



ADRENAL CORTICAL CANCER

What is cancer?

Cancer develops when cells in a part of the body begin to grow out of control. Although there are many kinds of cancer, they all start because of out-of-control growth of abnormal cells.

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

Because cancer cells continue to grow and divide, they are different from normal cells. Instead of dying, they outlive normal cells and continue to form new abnormal cells.

Cancer cells develop because of damage to DNA. This substance is in every cell and directs all its activities. Most of the time when DNA becomes damaged the body is able to repair it. In cancer cells, the damaged DNA is not repaired. People can inherit damaged DNA, which accounts for inherited cancers. Many times though, a person's DNA becomes damaged by exposure to something in the environment, like smoking.

Cancer usually forms as a tumor. Some cancers, like leukemia, do not form tumors. Instead, these cancer cells involve the blood and blood-forming organs and circulate through other tissues where they grow.

Often, cancer cells travel to other parts of the body, where they begin to grow and replace normal tissue. This process is called metastasis. Regardless of where a cancer may spread, however, it is always named for the place it began. For instance, breast cancer that spreads to the liver is still called breast cancer, not liver cancer.

Not all tumors are cancerous. Benign (non-cancerous) tumors do not spread (metastasize) to other parts of the body and, with very rare exceptions, are not life threatening.

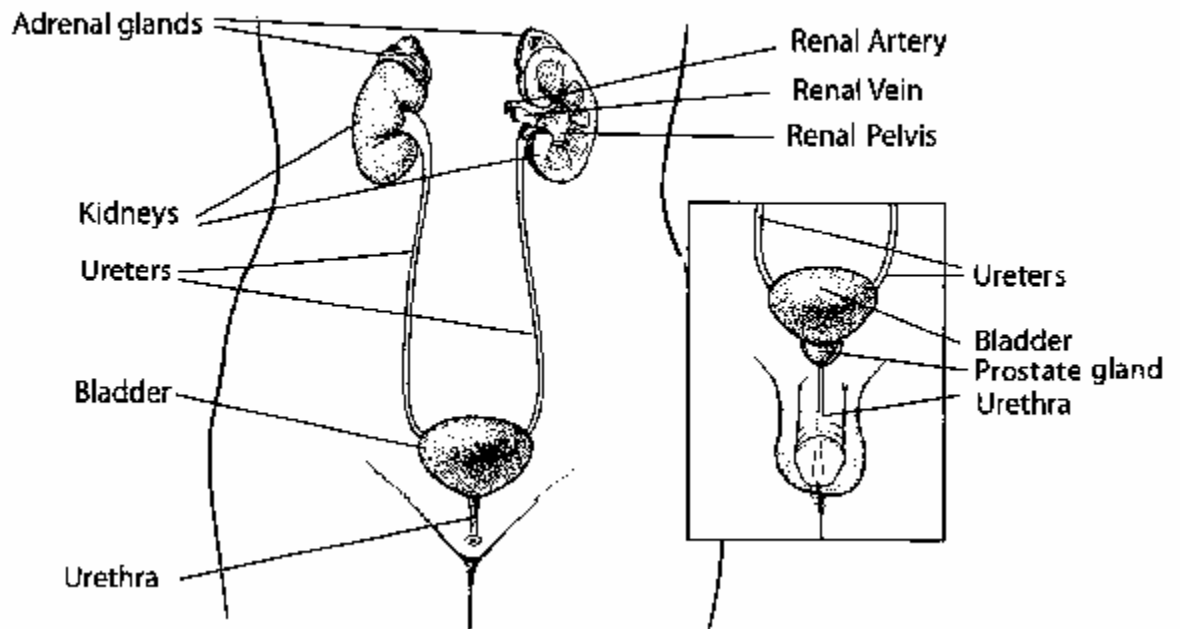
Different types of cancer can behave very differently. For example, lung cancer and breast cancer are very different diseases. They grow at different rates and respond to different treatments. That is why people with cancer need treatment that is aimed at their particular kind of cancer.

Cancer is the second leading cause of death in the United States. Nearly half of all men and a little over one third of all women in the United States will develop cancer during their lifetimes. Today, millions of people are living with cancer or have had cancer. The risk of developing most types of cancer can be reduced by changes in a person's lifestyle, for example, by quitting smoking and eating a better diet. The sooner a cancer is found and treatment begins, the better are the chances for living for many years.

What is adrenal cortical cancer?

About the adrenal glands

The adrenals are small glands that sit above each of the kidneys. The kidneys are located deep inside the upper part of the abdomen.



The adrenal gland is made up of 2 parts. The outer part, called the cortex, is where the adrenal cortical tumors develop. The function of the cortex is to produce certain hormones

for the body. These hormones all possess a similar chemical structure and are called steroids. They include:

- *cortisol* -- causes changes in metabolism that help the body to handle stress.
- *aldosterone* -- helps the kidneys regulate the amount of salt in the blood and tissues of the body.
- *androgens and estrogens* -- In men, the testicles produce most of the androgens (male hormones). The ovaries produce most of the estrogens (female hormones) in women. However, the adrenal glands produce small amounts of both androgens and estrogens in men and women.

The inner portion of the adrenal gland, called the medulla, is really an extension of the nervous system. Nervous system hormones such as *norepinephrine* and *epinephrine* (also called adrenaline) are made in the medulla. Tumors of the adrenal medulla, such as *pheochromocytomas* and *neuroblastomas*, are not discussed in this document. (For more information on neuroblastomas, see our document, *Neuroblastoma*.)

Adrenal cortex tumors

There are 2 main types of adrenal cortex tumors: benign (non-cancerous) and malignant (cancers). Most of these tumors are benign and are called adenomas. Cancers of the adrenal cortex are rare. These 2 types of tumors can be hard to tell apart when the cells are looked at under the microscope. There are certain features to look for, but the only way to know for sure that the tumor is a cancer is when it has spread. If it has spread to lymph nodes or other organs and tissues, it is a cancer. Adenomas do not spread outside the adrenal gland.

Adrenal cortex adenomas

Most tumors of the adrenal cortex are benign adenomas. These are small tumors, usually less than 5 centimeters (about 2 inches) in diameter. They usually occur in only 1 adrenal gland, but sometimes affect both glands.

Most people with adrenal adenomas have no symptoms and are unaware that they have an adrenal tumor. Some of these adenomas are discovered "incidentally" (by accident) when CT or MRI scans of the abdomen are done because of unrelated health problems. About 5% of people who have a CT scan of the abdomen are found to have an adrenal tumor that was not suspected. Most of these are nonfunctional -- meaning that they don't make adrenal hormones. Sometimes these tumors are known by the nickname "incidentalomas" because they aren't causing problems and were only found by accident.

Some adenomas produce too much adrenal steroid hormone. Sometimes the excess hormone can cause symptoms. Many of the hormone-related symptoms of adenomas are the same as those from adrenal carcinomas (cancers). These symptoms are discussed in the section, "Can

adrenal cortical cancer be found early?" Adenomas are much more likely than carcinomas to produce high levels of aldosterone, which can cause high blood pressure.

Treatment: Adenomas can be cured by removal of the adrenal gland that contains the adenoma. Some adrenal adenomas that cause hormone-related symptoms can be treated effectively with medications that block the production or actions of these hormones. This may be the best treatment choice for patients with other serious medical problems who might not be able to withstand a major operation.

The treatment of incidentalomas depends on the chance that it may be a cancer and whether or not it is raising hormone levels. When an adrenal tumor is found accidentally, tests are often done to see if it is making hormones. If it is, surgery is often recommended. Otherwise, surgery may only be recommended if it is likely to be a cancer. Tumors that are small are less likely to be cancer, and are often watched without treatment. The CT (or MRI) scan is repeated at least 6 months later to see if the tumor has grown. If it has, it may need to be removed. If it hasn't grown, hormone levels will be watched over the next few years. If the tumor remains small and doesn't make any hormones, it may not need to be treated at all.

The remainder of this document refers to adrenal cancers only, and not to adenomas.

Adrenal cortical cancer

The type of cancer that develops in the cortex of the adrenal gland is called adrenal cortical carcinoma. It is discovered for 1 of 2 reasons. The first is that it produces hormones that cause body changes such as weight gain and fluid retention, early puberty in children, or excess facial or body hair growth in women.

They can also be discovered when they start causing symptoms because they have gotten very large. Large tumors can press on other organs in the abdomen, causing pain, a feeling of fullness that causes people to eat less, and weight loss. Generally, adrenal cortical cancers are much larger than adrenal adenomas. An adrenal tumor larger than 5 or 6 centimeters (about 2 to 2 1/2 inches) is assumed to be a cancer. In one study, the average size of an adrenal cancer was about 13 cm (or 5 inches).

Most cancers found in the adrenal gland did not start there and are not adrenal cortical cancers. Instead, they start in other organs or tissues and then spread (metastasize) through the bloodstream to the adrenal glands. For example, lung cancers and breast cancers often spread to the adrenals. Even when other cancers spread to the adrenals; however, they are still named after the place they started and are treated like other cancers that start in the same place. They are not considered to be adrenal cancer. Their treatment is described in our documents on these cancers.

What are the key statistics about adrenal cortical cancer?

Adrenal cortical carcinomas are very rare and the real number diagnosed in the United States is not known. It is probably around 300 per year. They are much less common than adrenal adenomas, which are frequently found among middle aged and elderly people.

The average age of patients is around 45 to 50, but adrenal cortical cancer can occur at any age; even in children. It also seems to occur more often in females.

What are the risk factors for adrenal cortical cancer?

A risk factor is anything that changes a person's chance of getting a disease such as cancer. Different cancers have different risk factors. For example, unprotected exposure to strong sunlight is a risk factor for skin cancer and smoking is a risk factor for cancers of the lungs, mouth, larynx, esophagus, bladder, and several other organs. Scientists have found few risk factors that make a person more likely to develop adrenal gland tumors. Even if a patient does have one or more risk factors for adrenal gland tumors, it is impossible to know for sure how much that risk factor contributed to causing the cancer.

Genetic syndromes

The vast majority of adrenal cortex cancers are sporadic (not related to heredity), but some, particularly in children, are caused by a genetic defect.

Li-Fraumeni syndrome: The Li-Fraumeni syndrome is a very rare condition in which a genetic defect in a gene called p53 leads to a greatly increased risk of developing several types of cancers. These include breast cancer, bone cancers, brain cancer, and adrenal cortex cancer.

Beckwith-Wiedemann syndrome: People with this problem have large tongues, are large themselves, and can develop cancers of the kidney, liver, and adrenal cortex.

Multiple endocrine neoplasia (MEN-1): People with MEN-1 have a very high risk of developing tumors of three glands -- the pituitary, parathyroid, and pancreas. About one third of people with this condition also develop adrenal cortical adenomas, which tend to be small and usually do not cause any symptoms. This syndrome is caused by defects in a gene called MEN1. People who have a family history of MEN-1 or pituitary, parathyroid, pancreas, or adrenal cancers should ask their doctor whether they might benefit from genetic counseling.

Familial adenomatous polyposis (FAP): People with this syndrome develop hundreds of polyps in the large intestine. These polyps will lead to colon cancer if the colon is not removed. FAP also increases the risk of other cancers, including adrenal cortical cancer.

Lifestyle and environmental factors

Risk factors such as a high-fat diet, smoking, sedentary lifestyle, and exposure to cancer-causing substances in the environment have a great impact on a person's risk of developing many types of cancer. Although none of these factors has been definitely found to influence a person's risk of developing adrenal cortical cancer, smoking has been suggested as a risk factor by some researchers.

Do we know what causes adrenal cortical cancer?

Scientists do not know exactly what causes most adrenal cortical tumors. Over the past few years, they have made great progress in understanding how certain changes in a person's DNA can cause cells in the adrenal to become cancerous. DNA is the molecule that carries the instructions for nearly everything our cells do. We usually look like our parents because they are the source of our DNA. However, DNA affects more than our outward appearance. It also determines our risk for developing certain diseases, including some types of cancer.

Some *genes* (parts of our DNA) contain instructions for controlling when our cells grow and divide. Some genes that promote cell division are called *oncogenes*. Other genes that slow down cancer cell division or cause them to die are called tumor suppressor genes. We know that cancers can be caused by DNA mutations (changes) that turn on oncogenes or turn off tumor suppressor genes. Some people with cancer have DNA mutations they inherited from a parent, which increase their risk for developing the disease. But most DNA mutations that are seen in cancers happen during life rather than having been inherited. These mutations may result from exposure to radiation or carcinogens (cancer-causing chemicals). Most of these mutations, however, happen for no apparent reason.

The DNA mutations that cause tumors in people with the genetic syndromes discussed in the previous section have been identified. Defects in the gene that causes MEN-1 are responsible for most hereditary adrenal tumors.

The Li-Fraumeni syndrome is caused by inherited mutations that inactivate the p53 tumor suppressor gene. Only a small minority of adrenal cortical cancers are due to this syndrome. Many other adrenal cortical cancers have also been found to have abnormal p53 genes that were acquired after birth (not inherited).

Can adrenal cortical cancer be prevented?

Since people with adrenal cortical cancers have no known preventable risk factors, it is not now possible to prevent this disease, specifically. Not smoking is a way to lower the risk for many cancers, and perhaps even adrenal cortical cancer.

Can adrenal cortical cancer be found early?

It is hard to find adrenal cortical carcinomas early and they are often quite large when diagnosed. Adrenal cortical carcinomas are often found earlier in children than in adults because children are more sensitive to the hormones these tumors secrete. Children will show outward signs of excess hormone production early. In adults, these tumors may be found early by accident, when a CT scan is done for some other health concern.

The American Cancer Society has official recommendations for the early detection of several types of cancer. Because adrenal cancers occur so rarely, the Society does not recommend routine testing for this cancer in people without any symptoms.

How is adrenal cortical cancer diagnosed?

Medical history and physical exam

The first step is to take your complete medical history to check for any symptoms. Your doctor will want to know if anyone in your family has had adrenal cancer. Your doctor will also ask you about your menstrual or sexual function and about any other symptoms that you may be having. A physical exam will provide other information about signs of adrenal gland cancer and other health problems. Your abdomen will be thoroughly examined by your doctor for evidence of a tumor (or mass).

Imaging tests will also be done to look for a mass in the adrenal gland. If a mass is found, a biopsy sample may be taken and looked at under the microscope. Your blood and urine will be tested to look for high levels of the hormones produced by some adrenal cortical adenomas and carcinomas. If a cancer is found, more tests will be done to determine its stage (how far the cancer has spread).

Signs and symptoms of adrenal cortex cancers

In about half of people with adrenal cortex cancer, symptoms are caused by the hormones made by the tumor. In the other half, symptoms occur because the tumor has grown so large that it presses on nearby organs. If you or your child has any of the signs or symptoms described in this section, discuss them with your doctor without delay. These symptoms may be caused by an adrenal cortical tumor or by something else. Getting the proper medical tests is the only way to find out. The sooner you get a correct diagnosis, the sooner you can start treatment and the more effective your treatment will be.

Symptoms caused by androgen or estrogen production

In children, the symptoms are most often caused by the *androgens* (male-type hormones) that the tumor might secrete. The most common symptoms are excessive growth of facial and body hair (such as in the pubic and underarm area). The male hormones may also cause enlargement of the penis in boys or the clitoris in girls. If the tumor secretes *estrogens* (female-type hormones), it can cause girls to start puberty early. This can cause the breasts to develop and menstrual periods to start. In boys, estrogen-producing tumors may cause breast enlargement.

In adults, the symptoms from high levels of sex hormones are less noticeable because they have already gone through puberty and have breasts and adult patterns of body hair. Women with estrogen-producing tumors and men with androgen-producing tumors usually do not have any symptoms unless the tumor is large enough to press on nearby organs. Symptoms are more easy to notice if the tumor is making the hormone that is not usually present. For example, men with tumors that make estrogen (female hormone) may notice breast enlargement with tenderness. They may also have sexual problems such as erectile dysfunction (impotence) and loss of sex drive. Women with tumors that make androgens (male hormones) may notice excessive facial and body hair growth, receding hairline, irregular menstrual periods, and deepening of their voice.

Symptoms caused by cortisol production

Excessive levels of cortisol causes a problem known *Cushing syndrome*. Although some people have all of these symptoms, many people with high cortisol levels have only 1 or 2 symptoms. These signs and symptoms include:

- weight gain, usually greatest around the chest and abdomen
- fat deposits behind the neck and shoulders
- purple stretch marks on the abdomen.
- excessive hair growth on the face, chest, and back in women
- menstrual irregularities
- weakness in the legs
- easy bruising
- depression and/or moodiness
- weakened bones (osteoporosis), which can lead to fractures
- high blood sugar, often leading to diabetes
- high blood pressure

Cushing syndrome may be caused by an adrenal cancer or an adrenal adenoma that produces high levels of cortisol and/or related hormones. Benign pituitary gland tumors and several types of cancers, such as lung cancer, can produce high levels of another hormone called adrenal corticotrophic hormone (ACTH). ACTH in turn causes normal adrenal gland tissue to produce more cortisol.

Some people with immune system problems or some cancers, such as lymphomas, are treated with drugs chemically related to cortisol. Because there are so many causes of high cortisol levels that can lead to Cushing syndrome, doctors do a number of blood tests, urine tests, and imaging tests to find out whether the patient has an adrenal cortical tumor or some other cause of Cushing syndrome.

Symptoms caused by aldosterone production:

The main signs and symptoms caused by aldosterone-producing adrenal tumors are high blood pressure, weakness, muscle cramps, excessive thirst, and excessive urination. Adrenal adenomas often produce this hormone, but adrenal cancers rarely do so.

Symptoms caused by a large adrenal cancer pressing on nearby organs:

As an adrenal cortical carcinoma grows, it presses on nearby organs and tissues. This may cause pain near the tumor, a feeling of fullness in the abdomen, or trouble eating because of a feeling of filling up easily.

Imaging tests

Chest x-ray

This can tell if the cancer has spread to the lungs. It may also be useful to determine if there are any serious lung or heart diseases.

Ultrasound

Ultrasound tests use sound waves to take pictures of parts of the body. A device called a transducer produces the sound waves, which are reflected by tissues of nearby organs. The pattern of sound wave echoes is detected by the transducer and analyzed by a computer to create an image of these tissues and organs. This test can show if there is a tumor mass in the adrenal gland. It can also diagnose tumor masses in the liver if the cancer has spread there. In general, it is not used to look for adrenal tumors unless a CT scan isn't able to be done.

Computed tomography (CT)

The CT scan is an x-ray procedure that produces detailed cross-sectional images of your body. Instead of taking 1 picture, like a conventional x-ray, a CT scanner takes many pictures as the camera rotates around you. A computer then combines these pictures into an image of a slice of your body. The machine will take pictures of many slices of the part of your body that is being studied.

CT scans show the adrenal glands fairly clearly and often can confirm the location of the cancer. It can also help show whether your cancer has spread into your liver or other organs nearby. CT scans can also show lymph nodes and distant organs where metastatic cancer might be present. The CT scan can help to determine if surgery is a good treatment option.

Before any pictures are taken, you may be asked to drink 1 to 2 pints of a liquid called "oral contrast." This helps outline the intestine so that certain areas are not mistaken for tumors. You may also receive an IV (intravenous) line through which a different kind of contrast dye (IV contrast) is injected. This helps better outline structures in your body.

The injection can cause some flushing (redness and warm feeling that may last hours to days). A few people are allergic to the dye and get hives. Rarely, more serious reactions like trouble breathing and low blood pressure can occur. Medicine can be given to prevent and treat allergic reactions. Be sure to tell the doctor if you have ever had a reaction to any contrast material used for x-rays.

CT scans can also be used to precisely guide a biopsy needle into a suspected metastasis. For this procedure, called a CT-guided needle biopsy, the patient remains on the CT scanning table, while a radiologist moves a biopsy needle toward the location of the mass. CT scans are repeated until the doctors are sure that the needle is within the mass. A fine needle biopsy sample (tiny fragment of tissue) or a core needle biopsy sample (a thin cylinder of tissue about ½ inch long and less than 1/8 inch in diameter) is removed and examined under a microscope.

CT scans take longer than regular x-rays and you need to lie still on a table while they are being done. But modern machines allow the scans to be complete in just a few minutes. Also, you might feel a bit confined by the ring you lie within when the pictures are being taken.

Positron emission tomography (PET)

In this test, radioactive glucose (sugar) is injected into the patient's vein. Because cancer cells use sugar much faster than normal tissues, radioactivity will tend to concentrate in the cancer. A scanner can spot the radioactive deposits. This test can be helpful for spotting small collections of cancer cells and may be used to finding cancer that has spread.

A special type of PET scan uses a radioactive form of a substance called metomidate. This substance seems to concentrate in adrenal cortical tissue, particularly adenomas and carcinomas. PET scanning with metomidate can be helpful in distinguishing tumors that start in the adrenal cortex from cancers that started in other organs and then spread to the adrenals. It is also helpful in finding adrenal cortex cancer that has spread outside of the adrenals.

Magnetic resonance imaging (MRI)

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 tissue and by certain

diseases. A computer translates the pattern of radio waves given off by the tissues into a very detailed image of parts of the body. Not only does this produce cross sectional slices of the body like a CT scanner, it can also produce slices that are parallel with the length of your body. For some MRI scans, a contrast material called gadolinium is injected into a vein (IV). MRI may sometimes provide more information than CT scans because it can better distinguish adrenal cancers from benign tumors.

MRI scans are particularly helpful in examining the brain and spinal cord. In people with suspected adrenal tumors, an MRI may be done to examine the pituitary gland. Tumors of the pituitary gland, which lies underneath the front of the brain, can cause symptoms and signs similar to adrenal tumors.

MRI scans are a little more uncomfortable than CT scans. First, they take longer -- often up to an hour. Also, you have to be placed inside a tube, which is confining and can upset people who have become anxious in tight spaces (claustrophobia). If you have problems with tight spaces, tell your doctor before your MRI. Medication may be given before the scan to help with anxiety. If that doesn't work, the exam may be scheduled at an open MRI scanner. These machines are not so enclosing and are easier for patients with claustrophobia. Also, the machine makes a thumping noise that you may find disturbing. Some places will provide headphones with music to block this sound out.

Other tests

Laparoscopy

This procedure uses a laparoscope, a thin, flexible tube with a tiny video camera on the end. It is inserted through a small surgical opening in the patient's side to allow the surgeon to see where the cancer is growing. It can spot distant spread as well as enlarged lymph nodes. Sometimes it is combined with ultrasound to give a better picture of the cancer. Laparoscopy may be done to help predict whether it will be possible to completely remove the cancer by surgery. In addition to viewing adrenal tumors through the laparoscope, surgeons can sometimes remove small tumors through this instrument. This method is described in the section, "How is adrenal cortical cancer treated?"

Biopsy

Imaging tests may find tumors, but the only way to know for sure that it is cancer is to remove a sample of tumor tissue to look at under the microscope. This is called a *biopsy*. For adrenal cancer, a biopsy may be done before surgery using a needle to get a sample of tissue. If a thin needle that only removes tiny bits of tissue is used, it is called a fine needle aspiration, or FNA for short. When a larger needle that removes a thin cylindrical core of tissue is used, it is called a *core needle biopsy*. In either case, the biopsy is often done using a CT scan or ultrasound to guide the tip of the needle into the tumor. A needle biopsy can show whether a tumor started in the adrenal cortex, the adrenal medulla, or in another part of the

body. Since adrenal adenomas and cancers can look alike under the microscope, a biopsy may not be able to tell whether or not an adrenal cortical tumor is cancerous. For this reason, a biopsy may not be done before surgery if an adrenal tumor's size and certain features seen on imaging tests suggest it is cancer.

If the cancer appears to have metastasized (spread) to another part of the body such as the liver, then a needle biopsy of the metastasis may be done. If a patient is known to have an adrenal tumor and a liver biopsy shows adrenal cells are present in the liver, then the tumor is cancer.

Tests for adrenal hormones

Blood and urine tests to measure levels of adrenal hormones are important in deciding whether a patient with signs and symptoms of adrenal cancer has the disease. For urine tests, you may be asked to collect all of your urine for 24 hours. Blood and urine tests aren't as crucial to the diagnosis of adrenal cortical carcinoma as the imaging studies, but they can be very important. Doctors choose which tests to do based on the patient's symptoms. Doctors know which symptoms are associated with high levels of certain hormones, so they can focus on ways to look for the hormones most likely to be affected.

Tests for high cortisol levels

The tests used in this case include measuring levels of cortisol and 17-hydroxy steroids in the urine. If an adrenal tumor is making cortisol, these levels will be abnormally high. These tests may be done after giving the patient a dose of dexamethasone. Dexamethasone is a drug that acts like cortisol. If given to someone who does not have an adrenal tumor, it will decrease production of cortisol and similar hormones. In someone with an adrenal cortex tumor, these hormone levels will remain high after they receive dexamethasone. Blood levels of ACTH will also be measured to help distinguish adrenal tumors from other diseases that can cause high cortisol levels.

Tests high aldosterone levels

The level of aldosterone will be measured and will be high if the tumor is making aldosterone. Also, high aldosterone leads to low levels of potassium and renin (a hormone produced by the kidneys) in their blood.

Tests for high androgen or estrogen levels

Patients with androgen-producing tumors will have high levels of testosterone and patients with estrogen-producing tumors will have high levels of estrogen in their blood. In either case, they will also have high levels of 17-ketosteroids and 17-hydroxysteroids in their urine. Blood levels of follicle-stimulating hormone (FSH), a pituitary hormone, may also be measured. If FSH levels are increased, the problem is likely to come from the pituitary, not from an adrenal cortical tumor.

How is adrenal cortical cancer staged?

Staging is the process of finding out how far the cancer has spread. It's very important because treatment options and the course of the disease as well as prognosis (outlook) are determined by the stage of the cancer. Adrenal cortical cancer is staged by the TNM system, where T (followed by a number) is used to describe the tumor. N (followed by a number) is used to describe the state of the lymph nodes, and M is used to describe the presence of tumor spread. Then the values for T, N, and M are combined together to decide the stage. This is called stage grouping.

T is for tumor

- T1: the tumor is 5 cm (about 2 inches) or less in size; it has not grown into tissues outside of the adrenal gland
- T2: the tumor is greater than 5 cm (2 inches) in size; it has not grown into tissues outside of the adrenal gland
- T3: the tumor is growing in the fat that surrounds the adrenal gland
- T4: the tumor is growing into nearby organs

N is for lymph nodes

- N0: the cancer has not spread to nearby lymph nodes
- N1: the cancer has spread to nearby lymph nodes

M is for metastases (distant cancer spread)

- M0: the cancer has not spread to distant organs or tissues (like liver, bone, brain)
- M1: the cancer has spread to distant sites

Stage groupings for adrenal cortical cancer

Stage I: T1, N0, M0

The cancer is smaller than 5 cm (2 inches) and has not spread to surrounding tissues or organs, lymph nodes, or other body parts.

Stage II: T2, N0, M0

The cancer is larger than 5 cm (2 inches) but still has not spread to surrounding tissues or organs, lymph nodes, or other body parts.

Stage III (T1 or T2, N1, M0) OR (T3, N0, M0)

Either

- The cancer has spread to nearby lymph nodes (N1) but not to distant sites (M0). The tumor can be any size (T1 or T2).

OR

- The cancer has grown into the fat outside of the adrenal gland (T3). It has not spread to nearby lymph nodes or to distant sites.

Stage IV: (T3, N1, M0) OR (T4, N0 or N1, M0) OR (Any T, any N, M1)

Either

- the cancer has grown into the fat outside of the adrenal gland (T3) and it has spread to nearby lymph nodes (N1); it has not spread to distant body sites (M0)

OR

- the cancer has grown from the adrenal gland into tissues nearby (T4), it may (N1) or may not (N2) have spread to nearby lymph nodes, but it has not spread to distant sites (M0)

OR

- the cancer has spread to distant sites (M1). It can be any size and may or may not have spread to nearby tissues or lymph nodes

Survival rates by stage

The outlook for people with cancers of the adrenal cortex depends on many factors. The most important factor is the stage of the cancer. About 4 in 10 patients are diagnosed with early stage disease (includes stage I and stage II). The rest are divided between stage III and stage IV.

Early stage cancer, when the tumor is small and has not spread, has a very good outlook, and many patients can be cured with surgery alone. Relative 5-year survival rates for people with stage I and stage II are about 65%.

People with more advanced disease where the cancer has spread have a less favorable outlook for survival. Patients with stage III adrenal cortical cancers have relative 5-year survival rates of about 40%, while less than 10% of patients with stage IV live at least 5 years. These numbers are all approximate because there are not many reported cases. 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 of these patients live much longer than 5 years after diagnosis, but 5-year rates are used to produce a standard way of discussing outlook. Five-year relative survival rates assume that some people will die of other causes and compare the observed survival with that expected for people without the cancer. This is a more accurate way to see the effect of that cancer on survival. It's also important to remember that 5-year rates are based on patients diagnosed and initially treated more than 5 years ago. Improvements in treatment often result in a more favorable prognosis (outlook) for recently diagnosed patients.

The numbers presented above are from the National Cancer Institute's SEER program, which collects data about cancer cases from all over the country.

How is adrenal cortical 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.

After the cancer is found, your doctor will discuss your treatment options with you. It is important to take time and think about all of the choices. In choosing a treatment plan, factors to consider include your overall physical health and the stage of the cancer. Sometimes it is a good idea to get a second opinion. A second opinion can provide more information and help you feel more confident about the treatment plan that is chosen. Another reason for people with adrenal cortical cancer to get a second opinion is that, because these cancers are so rare, only large cancer centers will have much experience in treating them.

Surgery

Surgery to remove the adrenal gland is called an adrenalectomy. There are 2 major approaches to removing the adrenal gland. One way is to remove the gland through an incision in the back, just below the ribs. This works well for small tumors, but it can be hard to see larger tumors well. For most adrenal cortical carcinomas, the surgeon makes the incision through the front of the abdomen. This allows the surgeon to see the tumor more clearly and makes it easier to see if it has spread. It also allows room for the surgeon to remove a large cancer that has spread (locally) to tissues and organs near the adrenal gland. For example, if the cancer has grown into the kidney, the kidney must also be removed. If it has grown into the muscle and fat around the adrenal gland, these tissues will need to be removed as well.

Sometimes, the cancer has grown into the inferior vena cava, the large vein that carries blood from the lower body to the heart. Complete removal of these cancers requires a very extensive operation to remove the tumor and preserve the vein. To remove the tumor from the vein, the surgeon may need to bypass the body's circulation by putting the patient on a

heart-lung bypass pump like that used in heart surgery. If the cancer has grown into the liver, a part of that organ containing the cancer may need to be removed as well.

It is also possible to remove some adrenal tumors through a hollow lighted tube called a *laparoscope*. The laparoscope is a thin tube with a tiny video camera on the end that is inserted through a small surgical opening in the patient's side. Other instruments inserted through this tube or through other very small incisions are used to remove the adrenal gland. The main advantage of this method is that because the incisions are smaller, patients recover from surgery more quickly. Although laparoscopic surgery is often used to treat adenomas, it may not be an option to treat some larger adrenal cancers. That is because when adrenal cancers have grown into nearby tissues or lymph nodes, it may be hard to remove the entire tumor using laparoscopy.

Radiation therapy

Radiation therapy uses high-energy radiation to kill cancer cells. External beam radiation therapy focuses radiation on the cancer from a machine outside the body. Treatments are often given once or twice a day, 5 days a week for several weeks. Each treatment lasts only a few minutes, and is similar to having a regular x-ray test. As with a diagnostic x-ray, the radiation passes through the skin and other tissues before it reaches the tumor. The actual radiation exposure time is very short, and most of the treatment time is spent precisely positioning the patient so that the radiation is aimed accurately at the cancer.

Brachytherapy (internal radiation therapy) uses small pellets of radioactive material placed next to or directly into the cancer, sometimes in thin plastic tubes. The tubes containing the pellets are left in place for a few days and then removed. The actual time is determined by the strength of the radioactive pellets and the size of the tumor.

Radiation therapy is not often used as the main treatment for adrenal cortical carcinoma because the cancer cells are not easy to kill with x-rays. Radiation may be used after surgery to help keep the tumor from coming back -- this is called adjuvant therapy. Radiation may also be used to treat areas of cancer spread, such as in the bones or brain.

Chemotherapy

Systemic chemotherapy (chemo) uses drugs that are given into a vein or by mouth (in pill form). These drugs enter the bloodstream and reach throughout the body, making this treatment useful for cancer that has spread or metastasized to organs beyond the adrenal gland. It is only used for adrenal gland cancer that has become widespread. Chemo does not cure adrenal cancer. It is most often used for adrenal cancer that has spread or come back after surgery

The drug most often used for people with adrenal cortical carcinoma is a drug called *mitotane*. Mitotane blocks hormone production by the adrenal gland and also destroys adrenal cancer cells. Some studies have shown that starting mitotane treatment when all of the cancer is thought to be gone may delay the return of the cancer. If the cancer has not been completely removed by surgery or has come back, mitotane will shrink the cancer in about 30% of patients. On average, the response lasts about one year. But the response time can be longer for some patients.

Mitotane is particularly helpful for people with adrenal carcinomas who are suffering from the effects of excessive hormone production. Even when it doesn't shrink the tumor, mitotane can reduce abnormal hormone production and relieve symptoms. About 80% of patients with excess hormone secretion are helped by mitotane. This drug can cause major side effects, however. The most common are nausea, vomiting, diarrhea, rashes, and sleepiness. Sometimes lower doses of the drug can still be effective and cause fewer side effects. This drug comes as a pill and is taken 3 to 4 times a day.

Giving mitotane after the cancer is completely removed with surgery may help keep the cancer from growing back. This approach is called adjuvant chemotherapy. It works well for other types of cancer, but has not been studied well in adrenal cortex cancer.

Some of the other chemo drugs used to treat adrenal cortical cancer are:

- cisplatin
- doxorubicin (Adriamycin)
- paclitaxel (Taxol)
- 5-fluorouracil (5-FU)
- vincristine
- etoposide

These drugs may be given in different combinations and are often given with mitotane.

Chemotherapy drugs kill cancer cells but also damage some normal cells, which can cause some side effects. Careful attention must be given to avoid or minimize chemo side effects. Side effects from chemo depend on the type of drugs, the amount taken, and the length of treatment. Common side effects might include:

- nausea and vomiting
- loss of appetite
- loss of hair
- hand and foot rashes
- mouth sores
- low blood counts

Because chemotherapy can damage the blood-producing cells of the bone marrow, patients may have low blood cell counts. This can increase the chance of infection (due to a shortage

of white blood cells), bleeding or bruising after minor cuts or injuries (due to a shortage of blood platelets), and anemia (due to low red blood cell counts).

Most side effects disappear once treatment is stopped. Hair will grow back after treatment ends, though it may look different. There are good treatments for many of the temporary side effects of chemotherapy. For example, very good drugs called antiemetics are available to prevent or reduce nausea and vomiting.

Some chemo side effects may last a long time or even be permanent. For example, doxorubicin can damage the heart muscle over time. Your health care team will watch the dose of this drug closely, to make sure that the dose isn't high enough to cause this damage. Cisplatin and paclitaxel can both cause nerve damage, leading to painful tingling and numbness in the hands and feet.

Other drugs

Other medications besides mitotane may be used to block hormone production by the cancer. Ketoconazole, aminoglutethimide, and metyrapone can reduce adrenal steroid hormone production. This can help relieve symptoms caused by these hormones, but doesn't cause the cancer to shrink.

Clinical trials

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 Web site at <http://clinicaltrials.cancer.org>. 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 Web site 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. They are 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 Web site or call our toll-free number (1-800-ACS-2345) and have it sent to you.

Complementary and alternative therapies

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 Web sites 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 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-ACS-2345 (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.

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.

Treating adrenal cortical carcinoma by stage

Stages I and II

Surgery is the main treatment for stage I and stage II adrenal cortical carcinoma. The entire adrenal gland will be removed. Since there are 2 adrenal glands, removal of the diseased one does not cause problems for the patient. If nearby lymph nodes are enlarged, they will be removed as well. Most surgeons do not remove these lymph nodes if their size is normal. If the cancer is not producing hormones, no further treatment may be necessary. However, there will be periodic follow-up examinations since even stage I cancers can come back. Another option is to give radiation or mitotane after surgery to help keep the cancer from coming back.

Stage III

Surgery is also the main treatment for stage III adrenal cortical cancer. The goal of surgery is to remove all of the cancer. The adrenal gland containing the tumor is always removed, and the surgeon might also need to remove some tissue around the adrenal, including part (or all) of the nearby kidney and part of the liver. The lymph nodes near the adrenal gland will also be removed. After surgery, treatment with radiation or mitotane may be given to help keep the cancer from coming back.

Stage IV

If it is possible to remove all of the cancer, then surgery may be done. When the cancer has spread to other parts of the body, it usually cannot be cured with surgery. Still, some doctors

may still recommend surgery to remove as much of the tumor as possible. This type of surgery is called debulking. Removing most of the cancer may help reduce symptoms by lowering the production of hormones. Radiation therapy may also be used to treat any areas of cancer that are causing symptoms. For example, radiation can help people with painful bone metastases. Mitotane therapy may begin at the time of surgery or the doctors may wait until there are symptoms. Other chemotherapy drugs may also be used.

Recurrent adrenal cortical carcinoma

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). Recurrent adrenal cortical cancer is treated like stage IV disease. Surgery may be done to relieve symptoms. People with recurrent disease are often treated with mitotane. They may also receive chemotherapy and/or radiation therapy. If the mitotane doesn't work or cannot be tolerated, other drugs can be given to lower hormone production.

Most of the time, these treatments provide only temporary help because the tumor will eventually continue to grow. When this happens and these treatments are no longer helping, treatment aimed at providing as good a quality of life as possible may be the best choice. The best drugs to treat pain are morphine and other narcotic drugs. Many studies have shown that taking morphine for pain does not mean a person will become addicted.

There are many other ways your doctor can help maintain your quality of life and control your symptoms. This means that you must tell your doctor how you are feeling and what symptoms you are having. Many patients don't like to disappoint their doctors by telling them they are not feeling well. This does no one any good.

More treatment information

The National Cancer Institute (NCI) provides treatment guidelines via its telephone information center (1-800-4-CANCER) and its Web site (www.cancer.gov). Detailed guidelines intended for use by cancer care professionals are also available on www.cancer.gov.

What should you ask your doctor about adrenal cortical cancer?

As you deal with your adrenal gland cancer and the process of treatment, you need to have frank, open discussions with your cancer care team. You should feel free to ask any question that is on your mind, no matter how trivial it might seem. Among the questions you might want to ask are:

- Do I have a benign or malignant adrenal gland tumor?
- Has my cancer spread beyond the adrenal gland?
- Is my tumor secreting excessive amounts of hormones?
- How do we treat the hormone excess?
- Are the changes to my body permanent?
- How do we treat the changes to my body?
- Is this form of adrenal gland cancer hereditary? Does my adrenal gland cancer affect any other organs?
- What other treatment choices do I have?
- What side effects can I expect from my treatments?
- What are the other risks of treatments?
- How long will it take me to recover from treatment?
- When can I go back to work after treatment?
- What are the chances that my cancer will recur?
- What should I do to be ready for treatment?
- Do I need a second opinion?
- Based on what you've learned about my cancer, will it shorten my life?

You will no doubt have other questions about your personal situation. Be sure and write your questions down so you remember to ask them during each visit with your cancer care team. Keep in mind, too, that doctors are not the only ones who can provide you with information. Other health care professionals, such as nurses and social workers, may have the answers you seek.

What happens after treatment for adrenal cortical cancer?

Completing treatment can be both stressful and exciting. You will be relieved to finish treatment, yet it is hard not to worry about cancer coming back. (When cancer returns, it is called recurrence.) This is a very common concern among those who have had cancer.

It may take a while before your confidence in your own recovery begins to feel real and your fears are somewhat relieved. You can learn more about what to look for and how to learn to live with the possibility of cancer coming back in our document, *Living With Uncertainty: The Fear of Cancer Recurrence*, available at 1-800-ACS-2345 (1-800-227-2345).

Follow-up care

After your treatment is over, it is very important to keep all follow-up appointments. During these visits, your doctors will ask about symptoms, do physical exams, and order blood tests or imaging studies such as CT scans or x-rays. Follow-up is needed to check for cancer recurrence or spread, as well as possible side effects of certain treatments. This is the time for

you to ask your health care team any questions you need answered and to discuss any concerns you might have.

Almost any cancer treatment can have side effects. Some may last for a few weeks to several months, but others can be permanent. Don't hesitate to tell your cancer care team about any symptoms or side effects that bother you so they can help you manage them.

It is also important to keep medical insurance. Even though no one wants to think of their cancer coming back, it is always a possibility. If it happens, the last thing you want is to have to worry about paying for treatment. Should your cancer come back our document, *When Your Cancer Comes Back: Cancer Recurrence*, gives you information on how to manage and cope with this phase of your treatment. You can get this document by calling 1-800-ACS-2345.

If you are treated for adrenal carcinoma, your follow-up care will be very important. One reason for this is that regardless of your stage of disease, the cancer can recur. You should see your doctor frequently after treatment and less often later on. If you have stage II, III, or IV disease and are treated with mitotane your follow-up may need to be more frequent. Remember that mitotane will also suppress the usual adrenal steroid hormone production from your other, normal adrenal gland. As a result, you will need to take steroid hormone replacement tablets to protect you against this cortisol deficiency. CT scans may be done periodically to see if the cancer has returned or is continuing to grow. Periodic tests of your blood and urine hormone levels will be done to evaluate the success of medications in suppressing hormone production by the cancer.

Seeing a new doctor

At some point after your cancer diagnosis and treatment, you may find yourself in the office of a new doctor. Your original doctor may have moved or retired, or you may have moved or changed doctors for some reason. It is important that you be able to give your new doctor the exact details of your diagnosis and treatment. Make sure you have the following information handy and always keep copies for yourself:

- a copy of your pathology report from any biopsy or surgery
- if you had surgery, a copy of your operative report
- if you were hospitalized, a copy of the discharge summary that every doctor must prepare when patients are sent home from the hospital
- if you received radiation, a copy of your treatment summary
- finally, since some drugs can have long-term side effects, a list of your drugs, drug doses, and when you took them

Lifestyle changes to consider during and after treatment

Having cancer and dealing with treatment can be time-consuming and emotionally draining, but it can also 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 begin this process during cancer treatment.

Make healthier choices

Think about your life before you learned you had cancer. Were there things you did that might have made you less healthy? Maybe you drank too much alcohol, or ate more than you needed, or smoked, or didn't exercise very often. Emotionally, maybe you kept your feelings bottled up, or maybe you let stressful situations go on too long.

Now is not the time to feel guilty or to blame yourself. However, you can start making changes today that can have positive effects for the rest of your life. Not only will you feel better but you will also be healthier. What better time than now to take advantage of the motivation you have as a result of going through a life-changing experience like having cancer?

You can start by working on those things that you feel most concerned about. 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's Quitline[®] tobacco cessation program at 1-800-ACS-2345.

Diet and nutrition

Eating right can be a challenge for anyone, but it can get even tougher during and after cancer treatment. For instance, treatment often may change your sense of taste. Nausea can be a problem. You may lose your appetite for a while and lose weight when you don't want to. On the other hand, some people gain weight even without eating more. This can be frustrating, too.

If you are losing weight or have taste problems during treatment, do the best you can with eating and remember that these problems usually improve over time. You may want to ask your cancer team for a referral to a dietitian, an expert in nutrition who can give you ideas on how to fight some of the side effects of your treatment. You may also find it helps to eat small portions every 2 to 3 hours until you feel better and can go back to a more normal schedule.

One of the best things you can do after treatment is to put healthy eating habits into place. You will be surprised at the long-term benefits of some simple changes, like increasing the variety of healthy foods you eat. Try to eat 5 or more servings of vegetables and fruits each day. Choose whole grain foods instead of white flour and sugars. Try to limit meats that are high in fat. Cut back on processed meats like hot dogs, bologna, and bacon. Get rid of them

altogether if you can. If you drink alcohol, limit yourself to 1 or 2 drinks a day at the most. And don't forget to get some type of regular exercise. The combination of a good diet and regular exercise will help you maintain a healthy weight and keep you feeling more energetic.

Rest, fatigue, work, and exercise

Fatigue is a very common symptom in people being treated for cancer. This is often not an ordinary type of tiredness but a “bone-weary” exhaustion that doesn't get better with rest. For some, this fatigue lasts a long time after treatment, and can discourage them from physical activity.

However, exercise can actually help you reduce fatigue. Studies have shown that patients who follow an exercise program tailored to their personal needs feel physically and emotionally improved and can cope better.

If you are ill and need to be on bed rest during treatment, it is normal to expect your fitness, endurance, and muscle strength to decline some. Physical therapy can help you maintain strength and range of motion in your muscles, which can help fight fatigue and the sense of depression that sometimes comes with feeling so tired.

Any program of 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 3 times a week. If you haven't exercised in a few years but can still get around, you may want to think about taking short walks.

Talk with your health care team before starting, and get their opinion about your exercise plans. Then, try to get an exercise buddy so that 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, though, you will need to balance activity with rest. It is okay to rest when you need to. It is really hard for some people to allow themselves to do that when they are used to working all day or taking care of a household. (For more information about fatigue, please see the publication, *Cancer Related Fatigue and Anemia Treatment Guidelines for Patients*.)

Exercise can improve your physical and emotional health.

- It improves your cardiovascular (heart and circulation) fitness.
- It strengthens your muscles.
- It reduces fatigue.
- It lowers anxiety and depression.
- It makes you feel generally happier.

- It helps you feel better about yourself.

And long term, we know that exercise plays a role in preventing some cancers. The American Cancer Society, in its guidelines on physical activity for cancer prevention, recommends that adults take part in at least 1 physical activity for 30 minutes or more on 5 days or more of the week. Children and teens are encouraged to try for at least 60 minutes a day of energetic physical activity on at least 5 days a week.

How about your emotional health?

Once your treatment ends, you may find yourself overwhelmed by emotions. This happens to a lot of people. You may have been going through so much during treatment that you could only focus on getting through your treatment.

Now you may find that you think about the potential of your own death, or the effect of your cancer on your family, friends, and career. You may also begin to re-evaluate your relationship with your spouse or partner. Unexpected issues may also cause concern -- for instance, as you become healthier and have fewer doctor visits, you will see your health care team less often. That can be a source of anxiety for some.

This is an ideal time to seek out emotional and social 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 individual counselors.

Almost everyone who has been through cancer can benefit from getting some type of support. 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 realistic to go it all by yourself. And your friends and family may feel shut out if you decide 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-ACS-2345 and we can put you in touch with an appropriate group or resource.

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 healthy choices and feeling as well as possible, physically and emotionally.

What happens if treatment is no longer working?

If cancer continues to grow after one kind of treatment, or if it returns, it is often possible to try another treatment plan that might still cure the cancer, or at least shrink the tumors enough to help you live longer and feel better. On the other hand, when a person has received several different medical treatments and the cancer has not been cured, over time the cancer tends to become resistant to all treatment. At this time it's important to weigh the possible limited benefit of a new treatment against the possible downsides, including continued doctor visits and treatment side effects.

Everyone has his or her own way of looking at this. Some people may want to focus on remaining comfortable during their limited time left.

This is likely to be the most difficult time in your battle with cancer -- when you have tried everything medically within reason and it's just not working anymore. Although your doctor may offer you new treatment, you need to consider that at some point, continuing treatment is not likely to improve your health or change your prognosis or survival.

If you want to continue treatment to fight your cancer as long as you can, you still need to consider the odds of more treatment having any benefit. In many cases, your doctor can estimate the response rate for the treatment you are considering. Some people are tempted to try more chemotherapy or radiation, for example, even when their doctors say that the odds of benefit are less than 1%. In this situation, you need to think about and understand your reasons for choosing this plan.

No matter what you decide to do, it is important that you be as comfortable as possible. Make sure you are asking for and getting treatment for any symptoms you might have, such as pain. This type of treatment is called "palliative" treatment.

Palliative treatment helps relieve these symptoms, but is not expected to cure the disease; its main purpose is to improve your quality of life. Sometimes, the treatments you get to control your symptoms are similar to the treatments used to treat cancer. For example, radiation therapy might be given to help relieve bone pain from bone metastasis. Or chemotherapy might be given to help shrink a tumor and keep it from causing a bowel obstruction. But this is not the same as receiving treatment to try to cure the cancer.

At some point, you may benefit from hospice care. Most of the time, this can be given at home. Your cancer may be causing symptoms or problems that need attention, and hospice focuses on your comfort. You should know that receiving hospice care doesn't mean you can't have treatment for the problems caused by your cancer or other health conditions. It just means that the focus of your care is on living life as fully as possible and feeling as well as you can at this difficult stage of your cancer.

Remember also that maintaining hope is important. 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. In a way, pausing at this time in your cancer treatment is an

opportunity to refocus on the most important things in your life. This 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.

What's new in adrenal cortical cancer research and treatment?

Research focused on adrenal cortex cancer is currently underway. Imaging tests for diagnosing this cancer, medical laboratory tests to more accurately distinguish adenomas from carcinomas, and new treatments are being studied. Progress in this research is sometimes limited by the rarity of this type of cancer. Other studies of more general aspects of cancer that can be applied to adrenal cortex cancers as well as other types of cancers are also being done.

Some clinical trials currently underway are testing new combinations of chemotherapy drugs. One is testing the value of mitotane in preventing recurrence of localized adrenal cancers after surgery. Several clinical trials are studying targeted therapies, either by themselves, or in combination with other drugs such as mitotane. Targeted therapies are a group of newer drugs that take advantage of gene changes in cells that cause cancer. They generally cause fewer and less severe side effects than usual chemo. These drugs have been found to be effective for several more common types of cancer but their value for adrenal cancer is still not known.

Scientists are learning how changes in certain oncogenes and tumor suppressor genes can cause normal adrenal cortex cells to become cancerous. Understanding these genetic changes will help doctors develop better methods of diagnosing this disease as well as treatments that are more effective and have fewer side effects than ones currently available.

Additional resources

More information from your American Cancer Society

We have selected some related information that may also be helpful to you. The following document may be ordered from our toll-free number, 1-800-ACS-2345 (1-800-227-2345).

After Diagnosis: 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)

When Your Cancer Comes Back: Cancer Recurrence

The following books are available from the American Cancer Society. Call us at 1-800-ACS-2345 to ask about costs or to place your order.

American Cancer Society's Guide to Pain Control

Caregiving: A Step-By-Step Resource for Caring for the Person with Cancer at Home

Coming to Terms with Cancer: A Glossary of Cancer-Related Terms

Consumers Guide to Oncology Drugs

Informed Decisions, Second Edition: The Complete Book of Cancer Diagnosis, Treatment, and Recovery

National organizations and web sites*

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

National Cancer Institute

Toll-free number: 1-800-422-6237 (1-800-4-CANCER); TTY: 1-800-332-8615

Web site: www.cancer.gov

National Coalition for Cancer Survivorship

Toll-free number: 1-877-622-7937 (1-877-NCCS-YES)

Web site: www.canceradvocacy.org

**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-ACS-2345** or visit www.cancer.org.

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