



GASTROINTESTINAL (DIGESTIVE SYSTEM) CARCINOID TUMORS

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 often travel to other parts of the body where they begin to grow and replace normal tissue. This process, called **metastasis**, occurs as the cancer cells get into the bloodstream or lymph vessels of our body. But when cells from a cancer, like breast cancer, spread to another organ, like the liver, the cancer is still called breast cancer, not liver cancer.

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.

Not all tumors are cancerous. Benign (non-cancerous) tumors do not spread to other parts of the body (metastasize) 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 a Gastrointestinal Carcinoid Tumor?

The Gastrointestinal (Digestive) System

The digestive system processes food for energy and rids the body of solid waste. After food is chewed and swallowed, it enters the **esophagus**. This is a tube-shaped organ that carries food to the stomach through the neck and chest. The esophagus joins the **stomach** just beneath the diaphragm (the breathing muscle under the lungs). The stomach is a sac-like organ that holds food and begins the digestive process by secreting gastric juice. The food and gastric juices are mixed into a thick fluid, which is then emptied into the small intestine. The **small intestine** continues breaking down the food and absorbs most of the nutrients. It is the longest section of the gastrointestinal (GI) tract. The small intestine then joins the **colon**. This is a muscular tube about 5 feet long. The **appendix** is found near the junction of small intestine and colon. The colon continues to absorb water and mineral nutrients from the food matter and serves as a storage place for waste. The waste left after this process goes into the rectum. From there it passes out of the body through the anus.

The Diffuse Neuroendocrine System

Carcinoid tumors develop from cells of the **diffuse neuroendocrine system**. The diffuse neuroendocrine system consists of cells that are like **nerve cells** in certain ways and like **gland** (endocrine - hormone-producing) cells in other ways. These cells do not form an actual organ like the parathyroid, adrenal, or thyroid glands. Instead, they are scattered throughout other organs like the esophagus, stomach, intestines, and lungs. Because of the size of the digestive system, it has the most endocrine cells of any organ. This may explain why carcinoid tumors most often start in the digestive system.

Neuroendocrine cells of the digestive system help regulate the release of digestive juices, help control the speed at which food moves along the gastrointestinal tract, and may help control the growth of other types of digestive system cells.

Carcinoid Tumors

Like most cells of the body, gastrointestinal system neuroendocrine cells sometimes undergo certain changes that cause them to grow too much and form tumors. The tumors that develop from neuroendocrine cells are known as neuroendocrine tumors (or neuroendocrine cancers). There are many varieties of neuroendocrine tumors, but the most common are the carcinoid tumors or carcinoids.

Carcinoid tumors act like the cells they come from. They often release certain hormone-like substances into the bloodstream. In about 10% of people, the carcinoid tumors spread and grow very large and release high amounts of those hormones. These cause symptoms such as facial flushing (redness and warm feeling), wheezing, diarrhea, and a fast heartbeat. These symptoms are grouped together and called the "carcinoid syndrome." Most cancers cause symptoms only in the organs they start in or spread to. But carcinoid tumors can release substances into the blood that cause symptoms throughout the body.

Most tumors in the intestinal tract come from different **glandular cells** (the kind of gland cell that produce mucus rather than hormones) of the inner lining of the digestive system. These can be either **adenomas** (benign) or **adenocarcinomas** (malignant). These tumors are quite different from carcinoid tumors in their symptoms, their prognosis (course of the disease and outlook for survival), and their treatment. For these reasons, it is important for doctors to find out whether a patient has a carcinoid tumor, an adenoma, an adenocarcinoma,

some other type of tumor, or a non-cancerous condition. And it is important for patients to understand that carcinoids are not the same as other, more common types of tumors.

What Are the Key Statistics About Gastrointestinal Carcinoid Tumors?

About 11,000 to 12,000 carcinoid tumors are diagnosed each year in the United States. About half of these occur in the digestive system. About 30% of carcinoid tumors occur in the lungs. The rest develop in other organs. The number of carcinoid tumors diagnosed has been increasing at a rate of around 6% per year. The reason for this is unknown. Some think it is because they are found during the course of an exam, such as CT scan or endoscopy, when the doctor is looking for something else. There are probably many more people with carcinoid tumors. They never cause a problem and are only discovered at autopsy when a person dies of something else, or when someone has surgery for an unrelated digestive system condition.

Within the digestive system, the most common location of carcinoid tumors is the small intestine, often near the appendix (ileum). The next most common site is the rectum, followed by the colon (large intestine), the appendix, and the stomach. At one time, the appendix was a very common site, but that is no longer true. Only about 11% of all gastrointestinal carcinoid tumors begin in the appendix.

The average age of people diagnosed with carcinoids is 61. The average age of people with small bowel carcinoids is slightly older and those with appendiceal carcinoids, younger. Carcinoid tumors are slightly more common in females than males.

What Are the Risk Factors for Gastrointestinal Carcinoid Tumors?

A **risk factor** is anything that increases your chance of getting a disease such as a type of cancer. For example, unprotected exposure to strong sunlight is a risk factor for skin cancer and smoking is a risk factor for lung, mouth, throat, kidney, bladder, colorectal, and several other cancers. However, cancer can develop without any risk factors present. Risk factors for gastrointestinal carcinoid tumors include:

Family history of multiple endocrine neoplasia, type 1: Multiple endocrine neoplasia, type I (MEN1) is a hereditary condition that has a very high risk of developing tumors of 3 glands: the pituitary, parathyroid, and pancreas. Carcinoid tumors are a less common component of this condition. Some studies estimate that MEN1 is responsible for about 10% of carcinoid tumors. Most of these are gastric (stomach) carcinoids. MEN1 is inherited by about half of the children of each affected parent. If your family is affected by the MEN1 syndrome, you should speak with your doctor about the advantages and disadvantages of biochemical or genetic testing for this condition. The DNA mutations that cause tumors in people with multiple endocrine neoplasia, type I (MEN1) have been identified. Testing for these gene mutations currently is only available in research settings.

Smoking: Smoking may double the risk of developing a carcinoid tumor of the small intestine, according to a recent European study.

Race and gender: Carcinoid tumors are slightly more common among African Americans than whites. Outcomes are also poorer in black people. Researchers do not yet know why. Carcinoid tumors are also slightly more common in females than males.

Other stomach conditions: People with certain diseases that damage the stomach and reduce acid production by the stomach have a greater risk of developing stomach carcinoid tumors, but their risk for carcinoid tumors

of other organs is not affected.

Diet: Risk of developing gastrointestinal carcinoid tumors does not appear to be increased or decreased by any specific foods.

Do We Know What Causes Gastrointestinal Carcinoid Tumors?

Very little is known about the causes of gastrointestinal carcinoid tumors.

As with other cancers, scientists have recognized some changes in the DNA of carcinoid tumor cells that are probably responsible for their increased growth and abnormal spread. But the causes of these changes are not yet known.

Doctors do know that carcinoid tumors start out very small and grow slowly. When patients have parts of their stomach or small intestine removed to treat other diseases, close examination under the microscope often shows small groups of neuroendocrine cells that look like tiny carcinoids. Most of these miniature tumors, sometimes called **tumorlets**, never develop into an actual carcinoid tumor. Researchers still do not know why some remain so small and others begin to grow and become large enough to cause symptoms.

Can Gastrointestinal Carcinoid Tumors Be Prevented?

At this time, there is no known way to prevent gastrointestinal carcinoid tumors. Since smoking may increase the risk of carcinoid tumors of the small intestine, not starting or quitting smoking may reduce the risk for this disease.

Can Gastrointestinal Carcinoid Tumors Be Found Early?

Because carcinoid tumors usually grow and spread slowly, about half of all gastrointestinal carcinoid tumors are found at an early or localized stage.

Incidental Diagnosis of Carcinoid Tumors

In many cases, carcinoid tumors are found incidentally. This means that the tumors did not cause any symptoms but were discovered during tests done for other diseases or because parts of the gastrointestinal system were removed to treat other diseases. For example, a person with symptoms due to inflammation of the stomach may have a test called an **upper endoscopy**. During this test, the doctor viewing the stomach lining through a flexible lighted tube may incidentally notice a small bump in the stomach wall that turns out to be a carcinoid tumor.

Sometimes a routine **sigmoidoscopy** or **colonoscopy** (viewing the large bowel through a flexible lighted tube) for colorectal cancer screening will incidentally find a small carcinoid tumor.

Occasionally, when a person's appendix is removed to treat acute appendicitis (infection and inflammation of the appendix that causes abdominal pain), doctors may incidentally find a small carcinoid, as well as expected inflammation. Some studies have found that about 1 of every 300 people who have appendix surgery done for other diseases turn out to have a tiny carcinoid near the tip of their appendix. In most of these cases, the

carcinoid was still too small to have caused any problems. The number of these appendix carcinoids is dropping. Some people think that is because these days, surgeons are less likely to remove the appendix in the course of an abdominal operation for another problem.

How Are Gastrointestinal Carcinoid Tumors Diagnosed?

If there is a reason to suspect you may have a gastrointestinal carcinoid tumor, the doctor will use one or more methods to find out if the disease is really present.

Signs and Symptoms of Gastrointestinal Carcinoid Tumors

The symptoms a person develops from a carcinoid tumor often depends on where it is located. People with carcinoid tumors in their **appendix** often don't have symptoms. Often, it is discovered when they have their appendix removed during the course of an operation for some other problem. If the carcinoid starts in the **small intestine**, it can lead to abdominal pain caused by "kinking" or blockage of the intestines. This pain can be mild and last for a couple of years or more before the carcinoid tumor is found. Sometimes, carcinoid tumors may cause intestinal bleeding. Many are found in the course of a routine exam of the intestinal tract. The same is also true for carcinoid tumors that start in the **colon**. These can grow fairly large, though, before causing intestinal blockage. **Rectal** carcinoids are often found on routine exam, though they can cause pain and bleeding from the rectum.

Carcinoids that develop in the **stomach** are usually slow growing and often do not cause symptoms. They are discovered during routine exam of the stomach by endoscopy (see below). Some stomach carcinoids can cause symptoms such as the carcinoid syndrome (see below)

About 10% of the time, carcinoid tumors produce hormone-like substances that are released into the bloodstream. The **carcinoid syndrome** results from the effect of these substances. Symptoms include facial flushing (redness and warm feeling), severe diarrhea, wheezing, and fast heartbeat. Many patients find that stress, strenuous exercise, and drinking alcohol may make these symptoms worse. Over a long time, these hormone-like substances can damage heart valves, causing shortness of breath, weakness, and a heart murmur (abnormal heart sounds a doctor can hear through a stethoscope). Some carcinoid tumors may produce adrenocorticotrophic hormone (ACTH), a substance that stimulates the adrenal gland to produce excessive amounts of cortisol and related adrenal hormones. Symptoms of excessive amounts of adrenal hormones include weight gain, weakness, secondary diabetes, and increased body and facial hair.

Not all gastrointestinal carcinoid tumors can cause the carcinoid syndrome. Rectal carcinoids usually do not produce the hormone-like substances that cause these symptoms. Also, normal liver cells break down these substances. Because blood from the gastrointestinal tract flows through the liver, substances produced by gastrointestinal carcinoid tumors are broken down in the liver and do not cause symptoms. But spread of gastrointestinal carcinoids to the liver interferes with the breakdown of substances produced by the tumor cells. When spread to the liver occurs, carcinoid syndrome may result if the tumor cells produce large amounts of symptom-causing hormones.

Medical history and physical exam: A medical history is an interview in which the doctor asks questions about symptoms and risk factors you may have. If you have one or more symptoms that suggest this type of tumor, the doctor will specifically ask about other symptoms of the carcinoid syndrome, as well as symptoms caused by the presence of a mass in the stomach, intestines, or rectum.

Some patients with carcinoid tumors also have cancers or benign tumors of other organs, so doctors will ask

about symptoms that might suggest other tumors are present. A thorough physical exam will provide information about signs of carcinoid tumor and other health problems. The doctor will pay special attention to the abdomen, looking for a tumor mass or enlarged liver.

Imaging Tests

Barium x-rays: These x-ray studies use a barium-containing solution that coats the lining of the esophagus, stomach, and intestines. These are often useful for the diagnosis of some gastrointestinal carcinoid tumors. They are least effective in finding small intestine carcinoid tumors. The coating of barium helps find abnormalities of the lining of these organs. Barium studies can be used to examine the upper or lower parts of the digestive system.

A **barium swallow or upper gastrointestinal (GI) x-ray** study is used to examine the lining of the esophagus, stomach, and the first part of the small intestine. Patients undergoing this test drink a barium solution before the x-ray pictures are taken.

A **barium enema** with air contrast is used to examine the inner surface of the large intestine. Strong laxatives and enemas are used to cleanse the bowel the night before and the morning of the exam. For this test, the barium solution is given through the anus. When the colon is about half full of barium, the patient is turned on the x-ray table so the barium spreads throughout the colon. In addition to barium, air can be blown into the large intestine to help push the barium towards the wall of this organ and better coat its inner surface. When air is added to a barium x-ray study, it is called a **double-contrast study**. In general, barium x-rays are used less these days. They are being replaced by **endoscopy** -- where the doctor actually looks into the colon or stomach with a narrow fiberoptic scope.

Computed tomography (CT): The CT scan is an x-ray procedure that produces detailed cross-sectional images of your body. Instead of taking one picture, like a conventional x-ray, a CT scanner takes many pictures as it rotates around you. A computer then combines these pictures into an image of a slice of your body. The machine takes pictures of multiple slices of the part of your body that is being studied. This test can help tell if your carcinoid tumor has spread into lymph nodes or other organs such as your liver. Often after the first set of pictures is taken you will receive an intravenous (IV) injection of a contrast agent, or dye, which helps better outline structures in your body. A second set of pictures is then taken.

CT scans can also be used to guide a biopsy needle precisely 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 confident 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 are more tedious than regular x-rays because they take longer and you need to lie still on a table while they are being done. But just like other computerized devices, they are getting faster and your stay might be pleasantly short. Also, you might feel a bit confined by the ring you lay within when the pictures are being taken.

You will need an IV line through which the contrast dye is injected. The injection can cause some flushing. Some people are allergic and get hives, or rarely, more serious reactions like trouble breathing and low blood pressure can occur. Please be sure to tell the doctor if you have ever had a reaction to any contrast material used for x-rays.

Radioactive scans: Two procedures have been used. The older one is called **I¹³¹-MIBG scan**. This procedure uses a chemical called MIBG to which radioactive iodine (I¹³¹) is attached. This is injected into your vein and then your body is scanned to look for areas that picked up the radioactivity. These would most likely be carcinoid tumors, although other kinds of neuroendocrine tumors will also pick up this chemical.

A second kind of scan is **indium¹¹¹-labeled DTPA-octreotide scintigraphy** or **octreoscan**. Octreotide is a hormone-like substance that attaches to carcinoid cells. A small amount of this radioactive octreotide is injected into a vein. This material is attracted to carcinoid tumors. A special camera scans your body to show where the radioactivity collects. This procedure and the I¹³¹-MIBG scan are useful in detecting spread of gastrointestinal carcinoid tumors to other areas of the body.

Positron emission tomography (PET): This is a special kind of radioactive scan. PET scanning for carcinoid tumors uses a radioactive form of 5-hydroxytryptophan, a chemical that is taken up and used by carcinoid cells. A special camera can detect the radioactivity. PET scans are useful when your doctor thinks the cancer has spread but doesn't know where. PET scans can be used instead of several different x-rays because it scans your whole body. Some doctors have found it to be more accurate than a CT scan for detecting spread of disease.

Other Tests

Endoscopy: This test uses a flexible lighted tube with a video camera on the end. The camera is connected to a monitor, allowing the doctor to clearly see any masses in the lining of the digestive organs. If abnormalities are noted, small pieces of tissue can be removed through the endoscope (biopsy). The tissue can be examined under the microscope to find out if cancer is present and what kind of cancer it is.

For **upper endoscopy**, patients are sedated (made sleepy) and a tube is passed down through the mouth to view the esophagus, stomach, and first part of the small bowel. In a **colonoscopy**, a colonoscope (type of endoscope) is inserted through the anus up into the colon. The colonoscope allows the doctor to see the lining of the entire rectum and colon. If you have a colonoscopy, you will need to take a bowel preparation (laxative agent) beforehand to clean your colon. A colonoscopy should not cause pain because you will be given intravenous medication to make you feel relaxed and sleepy during the procedure. A colonoscopy may be done in a hospital outpatient department, in a clinic, or in a doctor's office. It usually takes 15 to 30 minutes, although it may take longer if a tumor is seen and/or a biopsy taken.

Endoscopic ultrasound: This is a new technique in which a special instrument is used in patients having endoscopy. For this test, the endoscope has a small ultrasound probe on the end. This probe releases high frequency sound waves and then detects the sound wave echoes that bounce off tissues of the stomach wall. A computer then translates the pattern of echoes into an image of the wall of the esophagus, stomach, intestine, or rectum.

Endoscopic ultrasound is sometimes useful in determining how far a tumor has spread through the wall of the esophagus, stomach, intestine, or rectum. The test can also help predict whether the tumor has spread beyond the wall of these organs to nearby tissues or lymph nodes.

None of the techniques described can readily find small intestine carcinoids. So far, it has not been possible to look very far into the small intestine with an endoscope. Barium x-rays are also not very good at finding tumors there either. That is why these carcinoids are often not discovered early.

Biopsy: Even if a barium x-ray and/or CT scan finds a mass, these imaging tests cannot tell if the mass is a

carcinoid tumor, some other type of tumor (benign or cancerous), or a localized infection. The only way to know for sure is to remove cells from the abnormal area and examine them under a microscope. This procedure is called a **biopsy**.

There are several ways to take a sample from a gastrointestinal tumor. One way is through the endoscope. When a tumor is found, the doctor can use a biopsy forceps (pincers or tongs) through the tube to take a small sample of the tumor. Even though the specimen the doctor takes will be very small, doctors can usually make an accurate diagnosis. Bleeding after a biopsy from a carcinoid tumor is a rare but potentially serious problem. If bleeding becomes a problem, doctors can sometimes inject drugs that constrict blood vessels through the endoscope into the tumor to stop the bleeding.

In rare cases, neither an endoscopic biopsy nor a CT scan-guided needle biopsy (refer to section on CT scan above) will be able to provide tissue to identify the type of tumor. A laparotomy (surgically opening the abdomen) to remove a tissue sample will be needed. For example, an endoscope cannot be passed into the small intestine and surgery may be needed to diagnose small intestinal carcinoids.

Blood and urine tests: Blood tests may be done to detect some of the hormone-like substances produced by carcinoid tumors, particularly if the patient has symptoms of the carcinoid syndrome, caused by excessive levels of such substances in the blood. The most commonly used blood tests measure levels of chromogranin A or neuron-specific enolase. Depending on the patient's symptoms, doctors may recommend additional blood tests.

Serotonin is one of the substances produced by some carcinoid tumors, especially those developing in the small intestine. Serotonin in the blood can be measured. Also, it is broken down to 5-hydroxyindoleacetic acid (abbreviated 5-HIAA), which is released into the urine. Urine tests to measure 5-HIAA levels are very useful in diagnosing carcinoid tumors that produce serotonin and have spread to the liver. However, localized gastrointestinal carcinoid tumors often do not have positive urine 5-HIAA results.

How Are Gastrointestinal Carcinoid Tumors Staged?

Staging -- or determining the **stage** of disease -- is the process of finding out how localized or widespread the carcinoid tumor is. It will show whether the tumor has spread and how far. The treatment and prognosis (the outlook for chances of survival) for a patient with a gastrointestinal carcinoid tumor depends, to a large extent, on the tumor's stage.

There is no standard system for describing the spread of gastrointestinal carcinoid tumors. Some doctors use the same systems that are used for other cancers of the same organs. Many doctors simply divide all gastrointestinal carcinoid tumors into 3 general stages: localized, regional spread, and distant spread. This approach is easy for patients to understand and is useful in considering treatment options.

Localized: The carcinoid tumor has not spread beyond the wall of the organ it developed in (for example, the stomach, intestine, or rectum).

Regional spread: The carcinoid tumor has spread through the wall of the organ it started in to involve nearby tissues such as fat, ligaments, muscle, or lymph nodes.

Distant spread: The carcinoid tumor has spread to tissues or organs that are not near the organ where the cancer started. Spread of a gastrointestinal carcinoid tumor to the liver, bones, or lungs, for example, is considered distant spread.

5-year Survival Rates by Stage and Primary Site

First, it is important to note that people are not statistics. Every person is different, and every person will have his/her own experience with cancer and its treatment. Second, cancer statistics are based upon patient data from 5 to 10 years ago. Today, newer treatments may offer more or more effective treatment options than those of more than 5 years ago.

Statistically, 5-year survival rates are calculated based on how many patients live at least 5 years after diagnosis of this disease. Many may live much longer than 5 years, but it is the 5-year mark that is used as a standard measurement for statistics and research.

About half the time carcinoids are localized to the organ they start in. Another fourth of the time they have spread to nearby surrounding tissues and another fourth of the time they have spread to distant sites, most often the liver.

Around 75% of people with carcinoids die of the cancer in 5 years. When the cancer is localized, the chance of surviving the cancer is 95%. If it has spread to nearby tissues, this drops to anywhere from 40% to 80%, depending on the site. For example, the survival rate is only 40% if the carcinoid started in the stomach, but 84% if it started in the small intestine. When the cancer has spread to distant sites, the chances of surviving the cancer at 5 years ranges from 15% to 51%.

How Are Gastrointestinal Carcinoid Tumors Treated?

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

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

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

After the carcinoid tumor is found and staged, the cancer care team will suggest one or more treatment plans. This is an important decision. It is also important for you to take time and think about all of the choices.

The main factors in selecting treatment options for gastrointestinal carcinoid tumors are the size and location of the tumor, whether it has spread to lymph nodes, liver, bones, or other organs, whether there are any other serious medical conditions, and whether the tumor is causing bothersome symptoms. It is often a good idea to seek a second opinion. A second opinion may provide more information and help the patient feel more confident about the treatment plan that is chosen.

Surgery

Most gastrointestinal carcinoid tumors are cured by surgery alone. The type of operation will depend on a

number of factors, including the size and location of the tumor, and whether the patient has any serious diseases of other organs, and whether the carcinoid tumor is causing the carcinoid syndrome.

In general, surgeons try to cure localized carcinoid tumors by removing them completely. This strategy is usually successful. The surgical treatment options for gastrointestinal carcinoid tumors with local or distant spread are more complex. Because most carcinoid tumors grow very slowly and some do not cause any symptoms, attempts at complete surgical removal of metastatic carcinoid tumors may not always be necessary. In some patients, surgery to remove all visible cancer is the best option. This is particularly true if removing most of the cancer will reduce the hormone-like substances responsible for symptoms.

There are several operations that may be used to treat gastrointestinal carcinoid tumors. Some of these operations remove the primary tumor (in the organ the cancer started). Other operations are intended to remove or destroy metastases in other organs.

Local excision: This operation removes the primary tumor and some surrounding normal tissue. The edges of the defect are then sewn together. This usually doesn't cause any prolonged problem with eating or bowel movements. This operation is usually done for small carcinoid tumors (no larger than 2 centimeters, or about 3/4 inch). The most common example of this is when an appendectomy is done and the carcinoid tumor is discovered after the surgery. Most doctors believe that if the carcinoid tumor was small, less than 1 centimeter (1/2 inch), the appendectomy is curative. Local excision (surgical removal of the area around the tumor) of rectal carcinoid tumors may be done through the anus, without cutting the skin. Local excision of other gastrointestinal system carcinoid tumors can sometimes be done through an endoscope but usually is done through a skin incision.

Electrofulguration: This treatment destroys a tumor by heating it with electric current. It is sometimes used for small rectal carcinoid tumors.

More extensive excision: When the carcinoid tumor is larger than 2 centimeters, then most surgeons prefer to do a larger operation to make sure they remove the entire tumor. This also gives them the opportunity to see whether the cancer has invaded other tissues so they can get to these areas and remove the invading tumor.

- **Segmental colon resection or hemicolectomy:** This operation removes between 1/3 and 1/2 of the colon, as well as nearby blood vessels and lymph nodes.
- **Low anterior resection:** This operation is used for some tumors of the upper part of the rectum. It removes some of the rectum and the remaining ends are sewn together, without much impact on digestive function.
- **Abdominoperineal resection:** This surgery is for large or very invasive cancers of the lower part of the rectum. After this operation, the end of the colon is connected to the surface of the front of the abdomen and waste is eliminated from the body through this opening called a colostomy.
- **Liver resection:** This is an operation to remove one or a few metastases from the liver. It is not usually expected to cure the cancer but is often helpful in reducing symptoms of carcinoid syndrome.

Procedures to destroy liver metastases: These methods are often useful in destroying carcinoid metastases that have spread to the liver, especially if the number or location of the liver metastases makes surgical removal difficult or impossible. CT scan images are used to guide a needle precisely into the tumor deposits. The cells

are then destroyed by injecting concentrated alcohol through the needle, or liquid nitrogen can be used to cool the needle and kill the carcinoid cells by freezing. One new approach, called **radiofrequency ablation**, uses high-energy radio waves for treatment. A thin, needle-like probe temporarily placed into the tumor releases these radio waves. Placement of the probe is accurately guided by CT scans. The probe releases high frequency alternating current that destroys the cancer cells.

Liver transplantation: This is a rarely used treatment that may be effective for young patients with carcinoid tumors that have only spread to the liver. Although this is very difficult treatment for patients to go through, it can be curative and should be considered in young patients. For more information see the American Cancer Society document, "Liver Cancer".

Medical Treatments

Chemotherapy: Chemotherapy uses anti-cancer drugs that are injected into a vein or a muscle or taken by mouth to kill cancer cells. These drugs enter the bloodstream and reach all areas of the body (called **systemic treatment**), making this treatment useful for some types of cancers that have spread or metastasized to organs other than the one where they started growing. Unfortunately, carcinoid tumors are often not very sensitive to chemotherapy. Because of this, chemotherapy is generally used only for carcinoid tumors that have spread to other organs, are causing severe symptoms, and have not responded to other medications. Some of the chemotherapy drugs used in this situation include 5-fluorouracil (5-FU), doxorubicin, etoposide, dacarbazine, streptozotocin, cisplatin, and cyclophosphamide. Many cancers are treated with combinations of chemotherapy drugs. But in carcinoid tumors, using more than one drug has not been shown to be any more effective than using a single drug.

Chemotherapy drugs kill cancer cells but also damage some normal cells. Therefore, your doctors will pay careful attention to avoiding or minimizing side effects. These depend on the type of drugs, amount taken, and length of treatment. Temporary side effects might include nausea and vomiting, loss of appetite, loss of hair, and mouth sores. Because chemotherapy can damage the blood-producing cells of the bone marrow, you may have low blood cell counts. This can result in an increased risk 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 fatigue or shortness of breath (due to low red blood cell counts).

Some side effects disappear within a few days after treatment. In addition, there are medicines that can help prevent or minimize treatment side effects. For example, your doctor can prescribe drugs to help prevent or reduce nausea and vomiting.

Chemoembolization and intra-arterial therapy: When the carcinoid tumor has spread to the liver, it is sometimes treated by directly injecting the chemotherapy drug into the artery that supplies blood to the liver. This approach exposes the liver metastases to high doses of the drug and limits exposure of the rest of the body. This can avoid many of the side effects described above. Sometimes the chemotherapy drug is injected together with a material that plugs up the artery. When the arteries leading to them are blocked, the tumors become starved for nutrients and oxygen and many die off. This can be more effective when combined with chemotherapy. This combined approach is called **chemoembolization**.

For more information on chemotherapy, see the American Cancer Society document, "Understanding Chemotherapy: A Guide for Patients and Families."

Other drugs for treating carcinoid tumors: Several medications are available for controlling the symptoms of carcinoid syndrome (problems arising from the release of substances produced by some of these tumors and found through blood and urine tests) in patients with metastatic carcinoid tumors. Octreotide and lanreotide are

agents chemically related to a natural hormone, somatostatin. They are very helpful in treating the flushing, diarrhea, and wheezing from carcinoid syndrome. Although these drugs rarely shrink carcinoid tumors, they often slow or stop their growth. Although this is not curative, it can prolong life. The main side effects of these medications are pain at the site of the injection, and rarely, stomach cramps, nausea, vomiting, headaches, dizziness, and fatigue. These drugs have become available in long-acting injections that need to be given only once a month. These longer acting preparations may help patients more than the short acting ones. A recent study found that patients taking them lived longer than patients on the short acting preparations.

Interferons are naturally occurring substances that normally activate the body's immune system. They also slow the growth of tumor cells. Alpha-interferon is helpful in occasionally shrinking some metastatic carcinoid tumors, slowing the growth of many others, and improving symptoms of carcinoid syndrome. Its usefulness is sometimes limited by its flu-like side effects, which may be severe. The drug is given by injection.

An agent called **cyproheptadine** can also relieve symptoms. In addition, an experimental drug called SOM230 has been found to help people who were no longer being helped by octreotide.

Other medications are also available to control specific symptoms. Please ask your doctor about these, or describe your symptoms to your doctor and ask about medications to control them.

Radiation Therapy

Radiation therapy uses high-energy radiation to kill cancer cells. Although most cases of carcinoid tumor are cured by surgery alone, radiation therapy may be an option for those who cannot undergo surgery.

External-beam radiation therapy is the type of radiation used most often for most types of gastrointestinal cancer. It is like having a regular x-ray except it takes longer and involves much higher amounts of radiation. Patients typically have treatments for 5 days a week for several weeks. Unfortunately, radiation therapy often is not very effective against most gastrointestinal carcinoid tumors. It is used primarily to treat pain from carcinoid tumors that have spread to the bones or other parts of the body.

The main side effects of gastrointestinal radiation therapy are fatigue (tiredness), nausea, vomiting, diarrhea, and mild temporary, sunburn-like skin changes.

For more information on radiation therapy, see the American Cancer Society document, "Understanding Radiation Therapy: A Guide for Patients and Families."

Clinical Trials

The purpose of clinical trials: Studies of promising new or experimental treatments in patients are known as clinical trials. A clinical trial is only done when there is some reason to believe that the treatment being studied may be valuable to the patient. Treatments used in clinical trials are often found to have real benefits.

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

- Is the treatment helpful?
- How does this new type of treatment work?
- Does it work better than other treatments already available?
- What side effects does the treatment cause?
- Are the side effects greater or less than the standard treatment?
- Do the benefits outweigh the side effects?

- In which patients is the treatment most likely to be helpful?

Types of clinical trials: There are 3 phases of clinical trials in which a treatment is studied before it is eligible for approval by the FDA (Food and Drug Administration).

Phase I clinical trials: The purpose of a phase I study is to find the best way to give a new treatment and how much of it can be given safely. The cancer care team watches patients carefully for any harmful side effects. The treatment has been well tested in lab and animal studies, but the side effects in patients are not completely known. Doctors conducting the clinical trial start by giving very low doses of the drug to the first patients and increasing the dose for later groups of patients until side effects appear. Although doctors are hoping to help patients, the main purpose of a phase I study is to test the safety of the drug.

Phase II clinical trials: These studies are designed to see if the drug works. Patients are given the highest dose that doesn't cause severe side effects (determined from the phase I study) and closely observed for an effect on the cancer. The cancer care team also looks for side effects.

Phase III clinical trials: Phase III studies involve large numbers of patients -- often several hundred. One group (the control group) receives the standard (most accepted) treatment. The other group receives the new treatment. All patients in phase III studies are closely watched. The study will be stopped if the side effects of the new treatment are too severe or if one group has had much better results than the others.

If you are in a clinical trial, you will have a team of experts taking care of you and monitoring your progress very carefully. The study is especially designed to pay close attention to you.

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

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

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

- Is there a clinical trial for which I would be eligible?
- What is the purpose of the study?
- What kinds of tests and treatments does the study involve?
- What does this treatment do? Has it been used before?
- Will I know which treatment I receive?
- What is likely to happen in my case with, or without, this new treatment?
- What are my other choices and their advantages and disadvantages?
- How could the study affect my daily life?
- What side effects can I expect from the study? Can the side effects be controlled?

- Will I have to be hospitalized? If so, how often and for how long?
- Will the study cost me anything? Will any of the treatment be free?
- If I am harmed as a result of the research, what treatment would I be entitled to?
- What type of long-term follow-up care is part of the study?
- Has the treatment been used to treat other types of cancers?

The American Cancer Society offers a clinical trials matching service for patients, their family, and friends. You can reach this service at 1-800-303-5691 or on our Web site at <http://clinicaltrials.cancer.org>. Based on the information you provide about your cancer type, stage, and previous treatments, this service can compile a list of clinical trials that match your medical needs. In finding a center most convenient for you, the service can also take into account where you live and whether you are willing to travel.

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 or by visiting the NCI clinical trials Web site at www.cancer.gov/clinical_trials/.

Complementary and Alternative Therapies

Complementary and alternative therapies are a diverse group of health care practices, systems, and products that are not part of usual medical treatment. They may include products such as vitamins, herbs, or dietary supplements, or procedures such as acupuncture, massage, and a host of other types of treatment. There is a great deal of interest today in complementary and alternative treatments for cancer. Many are now being studied to find out if they are truly helpful to people with cancer.

You may hear about different treatments from family, friends, and others, which may be offered as a way to treat your cancer or to help you feel better. Some of these treatments are harmless in certain situations, while others have been shown to cause harm. Most of them are of unproven benefit.

The American Cancer Society defines **complementary** medicine or methods as those that are used along with your regular medical care. If these treatments are carefully managed, they may add to your comfort and well-being. **Alternative** medicines are defined as those that are used instead of your regular medical care. Some of them have been proven not to be useful or even to be harmful, but are still promoted as "cures." If you choose to use these alternatives, they may reduce your chance of fighting your cancer by delaying, replacing, or interfering with regular cancer treatment.

Before changing your treatment or adding any of these methods, discuss this openly with your doctor or nurse. Some methods can be safely used along with standard medical treatment. Others, however, can interfere with standard treatment or cause serious side effects. That is why it's important to talk with your doctor. More information about specific complementary and alternative therapies used for cancer is available through our toll-free number or on our Web site.

Treatment of Gastrointestinal Carcinoid Tumor by Stage

Localized Disease

Treatment of localized carcinoid tumors is based mostly on their size. Experts in this field occasionally disagree on the exact size cutoffs for making treatment decisions, and there are some "gray zones" of size where many

admit that it has not been determined exactly what treatment is best.

Stomach: Small carcinoid tumors of the stomach can often be completely removed through an endoscope. For larger tumors, particularly those larger than 1 centimeter (slightly less than 1/2 inch), removal of the tumor and some surrounding stomach tissue through an incision in the abdomen may be needed. In some patients, formation growth of stomach carcinoid tumors may be stimulated by gastrin, a hormone released by cells of the antrum of the stomach (the part next to the small intestine). In these patients, doctors may recommend removing the antrum of the stomach.

Small intestine: Local excision (surgical removal of the tumor and some surrounding normal tissue) is the usual treatment for carcinoid tumors occurring in the small intestine that are smaller than 1 centimeter (slightly over 3/8 inch). Surgery for larger tumors involves taking more surrounding normal-appearing intestine tissue, as well as some surrounding blood vessels and lymph nodes.

Large intestine (other than appendix and rectum): The usual treatment is local excision. If the carcinoid tumor is smaller than 1 centimeter, this procedure can often be done through a colonoscope. If the tumor is larger, surgery usually involves an incision through the skin.

Appendix: Nearly all cancer specialists agree that an appendectomy (surgical removal of the appendix) is the only treatment needed for carcinoid tumors of the appendix that are smaller than 1.5 centimeters (slightly larger than 1/2 inch).

For carcinoid tumors in the appendix that are between 1.5 and 2 centimeters, most doctors believe that removing the appendix is all that is needed. But they also consider other factors, such as the patient's age, general health, and the patient's degree of worry about the possibility of the cancer coming back -- to determine whether additional treatment is needed.

Most specialists agree that more extensive surgery should be considered for tumors larger than 2 centimeters (slightly over 3/4 inch). For these tumors over 2 centimeters, removal of about a third of the colon next to the appendix, along with nearby blood vessels and lymph nodes is a consideration for patients younger than 60 years of age who are otherwise in good health. Because carcinoid tumors grow and spread slowly, people older than age 60 or people with other serious health problems (especially if these problems make surgery more risky) are not likely to benefit from more extensive surgery.

Rectum: Rectal carcinoid tumors that are smaller than 1 centimeter (about 3/8 inch) are usually treated by fulguration (destroying the cancer by burning it with an electrical current). Carcinoid tumors larger than 2 centimeters (slightly over 3/4 inch) have a high risk of aggressive growth and spread, so they are removed by the same operations used for adenocarcinomas (the usual type of rectal cancer). This treatment involves low anterior resection if the carcinoid is in the upper area of the rectum. If the lower part is involved, abdominoperineal resection and colostomy are used. But this is a very complex area. Because many of these tumors will have already spread, it is not certain that such an extensive operation is worthwhile.

For rectal carcinoid tumors between 1 and 2 centimeters, the best approach is often determined by how deeply the carcinoid tumor invades the wall of the rectum, as well as other details of each patient's medical situation.

Deeply invasive tumors measuring 1 to 2 centimeters are often treated the same as larger tumors. Less invasive tumors measuring 1 to 2 centimeters often are treated the same as smaller tumors, by local excision (removing the cancer and a margin of normal rectal tissue). If local excision is used, careful follow-up to check for recurrence is needed.

Pancreas: Pancreatic islet cell carcinoma is potentially curable with surgery. Lesions that are in the distal two thirds of the pancreas are usually managed with a distal pancreatectomy. This operation removes the tail and a portion of the body of the pancreas. The spleen is usually removed as well. Occasionally, patients who have tumors in the head of the pancreas that are causing obstruction will require a **pancreaticoduodenectomy (Whipple procedure)**. This operation removes the head of the pancreas and sometimes removes the body of the pancreas as well. It also removes part of the stomach, the entire duodenum, a small part of the jejunum and lymph nodes near the pancreas. The gallbladder and part of the common bowel duct are removed and the remaining bile duct is attached to the small intestine so that bile from the liver can enter the small intestine.

Regional Spread

If possible, the primary tumor and areas of spread to adjacent tissues and lymph nodes should all be removed by surgery. If this is not possible, surgery should remove as much cancer as possible without causing severe side effects. Surgery should also relieve symptoms such as intestinal blockage caused by the local growth of cancer. For example, surgery to redirect the flow of feces around a blocked area of intestine can be done by connecting adjacent areas of the intestine.

Distant Spread

Surgery in this situation does not attempt to cure the cancer, since this is not possible. Rather, the goal is to relieve symptoms and slow the course of the disease. For example, removing or bypassing areas blocked by cancer growth can relieve some symptoms. If distant metastases are not causing symptoms, treatment may not be needed, although chemotherapy or immunotherapy (with interferon) may help delay the onset of symptoms in some patients. If carcinoid syndrome is causing bothersome symptoms, treatment options include chemotherapy, immunotherapy, treatment with octreotide or lanreotide, or removing the metastatic tumors by surgery. If metastatic tumors cannot be removed without causing severe side effects from removing essential organs and tissues, ablative methods (removal without surgery) are used to destroy as much of the tumor tissue as possible. These ablative methods, used mostly for liver metastases, include chemoembolization, radiofrequency ablation, cryosurgery, and alcohol injection. Patients should also be advised to avoid alcoholic drinks, stress, strenuous exercise, spicy foods, and certain medications that can make the symptoms of carcinoid syndrome worse.

Carcinoid Heart Disease

The substances released into the blood by carcinoid tumors can damage the heart. Early symptoms are fatigue and shortness of breath on exertion. Eventually, patients accumulate fluid in their legs and even their abdomen. The major problem is damage to the valves of the heart. Doctors can usually make the diagnosis by listening to the heart and by an ultrasound exam of the heart called an echocardiogram.

The main treatment is with octreotide to block the cancer's secretion of the toxic substances. Also, drugs to strengthen the heart beat and to get rid of fluid (diuretics) are helpful. In some instances, heart surgery is needed to replace the damaged valves.

More Treatment Information

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

The NCCN, made up of experts from 19 of the nation's leading cancer centers, develops cancer treatment guidelines for doctors to use when treating patients. Those are available on the NCCN Web site

(www.nccn.org).

The American Cancer Society collaborates with the NCCN to produce a version of some of these treatment guidelines, written specifically for patients and their families. These less-technical versions are available on both the NCCN Web site (www.nccn.org) and the ACS Web site (www.cancer.org). A print version can also be requested from the ACS at 1-800-ACS-2345.

The 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 Gastrointestinal Carcinoid Tumors?

It is important to have honest, open discussions with your cancer care team. They want to answer all of your questions, no matter how trivial you might think they are. For instance, consider these questions:

- What kind of carcinoid tumor do I have?
- What is the stage of my carcinoid tumor and what does that mean to me?
- What treatment choices do I have?
- What do you recommend and why?
- Based on what you've learned about my carcinoid tumor, what is my prognosis?
- What risks or side effects are there to the treatments you suggest?
- What are the chances of recurrence of my carcinoid tumor with these treatment plans?
- What should I do to be ready for treatment?

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

What Happens After Treatment for Gastrointestinal Carcinoid Tumors?

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. Even with no recurrences, people who have had cancer learn to live with uncertainty.

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.

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:

- 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
- 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 our Quitline 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 to 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 Gastrointestinal Carcinoid Tumor Research and Treatment?

There is always research going on in the field of gastrointestinal cancer. Scientists are looking for the causes of, ways to prevent, and novel new approaches to diagnose and treat gastrointestinal carcinoid tumors.

Diagnosis: Because the outlook and treatment of gastrointestinal carcinoid tumors and gastrointestinal carcinomas are very different, accurate diagnosis is important. Researchers have made great progress in developing tests that can detect specific substances found in the cells of carcinoid tumors but not gastrointestinal cancers. Other substances may be found in both carcinoid tumors and carcinomas but higher levels are found in one type. Most of these tests involve treating tissue samples with special antibodies produced in the laboratory. The antibodies are designed to recognize specific substances in certain types of tumors.

Imaging: Researchers are testing ^{111}In -DTPA-octreotide scintigraphy (octreoscan) and other nuclear medicine methods for the early detection of carcinoid tumors.

Treatment: New chemotherapy drugs are being tested to find drugs that are active against carcinoid tumors.

Other drugs are being developed to prevent the release of substances that are responsible for the symptoms of carcinoid syndrome. Also, there is work being done using highly radioactive MIBG and somatostatin type compounds to kill carcinoid tumors.

The drug, imatinib (Gleevec), which is effective in chronic myelogenous leukemia and gastrointestinal stromal tumors, can help some people with carcinoid. It is a pill with few side effects. New, more powerful versions of this drug are being developed.

Additional Resources

More Information From Your American Cancer Society

We have selected some related information that may also be helpful to you. These materials may be viewed on our Web site or ordered from our toll-free number, 1-800-ACS-2345.

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

Caring for the Patient With Cancer at Home (also available in Spanish)

Pain Control: A Guide for People With Cancer and Their Families (also available in Spanish)

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

Cancer in the Family: Helping Children Cope With a Parent's Illness

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 Cancer 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 include:

National Cancer Institute

Telephone: 1-800-4-CANCER

Internet Addresses: www.cancer.gov

The Carcinoid Cancer Foundation, Inc.

Telephone: 1-888-722-3132 or 1-914-683-1001

Internet Address: www.carcinoid.org

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

The American Cancer Society is happy to address almost any cancer-related topic. If you have any more questions, please call us at 1-800-ACS-2345 at any time, 24 hours a day.

References

- Caplin ME, Buscombe JR, Hilson AJ, Jones AL, Watkinson AF, Burroughs AK. Carcinoid tumor. *Lancet*. 1998; 352:799-805.
- Jenson RT, Doherty GM. Carcinoid tumors and the carcinoid syndrome. In: DeVita VT, Hellman S, Rosenberg SA, eds. *Cancer: Principles and Practice of Oncology*. Philadelphia, Pa: Lippincott Williams & Wilkins; 2005: 1559-1580.
- Kvols LK. Neoplasms of the diffuse endocrine system (Carcinoid tumors). In: Kufe DW, Pollock RE, Weichselbaum RR, Bast RC, Gansler TS, Holland JF, Frei E, eds. *Cancer Medicine*. 6th ed. Hamilton, Ont: BC Decker; 2003:1276-1283.
- Maggard MA, O'Connell JB, Ko, CY. Updated population-based review of carcinoid tumors. *Ann Surg*. 2004;240:117-122.
- Modlin IM, Lye KD, Kidd M. A 5-decade analysis of 13,715 carcinoid tumors. *Cancer* 2003; 97:934-59
- PDQ database. Gastrointestinal carcinoid tumor. Bethesda, Md: National Cancer Institute. Available at:<http://www.cancer.gov/cancerinfo/types/gastrointestinalcarcinoid/>. Accessed December 2004.

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For additional assistance please contact your American Cancer Society
1 · 800 · ACS-2345 or www.cancer.org