

Carcinoid Syndrome

Carcinoid tumors are neuroendocrine tumors (NET) that originate from the hormone- and chemical-producing cells that normally are found in the digestive tract, lungs, and other organs.

Carcinoid tumors may cause local symptoms, but they may also produce and release various hormones into the systemic circulation causing carcinoid syndrome. Carcinoid syndrome is a constellation of signs and *symptoms* due to the secretion of chemicals predominately serotonin or other peptides such as histamine, bradykinin or tachykinin.

Symptoms

Symptoms of carcinoid syndrome include:

- Flushing (face, neck, or upper chest)
- Tachycardia
- Diarrhea
- Shortness of breath
- Wheezing due to bronchospasm
- Abdominal pain
- Carcinoid heart disease
- Unexplained weight gain
- Pellagra
- Neurosis & psychosis

Symptoms may be worsened by stress, and drinking alcohol

Diagnosis

Tests for carcinoid syndrome may include:

- 5-Hydroxyindoleacetic Acid (5-HIAA) Testing (Urine & Plasma)
- Urinary excretion of serotonin
- Chromogranin A (CgA) Testing
- Blood serotonin concentration

Imaging Studies

- Barium X-ray
- Computed tomography (CT) scan
- Magnetic resonance imaging (MRI)
- Somatostatin Receptor Scintigraphy (SRS) Testing (OctreoScan)
- Functional PET imaging with 68-Ga DOTATATE

Endoscopy – Depending on location, upper endoscopy, colonoscopy, proctoscopy, capsule endoscopy, double balloon enteroscopy and endoscopic ultrasound.

Treatment

- Surgery
- First-line therapy

Somatostatin analogs bind to somatostatin receptors on the surface of carcinoid tumors and inhibit the release of bioactive amines.

Octreotide

* Common side effects include constipation, nausea, increase in hepatic enzymes, depression, flatulence, decreased appetite, swelling of hands and feet, and fever

Lanreotide

* Common side effects include headache, dizziness and muscle spasm

Treatment

- Other agents

- *Interferon – To control refractory symptoms, this agent is often given along with SSA to help slow tumor growth.

- *Cyproheptadine, an antihistamine that can be used with carcinoid tumors that release histamines.

- *Telotristat ethyl – Is a tryptophan hydroxylase inhibitor used in combination with SSA therapy to treat adults with carcinoid syndrome diarrhea inadequately controlled by an SSA.