Nasopharyngeal Cancer

What is cancer?

The body is made up of trillions of living cells. Normal body cells grow, divide into new cells, and die in an orderly fashion. 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 always named for the place where it started. For example, breast cancer that has spread to the liver is still called breast cancer, not liver cancer. Likewise, prostate cancer that has spread to the bone is metastatic 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.

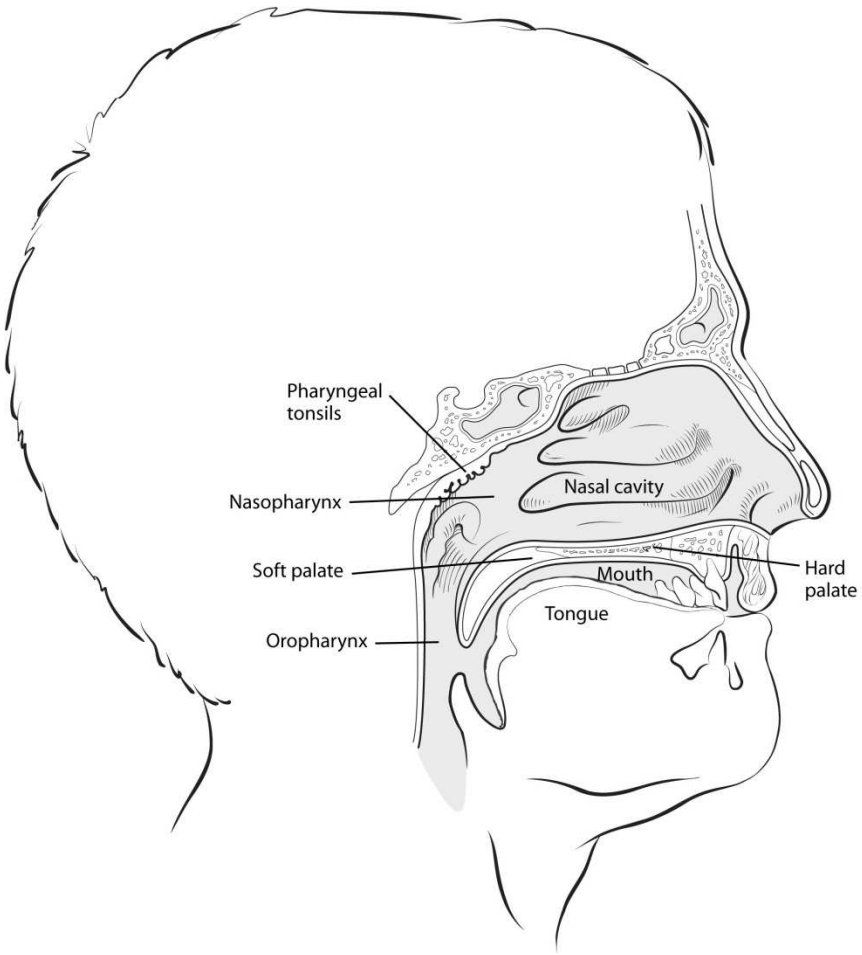
Not all tumors are cancerous. Tumors that aren't cancer are called benign. Benign tumors can cause problems – they can grow very large and press on healthy organs and tissues. But they cannot grow into (invade) other tissues. Because they can't invade, they also can't spread to other parts of the body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

What is nasopharyngeal cancer?

Nasopharyngeal cancer is a cancer that starts in the nasopharynx, the upper part of the throat behind the nose and near the base of skull. To understand nasopharyngeal cancer, it helps to know about the structure and function of the nasopharynx.

About the nasopharynx

The nasopharynx is the upper part of the throat (pharynx) that lies behind the nose. It is a box-like chamber about 1½ inches on each edge. It lies just above the soft part of the roof of the mouth (soft palate) and just in back of the nasal passages.



The nasopharynx serves as a passageway for air from the nose to the throat (and eventually to the lungs).

Nasopharyngeal tumors

Several types of tumors can develop in the nasopharynx. Some of these tumors are benign (non-cancerous), but others are malignant (cancerous). It is important to discuss what type of tumor you might have with your doctor.

Benign nasopharyngeal tumors

Benign tumors of the nasopharynx are fairly rare and tend to occur in children and young adults. These tumors do not spread to other parts of the body and are usually not life-threatening. They include tumors or malformations of the vascular (blood-carrying) system, such as *angiofibromas* and *hemangiomas*, and benign tumors of minor salivary glands within the nasopharynx.

Treatment of these tumors (if it is needed) is different from that for cancerous nasopharyngeal tumors and is not discussed further in this document. If you have one of these tumors, you and your doctor will talk about what treatments might be appropriate for you.

Nasopharyngeal cancers

These tumors can invade surrounding tissues and spread to other parts of the body.

Nasopharyngeal carcinoma (NPC): This is by far the most common malignant tumor of the nasopharynx. A *carcinoma* is a cancer that starts in epithelial cells – the cells lining the internal and external surfaces of the body. Most of the rest of this document refers to NPC.

There are 3 types of NPC, based on how the cancer cells look under the microscope:

- Keratinizing squamous cell carcinoma
- Non-keratinizing differentiated carcinoma
- Undifferentiated carcinoma

Each of these types is seen more often in some areas of the world than in others. In southern China, where NPC is much more common, nearly all cases are the undifferentiated type. In the United States, where NPC is rare, about 1 out of 4 cases are the keratinizing type.

Even though these types look different when seen under a microscope, studies have shown they start from the same cell type – the epithelial cells that line the surface of the nasopharynx. The treatment is also usually the same for all types of NPC. The stage of the cancer – how far it has grown and spread – is often more important than its type in predicting a person's outlook (prognosis).

Many NPCs also contain lots of immune system cells, especially lymphocytes. The term *lymphoepithelioma* is sometimes used to describe an undifferentiated NPC with many lymphocytes among the cancer cells. The presence of these cells does not usually affect the choice of treatment options. But they may be a clue to developing new treatments since they may represent the body's attempt to “reject” the tumor (for more information, see the section “What's new in nasopharyngeal cancer research and treatment?”).

Other cancers in the nasopharynx: Other types of cancers can arise in the nasopharynx.

Lymphomas can sometimes start in the nasopharynx. They are cancers of immune system cells called *lymphocytes*, which are found throughout the body, including in the nasopharynx. These cancers are discussed in our document *Non-Hodgkin Lymphoma*.

Adenocarcinoma and *adenoid cystic carcinoma* are cancers that can develop in the minor salivary glands in the nasopharynx, but these cancers are more commonly found in the nose (nasal cavity) or mouth (oral cavity). More information on these cancers can be found in our

documents *Oral Cavity and Oropharyngeal Cancer*, *Nasal Cavity and Paranasal Sinuses Cancer*, and *Salivary Gland Cancer*.

What are the key statistics about nasopharyngeal cancer?

Nasopharyngeal cancer (NPC) is fairly rare. In most parts of the world (including the United States), there is less than one case for every 100,000 people each year. In 2013, about 2,900 cases will occur in the United States.

This cancer is, however, much more common in certain parts of Asia and North Africa, particularly in southern China. It is also more common among Inuits of Alaska and Canada, and among some immigrant groups in the United States, such as recent Chinese and Hmong immigrants.

The risk of NPC increases slowly throughout life, but it can occur in people of any age, including children. About half of the people with NPC in the United States are younger than 55 years old.

What are the risk factors for nasopharyngeal cancer?

A risk factor is anything that affects a person’s 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 risk factors don’t tell us everything. Having a risk factor, or even several risk factors, does not mean that you will get the disease. And many people who get the disease may have few or no known risk factors.

Scientists have found several risk factors that make a person more likely to develop nasopharyngeal cancer (NPC).

Gender

NPC is found about twice as often in males as it is in females.

Race/ethnicity and where you live

NPC is most common in southern China (including Hong Kong), Singapore, Vietnam, Malaysia, and the Philippines. It is also fairly common in Northwest Canada and Greenland.

People of south China have a lower risk of NPC if they move to another area that has lower rates of NPC (like the US or Japan), but their risk is still higher than for people who are native to areas with lower risk. Over time, their risk seems to go down. The risk also goes down in new generations. Although whites born in the United States have a low risk of NPC, whites born in China have a higher risk.

In the United States, NPC is most common in Asian and Pacific Islanders (particularly Chinese Americans), followed by American Indian and Alaskan natives, African Americans, whites, and Hispanics/Latinos.

Diet

People who live in parts of Asia, northern Africa, and the Arctic region where NPC is common typically eat diets very high in salt-cured fish and meat. Indeed, the rate of this cancer is dropping in southeast China as people begin eating a more Westernized diet. In contrast, some studies have suggested that diets high in fruits and vegetables may lower the risk of NPC.

Epstein-Barr virus infection

Almost all NPC cells contain parts of the Epstein-Barr virus (EBV), and most people with NPC have evidence of infection by this virus in their blood. Infection with EBV is very common throughout the world, often occurring in childhood. In the United States, where infection with this virus tends to occur in slightly older children, it often causes infectious mononucleosis (“mono”), usually in teens.

The link between EBV infection and NPC is complex and not yet completely understood. EBV infection alone is not enough to cause NPC, since infection with this virus is very common and this cancer is rare. Other factors, such as a person’s genes, may affect how the body deals with EBV, which in turn may affect how EBV contributes to the development of NPC.

Genetic factors

A person’s genes may affect their risk for NPC. For example, just as people have different blood types, they also have different tissue types. Studies have found that people with certain inherited tissue types have an increased risk of developing NPC. Tissue types affect immune responses, so this may be related to how a person's body reacts to EBV infection.

Family history

Family members of people with NPC are more likely to get this cancer. It is not known if this is because of inherited genes, shared environmental factors (such as the same diet or living quarters), or some combination of these.

Other possible risk factors

Tobacco and alcohol use: Most (but not all) studies have found that smoking may contribute to the development of NPC, especially the keratinizing type. Some studies have linked heavy alcohol intake to this type of cancer. More research is needed to define these links, but they seem to be much weaker than the link between tobacco and alcohol use and most other types of cancers that start in the throat.

Workplace exposures: Some studies have suggested that workplace exposure to formaldehyde or wood dust may increase the risk of NPC. Still, not all studies have shown this and this link isn't clear.

Do we know what causes nasopharyngeal cancer?

The exact cause of most cases of nasopharyngeal cancer (NPC) is not known. But scientists have found that the disease is linked with certain diets, infections, and inherited characteristics, which are described in the section called "What are the risk factors for nasopharyngeal cancer? Research is being done to learn more about these causes.

In recent years, scientists have studied how the Epstein-Barr virus (EBV) may cause cells in the nasopharynx to become cancerous, but much still remains to be learned. In developed countries, most people infected with EBV develop only infectious mononucleosis (mono), and their immune system is able to recognize and destroy the virus. These people recover without any long-term problems. But in some cases, pieces of viral DNA mix with the DNA of cells in the nasopharynx.

DNA is the chemical in each of our cells that makes up our genes, the instructions for how our cells function. We usually look like our parents because they are the source of our DNA. But DNA affects more than how we look. Some genes contain instructions for controlling when cells grow and divide into new cells. Viruses such as EBV also contain DNA. When a cell is infected with the virus, the viral DNA may mix with the normal human DNA. EBV DNA may instruct the cells of the nasopharynx to divide and grow in an abnormal way.

But EBV infection only rarely results in NPC, so other factors probably play a role in whether or not it causes cancer. Eating a diet high in salt-cured fish and meat seems to increase the ability of EBV to cause NPC. Studies show that foods preserved in this way may

produce chemicals that can damage DNA. The damaged DNA alters a cell's ability to control its growth and replication.

Some studies suggest that inheriting certain tissue types may contribute to a person's risk of developing NPC. Because the tissue type plays a role in the function of the immune system, some scientists suspect that an abnormal immune reaction to EBV infection may be involved. The details of how certain tissue types might increase NPC risk are still being worked out.

Can nasopharyngeal cancer be prevented?

Most people in the United States who develop nasopharyngeal cancer (NPC) have no avoidable risk factors, so their cancers could not have been prevented. The possible links with tobacco and heavy alcohol use are not clear, so it's not known if avoiding these can lower a person's risk of NPC. However, both tobacco and alcohol use have clearly been linked to a number of other cancers, as well as other health problems, so avoiding them can have many health benefits.

Because certain dietary factors have been linked with NPC risk, reducing or eliminating some types of food may lower the number of cases in parts of the world where NPC is common, such as southern China, northern Africa, and the Arctic region. Descendants of Southeast Asians who immigrated to the United States and eat a typical American diet, for example, have a lower risk of developing NPC. But these dietary factors are not thought to account for all cases of NPC in most other parts of the world. Other factors, such as genetics, are likely to play a part as well.

Can nasopharyngeal cancer be found early?

In the United States and other countries where nasopharyngeal cancer (NPC) is fairly rare, most doctors do not recommend routine screening for this cancer. (Screening is testing for cancer in people without any symptoms.) There are no simple, non-invasive exams or blood tests that can reliably find this cancer early.

But in some parts of the world such as China, where NPC is common, some people are being screened routinely for this cancer. They are first selected because their blood shows evidence of infection with the Epstein-Barr virus, although EBV infection is much more common than NPC. They are given regular exams of the nasopharynx and neck. This approach can also be used in families when one member has developed NPC. It isn't known if this strategy lowers the death rate from this cancer.

Sometimes NPC can be found early if it causes symptoms that make patients seek medical attention. The symptoms may even seem unrelated to the nasopharynx (for example, a constant feeling of fullness in one ear). But in most cases, NPCs may not cause symptoms until they have reached an advanced stage.

Signs and symptoms of nasopharyngeal cancer

About 3 out of 4 people with NPC complain of a lump or mass in the neck when they first see their doctor. There may be lumps on both sides of the neck towards the back. The lumps are usually not tender or painful. This is caused by the cancer spreading to lymph nodes in the neck, causing them to become larger than normal. (Lymph nodes are glands or organs that contain collections of immune system cells that are found throughout the body. Normally, they are smaller than the size of a pea.)

Other possible symptoms of NPC include:

- Hearing loss, ringing in the ear, or feeling of fullness in the ear (especially on one side only)
- Ear infections that keep coming back
- Nasal blockage or stuffiness
- Nosebleeds
- Headache
- Facial pain or numbness
- Trouble opening the mouth
- Blurred or double vision

Ear infections are common in children, but are less common in adults. If you develop an infection in one ear and you have not had ear infections in the past, it is important that a specialist examine your nasopharynx. This is especially true if you don't have an upper respiratory tract infection (like a “cold”) along with the ear infection.

Many of the symptoms and signs of NPC are more often caused by other, less serious diseases. Still, if you have any of these problems, it's important to see your doctor right away so the cause can be found and treated, if needed.

How is nasopharyngeal cancer diagnosed?

Nasopharyngeal cancer (NPC) is most often diagnosed when a person goes to a doctor because of symptoms such as a lump in the neck. The doctor will take a history, do an exam, and then may refer the patient to a specialist and/or order some tests.

Medical history and physical exam

If you have any signs or symptoms that suggest you might have nasopharyngeal cancer, your doctor will want to get your complete medical history to learn about your symptoms and any possible risk factors, including your family history.

A physical exam will be done to look for signs of NPC or other health problems. During the exam, the doctor will pay special attention to the head and neck area, including the nose, mouth, and throat; the facial muscles, and the lymph nodes in the neck.

If your doctor suspects you may have a tumor or other problem in the nose or throat, he or she may order imaging tests (such as CT or MRI) to look at the head and neck area more closely. Your doctor may also refer you to an otolaryngologist (a doctor specializing in ear, nose, and throat problems, also sometimes called an ENT doctor), who will do a more thorough exam of the nasopharynx. The nasopharynx is a difficult area to examine. Most other kinds of doctors do not have the specialized training or equipment to do a thorough exam of this part of the body.

Exams of the nasopharynx

The nasopharynx is located deep inside the head and is not easily seen, so special techniques are needed to examine this area. There are 2 main types of exams used to look inside the nasopharynx for abnormal growths, bleeding, or other signs of disease. Both types of exams are usually done in the doctor's office.

- For *indirect nasopharyngoscopy*, the doctor uses special small mirrors and lights placed at the back of your throat to look at the nasopharynx and nearby areas.
- For *direct nasopharyngoscopy*, the doctor uses a fiber-optic scope known as a *nasopharyngoscope* (a flexible, lighted, narrow tube inserted through the nose) to look directly at the lining of the nasopharynx. You will have numbing medicine sprayed into your nose before the exam to make it easier. This is the method most often used to carefully examine the nasopharynx.

If a tumor starts under the lining of the nasopharynx (in the tissue called the submucosa), it may not be possible to see it directly on physical exam, which is why imaging tests such as CT scans (see below) may be needed as well.

Biopsy

Symptoms and the results of exams can suggest that a person might have NPC, but the actual diagnosis is made by removing cells from an abnormal area and looking at them under a microscope. This is known as a *biopsy*. Different types of biopsies may be done, depending on where the abnormal area is.

Endoscopic biopsy

If a suspicious growth is found in the nasopharynx during an exam, the doctor may remove a biopsy sample with small instruments and the aid of a fiber-optic scope. Often, biopsies of the nasopharynx are done in the operating room as an outpatient procedure. The sample is then sent to a lab, where a doctor called a pathologist looks at it under a microscope. If the biopsy sample contains cancer cells, the pathologist sends back a report describing the type of the cancer.

If the cancer is hidden beneath the surface of the nasopharynx, NPC may not be visible during an exam. If a person has symptoms suggesting NPC but nothing abnormal is seen on exam, the doctor may take several samples of normal-looking tissue, which may be found to contain cancer cells when looked at under the microscope.

Fine needle aspiration (FNA) biopsy

An FNA biopsy may be used if you have a suspicious lump in or near your neck. For this procedure, the doctor uses a thin, hollow needle attached to a syringe to aspirate (withdraw) a few drops of fluid containing cells and tiny fragments of tissue. A local anesthetic (numbing medicine) may be used on the skin where the needle will be inserted. In some cases, no anesthetic is needed.

The doctor places the needle directly into the mass for about 10 seconds and withdraws cells and a few drops of fluid. The cells are then looked at under a microscope to see if they are cancerous.

In patients with an enlarged lymph node in the neck area, an FNA biopsy can help determine if the enlargement is caused by a response to an infection, the spread of cancer from somewhere else (such as the nasopharynx), or a cancer that begins in lymph nodes – called a *lymphoma*. If the cancer started somewhere else, the FNA biopsy alone might not be able to tell where it started. But if a patient already known to have NPC has enlarged neck lymph nodes, FNA can help determine if the spread of NPC caused the lymph node swelling.

Imaging tests

Imaging tests use x-rays, magnetic fields, sound waves, or radioactive particles 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 cancerous, to learn how far cancer may have spread, and to help determine if treatment has been effective.

Chest x-ray

If you have been diagnosed with NPC, a plain x-ray of your chest may be done to see if the cancer has spread to your lungs. This is very unlikely unless your cancer is far advanced.

This x-ray can be done in any outpatient setting. If the results are normal, you probably don't have cancer in your lungs.

Computed tomography (CT) scan

The CT scan is an x-ray test that produces detailed cross-sectional images of your body. Instead of taking one x-ray, a CT scanner takes many pictures as it rotates around you. A computer then combines these into images of slices of the part of your body that is being studied.

Before the pictures are taken, you may get an IV (intravenous) line through which a kind of contrast dye (IV contrast) is injected. This helps better outline structures in your body. You may also 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. It may not be needed for CT scans of the nasopharynx.

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 a contrast material used for x-rays.

You need to lie still on a table while the scan is being done. During the test, the table slides in and out of the scanner, a ring-shaped machine that completely surrounds the table. You might feel a bit confined by the ring you have to lie in while the pictures are being taken.

A CT scan of the head and neck can provide information about the size, shape, and position of a tumor and can help find enlarged lymph nodes that might contain cancer. CT scans or MRIs are important in looking for cancer that may have grown into the bones at the base of the skull. This is a common place for nasopharyngeal cancer to grow. CT scans can also be used to look for tumors in other parts of the body.

Magnetic resonance imaging (MRI) scan

Like CT scans, MRI scans provide detailed images of soft tissues in the body. But MRI scans use radio waves and strong magnets instead of x-rays. The energy from the radio waves is absorbed and then released in a pattern formed by the type of body tissue and by certain diseases. A computer translates the pattern into very detailed images of parts of the body. A contrast material called *gadolinium* is often injected into a vein before the scan to better see details.

MRI scans may be a little more uncomfortable than CT scans. They take longer – often up to an hour. You may be asked to lie on a table that slides inside a large tube, which is confining and can upset people with a fear of enclosed spaces. Special, "open" MRI machines can sometimes help with this if needed. The MRI machine makes buzzing and clicking noises that you may find disturbing. Some places will provide earplugs to help block this noise out.

Like CT scans, MRIs can be used to try to determine if the cancer has grown into structures near the nasopharynx. MRIs are a little better than CT scans at showing the soft tissues in the nose and throat, but they're not quite as good for looking at the bones at the base of the skull, a common place for nasopharyngeal cancer to grow.

Positron emission tomography (PET) scan

For a PET scan, you receive an injection of a form of radioactive sugar (known as fluorodeoxyglucose or FDG). The amount of radioactivity used is very low. Because cancer cells in the body are growing rapidly, they absorb large amounts of the sugar. After about an hour, you are moved onto a table in the PET scanner. You lie on the table for about 30 minutes while a special camera creates a picture of areas of radioactivity in the body. The picture is not finely detailed like a CT or MRI scan, but it provides helpful information about your whole body.

Your doctor may use this test to see if the cancer has spread to your lymph nodes. It can also help give the doctor a better idea of whether an abnormal area on a chest x-ray may be cancer. A PET scan can also be useful if your doctor thinks the cancer may have spread but doesn't know where.

Some machines are able to do both a PET and CT scan at the same time (PET/CT scan). This lets the doctor compare areas of higher radioactivity on the PET with the more detailed appearance of that area on the CT.

Blood tests

Blood tests are not used to diagnose NPC, but they may be done for other reasons, such as to help determine whether the cancer may have spread to other parts of the body.

Routine blood counts and blood chemistry tests

Routine blood tests can help determine a patient's overall health. These tests can help diagnose malnutrition, anemia (low red blood counts), liver disease, and kidney disease. And they may suggest the possibility of spread of the cancer to the liver or bone, which may prompt further testing.

In people getting chemotherapy, blood tests are important to see if the treatment is damaging the bone marrow (where new blood cells are made), liver, and kidneys.

Epstein-Barr virus (EBV) DNA levels

In some patients, the blood level of EBV DNA may be measured before and after treatment to help show how effective treatment is.

How is nasopharyngeal cancer staged?

The stage of a cancer is a description of how far it has spread. The stage of nasopharyngeal cancer (NPC) is one of the most important factors in selecting treatment options and estimating a patient's outlook for recovery and survival (prognosis).

The stage of NPC is determined by the results of the physical exam, imaging tests (CT or MRI scan, etc.) and other tests, which are described in the section “How is nasopharyngeal cancer diagnosed?”, and by the results of surgery if it has been done.

If you have NPC, ask your cancer care team to explain the stage in a way you can understand. By knowing all you can about the stage of your cancer, you can take a more active role in making informed decisions about your treatment.

The American Joint Committee on Cancer (AJCC) TNM system

A staging system is a standard way for the cancer care team to sum up the extent of a cancer’s spread. The most common system used to describe the spread of NPC is the TNM system created by the American Joint Committee on Cancer (AJCC). This system contains 3 key pieces of information:

- **T** describes whether the main (primary) **tumor** has invaded other organs or tissues near the nasopharynx.
- **N** describes whether the cancer has spread to nearby (regional) lymph **nodes** (bean-sized collections of immune system cells throughout the body).
- **M** indicates whether the cancer has **metastasized** (spread) to other parts of the body. (The most common sites of distant NPC spread are the lungs, liver, and bones.)

The numbers or letters that appear after T, N, and M provide more details about each of these factors:

- The numbers 0 through 4 indicate further spread.
- The letter X means “cannot be assessed” because the information is not available.
- The letters “is” after the T stand for “in situ,” which means the tumor is still only in the layer of cells where it started and has not yet invaded deeper.

T groups for nasopharyngeal cancer

TX: Primary tumor cannot be assessed because of incomplete information.

T0: No evidence of a primary tumor.

Tis: Carcinoma in situ (cancer cells are found only in the surface layer of the nasopharynx but have not invaded into deeper layers).

T1: Tumor is in the nasopharynx. The cancer may also have grown into the oropharynx (the part of the throat in the back of the mouth) and/or nasal cavity but no farther.

T2: The cancer has grown into the tissues of left or right sides of the upper part of the throat (but not into bone).

T3: The tumor has grown into the sinuses and/or the bones nearby.

T4: The tumor has grown into the skull and/or cranial nerves (nerves in the head that lie near the nasopharynx and have special functions such as vision, smell, and eye movement), the hypopharynx (lower part of the throat), or the eye or its nearby tissues.

N groups for nasopharyngeal cancer

NX: Nearby lymph nodes cannot be assessed due to incomplete information.

N0: No spread to nearby lymph nodes.

N1: Spread to 1 or more lymph nodes on one side of the neck; OR spread to lymph nodes behind the throat (called *retropharyngeal lymph nodes*) on either side of the neck. In either case, no lymph node is larger than 6 cm (about 2½ inches) across.

N2: Spread to lymph nodes on both sides of the neck, none larger than 6 cm across.

N3: Spread to lymph nodes that are either:

- **N3a:** larger than 6 cm across
- **N3b:** located in the shoulder area just above the collarbone (this area is called the *supraclavicular fossa*)

M groups for nasopharyngeal cancer

M0: The cancer has not spread to tissues or organs far away from the nasopharynx.

M1: The cancer has spread to tissues or organs far away from the nasopharynx.

TNM stage grouping

Once the T, N, and M categories have been determined, this information is combined in a process called *stage grouping.* The stage is expressed as the number 0 (zero) or the Roman numerals I to IV (1-4). Some stages are subdivided with letters.

Stage 0: Tis, N0, M0: The cancer is “in situ.” Cancer cells are only in the surface layer of the nasopharynx but have not yet grown into deeper layers (Tis). The cancer has not spread to nearby lymph nodes (N0) or distant sites (M0).

Stage I: T1, N0, M0: The tumor is in the nasopharynx and may have spread to soft tissues of the nasal cavity and/or the oropharynx (T1). It has not spread to nearby lymph nodes (N0) or distant sites (M0).

Stage II: Either:

T2, N0, M0: The tumor has grown into the tissues of the left or right sides of the upper part of the throat (T2). It has not spread to nearby lymph nodes (N0) or distant sites (M0).

OR

T1 or T2, N1, M0: The tumor may still be confined to the nasopharynx, or it may have extended to the soft tissues of the nasal cavity or the oropharynx (T1), or the left or right sides of the upper part of the throat (T2). It has spread to one or more nearby lymph nodes, none of which is larger than 6 cm (about 2½ inches) across. These lymph nodes are either neck lymph nodes on one side or lymph nodes behind the throat (*retropharyngeal* lymph nodes) on either side (N1). The cancer has not spread to distant sites (M0).

Stage III: Either:

T3, N0 to N2, M0: The tumor has spread to the sinuses or the bones near the nasopharynx (T3). It may or may not have spread to lymph nodes in the neck or behind the throat, but none of these lymph nodes is larger than 6 cm across (N0 to N2). It has not spread to distant sites (M0).

OR

T1 or T2, N2, M0: The tumor may still be confined to the nasopharynx, or it may have grown into the soft tissues of the nasal cavity or the oropharynx (T1), or the left or right sides of the upper part of the throat (T2). The tumor has spread into nearby neck lymph nodes on both sides, with none of the lymph nodes larger than 6 cm across (N2). The cancer has not spread to distant sites. (M0).

Stage IVA: T4, N0 to N2, M0: The tumor has grown into the skull and/or cranial nerves, the hypopharynx (lower part of the throat), the eye, or its nearby tissues (T4). It may or may not have spread to nearby lymph nodes in the neck, none of which is larger than 6 cm across (N0 to N2). It has not spread to distant sites (M0).

Stage IVB: Any T, N3, M0: The tumor may or may not have extended into nearby soft tissues or bones (any T). It has spread to lymph nodes that are larger than 6 cm across and/or are located in the shoulder area above the collarbone (N3). The cancer has not spread to distant sites (M0).

Stage IVC: Any T, any N, M1: The tumor may or may not have extended into nearby soft tissues or bones (any T). It may or may not have spread to nearby lymph nodes (any N). It has spread to distant sites (M1).

Survival rates for nasopharyngeal cancer by stage

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 decide that you do not want to read about them, 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).

Relative survival rates (such as the numbers below) adjust for patients with nasopharyngeal cancer (NPC) who die from other causes, such as heart disease. They are considered to be a more accurate way to describe the outlook for patients with a particular type and stage of cancer.

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 more favorable outlook for people now being diagnosed with NPC.

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 other factors may affect a person's outlook, such as their age, overall health, the treatment received, and how well the cancer responds to treatment. Your doctor can tell you how the numbers above may apply to you, as he or she is familiar with your particular situation.

The numbers below were published in 2010 in the 7th edition of the AJCC Cancer Staging Manual and are based on people diagnosed between 1998 and 1999.

Stage	**Relative 5-year survival rates**
I	72%
II	64%
III	62%
IV	38%

How is nasopharyngeal cancer 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.

General treatment information for nasopharyngeal cancer

After the cancer is found and staged, your cancer care team will discuss treatment options (choices) with you. Depending on the stage of the cancer, your overall health, and other factors, your treatment options may include:

- Surgery
- Radiation therapy
- Chemotherapy
- Targeted therapy

Depending on the stage of the cancer, some of these treatments may be combined. For most nasopharyngeal cancers (NPCs), a combination of radiation therapy and chemotherapy is used.

Based on the stage of the cancer, you may have different types of doctors on your treatment team. These doctors may include:

- An otolaryngologist (also known as an ear, nose, and throat, or ENT doctor): a surgeon who treats certain diseases of the head and neck.
- A radiation oncologist: a doctor who treats cancer with radiation therapy.
- A medical oncologist: a doctor who treats cancer with medicines such as chemotherapy or targeted therapy.

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

It is important to discuss all of your treatment options, including goals and 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 nasopharyngeal cancer?" for some questions to ask.)

If time permits, it is often a good idea to get a second opinion. A second opinion can provide you with more information and help you feel confident about your chosen treatment plan.

The next few sections describe the various types of treatments used for nasopharyngeal cancers. This is followed by a description of the most common approaches used for these cancers, based on their stage.

Surgery for nasopharyngeal cancer

Because the nasopharynx is a hard place to operate on and because other types of treatment are often effective, surgery is seldom the main treatment for patients with nasopharyngeal cancer (NPC). When it is used, it is most often to remove lymph nodes in the neck that haven't responded to other treatments.

Removing the tumor

With newer endoscopic surgery techniques, doctors can use flexible fiber-optic scopes and long, thin surgical instruments to completely remove some nasopharyngeal tumors. But this is appropriate only for a small number of patients. These complex procedures are done only in specialized centers.

Surgery does have some advantages over other treatments such as radiation therapy – for example, it lets doctors look at the cancer (and nearby tissues) closely in the lab to make sure it has all been removed.

Removing lymph nodes

Cancers of the nasopharynx often spread to the lymph nodes in the neck. These cancers often respond well to treatment with radiation therapy (and sometimes chemotherapy). But if some cancer remains after these treatments, an operation called a *neck dissection* may be needed to remove these lymph nodes.

There are several types of neck dissection surgery. They differ in the amount of tissue removed from the neck.

- A *partial* or *selective neck dissection* removes only lymph nodes that are closest to the primary tumor site and most likely to have cancer spread.
- A *modified radical neck dissection* removes lymph nodes on one side of the neck between the jaw bone and collarbone, as well as some muscle and nerve tissue. The main nerve to the shoulder muscle is usually preserved.
- A *radical neck dissection* removes nearly all lymph nodes on one side as well as even more muscles, nerves, and veins.

Possible risks and side effects of surgery

The risks and side effects of any surgery depend on the extent of the operation and a person's general health before the surgery. If you are considering surgery, your doctor will discuss the likely side effects with you beforehand.

All surgeries carry some risk, including the possibility of bleeding, infections, complications from anesthesia, and pneumonia. Most people will have some pain for a while after the operation, although this can usually be controlled with medicines. Other possible side effects of surgery in the head and neck area can include problems with speech or swallowing.

The most common side effects of any neck dissection are numbness of the ear, weakness when raising the arm above the head, and weakness of the lower lip. These side effects are caused by the effects of the operation on certain nerves that supply these areas. After a selective neck dissection, the weakness of the shoulder and lower lip usually go away after a few months. But if either of the nerves that supply these areas is removed as part of a radical neck dissection or because of involvement with tumor, the weakness will be permanent.

After more extensive neck dissections, physical therapists can teach you exercises to improve neck and shoulder strength and mobility.

Radiation therapy for nasopharyngeal cancer

Radiation therapy uses high-energy x-rays or particles to destroy cancer cells or slow their rate of growth. It is usually at least part of the main treatment for nasopharyngeal cancer (NPC) because most of these cancers are very sensitive to radiation.

For many cases of NPC, chemotherapy is given with radiation to try to increase its effects. This treatment, known as *chemoradiation*, can be better than radiation alone at fighting the cancer, but it also tends to have more side effects. (This is discussed more in the section, “Chemotherapy for nasopharyngeal cancer.”)

Radiation therapy is usually given both to the main nasopharyngeal tumor and to nearby lymph nodes in the neck. Even if the lymph nodes are not abnormally firm or large, radiation is still used in case a few cancer cells have spread there. If the lymph nodes are known to have cancer cells, higher radiation doses are used.

Different types of radiation therapy can be used to treat NPC:

External beam radiation therapy (EBRT)

This type of radiation therapy uses x-rays from a machine outside the patient’s body to kill cancer cells. It is the most common form of radiation therapy for NPC.

Before your treatments start, the radiation team will take careful measurements to determine the correct angles for aiming the radiation beams and the proper dose of radiation. Radiation

therapy is much like getting an x-ray, but the radiation is stronger. The procedure itself is painless. Each treatment lasts only a few minutes, although the setup time – getting you in place for treatment – takes longer. Most often, radiation treatments are given 5 days a week for about 7 weeks.

Although standard (conventional) EBRT is helpful in treating NPC, it often has more side effects than newer techniques that focus the radiation better and lower the radiation exposure to nearby healthy tissues. The following newer techniques are the preferred way to give radiation to treat NPC:

Three-dimensional conformal radiation therapy (3D-CRT) uses the results of imaging tests such as MRI and special computers to precisely map the location of the tumor(s). Radiation beams are shaped and aimed at the tumor(s) from several directions, which makes it less likely to damage nearby normal tissues.

Intensity-modulated radiation therapy (IMRT) is an advanced form of 3D therapy. It uses a computer-driven machine that moves around the patient as it delivers radiation. Along with shaping the beams and aiming them at the tumor from several angles, the intensity (strength) of the beams can be adjusted to limit the dose reaching the most sensitive normal tissues.

Stereotactic radiosurgery is a type of radiation treatment that delivers a large, precise radiation dose to the tumor area in a single session. (There is no actual surgery involved in this treatment.) The radiation may be delivered in 2 ways.

In one approach, radiation beams are focused at the tumor from hundreds of different angles for a short period of time. One machine used to deliver this type of radiation is known as a *Gamma Knife*.

Another approach uses a movable linear accelerator (a machine that creates radiation) that is controlled by a computer. Instead of delivering many beams at once, this machine moves around the head to deliver radiation to the tumor from different angles. Several machines, with names such as *X-Knife*, *CyberKnife*, and *Clinac*, do stereotactic radiosurgery in this way.

Brachytherapy (internal radiation)

Another way to deliver radiation is to insert (implant) very thin metal rods or wires containing radioactive materials into or very near the cancer. The radiation travels a very short distance, so it affects the cancer without causing much harm to nearby healthy body tissues.

The implant is usually left in place for several days while the patient stays in a private hospital room. The length of time that visitors, nurses, and other caregivers can spend with the patient may be limited because of potential radiation exposure, but this depends on the type of radiation. The implant is removed before the patient goes home.

Brachytherapy may be used if the cancer comes back after external beam radiation therapy (although stereotactic radiosurgery may be used instead, as it is less invasive). Sometimes, internal and external beam radiation therapy are used together.

Possible side effects of radiation therapy

Common side effects of external beam to the head and neck include:

- Skin changes in the area where the radiation passes through, with redness or blistering
- Nausea and vomiting
- Fatigue (tiredness)
- Sore throat
- Sores in the mouth and throat which can lead to trouble swallowing and weight loss from not eating
- Hoarseness
- Loss of taste

These side effects get better once radiation has stopped. Other side effects may not get better over time, such as:

- Problems with hearing or vision because of damage to certain nerves
- Damage to bones in the skull
- Tooth problems
- Damage to the salivary glands

Radiation can make any tooth problems that you already have get worse and hard to fix. Most doctors advise that you have your teeth checked by a dentist before you have radiation therapy to the head or neck area. In some cases, the dentist may even advise removing some teeth before treatment to lessen the chance you will have problems later.

A major concern with radiation therapy for NPC is that it can damage the salivary glands, which can result in dry mouth that does not go away. This can cause problems with eating and swallowing and can lead to severe tooth decay. People treated with radiation to the head or neck area need to practice careful oral hygiene to help prevent this problem.

Newer radiotherapy techniques such as IMRT may also help reduce this side effect. Some of the damage to the salivary glands may also be lessened if a drug called amifostine (Ethyol®) is given before each radiation treatment.

The thyroid gland is often damaged if the neck area is treated with external beam radiation. The damage doesn't cause problems seen right way, so your doctor will watch your thyroid function with blood tests in the years after treatment. If your thyroid function goes down, pills to replace thyroid hormone may be needed.

The pituitary gland, which is responsible for controlling many hormones in the body, can be damaged by treatment of the tumor in the nasopharynx. This can also be found with blood tests. If the damage is serious enough, this might require taking certain hormones to replace the ones that are missing.

The carotid arteries, which are major blood vessels in the neck that deliver blood to the brain, can sometimes become narrowed after radiation, which could raise a person's risk of stroke or other problems. This usually takes several years to occur.

It is important to discuss the possible side effects of radiation therapy with your doctor before starting treatment, and to make sure everything is being done to try to limit these side effects as much as possible.

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

Chemotherapy for nasopharyngeal cancer

Chemotherapy (chemo) is the use of anti-cancer drugs to treat cancer. These drugs are most often given into a vein (IV) or by mouth. They enter the bloodstream and reach throughout the body, making this treatment useful for cancers that have spread beyond the head and neck.

Chemo may be used in different situations to treat nasopharyngeal cancer (NPC):

- Chemo is often used together with radiation therapy as the first treatment for more advanced stages of NPC because some chemo drugs make cancer cells more sensitive to radiation. This treatment is called *chemoradiation*.
- Chemo may also be given after radiation (or chemoradiation). This is known as *adjuvant treatment*.
- Chemo is used for patients whose NPC has spread to distant organs such as the lungs, bones, or liver. It may be used either alone or along with radiation.

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

Cisplatin is the chemo drug used most often to treat NPC. It is used alone as part of chemoradiation, but may be combined with another drug, 5-fluorouracil (5-FU) if given after radiation.

Some other drugs may also be helpful in treating cancer that has spread. These include:

- Carboplatin (Paraplatin®)
- Doxorubicin (Adriamycin®)
- Epirubicin (Ellence®)
- Paclitaxel (Taxol®)
- Docetaxel (Taxotere®)
- Gemcitabine (Gemzar®)
- Bleomycin
- Methotrexate

Often, combinations of 2 or more of these drugs are used.

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, 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 certain side effects.

The side effects of chemo depend on the type and dose of drugs you are given and how long they are taken. Common side effects include:

- Hair loss
- Mouth sores
- Loss of appetite
- Nausea and vomiting
- Diarrhea
- Increased chance of infections (due to low white blood cell counts)
- Easy bruising or bleeding (due to low blood platelet counts)
- Fatigue (due to low red blood cell counts)

These side effects are usually short-term and go away after treatment is finished. Once chemo is started, let your health care team know if you have side effects, so they can be treated. There are ways to prevent or treat many of the side effects of chemo. For example, many good drugs are available to help prevent or treat nausea and vomiting.

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

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 side effects from getting worse.

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

Targeted therapy for nasopharyngeal cancer

As researchers have learned more about the changes in cells that cause cancer, they have been able develop newer drugs that specifically target these changes. These targeted drugs work differently from standard chemotherapy (chemo) drugs. They may work in some instances when chemo drugs do not, or they may sometimes help chemo drugs work better. Targeted drugs also often have different (and less severe) side effects.

Cetuximab (Erbitux®)

Cetuximab is a monoclonal antibody (a man-made version of an immune system protein) that targets the epidermal growth factor receptor (EGFR). EGFR is a protein found on the surface of cells. It normally receives signals telling the cells to grow and divide. Nasopharyngeal cancer (NPC) cells sometimes have more than normal amounts of EGFR, which can help them grow faster. By blocking EGFR, cetuximab may slow or stop this growth.

The exact role of cetuximab in treating NPC is still being studied. It is most often used along with chemo and/or radiation in cases where the cancer has come back or continues to grow after initial chemo.

Cetuximab is given by IV infusion, usually once a week. A rare but serious side effect of cetuximab is an allergic reaction during the first infusion, which could cause breathing problems and low blood pressure. You may be given medicine before treatment to help prevent this. Many people develop skin problems such as an acne-like rash on the face and chest during treatment, which in some cases can lead to infections. Other side effects may include headache, tiredness, fever, and diarrhea.

More information about monoclonal antibodies can be found in our document *Immunotherapy*. For general information about targeted therapy, see our document *Targeted Therapy*.

You can also call us to learn more about individual drugs, or see our online Guide to Cancer Drugs.

Clinical trials for nasopharyngeal cancer

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.

If you would like to take part in a clinical trial, you should 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. 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.

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

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.

You can get a lot more information on clinical trials in our document called *Clinical Trials: What You Need to Know*. You can read it on our website or call our toll-free number (1-800-227-2345) and have it sent to you.

Complementary and alternative therapies for nasopharyngeal cancer

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 not to be helpful, and a few have even been found 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 read about them 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 options by stage of nasopharyngeal cancer

The treatment options your cancer care team will recommend depend on the extent of the cancer at its site of origin and how far the cancer has spread, if at all. Nasopharyngeal cancer (NPC) in children is treated largely the same way as NPC in adults.

Stages 0 and I

The usual treatment for these early stage cancers is radiation therapy aimed at the nasopharyngeal tumor.

Although the cancer has not yet spread to lymph nodes in these stages, the nearby lymph nodes in the neck are usually treated with radiation therapy as well. This is considered preventive (prophylactic) radiation. Some patients may have cancer cells in these lymph nodes that can't be detected. Although there are too few cancer cells in the lymph nodes to cause them to be enlarged, these cells could continue to grow and spread if not destroyed by radiation therapy.

Stages II, III, IVA and IVB

These cancers have spread outside of the nasopharynx and often to lymph nodes in the neck or above the collarbone. Patients with these stages of NPC usually receive chemoradiation (chemotherapy given along with radiation therapy to the nasopharynx and neck lymph nodes). The chemotherapy (chemo) drug most often used is cisplatin, but sometimes another drug is used as well. This is usually followed by more chemo, most often with cisplatin plus 5-FU. Most studies have found that chemoradiation helps patients live longer than just radiation therapy alone. But adding chemo leads to more side effects, which can affect quality of life. It's important to understand what the side effects are likely to be before starting this treatment.

If cancer is still in the lymph nodes after this treatment, surgery (neck dissection) may be done to remove the lymph nodes.

Stage IVC

These cancers have spread to distant parts of the body and can be hard to treat. The usual treatment is chemo, often with cisplatin and one other drug. If there is no sign of the cancer after chemotherapy, radiation therapy to the nasopharynx and the lymph nodes in the neck or chemoradiation is given to try to kill any remaining cancer cells. Another option in some cases is to give chemoradiation as the first treatment.

If there are still signs of cancer after the initial chemotherapy, another chemotherapy regimen using different drugs may be tried. Chemotherapy plus the targeted drug cetuximab (Erbitux) may be another option.

Because these cancers can be hard to treat effectively, taking part in a clinical trial of newer treatments may be a good option.

Recurrent nasopharyngeal cancer

Cancer is called *recurrent* when it come backs after treatment. Recurrence can be local (in or near the same place it started) or distant (spread to organs such as the lungs or bone). If your cancer returns after treatment, the choices available to you depend on the location and extent of the cancer, which treatments were used the first time around, and your overall health. It is important to understand the goal of any further treatment – whether it is to try to cure the cancer, to slow its growth, or to help relieve symptoms – as well as the likelihood of benefits and risks.

Some tumors that recur in the nasopharynx can be removed by surgery using an approach through the nose (called *endoscopic skull base surgery*). This is a specialized surgery that should only be done by a surgeon with a great deal of experience in this procedure, so it's not available at all medical centers.

Recurrent NPC in regional (neck area) lymph nodes can sometimes be treated by additional radiation therapy. But if doctors believe that more radiation would cause serious side effects or if the initial response to radiation was incomplete, surgery (neck dissection) may be used instead.

Cancer that recurs in distant sites is usually treated with chemotherapy. If chemotherapy has been given already, different chemo drugs may be tried. The targeted drug cetuximab may be given along with chemo, generally on a clinical trial.

New drug treatments being tested in clinical trials and new surgical procedures may help some patients with recurrent NPC, as well as improve knowledge that can help others with NPC in the future.

If the cancer can't be cured, further treatments may be aimed at slowing its growth or relieving symptoms caused by the distant spread of the cancer. For example, if the cancer has spread to the spine, radiation therapy may be given to the area to relieve pain and reduce the chances of further complications. Even if a cure is not possible, it is important to remember that there are many options to relieve symptoms of advanced cancer.

More treatment information for nasopharyngeal cancer

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

The NCI provides treatment information via telephone (1-800-4-CANCER) and its website (www.cancer.gov). Information for patients as well as more detailed information intended for use by cancer care professionals is also available on www.cancer.gov.

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). (Information on nasopharyngeal cancers is found in the NCCN Head and Neck Cancers guideline.)

What should you ask your doctor about nasopharyngeal cancer?

As you deal with your cancer and the process of treatment, you need to have honest, open discussions with your cancer care team. You should feel free to ask any question, no matter how minor it might seem. Among the questions you might want to ask are:

- What kind of nasopharyngeal cancer do I have? Does this affect my options?
- Has my cancer spread beyond the nasopharynx?
- What is the stage of my cancer? What does the stage 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?
- What treatment choices do I have?
- What do you recommend and why?
- What is the goal of the treatment?
- What are the chances my cancer can be cured with treatment?
- 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 involve? Where will it be done?
- How will treatment affect my daily activities?
- What risks and side effects can I expect? How long are they likely to last?
- What are the chances that my cancer will recur?

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

Along with these sample questions, you might want to write down some of your own. For instance, you might want more information about recovery times so you can plan your work or activity schedule. Or you may want to ask about second opinions or about clinical trials for which you may qualify. Keep in mind, too, that doctors are not the only ones who can give you information. Other health care professionals, such as nurses and social workers, may have the answers you seek.

What happens after treatment for nasopharyngeal cancer?

For many people with nasopharyngeal cancer (NPC), 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 questions about any problems you may have and may do exams and lab tests or imaging tests (such as MRI or CT scans) to look for signs of cancer or treatment side effects. Your health care team will discuss which tests should be done and how often based on the stage of your cancer and the type of treatment you received.

Most doctors recommend follow-up exams at least every few months for the first 2 years after treatment, then less often after this. If you had radiation therapy to the neck, your doctor will check your thyroid function with blood tests once or twice a year.

You may be advised to see your dentist after treatment to check on the health of your teeth. Your doctor will also want to keep a close eye on your hearing, speech, and swallowing, which can be affected by treatment. If you are having problems with any of these, your doctor may refer you to a therapist for help with rehabilitation.

Imaging tests such as CT or PET/CT scans may be done after treatment to get an idea of what the nasopharynx and neck area now look like. Further imaging tests may be done if you later develop any signs or symptoms that might be caused by a return of the cancer.

Almost any cancer treatment can have side effects. Some may last for a few weeks to months, but others can last the rest of your life. This is the time for you to talk to your cancer care team about any changes or problems you notice and any questions or concerns you have.

It is very important to report any new symptoms to the doctor right away, because they may prompt your doctor to do tests that could help find recurrent cancer as early as possible, when the chance of successful treatment is greatest.

If cancer does recur, further treatment will depend on the 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 options by stage of nasopharyngeal cancer." 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 is 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.

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 anything about your medical history. It is important that you be able to give your new doctor the details of your diagnosis and treatment. 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 targeted therapy, a list of your drugs, drug doses, and when you took them
- Copies of any imaging tests, such as CT scans and MRIs, which can often be placed on a DVD

The doctor may want copies of this information for his records, but always keep copies for yourself.

Lifestyle changes after having nasopharyngeal cancer

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 the 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. 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. This is especially true for cancers of the head and neck, such as nasopharyngeal cancer. The cancer or its treatment may affect how you swallow or cause dry mouth, changes in taste, or other problems. Nausea can be a problem from some treatments. You may not feel like eating and lose weight when you don't want to.

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.

For more information, see 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 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 exercised 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 exercise 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 fatigue and other treatment side effects, please see the “Physical Side Effects” section of our website or “Additional resources for nasopharyngeal cancer” to get a list of available information.)

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 can help 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 cancer progressing or coming back?

Most people want to know if there are specific lifestyle changes they can make to reduce their risk of their cancer progressing or coming back. For many 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 nasopharyngeal cancer to say for sure if there are things you can do that will be helpful. Tobacco and alcohol use have clearly been linked to most types of cancers of the head and neck, but the link with nasopharyngeal cancers is not as clear. We don't know for certain if not smoking or drinking can reduce your risk, but we do know that it can help improve your appetite and overall health. It can also reduce the chance of developing other types of cancer. If you want to quit smoking and need help, call the American Cancer Society at 1-800-227-2345.

Adopting other healthy behaviors such as eating well, getting regular physical activity, and maintaining a healthy weight may help as well, 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 cancer.

How does having nasopharyngeal cancer 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, as you feel better and have fewer doctor visits, you will see your health care team less often 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 treatment for nasopharyngeal cancer stops working

If cancer keeps growing or comes back after one kind of treatment, it is possible that another treatment plan might still cure the cancer, or at least shrink it 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, the cancer tends to become resistant to all treatment. If this happens, it's important to weigh the possible limited benefits of a new treatment against the possible downsides. 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 medical treatments and nothing's working anymore. Your doctor may 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. In many cases, your doctor can estimate how likely it is the cancer will respond to treatment you are considering. For instance, the doctor may 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 think about and understand your reasons for choosing this plan.

No matter what you decide to do, you need to 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 care.

Palliative care helps relieve symptoms, but is 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 purpose 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 bone pain caused by cancer that has spread to the bones. Or chemo 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. 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 Nearing the End of Life and Advance Directives..

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 the things you no longer want to do. Though the cancer may be beyond your control, there are still choices you can make.

What's new in nasopharyngeal cancer research and treatment?

Research into the causes, prevention, and treatment of nasopharyngeal cancer (NPC) is being done in many university hospitals, medical centers, and other institutions around the world.

Causes, prevention, and early detection

Many studies are looking at how Epstein-Barr virus (EBV) infection and other risk factors cause cells of the nasopharynx to become cancerous. Researchers hope these studies may eventually lead to vaccines to help prevent some cases of NPC by avoiding EBV infection.

Recent discoveries about EBV, its interaction with nasopharyngeal cells, and the immune system's reaction to EBV have led to new blood tests that may help detect NPC early and better predict the response to treatment. These tests are now being studied in areas of the world where this cancer is common.

Treatment

New surgical techniques

Advances in the field of skull base surgery such as the use of endoscopes in the nose now allow doctors to remove some tumors from hard to reach areas like the nasopharynx. This type of surgery requires a specialized team that has expertise in this field. It may offer hope for some patients with recurrent NPC and patients with the keratinizing type of NPC, which often doesn't respond to radiation therapy.

New radiation therapy techniques

Most types of radiation therapy use radiation in the form of x-rays. Another type of radiation uses protons to kill cancer cells, instead. Unlike x-rays, which release energy both before and after they hit their target, protons cause little damage to tissues they pass through and then release their energy after traveling a certain distance. This means that proton beam radiation may be able to deliver more radiation to the tumor and do less damage to nearby normal tissues. Although this approach is promising in theory, it hasn't been proven to be better than x-ray techniques like IMRT. Also, the machines needed for proton therapy are very expensive, and so this treatment is not widely available.

Doctors are also studying the best schedule for giving radiation therapy. External beam radiation treatments are usually given once a day, 5 days a week, for many weeks in a row. Studies are now under way to see if schedules that either give the doses over fewer days or give smaller doses twice a day might be more effective.

Chemotherapy

Researchers continue to develop new chemotherapy drugs, new drug combinations, and new ways to give drugs that might be more effective against advanced NPC. Several drugs that are already used to treat other cancers, such as capecitabine, oxaliplatin, and gemcitabine, have been studied for use against NPC as well.

Clinical trials are also testing ways to best combine chemotherapy with radiation therapy. For example, studies are comparing the effectiveness of chemotherapy given before, during, or after radiation therapy.

Targeted therapy

Drugs that target specific parts of cancer cells may prove to be useful against NPC and have fewer side effects than standard chemotherapy drugs.

The drug cetuximab (Erbitux), which targets the EGFR protein, is already being used in some cases of NPC that recur or keep growing after treatment with chemotherapy. Other drugs that target EGFR are also being studied for use against NPC, including nimotuzumab and icotinib.

Other newer drugs target a tumor's ability to develop new blood vessels, which they need in order to grow larger. These drugs are called angiogenesis inhibitors. Several of these drugs are now being tested for use against NPC, including bevacizumab (Avastin®), sorafenib (Nexavar®), and pazopanib (Votrient®).

Immunotherapy

NPC seems to be caused at least in part by infection with the Epstein-Barr virus (EBV). Although patients' immune systems can be shown to have reacted against EBV, this doesn't seem to be enough to kill the cancer. Researchers are trying to use different ways to boost the immune system or help it better target EBV-infected cells.

One way to do this is to remove T lymphocytes (immune system cells) from the blood of patients with NPC and alter them in the lab to increase their numbers and their power to kill EBV. The cells are then injected back into the patients. Early results with small numbers of patients have been promising, and larger studies of this technique are now under way.

Gene therapy

Scientists have recently discovered how certain gene mutations (changes) in nasopharyngeal cells may cause them to become cancerous. A clinical trial using a virus to replace the damaged tumor suppressor gene p53 in the cancer cells had some promising results. This approach is still being studied.

Additional resources for nasopharyngeal cancer

More information from your American Cancer Society

Here is more information you might find helpful. You also can order free copies of our documents from our toll-free number, 1-800-227-2345, or read them on our website, www.cancer.org.

Living with cancer

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

Distress in People With Cancer

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

Pain Control: A Guide for Those With Cancer and Their Loved Ones (also available in Spanish)

Sexuality for the Woman with Cancer (also available in Spanish)

Sexuality for the Man with Cancer (also available in Spanish)

Living with Uncertainty: The Fear of Cancer Recurrence

When Your Cancer Comes Back: Cancer Recurrence

Understanding cancer treatments

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

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

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

Cancer treatment side effects

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 available in Spanish)

Work, insurance, and finances

Health Insurance and Financial Assistance for the Cancer Patient

Returning to Work After Cancer Treatment

Working During Cancer Treatment

Your American Cancer Society also has books that you might find helpful. Call us at 1-800-227-2345 or visit our bookstore online to find out about costs or to place an order.

National organizations and Web sites*

In addition to the American Cancer Society, other sources of patient information and support include:

Support for People with Oral and Head and Neck Cancer (SPOHNC)
Toll-free number: 1-800-377-0928
Website: www.spohnc.org

> Offers information, resources, and some local support groups. For a fee, you can join the organization and get their newsletter (older newsletters are free on their website)

National Coalition for Cancer Survivorship
Toll-free number: 1-888-650-9127
1-877-NCCS-YES (1-877-622-7937) for some publications and Cancer Survivor Toolbox® orders
Website: www.canceradvocacy.org

> Offers information on work, health insurance, and more. The Cancer Survival Toolbox is a free, self-learning audio program to help cancer survivors and caregivers develop practical tools needed to deal with the diagnosis, treatment and challenges of cancer. Listen online or order CDs. Also in Spanish and Chinese

National Cancer Institute
Toll-free number: 1-800-4-CANCER (1-800-422-6237) TYY: 1-800-332-8615
Website: www.cancer.gov

> Provides information on all types of cancer, living with cancer, support information for families of people with cancer, research, and more

**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.

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1-800-227-2345 or www.cancer.org