids.

Typical and atypical carcinoid tumors look different under the microscope.

- **Typical carcinoids** tend to grow slowly and only rarely spread beyond the lungs. About 9 out of 10 lung carcinoids are typical carcinoids.
- **Atypical carcinoids** grow a little faster and are somewhat more likely to spread to other organs. They have more cells in the process of dividing and look more like a fast-growing tumor. They are much less common than typical carcinoids.

Carcinoids are sometimes also classified by where they form in the lung.

- **Central carcinoids** form in the walls of large airways (bronchi) near the center of the lungs. Most lung carcinoid tumors are central carcinoids, and nearly all of these are also typical carcinoids.
- **Peripheral carcinoids** develop in the smaller airways (bronchioles) toward the outer edges of the lungs. These are more likely than central carcinoids to be atypical, although most peripheral carcinoids are still typical carcinoids.

This distinction is important because the tumor's location can affect which symptoms a patient has (see Signs and Symptoms of Lung Carcinoid Tumors) and may also affect how the tumor is treated.

- References

See all references for Lung Carcinoid Tumor

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What Are the Key Statistics About Lung Carcinoid Tumors?

About 1% to 2% of all lung cancers are carcinoids. There are about 4,000 newly diagnosed lung carcinoid tumors in the United States each year.

Carcinoid tumors actually develop more often in the digestive tract than in the lungs. Only about 3 out of 10 carcinoid tumors start in the lungs.

Lung carcinoids tend to be diagnosed in people who are slightly younger than the typical age of people with other types of lung cancer. The average age at diagnosis is around 60 years.

Information on survival rates for lung carcinoids can be found in Survival Rates for Lung Carcinoid Tumors.

Visit the American Cancer Society's Cancer Statistics Center for more key statistics.

- References

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What’s New in Lung Carcinoid Tumor Research and Treatment?

Many medical centers around the world are researching the causes and treatment of lung carcinoid tumors. This disease is challenging to study because it is not common. But each year, scientists find out more about what causes the disease and how to improve treatment.

Genetics

Researchers have made great progress in understanding how certain changes in the DNA inside normal cells can cause them to become cancerous. DNA is the molecule that carries the instructions for nearly everything our cells do. We usually look like our parents because they are the source of our DNA. But DNA affects more than how we look.

Some genes (parts of our DNA) contain instructions for controlling when our cells grow and divide into new cells. Certain genes that cause cells to grow, divide, and stay alive are called *oncogenes*. Others that slow down cell division or cause cells to die at the right time are called *tumor suppressor genes*. Cancers can be caused by DNA changes that turn on oncogenes or turn off tumor suppressor genes.

Researchers have found many DNA changes in lung carcinoid cells in the past few years. The hope is that continued research in understanding these changes will lead to new tests for earlier diagnosis and new drugs for more effective treatment.

Diagnosis

Because the outlook and treatment of lung carcinoids and other types of lung cancer are very different, it’s important to be able to tell these cancers apart as soon as possible. Researchers have made great progress in developing tests that can detect specific substances found in the cells of carcinoid tumors but not other lung cancers. Most of these tests treat tissue samples with special man-made antibodies in the lab. The antibodies are designed to recognize specific substances in certain types of tumors.

Treatment

Doctors are learning how to treat lung carcinoids more effectively. For example, newer surgical techniques allow doctors to remove parts of the lung through smaller incisions, which can result in shorter hospital stays and less pain for patients. And new radiation therapy techniques help doctors focus the radiation more precisely on tumors, lowering the amount of radiation that normal tissues get and reducing side effects.

Carcinoid tumors that have spread remain hard to treat. Most carcinoid tumors grow fairly slowly. Because standard chemotherapy drugs work by attacking quickly growing cells, they are not very effective against carcinoid tumors.

Newer drugs called *targeted therapies* may prove to be more effective against carcinoids. Targeted therapies attack the parts of cancer cells that make them different from normal, healthy cells. Each type of targeted therapy works differently, but they alter the way a cancer cell grows, divides, repairs itself, or interacts with other cells in some way.

Two targeted therapy drugs, sunitinib (Sutent®) and everolimus (Afinitor®), have been shown to be helpful in treating neuroendocrine tumors that start in the pancreas. Studies of these drugs in carcinoid tumors (which are a type of neuroendocrine tumor) are in progress.

Targeted drugs called *angiogenesis inhibitors* affect the growth of new blood vessels, which tumors need to grow larger. Some of these drugs are already used to treat other types of cancer and are now being studied for use against carcinoid tumors. Examples of these drugs include bevacizumab (Avastin®), pazopanib (Votrient®), axitinib (Inlyta®), and cabozantinib (Cometriq®).

Researchers are also trying to improve upon drugs related to somatostatin, which help some people with carcinoid tumors. An example is pasireotide (Signifor®), which may prove to be more potent than current drugs such as octreotide and lanreotide. Another new approach is to pair drugs similar to octreotide with a radioactive atom. These drugs are like those used for some radionuclide scans, but the radiation is stronger. The drugs bind to the carcinoid cells, delivering radiation to those cells and limiting the effects on normal cells. In early studies, this type of treatment has helped some patients with advanced carcinoid tumors that were no longer responding to other treatments, but more studies of these new drugs are needed.

These and other new drugs are now being studied in clinical trials.

- References

See all references for Lung Carcinoid Tumor

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For additional assistance please contact your American Cancer Society
1-800-227-2345 or www.cancer.org