Background  The carcinoid syndrome (CS) is a symptom complex resulting from hormone secretion in patients with carcinoid tumors. Carcinoid tumors typically grow slowly, and patients may live for many years following diagnosis. CS becomes manifest only when sufficient concentration of hormones reach the systemic circulation, most commonly in the presence of liver metastases. This Fast Fact will focus on managing the symptoms of CS.

Symptoms and Causes  The CS includes the complex of: flushing, diarrhea, abdominal cramping, cyanosis, bronchoconstriction, and symptoms of right heart failure. Compared with the general population and other cancer patients, CS sufferers may also be at increased risk for cognitive impairment. Other specific symptoms depend on the hormones the tumor secretes. Serotonin-secreting tumors cause diarrhea and cramping; bradykinin and histamine secretion lead to flushing and cyanosis. Carcinoid tumors may also produce somatostatin, norepinephrine, dopamine, gastrin, vasoactive intestinal peptide, and other hormones. Drugs that block the hormonal secretion can help to control the symptoms of carcinoid syndrome.

Somatostatin Analogs  These drugs are the treatment of choice for CS. Three formulations are available: short-acting octreotide (continuous infusion or 50-500 mcg TID, IV or subcutaneously); depot octreotide (standard dose is 30 mg intramuscularly every 4 weeks; however, doses up to 40 to 60 mg every 4 weeks may offer added benefit); and lanreotide (standard dosing is 30 mg every other week intramuscularly). 50-70% of patients experience a significant reduction of diarrhea and flushing episodes within seven days. Efficacy and side effect profiles for the three preparations are similar. Side effects include pain at the injection site, abdominal bloating, fatigue, transient fever, elevated serum glucose, and asymptomatic biliary lithiasis.

Interferon  Interferon alpha is effective in controlling both diarrhea and flushing, although it is inferior to the somatostatin analogs. The dose is 3-9 mU subcutaneously three to seven times per week. Interferon alpha therapy is often limited by its side effects: fever, anemia, thrombocytopenia, neutropenia, fatigue, depression, and flu-like symptoms.

Other Drugs & Symptom-Specific Treatments
- Diarrhea: Cyproheptadine is an alternative treatment for carcinoid-associated diarrhea. 60% of patients report improvement within one week. The dosage is 4 mg TID given orally as a tablet; it can be titrated up to 0.5 mg/kg per day. Side effects include sedation, dry mouth, dizziness, mild blurring of vision, nausea, and vomiting. Loperamide and opioids are non-specific anti-diarrheal agents that can be used for mild symptoms.
- Wheezing: bronchodilators.
- Heart failure: diuretics; tricuspid valve replacement.

Non-Drug Treatments  Patients should be counseled to identify and eliminate stressors that reproducibly cause symptoms—this may include specific stressful situations, foods, or alcohol.

Other  Other treatments for refractory symptoms include systemic chemotherapy, hepatic artery embolization, hepatic chemoembolization, or debulking surgery of hepatic metastases.

References


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