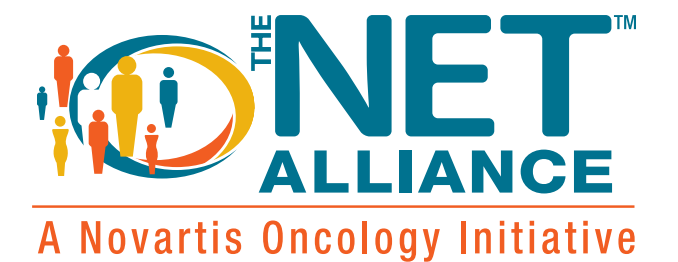
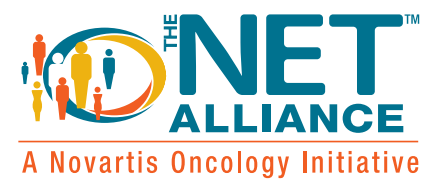




# Understanding Your NET

A patient's guide to living with a neuroendocrine tumor



## Stay informed, and stay healthy

*Understanding Your NET* is an educational brochure for patients and their loved ones affected by neuroendocrine tumors (NETs).

The brochure contains 8 sections covering a range of topics, including where a NET might originate, different types of NETs, symptoms of NETs, managing your condition, and communicating effectively with your healthcare team. These sections are tabbed so you can find the information you need easily.

Important medical terms are underlined throughout the brochure, and the definitions for these terms are included in the glossary in Section 9.

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We hope this brochure will help you stay informed about your condition and engaged with your healthcare team to ensure that you receive the best possible care.



# What is the neuroendocrine system?

## Major glands of the endocrine system

The neuroendocrine system is made up of the nervous system and the endocrine system. They work together to keep your body functioning regularly.

- Your brain sends signals to your nervous system that control various body functions
- Some messages are sent to the glands of the endocrine system. These include the pancreas, hypothalamus, thyroid and parathyroid, pituitary gland, adrenal glands, ovaries, and testes (see figure at right)
- These glands release hormones that control important body functions, such as growth, metabolism, reproduction, and response to stress or injury

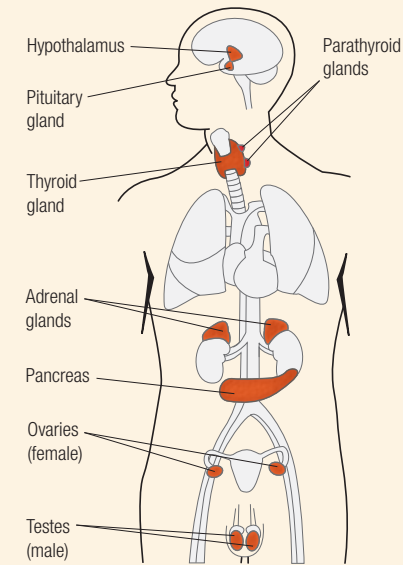


Figure reprinted from The Merck Manuals Online Medical Library.

## How does the neuroendocrine system work?

Your nervous system and endocrine system are made up of neuroendocrine cells.

Neuroendocrine cells are located throughout your body.

They release hormones that communicate messages to certain cells that are able to receive the messages.

- For example, when you need to digest food, your brain signals a hormone called serotonin to be released. Serotonin activates the muscles in the digestive tract

Other important hormones you may have heard about include insulin, adrenaline, estrogen, and testosterone.

Like other types of cells in your body, neuroendocrine cells can sometimes develop into cancer. This can lead to serious medical conditions, including neuroendocrine tumors (NETs).

## How do cells become tumors?

Cells that make up the tissue and organs in your body continually age, die, and are replaced by new cells.

This process normally occurs in 3 stages:

- Stage 1. Growth** – cells divide and multiply
- Stage 2. Differentiation** – cells change from unspecialized to specialized types of cells
- Stage 3. Apoptosis** – cells die normally

If this process malfunctions, cells can grow uncontrollably and may form tumors.

## Are all tumors cancerous?

Tumors can be benign or malignant (see figure below).

- Benign tumors are not considered to be cancer. They generally
  - are harmless and slow growing
  - are not very different from surrounding tissue
  - don't spread to nearby tissue
- Malignant tumors are considered to be cancer. They generally
  - grow uncontrollably
  - are not programmed to die normally
  - have a different structure than the cells in surrounding tissue
  - may invade nearby tissue or even spread (metastasize) to distant parts of the body

## What are some of the characteristics of NETs?

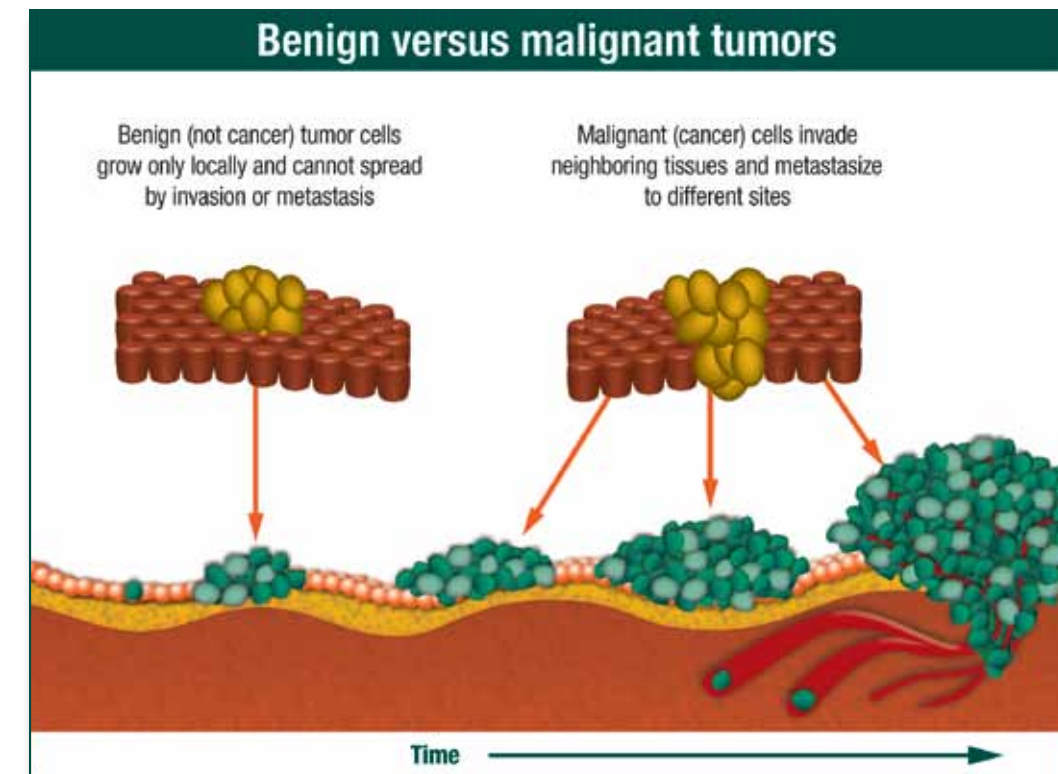
NETs vary in size and rate of growth, and they are generally considered to be malignant.

NETs originate from neuroendocrine cells throughout the body. They can spread to other parts of the body, most often to the liver or bone.

By the time they are diagnosed, more than 50% of NETs have already spread to other parts of the body. There are 3 main reasons for this:

- Some NETs are hard to find until they have grown or spread
- Some NETs don't cause symptoms until they have grown or spread
- Some NETs cause symptoms that are similar to those caused by more common conditions, so it may take longer for a NET to be diagnosed

Some NETs, however, may be found during routine exams and diagnosed at an early stage of disease.



Adapted from The National Cancer Institute. *Understanding Cancer Series: Cancer.*

# How are neuroendocrine tumors (NETs) characterized and managed?

“NET” is a term used for several specific types of tumors. Your doctor may use one of the following terms when discussing your NET:

- **Carcinoid**
  - A NET that *may* secrete serotonin and other chemicals that cause carcinoid syndrome
  - Carcinoid tumors usually originate in the gastrointestinal (GI) tract or the lungs
- **Gastroenteropancreatic NET (GEP-NET)**
  - A NET found in the GI tract or the pancreas
- **Gastrinoma, glucagonoma, insulinoma, or VIPoma (vasoactive intestinal peptide tumor)**
  - Types of GEP-NETs that secrete hormones that cause different symptoms and clinical syndromes
- **Islet cell tumor (or pancreatic neuroendocrine tumor)**
  - A GEP-NET found in the islet cells of the pancreas
- **Merkel cell carcinoma**
  - A type of skin cancer
- **Medullary thyroid carcinoma**
  - A NET found in the thyroid gland
- **Small-cell lung carcinoma**
  - A malignant form of lung cancer that may be classified as a NET

## Where do NETs originate?

NETs originate in neuroendocrine cells located throughout your body. Most NETs first appear in your GI tract or your pancreas. They are called GEP-NETs.

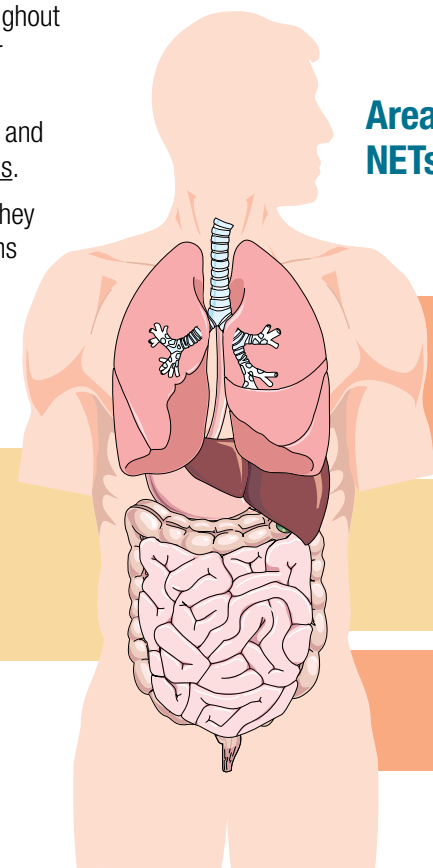
NETs can also appear in your lungs, ovaries, testes, and other glands, such as the thymus and adrenal glands.

One of the ways doctors classify NETs is by where they originate in your body. Your doctor may use the terms “foregut,” “midgut,” and “hindgut” when talking with you about your NET (see figure).

### Pancreatic NETs

- Gastrinoma
- Insulinoma
- Glucagonoma
- VIPoma
- Somatostatinoma
- Pancreatic polypeptidoma

Like other NETs, pancreatic NETs can also be nonfunctional tumors.



### Areas in your body where NETs may appear

#### Other NETs\*

- Lungs
- Thymus
- Stomach
- First part of small intestine

- Appendix
- Second part of small intestine
- Large intestine

- Colon
- Rectum

\*Well- to moderately differentiated NETs of the lung and GI tract can also be referred to as carcinoid tumors.

## What are other ways that NETs are classified?

Your doctor may talk about your NET in terms of whether it does or does not secrete hormones.

- NETs that produce hormones are called “functional” or “secretory” tumors
  - If a NET secretes large amounts of hormones, it may cause a variety of symptoms
- NETs that don’t produce hormones are called “nonfunctional” or “nonsecretory” tumors
  - These may only cause symptoms produced by the tumor’s growth, such as pain, intestinal blockage, or bleeding

Doctors also describe a NET in terms of what it looks like under a microscope (how it is structured). This includes how differentiated the NET is from the surrounding tissue.

- A NET may be described as being “well” or “poorly” differentiated
  - Well-differentiated NETs can be low or intermediate grade. They generally are slower growing
  - Poorly differentiated NETs are high grade. They generally are aggressive and grow and spread rapidly

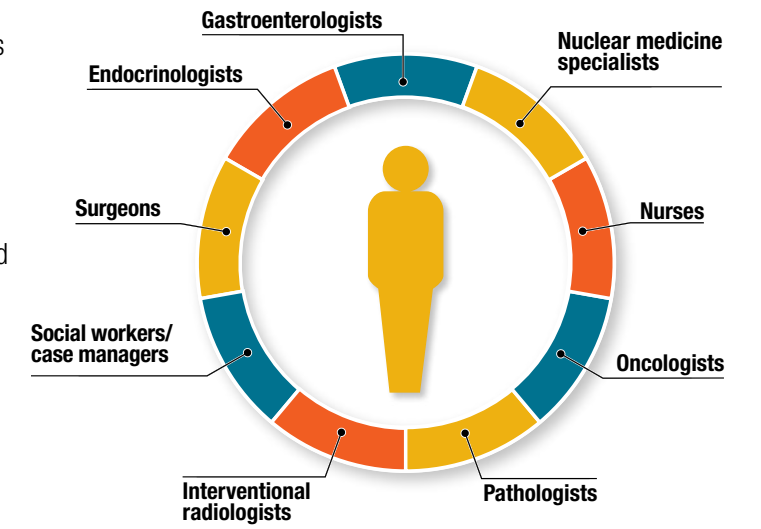
## What specialists may be involved in my care?

Your doctor or your local oncologist may collaborate with experts who specialize in NETs. In some cases, a team of healthcare professionals from various specialties will utilize a multidisciplinary approach so that they may provide the best care for patients with NETs.

A multidisciplinary approach involves using different medical specialists, each of whom has expertise in a specific area (see figure). Specialists whom patients with NETs might see include, but are not limited to

- **Oncologist:** specializes in tumors and how to manage them
- **Gastroenterologist:** specializes in diseases of the GI tract and their symptoms
- **Endocrinologist:** specializes in diseases of the endocrine system
- **Surgeon:** performs surgery, including the removal of tumors

- **Pathologist:** specializes in diagnosing diseases by using a powerful microscope to examine samples taken during a tumor biopsy
- **Nuclear medicine specialist:** has expertise in imaging (taking medical pictures of) tumors
- **Interventional radiologist:** uses radiology treatments on tumors that have spread to other parts of the body
- **Nurses/social workers/case managers:** skilled in caring for patients with NETs



## Why may it be best to manage NETs using a multidisciplinary approach?

Here are just a few reasons why NETs may be best managed in collaboration with a team of medical experts:

- NETs occur in different areas of the body
- NETs appear and grow differently
- NETs cause different symptoms and comorbidities
- NETs may or may not spread to other parts of the body

# Do all neuroendocrine tumors (NETs) cause the same symptoms?

Different types of NETs cause different symptoms. The symptoms you may experience depend on whether or not your NET is *functional* or *nonfunctional* and on the location of the tumor.

- *Functional* NETs produce symptoms caused by the secretion of hormones (see table on right)
- *Nonfunctional* NETs do not secrete hormones. They may only produce symptoms caused by the tumor's growth, such as intestinal blockage, pain, and bleeding

## What symptoms can be caused by functional NETs?

The hormones and other products secreted by functional NETs sometimes cause clinical syndromes. A syndrome is a set of symptoms that occur together due to the presence of a disease.

The syndrome caused by a functional NET depends on the hormones it secretes. Some key types of functional NETs and the syndromes they may cause are shown in the table on the right.

## What can I do to better manage my symptoms?

Consider seeking out resources that may help you understand your condition. Some patients have found the following approaches helpful:

- Research reliable sources on the Internet
- Learn about local, regional, or national patient advocacy and support groups, and join the ones that seem right for you

Keep track of your symptoms

- Keep accurate records about all symptoms you experience
- Keep track of any changes in how frequent or severe your symptoms are, or any new symptoms
- Keep track of any changes in your eating habits or daily routines

Tell your healthcare team about

- Your symptoms
- Your allergies, eating habits, and any prescription or over-the-counter medications you are taking
- Any changes in your daily routines

## What can I do to live more comfortably with my condition?

Consider making changes in your lifestyle and eating habits that may help alleviate symptoms. For example:

- Follow a nutritious diet
- Perform regular, mild exercise, such as walking
- Get plenty of sleep, and take naps if necessary
- Avoid stress

## Syndromes and symptoms caused by functional NETs

Type of NET	Typical location	Hormone produced	Associated syndrome	Characteristic symptoms*
<u>Carcinoid</u>	Stomach, bowel, small intestine, <u>pancreas</u>	<u>Serotonin</u>	<u>Carcinoid syndrome</u>	<ul style="list-style-type: none"> <li>• Diarrhea (including nighttime)</li> <li>• Cramping</li> <li>• Flushing</li> <li>• Wheezing</li> <li>• <u>Pellagra</u> (causes abdominal pain; redness and swelling of the mouth and tongue; rash; and memory loss)</li> <li>• May also cause <u>lesions</u> in the heart valves</li> </ul>
Gastrinoma	<u>Duodenum</u> , pancreas	<u>Gastrin</u>	Zollinger-Ellison syndrome	<ul style="list-style-type: none"> <li>• Ulcers</li> <li>• Diarrhea</li> </ul>
Insulinoma	Pancreas	<u>Insulin</u>	<u>Hypoglycemia syndrome</u> (Whipple's triad)	<ul style="list-style-type: none"> <li>• Hypoglycemia</li> <li>• Weight gain</li> <li>• Poor or abnormal absorption of nutrients</li> <li>• Insulin resistance</li> </ul>
VIPoma (vasoactive intestinal peptide tumor)	Pancreas	<u>Vasoactive intestinal peptide (VIP)</u>	Verner-Morrison syndrome	<ul style="list-style-type: none"> <li>• Watery diarrhea (also known as pancreatic cholera), which can cause <u>hypokalemia</u> or <u>hypochlorhydria</u></li> </ul>
Glucagonoma	Pancreas	<u>Glucagon</u>	–	<ul style="list-style-type: none"> <li>• Rash</li> <li>• Glucose intolerance (diabetes)</li> <li>• Weight loss</li> </ul>
Somatostatinoma	Pancreas, duodenum	<u>Somatostatin</u>	–	<ul style="list-style-type: none"> <li>• <u>Hyperglycemia</u></li> <li>• Diarrhea (bulky, greasy, odorous)</li> <li>• Hypochlorhydria</li> <li>• Weight loss</li> </ul>
ACTHoma	Pancreas	<u>Adrenocorticotrophic hormone (ACTH)</u>	Cushing syndrome	<ul style="list-style-type: none"> <li>• Excessive torso fat</li> <li>• Large, round face</li> <li>• Muscle fatigue</li> <li>• Thin skin</li> </ul>
GRFoma	Pancreas	<u>Growth hormone-releasing factor (GRF)</u>	Acromegaly	<ul style="list-style-type: none"> <li>• Bone deformity</li> <li>• Heart failure</li> <li>• Weakness</li> <li>• Vision problems</li> </ul>

\*Symptoms may vary among patients.



# What tests might my healthcare provider order?

A number of tests are commonly used to help identify or confirm the presence of a neuroendocrine tumor (NET), and to track any changes in your NET that might occur over time.

- Your family doctor may conduct some tests. You may be referred to a specialist for others
- Your doctor will determine which tests are appropriate for you and how often you need them

Keep accurate records about any tests you've had scheduled and the results of the tests.

Ask your doctor about any changes in your test results and what they mean. Record your doctor's answers.

## Biochemical tests

A biochemical test measures the amount or activity of a substance, such as an enzyme or protein, in a sample of blood, urine, or tissue.

### CgA test

**Type of test:** Blood sample

**How is it used?** Chromogranin A (CgA) is a substance produced by NETs that can be detected in your blood. Because the test is sensitive for many types of NETs, your doctor may use CgA testing to monitor your tumor over time.

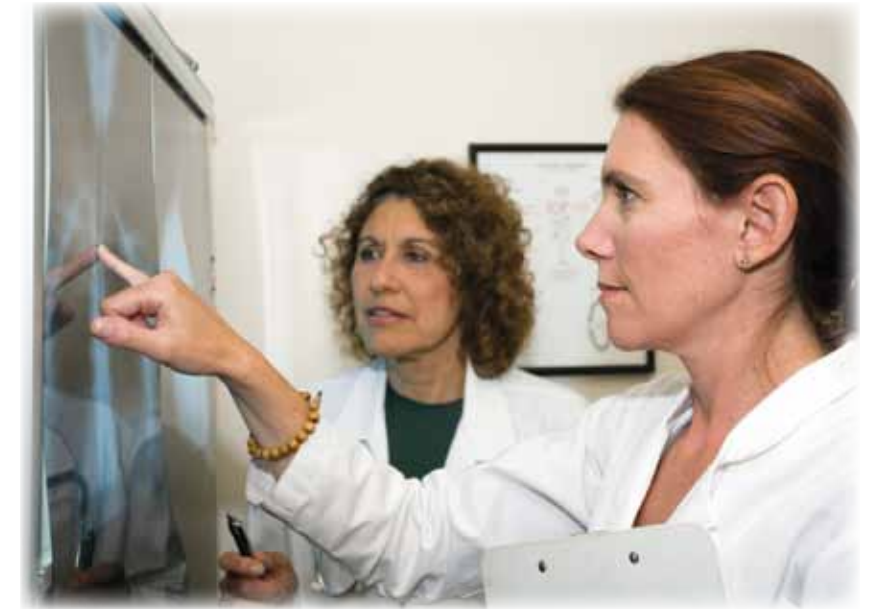
**When is it scheduled?** A CgA test may be scheduled along with other tests to confirm a diagnosis of a NET. Testing may be repeated on a regular basis (as determined by your doctor) to help monitor changes in the tumor, by comparing changes in your CgA levels to the original test results.

### 5-HIAA test

**Type of test:** 24-hour urine sample

**How is it used?** 5-HIAA (hydroxyindoleacetic acid) is a substance that occurs naturally in your body. Usually only small amounts are present in your urine. Elevated levels may indicate the presence of certain types of NETs that display groups of symptoms known as "carcinoid syndrome" (see section 3). A 5-HIAA test may be ordered alone or, less commonly, with a test measuring your blood serotonin level. A 24-hour urine sample is usually taken for the 5-HIAA test because the levels in the urine can vary during the day.

**When is it scheduled?** Your doctor may schedule the 5-HIAA test at certain intervals to help monitor your disease for a type of tumor most often associated with carcinoid syndrome. Certain serotonin-rich foods (bananas, avocados, plums, eggplant, tomatoes, plantains, pineapples, and walnuts) can increase levels of 5-HIAA in your urine and may cause false results.



## Imaging tests

An imaging test uses special instruments or techniques to visualize internal organs or tissues.

### Computed tomography (CT)

**How is it used?** CT images show the shape, size, and position of internal organs and tissues. CT is often used to establish the location of tumors. Before you have a CT scan, you may receive an injection or drink a fluid containing dye that shows up on the scan.

**When is it scheduled?** Your doctor may schedule a CT scan of your tumor when it is first diagnosed. Additional scans may then be taken to see if your tumor is growing or has otherwise changed over time.

### Magnetic resonance imaging (MRI)

**How is it used?** MRI produces soft-tissue images. This allows your doctor to identify different types of body tissue, and to see the difference between normal, healthy tissue and diseased tissue, such as a tumor.

**When is it scheduled?** Your doctor may schedule an MRI to diagnose a tumor or monitor tumor growth or to see if the tumor is spreading. An MRI may also be ordered to provide a higher level of detail than CT.

### Octreoscan™

**How is it used?** Octreoscan is used to identify specific types of NETs and disease that has spread to other parts of the body. For this test, you are injected with a tracer and scans are usually taken 24 hours later. Roughly 80% to 90% of NETs can be identified with the specific tracer used in this test.

**When is it ordered?** Octreoscan is ordered when your doctor wants to identify tumors that may have been missed by other tests, and to see how much your disease has spread.

### How are these imaging tests different?

MRI is usually used for imaging soft tissue, while CT is the preferred technique for imaging bone.

CT and Octreoscan use a radiation source, whereas MRI does not.

Octreoscan is a trademark of Covidien AG or one of its affiliates.

# How will my doctor decide the best way to manage my neuroendocrine tumor (NET)?

How your healthcare team decides to manage your NET depends on a number of factors. These include:

- The size and location of your tumor
- The grade and stage of your tumor
- Whether your tumor is functional or nonfunctional
- Any symptoms you may be experiencing
- Quality of life, overall health, and personal considerations

## What strategies might my doctor use as part of my care plan?

Options your doctor or other members of your healthcare team may discuss with you are listed below.

### Surgery

Surgery is the first option for many patients with NETs.

The goal of surgery is to fully remove a NET or reduce the tumor burden.

- For patients who are diagnosed early, surgeons attempt to remove the NET or manage the growth of the tumor. The goal is to keep the patient disease- and symptom-free for as long as possible
- For patients whose disease has spread to other parts of the body, if surgery is an option, it may help ease symptoms

### Chemotherapy

Some chemotherapy agents are routinely used to manage certain types of NETs. However, the use of some newer chemotherapy agents and other systemic agents in managing NETs is still being studied.

Chemotherapy may also be used to ease symptoms and control tumor growth.

### Chemoembolization

Chemoembolization is a procedure in which the blood supply to a tumor is temporarily blocked and chemotherapy drugs are administered directly into the tumor.

Chemoembolization may be used when disease has spread to the liver.

### Radiofrequency ablation

Radiofrequency ablation is a technique in which radio waves are used to heat and destroy abnormal cells, such as tumor cells. The radio waves travel through electrodes that can be positioned near the tumor.

Radiofrequency ablation may be used to reduce tumor size or when disease has spread to the liver and cannot be removed with surgery.

### Radiotherapy

Radiotherapy may be used when disease has spread and may help ease symptoms when disease has spread to the bone.

### Targeted radionuclide therapy

A radionuclide is a chemical element that releases radiation. In targeted radionuclide therapy, radionuclides are combined with a substance that is injected into your body and collects in specific areas.

A technique called <sup>131</sup>I-MIBG can be used to help ease symptoms in some patients who have disease that has spread or cannot be removed with surgery. Other forms of targeted radionuclide therapy are currently under development.

### Medicines

Medical treatments are used to treat certain NET patients. Your doctor may discuss these options with you as part of your personalized management plan.

### Waiting or observing

Because certain NETs might not spread or cause symptoms for some time, your doctor may recommend a gradual period of observation as a strategy for your disease.

Your doctor may suggest this strategy, at least initially, if your NET is causing occasional symptoms that can be controlled with diet and lifestyle adjustments, or if the risk of surgical or medical intervention outweighs the benefit. During this time, your doctor may continue to order tests to monitor your disease.

Your doctor will work with other members of your healthcare team to determine the best management plan for you.



## Are other treatment options being tested or developed?

Clinical studies may be an option for you.

You can learn more about clinical studies involving NETs on Web sites such as ClinicalTrials.gov ([www.clinicaltrials.gov](http://www.clinicaltrials.gov)), which lists all clinical studies currently being conducted throughout the world. The site gives you information about a study's purpose, who may participate, locations, and phone numbers for more details. This information should only be used in conjunction with advice from your doctor.

# What can I do to live more comfortably with my disease?

For many patients, neuroendocrine tumors (NETs) are a lifelong condition. Like any cancer, NETs can be life changing.

It's important to learn to live comfortably with your disease. Here are some tips.

## Understand your patient journey

Your journey as a person with a NET encompasses initial diagnosis and ongoing management of your condition.

Take an active role in your journey. All it takes is dedication and openness. Staying engaged and informed can help you

- Receive the best possible care
- Monitor your care plan
- Stay constantly aware of your disease state
- Talk comfortably about your symptoms
- Feel confident about your health
- Keep a positive outlook

## Talk openly with your doctors and nurses

One way to take control of your health is to establish a strong, open relationship with your healthcare team. You should

- Provide accurate information about your symptoms, including changes in frequency and severity, and any new symptoms you may experience
- Provide information about allergies, eating habits, and all medications you are currently taking
- Ask any questions you have about your NET and your health. Write down the answers carefully

Develop a plan with your healthcare team and stick to it.

## Make lifestyle and dietary changes

Depending on the type of NET you have, your doctor may recommend things you can do to minimize interruptions to your daily routines caused by your condition. Be sure to discuss all recommendations for your personal management plan with your doctor. You may want to

- Follow a nutritious diet
- Perform regular, mild exercise, such as walking
- Get plenty of sleep, and take naps if necessary
- Avoid stress

## Connect with others

Remember, you are not alone. There are other patients like you and a network of advocacy and support groups that want to help.

A list of these groups and how to contact them is included in section 7 of this brochure. Your doctor or nurse may also have information about local groups that you can contact.

## Manage your health coverage

The bills and insurance paperwork you receive from your doctor and hospital visits can be confusing. It's important to keep track of this information and file it in an organized manner.

To get a better handle on the insurance issues surrounding your care, you should

- Be sure you understand your health insurance coverage, your financial obligations, and the process for appealing reimbursement decisions
- Ask your insurance company to have a case manager or a single person assigned to you. This way, the same person will handle your questions, approvals, bills, reimbursements, and other issues
- Have your insurance company explain how to read its bills. Be sure you understand which bills will come from your insurance company, your doctor, the hospital, or other providers or specialists involved in your care

- Take notes during conversations that you have, either on the phone or in person, with your insurance company or healthcare providers



# How can I make the most of my doctor visits?

Many patients with NETs will see their doctors and other members of their healthcare team regularly, sometimes over many years.

Each of these visits is an opportunity to keep your healthcare team informed about your condition. Each visit also helps you stay informed about changes in your disease and your management plan. Here are some ways you can make the most of your visits.

## Prepare for each visit

- Bring records of previous visits/tests with you
- Write down your questions or concerns before you go

## Take notes and ask questions

- Record the answers to your questions accurately and completely
- Ask for information to be repeated or clarified if it seems unclear

## Have a caregiver attend with you, if possible

- A caregiver can help provide emotional support as well as record any information you may have missed

## Know your healthcare team

- Know what each specialist's role and focus are
- Ask why certain referrals or consultations are being requested
- Get a second opinion or schedule a visit with a NET specialist, if you feel it is necessary

## Familiarize yourself with important terms

- Use the glossary provided in this brochure
- Educate yourself by using other reliable resources, such as the Internet and patient support groups. Use the Internet carefully because not all of the available information about NETs is accurate

It is important to communicate all changes in your symptoms and general well-being to your healthcare team. This way, they are better informed about your condition and can make better decisions about your care.

# What resources are available to me as a patient with a neuroendocrine tumor (NET)?

Educational and emotional support are important for patients who have a less common condition, such as a NET.

Your healthcare team should be your first source of information. They understand your individual medical needs. But other useful resources are available to learn more about NETs and to reach out to other people who share your condition.

Whether you browse the Internet, contact a national or regional patient advocacy group, or join a local support group, you are not alone. There are many people who can help you cope with the challenges of living with a NET.

## US NET advocacy groups

**Carcinoid Cancer Awareness Network (CCAN)**  
www.carcinoidawareness.org

1-866-850-9555

CCAN is a nonprofit organization dedicated to increasing awareness of carcinoid cancer and other NETs. Focused on education and support, the group supports patients and groups across the United States and abroad.

**Carcinoid Cancer Foundation (CCF)**  
www.carcinoid.org

1-888-722-3132

CCF has a dual mission: to educate healthcare professionals and the public about carcinoid cancer and related NETs; and to encourage, conduct, and support research into these rare cancers.

**Caring for Carcinoid Foundation (CFCF)**  
www.caringforcarcinoid.org

1-617-848-3977

CFCF's signature is the aggressive pursuit of a cure through scientific funding. Its mission is to cure carcinoid and related NETs. Its vision is to eliminate the suffering of patients and family members affected by these tumors.

**Pancreatic Cancer Action Network (PanCAN)**  
www.pancan.org

1-877-272-6226

PanCAN is a nationwide network dedicated to advancing research, supporting patients, and creating hope for those affected by pancreatic cancer, including pancreatic NETs. The PanCAN Patient and Liaison Services (PALS) program is a comprehensive information service for patients with pancreatic cancer, their families, and healthcare professionals.

## International NET advocacy groups

**Worldwide NET Cancer Awareness Day**  
www.netcancerday.org

World NET Community is an independent group whose aim is to raise awareness about NET cancers on a global level. The community members include charitable organizations and patient groups from around the world working to expand to include new members to broaden the awareness campaign.

A list of international advocacy groups can be found on the World NET Community Web site: <http://netcancerday.org/community/steering-committee/>.

## Medical organizations

**Carcinoid NeuroEndocrine Tumour Society Canada (CNETS)**  
www.cnetscanada.org

CNETS Canada is committed to supporting NET cancer research and better clinical management practices that will achieve an ultimate goal of early accurate diagnosis and best possible treatment for NET patients.



**European Neuroendocrine Tumor Society (ENETS)**  
www.enets.org

ENETS is an international professional association composed of physicians and researchers whose main area of study is related to neuroendocrine tumors.

**American Society of Clinical Oncology (ASCO)**  
www.cancer.net

ASCO provides oncologist-approved information about cancer. The Web site can be viewed in English or Spanish.

**North American Neuroendocrine Tumor Society (NANETS)**  
www.nanets.net

NANETS is a nonprofit organization established by a group of doctors dedicated to improving the management of patients with NETs. The group sponsors educational and research programs for doctors and researchers working with NETs.

## Other resources

**CarcinoidLink**  
www.carcinoid.com

CarcinoidLink is an informative site sponsored by Novartis Oncology that focuses on carcinoid syndrome. It provides a medical explanation of the condition, management strategies, patient stories, and other resources.

**The NET Community**  
www.thenetalliance.com

The NET Community, sponsored by Novartis Oncology, is a comprehensive resource for patients who have been diagnosed with a NET. It includes videos, downloadable information, expert advice, and stories from other patients with NETs.

# For caregivers and family members

## What can friends or family members expect when caring for a loved one with a NET?

For many patients, NETs are a lifelong condition. Like any cancer, NETs can be life changing. Your loved one may undergo surgery or other treatments, go long periods of time without symptoms, or experience sudden worsening of symptoms.

You will be with your loved one through the ups and downs of this journey. It is important to let your loved one know often that you are concerned for his or her well-being and that you are there to help.

Caregiving can mean helping your loved one perform day-to-day activities or cope with feelings about his or her condition.

## Understand the condition

NETs are all very different and affect people differently. It may be helpful for you to learn about these tumors. Your loved one's doctor can provide you with information, and several Internet resources that may be helpful are listed in this section.

It can be a comfort to patients with NETs to have a caregiver accompany them to their doctor and hospital visits. You can provide emotional support and also help by taking notes about the visit. You can help your loved one keep track of any appointments and paperwork.

## Encourage a healthy lifestyle

Your loved one's doctor has probably recommended that he or she do certain things to minimize interruptions to daily routines. You can help your loved one make some of these changes and stick with them.

Some suggestions are to:

- Follow a nutritious diet
- Perform regular, mild exercise, such as walking
- Get plenty of sleep, and take naps if necessary
- Avoid stress

Be sure to take care of yourself, too. Caregiving can be emotionally and physically tiring.

## Connect with others like yourself

There are several national and regional support groups for people who care for cancer patients. You can ask your loved one's doctor or local hospital about getting in touch with these groups.

You can also search for caregiver information on the Internet. Use the Internet carefully, because not all of the available information is accurate. Some of the advocacy groups listed in this section also provide resources for caregivers.

**Remember: You are not alone, and help is available!**

These organizations and Web sites are independently operated by third parties over whom Novartis Pharmaceuticals Corporation has no control. Novartis assumes no responsibility for any information they may provide.

# Communication is key

It is important to keep detailed notes about your condition, your symptoms, and your overall health. Sharing this information with your healthcare team may better prepare them to help you.

You may have questions that you want to ask the members of your healthcare team. Perhaps you want to know about a new symptom you are experiencing. Or you may need to have test results explained to you more clearly. It is important to ask these questions and carefully record the answers.

Here are some sample questions that can help you begin the discussion with your doctor. You can ask them as they are written here, or they may prompt further questions about your specific situation.

## Questions to ask my oncologist

### About my condition

- Where can I find more information about neuroendocrine tumors (NETs)?
- How big is my tumor?
- Are you able to find my primary tumor? Where is it located?
- Has my disease spread to other parts of my body?
- What do the terms “grade” and “stage” mean? What are the grade and stage of my disease?
- How much experience do you have with NETs?
- Should I get a second opinion?

### About my care

- What tests or procedures are necessary? How often will I need them? How will they be used in developing my care plan?
- Are there things I need to do, or not do, before each test or procedure?
- What is the goal of my care plan? What is the timeline for my care plan?
- What can I do to get ready for my care?
- How will my care plan affect my daily life? Will I be able to work and perform my usual activities?
- How can I keep myself as healthy as possible?
- Could my care plan affect my ability to have children?

### About ongoing follow-up

- How often will I need to see a doctor?
- What tests will I need? How often will I need these tests?
- How can I get copies of my laboratory test results?

- What support services are available to me and my family?
- Whom do I contact about problems or questions I may have about my care plan?
- Where can I find help with handling the cost of my cancer care?

## Questions to ask my surgeon

### Before surgery

- How much of your surgery is focused on cancer? Do you have experience with NETs?
- Do I need tests before this surgery? Can you explain them to me?
- Do I need a biopsy? Can you explain what this is? What is a biopsy used for?
- Will there be a pathology report? Can I get a copy? Who will explain it to me?
- How soon do I have to make a decision about surgery?
- What should I do to get ready for the surgery? Are there any foods or medicines I should stop taking?
- How long will my surgery take?
- Can you describe what you will do during the surgery?
- What are the risks and side effects of this surgery? How will I know if there is a problem? Should I call you?
- Will I need to stay in the hospital for this surgery? If so, how long?
- Will I have stitches, staples, or bandages?
- Will there be a scar? Where will it be located?
- Are there any permanent effects from this surgery?

- What does it mean to have a surgical drain? Will I need one?
- When will I need to return for a follow-up appointment?

### After surgery

- Will I have side effects after surgery? What can be done to ease them?
- How long will it take me to recover after the surgery?
- Are there instructions or materials about postoperative care that I can take home with me?
- Do I need additional surgery?

## Questions to ask about my health insurance

It's not always clear who the best person is to answer questions about your health insurance.

Your doctor or other members of your healthcare team may be able to answer some of your questions. Doctor's office support staff, nurses, case managers, and patient advocacy groups may also be able to help. And, of course, representatives from your insurance company can help answer questions about your specific health plan.

The following questions can help begin the discussion about your health insurance coverage.

### About my doctor visits

- Who at this office will answer my questions about health insurance?
- Will this person help me work with my insurance company?
- Can this person help me understand my medical bills and the billing codes to make sure they are correct?
- How much is my co-pay for each doctor visit? When is this payment due?
- Do you offer any payment plans?
- Will I be billed separately for laboratory tests? Are these tests covered under my health insurance?
- If an insurance claim is denied, who can help me file an appeal?



### About my hospital visits

- Is the hospital or facility being recommended to me in my insurance plan's network?
- If I need to be admitted to the hospital, what will be covered under my medical insurance?
- Will there be a co-pay for each individual visit? When are these payments due?
- Do you offer any payment plans?
- If an insurance claim is denied, who can help me file an appeal?

### About my insurance coverage

- How can a nurse case manager help me? How can I have one assigned to me?
- Is there a limit to how much my insurance will cover for my care? If so, are my medical bills likely to reach that amount?
- Who can help me understand how to read my medical bills and explanations of benefits?
- Are outpatient visits covered under my medical insurance?
- Does my insurance company need to approve any or all of my management plan before I begin?
- Does my insurance cover other doctor visits, such as for a consultation or second opinion?

## Glossary of terms

Use this glossary to look up the definitions of important terms that are underlined throughout this brochure.

**Adrenal gland:** A gland near the top of the kidney that produces hormones, such as adrenaline and cortisol. Your body has 2 adrenal glands.

**Adrenocorticotrophic hormone (ACTH):** A hormone also known as corticotropin that is produced by the adrenal gland.

**Aggressive:** A term used to describe the speed at which, or the extent to which, a tumor grows and spreads.

**Biopsy:** A procedure where a small amount of tissue is removed from the body and examined under a microscope or using other tests to determine whether a tumor is present.

**Carcinoid:** A term that may be used interchangeably with NET or GEP-NET (see below).

**Carcinoid syndrome:** A set of symptoms that occur when a functional NET that secretes serotonin begins to spread or metastasize. The symptoms may be sudden or severe. Refer to section 3 for more information.

**Carcinoma:** Cancer that begins in the skin or in tissues that line or cover internal organs.

**Chemotherapy:** The use of drugs to destroy cancer cells, usually by affecting their ability to grow.

**Comorbidity:** Coexisting medical conditions or diseases in a patient that are additional to an initial diagnosis.

**Differentiation:** In normal cells, differentiation is the process that changes a less specialized cell to a more specialized cell type. In tumor cells, differentiation refers to how developed the cells are. Differentiated tumor cells resemble normal cells. Undifferentiated or poorly differentiated tumor cells lack the structure and function of normal cells. Poorly differentiated tumor cells are considered to have a greater chance of being malignant.

**Duodenum:** The first part of the small intestine, connected to the stomach. The duodenum receives enzymes from the pancreas, along with chemicals from the liver and the gallbladder, that also aid digestion.

**Endocrine system:** A group of glands and organs that regulate and control various body functions by producing and secreting hormones.

**Functional NET:** A NET that secretes hormones and may cause a variety of symptoms. Also called a secretory NET.

**Gastrin:** A hormone secreted by the pancreas that signals your stomach to produce digestive acids and enzymes.

**Gastroenteropancreatic NET (GEP-NET):** A NET that most often originates in the gastrointestinal tract or pancreas.

**Gastrointestinal (GI) tract:** Another name for the digestive system, which includes the mouth, throat, esophagus, stomach, small intestine, large intestine, rectum, and anus.

**Glucagon:** A hormone secreted by the pancreas that raises glucose (sugar) levels in your blood.

**Grade:** A system of classifying tumor cells. The cells are graded based on how abnormal they look under a microscope and how quickly the tumor is likely to grow and spread. Low-grade tumors (grades 1 and 2) closely resemble surrounding tissue and are less aggressive. High-grade tumors (grades 3 and 4) do not resemble surrounding tissue and are more aggressive.

**Growth hormone-releasing factor (GRF):** A chemical released by the brain that signals the pituitary gland to produce growth hormone.

**Hormone:** A secreted substance, usually a protein, that is transported through the bloodstream to different organs. Hormones help regulate how some of the organs in the body work.

**Hyperglycemia:** The condition of having high levels of glucose (sugar) in your blood. Symptoms include dry mouth; thirst; frequent urination, including at night; blurry vision; and dry, itchy skin.

**Hypochlorhydria:** The condition of having low levels of hydrochloric acid in your stomach. Symptoms include halitosis (bad breath), a metallic taste in the mouth, heartburn, bloating or belching immediately after eating, and indigestion.

**Hypoglycemia:** The condition of having low levels of glucose (sugar) in your blood. Symptoms include dizziness, headache, fatigue, and confusion.

**Hypokalemia:** The condition of having low levels of potassium (salt) in your blood that may be due to excessive diarrhea.

**Insulin:** A hormone secreted by the pancreas that lowers glucose (sugar) levels in your blood.

**Lesion:** An area of abnormal tissue that may or may not be cancerous.

**Metastasize:** To spread from one part of the body to another. The terms “localized,” “regional,” and “distant” are sometimes used to describe how extensively a NET has metastasized.

**Nonfunctional NET:** A NET that doesn’t secrete hormones and may only cause symptoms due to tumor growth. Also called a nonsecretory NET.

**Pancreas:** An organ that produces hormones and enzymes that aid in the digestion of food.

**Pellagra:** A condition caused by low levels of niacin (a B vitamin) in your blood. Symptoms include poor appetite and weight loss, diarrhea, scaly skin rash, mouth ulcers, restlessness, and anxiety.

**Radiology:** The use of radiation to treat or diagnose disease.

**Radiotherapy:** The use of high-energy radiation to destroy cancer cells and shrink tumors. The radiation may come either from a machine outside the body or from radioactive material that is placed in the body near cancer cells.

**Sensitivity:** In medical testing, sensitivity refers to how much of a certain disease must be present in order for the test to measure it. A test with high sensitivity, for example, is able to detect small amounts or low levels of a disease. Sensitivity is expressed as the percentage of people who test positive for the disease among a group of people who have the disease. No test is 100% sensitive, because some people who have the disease will test negative for it. This is called a false negative result.

**Serotonin:** A hormone made by certain types of cells in the body, mostly in the gastrointestinal tract. Serotonin helps with various functions, including reducing gastric secretions and stimulating smooth muscles, such as those involved in digestion.

**Somatostatin:** A hormone that stops the release of other hormones, including gastrin, insulin, and glucagon.

**Specialized cells:** Specialized cells have specific functions in the body. They originate from unspecialized cells, also known as stem cells, which are present in the fetus and have the ability to develop into any kind of cell (see below). During fetal development, DNA coding determines the kind of cells these unspecialized cells will be. The cells then grow and change shape, becoming specialized cells.

**Stage:** The extent of a cancer in a person’s body. Staging is usually based on the size of the tumor, whether lymph nodes contain cancer, and whether the cancer has spread from the original site to other parts of the body.

**Syndrome:** A set of symptoms that occur together and suggest the presence of a certain disease or an increased chance of developing the disease. For example, hypoglycemia syndrome may be caused by a type of NET called an insulinoma, and Zollinger-Ellison syndrome may be caused by a type of NET called a gastrinoma. Refer to section 3 for more information.

**Thymus:** An organ near the base of the neck that produces infection-fighting cells.

**Thyroid:** A small gland in the neck, just under the skin below the Adam’s apple. It produces thyroid hormones, which help control metabolism.

**Tumor:** An abnormal growth or mass in the body caused when cells grow uncontrollably or fail to die when they are supposed to.

**Tumor burden:** A term used to describe the number of cancer cells, the size of a tumor, or the amount of cancer in a person’s body.

**Unspecialized cells:** Unspecialized cells, also known as stem cells, are present in the fetus. They have the ability to develop into any kind of cell. During fetal development, DNA coding determines the kind of cells these unspecialized cells will be. The cells then grow and change shape, becoming specialized cells with specific functions in the body.

**Vasoactive intestinal peptide (VIP):** A substance secreted by the pancreas that stimulates the secretion of water and electrolytes in your intestines.