Islet Cell Tumor - Treatment Options [1]

This section has been reviewed and approved by the Cancer.Net Editorial Board [2], 06/2015

ON THIS PAGE: You will learn about the different ways doctors use to treat people with this type of tumor. To see other pages, use the menu on the side of your screen.

This section outlines treatments that are the standard of care (the best known treatments available) for an islet cell tumor. When making treatment plan decisions, patients are also encouraged to consider clinical trials as an option. A clinical trial is a research study to test a new approach to treatment to evaluate whether it is safe, effective, and possibly better than the standard treatment. Clinical trials may test such approaches as a new drug, a new combination of standard treatments, or new doses of current therapies. Your doctor can help you review all treatment options. For more information, see the About Clinical Trials [3] and Latest Research [4] sections.

Treatment overview

In caring for a person with a tumor, different types of doctors often work together to create a patient?s overall treatment plan that combines different types of treatments. This is called a multidisciplinary team [5]. Care teams also include a variety of other health care professionals, including physician assistants, oncology nurses, social workers, pharmacists, counselors, dietitians, and others.

Descriptions of the most common treatment options for an islet cell tumor are listed below. Treatment options and recommendations depend on several factors, including the type, grade, and stage of the tumor, possible side effects, and the patient?s preferences and overall health. Your care plan may also include treatment for symptoms and side effects, an important part of cancer care. Take time to learn about all of your treatment options and be sure to ask questions about things that are unclear. Also, talk about the goals of each treatment with your doctor and what you can expect while receiving the treatment. Learn more about making treatment decisions [6].

Active surveillance

Sometimes active surveillance, which is also called watchful waiting or watch-and-wait, may be recommended. This is because many islet cell tumors are slow growing and may not grow, spread, or cause problems for many months or years. With this approach, the tumor is monitored closely with regular imaging tests, usually CT scans or sometimes MRI (see the Diagnosis [7] section); blood tests; and physical exams. Active treatment usually only begins if the tumor shows signs of growing and/or spreading.

Surgery

Surgery is the most common treatment for an islet cell tumor that is localized, meaning it has not spread outside the pancreas. Surgery is the removal of the tumor and some surrounding healthy tissue during an operation. During surgery, the doctor may need to remove most or part of the pancreas, depending on the location and size of the tumor. A surgical oncologist is a doctor who specializes in treating cancer using surgery.

Common surgical procedures that may be used to treat an islet cell tumor include:

- **Enucleation.** During this surgery, only the tumor is removed.
- **Whipple procedure.** During this procedure, the surgeon removes the head of the pancreas and part of the small intestine, bile duct, and stomach, and then reconnects the digestive tract and biliary system.
- **Distal pancreatectomy.** If the cancer is located in the tail of the pancreas, the most common operation is a distal pancreatectomy. During this operation, the surgeon removes the tail and body of the pancreas, as well as the spleen.
- **Splenectomy.** This is the removal of the spleen.
- **Gastrectomy.** For a gastrinoma, it may also be necessary to take out the stomach to remove ulcers, a procedure called a gastrectomy.

Side effects of surgery include weakness, fatigue, and pain for the first few days following the procedure. The doctor may prescribe medication to help manage these side effects. The patient will need to stay in the hospital for several days and will probably need to rest at home for about one month. Learn more about the basics of cancer surgery [8].

If all or part of the pancreas was removed during surgery, it may be difficult to digest food. A special diet and medicine may help. Also, the doctor can prescribe hormones and enzymes to replace those lost by the removal of the pancreas. Diabetes is another side effect of pancreas removal due to the loss of insulin, which is made by the pancreas. For this, the doctor can prescribe insulin.

Hormone therapy

Hormone therapy may be given to relieve symptoms caused by the tumor (see the Symptoms and Signs [9] section). The most common hormone used is octreotide (Sandostatin, OncoLAR), which is similar to one of the hormones normally produced by the body, somatostatin.

Many islet cell tumors have receptors for somatostatin on the surface of their cells. When octreotide attaches to these receptors, it blocks the release of hormones by the tumor and also can help stop the tumor from growing.

Octreotide can be given either as daily injections under the skin or as monthly injections into a muscle, called Sandostatin LAR Depot. Common side effects of octreotide include gallstones and/or biliary sludge and gastrointestinal problems, such as diarrhea and abdominal discomfort.

Another hormone therapy, lanreotide (Somatuline), was also recently approved for the treatment of islet cell tumors. Similar to octreotide, it is also a somatostatin analogue.

Targeted therapy

Targeted therapy is a treatment that targets the tumor?s specific genes, proteins, or the tissue environment that contributes to a tumor's growth and survival. This type of treatment blocks the growth and spread of tumor cells while limiting damage to healthy cells. Targeted therapies still have a number of side effects, but they are different from those caused by traditional chemotherapy (see below).

Recent studies show that not all tumors have the same targets. To find the most effective treatment, your doctor may run tests to identify the genes, proteins, and other factors in your tumor. This helps doctors better match each patient with the most effective treatment whenever possible. In addition, many research studies are taking place now to find out more about specific molecular targets and new treatments directed at them. Learn more about the basics of targeted treatments [10].

In 2011, the U.S. Food and Drug Administration (FDA) approved two targeted treatments for locally advanced or metastatic islet cell tumors. These treatments have been shown to significantly delay the growth of islet cell tumors. Both drugs are taken orally each day.

- **Everolimus (Afinitor).** This drug targets a protein called mTOR that is important for cell growth and survival. Common side effects associated with everolimus include mouth sores, fatigue, diarrhea, and rash.
- **Sunitinib (Sutent).** This drug?s main target is the receptor of a protein called VEGF. VEGF is important in angiogenesis, the process of making new blood vessels. Because a tumor needs the nutrients delivered by blood vessels to grow and spread, the goal of anti-angiogenesis therapies like sunitinib is to ?starve? the tumor. Common side effects associated with this drug include diarrhea, nausea/vomiting, fatigue, and high blood pressure.

Chemotherapy

Chemotherapy is the use of drugs to destroy tumor cells, usually by stopping the cells? ability to grow and divide. Chemotherapy is given by a medical oncologist, a doctor who specializes in treating tumors with medication. Generally, chemotherapy is used to treat islet cell tumors that are moderately to poorly differentiated, quite large, or are no longer responding to hormonal or targeted therapies.

Systemic chemotherapy gets into the bloodstream to reach cancer cells throughout the body. Common ways to give chemotherapy include an intravenous (IV) tube placed into a vein using a needle or in a pill or capsule that is swallowed (orally).

A chemotherapy regimen (schedule) usually consists of a specific number of cycles given over a set period of time. A patient may receive one drug at a time or combinations of different drugs at the same time. Common drug combinations used for islet cell tumors include capecitabine (Xeloda) and temozolomide (Methazolastone,

Temodar), which are given orally, or either carboplatin (Paraplatin) or cisplatin (Platinol) combined with etoposide (Toposar, VePesid), which is given intravenously.

The side effects of chemotherapy depend on the individual and the dose used, but they can include fatigue, risk of infection, nausea and vomiting, hair loss, loss of appetite, and diarrhea. These side effects usually go away once treatment is finished.

Learn more about the basics of chemotherapy [11] and preparing for treatment [12]. The medications used to treat cancer are continually being evaluated. Talking with your doctor is often the best way to learn about the medications prescribed for you, their purpose, and their potential side effects or interactions with other medications. Learn more about your prescriptions by using these searchable drug databases [13].

Immunotherapy

Immunotherapy, also called biologic therapy, is designed to boost the body's natural defenses to fight the tumor. It uses materials made either by the body or in a laboratory to improve, target, or restore immune system function.

Alpha interferon is a type of immunotherapy given as injections under the skin. It is sometimes used to help relieve symptoms caused by an islet cell tumor. However, it can have significant side effects including fatigue, depression, and flu-like symptoms. Alpha interferon is not used as frequently nowadays because of the development of other more effective therapies.

Learn more about the basics of immunotherapy [14].

Hepatic artery embolization

Generally used for patients with cancer that has spread to the liver, hepatic artery embolization helps stop cancer growth and spread by temporarily sealing off the blood vessels that supply oxygen and nutrients to the tumor(s). These are typically branches of the hepatic artery. Whether this procedure is recommended for a patient depends on the size, number, and location of the tumor(s) in the liver

Sometimes this procedure also involves delivering chemotherapy directly into the tumor before blocking the blood supply to that portion of the liver. The drug(s) used during this procedure are either mixed with an oil called lipiodol or loaded into beads/spheres. This is called chemoembolization or transarterial chemoembolization (TACE). Alternatively, radioactively labeled microbeads, such as TheraSpheres or SIR-Spheres, can be delivered during this procedure. This is known as radioembolization or selective internal radiation therapy (SIRT).

These procedures are usually performed by an experienced interventional radiologist and may require an inpatient hospital stay. Side effects include pain around the liver, fever, and temporarily higher levels of liver enzymes as measured by blood tests.

Getting care for symptoms and side effects

A tumor and its treatment often cause side effects. In addition to treatment to slow, stop, or eliminate the tumor, an important part of care is relieving a person?s symptoms and side effects. This approach is called palliative or supportive care, and it includes supporting the patient with his or her physical, emotional, and social needs.

Palliative care is any treatment that focuses on reducing symptoms, improving quality of life, and supporting patients and their families. Any person, regardless of age or type and stage of cancer, may receive palliative

care. It works best when palliative care is started as early as needed in the cancer treatment process.

People often receive treatment for the tumor and treatment to ease side effects at the same time. In fact, patients who receive both often have less severe symptoms, better quality of life, and report they are more satisfied with treatment.

Palliative treatments vary widely and often include medication, nutritional changes, relaxation techniques, emotional support, and other therapies. You may also receive palliative treatments similar to those meant to eliminate the cancer, such as chemotherapy or surgery. Talk with your doctor about the goals of each treatment in your treatment plan.

Before treatment begins, talk with your health care team about the possible side effects of your specific treatment plan and palliative care options. And during and after treatment, be sure to tell your doctor or another health care team member if you are experiencing a problem so it can be addressed as quickly as possible. Learn more about palliative care [15].

Progressive islet cell tumor

Progressive disease is when the tumor becomes larger or spreads, usually while the patient is having treatment. Patients with this diagnosis are encouraged to talk with doctors who are experienced in treating this stage of disease because there can be different opinions about the best treatment plan. Learn more about getting a second opinion [16] before starting treatment so you are comfortable with the treatment plan chosen. This discussion may include clinical trials [3].

Your health care team may recommend a treatment plan that includes a combination of surgery, hormone therapy, chemotherapy, immunotherapy, or targeted therapy. As noted above, hepatic artery occlusion or embolization may be used if cancer has spread to the liver. Palliative care will also be important to help relieve symptoms and side effects.

For most patients, a diagnosis of progressive disease is very stressful and, at times, difficult to bear. Patients and their families are encouraged to talk about the way they are feeling with doctors, nurses, social workers, or other members of the health care team. It may also be helpful to talk with other patients, including through a support group.

Remission and the chance of recurrence

A remission is when a tumor cannot be detected in the body and there are no symptoms. This may also be called having ?no evidence of disease? or NED.

A remission may be temporary or permanent. This uncertainty causes many people to worry that the tumor will come back. While many remissions are permanent, it?s important to talk with your doctor about the possibility of the tumor returning. Understanding your risk of recurrence and the treatment options may help you feel more prepared if it does return. Learn more about coping with the fear of recurrence [17].

If an islet cell tumor does return after the original treatment, it is called a recurrent tumor. It may come back in the same place (called a local recurrence), nearby (regional recurrence), or in another place (distant recurrence).

When this occurs, a cycle of testing will begin again to learn as much as possible about the recurrence. After testing is done, you and your doctor will talk about your treatment options. Often the treatment plan will include the treatments described above, such as surgery, chemotherapy, immunotherapy, and targeted therapy, but they may be used in a different combination or given at a different pace. Your doctor may also suggest clinical trials

that are studying new ways to treat this type of recurrent tumor. Whichever treatment plan you choose, palliative care will be important for relieving symptoms and side effects.

People with a recurrent tumor often experience emotions such as disbelief or fear. Patients are encouraged to talk with their health care team about these feelings and ask about support services to help them cope. Learn more about dealing with recurrence [18].

If treatment fails

Recovery from an islet cell tumor is not always possible. If the tumor cannot be cured or controlled, the disease may be called advanced or terminal.

This diagnosis is stressful, and advanced disease is difficult to discuss for many people. However, it is important to have open and honest conversations with your doctor and health care team to express your feelings, preferences, and concerns. The health care team is there to help, and many team members have special skills, experience, and knowledge to support patients and their families. Making sure a person is physically comfortable and free from pain is extremely important.

Patients who have advanced disease and who are expected to live less than six months may want to consider a type of palliative care called hospice care. Hospice care is designed to provide the best possible quality of life for people who are near the end of life. You and your family are encouraged to think about where you would be most comfortable: at home, in the hospital, or in a hospice environment. Nursing care and special equipment can make staying at home a workable alternative for many families. Learn more about advanced cancer care planning [19].

After the death of a loved one, many people need support to help them cope with the loss. Learn more about grief and loss [20].

The next section in this guide is About Clinical Trials [3]*, and it offers more information about research studies that are focused on finding better ways to care for people with islet cell tumors. Or, use the menu on the side of your screen to choose another section to continue reading this guide.*

Links:
[1] http://www.cancer.net/cancer-types/islet-cell-tumor/treatment-options
[2] http://www.cancer.net/about-us
[3] http://www.cancer.net/node/18949
[4] http://www.cancer.net/node/18952
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