# Zollinger-Ellison Syndrome Diagnosis and Treatment

Zollinger-Ellison syndrome (ZES) is caused by a gastrin-secreting tumor (gastrinoma) of the pancreas or duodenum, which stimulates acid-secreting cells of the stomach. This leads to recurrent ulcers of the duodenum and jejunum, and the symptoms of abdominal pain, heartburn and diarrhea. Diagnosis of this disease can be made with serum gastrin levels greater than 1000 pg/mL, or with a positive secretin stimulation test. ZES is treated with a high-dose proton pump inhibitor (PPI), and with octreotide in metastatic disease. Surgical resection is the only curative measure.




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## Diagnosis

### Gastrin > 1000 pg/mL

Gas greater than 1000 pg/mL

Normal gastrin levels in healthy patients rarely exceed 100pg/mL. Patients with Zollinger-Ellison syndrome (ZES) have fasting serum gastrin levels that exceed this, and often go beyond 1000 pg/dL. If gastrin levels are found to be this high, it is diagnostic of ZE syndrome. Another, rather sensitive and specific, diagnostic method is to measure basal gastric acid output, or to measure gastric pH. Aberrantly high levels of gastric acid output, or an extremely low pH.

### Positive Secretin Stimulation Test

Positive Secret-tin-agent

The secretin stimulation test is the most specific and sensitive test for identifying gastrinomas causative of Zollinger-Ellison syndrome (ZES). The secretin stimulation test is considered a provocative test, as the response to administered secretin is measured, and the patient's response aids in diagnosis. In this test, secretin is administered as a bolus after an overnight fast, and gastrin levels are measured in response at different time intervals. An increase in serum gastrin of greater than 200 pg/mL yields a positive test result and is diagnostic of ZES.

## Treatment

### High-Dose PPI

Large-needle Proton Pump with Inhibiting-chains

High-dose proton pump inhibitors (PPIs) are used to slow down acid secretion by parietal cells in Zollinger-Ellison syndrome (ZES), and are considered a first-line therapy. Extended use of these medications in older patients, however, is linked to an increased risk of fracture. It should be noted PPI use is not curative, but alleviates the complications of excess acid secretion in patients (GERD, heartburn, abdominal pain, ulcer formation).

### Octreotide

Octo-tree-ride

Octreotide is a somatostatin analog that is used as a second-line agent in treating Zollinger-Ellison syndrome (ZES). This medication is very effective in treating VIPomas, carcinoid tumors and glucagonomas, but has unpredictable results with the gastrinomas seen with ZES. Despite its unpredictability, this drug can reduce gastrin levels and may slow tumor growth, and can be used in patients who have unsuccessful results with high-dose proton pump inhibitors.

### Surgical Resection

Surgeon

Surgery is the only modality of treatment which may lead to a cure. This, however, is hinged on the tumor being in a resectable area and non-metastatic. All sporadic cases of Zollinger-Ellison syndrome (ZES) should be surgically explored (including duodenotomy) even with negative imaging results, because of the high likelihood of finding and removing a tumor for potential cure. Surgery for multiple endocrine neoplasia type 1 (MEN 1) related cases should be focused on prevention of metastatic disease, with surgery being recommended when pancreatic tumors are greater than 2 cm.

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