

Understanding Neuroendocrine Cancer

A Fact Sheet for Patients and Their Loved Ones



Neuroendocrine cancer is a rare type of cancer that starts in neuroendocrine cells. It can be hard to diagnose because symptoms often look like other common health problems. Treatment depends on the type and location of the cancer. Many people with neuroendocrine cancer live long, full lives. Support from your care team, loved ones, and support groups can help. The purpose of this fact sheet is to educate patients and their loved ones about neuroendocrine cancer.

The main topics include:

- **Overview of neuroendocrine cancer**
- **Diagnosis process**
- **Options for treatment**
- **Side effect management**
- **Ways to find support and live well with neuroendocrine cancer**



To learn more about neuroendocrine cancer and additional support resources, visit www.CancerSupportCommunity.org/Neuroendocrine-Cancer



WHAT IS NEUROENDOCRINE CANCER?

Neuroendocrine cancer is an uncommon cancer that starts in neuroendocrine cells. This type of tumor is called a neuroendocrine neoplasm (NEN). Neuroendocrine cells are found throughout your body. They help your body respond to stress, control hormones, digest food, and keep many organs working. Hormones are chemical messengers made by the body. They travel through the bloodstream to tell organs and tissues what to do.

The areas in the body where neuroendocrine cancers are found most often include:

- Digestive system (includes small intestine, stomach, colon, appendix, and rectum)
- Lungs
- Endocrine organs (adrenal glands and thyroid – these make hormones that help control how the body works)

TYPES OF NEUROENDOCRINE CANCER

Understanding a few key terms about neuroendocrine cancer can help you and your care team talk more clearly about your specific diagnosis. These cancer cells are described based on whether they make hormones, how they look under a microscope, and how fast they are growing. Together, these features help explain what symptoms may happen and what treatment options are available.

Feature	Description	Types
Functioning	Whether cancer cells produce hormones	<ul style="list-style-type: none">• Functional – produce hormones; symptoms related to increased hormone activity• Non-functional – don't produce hormones; either no symptoms, or symptoms are related to the cancer cell location and effect on nearby organs
Differentiation	How abnormal the cancer cells look under a microscope	<ul style="list-style-type: none">• Well-differentiated – cancer cells look well-organized• Poorly differentiated – cancer cells look disorganized and messy, with dead cells mixed in
Grade	How quickly the cancer cells are growing and dividing	<ul style="list-style-type: none">• Low grade – cancer cells grow slowly• High grade – cancer cells grow quickly

Understanding NETs vs. NECs

- Slower growing (low-grade), well-differentiated NENs are called **neuroendocrine tumors (NETs)**
- Faster growing (high-grade), poorly differentiated NENs are called **neuroendocrine carcinomas (NECs)**

The difference is important because it determines your treatment options.

RISK FACTORS

The cause of neuroendocrine cancer is still being researched, and NENs can affect people of any age. It is difficult to know what causes neuroendocrine cells to become cancerous tumors. Only a very small percentage of NENs are caused by a hereditary genetic mutation (passed down through family members).



HOW IS NEUROENDOCRINE CANCER DIAGNOSED?

Neuroendocrine cancer often causes symptoms that look like other common health problems. This can make diagnosis challenging and delayed. Many patients don't experience any symptoms. In this case, the cancer can be diagnosed seemingly by accident. Many NENs patients go months or years before their diagnosis is confirmed. Not all instances of neuroendocrine cancer have the same symptoms. Symptoms depend on the cancer's location and if it produces hormones.

General Symptoms

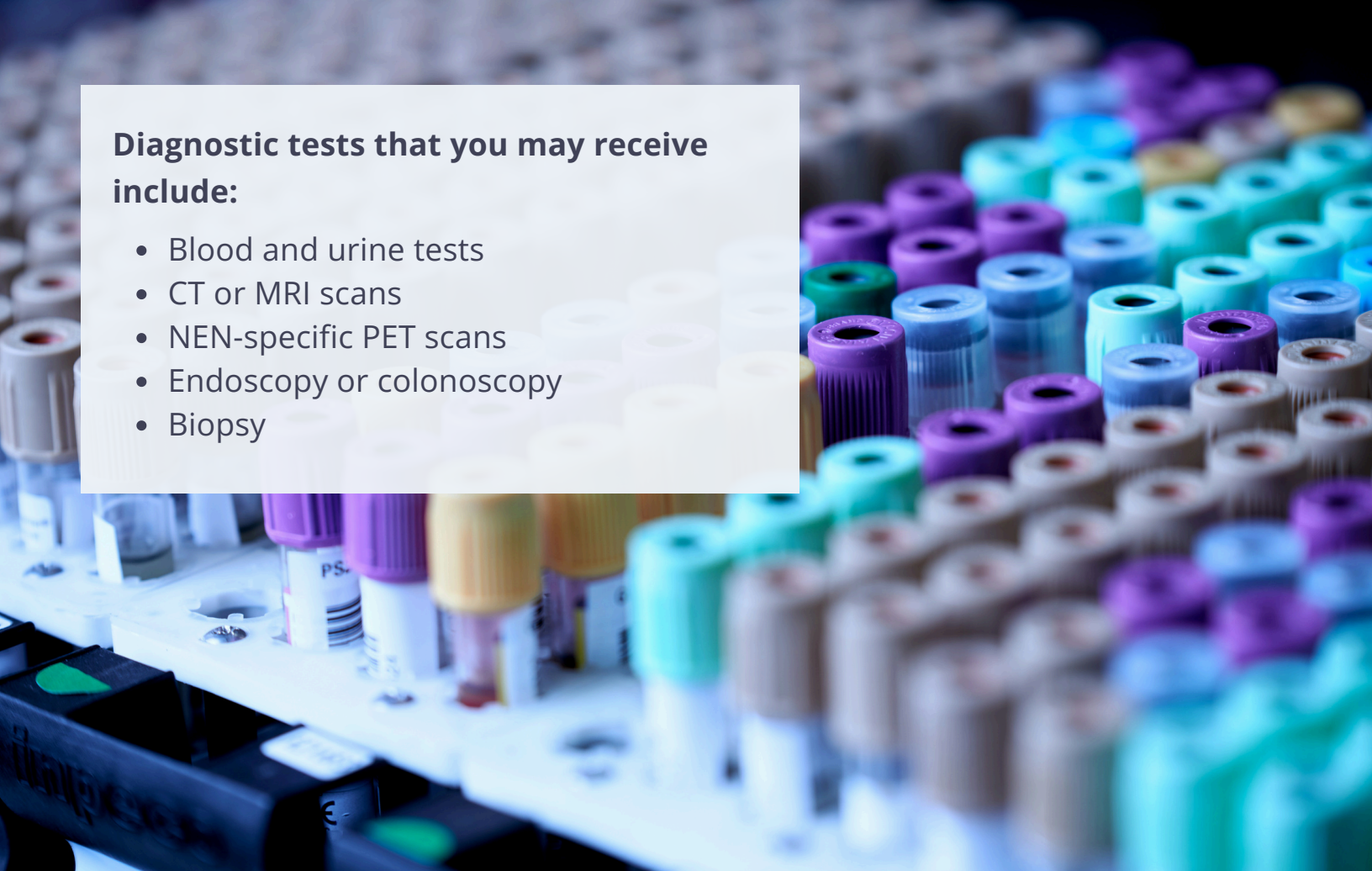
- Fatigue
- Unexplained weight loss
- Rashes

Digestive Symptoms

- Belly pain or cramps
- Diarrhea
- Blood in your stool
- Nausea/vomiting
- Feeling full quickly

Hormone-related Symptoms

- Flushing (red, warm skin on face or neck)
- Trouble breathing
- Feeling anxious
- Diarrhea
- Carcinoid syndrome (when a group of these hormone-related symptoms occur)



Diagnostic tests that you may receive include:

- Blood and urine tests
- CT or MRI scans
- NEN-specific PET scans
- Endoscopy or colonoscopy
- Biopsy

If you have symptoms that do not improve or seem unusual for you, ask your care team about neuroendocrine cancer. Further, the size of a NEN can vary — a small tumor can be missed in some imaging tests. You may need to see more than one doctor before a diagnosis is confirmed.

Once a NEN is diagnosed, or suspected, ask to see a neuroendocrine cancer specialist. This specialist focuses on NENs and has experience with diagnosing and treating them. Seeking a specialist is a common step with a rare cancer. A neuroendocrine cancer specialist can help with:

- Choosing the right tests
- Finding appropriate treatment options
- Managing symptoms
- Working together with your local care team to get appropriate treatment close to home

You can call the Cancer Support Helpline at CSC-867-5309 or visit **Neuroendocrine Tumor Research Foundation's page** if you would like help finding a specialist.



TREATMENT FOR NEUROENDOCRINE CANCER

Even though NENs are rare, there are many options to treat them. There is often more than one way to treat NENs. This is especially true if the cancer has spread (metastasized). Work with your care team to determine the treatment options best for you.

Your treatment plan will be based on:

- Your specific NEN diagnosis
 - Type
 - Stage (whether the cancer is localized or has spread to other organs)
 - Grade
 - Location of primary tumor
- Your general health
- Your symptoms
- Your personal and treatment goals



TREATMENT OPTIONS FOR NENS

Surgery	Surgery can remove the entire tumor or as much of the tumor as possible. It is the only way to cure localized cancer that has not spread.
Chemotherapy	Also known as chemo. This treatment uses drugs to destroy or damage fast-growing cells, like cancer cells. Chemotherapy can be given through a vein or in pills. Chemotherapy is only used for more aggressive NENS.
Ablative or embolic therapies	These treatments use thermal heat, cold, or radiation to destroy tumor tissue or block the blood supply to a tumor. They can decrease the tumor's size, reduce symptoms, and/or slow the growth of a tumor.
Hormone therapy	Somatostatin analogs (SSAs) are used to control carcinoid syndrome (seen as diarrhea and flushing) and to slow down tumor growth. Somatostatin regulates the endocrine system and can slow the release of other hormones.
Targeted therapy	Uses drugs to target specific changes in cancer cells that help them grow, divide, and spread. There are different types of targeted therapies to treat neuroendocrine cancer.
Radiation therapy	Radiation therapy, or external beam radiation therapy (EBRT), uses a machine to deliver high-energy rays to kill or damage cancer cells in a specific area of the body.
Peptide receptor radionuclide therapy (PRRT)	PRRT delivers radiation directly to the neuroendocrine cancer cells that have a specific receptor. This radiation helps to shrink and kill the tumor while causing less harm to other cells in the body. This type of treatment is also known as radioligand therapy (RLT) .
Immunotherapy	Works with the body's immune system to find, attack, and kill cancer cells. These drugs can work in different ways. Some boost the immune system, so it can fight cancer better. Other drugs help the immune system better recognize cancer cells. Immunotherapy is not a common treatment for NENS.

As you are making treatment decisions, think about what is important to you and any personal goals you may have. Talk with your care team about your needs and preferences. Ask about the goals of the treatment and how they will affect your quality of life. Make sure you understand why one treatment might work better for you than another.

For more information about making treatment decisions, visit www.CancerSupportCommunity.org/Treatment-Decision-Process.

WHAT ARE CLINICAL TRIALS?

Consider talking to your care team about clinical trials. Clinical trials are research studies. They help clinicians learn new and better ways to treat cancer. Clinical trials may aim to improve a current treatment or test a new one. Clinical trials may be an option at any point during your care. A clinical trial may be the only way to gain access to a promising new treatment option.

For more information about clinical trials, visit www.CancerSupportCommunity.org/Clinical-Trials.

Visit **NETRF's Clinical Trial Finder** to search for clinical trials for neuroendocrine cancer.

Your voice and wishes are very important throughout the decision-making process. You might help choose the type of treatment, the timing of treatment, or the order of treatments.



TREATMENT SIDE EFFECTS

It helps to learn about the side effects of treatment before you begin, so you will know what to expect. Ask about side effects that may show up during or immediately after treatment, as well as long-term side effects. When you know more, you can work with your care team to manage your quality of life during and after treatment.



SIDE EFFECTS THAT YOU MAY EXPERIENCE INCLUDE:

- Diarrhea
- Flushing of the face
- Fever/infection
- Low blood counts
- High blood sugar levels
- Upset stomach/nausea, bloating
- Fatigue
- Swelling
- Hair loss
- Skin reactions
- Pain
- Appetite changes
- Anxiety or distress

MANAGING SIDE EFFECTS AND SEEKING SUPPORT

Getting a neuroendocrine cancer diagnosis and the proper treatment can be challenging. Still, there are many treatment options and support resources available for you and your loved ones. Navigating a neuroendocrine cancer diagnosis may bring on both physical and emotional side effects. You may experience different side effects at different times. They may be because of treatment or as an adjustment to living with a rare cancer.

Support options include:

- Counseling
- Peer support groups
- Organizations such as Cancer Support Community (CSC), Neuroendocrine Tumor Research Foundation (NETRF), and Neuroendocrine Cancer Foundation (NCF)
- Coping skills, such as meditation, journaling, or physical activity



Side effects can also be related to nutrition, so talk with an oncology registered dietician early. They can recommend foods that may be easier to consume during treatment to help you stay nourished and satisfied. For more information about supporting your nutrition, visit www.CancerSupportCommunity.org/Diet-Nutrition.

It is important to let your care team know about any emotional or physical changes you experience. Your care team can offer medications and strategies to manage side effects and reduce discomfort. Visit www.CancerSupportCommunity.org/Cancer-Treatment-Side-Effects for more information on managing treatment side effects.

Many people with neuroendocrine cancer live long, full lives. Many NENs grow slowly, and treatments continue to improve. Managing your symptoms, meeting with specialists, and staying informed can help you take control of your care.



SUPPORTING YOUR MENTAL HEALTH

It may take time to understand your diagnosis, especially if your path to answers was long or confusing. Delayed diagnosis can lead to frustration, fear, or feeling dismissed. No level of distress or anxiety is too small to ask for help. Talk to your care team about how you are feeling. They can provide options for support.

To learn more about coping with mental health and cancer, visit www.CancerSupportCommunity.org/Coping-Mental-Health-Concerns.



SUPPORT FOR CAREGIVERS

Caregivers play such an important role in the cancer experience. They provide important physical, emotional, and logistical support. While this role can be rewarding, the responsibilities can often be stressful and take up a great deal of time. It is important that your caregiver(s) feel supported every step of the way.

As your caregiver navigates the cancer experience, remember:

- Your care team can provide resources. They can connect your caregiver(s) to other forms of support to help manage their duties.
- It is important for your caregiver(s) to keep up with their own health needs. Setting time to care for themselves will greatly benefit you as well.
- Caregivers can connect with others and exchange ideas on CSC's Caregiver Support discussion forum. Visit www.MyLifeLine.org.

For more information and resources to support your caregiver(s), call CSC-867-5309 (or outside of the U.S., toll-free 888-793-9355) or visit www.CancerSupportCommunity.org/Caregivers.



Receiving and navigating a neuroendocrine cancer diagnosis can feel challenging at times. You are not alone! Stay connected with your support system, and focus on things within your control. Share your concerns and questions with your care team. Your needs may change over time, and that is OK. Having support during and after cancer treatment can make a big difference. Feel empowered to seek the care and help that you or your loved ones need.

Know that there are many resources available to you as you navigate your cancer experience. Below, you will find a list of trusted organizations that can help. Cancer Support Community and many other organizations offer helplines, support groups, online discussion boards, and other ways to connect with others who have cancer.

RESOURCES

NEUROENDOCRINE CANCER RESOURCES

Cancer Support Community

CSC-867-5309 (or outside of the U.S., toll-free 888-793-9355)

www.CancerSupportCommunity.org/Neuroendocrine-Cancer

Neuroendocrine Tumor Research Foundation (NETRF)

NETRF.org

American Cancer Society

www.Cancer.org/Cancer/Types/Carcinoid-Neuroendocrine-Tumors.html

National Cancer Institute (NCI)

800-422-6237

www.Cancer.gov/Types/GI-Neuroendocrine-Tumors

NCI's Clinical Trials Registry

800-422-6237

www.Cancer.gov/ClinicalTrials

Neuroendocrine Cancer Foundation

www.NCF.net



Cancer Support Community Resources

Cancer Support Helpline® — Have questions, concerns, or looking for resources? Call CSC's toll-free Cancer Support Helpline at CSC-867-5309 (or outside of the U.S., toll-free 888-793-9355), available in 200 languages.

Open to Options® — Preparing for your next appointment? Our trained specialists can help you create a list of questions to share with your doctor. Make an appointment by calling 888-793-9355 or by contacting your local CSC or Gilda's Club.

Frankly Speaking About Cancer® — Trusted information for cancer patients and their loved ones is available through publications, online, and in-person programs.

Services at Local CSCs and Gilda's Clubs — With the help of over 200 locations, in 50 markets, CSC and Gilda's Club centers provide services free of charge to people impacted by cancer. Attend support groups, educational sessions, wellness programs, and more
www.CancerSupportCommunity.org/FindLocation.

Cancer Experience Registry® — Help others by sharing your cancer patient or cancer caregiver experience via survey at **www.CancerExperienceRegistry.org**.

MyLifeLine® — CSC's secure, online community welcomes anyone impacted by cancer to easily connect with community to reduce stress, anxiety, and isolation. Create a personal network site and invite friends & family to follow your journey. And participate in our discussion forums any time of day to meet others like you who understand what you are experiencing. Join now at **www.MyLifeLine.org**.

Grassroots Network — Make sure your voice is heard by federal and state policy makers on issues affecting cancer patients and survivors by joining our Network at **www.CancerSupportCommunity.org/Become-Advocate**.

This publication is available to download and print yourself at **www.CancerSupportCommunity.org/Neuroendocrine-Cancer**.

For print copies of this publication or other information about coping with cancer, visit **Orders.CancerSupportCommunity.org**.

Frankly Speaking About Cancer:
Neuroendocrine Cancer Program Partners:



This publication was made possible through generous support from:
Exelixis

The Cancer Support Community and its partners provide this information as a service. This publication is not intended to take the place of medical care or the advice of your doctor. We strongly suggest consulting your doctor or other healthcare professionals to answer questions and learn more.

Photos are stock images posed by models.

Brought to you by:

