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# Lung Carcinoid Tumor Early Detection, Diagnosis, and Staging

Know the signs and symptoms of lung carcinoid tumors. Find out how lung carcinoid tumors are tested for, diagnosed, and staged.

#### **Detection and Diagnosis**

Catching cancer early often allows for more treatment options. Some early cancers may have signs and symptoms that can be noticed, but that is not always the case.

- Can Lung Carcinoid Tumors Be Found Early?
- Signs and Symptoms of Lung Carcinoid Tumors
- Tests for Lung Carcinoid Tumors
- Understanding Your Pathology Report

#### **Stages of Lung Carcinoid Tumors**

After a cancer diagnosis, staging provides important information about the extent of cancer in the body and anticipated response to treatment.

Lung Carcinoid Tumor Stages

#### **Outlook (Prognosis)**

Doctors often use survival rates as a standard way of discussing a person's outlook (prognosis). These numbers can't tell you how long you will live, but they might help you better understand your prognosis. Some people want to know the survival statistics for people in similar situations, while others might not find the numbers helpful, or might

even not want to know them.

Survival Rates for Lung Carcinoid Tumors

#### **Questions to Ask About Lung Carcinoid Tumors**

Here are some questions you can ask your cancer care team to help you better understand your cancer diagnosis and treatment options.

Questions to Ask Your Doctor About Lung Carcinoid Tumors

# Can Lung Carcinoid Tumors Be Found Early?

Screening is the use of tests or exams to find a disease in people who don't have symptoms. Lung carcinoid tumors are not common, and there are no widely recommended screening tests for these tumors in most people.

People with multiple endocrine neoplasia type 1 (MEN1) are at increased risk for these tumors, and some doctors recommend they have CT scans of the chest every 2-5 years.

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

However, many people with peripheral carcinoid tumors or small central carcinoid tumors have no symptoms. Carcinoids that are not causing symptoms are often found when a chest x-ray or CT scan done for other reasons.

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# Signs and Symptoms of Lung Carcinoid Tumors

- Central carcinoids
- Peripheral carcinoids
- Symptoms caused by hormones from the tumor
- If you have 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 when tests are done for other reasons.

#### Central carcinoids

Most carcinoid tumors start in the large bronchial tubes leading into the lung. Symptoms can include:

- Cough, which can sometimes be bloody
- Wheezing
- Shortness of breath
- 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 doesn't 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 <u>gastrointestinal carcinoid tumors</u><sup>1</sup>.

**Carcinoid syndrome:** Rarely, lung carcinoid tumors release enough hormone-like substances into the bloodstream to cause symptoms. This causes carcinoid syndrome. Symptoms can include:

- Facial flushing (redness and warm feeling)
- Diarrhea
- Wheezing
- Fast heartbeat

Many people with carcinoid syndrome 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
- 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 (a steroid hormone) and other hormones. This can lead to:

- Weight gain
- Easy bruising
- Weakness
- Drowsiness
- High blood sugar (or even diabetes)
- High blood pressure
- Increased body and facial hair

# If you have symptoms of lung carcinoid tumors

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.

# **Hyperlinks**

1. www.cancer.org/cancer/types/gastrointestinal-carcinoid-tumor.html

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# **Tests for Lung Carcinoid Tumors**

- Medical history and physical exam
- Imaging tests
- Sputum cytology
- Biopsy
- Blood and urine tests
- Pulmonary function tests

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 take a complete medical history, including your family history, to learn about your symptoms and possible <u>risk factors</u><sup>1</sup>.

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**

Doctors use imaging tests to take pictures of the inside of your body. <u>Imaging tests</u><sup>2</sup> are done for a number of reasons, including to help find a suspicious area that might be cancer or to learn how far cancer may have spread (metastasized).

#### **Chest x-ray**

A chest <u>x-ray</u><sup>3</sup> 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 <u>CT scan</u><sup>4</sup>uses x-rays taken from different angles, which are combined by a computer to make detailed pictures of the organs. This test is most often used to look at the chest and/or belly (abdomen) to see if carcinoid has spread to other organs. It can also be used to guide a biopsy needle into an area of concern.

A CT scan is more likely to show small lung tumors than routine chest x-rays. It 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.

#### 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 where cancer might have spread.

**Positron emission tomography (PET) scan**: For most types of cancer, PET scans use a form of radioactive glucose (sugar) to find tumors. This type of PET scan is not very useful in finding atypical carcinoid tumors, but a newer type of PET scan called a gallium-68 dotatate PET/CT scan is being used more often for typical carcinoid tumors. It uses the radioactive agent <sup>68</sup>Ga-dotatate which attaches to the somatostatin receptors on carcinoid cells. A special camera can detect the radioactivity. This gallium-68 dotatate PET/CT scan is becoming more widely available since it was approved by the FDA in 2016. It is able to find carcinoid tumors better than OctreoScan (described below).

Somatostatin receptor scintigraphy (OctreoScan): This test 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 scan:** This test is used much 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 areas would most likely be carcinoid tumors, but other kinds of neuroendocrine tumors will also pick up this chemical.

# **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. More tests may be needed to get a sample of the abnormal cells to be looked at in the lab.

One way to do this is called *sputum cytology*. A sample of sputum (mucus you cough up from the lungs) is looked at in the 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.

#### **Bronchoscopy and 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 your windpipe and bronchi to look at the lining of the lung's main airways. Your 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 will be ready to return home within hours. A disadvantage is that this type of biopsy may not always be able to remove enough tissue 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 serious. 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) and biopsy

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 (carina), 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 be looked at in a lab.

#### **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 get 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 a local anesthetic 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 in 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 is called a *pneumothorax*. If the air leak is minimal, it often gets better without any treatment. A large pneumothorax is treated by putting a small tube into the chest space and sucking out the air over a few days, 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:

- Transtracheal FNA or transbronchial FNA is done by passing the needle through the wall of the windpipe (trachea) or bronchi (the large airways leading into the lungs) during bronchoscopy or EBUS (already 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 in the lab. Thoracoscopy can also be used to sample lymph nodes and fluid and find out if a tumor is growing into nearby tissues or organs. This procedure is also known as video-assisted thoracoscopic surgery (VATS).

**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 <u>carcinoid syndrome</u>, which is caused by excess levels of these chemicals in the blood.

Serotonin is 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 the patient's symptoms and where the tumor might be located, 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, like the gastrointestinal (GI) tract.

# **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 <u>surgery</u><sup>5</sup> might be used to treat the cancer, because surgery will remove part or all of the lung. 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.

For a PFT, you will need to breathe in and out through a tube that is connected to a machine that measures airflow.

# **Hyperlinks**

- 1. <a href="www.cancer.org/cancer/types/lung-carcinoid-tumor/causes-risks-prevention/risk-factors.html">www.cancer.org/cancer/types/lung-carcinoid-tumor/causes-risks-prevention/risk-factors.html</a>
- 2. <u>www.cancer.org/cancer/diagnosis-staging/tests/imaging-tests/imaging-radiology-tests-for-cancer.html</u>
- 3. <u>www.cancer.org/cancer/diagnosis-staging/tests/imaging-tests/x-rays-and-other-radiographic-tests.html</u>
- 4. <u>www.cancer.org/cancer/diagnosis-staging/tests/imaging-tests/ct-scan-for-cancer.html</u>
- 5. www.cancer.org/cancer/types/lung-carcinoid-tumor/treating/surgery.html

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# **Lung Carcinoid Tumor Stages**

- How is the stage determined?
- Stages of lung carcinoid tumors

After someone is diagnosed with a lung carcinoid tumor, doctors will try to figure out if it has spread, and if so, how far. This process is called *staging*. The stage of a cancer describes how much cancer is in the body. It helps determine how serious the cancer is and how best to <u>treat</u><sup>1</sup> it. Doctors also use a cancer's stage when talking about survival statistics.

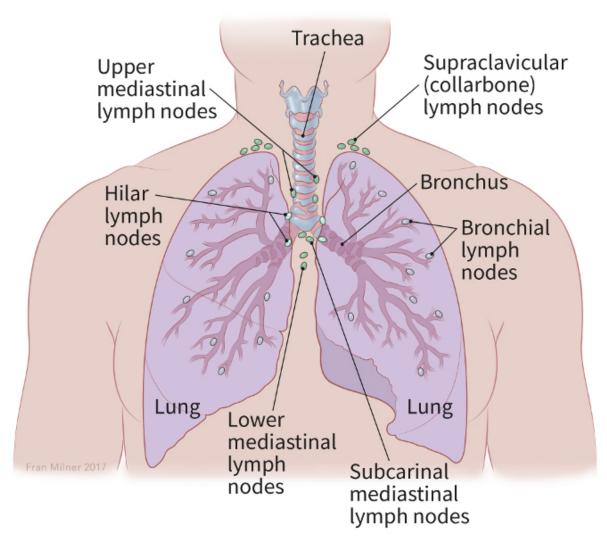
The earliest stage is stage 0. The other main stages range from I (1) through IV (4). Some of these are divided further using letters or numbers. As a rule, the lower the stage, the less the cancer has spread. A higher number, such as stage IV, means cancer has spread more. And within a stage, an earlier letter (or number) means a lower stage. Although each person's cancer experience is unique, cancers with similar stages tend to have a similar outlook and are often treated in much the same way.

# How is the stage determined?

The staging system most often used for lung carcinoid tumors is the American Joint Committee on Cancer (AJCC) **TNM** system, which is based on 3 key pieces of information:

• The size and extent of the main **tumor (T)**: How large is the tumor? Has it grown into nearby structures or organs?

- The spread to nearby lymph **nodes (N)**: Has the cancer spread to nearby lymph nodes? (See image.)
- The spread (**metastasis**) to distant sites (**M**): Has the cancer spread to distant organs? (The most common site of spread is the liver.)



Numbers or letters after T, N, and M provide more details about each of these factors. Higher numbers mean the cancer is more advanced. Once a person's T, N, and M categories have been determined, this information is combined in a process called *stage grouping* to assign an overall stage. For more information, see <u>Cancer Staging</u><sup>2</sup>.

There is not a separate staging system for lung carcinoid tumors. It uses the same classification as non-small cell lung cancer and small cell lung cancer. The system described below is the most recent version of the AJCC system, effective as of January

#### 2018.

Lung carcinoid tumors are typically given a *clinical stage* based on the results of physical exams, biopsies, imaging tests, and any other tests that have been done. (See Tests for Lung Carcinoid Tumors.) If surgery is done, the *pathologic stage* (also called the *surgical stage*) is determined by examining tissue removed during the operation.

Staging for lung carcinoid tumors can be complex, so ask your doctor to explain it to you in a way you understand.

# Stages of lung carcinoid tumors

AJCC Stage	Stage grouping	Stage description*	
Occult (hidden) cancer	TX NO MO	The main tumor can't be assessed for some reason, or 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 (TX). The cancer is not thought to have spread to nearby lymph nodes (N0) or to distant parts of the body (M0).	
0	Tis N0 M0	The tumor is found only in the top layers of cells lining the air passages, but it has not invaded deeper into other lung tissues (Tis). The cancer has not spread to nearby lymph nodes (N0) or to distant parts of the body (M0).	
IA1	T1a N0 M0	The tumor is no larger than 1 cm across, it has not reached the membranes that surround the lungs, and it does not affect the main branches of the bronchi (T1a). The cancer has not spread to nearby lymph nodes (N0) or to distant parts of the body (M0).	
IA2	T1b N0 M0	The tumor is larger than 1 cm but no larger than 2 cm across. It has not reached the membranes that surround the lungs, and it does not affect the main branches of the bronchi (T1b). The cancer has not spread to nearby lymph nodes (N0) or to distant parts of the body (M0).	
IA3	T1c N0 M0	The tumor is larger than 2 cm but no larger than 3 cm across. It has not reached the membranes that surround the lungs, and it does not affect the main branches of the bronchi (T1c). The cancer has not spread to nearby lymph nodes (N0) or to distant parts of the body (M0).	

	The tumor has one or more of the following features (T2a):
T2a N0 M0	<ul> <li>It is larger than 3 cm but not larger than 4 cm across.</li> <li>It has grown into a main bronchus, but is not within 2 cm of the carina (the point where the windpipe splits into the left and right main bronchi) and it is not larger than 4 cm across.</li> <li>It has grown into the visceral pleura (the membranes surrounding the lungs) and is not larger than 4 cm across.</li> <li>It is partially clogging the airways (and is not larger than 4 cm across).</li> </ul> The cancer has not spread to nearby lymph nodes (N0) or to
	distant parts of the body (M0).
T2b N0 M0	<ul> <li>It is larger than 4 cm but not larger than 5 cm across.</li> <li>It has grown into a main bronchus, but is not within 2 cm of the carina (the point where the windpipe splits into the left and right main bronchi) and it is larger than 4 cm but not larger than 5 cm across.</li> <li>The tumor has grown into the visceral pleura (the membranes surrounding the lungs) and is larger than 4 cm but not larger than 5 cm across.</li> <li>The tumor is partially clogging the airways (and is larger than 4 cm but not larger than 5 cm across).</li> </ul> The cancer has not spread to nearby lymph nodes (N0) or to distant parts of the body (M0).
N1 M0	The tumor 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 (T1). 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 (N1). The cancer has not spread to distant parts of the body (M0).
OR	

		The tumor has one or more of the following features (T2):	
IID	T2a/T2b N1 M0	<ul> <li>It is larger than 3 cm but not larger than 5 cm across.</li> <li>It has grown into a main bronchus, but is not within 2 cm of the carina (the point where the windpipe splits into the left and right main bronchi) and it is not larger than 5 cm across).</li> <li>It has grown into the visceral pleura (the membranes surrounding the lungs) and is not larger than 5 cm.</li> <li>It is partially clogging the airways (and is not larger than 5 cm).</li> </ul> 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 (N1). The cancer has not spread to distant parts of the body (M0).	
OR OR			
	T3 N0 M0	<ul> <li>The tumor has one or more of the following features (T3):</li> <li>It is larger than 5 cm but not larger than 7 cm across.</li> <li>It has grown into the chest wall, the inner lining of the chest wall (parietal pleura), the phrenic nerve, or membranes of the sac surrounding the heart (parietal pericardium).</li> <li>There are 2 or more separate tumor nodules in the same lobe of a lung.</li> </ul>	
		The cancer has not spread to nearby lymph nodes (N0) or distant parts of the body (M0).	
	T1a/T1b/T1c N2 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 (T1). The cancer has 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 (N2). The cancer has not spread to distant parts of the body (M0).	
	OR		

		The tumor has one or more of the following features (T2):
	T2a/T2b N2 M0	<ul> <li>It is larger than 3 cm but not larger than 5 cm across.</li> <li>It has grown into a main bronchus, but is not within 2 cm of the carina (the point where the windpipe splits into the left and right main bronchi) and it is not larger than 5 cm across).</li> <li>It has grown into the visceral pleura (the membranes surrounding the lungs) and is not larger than 5 cm.</li> <li>It is partially clogging the airways (and is not larger than 5 cm).</li> </ul>
		The cancer has 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 (N2). The cancer has not spread to distant parts of the body (M0).
	OR	
		The tumor has one or more of the following features (T3):
IIIA	T3 N1 M0	<ul> <li>It is larger than 5 cm but not larger than 7 cm across.</li> <li>It has grown into the chest wall, the inner lining of the chest wall (parietal pleura), the phrenic nerve, or membranes of the sac surrounding the heart (parietal pericardium).</li> <li>There are 2 or more separate tumor nodules in the same lobe of a lung.</li> </ul>
		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 (N1). The cancer has not spread to distant parts of the body (M0).
	OR	•
	T4	The tumor has one or more of the following features (T4):
	N0 or N1	It is larger than 7 cm across.
	МО	<ul> <li>It has grown into the space between the lungs (mediastinum), the heart, the large blood vessels near the</li> </ul>

	T1a/T1b/T1c N3 M0	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 (N0 or N1). The cancer has not spread to distant parts of the body (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 (T1). The cancer has spread to lymph nodes near the collarbone on either side of the body, and/or has spread to hilar or mediastinal lymph nodes on the other side of the body from the main tumor (N3). The cancer has not spread to distant parts of the body (M0).  The tumor has one or more of the following features (T2):  • It is larger than 3 cm but not larger than 5 cm across.
IIIB		<ul> <li>It has grown into a main bronchus, but is not within 2 cm of the carina (the point where the windpipe splits into the left and right main bronchi) and it is not larger than 5 cm across.</li> <li>It has grown into the visceral pleura (the membranes surrounding the lungs) and is not larger than 5 cm.</li> <li>It is partially clogging the airways (and is not larger than 5 cm).</li> <li>The cancer has spread to lymph nodes near the collarbone on either side of the body, and/or has spread to hilar or mediastinal lymph nodes on the other side of the body from the main tumor (N3). The cancer has not spread to distant parts of the body (M0).</li> </ul>

OR	OR	
	The tumor has one or more of the following features (T3):	
T3 N2	<ul> <li>It is larger than 5 cm but not larger than 7 cm across.</li> <li>It has grown into the chest wall, the inner lining of the chest wall (parietal pleura), the phrenic nerve, or membranes of the sac surrounding the heart (parietal pericardium).</li> <li>There are 2 or more separate tumor nodules in the same lobe of a lung.</li> </ul>	
MO	The cancer has 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 (N2). The cancer has not spread to distant parts of the body (M0).	
OR		
T4 N2 M0	<ul> <li>The tumor has one or more of the following features (T4):</li> <li>It is larger than 7 cm across.</li> <li>It has grown into the space between the lungs (mediastinum), the heart, the large blood vessels near the heart (such as the aorta), the windpipe (trachea), the tube connecting the throat to the stomach (esophagus), the thin muscle separating the chest from the abdomen (diaphragm), the backbone (spine), or the carina (the point where the windpipe splits into the left and right bronchi).</li> <li>There are 2 or more separate tumor nodules in different lobes of the same lung.</li> </ul>	
	The cancer has 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 (N2). The cancer has not spread to distant parts of the body (M0).	
Т3	The tumor has one or more of the following features (T3):	
N3	<ul> <li>It is larger than 5 cm but not larger than 7 cm across.</li> <li>It has grown into the chest wall, the inner lining of the chest</li> </ul>	

	MO	<ul> <li>wall (parietal pleura), the phrenic nerve, or membranes of the sac surrounding the heart (parietal pericardium).</li> <li>There are 2 or more separate tumor nodules in the same lobe of a lung.</li> <li>The cancer has spread to lymph nodes near the collarbone on either side of the body, and/or has spread to hilar or mediastinal lymph nodes on the other side of the body from the main tumor (N3). The cancer has not spread to distant parts of the body (M0).</li> </ul>
	OR	•
		The tumor has one or more of the following features (T4):
IIIC	T4 N3 M0	<ul> <li>It is larger than 7 cm across.</li> <li>It has grown into the space between the lungs (mediastinum), the heart, the large blood vessels near the heart (such as the aorta), the windpipe (trachea), the tube connecting the throat to the stomach (esophagus), the thin muscle separating the chest from the abdomen (diaphragm), the backbone (spine), or the carina (the point where the windpipe splits into the left and right bronchi).</li> <li>There are 2 or more separate tumor nodules in different lobes of the same lung.</li> </ul>
		The cancer has spread to lymph nodes near the collarbone on either side of the body, and/or has spread to hilar or mediastinal lymph nodes on the other side of the body from the main tumor (N3). The cancer has not spread to distant parts of the body (M0).
		The cancer can be any size and may or may not have grown into nearby structures (any T). It may or may not have reached
	Any T	nearby lymph nodes (any N). In addition, any of the following is
	true (M1a):	
		The cancer has spread to the other lung.
• Cancer cells are for malignant pleural e		<ul> <li>Cancer cells are found in the fluid around the lung (called a malignant pleural effusion).</li> </ul>
		Cancer cells are found in the fluid around the heart (called)

	a malignant pericardial effusion).	
	OR	
into nearby structures (any T). It may or may not h <b>Any N</b> nearby lymph nodes (any N). It has spread as a si		
IVB	Any T Any N M1c	The cancer can be any size and may or may not have grown into nearby structures (any T). It may or may not have reached nearby lymph nodes (any N). It has spread as more than one tumor outside the chest, such as to distant lymph nodes and/or to other organs such as the liver, bones, or brain (M1c).

<sup>\*</sup>The following additional categories are not listed in the table above:

- **T0**: There is no evidence of a primary tumor.
- NX: Nearby lymph nodes cannot be assessed due to lack of information.

# **Hyperlinks**

- 1. www.cancer.org/cancer/types/lung-carcinoid-tumor/treating.html
- 2. www.cancer.org/cancer/diagnosis-staging/staging.html

#### References

American Joint Committee on Cancer. Lung. In: *AJCC Cancer Staging Manual*. 8th ed. New York, NY: Springer; 2017: 431-456.

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# **Survival Rates for Lung Carcinoid Tumors**

- What is a 5-year relative survival rate?
- Where do these numbers come from?
- 5-year relative survival rates for lung carcinoid tumor
- Understanding the numbers

Survival rates can give you an idea what percentage of people with the same type and stage of cancer are still alive a certain amount of time (usually 5 years) after they were diagnosed. They can't tell you how long you will live, but they may help give you a better understanding of how likely it is that your treatment will be successful.

Keep in mind that survival rates are estimates and are often based on previous outcomes of large numbers of people who had a specific cancer, but they can't predict what will happen in any particular person's case. These statistics can be confusing and may lead you to have more questions. Your doctor is familiar with your situation; ask how these numbers may apply to you.

# What is a 5-year relative survival rate?

A **relative survival rate** compares people with the same type and stage of lung carcinoid tumor to people in the overall population. For example, if the **5-year relative survival rate** for a specific stage of lung carcinoid tumor is 90%, it means that people who have that cancer are, on average, about 90% as likely as people who don't have that cancer to live for at least 5 years after being diagnosed.

#### Where do these numbers come from?

The American Cancer Society relies on information from the Surveillance, Epidemiology, and End Results (SEER) database, maintained by the National Cancer Institute (NCI), to provide survival statistics for different types of cancer.

The SEER database tracks 5-year relative survival rates for lung carcinoid tumor in the United States, based on how far the cancer has spread. The SEER database, however, does not group cancers by AJCC TNM stages (stage 1, stage 2, stage 3, etc.). Instead, it groups cancers into localized, regional, and distant stages:

- Localized: There is no sign that the cancer has spread outside of the lung.
- **Regional:** The cancer has spread outside of the lung to nearby structures or lymph nodes.
- **Distant:** The cancer has spread to distant parts of the body such as the brain, liver, bones, or other lung.

### 5-year relative survival rates for lung carcinoid tumor

These numbers are based on people diagnosed with lung carcinoid tumors between 2012 and 2018.

SEER Stage	5-Year Relative Survival Rate
Localized	98%
Regional	86%
Distant	55%

All SEER stages combined 89%

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.

# **Understanding the numbers**

- These numbers apply only to the stage of the cancer when it is first diagnosed. They do not apply later on if the cancer grows, spreads, or comes back after treatment.
- These numbers don't take everything into account. Survival rates are grouped based on how far the cancer has spread, but your age and overall health, type of lung carcinoid tumor<sup>1</sup>, how well it responds to treatment, and other factors can also affect your outlook.
- People now being diagnosed with lung carcinoid tumors may have a better outlook than these numbers show. Treatments improve over time, and these numbers are based on people who were diagnosed and treated at least 5 years earlier.

#### **Hyperlinks**

1. <u>www.cancer.org/cancer/types/lung-carcinoid-tumor/about/what-is-lung-carcinoid-tumor.html</u>

#### References

Ruhl JL, Callaghan C, Hurlbut, A, Ries LAG, Adamo P, Dickie L, Schussler N (eds.) Summary Stage 2018: Codes and Coding Instructions, National Cancer Institute, Bethesda, MD, 2018.

SEER\*Explorer: An interactive website for SEER cancer statistics [Internet]. Surveillance Research Program, National Cancer Institute. Accessed at https://seer.cancer.gov/explorer/ on February 23, 2023.

Thomas CF, Jett JR, Strosberg JR. Lung neuroendocrine (carcinoid) tumors: Treatment and prognosis. UpToDate website. https://www.uptodate.com/contents/lung-neuroendocrine-carcinoid-tumors-treatment-and-prognosis. Updated Feb. 6, 2018. Accessed July 17, 2018.

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# Questions to Ask Your Doctor About Lung Carcinoid Tumors

- When you're told you have a lung carcinoid tumor
- When deciding on a treatment plan
- During treatment
- After treatment

It is important to have honest, open discussions with your cancer care team. They want to answer your questions, so that you can make informed treatment and life decisions. For instance, consider these questions:

# When you're told you have a lung carcinoid tumor

- What kind of carcinoid tumor<sup>1</sup> do I have?
- Where is the cancer located?
- Has the carcinoid tumor spread beyond the lungs?
- What is the stage (extent) of the carcinoid tumor, and what does that mean?
- Will I need other tests before we can decide on treatment?
- Do I need to see any other doctors or health professionals?
- If I'm concerned about the costs and insurance coverage for my diagnosis and treatment, who can help me?

### When deciding on a treatment plan

- What are my <u>treatment</u><sup>2</sup> options?
- What do you recommend and why?
- How much experience do you have treating this type of cancer?
- Should I get a second opinion<sup>3</sup>? How do I do that? Can you recommend someone?
- What would the goal of the treatment be?
- 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?
- What risks or side effects are there to the treatments you suggest? Are there things I can do to reduce these side effects?
- How might treatment affect my daily activities? Can I still work full time?
- What are the chances the cancer will <u>recur</u><sup>4</sup> (come back) with these treatment plans?
- What will we do if the treatment doesn't work or if the cancer recurs?
- What if I have transportation problems getting to and from treatment?

# **During treatment**

Once treatment begins, you'll need to know what to expect and what to look for. Not all of these questions may apply to you, but asking the ones that do may be helpful.

- How will we know if the treatment is working?
- Is there anything I can do to help manage side effects?
- What symptoms or <u>side effects</u><sup>5</sup> should I tell you about right away?
- How can I reach you on nights, holidays, or weekends?
- Do I need to change what I eat during treatment?
- Are there any limits on what I can do?
- Can I exercise during treatment<sup>6</sup>? If so, what kind should I do, and how often?
- Can you suggest a mental health professional I can see if I start to feel overwhelmed, depressed, or distressed?

#### After treatment

- Do I need a special diet after treatment?
- Are there any limits on what I can do?
- What other symptoms should I watch for?
- What kind of exercise should I do now?
- What type of follow-up will I need after treatment?
- How often will I need to have follow-up exams and imaging tests?
- Will I need any blood tests?
- How will we know if the cancer has come back<sup>7</sup>? What should I watch for?
- What will my options be if the cancer comes back?

Along with these sample questions, be sure to write down some of your own. For instance, you might want more information about recovery time. Or you may want to ask about <u>clinical trials</u><sup>8</sup>.

Keep in mind that doctors aren't the only ones who can give you information. Other health care professionals, such as nurses and social workers, may have the answers to some of your questions. You can find more information about speaking with your health care team in <a href="https://example.com/The-Doctor-Patient Relationship">The Doctor-Patient Relationship</a>9.

# **Hyperlinks**

- 1. <a href="www.cancer.org/cancer/types/lung-carcinoid-tumor/about/what-is-lung-carcinoid-tumor.html">www.cancer.org/cancer/types/lung-carcinoid-tumor/about/what-is-lung-carcinoid-tumor.html</a>
- 2. www.cancer.org/cancer/types/lung-carcinoid-tumor/treating.html
- 3. <u>www.cancer.org/cancer/managing-cancer/finding-care/seeking-a-second-opinion.html</u>
- 4. www.cancer.org/cancer/survivorship/long-term-health-concerns/recurrence.html
- 5. www.cancer.org/cancer/managing-cancer/side-effects.html
- 6. <u>www.cancer.org/cancer/survivorship/be-healthy-after-treatment/physical-activity-and-the-cancer-patient.html</u>
- 7. www.cancer.org/cancer/survivorship/long-term-health-concerns/recurrence.html
- 8. <u>www.cancer.org/cancer/managing-cancer/making-treatment-decisions/clinical-trials.html</u>
- 9. <u>www.cancer.org/cancer/managing-cancer/finding-care/the-doctor-patient-relationship.html</u>

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