NEUROENDOCRINE TUMORS FACT SHEET

Neuroendocrine tumors (NETs) arise from cells that can produce and secrete a variety of hormones that regulate bodily functions\(^1\). There are many types of NETs that can occur throughout the body; however, most are found in the gastrointestinal tract, pancreas and lungs. For example, carcinoid tumors are a type of NETs that frequently originate in the gastrointestinal tract or lungs\(^2\). Another type, pancreatic NETs, sometimes called islet cell tumors, is an uncommon NET which can be difficult to treat when diagnosed at an advanced stage\(^3,4\).

Neuroendocrine tumors can be categorized as either functional (hormone secreting) or non-functional (non-hormone secreting)\(^6\). Patients with functional NETs may experience a range of symptoms and syndromes due to the excess release of hormones from the tumor cells, such as carcinoid syndrome which can cause diarrhea and flushing\(^4,6\). Patients with non-functional NETs do not experience these symptoms associated with hormone secretion and are more likely to be diagnosed at an advanced stage, often accidentally during an unrelated examination, radiological test or surgery\(^5,6\).

Incidence, Risk Factors and Prognosis

With an incidence rate of five cases per 100,000 population, NETs are a relatively uncommon cancer\(^2\). However, the incidence of NETs are increasing dramatically, having more than quadrupled in the past 30 years\(^2,6\). People with a family history of cancer and women are at an increased risk of developing a NET, and those with diabetes are at an increased risk of developing NETs specific to the gastrointestinal tract and pancreas\(^7\).

The amount of time a patient can live with a NET varies depending on several factors, including the stage of the disease and the primary tumor site. For example, patients with well-differentiated NETs of the lung with advanced disease have a 5-year survival rate of 27%, whereas those with localized disease have a 5-year survival rate of 84%\(^2\).

Diagnosis and Treatment

Some patients with NETs may remain undiagnosed for years due to vague signs or symptoms\(^5\). The estimated time to diagnosis for NETs is five to seven years, and even symptomatic patients may be misdiagnosed because symptoms can be similar to other diseases and conditions (e.g., irritable bowel syndrome)\(^6,8\). Unfortunately, NETs are often initially diagnosed at an advanced stage. For instance, approximately 60% of patients with pancreatic NETs had advanced disease at time of diagnosis in a 2008 study published in the *Annals of Oncology* that measured 1483 cases of pancreatic NETs from the Surveillance, Epidemiology and End Results (SEER) Program database\(^8\).

There is no routine screening for NETs; however, computed tomography (CT) scans, magnetic resonance imaging (MRI), ultrasound endoscopy and somatostatin receptor scintigraphy are common tools used to diagnose NETs. Also helpful in diagnosis of NETs are blood and urine tests that evaluate biomarkers such as chromogranin A (CgA) or 5-HIAA, a byproduct of serotonin\(^5\).

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**Signs and Symptoms**\(^3,5\)

Symptoms include, but are not limited to:
- Flushing
- Diarrhea
- Intermittent abdominal pain
- Wheezing
- Cough
- Bloody sputum
- Gastrointestinal bleeding
The treatment of NETs depends on the size and location of the cancer, whether the cancer has spread to other parts of the body and the patient's overall health. Current therapeutic options include surgery, radiation therapy, chemotherapy and medical therapies.

References