



# Lung Carcinoid Tumor

## What is cancer?

The body is made up of trillions of living cells. Normal body cells grow, divide to make new cells, and die in an orderly way. During the early years of a person's life, normal cells divide faster to allow the person to grow. After the person becomes an adult, most cells divide only to replace worn-out or dying cells or to repair injuries.

Cancer begins when cells in a part of the body start to grow out of control. There are many kinds of cancer, but they all start because of out-of-control growth of abnormal cells.

Cancer cell growth is different from normal cell growth. Instead of dying, cancer cells continue to grow and form new, abnormal cells. Cancer cells can also invade (grow into) other tissues, something that normal cells cannot do. Growing out of control and invading other tissues are what makes a cell a cancer cell.

Cells become cancer cells because of damage to DNA. DNA is in every cell and directs all its actions. In a normal cell, when DNA gets damaged the cell either repairs the damage or the cell dies. In cancer cells, the damaged DNA is not repaired, but the cell doesn't die like it should. Instead, this cell goes on making new cells that the body does not need. These new cells will all have the same damaged DNA as the first cell does.

People can inherit damaged DNA, but most DNA damage is caused by mistakes that happen while the normal cell is reproducing or by something in our environment. Sometimes the cause of the DNA damage is something obvious, like cigarette smoking. But often no clear cause is found.

In most cases the cancer cells form a tumor. Some cancers, like leukemia, rarely form tumors. Instead, these cancer cells involve the blood and blood-forming organs and circulate through other tissues where they grow.

Cancer cells often travel to other parts of the body, where they begin to grow and form new tumors that replace normal tissue. This process is called *metastasis*. It happens when the cancer cells get into the bloodstream or lymph vessels of our body.

No matter where a cancer may spread, it is named (and treated) based on the place where it started. For example, breast cancer that has spread to the liver is still breast cancer, not liver cancer. Likewise, prostate cancer that has spread to the bone is still prostate cancer, not bone cancer.

Different types of cancer can behave very differently. For example, lung cancer and breast cancer are very different diseases. They grow at different rates and respond to different treatments. That is why people with cancer need treatment that is aimed at their particular kind of cancer.

Not all tumors are cancerous. Tumors that aren't cancer are called *benign*. Benign tumors can cause problems – they can grow very large and press on healthy organs and tissues. But they cannot grow into (invade) other tissues. Because they can't invade, they also can't spread to other parts of the body (metastasize). These tumors are almost never life threatening.

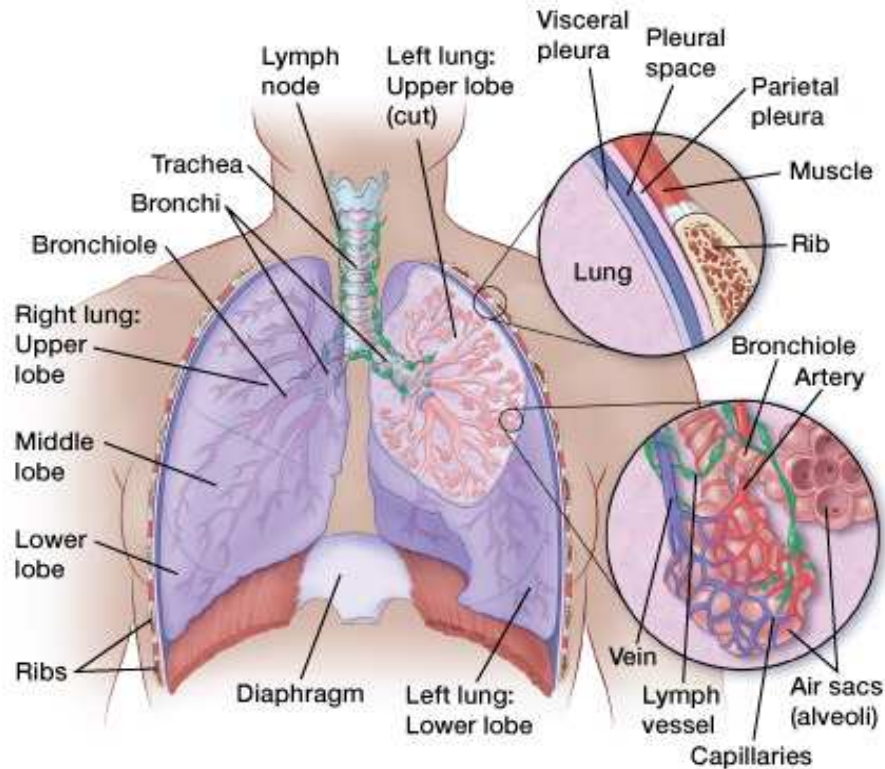
## What are lung carcinoid tumors?

Lung carcinoid tumors (also known as *lung carcinoids*) are uncommon tumors that start in the lungs. They 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 more 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 can 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 our documents *Gastrointestinal Carcinoid Tumors* and *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. It is discussed in our document *Lung Cancer (Small Cell)*.

## Large cell neuroendocrine carcinoma

Large cell neuroendocrine carcinoma (LCNEC) is a rare cancer. It is a subtype of large cell carcinoma, which is a type 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. For treatment information, see *Lung Cancer (Non-Small Cell)*.

## Typical and atypical carcinoid tumors

The other 2 types of lung neuroendocrine tumors are carcinoids. **The rest of this document will only cover these 2 types of tumors.**

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. Seen under a microscope, 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 the section "Signs and symptoms of lung carcinoid tumors?") and may also affect how the tumor is treated.

## 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 are actually more common 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 for 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 the section “Survival rates for lung carcinoid tumors.”

## **What are the risk factors for lung carcinoid tumors?**

A risk factor is anything that affects your chance of getting a disease such as cancer. Different cancers have different risk factors. Some risk factors, like smoking, can be changed. Others, like a person’s age or family history, can’t be changed.

But having a known risk factor, or even several risk factors, does not mean that you will get the disease. And some people who get the disease may have few or no known risk factors.

Not much is known about why lung carcinoid tumors develop in some people but not in others.

### **Gender**

Carcinoids occur more often in women than in men. The reasons for this are not known.

### **Race/ethnicity**

Lung carcinoids are more common in whites than in African Americans, Asian Americans, or Hispanics/Latinos.

### **Age**

These tumors are usually found in people about 60 years old, which is slightly younger than the average age for other types of lung cancer. But carcinoids can occur in people of almost any age. Although it’s rare, lung carcinoid tumors are sometimes even found in children.

### **Multiple endocrine neoplasia type 1**

People with multiple endocrine neoplasia type 1 (MEN1), an inherited syndrome, are at high risk for tumors in certain endocrine organs, such as the pancreas and the pituitary and parathyroid glands. These people also seem to be at increased risk for lung carcinoid tumors.

## Family history

Most people with lung carcinoid tumors do not have a family history of this type of cancer, but having a family history of lung carcinoid tumors can increase your risk. In rare cases, several family members have been diagnosed with this cancer. The overall risk is still low because this cancer is so uncommon.

## Tobacco smoke

Typical lung carcinoid tumors do not seem to be linked with smoking or with any known chemicals in the environment or workplace. But some studies have found that atypical lung carcinoids may be more common in people who smoke.

## Do we know what causes lung carcinoid tumors?

Not much is known about what causes lung carcinoid tumors. Researchers have learned a lot about how certain risk factors like cancer-causing chemicals or radiation can cause changes in lung cells that lead to carcinomas, the more common type of lung cancer. But these factors are not thought to play a large role in the development of lung carcinoid tumors.

Carcinoid tumors probably develop from tiny clusters of neuroendocrine cells in the lung airways called *carcinoid tumorlets*. Tumorlets are sometimes found unexpectedly in lung biopsies done to treat or diagnose other conditions. Under the microscope, tumorlets resemble carcinoid tumors, except that they are much smaller – less than 5 mm (about ¼ inch) across. Most tumorlets never grow any bigger, but some may eventually become carcinoid tumors.

Researchers have found some chromosome and gene changes that are common in lung carcinoid tumor cells. Chromosomes are long strands of DNA in each of our cells that contain our genes – the instructions for how our cells function. But it is still not clear exactly how these changes cause carcinoid tumorlets to develop from lung neuroendocrine cells or how they might cause tumorlets to grow to become carcinoid tumors.

## Can lung carcinoid tumors be prevented?

Because we do not yet know what causes most lung carcinoid tumors, it is not possible to know how to prevent them.

Smoking has been linked with an increased risk of atypical carcinoids in some studies, so quitting (or not starting) might reduce a person's risk.

# Can lung carcinoid tumors be found early?

Lung carcinoid tumors are not common, and there are no widely recommended screening tests for these tumors in most people. (Screening is testing for cancer in people without any symptoms.)

People with multiple endocrine neoplasia type 1 (MEN1) are at increased risk for these tumors, and some doctors recommend they have computed tomography (CT) scans of the chest every 3 years starting when they are age 20.

Because carcinoid tumors usually grow and spread slowly, most are found at an early or localized stage, even if they have been causing symptoms for some time.

Many patients with peripheral carcinoid tumors or with small central carcinoid tumors have no symptoms. Carcinoids that do not cause symptoms often are found on a chest x-ray or CT scan done for other reasons.

## Signs and symptoms of lung carcinoid tumors

About 2 out of 3 people with carcinoid tumors will have signs or symptoms that will lead to the diagnosis of the disease. But because carcinoids tend to grow slowly, they may not cause symptoms for several years in some people, or they may be found by medical tests done for other reasons.

### Central carcinoids

Most carcinoid tumors start in the large bronchial tubes leading into the lung. People with these tumors may have a cough, which can sometimes be bloody, or they may have wheezing. Other possible symptoms include shortness of breath and chest pain, especially when taking deep breaths.

Large carcinoids can cause partial or complete blockage of an air passage, which can lead to pneumonia (an infection in the lung). Sometimes a doctor may suspect a tumor only after treatment with antibiotics fails to cure the pneumonia.

### Peripheral carcinoids

Some tumors start in the smaller airways toward the outer edges of the lungs. They rarely cause any symptoms unless there are so many of them that they cause trouble breathing. Usually they are found as a spot on a chest x-ray or CT scan that is done for an unrelated problem.



## Symptoms caused by hormones from the tumor

Some carcinoid tumors can make hormone-like substances that are released into the bloodstream. Lung carcinoids do this far less often than gastrointestinal carcinoid tumors.

**Carcinoid syndrome:** Rarely, lung carcinoid tumors release enough hormone-like substances into the bloodstream to cause symptoms. This results in the carcinoid syndrome. Symptoms can include facial flushing (redness and warm feeling), severe diarrhea, wheezing, and fast heartbeat. Many patients find that stress, heavy exercise, and drinking alcohol can bring on these symptoms or make them worse. Over a long time, these hormone-like substances can damage heart valves, causing shortness of breath, weakness, and a heart murmur (an abnormal heart sound).

**Cushing syndrome:** In rare cases, lung carcinoid tumors may make a hormone called ACTH. This causes the adrenal glands to make too much cortisol and other hormones. This can lead to weight gain, easy bruising, weakness, drowsiness, high blood sugar (or even diabetes), high blood pressure, and increased body and facial hair.

The symptoms and signs above may be caused by lung carcinoid tumors, but they can also be caused by other conditions. Still, if you have any of these problems, it's important to see your doctor so the cause can be found and treated, if needed.

## How are lung carcinoid tumors diagnosed?

Certain signs and symptoms might suggest that a person could have a lung carcinoid tumor, but tests are needed to confirm the diagnosis.

### Medical history and physical exam

If you have any signs or symptoms that suggest you might have a lung carcinoid tumor (or another type of lung tumor), your doctor will want to take a complete medical history, including your family history, to learn about your symptoms and possible risk factors.

A physical exam can give your doctor information about your general health, possible signs of lung carcinoid tumor, and other health problems. During your exam, your doctor will pay close attention to your chest and lungs.

If your symptoms or the results of the exam suggest you might have a lung carcinoid tumor (or another type of tumor), more tests will be done. These might include imaging tests, lab tests, and other procedures.

## Imaging tests

Imaging tests use x-rays, radioactive particles, or other means to create pictures of the inside of your body. Imaging tests may be done for a number of reasons, including to help find a suspicious area that might be cancer, to learn how far cancer may have spread, and to help determine if treatment has been effective.

### **Chest x-ray**

A chest x-ray is often the first imaging test a doctor orders if a lung problem is suspected. It might be able to show if there is a tumor in the lung. But some carcinoids that are small or are in places where they are covered by other organs in the chest may not show up on a chest x-ray. If your doctor is still suspicious or if something is seen on the chest x-ray, a CT scan may be ordered.

### **Computed tomography (CT) scan**

A CT (or CAT) scan is more likely to show lung tumors than routine chest x-rays. A CT scan can also provide precise information about the size, shape, and position of any lung tumors and can help find enlarged lymph nodes that might contain cancer that has spread from the lung.

The CT scan uses x-rays to produce detailed cross-sectional images of your body. Instead of taking one picture, like a regular x-ray, a CT scanner takes many pictures as it rotates around you while you are lying on a narrow platform. A computer then combines these into images showing slices of the part of your body that is being studied.

Before any pictures are taken, you may be asked to drink 1 to 2 pints of a liquid called *oral contrast*. This helps outline the intestine so that certain areas are not mistaken for tumors. This is not needed if the CT scan is only looking at the chest and lungs. You may also receive an IV (intravenous) line through which a different kind of contrast dye (IV contrast) is injected. This helps better outline structures in your body. The injection can cause some flushing (redness and warm feeling). Some people are allergic and get hives or, rarely, more serious reactions like trouble breathing and low blood pressure. Be sure to tell the doctor if you have any allergies or have ever had a reaction to any contrast material used for x-rays.

A CT scanner has been described as a large donut, with a narrow table that slides in and out of the middle opening. You will need to lie still on the table while the scan is being done. CT scans take longer than regular x-rays, and you might feel a bit confined by the ring while the pictures are being taken.

CT scans can have several uses:

- CT scans of the chest can spot very small lung tumors and help determine the exact location and extent of the tumors.

- CT scans can be helpful in staging a cancer (determining the extent of its spread). For example, CT scans of the abdomen can show if the cancer has spread to the liver or other organs. This can help to determine if surgery is a good treatment option.
- CT scans can also be used to guide a biopsy needle precisely into a suspected tumor. For this procedure, called a *CT-guided needle biopsy*, you stay on the CT scanning table while the doctor advances a biopsy needle through the skin and toward the mass. CT scans are repeated until the needle is within the mass. A biopsy sample is then removed and looked at under a microscope.
- CT scans can be done during or after treatment to see how effective the treatment has been.

## Radionuclide scans

Scans using small amounts of radioactivity and special cameras may be helpful in looking for carcinoid tumors. They can help find tumors or look for areas of cancer spread if doctors aren't sure where they are in the body.

**Somatostatin receptor scintigraphy:** The most common scan is somatostatin receptor scintigraphy (SRS), also known as the *OctreoScan*. It uses a drug called octreotide bound to radioactive indium-111. Octreotide is a hormone-like substance that attaches to carcinoid cells. A small amount is injected into a vein. It travels through the blood and is attracted to carcinoid tumors. A few hours after the injection, a special camera can be used to show where the radioactivity has collected in the body. More scans may be done in the following few days as well. Along with showing where tumors are, this test can help tell whether treatment with certain drugs such as octreotide and lanreotide is likely to be helpful.

**I-131 MIBG:** This test is used less often. It uses a chemical called MIBG attached to radioactive iodine (I-131). This substance is injected into a vein, and the body is scanned several hours or days later with a special camera to look for areas that picked up the radioactivity. These would most likely be carcinoid tumors, but other kinds of neuroendocrine tumors will also pick up this chemical.

**Positron emission tomography (PET):** For most types of cancer, PET scans use a form of radioactive glucose (sugar) to find tumors. But this type of PET scan is not very useful in finding carcinoid tumors. Instead, PET scanning for carcinoid tumors usually uses a radioactive form of 5-hydroxytryptophan, a chemical that is taken up and used by carcinoid cells. A special camera can detect the radioactivity. The usefulness of this test for lung carcinoid tumors is still being studied. This special type of PET scan is not available in every hospital.

## Sputum cytology

Even if an imaging test such as a chest x-ray or CT scan shows a mass, it's often hard for doctors to tell if the mass is a carcinoid tumor, another type of lung cancer, or an area of infection. Tests may be needed to get a sample of the abnormal cells to be looked at under a microscope.

One way to do this is called *sputum cytology*. A sample of sputum (mucus you cough up from the lungs) is looked at under a microscope to see if it contains cancer cells. The best way to do this is to get samples taken early in the morning, 3 days in a row.

This test is not as good at finding lung carcinoids as it is at finding other types of lung cancers.

## Biopsy

In many cases, the only way to know for sure if a person has some type of lung cancer is to remove cells from the tumor and look at them under a microscope. This procedure is called a *biopsy*. There are several ways to take a sample from a lung tumor.

### **Bronchoscopic biopsy**

This approach is used to view and sample tumors in large airways. The doctor passes a long, thin, flexible, fiber-optic tube called a *bronchoscope* down the throat and through the windpipe and bronchi to look at the lining of the lung's main airways. The mouth and throat are sprayed first with a numbing medicine. You may also be given medicine through an intravenous (IV) line to make you feel relaxed.

If a tumor is found, the doctor can take biopsies (small samples of the tumor) through the tube. The doctor can also sample cells from the lining of the airways by wiping a tiny brush over the surface of the tumor (*bronchial brushing*) or by rinsing the airways with sterile saltwater and then collecting it (*bronchial washing*). Brushing and washing samples are sometimes helpful additions to the bronchial biopsy, but they are not as helpful in diagnosing carcinoids as they are with other lung cancers.

An advantage of this type of biopsy is that no surgery or hospital stay is needed, and you are ready to return home within hours. A disadvantage is that this type of biopsy may not always be able to remove enough of a sample to be certain that a tumor is a carcinoid. But with recent advances in the lab testing of lung tumors, doctors can usually make an accurate diagnosis even with very small samples.

Bleeding from a carcinoid tumor after a biopsy is rare but it can be a 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 aimed through the bronchoscope.

## Endobronchial ultrasonography (EBUS)

If a CT scan shows lymph nodes are enlarged on either side of the trachea or in the area just below where the trachea divides, this test can be used to biopsy these nodes to see if they contain cancer.

Ultrasound is a type of imaging test that uses sound waves to create pictures of the inside of your body. For this test, a small, microphone-like instrument called a *transducer* gives off sound waves and picks up the echoes as they bounce off body tissues. The echoes are converted by a computer into a black and white image on a computer screen.

For endobronchial ultrasound, a bronchoscope is fitted with an ultrasound transducer at its tip and is passed down into the windpipe. This is done with numbing medicine (local anesthesia) and light sedation.

The transducer can be pointed in different directions to look at lymph nodes and other structures in the mediastinum (the area between the lungs). A hollow needle can be passed through the bronchoscope to get biopsy samples of enlarged lymph nodes or other abnormal areas. The samples are then sent to a lab to be looked at under a microscope.

## Needle biopsies

Doctors can often use a hollow needle to get a small sample from a suspicious area (mass). An advantage of needle biopsies is that they don't require a surgical incision, but in some cases they might not provide enough of a sample to make a diagnosis. There are two types of needle biopsies, based on the type of needle used:

- In a *fine needle aspiration (FNA)* biopsy, the doctor uses a syringe with a very thin, hollow needle (thinner than the ones used for blood tests) to withdraw (aspirate) cells and small fragments of tissue.
- In a *core biopsy*, a larger needle is used to remove one or more small cylinders (cores) of tissue. Core biopsies provide a larger sample than FNA biopsies.

If the suspected tumor is in the outer part of the lungs, either kind of biopsy needle can be inserted through the skin on the chest wall. This is called a *transthoracic needle biopsy*. The area where the needle is to be inserted may be numbed with local anesthesia first. The doctor then guides the needle into the area while looking at the lungs with either fluoroscopy (which is like an x-ray, but the image is shown on a screen rather than on film) or CT scans. Unlike fluoroscopy, CT doesn't give a constant picture, so the needle is inserted toward the mass, a CT image is taken, and the direction of the needle is guided based on the image. This is repeated a few times until the needle is within the mass.

A possible complication of this procedure is that air may leak out of the lung at the biopsy site and into the space between the lung and the chest wall. This can cause part of the lung to collapse and could cause trouble breathing. This complication, called a *pneumothorax*, often

gets better without any treatment. If not, it's treated by putting a small tube into the chest space and sucking out the air over a day or two, after which it usually heals on its own.

An FNA biopsy may also be done to check for cancer in the lymph nodes between the lungs. This can be done in two ways:

- *Transtacheal FNA* or *transbronchial FNA* is done by passing the needle through the wall of the trachea (windpipe) or bronchi (the large airways leading into the lungs) during bronchoscopy or EBUS (described above).
- In some cases an FNA biopsy is done during endoscopic esophageal ultrasound (which is similar to EBUS, except that the scope is passed down the esophagus instead of the windpipe) by passing the needle through the wall of the esophagus.

## **Surgical biopsies**

In some cases, the types of biopsies above can't remove enough tissue to identify the type of tumor, and your doctor may need to do surgery to get a biopsy sample. Different types of operations may be used. They are most often done in the operating room while you are under general anesthesia (in a deep sleep).

**Thoracotomy:** For a thoracotomy, the surgeon makes an incision (cut) in the chest wall between the ribs to get to the lungs and to the space between the lungs and the chest wall. In some cases if the doctor strongly suspects a carcinoid or some other type of lung cancer, they may do a thoracotomy and remove the entire tumor without first doing a biopsy.

**Thoracoscopy:** This procedure is also used to look at the space between the lungs and the chest wall, but it does not require a long incision like a thoracotomy. The doctor inserts a thin, lighted scope with a small video camera on the end through a small cut made in the chest wall to look at the outside of the lungs and the space between the lungs and the chest wall. (Sometimes more than one cut is made.) Using this scope, the doctor can see potential areas of cancer and remove small pieces of tissue to look at under the microscope. Thoracoscopy can also be used to sample lymph nodes and fluid and find out whether a tumor is growing into nearby tissues or organs.

**Mediastinoscopy:** This procedure may be done if imaging tests such as a CT scan suggest that the cancer may have spread to the lymph nodes in the mediastinum (the space between the lungs). A small cut is made in the front of the neck and a thin, hollow, lighted tube is inserted behind the sternum (breast bone) and in front of the windpipe to look at the area. Instruments can be passed through this tube to take tissue samples from the lymph nodes along the windpipe and the bronchi.

## Blood and urine tests

Because carcinoid tumors can secrete hormone-like chemicals into the blood, these tumors can sometimes be found with blood or urine tests. This is especially true if you have symptoms of the carcinoid syndrome, which is caused by excess levels of these substances in the blood.

Serotonin is a substance made by some carcinoid tumors, and probably causes some of the symptoms. It is broken down by the body into 5-hydroxyindoleacetic acid (5-HIAA), which is released into the urine. A common test to look for carcinoid syndrome measures the levels of 5-HIAA in a urine sample collected over 24 hours. Measuring the serotonin levels in the blood or urine may also give useful information. These tests can help diagnose some carcinoid tumors, but they are not always accurate. Some other medical conditions, as well as foods and medicines, can affect the results, and some carcinoid tumors may not release enough of these substances to give a positive test result.

Other tests used to look for carcinoids include blood tests for chromogranin A (CgA), neuron-specific enolase (NSE), cortisol, and substance P. Depending on where the tumor might be located and on the patient's symptoms, doctors may do other blood tests as well.

These tests are less likely to be helpful with lung carcinoid tumors than with carcinoid tumors that start elsewhere in the body.

## Pulmonary function tests

If a lung carcinoid is found, pulmonary function tests (PFTs) are often done to see how well your lungs are working. This is especially important if surgery might be used to treat the cancer. Because surgery will remove part or all of the lung, it's important to know how well your lungs are working beforehand. These tests can give the surgeon an idea of whether surgery is a good option, and if so, how much lung can be removed safely.

There are a few different types of PFTs, but they all basically have you breathe in and out through a tube that is connected to a machine that measures airflow.

## How are lung carcinoid tumors staged?

The stage of a cancer describes how far it has spread. Your treatment and prognosis (outlook) depend, to a large extent, on the cancer's stage.

The stage of the cancer is based on the results of the physical exam, imaging tests (CT scan, PET scan, etc.), and other tests, which are described in the section "How are lung carcinoid tumors diagnosed?"

## The TNM staging system

A staging system is a standard way for the cancer care team to sum up how large a cancer is and how far it has spread. The staging system for lung carcinoid tumors is the same one used to stage other types of lung cancer – the American Joint Committee on Cancer (AJCC) **TNM** staging system. The TNM system describes 3 key pieces of information:

- **T** indicates the size of the main (primary) **tumor** and whether it has grown into nearby areas.
- **N** describes the spread of cancer to nearby (regional) lymph **nodes**. Lymph nodes are small bean-shaped collections of immune system cells to which cancers often spread.
- **M** indicates whether the cancer has spread (**metastasized**) to other organs of the body. (The most common site is the liver.)

Numbers or letters appear after T, N, and M to provide more details about each of these factors. The numbers 0 through 4 indicate increasing severity.

The TNM staging system is complex and can be hard for patients (and even some doctors) to understand. If you have any questions about the stage of your cancer, ask your doctor to explain it to you.

### T categories

**TX:** The main (primary) tumor can't be assessed, or cancer cells were seen on sputum cytology or bronchial washings but no tumor can be found.

**T0:** There is no evidence of a primary tumor.

**Tis:** The cancer is found only in the top layers of cells lining the air passages. It has not invaded into deeper lung tissues. This stage is also known as *carcinoma in situ*.

**T1:** The tumor is no larger than 3 centimeters (cm) – slightly less than 1¼ inches – across. It has not reached the membranes that surround the lungs (visceral pleura), and it does not affect the main branches of the bronchi.

If the tumor is 2 cm (about 4/5 of an inch) or less across, it is called **T1a**. If the tumor is larger than 2 cm but not larger than 3 cm across, it is called **T1b**.

**T2:** The tumor has 1 or more of the following features:

- It is larger than 3 cm but not larger than 7 cm across. If the tumor is 5 cm or less across (but still larger than 3 cm), it is called **T2a**. If the tumor is larger than 5 cm across (but not larger than 7 cm), it is called **T2b**.
- It involves a main bronchus, but is not closer than 2 cm (about ¾ inch) to the carina (the point where the windpipe splits into the left and right main bronchi).



- It has grown into the membranes that surround the lungs (visceral pleura).
- The tumor partially clogs the airways, but this has not caused the entire lung to collapse or develop pneumonia.

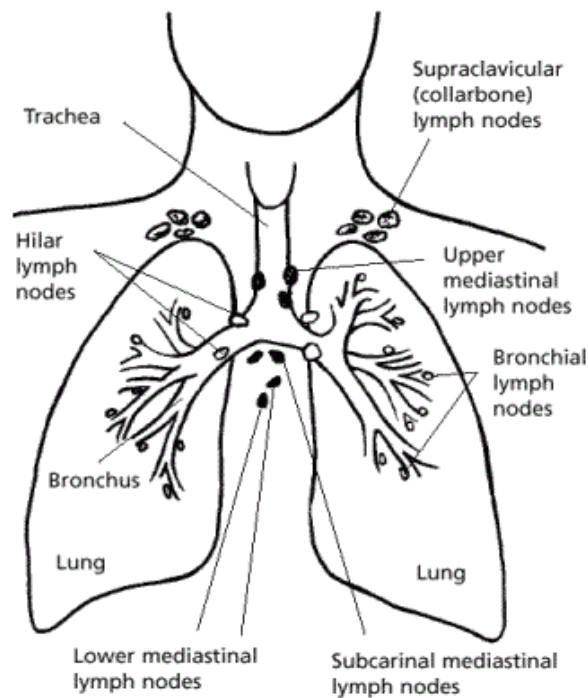
**T3:** The tumor has 1 or more of the following features:

- It is larger than 7 cm across.
- It has grown into the chest wall, the breathing muscle that separates the chest from the abdomen (diaphragm), the membranes surrounding the space between the lungs (mediastinal pleura), or membranes of the sac surrounding the heart (parietal pericardium).
- It invades a main bronchus and is closer than 2 cm (about  $\frac{3}{4}$  inch) to the carina, but it does not involve the carina itself.
- It has grown into the airways enough to cause an entire lung to collapse or to cause pneumonia in the entire lung.
- Two or more separate tumor nodules are present in the same lobe of a lung

**T4:** The cancer has 1 or more of the following features:

- A tumor of any size has grown into the space between the lungs (mediastinum), the heart, the large blood vessels near the heart (such as the aorta), the windpipe, the esophagus (tube connecting the throat to the stomach), the backbone, or the carina.
- Two or more separate tumor nodules are present in different lobes of the same lung.

## N categories



**NX:** Nearby lymph nodes cannot be assessed.

**N0:** There is no spread to nearby lymph nodes.

**N1:** The cancer has spread to lymph nodes within the lung and/or around the area where the bronchus enters the lung (hilar lymph nodes). Affected lymph nodes are on the same side as the primary tumor(s).

**N2:** The cancer has spread to lymph nodes around the carina (the point where the windpipe splits into the left and right bronchi) or to lymph nodes in the space between the lungs (mediastinum) on the same side as the primary tumor.

**N3:** The cancer has spread to lymph nodes near the collarbone on either side, and/or spread to hilar or mediastinal lymph nodes on the side opposite the primary tumor.

## M categories

**M0:** No spread to distant organs or areas. This includes the other lung, lymph nodes further away than those mentioned in the N stages above, and other organs or tissues such as the liver, bones, or brain.

**M1a:** Any of the following:

- The cancer has spread to the other lung.
- Cancer cells are found in the fluid around the lung (called a *malignant pleural effusion*).
- Cancer cells are found in the fluid around the heart (called a *malignant pericardial effusion*).

**M1b:** The cancer has spread to distant lymph nodes or to organs such as the liver, bones, or brain.

## Stage grouping

Once the T, N, and M categories have been determined, this information is combined to assign an overall stage of 0, I, II, III, or IV. This process is called *stage grouping*. Some stages are subdivided into A and B. The stages identify tumors that have a similar outlook (prognosis) and so they are treated in a similar way. Patients with lower stage numbers tend to have a better outlook.

### Occult (hidden) cancer

**TX, N0, M0:** Cancer cells are seen in a sample of sputum or other lung fluids, but the cancer isn't found with other tests, so its location can't be determined.

### Stage 0

**Tis, N0, M0:** The cancer is found only in the top layers of cells lining the air passages. It has not invaded deeper into other lung tissues and has not spread to lymph nodes or distant sites.

### Stage IA

**T1, N0, M0:** The cancer is no larger than 3 cm across, has not reached the membranes that surround the lungs, and does not affect the main branches of the bronchi. It has not spread to lymph nodes or distant sites.

### Stage IB

**T2a, N0, M0:** The cancer has 1 or more of the following features:

- The main tumor is larger than 3 cm across but not larger than 5 cm.
- The tumor involves a main bronchus, but is not within 2 cm of the carina and is not larger than 5 cm.
- The tumor has grown into the visceral pleura (the membranes surrounding the lungs) and is not larger than 5 cm.
- The cancer is partially clogging the airways and is not larger than 5 cm.

The cancer has not spread to lymph nodes or distant sites.

## Stage IIA

Three main combinations of categories make up this stage.

**T1, N1, M0:** The cancer is no larger than 3 cm across, has not grown into the membranes that surround the lungs, and does not affect the main branches of the bronchi. It has spread to lymph nodes within the lung and/or around the area where the bronchus enters the lung (hilar lymph nodes). These lymph nodes are on the same side as the cancer. It has not spread to distant sites.

OR

**T2a, N1, M0:** The cancer has 1 or more of the following features:

- The main tumor is larger than 3 cm across but not larger than 5 cm.
- The tumor has grown into a main bronchus, but is not within 2 cm of the carina and is not larger than 5 cm.
- The tumor has grown into the visceral pleura (the membranes surrounding the lungs) and is not larger than 5 cm.
- The cancer is partially clogging the airways and is not larger than 5 cm.

The cancer has also spread to lymph nodes within the lung and/or around the area where the bronchus enters the lung (hilar lymph nodes). These lymph nodes are on the same side as the cancer. It has not spread to distant sites.

OR

**T2b, N0, M0:** The cancer has 1 or more of the following features:

- The main tumor is larger than 5 cm across but not larger than 7 cm.
- The tumor involves a main bronchus and is between 5 and 7 cm across, but it is not within 2 cm of the carina.
- The tumor has grown into the visceral pleura (the membranes surrounding the lungs) and is between 5 and 7 cm across.
- The cancer is partially clogging the airways and is between 5 and 7 cm across.

The cancer has not spread to lymph nodes or distant sites.

## Stage IIB

Two combinations of categories make up this stage.

**T2b, N1, M0:** The cancer has 1 or more of the following features:

- The main tumor is larger than 5 cm across but not larger than 7 cm.
- The tumor has grown into a main bronchus and is between 5 and 7 cm across, but it is not within 2 cm of the carina.
- The tumor has grown into the visceral pleura (the membranes surrounding the lungs) and is between 5 and 7 cm across.
- The tumor is partially clogging the airways and is between 5 and 7 cm across.

The cancer has also spread to lymph nodes within the lung and/or around the area where the bronchus enters the lung (hilar lymph nodes). These lymph nodes are on the same side as the cancer. It has not spread to distant sites.

OR

**T3, N0, M0:** The main tumor has 1 or more of the following features:

- It is larger than 7 cm across.
- It has grown into the chest wall, the breathing muscle that separates the chest from the abdomen (diaphragm), the membranes surrounding the space between the lungs (mediastinal pleura), or membranes of the sac surrounding the heart (parietal pericardium).
- It invades a main bronchus and is closer than 2 cm (about  $\frac{3}{4}$  inch) to the carina, but it does not involve the carina itself.
- It has grown into the airways enough to cause an entire lung to collapse or to cause pneumonia in the entire lung.
- Two or more separate tumor nodules are present in the same lobe of a lung.

The cancer has not spread to lymph nodes or distant sites.

### **Stage IIIA**

Three main combinations of categories make up this stage.

**T1 to T3, N2, M0:** The tumor can be any size or have any of the following features:

- The tumor involves a main bronchus without growing into the carina.
- The tumor has grown into the visceral pleura (the membranes surrounding the lungs).
- The tumor is partially clogging the airways, or it has grown into the airways enough to cause an entire lung to collapse or to cause pneumonia in the entire lung.
- It has grown into the chest wall, the breathing muscle that separates the chest from the abdomen (diaphragm), the membranes surrounding the space between the two lungs

(mediastinal pleura), or membranes of the sac surrounding the heart (parietal pericardium).

- Two or more separate tumor nodules are present in the same lobe of a lung.

The cancer has also spread to lymph nodes around the carina (the point where the windpipe splits into the left and right bronchi) or in the space between the lungs (mediastinum). These lymph nodes are on the same side as the main lung tumor. The cancer has not spread to distant sites.

OR

**T3, N1, M0:** The tumor has 1 or more of the following features:

- It is larger than 7 cm across.
- It has grown into the chest wall, the breathing muscle that separates the chest from the abdomen (diaphragm), the membranes surrounding the space between the two lungs (mediastinal pleura), or membranes of the sac surrounding the heart (parietal pericardium).
- It invades a main bronchus and is closer than 2 cm to the carina, but it does not involve the carina itself.
- Two or more separate tumor nodules are present in the same lobe of a lung.
- It has grown into the airways enough to cause an entire lung to collapse or to cause pneumonia in the entire lung.

The cancer has also spread to lymph nodes within the lung and/or around the area where the bronchus enters the lung (hilar lymph nodes). These lymph nodes are on the same side as the cancer. It has not spread to distant sites.

OR

**T4, N0 or N1, M0:** The cancer has 1 or more of the following features:

- A tumor of any size has grown into the space between the lungs (mediastinum), the heart, the large blood vessels near the heart (such as the aorta), the windpipe, the esophagus (tube connecting the throat to the stomach), the backbone, or the carina.
- Two or more separate tumor nodules are present in different lobes of the same lung.

The cancer may or may not have spread to lymph nodes within the lung and/or around the area where the bronchus enters the lung (hilar lymph nodes). Any affected lymph nodes are on the same side as the cancer. It has not spread to distant sites.

**Stage IIIB**

Two combinations of categories make up this stage.

**Any T, N3, M0:** The cancer can be of any size. It may or may not have grown into nearby structures or caused pneumonia or lung collapse. It has spread to lymph nodes near the collarbone on either side, and/or has spread to hilar or mediastinal lymph nodes on the side opposite the primary tumor. The cancer has not spread to distant sites.

OR

**T4, N2, M0:** The cancer has 1 or more of the following features:

- A tumor of any size has grown into the space between the lungs (mediastinum), the heart, the large blood vessels near the heart (such as the aorta), the windpipe, the esophagus (tube connecting the throat to the stomach), the backbone, or the carina.
- Two or more separate tumor nodules are present in different lobes of the same lung.

The cancer has also spread to lymph nodes around the carina or in the space between the lungs (mediastinum). Affected lymph nodes are on the same side as the main lung tumor. It has not spread to distant sites.

#### **Stage IV**

Two combinations of categories make up this stage.

**Any T, any N, M1a:** The cancer can be any size and may or may not have grown into nearby structures or reached nearby lymph nodes. In addition, any of the following is true:

- The cancer has spread to the other lung.
- Cancer cells are found in the fluid around the lung (called a *malignant pleural effusion*).
- Cancer cells are found in the fluid around the heart (called a *malignant pericardial effusion*).

OR

**Any T, any N, M1b:** The cancer can be any size and may or may not have grown into nearby structures or reached nearby lymph nodes. It has spread to distant sites, such as the liver.

## **Survival rates for lung carcinoid tumors**

Survival rates are often used by doctors as a standard way of discussing a person's prognosis (outlook). Some patients with cancer may want to know the survival statistics for people in similar situations, while others may not find the numbers helpful, or may even not want to know them. If you do not want to read about survival rates for lung carcinoid tumors, stop reading here and skip to the next section.

The 5-year survival rate refers to the percentage of patients who live *at least 5 years* after their cancer is diagnosed. Of course, many people live much longer than 5 years (and many are cured).

In order to get 5-year survival rates, doctors have to look at people who were treated at least 5 years ago. Improvements in treatment since then may result in a better outlook for people now being diagnosed.

Overall, the 5-year survival rate for patients with typical lung carcinoids is around 85% to 90%, and the 5-year survival rate for patients with atypical lung carcinoids is around 50% to 70%. These ranges reflect different survival rates quoted by several different studies in medical journals.

Lung carcinoids are uncommon tumors, so it's hard to get accurate, up-to-date survival statistics for these cancers based on stage. The numbers below come from a study of more than 1400 people in the United States who were diagnosed with lung carcinoid tumors between 1990 and 2002 and were treated with surgery. They include some people who died from causes other than their cancer.

<b>Stage</b>	<b>5-year Survival Rate</b>
I	93%
II	85%
III	75%
IV	57%

These numbers include people with both typical and atypical carcinoids, but survival rates would be expected to be slightly better for typical carcinoids and not as good overall for atypical carcinoids.

Survival rates are often based on previous outcomes of large numbers of people who had the disease, but they cannot predict what will happen in any particular person's case. Many factors can affect a person's outlook, such as the type of treatment used, how well the cancer responds to treatment, and your general health. Your doctor can tell you how the numbers above might apply to you, as he or she is familiar with your situation.

Even with carcinoids that appear to have been treated successfully, in a small number of cases the cancer can come back (recur) many years later, which is why doctors often advise close follow-up for at least 10 years.



# How are lung carcinoid tumors treated?

*This information represents the views of the doctors and nurses serving on the American Cancer Society's Cancer Information Database Editorial Board. These views are based on their interpretation of studies published in medical journals, as well as their own professional experience.*

*The treatment information in this document is not official policy of the Society and is not intended as medical advice to replace the expertise and judgment of your cancer care team. It is intended to help you and your family make informed decisions, together with your doctor.*

*Your doctor may have reasons for suggesting a treatment plan different from these general treatment options. Don't hesitate to ask him or her questions about your treatment options.*

## Making treatment decisions for lung carcinoid tumors

After the tumor is found and staged, your cancer care team will discuss your treatment options with you. The main factors in selecting a treatment for lung carcinoid tumors are the type of carcinoid, the size and location of the tumor, whether it has spread to lymph nodes or other organs, and if you have any other serious medical conditions. Based on these factors, the main treatment options for people with lung carcinoid tumors can include:

- Surgery
- Chemotherapy
- Other drug treatments
- Radiation therapy

In some cases, more than one of these treatments may be used.

Selecting a treatment plan is an important decision, and you should take the time to think about all of your choices. Be sure to discuss all of your treatment options as well as their possible side effects with your doctors to help make the decision that best fits your needs. (See the section “What should you ask your doctor about lung carcinoid tumors?” for some questions to ask.)

Seeking a second opinion is often a good idea if time permits. It can give you more information and help you feel more confident about the treatment plan you choose.

You may have different types of doctors on your treatment team, depending on the stage of your cancer and your treatment options. These doctors may include:

- A thoracic surgeon: a doctor who treats diseases of the lungs and chest with surgery
- A medical oncologist: a doctor who treats cancer with medicines such as chemotherapy
- A pulmonologist: a doctor who specializes in medical treatment of diseases of the lungs

- A radiation oncologist: a doctor who treats cancer with radiation therapy

Many other specialists may be involved in your care as well, including physician assistants, nurse practitioners, nurses, respiratory therapists, social workers, and other health professionals.

The next few sections describe the various types of treatments used for lung carcinoids. This is followed by a description of the most common approaches based on the type and extent of the disease.

## **Surgery and other procedures for lung carcinoid tumors**

Surgery is the main treatment for lung carcinoid tumors whenever possible. If the tumor has not spread, it can often be cured by surgery alone.

Several types of surgery are used to treat people with lung carcinoid tumors. The type of surgery will depend on a number of factors, including the size and location of the tumor and whether you have any other lung problems or serious diseases. Thoracic and cardiothoracic surgeons are likely to have the most experience with these operations.

Surgeons usually have to remove some normal lung tissue along with the tumor, but they try not to remove any more normal tissue than they need to.

These operations require general anesthesia (where you are in a deep sleep) and often require a surgical incision between the ribs in the chest (thoracotomy). You will generally need to spend about 5 to 7 days in the hospital after the surgery.

### **Sleeve resection**

To treat a central carcinoid in a large airway (such as a bronchus), the surgeon may do a sleeve resection. If you think of the large airway with a tumor to be like the sleeve of a shirt with a stain an inch or 2 above the wrist, the sleeve resection would be like cutting across the sleeve above and below the stain and sewing the cuff back onto the shortened sleeve. The surgeon may be able to do this type of operation instead of removing part or all of the lung, in order to preserve more lung function.

### **Wedge resection**

For small carcinoids found at the outer edges of the lungs away from the large airways, the surgeon may remove a wedge-shaped piece of the lung in an operation called a wedge resection or segmental resection.

## **Lobectomy**

If it's not possible to do a wedge resection because of the size or location of the tumor, the surgeon will usually do a lobectomy, in which an entire lobe of the lung is removed. In some cases 2 lobes may be removed (bilobectomy).

## **Pneumonectomy**

In rare cases where the cancer is in many spots in a lung or is in a place that makes it hard to remove, the entire left or right lung may need to be removed in an operation called a pneumonectomy.

## **Lymph node sampling**

With any of these operations, lymph nodes near the lungs are usually removed to look for possible spread of the cancer. This is important because the carcinoid might have spread to lymph nodes by the time it is diagnosed. (This risk is higher for atypical carcinoids than for typical carcinoids.) If lymph nodes containing cancer are not removed, it will increase the risk of the carcinoid tumor spreading even farther, to other organs. If this happens, you may no longer be able to be cured by surgery. Checking for cancer cells in the lymph nodes can also provide some indication of your risk of having the cancer come back.

## **Video-assisted thoracic surgery (VATS)**

This is a less invasive type of surgery for treating some cancers in the lungs. During this operation, a thin, rigid tube with a tiny video camera on the end is placed through a small cut in the side of the chest to help the surgeon see inside the chest. One or two other small cuts are created in the skin, and long instruments are passed through these cuts to do the same operation that would be done using an open approach (thoracotomy). Because only small incisions are needed, there is less pain after the surgery and a shorter hospital stay – usually around 4 to 5 days.

Most experts recommend that only smaller tumors near the outside of the lung be treated with this method. The cure rate after this surgery seems to be the same as with surgery done with a larger incision. But it is important that the surgeon doing this operation be experienced because it requires a great deal of technical skill.

## **Possible risks and side effects of lung surgery**

Possible complications depend on the extent of the surgery and the person's health beforehand. Serious complications can include excessive bleeding, wound infections, and pneumonia.

Lung surgery is a major operation, and recovering from the operation typically takes weeks to months. If the surgery is done through a thoracotomy, the surgeon must spread the ribs to get to the lung, so the area near the incision will hurt for some time after surgery. Your activity will be limited for at least a month or two. People who have VATS instead of thoracotomy have less pain after surgery and tend to recover more quickly.

If your lungs are in good condition (other than the presence of the cancer) you can usually return to normal activities after a lobe or even an entire lung has been removed. If you also have non-cancerous diseases such as emphysema or chronic bronchitis (which are common among heavy smokers), you may become short of breath with activity after surgery.

## **Procedures to relieve symptoms from lung carcinoid tumors**

If you can't have major surgery because you have reduced lung function or other serious medical problems, or if the cancer has spread too far to be removed, other treatments may be used to relieve some symptoms.

These treatments, called *palliative procedures*, can relieve symptoms, but they do not cure the cancer and are recommended only if you cannot have surgery to completely remove the tumor. If you are treated with these procedures you may also get radiation therapy.

### **Treating airway blockage**

If the tumor is blocking airways in the lung, it might lead to pneumonia or shortness of breath. Removing most of the tumor through a bronchoscope or destroying most of it with a laser (on the end of a bronchoscope) can be helpful. In some cases, a bronchoscope may be used to place a stent (a stiff tube) made of metal or silicone in the airway after treatment to help keep it open.

### **Treating fluid buildup**

Sometimes fluid can build up inside the chest (outside of the lungs), which can press on the lungs and affect breathing. Usually, a hollow needle is put through the skin and into the pleural space to remove the fluid. (This is known as a *thoracentesis*.) Removing the fluid can relieve breathing problems right away in most patients, but the fluid will often build up again quickly if nothing else is done.

**Pleurodesis:** To remove the fluid and keep it from coming back, doctors sometimes do a procedure called *pleurodesis*. A small cut is made in the skin of the chest wall, and a hollow tube is placed into the chest to remove the fluid. Either talc or a drug such as doxycycline or a chemotherapy drug is then instilled into the chest cavity. This causes the linings of the lung (visceral pleura) and chest wall (parietal pleura) to stick together, sealing the space and limiting further fluid buildup. The tube is often left in for a day or two to drain any new fluid that might collect.

**Catheter placement:** This is another way to control the buildup of fluid. One end of the catheter (a thin, flexible tube) is placed in the chest through a small cut in the skin, and the

other end is left outside the body. This is done in a doctor's office or hospital. Once in place, the catheter can be attached to a special bottle or other device to allow the fluid to drain out on a regular basis.

## **Procedures to relieve symptoms of liver metastases**

If the cancer spreads to the liver, treating the liver tumors may help with symptoms. When there are only 1 or 2 tumors in the liver, they may be removed with surgery. If there are more than just a few liver tumors (or if a person is too sick for surgery), other techniques may be used.

### **Ablation**

Ablation techniques destroy tumors without removing them. They are generally not used for large tumors, and are best for tumors no more than about 2 cm (a little less than an inch) across.

**Radiofrequency ablation (RFA)** uses high-energy radio waves for treatment. A thin, needle-like probe is placed through the skin and into the tumor. Placement of the probe is guided by ultrasound or CT scans. The tip of the probe releases a high-frequency current that heats the tumor and destroys the cancer cells.

**Ethanol (alcohol) ablation** (also known as *percutaneous ethanol injection*) kills the cancer cells by injecting concentrated alcohol directly into the tumor. This is usually done through the skin using a needle guided by ultrasound or CT scans.

**Microwave thermotherapy** uses microwaves to heat and destroy the abnormal tissue.

**Cryosurgery (cryotherapy)** destroys a tumor by freezing it with a metal probe. The probe is guided through the skin and into the tumor using ultrasound. Then very cold gasses are passed through the probe to freeze the tumor, killing the cancer cells. This method may be used to treat larger tumors than the other ablation techniques, but it sometimes requires general anesthesia (where you are asleep).

### **Embolization**

**Arterial embolization**, also known as *transarterial embolization* (or TAE), is another option for tumors that cannot be removed. Embolization can be used for larger tumors – up to about 5 cm (2 inches) across – than the ablative methods. This technique is used to reduce the blood flow to the cancer cells by blocking the branch of the hepatic artery feeding the area of the liver containing the tumor. Blood flow is blocked (or reduced) by injecting materials that plug up the artery. Most of the healthy liver cells will not be affected because they get their blood supply from the portal vein.

In this procedure a catheter is put into an artery in the inner thigh and threaded up into the liver. A dye is usually injected into the bloodstream at this time to allow the doctor to

monitor the path of the catheter via angiography, a special type of x-ray. Once the catheter is in place, small particles called *microspheres* are injected into the artery to plug it up.

**Radioembolization** combines embolization with radiation therapy. In the United States, this is done by injecting small radioactive beads into the hepatic artery. The beads travel to the tumor and give off small amounts of radiation only at the tumor sites.

For more general information about surgery, please see our document *Understanding Cancer Surgery: A Guide for Patients and Families*.

## Chemotherapy for lung carcinoid tumors

Chemotherapy (chemo) is the use of anti-cancer drugs that are injected into a vein 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 have spread to organs beyond the lungs.

Unfortunately, carcinoid tumors usually do not respond very well to chemo. It is mainly used for carcinoid tumors that have spread to other organs, are causing severe symptoms, and have not responded to other medicines. In some cases it may be given after surgery.

Because chemo does not always shrink carcinoid tumors, it is important to ask your doctors about the chances of it helping and if the benefits are likely to outweigh the risk of side effects.

Some of the chemo drugs that may be used for advanced lung carcinoids include:

- Streptozocin
- Etoposide (VP-16)
- Cisplatin
- Temozolomide
- Cyclophosphamide (Cytosan<sup>®</sup>)
- 5-fluorouracil (5-FU)
- Doxorubicin (Adriamycin<sup>®</sup>)
- Dacarbazine (DTIC)

In most cases, chemo drugs are used together, often along with other types of medicines.

Doctors give chemo in cycles, with each period of treatment followed by a rest period to allow the body time to recover. Chemo cycles generally last about 3 to 4 weeks, and initial treatment typically involves 4 to 6 cycles. Chemo is often not recommended for patients in poor health, but advanced age by itself is not a barrier to getting chemo.

## Possible side effects of chemotherapy

Chemo drugs attack cells that are dividing quickly, which is why they work against cancer cells. But other cells in the body, such as those in the bone marrow (where new blood cells are made), the lining of the mouth and intestines, and the hair follicles, also divide quickly. These cells are also likely to be affected by chemo, which can lead to side effects.

The side effects of chemo depend on the type and dose of drugs given and the length of time they are taken. Common side effects can include:

- Hair loss
- Mouth sores
- Loss of appetite
- Nausea and vomiting
- Diarrhea or constipation
- Increased chance of infections (from having too few white blood cells)
- Easy bruising or bleeding (from having too few blood platelets)
- Fatigue (from having too few red blood cells)

These side effects are usually short-term and go away after treatment is finished. There are often ways to lessen these side effects or keep them from occurring. For example, drugs can be given to help prevent or reduce nausea and vomiting.

Some drugs can have other side effects. For example, cisplatin can damage nerve endings (a condition called *neuropathy*). This may lead to symptoms (mainly in the hands and feet) such as pain, burning or tingling sensations, sensitivity to cold or heat, or weakness. In most cases this goes away once treatment is stopped, but it may last a long time in some people. For more information, see our document *Peripheral Neuropathy Caused by Chemotherapy*.

You should report any side effects or changes you notice while getting chemotherapy to your medical team so that they can be treated promptly. In some cases, the doses of the chemo drugs may need to be reduced or treatment may need to be delayed or stopped to prevent the effects from getting worse.

For more general information about chemotherapy, please see the “Chemotherapy” section of our website, or our document *Understanding Chemotherapy: A Guide for Patients and Families*.

## Other drugs for treating lung carcinoid tumors

For people with metastatic lung carcinoid tumors, several medicines can help control symptoms and may help keep the tumor from growing for a time.

### **Somatostatin analogs**

These drugs are related to somatostatin, a natural hormone that seems to help slow the growth of neuroendocrine cells. They are especially useful in people who have carcinoid syndrome (facial flushing, diarrhea, wheezing, rapid heart rate) and in people whose tumors show up on a somatostatin receptor scintigraphy (SRS) scan.

**Octreotide:** This drug is very helpful in treating the symptoms of carcinoid syndrome. Sometimes octreotide can temporarily shrink carcinoid tumors, but it does not cure them. Side effects can include pain or burning at the injection site, stomach cramps, nausea, vomiting, headaches, dizziness, and fatigue.

The original version of octreotide (Sandostatin<sup>®</sup>) is given as an injection under the skin (subcutaneously) at least twice daily. Some people can learn to give this injection themselves at home. A newer, long-acting version of the drug (Sandostatin LAR<sup>®</sup>) is given as an injection into a muscle once a month by your doctor or nurse. When first starting treatment, most people are given injections every day. Once the doctor finds the correct dose, the longer-acting monthly injection may be used.

**Lanreotide:** Lanreotide (Somatuline<sup>®</sup>) is a drug similar to octreotide. It is given as an injection under the skin once a month. It may be given by your doctor or nurse, or you may learn how to give the injection at home. Side effects are similar to those of octreotide, although pain at the injection site is less common.

### **Interferons**

These drugs are natural substances in the body that normally help activate the immune system. They also suppress the growth of some tumors. Interferon alfa is sometimes helpful in shrinking or slowing the growth of metastatic carcinoid tumors and improving symptoms of carcinoid syndrome. But its usefulness is limited by its flu-like side effects, which can be severe. It can also cause depression. Interferon alfa is given by injection, either daily or several times a week.

### **Targeted drugs**

In recent years, anti-cancer drugs that work differently from standard chemotherapy drugs have been developed for some types of cancer. These drugs target specific parts of cancer cells. They are sometimes helpful when chemotherapy is not, and they often have less severe side effects.



Two targeted drugs, sunitinib (Sutent<sup>®</sup>) and everolimus (Afinitor<sup>®</sup>), have been shown to help treat neuroendocrine tumors that start in the pancreas. These drugs may also be helpful against carcinoid tumors, which are a type of neuroendocrine tumor. Studies are now in progress to try to prove this, but some doctors are already using these drugs for carcinoid tumors.

Other medicines can be used to help control specific symptoms. It is important to describe your symptoms to your doctor so that they can be treated effectively.

## Radiation therapy for lung carcinoid tumors

Radiation therapy is the use of high-energy rays (such as x-rays) or radioactive particles to kill cancer cells. Unfortunately, radiation therapy usually has only a limited effect on lung carcinoid tumors.

Surgery is the main treatment for most carcinoid tumors, but radiation therapy may be an option for those who can't have surgery for some reason. It may also be given after surgery in some cases if there's a chance some of the tumor was not removed. Radiation therapy can also be used to help relieve symptoms such as pain if the cancer has spread to the bones or other areas.

### **External beam radiation therapy**

External beam radiation therapy uses a machine that delivers a beam of radiation to a specific part of the body. This is the type of radiation used most often for lung carcinoid tumors.

Before your treatments start, the radiation team will determine the correct angles for aiming the radiation beams and the proper dose of radiation. Treatment is much like getting an x-ray, but the radiation dose is stronger. The procedure itself is painless. Each treatment lasts only a few minutes, but the setup time – getting you into place for treatment – usually takes longer. Most often, radiation treatments are given 5 days a week for several weeks, but this can vary based on the reason it's being given.

The main side effects of lung radiation therapy are fatigue (tiredness) and temporary sunburn-like skin changes where the radiation passed through the skin. If high doses are given, radiation therapy can cause scar tissue to form in the lungs over time, which might lead to trouble breathing and an increased risk of pneumonia.

### **Radioactive drugs**

Drugs containing radioactive particles may be useful in treating some widespread carcinoid tumors. For this type of treatment, doctors use some of the same drugs used in radionuclide scans (see the section "How are lung carcinoid tumors diagnosed?"), such as MIBG and octreotide, but they are attached to more strongly radioactive particles than are used in the scans. Once injected into the body, these drugs attach to carcinoid tumor cells. This lets

doctors deliver high doses of radiation directly to the tumors. Some early results have been promising, but this type of treatment is not widely used at this time (see “What’s new in lung carcinoid tumor research and treatment?”).

For more general information about radiation therapy, please see the “Radiation Therapy” section of our website, or our document *Understanding Radiation Therapy: A Guide for Patients and Families*.

## Clinical trials for lung carcinoid tumors

You may have had to make a lot of decisions since you’ve been told you have cancer. One of the most important decisions you will make is choosing which treatment is best for you. You may have heard about clinical trials being done for your type of cancer. Or maybe someone on your health care team has mentioned a clinical trial to you.

Clinical trials are carefully controlled research studies that are done with patients who volunteer for them. They are done to get a closer look at promising new treatments or procedures.

Clinical trials are one way to get state-of-the-art cancer treatment. Sometimes they may be the only way to get access to some newer treatments. They are also the only way for doctors to learn better methods to treat cancer. Still, they are not right for everyone.

If you would like to learn more about clinical trials that might be right for you, start by asking your doctor if your clinic or hospital conducts clinical trials. You can also call our clinical trials matching service for a list of clinical trials that meet your medical needs. You can reach this service at 1-800-303-5691 or on our website at [www.cancer.org/clinicaltrials](http://www.cancer.org/clinicaltrials). You can also get a list of current clinical trials by calling the National Cancer Institute’s Cancer Information Service toll-free at 1-800-4-CANCER (1-800-422-6237) or by visiting the NCI clinical trials website at [www.cancer.gov/clinicaltrials](http://www.cancer.gov/clinicaltrials).

There are requirements you must meet to take part in any clinical trial. If you do qualify for a clinical trial, it’s up to you whether or not to enter (enroll in) it.

You can get a lot more information on clinical trials in our document *Clinical Trials: What You Need to Know*. You can read it on our website or call us at 1-800-227-2345 to have it sent to you.

## Complementary and alternative therapies for lung carcinoid tumors

When you have cancer you are likely to hear about ways to treat your cancer or relieve symptoms that your doctor hasn’t mentioned. Everyone from friends and family to Internet groups and websites may offer ideas for what might help you. These methods can include

vitamins, herbs, and special diets, or other methods such as acupuncture or massage, to name a few.

## **What exactly are complementary and alternative therapies?**

Not everyone uses these terms the same way, and they are used to refer to many different methods, so it can be confusing. We use *complementary* to refer to treatments that are used *along with* your regular medical care. *Alternative* treatments are used *instead of* a doctor's medical treatment.

**Complementary methods:** Most complementary treatment methods are not offered as cures for cancer. Mainly, they are used to help you feel better. Some methods that are used along with regular treatment are meditation to reduce stress, acupuncture to help relieve pain, or peppermint tea to relieve nausea. Some complementary methods are known to help, while others have not been tested. Some have been proven to not be helpful, and a few have even been found to be harmful.

**Alternative treatments:** Alternative treatments may be offered as cancer cures. These treatments have not been proven safe and effective in clinical trials. Some of these methods may pose danger, or have life-threatening side effects. But the biggest danger in most cases is that you may lose the chance to be helped by standard medical treatment. Delays or interruptions in your medical treatments may give the cancer more time to grow and make it less likely that treatment will help.

## **Finding out more**

It is easy to see why people with cancer think about alternative methods. You want to do all you can to fight the cancer, and the idea of a treatment with few or no side effects sounds great. Sometimes medical treatments like chemotherapy can be hard to take, or they may no longer be working. But the truth is that most of these alternative methods have not been tested and proven to work in treating cancer.

As you consider your options, here are 3 important steps you can take:

- Look for “red flags” that suggest fraud. Does the method promise to cure all or most cancers? Are you told not to have regular medical treatments? Is the treatment a “secret” that requires you to visit certain providers or travel to another country?
- Talk to your doctor or nurse about any method you are thinking about using.
- Contact us at 1-800-227-2345 to learn more about complementary and alternative methods in general and to find out about the specific methods you are looking at. You can also check them out in the “Complementary and Alternative Medicine” section of our website.

## **The choice is yours**

Decisions about how to treat or manage your cancer are always yours to make. If you want to use a non-standard treatment, learn all you can about the method and talk to your doctor about it. With good information and the support of your health care team, you may be able to safely use the methods that can help you while avoiding those that could be harmful.

## **Treatment of lung carcinoid by type and extent of disease**

The treatment of lung carcinoid tumors depends to a large extent on the type (typical versus atypical) and extent of the cancer. Other factors, such as a person's overall health and ability to withstand surgery, are also important.

Many doctors use the TNM staging system (see the section "How are lung carcinoid tumors staged?") to formally describe the extent of these cancers. But for treatment purposes most doctors use a simpler system, dividing these tumors into 2 groups: those that can be treated with surgery (resectable cancers) and those that can't be removed completely (unresectable cancers).

### **Resectable carcinoid tumors**

Resectable carcinoid tumors haven't spread far beyond where they started and can be removed completely. In the TNM staging system, this includes most stage I, II, and IIIA cancers.

These cancers are treated with surgery. The extent of the surgery depends on the type of carcinoid tumor and the size and location of the cancer. Atypical carcinoids may need more extensive surgery than typical carcinoids. Nearby lymph nodes are usually removed as well, especially if you have an atypical carcinoid.

Most patients with resectable lung carcinoid tumors are cured with surgery alone and don't need other treatments. Some experts recommend additional treatment for people with an atypical carcinoid that has spread to lymph nodes. This can be chemotherapy, radiation therapy, or both. But it's not clear if the added therapy lowers the chance of the cancer coming back, or if it helps people live longer.

### **Unresectable carcinoid tumors**

Unresectable carcinoid tumors include those that have grown too much or spread too far to be removed completely by surgery (including most stage IIIB and stage IV cancers), as well as tumors in people who are not healthy enough for surgery.

Treatment depends on where the cancer is and whether you have symptoms of the carcinoid syndrome. In general, this is a slow-growing cancer, and chemotherapy has not proven to be

very successful. If you have only a small number of tumors that can be removed, surgery (on both the lung and at the site of metastasis) is likely to be your best option.

Lung carcinoid tumors usually spread to the liver first. If the carcinoid has spread only to your liver but can't be removed there with standard surgery, another option might be to have a liver transplant. This is a very involved operation that most doctors still consider experimental. It is done at only a few transplant centers.

If the carcinoid is in your liver and is causing symptoms, procedures such as ablation or hepatic artery embolization may be helpful. They may relieve symptoms or slow the growth of the cancer, but are very unlikely to result in a cure. These treatments are discussed in detail in the section on surgery.

If tumors in your liver are too large or numerous to be treated directly, or if the carcinoid has spread to other parts of your body, then drug therapy can be helpful. The drugs octreotide and lanreotide, which can slow the growth of the cancer and stop the secretion of the chemicals that cause the carcinoid syndrome, are often helpful. Sometimes these drugs can actually shrink the cancers. Another drug, interferon alfa, can act the same way. Sometimes these 2 types of drugs are given together. Newer targeted drugs such as sunitinib (Sutent) and everolimus (Afinitor) may be helpful as well, although this still needs to be proven. Adding chemotherapy may also help reduce symptoms, but it seldom shrinks the tumor much more.

For people with earlier stage cancers who can't have surgery, most doctors recommend radiation therapy for typical carcinoids and chemotherapy plus radiation therapy for atypical carcinoids.

External radiation therapy can also be used to relieve symptoms caused by tumors such as bone pain. For more widespread disease, radioactive drugs may be helpful.

## **Recurrent carcinoid tumors**

When cancer comes back after treatment, it is called a *recurrence*. Recurrence can be local (in or near the same place it started) or distant (spread to organs such as the liver or bone).

Carcinoid tumors can sometimes come back, even several years after the initial treatment. If this happens, further treatment options depend on where the cancer is and what treatments have already been used. Cancers that recur locally or in only 1 or 2 areas can sometimes be treated with further surgery. If surgery is not an option, radiation therapy, chemotherapy, or other drugs may be tried. Because recurrent cancers can often be hard to treat, clinical trials of new types of treatment may be a good option.

## **More treatment information for lung carcinoid tumors**

For more details on treatment options – including some that may not be addressed in this document – the National Comprehensive Cancer Network (NCCN) and the National Cancer Institute (NCI) are good sources of information.

The NCCN, made up of experts from many of the nation's leading cancer centers, develops cancer treatment guidelines for doctors to use when treating patients. These are available on the NCCN website ([www.nccn.org](http://www.nccn.org)). (Information on treating lung carcinoid tumors can be found in the Small Cell Lung Cancer guideline.)

The NCI provides treatment guidelines via its telephone information center (1-800-4-CANCER) and its website ([www.cancer.gov](http://www.cancer.gov)). Detailed guidelines intended for use by cancer care professionals are also available on [www.cancer.gov](http://www.cancer.gov).

## **What should you ask your doctor about lung carcinoid tumors?**

It is important to have honest, open discussions with your cancer care team. You should feel free to ask any question, no matter how small it might seem. Nurses, social workers, and other members of the treatment team may also be able to answer many of your questions. Here are some questions you might want to ask:

- What kind of carcinoid tumor do I have?
- Has my carcinoid tumor spread beyond the lungs?
- What is the stage (extent) of my carcinoid tumor, and what does that mean in my case?
- Are there other tests that need to be done before we can decide on treatment?
- Are there other doctors I need to see?
- How much experience do you have treating this type of cancer?
- Should I get a second opinion?
- What treatment choices do I have?
- What do you recommend and why?
- What risks or side effects are there to the treatments you suggest?
- How quickly do we need to decide on treatment?
- What should I do to be ready for treatment?
- How long will treatment last? What will it be like? Where will it be done?
- How will treatment affect my daily activities?
- What are the chances of my carcinoid tumor coming back with these treatment plans?

- What would we do if the treatment doesn't work or if the cancer comes back?
- What type of follow-up might I need after treatment?

Along with these sample questions, be sure to write down some of your own. For instance, you might want more information about recovery time so that you can plan your work schedule. Or you may want to ask about clinical trials for which you may qualify. You can find more information about communicating with your health care team in our document *Talking With Your Doctor*.

## What happens after treatment for lung carcinoid tumors?

For many people with carcinoid tumors, treatment may remove or destroy the cancer. Completing treatment can be both stressful and exciting. You may be relieved to finish treatment, but find it hard not to worry about cancer coming back. (When cancer comes back after treatment, it is called *recurrence*.) This is a very common concern in people who have had cancer.

It may take a while before your fears lessen. But it may help to know that many cancer survivors have learned to live with this uncertainty and are living full lives. Our document *Living With Uncertainty: The Fear of Cancer Recurrence* gives more detailed information on this.

For other people, the cancer may never go away completely. These people may get regular treatments with chemotherapy, radiation therapy, or other therapies to try to help keep the cancer in check. Learning to live with cancer that does not go away can be difficult and very stressful. It has its own type of uncertainty. Our document *When Cancer Doesn't Go Away* talks more about this.

### Follow-up care

If you have completed treatment, your doctors will still want to watch you closely. It is very important to go to all of your follow-up appointments. During these visits, your doctors will ask about any problems you are having and may do exams, lab tests, or imaging tests (such as x-rays or CT scans) to look for signs of cancer or treatment side effects.

Your doctor will most likely want to see you fairly often (every couple of months or so) at first. The time between visits may be extended if there are no problems. Lung carcinoid tumors are often cured by the initial treatment, but sometimes they can come back (recur) many years later, which is why doctors often advise close follow-up for at least 10 years.

Almost any cancer treatment can have side effects. Some may last for a few weeks or months, but others can last the rest of your life. During your doctor visits, talk to your cancer

care team about any changes or problems you notice and let them know about any questions or concerns you have.

It's important to keep health insurance. Tests and doctor visits cost a lot, and even though no one wants to think of their cancer coming back, this could happen.

If the cancer does recur at some point, further treatment will depend on the type and location of the cancer, what treatments you've had before, and your health. For more information on how recurrent cancer is treated, see the section "Treatment of lung carcinoid by type and extent of disease." For more general information on dealing with a recurrence, you may also want to see our document *When Your Cancer Comes Back: Cancer Recurrence*. It can be read online, or call us at 1-800-227-2345 to have a free copy sent to you.

## Seeing a new doctor

At some point after your cancer diagnosis and treatment, you may find yourself seeing a new doctor who does not know about your medical history. It's important that you be able to give your new doctor the details of your diagnosis and treatment. Gathering these details during and soon after treatment may be easier than trying to get them at some point in the future. Make sure you have the following information handy:

- A copy of your pathology report(s) from any biopsies or surgeries
- If you had surgery, a copy of your operative report(s)
- If you stayed in the hospital, a copy of the discharge summary that doctors prepare when patients are sent home
- If you had radiation therapy, a copy of your treatment summary
- If you had chemotherapy or other medicines, a list of your drugs, drug doses, and when you took them
- A copy of your x-rays or other imaging tests (which can often be stored digitally on a DVD, etc.)

## Lifestyle changes after lung carcinoid tumor

You can't change the fact that you have had cancer. What you can change is how you live the rest of your life – making choices to help you stay healthy and feel as well as you can. This can be a time to look at your life in new ways. Maybe you are thinking about how to improve your health over the long term. Some people even start during cancer treatment.



## **Making healthier choices**

For many people, a diagnosis of cancer helps them focus on their health in ways they may not have thought much about in the past. Are there things you could do that might make you healthier? Maybe you could try to eat better or get more exercise. Maybe you could cut down on alcohol, or give up tobacco. Even things like keeping your stress level under control may help. Now is a good time to think about making changes that can have positive effects for the rest of your life. You will feel better and you will also be healthier.

You can start by working on those things that worry you most. Get help with those that are harder for you. For instance, if you are thinking about quitting smoking and need help, call the American Cancer Society for information and support at 1-800-227-2345. A tobacco cessation and coaching service can help increase your chances of quitting for good.

## **Eating better**

Eating right can be hard for anyone, but it can get even tougher during and after cancer treatment. Treatment may change your sense of taste. Nausea can be a problem. You may not feel like eating and lose weight when you don't want to. Or you may have gained weight that you can't seem to lose. All of these things can be very frustrating.

If treatment caused weight changes or eating or taste problems, do the best you can and keep in mind that these problems usually get better over time. You may find it helps to eat small portions every 2 to 3 hours until you feel better. You may also want to ask your cancer team about seeing a dietitian, an expert in nutrition who can give you ideas on how to deal with these treatment side effects.

One of the best things you can do after cancer treatment is put healthy eating habits into place. You may be surprised at the long-term benefits of some simple changes, like increasing the variety of healthy foods you eat. Getting to and staying at a healthy weight, eating a healthy diet, and limiting your alcohol intake may lower your risk for a number of types of cancer, as well as having many other health benefits.

You can get more information in our document *Nutrition and Physical Activity During and After Cancer Treatment: Answers to Common Questions*.

## **Rest, fatigue, and exercise**

Extreme tiredness, called fatigue, is very common in people treated for cancer. This is not a normal tiredness, but a bone-weary exhaustion that often doesn't get better with rest. For some people, fatigue lasts a long time after treatment, and can make it hard for them to exercise and do other things they want to do. But exercise can help reduce fatigue. Studies have shown that patients who follow an exercise program tailored to their personal needs feel better physically and emotionally and can cope better, too.

If you were sick and not very active during treatment, it is normal for your fitness, endurance, and muscle strength to decline. Any plan for physical activity should fit your own situation. An older person who has never exercised will not be able to take on the same amount of exercise as a 20-year-old who plays tennis twice a week. If you haven't been active in a few years, you will have to start slowly – maybe just by taking short walks.

Talk with your health care team before starting anything. Get their opinion about your exercise plans. Then, try to find an exercise buddy so you're not doing it alone. Having family or friends involved when starting a new activity program can give you that extra boost of support to keep you going when the push just isn't there.

If you are very tired, you will need to balance activity with rest. It is OK to rest when you need to. Sometimes it's really hard for people to allow themselves to rest when they are used to working all day or taking care of a household, but this is not the time to push yourself too hard. Listen to your body and rest when you need to. (For more information on dealing with fatigue and other treatment side effects, please see the "Physical Side Effects" section of our website or "Additional resources for lung carcinoid" section.)

Keep in mind exercise can improve your physical and emotional health.

- It improves your cardiovascular (heart and circulation) fitness.
- Along with a good diet, it will help you get to and stay at a healthy weight.
- It makes your muscles stronger.
- It reduces fatigue and helps you have more energy.
- It can help lower anxiety and depression.
- It can make you feel happier.
- It helps you feel better about yourself.

And long term, we know that getting regular physical activity plays a role in helping to lower the risk of some cancers, as well as having other health benefits.

### **Can I lower my risk of the carcinoid tumor progressing or coming back?**

Most people want to know if there are specific lifestyle changes they can make to reduce their risk of cancer progressing or coming back. Unfortunately, for most cancers there is little solid evidence to guide people. This doesn't mean that nothing will help – it's just that for the most part this is an area that hasn't been well studied. Most studies have looked at lifestyle changes as ways of preventing cancer in the first place, not slowing it down or preventing it from coming back.

At this time, not enough is known about lung carcinoid tumors to say for sure if there are things you can do that will be helpful. Adopting healthy behaviors such as not smoking,

eating well, being active, and staying at a healthy weight may help, but no one knows for sure. However, we do know that these types of changes can have positive effects on your health that can extend beyond your risk of carcinoid tumors or other cancers.

## How might having a lung carcinoid tumor affect your emotional health?

During and after treatment, you may find yourself overcome with many different emotions. This happens to a lot of people.

You may find yourself thinking about death and dying. Or maybe you're more aware of the effect the cancer has on your family, friends, and career. You may take a new look at your relationships with those around you. Unexpected issues may also cause concern. For instance, you might be stressed by financial concerns resulting from your treatment. You might also see your health care team less often after treatment and have more time on your hands. These changes can make some people anxious.

Almost everyone who is going through or has been through cancer can benefit from getting some type of support. You need people you can turn to for strength and comfort. Support can come in many forms: family, friends, cancer support groups, church or spiritual groups, online support communities, or one-on-one counselors. What's best for you depends on your situation and personality. Some people feel safe in peer-support groups or education groups. Others would rather talk in an informal setting, such as church. Others may feel more at ease talking one-on-one with a trusted friend or counselor. Whatever your source of strength or comfort, make sure you have a place to go with your concerns.

The cancer journey can feel very lonely. It is not necessary or good for you to try to deal with everything on your own. And your friends and family may feel shut out if you do not include them. Let them in, and let in anyone else who you feel may help. If you aren't sure who can help, call your American Cancer Society at 1-800-227-2345 and we can put you in touch with a group or resource that may work for you. You can also read our document *Distress in People with Cancer* or see the "Emotional Side Effects" section of our website for more information.

## If lung carcinoid tumor treatment stops working

If cancer keeps growing or comes back after one kind of treatment, it is possible that another treatment plan might still control the cancer, or at least keep it in check enough to help you live longer and feel better. But when a person has tried many different treatments and the cancer has not gotten any better, even newer treatments might no longer be effective. If this happens, it's important to weigh the possible limited benefits of trying a new treatment against the possible downsides, including treatment side effects. Everyone has their own way of looking at this.

This is likely to be the hardest part of your battle with cancer – when you have been through many treatments and nothing’s working anymore. Your doctor might offer you new options, but at some point you may need to consider that treatment is not likely to improve your health or change your outcome or survival.

If you want to continue to get treatment for as long as you can, you need to think about the odds of treatment having any benefit and how this compares to the possible risks and side effects. Your doctor can estimate how likely it is the cancer will respond to treatment you are considering. For instance, the doctor might say that more treatment might have about a 1 in 100 chance of working. Some people are still tempted to try this. But it is important to have realistic expectations if you do choose this plan.

## **Palliative or supportive care**

No matter what you decide to do, it is important that you feel as good as you can. Make sure you are asking for and getting treatment for any symptoms you might have, such as nausea or pain. This type of treatment is called *palliative* or *supportive care*.

Palliative care helps relieve symptoms, but it’s not expected to cure the disease. It can be given along with cancer treatment, or can even be cancer treatment. The difference is its purpose – the main goal of palliative care is to improve the quality of your life, or help you feel as good as you can for as long as you can. Sometimes this means using drugs to help with symptoms like pain or nausea. Sometimes, though, the treatments used to control your symptoms are the same as those used to treat cancer. For instance, radiation might be used to help relieve pain caused by a large tumor. Or chemo or other drugs might be used to help shrink a tumor and keep it from blocking the bowels. But this is not the same as treatment to try to cure the cancer.

## **Hospice care**

At some point, you may benefit from hospice care. This is special care that treats the person rather than the disease; it focuses on quality rather than length of life. Most of the time, it is given at home. Your cancer may be causing problems that need to be managed, and hospice focuses on your comfort. You should know that while getting hospice care often means the end of treatments such as chemo and radiation, it doesn’t mean you can’t have treatment for the problems caused by your cancer or other health conditions. In hospice the focus of your care is on living life as fully as possible and feeling as well as you can at this difficult time. You can learn more about hospice in our document called *Hospice Care*.

Staying hopeful is important, too. Your hope for a cure may not be as bright, but there is still hope for good times with family and friends – times that are filled with happiness and meaning. Pausing at this time in your cancer treatment gives you a chance to refocus on the most important things in your life. Now is the time to do some things you’ve always wanted to do and to stop doing things you no longer want to do. Though the cancer may be beyond your control, there are still choices you can make.

You can learn more about the changes that occur when curative treatment stops working, and about planning ahead for yourself and your family, in our documents *Advanced Cancer and Nearing the End of Life*. You can read them online or call us at 1-800-227-2345 to have free copies mailed to you.

## 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 is a challenging disease 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<sup>®</sup>) and everolimus (Afinitor<sup>®</sup>), 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<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.

## **Additional resources for lung carcinoid tumors**

### **More information from your American Cancer Society**

We have some related information that may also be helpful to you. These materials can be ordered from our toll-free number 1-800-227-2345.

#### **Living with cancer**

After Diagnosis: A Guide for Patients and Families (also in Spanish)

Caring for the Patient With Cancer at Home: A Guide for Patients and Families (also in Spanish)

Living With Uncertainty: The Fear of Cancer Recurrence

When Your Cancer Comes Back: Cancer Recurrence

Distress in People With Cancer

Anxiety, Fear, and Depression

## **Understanding cancer treatments**

Understanding Cancer Surgery: A Guide for Patients and Families (also in Spanish)

Understanding Chemotherapy: A Guide for Patients and Families (also in Spanish)

Understanding Radiation Therapy: A Guide for Patients and Families (also in Spanish)

Lasers in Cancer Treatment

## **Cancer treatment side effects**

Guide to Controlling Cancer Pain (also in Spanish)

Nausea and Vomiting

Anemia in People With Cancer

Fatigue in People With Cancer

Peripheral Neuropathy Caused by Chemotherapy

## **Family and caregiver concerns**

Talking With Friends and Relatives About Your Cancer (also in Spanish)

What It Takes to Be a Caregiver

Helping Children When a Family Member Has Cancer: Dealing With Diagnosis (also in Spanish)

## **Work, insurance, and finances**

Health Insurance and Financial Assistance for the Cancer Patient (also in Spanish)

Working During Cancer Treatment

Returning to Work After Cancer Treatment

In Treatment: Financial Guidance for Cancer Survivors and Their Families (also in Spanish)

Your American Cancer Society also has books that you might find helpful. Call us at 1-800-227-2345 or visit our bookstore online at [cancer.org/bookstore](http://cancer.org/bookstore) to find out about costs or to place an order.

## National organizations and websites\*

Along with the American Cancer Society, other sources of information and support include:

### **The Carcinoid Cancer Foundation, Inc. (CCF)**

Toll-free number: 1-888-722-3132

Website: [www.carcinoid.org](http://www.carcinoid.org)

Offers literature on carcinoid cancer and neuroendocrine tumors; an international listing of doctors and centers with expertise in these rare diseases; answers to general questions regarding all aspects of carcinoid tumors (including nutritional issues, quality of life and psychosocial issues) provided by medical professionals; a national and international listing of carcinoid cancer support groups; information on National Carcinoid Symposiums, including webcasts of video and transcripts of past symposium lectures; and information on current related clinical trials.

### **Lung Cancer Alliance**

Toll-free number: 1-800-298-2436

Website: [www.lungcanceralliance.org](http://www.lungcanceralliance.org)

Offers free information by phone or through the website; has an online support community, a Telephone Support Group for Spouse/Partner Caregivers, makes referrals to lung cancer support groups, and has 'Phone Buddies' -- a peer-to-peer telephone support program for patients, families and friends.

### **National Cancer Institute (NCI)**

Toll-free number 1-800-4-CANCER (1-800-422-6237)

TTY: 1-800-332-8615

Website: [www.cancer.gov](http://www.cancer.gov)

Offers a wide variety of free, accurate, up-to-date information about cancer to patients, their families, and the general public; also can help people find clinical trials in their area.

### **National Coalition for Cancer Survivorship**

Toll-free number: 1-888-650-9127

Website: [www.canceradvocacy.org](http://www.canceradvocacy.org)

Has publications on many cancer-related topics; also offers the Cancer Survival Toolbox – a free program that teaches skills that can help people with cancer meet the challenges of their illness.



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

No matter who you are, we can help. Contact us anytime, day or night, for information and support. Call us at 1-800-227-2345 or visit [www.cancer.org](http://www.cancer.org).

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