Treating Gastrointestinal Carcinoid Tumors

General approach to treatment

Once a gastrointestinal (GI) carcinoid tumor is found and staged, the cancer care team will discuss your treatment options with you. Depending on your situation, you may have different types of doctors on your treatment team. These doctors may include:

- A **surgeon**: a doctor who treats diseases with surgery.
- A **radiation oncologist**: a doctor who treats cancer with radiation therapy.
- A **medical oncologist**: a doctor who treats cancer with medicines such as chemotherapy.
- A **gastroenterologist**: a doctor who specializes in diagnosing and treating diseases of the digestive system.
- An **endocrinologist**: a doctor who specializes in the diagnosis and treatment of diseases related to hormones.

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

The main types of treatment for GI carcinoid tumors are:

- Surgery
- Chemotherapy and other drugs
- Radiation

In some cases, doctors may recommend combining more than one of these treatments (see the [most common approaches used based on the stage and primary site](#)).

It’s important to discuss all of your treatment options, including their goals and possible
side effects, with your doctors to help make the decision that best fits your needs. Some of the factors that might influence your treatment options are:

- The tumor size and location
- Whether it has spread to lymph nodes, liver, bones, or other organs
- Whether you have any other serious medical conditions
- Whether the tumor is causing bothersome symptoms

Take your time and think about all of your options when you make this important decision. If time allows, it’s often a good idea to get a second opinion. A second opinion may give you more information and help you feel more confident about the treatment plan you choose.

It’s also very important to ask questions if you’re not sure about something. You can find some good questions to ask in What should you ask your doctor about gastrointestinal carcinoid tumors?

Thinking about taking part in a clinical trial

Clinical trials are carefully controlled research studies that are done to get a closer look at promising new treatments or procedures. Clinical trials are one way to get state-of-the-art cancer treatment. In some cases they may be the only way to get access to newer treatments. They are also the best way for doctors to learn better methods to treat cancer. Still, they are not right for everyone.

If you would like to learn more about clinical trials that might be right for you, start by asking your doctor if your clinic or hospital conducts clinical trials. You can also call our clinical trials matching service at 1-800-303-5691 for a list of studies that meet your medical needs, or see the Clinical Trials section to learn more.

Considering complementary and alternative methods

You may hear about alternative or complementary methods that your doctor hasn’t mentioned to treat your cancer or relieve symptoms. These methods can include vitamins, herbs, and special diets, or other methods such as acupuncture or massage, to name a few.

Complementary methods refer to treatments that are used along with your regular medical care. Alternative treatments are used instead of a doctor’s medical treatment. Although some of these methods might be helpful in relieving symptoms or helping you feel better, many have not been proven to work. Some might even be dangerous.
Be sure to talk to your cancer care team about any method you are thinking about using. They can help you learn what is known (or not known) about the method, which can help you make an informed decision. See the Complementary and Alternative Medicine section to learn more.

Help getting through cancer treatment

Your cancer care team will be your first source of information and support, but there are other resources for help when you need it. Hospital- or clinic-based support services are an important part of your care. These might include nursing or social work services, financial aid, nutritional advice, rehab, or spiritual help.

The American Cancer Society also has programs and services – including rides to treatment, lodging, support groups, and more – to help you get through treatment. Call our National Cancer Information Center at 1-800-227-2345 and speak with one of our trained specialists on call 24 hours a day, every day.

The treatment information given here is not official policy of the American Cancer 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.

Surgery for Gastrointestinal Carcinoid Tumors

Most gastrointestinal (GI) carcinoid tumors can be cured by surgery alone. The type of operation will depend on a number of factors, including the size and location of the tumor, whether the person has any other serious diseases, and whether the tumor is causing the carcinoid syndrome.

Surgeons often try to cure localized carcinoid tumors by removing them completely, which is usually successful.

The options for GI carcinoid tumors that have spread locally or to distant parts of the body are more complex. Because most carcinoid tumors grow very slowly and some do not cause any symptoms, completely removing all metastatic carcinoid tumors may not always be needed. But 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 level of hormone-like substances causing symptoms.

Several types of operations can be used to treat GI carcinoid tumors. Some of these remove the primary tumor (where the cancer started), while others remove or destroy cancer that has spread (metastasized) to other organs.

**Fulguration (electrofulguration)**

This treatment destroys a tumor by heating it with electric current. It is sometimes used for small rectal carcinoid tumors, which can be reached fairly easily.

**Endoscopic mucosal resection**

In this procedure, the cancer is removed through an endoscope. This is most often used to treat small carcinoid tumors of the stomach and duodenum (the first part of the small intestine) and it can be used to remove small carcinoid tumors of the rectum.

**Local excision**

This operation removes the primary tumor and some normal tissue around it. The edges of the defect are then sewn together. This usually doesn’t cause any prolonged problem with eating or bowel movements. This operation may be done for small carcinoid tumors (no larger than 2 cm, or a little less than an inch).

Carcinoid tumors are sometimes removed during an operation for some other reason. This often occurs with carcinoid tumors of the appendix. When the appendix is removed (for some other reason), it is examined after surgery, and sometimes a carcinoid tumor is found. Most doctors believe that if the tumor is small — 2 cm or less — removing the appendix (appendectomy) is curative and no other surgery is needed. If the tumor is larger than 2 cm, more surgery may be needed.

Rectal carcinoid tumors may be excised (taken out) through the anus, without cutting the skin. Other GI carcinoid tumors can sometimes be locally excised through an endoscope but usually it is done through an incision (cut) in the skin.

**More extensive surgeries**

For larger tumors, a larger incision (cut) is needed to remove the tumor along with nearby tissues. This also gives the surgeon the chance to see whether the tumor has
grown into other tissues in the abdomen (belly). If it has, the surgeon may be able to remove the areas of cancer spread.

**Partial gastrectomy:** In this operation, part of the stomach is removed. If the upper part is removed, sometimes part of the esophagus is removed as well. If the lower part of the stomach is removed, sometimes the first part of the small intestine (the duodenum) is also taken. Nearby lymph nodes are also removed. This operation is also known as a *subtotal gastrectomy*.

**Small bowel (intestine) resection:** This is an operation to remove a piece of the small intestine (also called the *small bowel*). When it is used to treat a small bowel carcinoid, this surgery includes removing the tumor and some of the small bowel around it (called a *wide margin resection*), plus removing nearby (regional) lymph nodes and the supporting connective tissue (called the *mesentery*) that contains lymph nodes and vessels that carry blood to and from the intestine. Tumors in the terminal ileum (the last part of the small bowel) may require removing the right side of the colon (hemicolecotomy).

**Pancreaticoduodenectomy (Whipple procedure):** This operation is most often used to treat pancreatic cancer, but it is also used to treat cancers of the duodenum. It removes the duodenum, part of the pancreas, nearby lymph nodes and part of the stomach. The gallbladder and part of the common bile duct are removed and the remaining bile duct is attached to the small intestine so that bile from the liver can continue to enter the small intestine. This is a complex operation that requires a lot of skill and experience. It carries a relatively high risk of complications that could even be fatal.

**Segmental colon resection or hemicolecotomy:** This operation removes between and ½ of the colon, as well as the nearby mesentery (the layers of tissue that hold and connect the intestines), which includes blood vessels and lymph nodes.

**Low anterior resection:** This operation can be used for some tumors in the upper part of the rectum. It removes some of the rectum and the remaining ends are sewn together. This does not have much effect on digestive function.

**Abdominoperineal (AP) resection:** This surgery is done for large or very invasive cancers in the lower part of the rectum. It removes the anus, rectum, and lower part of the colon. After this operation, the end of the colon is connected to an opening on the skin on the abdomen (called a *colostomy*). A bag attached over this opening collects stool (feces) as it leaves the body. (For more information, see *Colostomy: A Guide*).

For more information on surgery for treating cancer, see *A Guide to Cancer Surgery*. 
Surgery for carcinoid tumors that have spread to the liver

The liver is a common site of spread for GI carcinoid tumors. Treating cancer that has spread to the liver can help with symptoms and can even help some people live longer.

Liver resection

In this operation, one or more pieces of the liver that contain areas of cancer are removed. If it isn’t possible to remove all areas of cancer, surgery may still be done to remove as much tumor as is possible to help reduce symptoms of carcinoid syndrome. This is sometimes called cytoreductive surgery. Removing liver metastases may help some people with carcinoid tumors live longer, but most people who have this surgery will eventually develop new liver metastases.

Liver transplant

This operation removes the patient’s liver and puts a liver (or a piece of a liver) from someone else in its place. It can be used to treat some neuroendocrine cancers that have only spread to the liver, after the primary tumor has been removed. A liver transplant is generally only an option for people who are young and otherwise healthy. Although this is a very difficult treatment to go through, it can be curative. For more information on liver transplants see Liver Cancer.

- References

See all references for Gastrointestinal Carcinoid Tumor

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Chemotherapy and Other Drugs for Gastrointestinal Carcinoid Tumors

Chemotherapy (chemo) 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, making this treatment useful for some types of cancers that have...
spread to other organs.

Unfortunately, gastrointestinal (GI) carcinoid tumors often do not respond well to chemo. Because of this, chemo generally is used only for tumors that have spread to other organs, are causing severe symptoms, and have not responded to other medicines.

Some of the chemo drugs used to treat GI carcinoid tumors include:

- Capecitabine (Xeloda)
- 5-fluorouracil (5-FU)
- Doxorubicin (Adriamycin)
- Etoposide (VP-16)
- Dacarbazine (DTIC)
- Streptozocin
- Temozolomide
- Cisplatin
- Cyclophosphamide (Cytoxan)

Some tumors, especially high-grade tumors, may be treated with more than one drug. For these, combinations of etoposide plus carboplatin or temozolomide plus capecitabine may be used.

Chemo drugs kill cancer cells but also damage some normal cells, which can cause some side effects. Side effects depend on the type of drugs, the amount taken, and the length of treatment. Short-term side effects might include:

- Nausea and vomiting
- Loss of appetite
- Hair loss
- Mouth sores
- Low blood counts

Because chemo can damage the blood-making cells of the bone marrow, you may have low blood cell counts. This can result in:

- Increased risk of infection (from too few white blood cells)
- Bleeding or bruising after minor cuts or injuries (from a shortage of blood platelets)
- Fatigue or shortness of breath (from too few red blood cells)

Most side effects go away a short time after treatment. Often medicines can help prevent or minimize many of the side effects. For example, your doctor can prescribe drugs to help prevent or reduce nausea and vomiting.
Intra-arterial therapy and chemoembolization

Normally, chemo drugs enter the bloodstream and can travel throughout the body. When carcinoid cancer has spread to the liver, it is sometimes treated by directly injecting the chemo drug into the hepatic artery, which supplies blood to parts of the liver. This exposes the liver tumors to high doses of the drug but limits exposing the rest of the body. This lets patients avoid some side effects. Sometimes the chemo drug is injected together with a material that plugs up the artery (an approach called chemoembolization). When the arteries leading to them are blocked, the tumors become starved for nutrients and oxygen and many die off.

For more information on chemo, see Chemotherapy.

Other drugs for treating carcinoid tumors

Somatostatin analogs are man-made drugs chemically related to the natural hormone, somatostatin. These drugs can help control the symptoms caused by the hormones released from carcinoid tumors, including carcinoid syndrome. They can also help slow or stop tumor growth for a time in patients with metastatic neuroendocrine cancers.

The main somatostatin analogs used to treat carcinoid tumors are:

- Octreotide (Sandostatin)
- Lanreotide (Somatuline Depot)

These drugs can help treat flushing, diarrhea, and wheezing from carcinoid syndrome. They can also help slow or stop the growth of carcinoid tumors that have spread, which might help prolong life. Octreotide is also sometimes given to prevent and treat problems caused by the release of serotonin or other hormones when patients with carcinoid tumors have surgery.

The main side effects of these drugs are pain at the site of the injection and rarely stomach cramps, nausea, vomiting, headaches, dizziness, and fatigue. In patients that don’t have carcinoid syndrome, they can also cause diarrhea. These drugs can also cause sludging (thickening or crystallizing) of bile in the gallbladder, which can lead to gallstones. They can also raise blood sugar levels and make pre-existing diabetes more difficult to control.

Octreotide comes in a short-acting version that is given 2 to 4 times a day. It is also available as a long-acting injection (Sandostatin LAR) given only once a month, which may help patients more than the short-acting version.
Lanreotide only comes as a monthly injection.

A newer somatostatin analog, pasireotide (Signifor), is more often used to treat the symptoms of Cushing syndrome.

**Interferons** are natural substances that normally activate the body's immune system. They also slow the growth of tumor cells. Interferon-alfa is sometimes helpful in shrinking or slowing the growth of metastatic neuroendocrine cancers 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.

**Cyproheptadine** is an antihistamine that can help relieve some of the symptoms of carcinoid syndrome.

Other medicines are also available to control specific symptoms. For example, **telotristat (Xermelo)** can be used along with a somatostatin analog to help control carcinoid syndrome diarrhea.

Be sure to describe your symptoms to your doctor and ask about medicines to control them.

- References
  See all references for Gastrointestinal Carcinoid Tumor

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**Radiation Therapy for Gastrointestinal Carcinoid Tumors**

**External beam radiation therapy (EBRT)**

In this form of radiation therapy, a machine aims a beam of high-energy rays (or particles) to kill cancer cells. It is like having a regular x-ray except it takes longer and uses much higher amounts of radiation. Patients typically have treatments 5 days a
week for several weeks.

This is the type of radiation used most often to treat cancer. Unfortunately, EBRT is not very effective against most gastrointestinal (GI) carcinoid tumors. It is used mainly to treat pain from cancers that have spread to the bones or other parts of the body. Although surgery is the first option for most carcinoid tumors, those who can't have surgery might choose radiation therapy.

The main side effects of GI radiation therapy are:

- Tiredness (fatigue)
- Nausea and vomiting
- Diarrhea (if the belly or pelvis is treated)
- Skin changes, which can range from mild redness to blistering and peeling
- Hair loss in the area being treated

**Radioembolization**

This technique combines embolization with radiation therapy and is used to treat liver metastases. Embolization reduces blood flow to a tumor by injecting materials that plug up the artery feeding the area of the liver containing the tumor. This artery that is blocked is a branch of the hepatic artery, the artery that feeds the liver. Most of the healthy liver cells will not be affected because they get their blood supply from another blood vessel (the portal vein).

In this procedure a catheter is put into an artery in the inner thigh and threaded up into the liver. A dye is usually injected into the bloodstream at the same time to allow the doctor to monitor the path of the catheter via angiography, a special type of x-ray. Once the catheter is in place, small particles called microspheres are injected into the artery to plug it up.

In radioembolization, microspheres that are attached to a radioactive element called yttrium-90 (or 90Y) are used (some brand names for these beads include TheraSphere® and SIR-Spheres®). After they are injected, the beads travel in the liver blood vessels until they get stuck in small blood vessels near the tumor. There they give off radioactivity for a short while, killing tumor cells. The radiation travels a very short distance, so its effects are limited mainly to the tumor.

**Radiopharmaceuticals**
In this form of radiation therapy, a drug is linked to a radioactive element. The drug travels throughout the body, attaches to the cancer cells, and gives off radiation to kill them. This type of treatment has been used for a long time to treat some cancers, but is now also being used to treat carcinoid tumors. One option is to use I-131 MIBG in higher doses than are normally used to image carcinoid tumors (see “Imaging tests” in the section “How are gastrointestinal carcinoid tumors diagnosed?” Treatment using I-131 MIBG is available in Europe, but is not available in the United States.

Another being studied is a drug like octreotide called edotreotide linked with a radioactive form of the element yttrium. This is discussed in more detail in “What's new in gastrointestinal carcinoid tumor research and treatment?”

For more information on radiation therapy, see the Radiation Therapy section of our website, or read Understanding Radiation Therapy: A Guide for Patients and Families.

- References
  See all references for Gastrointestinal Carcinoid Tumor

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Treatment of Gastrointestinal Carcinoid Tumors by Stage

Localized disease

A tumor is localized when it is found only in the organ where it started. Treatment of localized carcinoid tumors is based mostly on their size. Experts sometimes disagree on the exact size for making treatment decisions, and there are some sizes where it has not been determined exactly what treatment is best.

Stomach

Carcinoid tumors of the stomach are grouped by the level of a hormone called gastrin
and the amount of acid in the stomach (measured before surgery). Certain conditions linked to high gastrin levels are also linked to getting many carcinoid tumors of the stomach. Patients with these conditions who have stomach carcinoid tumors are treated differently from patients without these conditions.

**Type 1:** Patients with high gastrin levels but low levels of stomach acid are said to have type 1 tumors. These carcinoid tumors tend to be small, but there may be many of them. The tumors don’t tend to grow into the deeper layers of the stomach or spread to other organs or tissues, and are often treated by removing them completely through an endoscope. The other option is to watch the tumors closely (by endoscopy), and only removing them if they start growing.

**Type 2:** Patients who have high gastrin levels and high stomach acid levels are said to have type 2 tumors. Like type 1 tumors they are often small and there may be more than one at a time. Type 2 tumors also don’t tend to grow into deeper layers of the stomach or spread to other organs. Small tumors can be watched closely without treatment, removed with endoscopy, or treated with a medicine like octreotide (Sandostatin) or lanreotide (Somatuline) that will lower both gastrin and stomach acid. For tumors larger than 2 cm (slightly less than an inch), just watching the tumor closely isn’t usually an option. These tumors need to be removed, either through an endoscope or in a regular operation through an incision in the abdomen that removes the tumor and some surrounding stomach tissue.

**Type 3:** Patients who don’t have high gastrin levels (and have normal levels of stomach acid) are said to have type 3 tumors. There is usually only one tumor, and the tumor tends to grow into deeper layers of the stomach or even spread to nearby lymph nodes or other organs (like the liver). If the tumor is small, endoscopic surgery may still be an option. More, often, though, more extensive surgery is needed: A piece of the stomach is removed (a partial gastrectomy) along with nearby lymph nodes.

**Small intestine**

Some small tumors in the duodenum (the first part of the small intestine) can often be removed through the endoscope (endoscopic resection). Depending on the size of the tumor and whether it is growing into nearby tissues, other options include surgery to remove the tumor (local excision), removing all or part of the duodenum with nearby lymph nodes, and removing the duodenum and part of the pancreas (a pancreatoduodenectomy).

For tumors in other parts of the small intestine, treatment is either local excision for small tumors or small bowel resection (removal of a piece of intestine as well as some
surrounding blood vessels and lymph nodes) for larger tumors.

**Large intestine (other than appendix and rectum)**

The usual treatment is hemicolecotmy (removal of a section of colon along with nearby lymph nodes and blood vessels). Because many patients have more than one carcinoid tumor, the surgeon will often check the rest of the colon for other tumors during surgery. For very small tumors, sometimes the tumor can be removed without surgery using a colonoscope.

**Appendix**

Most often, an appendectomy (surgical removal of the appendix) is the only treatment needed for carcinoid tumors that are 2 cm (a little less than an inch) across or smaller. Still, other factors, such as the way the tumor cells look under the microscope, the patient's age, general health, and the patient's degree of worry about the possibility of the cancer coming back, might also be used to determine whether more treatment is needed.

Tumors larger than 2 cm are more likely to have already spread to nearby tissues and lymph nodes, so more extensive surgery is usually recommended for these tumors. This means removal of about a third of the colon next to the appendix (a hemicolecotmy), along with nearby blood vessels and lymph nodes. This procedure might not be recommended for people who are older or have other serious health problems (especially if these problems make surgery more risky), because the benefit might not outweigh the risks.

**Rectum**

Doctors check how deep the cancer is growing into the wall of the rectum before surgery by using endoscopic ultrasound (discussed in the section about diagnosis). Most rectal carcinoid tumors that are smaller than 2 cm (slightly less than an inch) can be removed through an endoscope or local excision through the anus.

The best approach for rectal carcinoid tumors between 1 and 2 cm, depends on how deeply the tumor has grown into the wall of the rectum, as well as other details of each patient's medical situation. If the tumor has grown into the thick muscle layer of the rectum (the muscularis propria) or deeper, it needs to be treated the same as a larger tumor.
Carcinoid tumors larger than 2 cm (and those that have grown deep into the wall of the rectum) have a higher risk of growing and spreading, so they are removed by the same operations used for adenocarcinomas (the usual type of rectal cancer). This operation is a low anterior resection if the carcinoid is in the upper part of the rectum. If the lower part is involved, abdominoperineal (AP) resection and colostomy are used.

**Regional spread**

Regional spread means that the cancer has either spread to nearby lymph nodes or it has grown through the wall of the organ where it started and has invaded nearby tissues such as fat, ligaments, and muscle.

If possible, the primary (main) tumor and any areas of cancer spread should all be removed by surgery. Nearby lymph nodes should be removed and checked for signs of cancer spread. This provides the best chance of cure. If this can't be done, surgery should remove as much cancer as possible without causing severe side effects. Surgery should also be done to relieve symptoms such as intestinal blockage caused by the local growth of the tumor.

**Distant spread**

At this stage, a cure is not usually possible, although treatment is not always needed right away, depending on how quickly the tumors are growing. The goal of surgery in this situation is usually 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 interferon-alfa may help delay symptoms in some patients. If the cancer has spread to the liver, even when it isn’t causing symptoms, some doctors recommend treatment with octreotide or lanreotide because it may slow tumor growth.

If carcinoid syndrome is causing bothersome symptoms, treatment options include chemotherapy, immunotherapy, treatment with octreotide or lanreotide, or surgery to remove the metastatic tumors. If metastatic tumors in the liver cannot be removed by surgery without causing severe side effects, ablation or embolization can be used to destroy as much of the tumors as possible. Patients should also be advised to avoid alcoholic drinks, stress, strenuous exercise, spicy foods, and certain medicines that can make the symptoms of carcinoid syndrome worse.
Recurrent carcinoid tumors

When cancer come backs after treatment it is called a recurrence. Recurrence can be local (in or near the same place it started) or distant (spread to organs such as the lungs or bone). Patients with recurrent carcinoid tumors are treated with surgery to remove all signs of tumor whenever possible. This provides the best chance for a good long-term outcome. If surgery is not possible, the treatments used for distant spread may be helpful.

Carcinoid heart disease

The substances released into the blood by some carcinoid tumors can damage the heart. Early symptoms are fatigue and shortness of breath. Eventually, patients get fluid in their legs and even their abdomen. The major cause 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 a somatostatin analog like octreotide or lanreotide to block the cancer’s secretion of the toxic substances. Drugs (diuretics) to strengthen the heartbeat and to get rid of fluid can also help. In some instances, heart surgery may be needed to replace the damaged valves.

References
See all references for Gastrointestinal Carcinoid Tumor

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