THYMUS CANCER

What Is Cancer?

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

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

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

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

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

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

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

What Is Thymus Cancer?

The *thymus* is a small organ located in the upper/front portion of the chest, extending from the base of the throat to the front of the heart. The thymus is composed of an inner *medulla* and an outer *cortex,* and is surrounded by a thin covering called the *capsule.* The thymus reaches its maximum weight of about 1 ounce during puberty, then slowly decreases in size during adulthood and is gradually replaced by fat tissue.

During fetal development and childhood, the thymus is involved in the production and maturation of *T-lymphocytes,* a type of white blood cell important in the immune system. T-lymphocytes develop in the thymus and then travel to *lymph nodes* (bean-sized collections of immune system cells) throughout the body. There they help the immune system protect the body from viruses, fungus, and other types of infections.

The thymus contains two main types of cells, *thymic epithelial cells* and *lymphocytes.* Thymic epithelial cells can give origin to *thymomas* and *thymic carcinomas.* Lymphocytes, whether in the thymus or in the lymph nodes, can become malignant and develop into cancers called *Hodgkin's disease* and *non-Hodgkin's lymphomas*. Information about these cancers is available in additional documents from the American Cancer Society.

The thymus also contains another, much less common type of cells called *Kulchitsky cells,* or *neuroendocrine cells,* which normally release certain hormones. These cells can give rise to cancers, called *carcinoids* or *carcinoid tumors,* that often release the same type of hormones, and are similar to other tumors arising from neuroendocrine cells elsewhere in the body.

Thymomas, thymic carcinomas, and thymic carcinoid tumors are distinguished from one another and from other tumors of the thymus (such as Hodgkin's disease and non-Hodgkin's lymphoma) by their appearance under the microscope and results of other laboratory tests performed on tissue samples.

There is some disagreement among doctors about the best way to describe and classify thymomas. In the past, thymomas were sometimes divided into *benign thymomas* (not cancerous) and *malignant thymomas* (considered cancerous because of evidence they had invaded or penetrated beyond the thymus into other tissues or organs). The view currently held by most doctors is that all thymomas are potentially cancerous, and the best way to predict their *prognosis* (the outlook for chances of survival) is to describe whether they have invaded beyond the thymus and, if so, how far. Invasion is identified by the surgeon who notes whether or not the tumor appears attached to nearby organs, and by the pathologist who looks at samples from the *margins* (edges) of the tumor under the microscope, to see whether thymoma cells have spread among cells from other tissues or organs.

Some doctors recommend classifying thymomas based on whether the cells of the tumor more closely resemble normal cells of the *cortex* (outer part) of the thymus or the *medulla* (inner part). Several studies have suggested that *cortical thymomas* and *predominantly cortical thymomas* are more likely to be invasive and/or *recur* (come back) after surgery, while *medullary thymomas* and *mixed thymomas* are more likely to be cured by surgery. However, not all studies have confirmed the value of this classification and its use remains a topic of uncertainty and disagreement.

In general, most doctors prefer to use the system based on whether there is invasion and, how much invasion there is.

Thymic carcinomas are formed by cells that have certain abnormalities that are clearly recognized under the microscope. Cells with these abnormalities always have the potential to invade nearby tissues and can often *metastasize* (spread to distant tissues and organs). Thymic carcinomas are further divided into low-grade (better prognosis) and high-grade (worse prognosis, that is, more likely to grow and spread

quickly) categories. Low-grade thymic carcinomas include *well-differentiated squamous cell, mucoepidermoid,* and *basaloid* types. High-grade thymic carcinomas include *poorly-differentiated squamous cell, small cell/neuroendocrine, clear cell, anaplastic/undifferentiated,* and *sarcomatoid types.*

Thymic carcinoids are considered malignant, since most will invade and metastasize.

Although thymomas, thymic carcinomas, and thymic carcinoid tumors are all considered cancerous, many, if found at early stages, can be cured. Even some later-stage tumors of the thymus can be treated successfully, leading to cure or long *remissions* (periods of time before the cancer comes back).

What Are The Key Statistics About Thymus Cancers?

Approximately 90% of thymic tumors are thymomas. Most of the remainder are thymic carcinomas, lymphomas, or carcinoid tumors.

Thymomas are rare, although we do not know exactly how rare. Because not all doctors classify them as true cancers, most hospitals have not kept detailed records of the number of patients treated for thymomas.

Thymic carcinomas and thymic carcinoid tumors are extremely rare. Less than 200 cases of each type have been reported in medical journal articles, but the actual number of patients with these cancers is likely to be somewhat larger.

What Are The Risk Factors For Thymus Cancer?

A *risk factor* is anything that increases a person's chance of getting a disease such as cancer. Different cancers have different risk factors. For example, unprotected exposure to strong sunlight is a risk factor for skin cancer. Smoking is a risk factor for lung, oral, laryngeal, bladder, and several other cancers.

No specific inherited, environmental, or lifestyle risk factors have been identified for the development of a thymoma, thymic carcinoid tumor, or thymic carcinoma. Some studies have suggested a possible association with a previous exposure to radiation to the upper chest area, but this has not been confirmed.

Men and women have about the same likelihood of developing tumors of the thymus. These tumors are more common in middle-aged or elderly people than in children and young adults.

Can Cancers Of The Thymus Be Prevented?

Since research has not yet found any controllable risk factors, there are no known ways to prevent these cancers.

Can Thymus Cancer Be Found Early?

So far, there are no screening tests that are suitable for early detection of thymic tumors in people without symptoms. The only strategy available for finding these cancers earlier is prompt medical evaluation of people with symptoms suggesting they may have a tumor of the thymus.

Approximately 25% of patients with thymic tumors have had symptoms for at least six months prior to seeking medical attention, and the majority has had symptoms for 2-3 months prior to diagnosis. About 40% of patients with thymomas have no symptoms. In these patients, chest x-rays done to evaluate unrelated medical conditions show evidence of a tumor in the region of the thymus.

Symptoms of Thymic Cancers

Thymic tumors are usually not apparent until they are large enough to compress nearby, air passages or blood vessels. Compression of the *trachea* (windpipe) causes shortness of breath, compression of veins that return blood from the head and neck to the heart can cause severe facial swelling.

Thymomas are often associated with the development of symptoms that are not directly related to pressure from the tumor mass itself. These *paraneoplastic syndromes* (tumor-related conditions) include *myasthenia gravis, red cell aplasia*, and *hypogammaglobulinemia*. These conditions can be very important in the early diagnosis of thymomas because they may be prominent while the tumor is still at an early stage. Cells of the immune system normally produce antibodies that specifically attack disease-causing microbes such as bacteria. Sometimes, however, the immune system produces antibodies that attack or interfere with functioning of normal cells, and an *autoimmune disease* can result. Although paraneoplastic syndromes associated with certain other types of cancer have a variety of causes, the syndromes that commonly affect patients with thymomas are types of autoimmune diseases.

Myasthenia gravis is one such autoimmune disease. The body forms antibodies that block the receptors on the surface of muscle fibers that stimulate muscles to move, resulting in severe muscle weakness. Indeed, *myasthenia gravis* is Latin for "severe muscle weakness", an accurate description of this disorder. Patients have reduced muscular strength throughout the body, but most severely involved are the muscles of the eyes, neck, and chest, causing blurred or double vision and problems with swallowing and breathing. Myasthenia gravis develops in 30%-50% of people with a thymoma. But, only 15% of patients diagnosed with myasthenia gravis have a thymoma. An additional 55% of people with the disorder have other, noncancerous abnormalities of the thymus gland. Myasthenia gravis can be treated by surgically removing the thymus (whether or not a thymoma is present). Medicines that reduce formation of anti-receptor antibodies, or strengthen the muscles to nerve signals are also used.

Red cell aplasia, which means "lack of red blood cell formation", occurs in only 5% of thymoma patients. But 30%-50% of people with red cell aplasia have a thymoma. Red blood cells are essential in carrying oxygen from the lungs to other tissues of the body. Reduced red blood cell production causes *anemia* (low red blood cell counts). Symptoms of anemia include weakness, dizziness, shortness of breath, and reduced exercise tolerance. Red cell aplasia typically occurs in men and women older than 40 years of age. Treatment is removal of the thymus gland.

Hypogammaglobulinemia is a disorder in which the body produces an abnormally low amount of normal infection-fighting antibodies, leaving the individual susceptible to infections. Hypogammaglobulinemia develops in 5%-10% of thymoma patients. About 10% of patients with hypogammaglobulinemia have a thymoma. Unlike myasthenia gravis and red cell aplasia however, removal of the thymus does not help correct this disease.

Several other conditions caused by immune system problems are also more common in people with thymomas than in the general population. However, they are much less common than myasthenia gravis, pure red cell aplasia, or hypogammaglobulinemia.

Thymic carcinomas usually cause symptoms by compressing and invading important blood vessels or airways in the chest. Unlike thymomas, they very rarely cause paraneoplastic syndromes.

Thymic carcinoids usually cause symptoms by compressing veins or airways in the chest. While they do not cause any of the paraneoplastic syndromes associated with thymomas, they sometimes cause a condition called the *carcinoid syndrome.* This syndrome is caused by certain hormones released by carcinoid tumors. Symptoms include *flushing* (redness and warmth of the skin due to increased blood flow), diarrhea, and asthma. Most patients with carcinoid syndrome do not have all of these symptoms, and most thymic carcinoids do not cause the carcinoid syndrome.

How Is Thymus Cancer Diagnosed?

If there is a reason to suspect a tumor of the thymus, the doctor will use one or more examinations and tests to find out if the disease is really present.

Medical History and Physical Examination

A complete medical history will reveal symptoms commonly associated with these tumors. However, most symptoms caused by thymomas are not specific and can be caused by other diseases as well. A physical exam will provide information about signs of thymic cancer and other health problems. Patients with thymic cancer will sometimes have a fullness that the doctor can feel in the lower neck area. Thymomas are more often diagnosed by recognizing the signs and symptoms associated with myasthenia gravis, hypogammaglobulinemia, and red cell aplasia described in Can Thymus Cancer Be Found Early?

Imaging Studies

Chest x-rays, *computed tomography* (*CT)* scans, and *magnetic resonance imaging* (MRI) scans are essential in determining the location, size, and extent of a tumor.

Computed tomography (CT): CT scans are a type of x-ray procedure in which multiple images are taken as an x-ray beam rotates around the body. A computer combines these images to produce a detailed cross-sectional image that is useful in identifying many types of thymic tumors. Injection of *contrast material* into a vein before CT scanning can help produce clearer images that better distinguish tumors from normal tissue. CT scanning is one of the most useful tests in finding a mass inside the thymus. It is also useful in checking whether or not a cancer has spread to organs and tissues beyond the thymus.

Magnetic resonance imaging (MRI): An MRI scan uses radio waves and strong magnets instead of x-rays. Tissues of the body absorb energy from the radio waves, then the energy is released in a pattern influenced by the type of tissue and by certain diseases. A computer then translates the pattern of radio waves given off by tissues into a very detailed image of parts of the body.

Biopsy

Although signs, symptoms, and imaging studies can suggest that a thymic tumor is likely to be present, a *biopsy* (sample of tumor tissue removed for examination under the microscope) is needed to confirm whether a tumor is present and, if so, to determine its type. This is important because other tumors that start near the thymus or start in other organs and spread to lymph nodes near the thymus, have a different prognosis and are treated in different ways. There are several types of biopsies that can be used to obtain samples of masses suspected of being thymic tumors.

Mediastinoscopy: A hollow lighted tube is inserted into a small surgical incision at the base of the throat and advanced behind the chest bone. (This area is called the *mediastinum*, hence the name of the procedure.) The surgeon can view a tumor of the thymus through the tube and can operate small instruments through the tube to remove samples of the tumor.

Bronchoscopy: For patients suspected of having *compression* (squeezing) of the *trachea* (windpipe) by the thymic tumor, the doctor may also do a *bronchoscopy*, a procedure in which a flexible lighted tube is inserted through the mouth, and into the airways bronchi to check for tumors and narrowing.

Needle biopsy: A long thin needle (about the same width as a needle used for blood tests) is used in a fine needle biopsy to remove small fragments of tissue. A slightly wider needle may be used in a core needle biopsy to remove a cylindrical piece of tissue (about 1/16 inch wide). The needle is inserted through the skin, and imaging procedures such as CT scans are used by the doctor to guide the needle into the correct location. Many doctors prefer the needle biopsy when they suspect a thymoma. They are concerned that taking a bigger piece of the tumor may allow it to spread. If the person with the suspected thymoma has one of the "paraneoplastic" syndromes that were described, then the surgeon may proceed to operate without any biopsy, since the treatment (removal of the thymus) is the same, whether or not a thymoma is present.

The doctor may recommend other tests and procedures as well.

How Is Thymus Cancer Staged?

Staging is a process that tells the doctor how widespread the cancer may be. Staging of thymic cancer is based on:

- Extent of disease as seen on CT or MRI scans.
- Whether the surgeon finds the tumor difficult to separate from nearby tissues (indicating invasion).
- Whether the *pathologist* (a doctor specializing in diagnosing diseases using laboratory tests) sees spread of tumor cells beyond the thymus during examination of the tumor sample under the microscope.

The treatment and outlook for thymic cancers depend to a large extent on their stage.

The system most often used in staging thymomas is called the *Masaoka system*, after the doctor who, in 1981, first proposed it. This system separates thymomas into four stages.

Stage I: The thymoma is noninvasive. That is, it has not spread beyond the thymus.

Stage II: The thymoma invades beyond the *capsule* (outer boundary of the thymus) and into the nearby fatty tissue or to the *pleura* (outer covering of the lung).

Stage III: The thymoma extends into the neighboring tissues or organs of the lower neck or upper chest area, including the *pericardium* (covering of the heart), the lungs, or the main blood vessels leading into or exiting from the heart.

Stage IVA: The thymoma has spread widely throughout the pleura and/or pericardium.

Stage IVB: The thymoma has spread to distant organs.

The *prognosis* (the outlook for chances of survival)) following diagnosis and treatment of a thymoma depends to a large extent on its stage. For Stage I tumors, 5-year survival rates range from 89%-95%; Stage II for approximately 86%; Stage III for 70%-80%; and Stage IV disease for 50%-60%.

Because thymic carcinoid tumors and thymic carcinomas are so rare, no staging system has been developed for them. Some doctors use the Masaoka system for thymic carcinoma as well as for thymomas. The overall 5-year survival rate for thymic carcinoids is about 60% and the overall 5-year survival rate for thymic carcinomas is about 35%.

The 5-year survival rate refers to the percent of patients who live at least 5 years after their cancer is diagnosed. Many of these patients live much longer than 5 years after diagnosis, and 5-year rates are used to produce a standard way of discussing prognosis. Five-year *relative* survival rates exclude from the calculations patients dying of other diseases, and are considered to be a more accurate way to describe the prognosis for patients with a particular type and stage of cancer. Of course, 5-year survival rates are

based on patients diagnosed and initially treated more than 5 years ago. Improvements in treatment often result in a more favorable outlook for recently diagnosed patients.

How Is Thymus Cancer Treated?

After a thymic tumor is found and staged, the cancer care team will recommend one or more treatment options. Factors important in choosing a treatment include the type and stage of the cancer, the patient's overall physical health, and other medical problems. In the case of thymic cancer, the rarity with which this tumor occurs has made it difficult to accurately predict the effectiveness of treatment strategies. Much about the best way to treat this cancer remains to be determined.

It is often a good idea to seek a second opinion. A second opinion can provide more information and help the patient feel more certain about the treatment plan that is chosen.

Surgery for Thymic Cancers

Depending on the stage of a thymic tumor (either thymoma, thymic carcinoid, or thymic carcinoma), surgery may be used to remove the cancer and some of the surrounding tissue. Surgical *resection* (removal), is typically performed through a *median sternotomy*, an incision that splits the *sternum* (breast bone), permitting thorough removal of the thymus and tumor. Total resection of the thymus with complete removal of the thymoma is possible in nearly all patients with Stage I and II thymomas, and in 27%-44% of Stage III thymomas. Patients with Stage II or III thymomas may also undergo removal of nearby structures such as the pleura, pericardium, lung, nerves, and segments of the *superior vena cava* (a large vein leading to the heart), if involved with disease. Stage IV thymomas are sometimes partially resected, but complete removal of the tumor is not possible.

Chemotherapy for Thymic Cancers

Chemotherapy uses anticancer drugs that are given *intravenously* (in a vein) or by mouth. These drugs enter the bloodstream and reach all areas of the body, making this treatment useful for cancer that has spread or metastasized to organs beyond the thymus. Thymic tumor cells are particularly susceptible to some of these anticancer drugs. They may be given as *adjuvant* therapy, which means along with other treatment to improve overall effectiveness. In the case of thymic cancers, chemotherapy as adjuvant therapy is given following surgery.

Sometimes chemotherapy is given before surgery to shrink a tumor. Given before surgery, it's referred to as *neoadjuvant* (preliminary therapy).

Several anticancer drugs have been used in the treatment of thymomas and thymic carcinomas. The agents most effective when given alone are doxorubicin (Adriamycin), cisplatin, ifosfamide, and corticosteroids. Often, these drugs are given in combination to increase their effectiveness. Combinations used in the treatment of thymic cancer include cisplatin, doxorubicin, and cyclophosphamide, and the combination of cisplatin, doxorubicin, cyclophosphamide, and vincristine.

These chemotherapy drugs kill cancer cells but can also damage normal cells. Careful attention is given to avoid or reduce the side effects of chemotherapy. These side effects depend on the type and dose of drugs given and the length of time they are taken. Drugs used in cancer chemotherapy specifically attack cells that are rapidly dividing. These drugs are useful because cancer cells spend more of their time dividing and reproducing than normal cells do.

However, there are some normal tissues such as the bone marrow, the lining of the mouth and intestines, and the hair follicles which also grow rapidly in order to replace cells that wear out. These rapidly reproducing cells are the ones most likely to be affected by chemotherapy. As a result, a patient may have hair loss, mouth sores, lowered resistance to infection (due to low white blood cell counts), easy bruising or bleeding (due to low blood platelets), and fatigue (due to low red blood cells). Loss of appetite, nausea, and vomiting result in part from damage to intestinal cells, but effects of certain drugs on areas of the brain controlling appetite and vomiting also contribute to these problems. These side effects are temporary and go away after treatment is finished.

If you have side effects, your cancer care team can suggest steps to ease their impact. For example, drugs can be given along with the chemotherapy to prevent or reduce nausea and vomiting.

In addition to these temporary side effects, some chemotherapy drugs can permanently damage certain organs such as the heart or kidneys. These possible risks are carefully balanced against their benefits, and, the health of these organs is carefully monitored during treatment. If serious organ damage occurs, the responsible drug is discontinued and replaced with another.

Radiation Therapy for Thymic Cancers

Radiation therapy uses high energy radiation to kill cancer cells. *External beam radiation therapy* uses radiation focused on the cancer, delivered from outside the body. This type of radiation therapy is often used to treat thymic cancer. After surgery, radiation therapy can be used as adjuvant therapy to kill very small deposits of cancer that could not be seen and removed during surgery, particularly in cases of Stage II, III, and IV disease. Radiation therapy may also be used to decrease the size of larger tumors before surgery. Radiation therapy is not as effective when used alone to treat thymic cancer, although it may be the only option for patients too ill to tolerate an operation.

Side effects of radiation therapy may include mild skin problems, upset stomach, or tiredness. Often these side effects go away after a short while. Radiation may also make the side effects of chemotherapy worse. Chest radiation therapy may cause lung damage and lead to breathing difficulty and shortness of breath. Also, because the *esophagus* (swallowing tube) goes from the mouth to the stomach through the middle of the chest, it will be included in the radiation field. The radiation may make swallowing uncomfortable. This goes away two to three weeks after the radiation treatments are finished.

Do not hesitate to talk with your doctor about these possible side effects since there are often ways to remedy them.

Clinical Trials

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

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

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

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

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

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

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

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

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

- What is the purpose of the study?
- What kinds of tests and treatments does the study involve?
- What does this treatment do?
- What is likely to happen in my case with, or without, this new research treatment?
- What are my other choices and their advantages and disadvantages?
- How could the study affect my daily life?
- What side effects can I expect from the study? Can the side effects be controlled?
- Will I have to be hospitalized? If so, how often and for how long?
- Will the study cost me anything? Will any of the treatment be free?
- If I am harmed as a result of the research, what treatment would I be entitled to?
- What type of long-term follow-up care is part of the study?
- Has the treatment been used to treat other types of cancers?

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

Treatment of Thymus Cancers by Stage and Type

Stage I Thymoma: Most patients with Stage I thymic cancer have their cancer surgically removed through median sternotomy as described in the section for Thymus Cancers, above. Long-term survival following complete removal of a Stage I thymoma approaches 100%.

Radiation therapy is not used in treating Stage I thymomas except for cases in which complete removal is not possible (for example, in patients with other serious medical conditions who are unable to undergo a major operation). For these patients, radiation after partial removal, or radiation instead of surgery are options. However, the best outlook for cure is provided by complete removal whenever possible.

Stages II, III, IV Thymoma: Patients with Stage II and III thymic cancer are typically treated with surgical removal of the thymus and, when possible, any other tissues to which the tumor has spread. *Postoperative* (after surgery) radiation is then given to reduce the risk of recurrence and spread of the tumor.

Some Stage III and most Stage IV thymomas have spread too widely to be completely removed. These tumors are usually treated by radiation therapy. Sometimes the tumor is wrapped around the major artery (a*orta*) or vein (*vena cava*) in the middle of the chest. Many doctors feel it is important to remove as much of the tumor as is possible in people with stage III disease (where the tumor has not spread widely). This gives the post-operative radiation a better chance of getting rid of the remaining cancer. For stage IV disease (where the cancer has spread more widely), some studies suggest that *debulking* (removing as much tumor as possible) before radiation therapy improves survival rates, but the value of debulking has not yet been proven.

Other options for *unresectable* (unable to be removed) thymomas include chemotherapy, or chemotherapy plus radiation therapy. In some cases, *neoadjuvant* chemotherapy, given before surgery, will shrink a tumor enough so that it can then be completely resected After the surgery, radiation therapy will be given.

Recurrent Thymoma: Treatment for thymoma that has recurred after initial treatment depends on what the original treatment was. For example, in patients originally treated with radiation, use of more radiation after recurrence might cause severe damage to normal tissues. In most cases, options for recurrent thymoma include radiation or chemotherapy. The value of surgery in this situation is debated, but some studies suggest that if recurrences are not too widespread, surgical removal offers the best chance of long-term survival.

Treatment of Thymic Carcinoma: The usual treatment is removal of as much of the thymic tumor as possible, followed by radiation and chemotherapy. Sometimes, chemotherapy is given before surgery.

Treatment of Thymic Carcinoid: Complete surgical removal, when possible, is the main strategy. If complete removal is not possible, radiation therapy is given after debulking. Radiation is also used in treating local recurrences. Chemotherapy is sometimes used in treating thymic carcinoids with distant spread.

What Should You Ask Your Doctor About Thymus Cancer?

As noted earlier, it is important to have frank, open discussions with your cancer care team. They want to answer all of your questions, no matter how trivial they might seem.
For instance, consider these questions:

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

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

What Happens After Treatment For Thymus Cancer?

Follow-up Tests: The health care team will decide which tests should be done and how often. Chest x-rays and other imaging studies, such as CT scans and MRI scans, may be done to watch for local or distant recurrences.

New Symptoms : It is important for the patient to report any new symptoms to the doctor right away since these may be a sign of a recurrence or of side effects of treatment.

What's New In Thymus Cancer Research And Treatment?

There is always research going on in the area of thymic tumors. Scientists are looking for causes and ways to prevent thymic tumors, and doctors are working to improve treatments. Because thymic tumors are relatively rare, more information from clinical trials is needed to decide which of the currently available treatments are best for each type and stage. For example, the role of chemotherapy in treating thymomas is still being explored.

In addition, new treatments are being developed and tested.

More accurate ways of predicting the aggressiveness of each tumor are being studied so that treatment can be more appropriately selected for each patient.

Removing or destroying all of the cancer cells is not the only consideration in treating patients with thymomas. Some paraneoplastic syndromes may persist even after the tumor has been treated. Researchers are studying the causes of these syndromes and the best ways to treat them.

Additional Resources

National Organizations and Web Sites

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

National Cancer Institute
Office of Cancer Communications
Telephone: 1-800-4-CANCER

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

Additional American Cancer Society Information

After Diagnosis: A Guide for Patients and Families (Booklet Code #9440.00)
Questions and Answers About Pain Control (Booklet; Code #4518.00)

Other Publications*

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

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

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

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