



Deer in a Wheat Field

PIC QUESTION OF THE WEEK: 10/04/10

Q: Briefly describe Zollinger-Ellison syndrome and methods for its management?

A: In 1955, Zollinger and Ellison described the rare condition that now bears their name. The distinguishing characteristic of Zollinger-Ellison syndrome (ZES) is acid hypersecretion by gastric parietal cells. The pathobiology of hypersecretion is based on the presence of gastrin-releasing endocrine tumor (s) located in the pancreas or duodenum. Tumors are two to five times more common in the duodenum than the pancreas and generally smaller in size. Ulcers can also appear in unusual locations such as the jejunum. The syndrome most frequently occurs between the ages of 35 and 65 years and is more common in men (~60%). ZES usually presents with the clinical signs and symptoms of gastric acid hypersecretion such as diarrhea, peptic ulceration, and gastro-esophageal reflux disease (GERD). Given the nonspecific nature of these symptoms, ZES is sometimes misinterpreted as GERD or peptic ulcer disease. A diagnosis of ZES should be considered in patients presenting with the triad of abdominal pain, diarrhea, and weight loss or those suffering from recurrent or refractory ulcers. In addition, patients with multiple endocrine neoplasia type 1 (MEN-1), a type of gastrin secreting tumor that accounts for 20% of all gastrinomas, should prompt the clinician to consider ZES. Fasting serum gastrin levels greater than 100 pg/mL support a diagnosis of ZES. There are two primary methods of managing ZES: drug therapy and surgical intervention. The objective of drug therapy is control of gastric acid secretion while the aim of surgical therapy is removal of the carcinoma and reduction of gastrin secretion. Prior to the availability of proton pump inhibitors (PPIs), histamine H₂- antagonists were the drugs of choice for ZES. Famotidine, in doses of up to 800 mg daily, has been effective in the treatment of ZES. Today, PPIs are considered the preferred agents for gastric hypersecretory states. Maximum labeled daily doses of PPIs for ZES are: omeprazole 360 mg; 240 mg for esomeprazole and pantoprazole; rabeprazole 120 mg; and lansoprazole 180 mg. Some of these drugs have been proven effective up to twenty years after therapy was initiated. Long term treatment with PPIs may necessitate supplementation with vitamin B₁₂. Surgical exploration for cure is recommended for patients without liver metastases, MEN-1, or complicating medical conditions. Experienced endocrine surgeons can locate the tumors in 95% of patients. Removal of these tumors results in a 5 year cure rate of about 30%. Prognosis for patients with ZES is generally based on the presence and degree of liver metastases.

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Ryan M. Walkovich and Stephen A. Novicki, Pharm.D. Clerkship Students

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