

Neuroendocrine Tumors Backgrounder

What are neuroendocrine tumors?

- Neuroendocrine tumors (NET) are a diverse mixture of rare tumors formed from cells that have roles in both the endocrine and the nervous system. These cells produce and secrete a variety of regulatory hormones or neuropeptides¹.
- NET may secrete excess amounts of hormones, leading to symptoms¹.
- There are many different types of NET and these tumors are found throughout the body¹.
- Most NET occur in the digestive system and are collectively called gastroenteropancreatic neuroendocrine tumors (GEP-NET)¹.
- GEP-NET include carcinoid tumors and pancreatic neuroendocrine tumors (pNET).

Neuroendocrine tumor terminology

GEP-NET: a NET arising in the gastroenteropancreatic system (stomach, intestines, pancreas). These tumors include carcinoid and pNET.

Carcinoid: a type of slow-growing tumor, often found in the digestive system

pNET: pancreatic endocrine tumor; also known as islet cell tumors

How common are NET?

- NET are now ranked as the second most prevalent GI malignancy (behind colon cancer)².
- NET may be underdiagnosed. Often patients with NET do not experience symptoms and their tumor type may be labeled as non-functioning. Many asymptomatic NET are only discovered during surgery for other conditions².

Can NET spread?

- NET can either be benign or malignant (cancerous). When a NET is cancerous it has the potential to spread (metastasize), even though the tumor may be considered slow-growing¹.
- Overall, approximately 20% of NET in the small intestine will spread to distant sites such as the liver, lungs, bone³.
- If distant metastases are present when a NET is first diagnosed, five-year survival rate is estimated to be approximately 27% for untreated patients³.
- Up to 75% of patients with NET already have liver metastases at diagnosis⁴.

What is carcinoid syndrome?

- Affecting about 10% of people with NET, carcinoid syndrome is a set of symptoms caused by excessive amounts of serotonin and other hormones secreted by some NET, usually after the cancer has already spread to other parts of the body⁵.
- Common symptoms include diarrhea, hot red flushing of the face and asthma-like wheezing attacks. Less frequent symptoms include abdominal pain, alone or in combination with diarrhea and heart disease, which is the result of tissue build-up associated with carcinoid tumors⁵.
- Over 75% of people with carcinoid syndrome experience diarrhea, which can occur with flushing or by itself. Stools are watery and the diarrhea can be mild to severe. Episodes can occur several times a day and can interfere with daily life. Patients with severe cases of diarrhea often have trouble leaving their homes for work, social functions and other activities⁶.

How are NET diagnosed?

- NET can remain undiagnosed for years due to vague abdominal symptoms that are often attributed to irritable bowel or spastic colon. At least one-third of patients with small bowel NET experience several years of intermittent abdominal pain⁷.
- Imaging studies such as CT scans, PET scans, MRIs, ultrasound, endoscopy and radionuclide scans such as somatostatin receptor scintigraphy are common tools used to diagnose NET. Also helpful in diagnosis are: blood and urine tests that test for biomarkers such as chromogranin A (CgA) or 5-HIAA, a byproduct of serotonin⁵.
- Since these tumors are relatively rare cancers, there is no routine screening.

How are NET treated?

- Since tumors and symptoms vary widely, so does treatment.

Surgery and medication

NET therapeutic strategies include surgery for cure (which is achieved rarely), radiation treatment, chemotherapy and somatostatin analogues to control the symptoms of carcinoid syndrome⁸. Delays in diagnosis may result in a metastatic NET, which may make patients ineligible for surgery. No treatment regimen has been established for pNET patients with liver metastasis⁹.

Symptomatic relief

Somatostatin analogues bind to hormone receptors found on NET to regulate the excess hormonal secretions¹⁰. A patient with diarrhea caused by a secretory tumor may experience improved outcomes as a result of this treatment¹¹.

About Sandostatin LAR

Sandostatin LAR is a long-acting, injectable depot formulation of octreotide acetate, that is approved for the treatment of acromegaly, diarrhea/flushing episodes associated with advanced carcinoid tumors and profuse watery diarrhea associated with VIP-secreting tumors. Based on these attributes, octreotide has been used to treat symptoms associated with metastatic carcinoid tumors (flushing and diarrhea) and vasoactive intestinal peptide (VIP) secreting adenomas (watery diarrhea). In addition, octreotide substantially reduces and in many cases can control growth hormone and/or normalize IGF-1 levels in patients with acromegaly, a disease caused by a GH-secreting pituitary adenoma¹². Sandostatin LAR is not indicated as a treatment for tumor control. The clinical trials that supported approval of Sandostatin LAR did not study effect on tumor size or rate of growth.

The active ingredient in Sandostatin LAR, octreotide acetate, was approved in the United States in October of 1988. In November of 1998, the FDA approved the long-acting formulation of octreotide acetate which Novartis markets as Sandostatin LAR. Through more than a decade and 600,000 patient years of experience, the active ingredient in Sandostatin LAR has achieved a long-standing track record of sustained efficacy and a well-established safety profile¹².

Sandostatin LAR important safety information

The most frequently reported drug-related adverse events were biliary disorders (62%), gastrointestinal disorders (14% to 38%), and injection-site pain (20% to 50%). Hypoglycemia (4%), hyperglycemia (27%), sinus bradycardia (19%), conduction abnormalities (9%) and arrhythmias (3%) have been reported. Additional adverse reactions identified in clinical studies include nausea, abdominal pain, gas, constipation, vomiting, pain on injection, high or low blood sugar levels and slow or irregular heart rate. Many patients developed gallstones, although few patients required treatment.

For full prescribing information, please visit www.us.sandostatin.com¹².

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References

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