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# **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.

- Key Statistics for Lung Carcinoid Tumor
- What's New in Lung Carcinoid Tumor Research?

# What Are Lung Carcinoid Tumors?

Lung carcinoid tumors (also known as *lung carcinoids*) are a type of lung cancer. 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. (To learn more about how cancers start and spread, see <u>What Is Cancer?</u>)<sup>1</sup>

Lung carcinoid tumors are uncommon and tend to grow slower than other types of lung cancers.

## Where lung carcinoid tumors start

Lung carcinoid tumors start in neuroendocrine cells, a special kind of cell found in the lungs. Neuroendocrine cells are also found in other areas of the body, but only cancers that form from neuroendocrine cells in the lungs are called lung carcinoid tumors.

## The neuroendocrine system

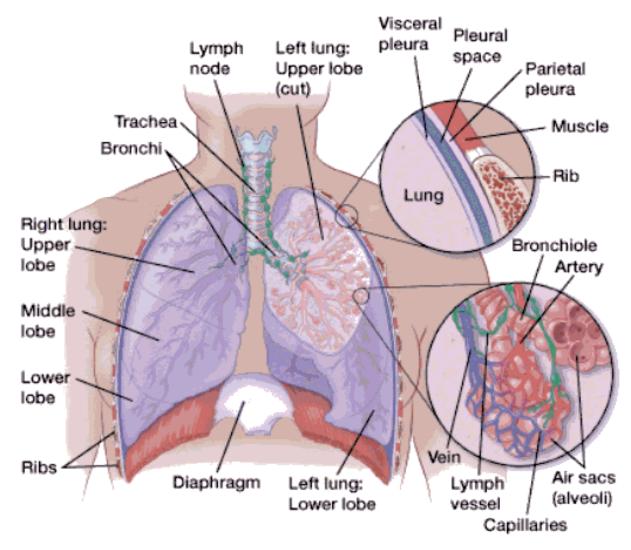
Neuroendocrine cells are like nerve cells in some way, and like hormone-making endocrine cells in other ways. They make hormones like adrenaline and similar substances. Cells in this system don't form actual organs. Instead they are scattered throughout the body in organs like the lungs, stomach, and intestines.

Neuroendocrine cells in the lungs can have several different functions. They may:

- Help control air flow and blood flow in the lungs
- Help control the growth of other types of lung cells
- 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.)

## The lungs

Lung carcinoid tumors start in the lungs – 2 sponge-like organs in your chest. Their main functions are to take in oxygen and get rid of carbon dioxide.



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 where it is expelled from the body when you exhale.

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.

Carcinoid tumors are sometimes classified by where they form in the lung. The tumor's location is important because it can affect which symptoms a patient has and how the tumor is treated.

- **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 are typical carcinoids. (See types of lung carcinoid tumors below.)
- Peripheral carcinoids develop in the smaller airways (bronchioles) toward the outer edges of the lungs. Most peripheral carcinoids are typical carcinoids. (See below.)

## Types of lung carcinoid tumors

Lung carcinoid tumors are a type of neuroendocrine tumor. Neuroendocrine tumors are more common in the digestive system (see <u>Gastrointestinal Carcinoid</u><sup>2</sup><u>Tumors</u><sup>3</sup>and Pancreatic Cancer)<sup>4</sup>, but the second most common place is in the lungs.

There are 2 types of lung carcinoid tumors:

- **Typical carcinoids** tend to grow slowly and rarely spread beyond the lungs. About 9 out of 10 lung carcinoids are typical carcinoids. They also do not seem to be linked with smoking.
- Atypical carcinoids grow a little faster and are somewhat more likely to spread to
  other organs. They have more cells that are dividing and look more like a fastgrowing tumor. They are much less common than typical carcinoids and may be
  found more often in people who smoke.

In addition to lung carcinoid tumors, there are other types of neuroendocrine tumors that start in the lungs: small cell lung cancer and large cell neuroendocrine carcinoma, which is a type of non-small cell lung cancer. These lung cancers are treated differently, so it's important to know exactly what type you have. (See <u>Small Cell Lung Cancer</u><sup>5</sup> and <u>Non-Small Cell Lung Cancer</u><sup>6</sup>.)

# **Hyperlinks**

- 1. www.cancer.org/cancer/cancer-basics/what-is-cancer.html
- 2. www.cancer.org/cancer/gastrointestinal-carcinoid-tumor.html
- 3. www.cancer.org/cancer/gastrointestinal-carcinoid-tumor.html
- 4. /content/cancer/en/cancer/pancreaticcancer/detailedguide/pancreatic-cancer-detailed-guide-toc.html
- 5. www.cancer.org/cancer/small-cell-lung-cancer.html
- 6. www.cancer.org/cancer/non-small-cell-lung-cancer.html

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Pelosi G, Sonzogni A, Harari S, et al. Classification of pulmonary neuroendocrine tumors: new insights. *Translational Lung Cancer Research*. 2017;6(5):513-529. doi:10.21037/tlcr.2017.09.04.

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# Key Statistics for Lung Carcinoid Tumor

About 1% to 2% of all lung cancers are carcinoids. This means there are about 2,000 to 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 2 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 45 years for typical carcinoids and 55 years for atypical carcinoids.

Information on survival rates for lung carcinoids can be found in <u>Survival Rates for Lung Carcinoid Tumors</u><sup>1</sup>.

Visit the American Cancer Society's Cancer Statistics Center<sup>2</sup> for more key statistics.

# **Hyperlinks**

- 1. <u>www.cancer.org/cancer/lung-carcinoid-tumor/detection-diagnosis-staging/survival-rates.html</u>
- 2. http://cancerstatisticscenter.cancer.org/

#### References

American Cancer Society. *Cancer Facts & Figures 2019*. Atlanta: American Cancer Society; 2019.

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# What's New in Lung Carcinoid Tumor Research?

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.

The targeted therapy drug sunitinib (Sutent<sup>®</sup>) has been shown to be helpful in treating neuroendocrine tumors that start in the pancreas. More studies of this drug in carcinoid tumors (including lung carcinoid) 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<sup>®</sup>), pazopanib (Votrient<sup>®</sup>), axitinib (Inlyta<sup>®</sup>), and cabozantinib (Cometriq<sup>®</sup>).

Researchers are also trying to improve upon drugs related to somatostatin, which help some people with carcinoid tumors. An example is pasireotide (Signifor<sup>®</sup>), 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<sup>1</sup>.

# **Hyperlinks**

1. www.cancer.org/treatment/treatments-and-side-effects/clinical-trials.html

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