

Zollinger-Ellison syndrome (ZES) is characterized by the development of a tumor (gastrinoma) or tumors that secrete excessive levels of gastrin, a hormone that stimulates production of acid by the stomach. Many affected individuals develop multiple gastrinomas, which are thought to have the potential to be cancerous (malignant). In most patients, the tumors arise within the pancreas and/or the upper region of the small intestine (duodenum). Due to excessive acid production (gastric acid hypersecretion), individuals with ZES may develop peptic ulcers of the stomach, the duodenum, and/or other regions of the digestive tract. Peptic ulcers are sores or raw areas within the digestive tract where the lining has been eroded by stomach acid and digestive juices. Symptoms and findings associated with ZES may include mild to severe abdominal pain; diarrhea; increased amounts of fat in the stools (steatorrhea); and/or other abnormalities. In most affected individuals, ZES appears to develop randomly (sporadically) for unknown reasons. In approximately 25 percent of patients, ZES occurs in association with a genetic syndrome known as multiple endocrine neoplasia type 1 (MEN-1). All of the tumors are considered to have malignant potential. Prognosis is related to tumor size and the presence of distant metastases. ZES may become apparent at any age. However, symptom onset usually occurs between ages 30 and 60 years. The exact frequency of ZES in the general population is unknown. However, some researchers estimate that ZES represents less than one percent of peptic ulcers.