

# LUNG CARCINOID TUMOR

## What Is Cancer?

Cancer is a group of many related diseases. All forms of cancer involve out-of-control growth and spread of abnormal cells.

Normal body cells grow, divide, and die in an orderly fashion. During the early years of a person's life, normal cells divide more rapidly until the person becomes an adult. After that, normal cells of most tissues divide only to replace worn-out or dying cells and to repair injuries.

Cancer cells, however, continue to grow and divide, and can spread to other parts of the body. These cells accumulate and form *tumors* (lumps) that may compress, invade, and destroy normal tissue. If cells break away from such a tumor, they can travel through the bloodstream, or the lymph system to other areas of the body. There, they may settle and form "colony" tumors. In their new location, the cancer cells continue growing. The spread of a tumor to a new site is called *metastasis*. When cancer spreads, though, it is still named after the part of the body where it started. For example, if prostate cancer spreads to the bones, it is still prostate cancer, and if breast cancer spreads to the lungs it is still called breast cancer.

Leukemia, a form of cancer, does not usually form a tumor. Instead, these cancer cells involve the blood and blood-forming organs (bone marrow, lymphatic system, and spleen), and circulate through other tissues where they can accumulate.

It is important to realize that not all tumors are cancerous. Benign (noncancerous) tumors do not metastasize and, with very rare exceptions, are not life-threatening.

Cancer is classified by the part of the body in which it began, and by its appearance under a microscope. Different types of cancer vary in their rates of growth, patterns of spread, and responses to different types of treatment. That's why people with cancer need treatment that is aimed at their specific form of the disease.

In America, half of all men and one-third of all women will develop cancer during their lifetimes. Today, millions of people are living with cancer or have been cured of the disease. The risk of developing most types of cancer can be reduced by changes in a person's lifestyle, for example, by quitting smoking or eating a better diet. The sooner a cancer is found, and the sooner treatment begins, the better a patient's chances are of a cure.

# What Is A Lung Carcinoid Tumor?

## The Lungs

The lungs are two sponge-like organs found in the chest cavity. The right lung has three sections, called lobes. The left lung has two lobes. It is smaller because the heart takes up more room on that side of the body. The lungs bring air in and out, taking in oxygen and getting rid of carbon dioxide gas, a waste product of the body.

The lining, which surrounds the lungs and helps to protect them and to facilitate the sliding motion during breathing, is called the *pleura*. The chest cavity is called the *pleural cavity*. The *trachea* (windpipe) brings air down into the lungs. It divides into tubes called the *bronchi*, which divide into smaller branches called the *bronchioles*. At the end of the bronchioles are tiny air sacs known as *alveoli*.

## The Diffuse Neuroendocrine System

The *diffuse neuroendocrine system* consists of cells that are in certain ways like nerve cells and in other ways like cells of *endocrine* (hormone-producing) glands. These cells do not form an actual organ like the pancreas, adrenal, or thyroid. Instead, they are scattered throughout other organs like the lungs, stomach, and intestines.

Neuroendocrine cells of the lungs help control air flow and blood flow in the lungs and may help control growth of other types of lung cells. These neuroendocrine cells may detect decreased oxygen or increased carbon dioxide in the air we breathe and then release chemical messages to help the lungs adjust to changes in air composition. People who live at higher altitudes have an increased number of lung neuroendocrine cells, apparently because there is less oxygen in the air they breathe.

## Neuroendocrine Cancers

Like most cells of the body, lung neuroendocrine cells sometimes undergo certain changes that cause them to grow too much and form tumors. The tumors that develop from neuroendocrine cells are known as *neuroendocrine tumors* or *neuroendocrine cancers*.

There are four types of neuroendocrine lung tumors. The most serious type, *small cell lung cancer* (SCLC), is among the most rapidly growing and spreading of all cancers. It is discussed in a separate American Cancer Society lung cancer document. *Large cell neuroendocrine carcinoma* is a rare cancer that, with the exception of the size of the cells forming the cancer, is very similar to SCLC in its prognosis and in how patients are treated. *Carcinoid tumors*, also known as *carcinoids*, comprise the other 2 types of lung neuroendocrine cancer. These two

types are *typical carcinoid* and *atypical carcinoid*. This discussion will only cover these two tumors.

## **Carcinoid Tumors**

*Typical carcinoids* grow slowly and only rarely spread beyond the lungs. *Atypical carcinoids* grow a little faster and are somewhat more likely to spread to other organs. Typical carcinoids are nine times as common as atypical ones. The two types are distinguished from each other by their appearance under the microscope. Atypical carcinoids in their most malignant form resemble small cell lung cancers in their appearance and behavior.

In addition to being classified as typical or atypical based on how they look under a microscope, carcinoids are sometimes also classified according to where they form within the lung. *Central carcinoids* form in the walls of large airways near the center of the lungs. *Peripheral carcinoids* develop in the narrower airways toward the edges of the lungs. This distinction is important because the tumor's location affects which symptoms a patient will have (refer to How Are Lung Carcinoid Tumors Diagnosed?). Nearly all central carcinoids are also typical carcinoids. The majority of peripheral carcinoids are typical carcinoids but atypical ones are a significant minority.

## **What Are The Key Statistics About Lung Carcinoid Tumor?**

Lung carcinoid tumors account for 1% to 3% of all lung tumors. Most lung carcinoids vary from 0.5 cm (slightly smaller than  $\frac{1}{4}$  inch) to 2 cm (a little over  $\frac{3}{4}$  inch) at the time of diagnosis. Patients with carcinoids larger than 3 cm (almost 1  $\frac{1}{4}$  inch), atypical carcinoids, or carcinoids that have spread to lymph nodes have a worse *prognosis* (the outlook for chances of survival).

The 5-year survival rates for patients with typical and atypical lung carcinoids are 90% to 100% and 40% to 76% respectively. The ranges reflect different survival rates quoted by several medical journal articles. For both types of carcinoids, the ten-year survival rates are about 10% lower than the 5-year rates. The ranges of 5-year survival rates for patients without and with spread to lymph nodes are 90%-100% and 38%-74%.

## **What Are The Risk Factors For Lung Carcinoid Tumor?**

A *risk factor* is anything that increases a person's chance of getting a disease such as a type of cancer. For example, smoking is responsible for about 80% of cases of *lung carcinoma* (the usual and more serious type of lung cancer). In contrast, very little is known about why lung carcinoid tumors develop in some people but not in others.

**Chemical exposure:** Lung carcinoid tumors are not associated with smoking or with any known chemicals in the environment or workplace.

**Gender:** Carcinoids occur equally in men and women.

**Age:** These tumors are usually found in people between 45 and 55 years old. Children are rarely affected.

## **Do We Know What Causes Lung Carcinoid Tumor?**

Very little is known about the causes of lung carcinoid tumors. Researchers have learned much about how certain risk factors like cancer-causing chemicals or radiation cause lung cells to become carcinomas, the usual type of lung cancer. But, similar studies of lung carcinoid tumors have not found any risk factors.

Sometimes, tiny clusters of neuroendocrine cells that are similar to those seen in lung carcinoid tumors are seen under the microscope as an unexpected finding in lung biopsies done to treat or diagnose other conditions. These lesions, termed *carcinoid tumorlets* develop in small airways and are more common in people with certain lung diseases that cause formation of scar tissue in the lungs. Under the microscope, tumorlets have a striking resemblance to peripheral carcinoid tumors, except that they are usually much smaller (¼ inch). Central carcinoid tumors are believed to develop from the glands beneath the surface of the large air passages. Researchers still do not understand how carcinoid tumorlets develop from lung neuroendocrine cells or why some tumorlets may eventually grow to become carcinoid tumors.

## **Can Lung Carcinoid Tumors Be Prevented?**

Since research has not found any avoidable risk factors yet, there is no way to prevent carcinoid tumors.

## **Can Lung Carcinoid Tumors Be Found Early?**

Because carcinoid tumors usually grow and spread slowly, most are found at an early or localized stage. Most patients with peripheral carcinoid tumors or with small central carcinoid tumors have no symptoms. Carcinoids that do not cause symptoms often are found by chest x-rays done during a routine examination or done to look into unrelated medical problems such as some heart diseases.

## **Signs and Symptoms**

Some patients with central carcinoid tumors may have a cough, may cough up bloody sputum, or have wheezing symptoms like asthma. When a large carcinoid

causes partial or complete blockage of a large air passage, the patient may develop an infection of the lung called *post-obstructive pneumonia*. Sometimes the possibility of a tumor is considered by the doctor only after treatment with antibiotics fails to cure the patient's pneumonia.

Some carcinoid tumors can produce hormone-like substances that are released into the bloodstream. Lung carcinoids do this far less often (10% to 20% of the time) than do carcinoid tumors in the gastrointestinal system. The *carcinoid syndrome* results from the effect of these substances. Symptoms include facial *flushing* (redness and warm feeling that may last hours to days), sweating, diarrhea, and fast heartbeat. Some carcinoid tumors may produce adrenocorticotrophic hormone (ACTH), a hormone that stimulates the adrenal gland to produce excessive amounts of cortisol and related hormones. Symptoms of excessive amounts of these hormones include weight gain, weakness, secondary diabetes, and increased body and facial hair.

## **How Are Lung Carcinoid Tumors Diagnosed?**

If there is a reason to suspect you may have a lung carcinoid tumor, the doctor will use one or more methods to find out if the disease is really present. About two-thirds of the time, this suspicion is based on the patient's symptoms. If you have one or more symptoms that suggest this type of tumor, the doctor will specifically ask about other symptoms such as cough, chest pain, wheezing, asthma, blood-tinged sputum, pneumonia that is not cured by antibiotics, recent weight gain, facial flushing (redness), or diarrhea.

A thorough physical exam will provide information about signs of carcinoid tumor such as the carcinoid syndrome and other health problems.

### **Imaging Tests**

Imaging tests produce pictures or images of the inside of the body. Imaging tests such as chest x-rays, computed tomography (CT) scans, octreoscans, and MIBG scans are useful in finding carcinoid tumors and determining how far they have spread.

Chest x-rays will be taken to look for a tumor in the lungs. However, some carcinoids that are small or in locations where they are covered by other organs in the chest may not show up on a chest x-ray. If the doctor is still suspicious or there is a vague abnormality on the chest x-ray, a CT scan may be ordered.

Sometimes a *computed tomographic (CT) scan* is used to get a more detailed view of something seen on the chest x-ray or to look for a mass if none were seen on the chest x-rays. The CT scan is a special x-ray procedure in which the x-ray beam moves around the body, taking pictures from different angles. These

pictures are then combined by a computer to produce a detailed cross-sectional view of organs inside the body.

About one-third of carcinoids are diagnosed by accident, in people without any symptoms to suggest a carcinoid. In these cases, a chest x-ray done to evaluate an unrelated medical problem finds a lung mass.

Even if imaging tests such as a chest x-ray and/or CT scan find a mass, these imaging tests cannot tell if the mass is a carcinoid tumor, a lung carcinoma, or a localized infection. The only way to tell is to remove cells from the tumor and examine them under a microscope. This procedure is called a *biopsy*.

A procedure called *indium-111-labeled DTPA-octreotide scintigraphy* or *octreoscan* may be used in some cases. A small amount of this radioactive hormone-like substance (octreotide is a hormone that attaches to carcinoid tumors) is injected into a vein. This material is attracted to carcinoid tumors. A special radioactivity-detecting camera is used to show where the radioactivity accumulates. This test is useful in detecting spread of lung carcinoid tumors to other areas of the body. A similar test uses radioactive MIBG. MIBG is a chemical that is taken up by carcinoid tumors. The MIBG is attached to radioactive iodine and injected in the blood stream. If there is a carcinoid tumor, the radioactivity will go there and the scanner will detect it.

## **Biopsy**

There are several ways to take a sample from a lung tumor. Tumors of large airways, such as central carcinoids, can be sampled by *bronchoscopic biopsy*. The doctor passes a long, lighted tube called a *bronchoscope* down the throat to look at the lining of the lung's main airways. When a tumor is found, the doctor can operate a biopsy *forceps* (pincers or tongs) through the tube to take a small sample of the tumor. The advantage of this approach is that no surgical incision is needed, no hospital stay is needed, and the patient is ready to return home within hours. One disadvantage is that a bronchial biopsy may not always be able to remove enough tissue to be certain the tumor is a carcinoid. But, thanks to recent advances in laboratory testing of lung tumors, doctors can usually make a more accurate diagnosis even with very small samples. Bleeding after a biopsy from a carcinoid tumor is a rare but potentially serious problem. If bleeding becomes a problem, doctors can inject drugs through the bronchoscope into the tumor to narrow its blood vessels, or they can seal off the bleeding vessels with a laser beam aimed through the bronchoscope. Doctors can also take a *brushing* sample through the bronchoscope. They wipe a tiny brush over the surface of the tumor. The cells removed by the brush are smeared onto a microscope slide and examined in the lab. Brushing samples are sometimes a helpful addition to the bronchial biopsy but are not as helpful with diagnosing carcinoids as they are with lung carcinomas.

Tumors that are not near the large airways are often sampled by needle biopsy. A long needle is passed between the ribs into the lung. CT scan images are used to guide the needle into the tumor so a small tissue sample can be removed for examination under the microscope. This procedure is also done without a surgical incision or overnight hospital stay. Some patients may experience a collapse of one lung, called *pneumothorax*, after this procedure. But, this complication can be treated effectively by temporarily placing a suction tube into the chest.

In some cases, neither a bronchoscopic biopsy nor a needle biopsy can provide enough tissue to identify the type of tumor, and a *thoracotomy* (surgically opening the chest cavity) is needed. In other cases, when the doctors strongly suspect a carcinoid or some other type of lung cancer, they may do a thoracotomy and remove the entire mass without first doing a bronchoscopic biopsy or needle biopsy.

### **Blood Tests**

Blood tests may be done to detect some of the hormone-like substances produced by carcinoids, particularly if the patient has symptoms of the *carcinoid syndrome*, caused by excessive levels of such substances in the blood.

## **How Are Lung Carcinoid Tumors Staged?**

Staging is a process of finding out how localized or widespread the lung carcinoid tumor is. It will show if the tumor has spread and how far. The treatment and *prognosis* (the outlook for chances of survival) for a lung carcinoid tumor depends, to a large extent, on its stage. The staging of lung carcinoid tumor is identical to the staging of non-small cell lung cancer.

The system most often used to describe the growth and spread of carcinoids and of non-small cell lung cancers is the **TNM** staging system, also known as the American Joint Committee on Cancer or AJCC system. **T** stands for tumor (its size and how far it has spread within the lung and to nearby organs), **N** stands for spread to lymph nodes, and **M** is for *metastasis* (spread to distant organs). In TNM staging, information about the tumor, lymph nodes, and metastasis is combined and a stage is assigned to specific TNM groupings. The grouped stage is described using Roman numerals from 0 to IV.

### **T Stages**

**T1:** The cancer is no larger than 3 centimeters (slightly less than 1¼ inches), has not spread to the *visceral pleura* (membranes that surround the lungs), and does not affect the main branches of the bronchi.

T2: The cancer has one or more of the following features:

- it is larger than 3 cm
- it involves a main bronchus, but is not closer than 2 cm (about ¾ inch) to the point where the trachea (windpipe) branches into the left and right main bronchi
- it has spread to the *visceral pleura*
- the cancer may partially clog the airways, but this has not caused the entire lung to collapse or develop pneumonia

T3: The cancer has one or more of the following features:

- spread to the chest wall, the *diaphragm* (breathing muscle that separates the chest from the abdomen), the *mediastinal pleura* (membranes surrounding the space between the two lungs), or *parietal pericardium* (membranes of the sac surrounding the heart)
- involves a main bronchus and is closer than 2 cm (about ¾ inch) to the point where the *trachea* (windpipe) branches into the left and right main bronchi, but does not involve this area
- has grown into the airways enough to cause one lung to entirely collapse or to cause pneumonia of the entire lung

T4: The cancer has one or more of the following features:

- spread to the *mediastinum* (space behind the chest bone and in front of the heart), the heart, the *trachea* (windpipe), the *esophagus* (tube connecting the throat to the stomach), the backbone, or the point where the windpipe branches into the left and right main bronchi
- two or more separate tumor nodules are present in the same lobe
- there is a *malignant pleural effusion* (fluid containing cancer cells in the space surrounding the lung)

## **N Stages**

N0: No spread to lymph nodes

N1: Spread to lymph nodes within the lung, *hilar lymph nodes* (located around the area where the bronchus enters the lung). Metastases affect lymph nodes only on the same side as the cancerous lung.

N2: Spread to lymph nodes around the point where the windpipe branches into the left and right bronchi or to lymph nodes in the *mediastinum* (space behind the chest bone and in front of the heart). Affected lymph nodes are on the same side of the cancerous lung.

N3: Spread to lymph nodes near the collar bone on either side, to hilar or mediastinal lymph nodes on the side opposite the cancerous lung.

## **M Stages**

M0: No distant spread

M1: Distant spread is present. Sites considered distant include other lobes of the lungs, lymph nodes further than those mentioned in N stages, and other organs or tissues such as the liver, bones, or brain.

### Stage Grouping

Once the T, N, and M categories have been assigned, this information is combined (stage grouping) to assign an overall stage of I, II, III, or IV. Patients with lower stage numbers have a more favorable outlook for survival.

<u>Overall Stage</u>	<u>T Stage</u>	<u>N Stage</u>	<u>M Stage</u>
Stage IA	T1	N0	M0
Stage IB	T2	N0	M0
Stage IIA	T1	N1	M0
Stage IIB	T2	N1	M0
	T3	N0	M0
Stage IIIA	T1	N2	M0
	T2	N2	M0
	T3	N1	M0
	T3	N2	M0
Stage IIIB	Any T	N3	M0
	T4	Any N	M0
Stage IV	Any T	Any N	M1

### How Are Lung Carcinoids Treated?

After the tumor is found and staged, the cancer care team will suggest one or more treatment plans. This is an important decision, and it is important for you to take time and think about all of the choices.

The main factors in selecting treatment options for lung carcinoid tumors are the size and location of the tumor, whether it has spread to lymph nodes or other organs, whether there are any other serious medical conditions. It is often a good idea to seek a second opinion. A second opinion may provide more information and help the patient feel more certain about the treatment plan that is chosen.

### Surgery

Most lung carcinoid tumors are cured by surgery alone. The patient will be referred to a thoracic or cardiothoracic surgeon who will discuss the surgical options. The type of operation will depend on a number of factors, including the size and location of the tumor, and whether the patient has any other lung problems or serious diseases of other organs.

There are several operations that may be used to treat people with lung carcinoid tumors. It is usually necessary to remove some normal lung tissue along with the tumor, but surgeons try not to remove any more normal tissue than they need to.

To treat central carcinoids of a large airway, the surgeon may do a *sleeve resection*. If you think of the large airway with a tumor as similar to the sleeve of a shirt with a stain an inch or two above the wrist, the sleeve resection would be like cutting across the sleeve above and below the stain and sewing the cuff back into the shortened sleeve. If it is not possible to do a sleeve resection because of the size of the tumor and its exact location in a large airway, the surgeon will usually do a *lobectomy* (remove an entire lobe). Less often, it may be necessary to remove two lobes or, rarely, perform a *pneumonectomy* (remove the whole left or right lung).

Peripheral carcinoids, found at the edges of the lungs away from the large airways, are usually treated by lobectomy. If the tumor is very small, the surgeon may do a *wedge resection*, removing a wedge-shaped piece of the lung.

In most cases, the surgeon will also remove some lymph nodes near the lungs. This is important because about 10% of typical carcinoids and 30% to 50% of atypical carcinoids will have spread to lymph nodes by the time they are diagnosed. Not removing these nodes might increase the risk of the carcinoid tumor spreading even further, to other organs. At that point, the patient could no longer be cured by surgery. Removing the lymph nodes also provides some indication of risk of recurrence of distant metastases.

Some patients who also have lung diseases such as severe emphysema or chronic bronchitis may not be able to have their carcinoid treated surgically, because removing some normal lung tissue along with the cancer would cause severe shortness of breath. Patients with other medical problems, such as severe heart disease, also may not be able to have curative surgery. For these patients, palliative procedures, such as removing most of the tumor through a bronchoscope or vaporizing most of it with a laser can be helpful. These treatments can relieve symptoms caused by blockage of airways, but they cannot cure the cancer and are recommended only for patients that cannot have surgery to completely remove the tumor. Such treatments are often supplemented by external or intrabronchially-administered radiation (See essay on radiation therapy).

## Medical Treatments

**Chemotherapy:** Chemotherapy uses anticancer drugs that are injected into a vein or a muscle or taken by mouth. These drugs enter the bloodstream and reach all areas of the body, making this treatment useful for some types of lung cancer that has spread or metastasized to organs beyond the lungs. Unfortunately, carcinoid tumors are usually not sensitive to chemotherapy. Chemotherapy is generally used only for carcinoid tumors that have spread to other organs, are causing severe symptoms, and have not responded to other medications. Some of the chemotherapy drugs used in this situation include etoposide, cisplatin, cyclophosphamide, 5-fluorouracil, doxorubicin (Adriamycin), and dacarbazine. Several chemotherapeutic drugs are sometimes used together to treat metastatic carcinoid tumor, often in combination with other types of medications.

Drugs used in chemotherapy kill some cancer cells but can also affect some of the normal, healthy cells in your body, causing side effects. Rapidly growing cells, such as the blood-producing cells of bone marrow, and the cells of hair follicles and the lining of the mouth are particularly sensitive to chemotherapy. Possible side effects include:

- Nausea, vomiting, and decrease in appetite
- Temporary loss of hair
- Mouth sores
- Increased risk of infections (due to low white blood cell counts) or bleeding (due to low blood platelet counts)
- Fatigue

If you have side effects, your cancer care team can suggest steps to ease them. For example, there are drugs available to help control nausea and vomiting. Sometimes changing the dosage or the time of day at which you take your medications can reduce side effects. Fortunately, most side effects will disappear when your course of treatment ends.

Chemotherapy is only occasionally effective in shrinking carcinoid tumors. Patients should discuss with their doctors whether the side effects they experience are worth the small chance they will get better.

**Other drugs for treating carcinoid tumors:** Several medications are available for controlling symptoms of carcinoid syndrome (problems arising from release of substances produced by some of these tumors and recognized through blood and urine tests) in patients with metastatic carcinoid tumors. *Octreotide* is a drug chemically related to a natural hormone, somatostatin. It is very helpful in treating the flushing (skin redness and feeling hot), diarrhea, and wheezing from carcinoid syndrome. Sometimes octreotide can temporarily shrink carcinoid tumors, but it does not cure them. The medication's side effects principally include pain at the site of its injection, and rarely, stomach cramps, nausea, vomiting, headaches, dizziness, and fatigue. *Interferons* are substances that activate the body's immune

system. Alpha-interferon is helpful in shrinking some metastatic carcinoid tumors and improving symptoms of carcinoid syndrome.

Other medications are also available for controlling specific *syndromes*. Ask your doctor about them, or describe your symptoms to your doctor and ask about medications to control them.

## **Radiation Therapy**

Radiation therapy uses high-energy radiation to kill cancer cells. Although most cases of carcinoid tumor are cured by surgery alone, if for some reason the patient is unable to have an operation, radiotherapy may be an option.

*External beam radiation therapy* is the type of radiation used most often for lung cancer. It is like having a regular x-ray except it lasts a little longer. Patients typically have treatments for five days a week for several weeks. Unfortunately, radiation therapy is not usually very effective against most lung carcinoid tumors.

The main side effects of lung radiation therapy are fatigue (tiredness) and mild temporary, sunburn-like skin changes. If high doses are given, radiation damage to normal lung tissue can cause scar tissue formation, trouble with breathing, and increased susceptibility to infection.

## **Complementary and Alternative Therapies**

If you are considering any unproven alternative or complementary treatments, it is best to discuss this openly with your cancer care team and request information from the American Cancer Society or the National Cancer Institute. Some unproven treatments can interfere with standard medical treatments or may cause serious side effects.

## **Clinical Trials**

Studies of promising new or experimental treatments in patients are known as clinical trials. A clinical trial is only done when there is some reason to believe that the treatment being studied may be of value to the patient. Treatments used in clinical trials are often found to have real benefits. There are three phases of clinical trials in which a treatment is studied before the treatment is eligible for approval by the FDA (Food and Drug Administration).

The purpose of a Phase I study is to find the best way to give a new treatment and how much of it can be given safely. Physicians watch patients carefully for any harmful side effects. The research treatment has been well tested in laboratory and animal studies, but the side effects in patients are not completely predictable.

Phase II trials determine the effectiveness of a research treatment after safety has been evaluated in a Phase I trial. Patients are closely observed for an anticancer effect by careful measurement of cancer sites present at the beginning of the trial. In addition to monitoring patients for response, any side effects are carefully recorded and assessed.

Phase III trials require entry of large numbers of patients. Some trials enroll thousands of patients. One of the groups may receive standard (the most accepted) treatment, so the new treatments can be directly compared. The group that receives the standard treatment is called the "control group." For example, one group of patients (the control group) may receive the standard chemotherapy for a certain type of cancer, while another patient group may receive a different type of chemotherapy, that may or may not contain an investigational drug, to see if this improves survival. All patients in Phase III trials are monitored closely for side effects, and treatment is discontinued if the side effects are too severe.

Researchers conduct studies of new treatments to answer the following questions:

- Is the treatment likely to be helpful?
- Does this new type of treatment work?
- Does it work better than other treatments already available?
- What side effects does the treatment cause?
- Do the benefits outweigh the risks, including side effects?
- In which patients is the treatment most likely to be helpful?

However, there are some risks. No one involved in the study knows in advance whether the treatment will work or exactly what side effects will occur. That is what the study is designed to discover. While most side effects will disappear in time, some can be permanent or even life-threatening. Keep in mind that even standard treatments have side effects. Depending on many factors, a patient may decide that a clinical trial will be beneficial.

Enrollment in any clinical trial is completely up to you. Your doctors and nurses will explain the study to you in detail and will give you a form to read and sign indicating your desire to take part. This process is known as giving your *informed consent*. Even after signing the form and after the clinical trial begins, you are free to leave the study at any time, for any reason. Taking part in the study does not prevent you from getting other medical care you may need.

To find out more about clinical trials, ask your cancer care team. Among the questions you should ask are:

- What is the purpose of the study?
- What kinds of tests and treatments does the study involve?
- What does this treatment do?
- What is likely to happen in my case with, or without, this new research treatment?
- What are my other choices and their advantages and disadvantages?

- How could the study affect my daily life?
- What side effects can I expect from the study? Can the side effects be controlled?
- Will I have to be hospitalized? If so, how often and for how long?
- Will the study cost me anything? Will any of the treatment be free?
- If I am harmed as a result of the research, what treatment would I be entitled to?
- What type of long-term follow-up care is part of the study?
- Has the treatment been used to treat other types of cancers?

You can get a list of current clinical trials by calling the National Cancer Institute's Cancer Information Service toll free at 1-800-4-CANCER or visiting the NCI clinical trials website for patients ([cancertrials.nci.nih.gov](http://cancertrials.nci.nih.gov)) or healthcare professionals ([cancer.net.nci.nih.gov/prot/protsrch.shtml](http://cancer.net.nci.nih.gov/prot/protsrch.shtml)).

## **What Should You Ask Your Doctor About Lung Carcinoid Tumor?**

It is important to have frank, open discussions with your cancer care team. They want to answer all of your questions, no matter how trivial you might think they are. For instance, consider these questions:

- What kind of carcinoid tumor do I have?
- Has my carcinoid tumor spread beyond the lungs?
- What is the stage of my carcinoid tumor and what does that mean in my case?
- What treatment choices do I have?
- What do you recommend and why?
- Based on what you've learned about my carcinoid tumor, how long do you think I'll survive?
- What risks or side effects are there to the treatments you suggest?
- What are the chances of recurrence of my carcinoid tumor with these treatment plans?
- What should I do to be ready for treatment?

In addition to these sample questions, be sure to write down some of your own. For instance, you might want more information about recovery times so you can plan your work schedule. Or, you may want to ask about second opinions or about clinical trials for which you may qualify.

## What Happens After Treatment For Lung Carcinoid Tumor?

After treatment, you will be asked to return to the doctor for regular physical exams and, in some cases, x-rays and blood tests. These exams and tests will help find any recurrences of the tumor as early as possible.

It is important for the patient to report any new symptoms to the doctor right away. They might be signs of the tumor's recurrence or of side effects from treatment.

If you smoke, it is very important to quit. Quitting improves the health of your heart and lungs. After surgery to remove some lung tissue, some people have breathing problems and feel weak. Smoking makes this situation worse. Once you have recovered from surgery, ask your doctors and nurses about physical activity. A good exercise plan may help you recover faster and regain your energy level and reduce shortness of breath.

## What's New In Lung Carcinoid Tumor Research And Treatment?

There is always research going on in the field of lung cancer. Scientists are looking for causes and ways to prevent carcinoid tumors.

Present and current carcinoid tumor research is focused on identifying causes, and improving diagnosis and treatment of metastatic tumors.

**Genetics:** Researchers have made great progress in understanding how certain changes in DNA can cause normal cells to become cancerous. DNA is the molecule that carries the instructions for nearly everything our cells do. We usually resemble our parents because they are the source of our DNA. However, DNA affects more than our outward appearance. Some *genes* (parts of our DNA) contain instructions for controlling when our cells grow and divide. Certain genes that promote cell division are called *oncogenes*. Others that slow down cell division or cause cells to die at the appropriate time are called *tumor-suppressor genes*. It is known that cancers can be caused by DNA *mutations* (defects) that turn on oncogenes or turn off tumor-suppressor genes. Many of the DNA changes in lung carcinoids have been characterized by researchers in the past few years.

However, much less is known about DNA changes in carcinoid tumors. It is expected 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 lung carcinomas are very different, accurate diagnosis is important. Researchers have made great progress in developing tests that can detect specific substances found in the cells of carcinoid tumors but not lung cancers. Other substances may be

found in both carcinoids and carcinomas but higher levels are found in one type. Most of these tests involve treating tissue samples with special antibodies produced in the laboratory. The antibodies are designed to recognize specific substances in certain types of tumors.

**Imaging:** Researchers are testing <sup>111</sup>In-DTPA-octreotide scintigraphy (octreoscan) scan and other nuclear medicine methods for the early detection of carcinoid tumors.

**Treatment:** New chemotherapeutic agents are being tested to find treatments that are active against carcinoids. Other drugs are being developed to prevent release of substances responsible for symptoms of carcinoid syndrome. A new somatostatin-like drug called Lanreotide has been developed. It has proved effective in shrinking tumors in some patients and relieving symptoms of the carcinoid syndrome in over 50% of patients. Another drug, MIBG, a chemical taken up by the carcinoid tumor also damages them. It has also been helpful in some patients. When radioactive iodine is attached to the molecule it is even more helpful and may, in the future, become a very useful treatment. It has few side effects.

## **Additional Resources**

### **National Organizations and Web Sites**

The following organizations can also provide additional information and resources . \*

Alliance for Lung Cancer Advocacy, Support & Education (ALCASE)  
1601 Lincoln Avenue  
Vancouver, Washington 98660  
Telephone: 1-800-298-2436 or 360-696-2436  
Internet Address: [www.alcase.com](http://www.alcase.com)

American Lung Association  
1740 Broadway  
New York, New York 10019  
Telephone: 1-800-586-4872 or (212) 315-8700  
Internet Address: [www.lungusa.org](http://www.lungusa.org)

National Cancer Institute  
International Cancer Information Center  
Telephone: 1-800-4-CANCER  
Internet Addresses: [www.nci.nih.gov](http://www.nci.nih.gov) and [cancernet.nci.nih.gov](http://cancernet.nci.nih.gov)

The Carcinoid Cancer Foundation, Inc.

171 York Avenue  
New York, NY 10128  
Telephone: 1-212-722-3132  
Internet Address: www.carcinoid.org

*\*Inclusion on this list does not imply endorsement by the American Cancer Society*

### **Additional American Cancer Society Information**

*After Diagnosis: A Guide for Patients and Families* (Booklet; Code #9440)

*Caregiving: A Step-By-Step Resource for Caring for the Person with Cancer at Home* (Book; Code #9422)

*Caring for the Patient with Cancer at Home* (Booklet; Code #4545)

*Questions About Smoking, Tobacco, and Health* (Booklet; Code #2023)

*Questions and Answers About Pain Control* (Booklet; Code #4518)

### **Other Publications\***

*\*Inclusion on this list does not imply endorsement by the American Cancer Society*

*A Cancer Survivor's Almanac: Charting Your Journey*. Edited by Barbara Hoffman, JD. National Coalition for Cancer Survivorship. Chronimed Publishing, 1996.

Capossela, Cappy, Warnock Sheila. *Share the Care: How to Organize a Group for Someone Who Is Seriously Ill*. New York: Simon and Schuster, 1995.

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