

Surgery. 2004 Dec;136(6):1267-74.

Gastric carcinoid tumors in multiple endocrine neoplasia-1 patients with Zollinger-Ellison syndrome can be symptomatic, demonstrate aggressive growth, and require surgical treatment.

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BACKGROUND: Gastric carcinoid tumors occur in 15% to 50% of patients with multiple endocrine neoplasia-1/Zollinger-Ellison syndrome (MEN-1/ZES) but are thought to be benign. We report 5 patients with MEN-1/ZES with symptomatic, aggressive gastric carcinoid tumors that required surgical procedures.

METHODS: This was a retrospective chart review. **RESULTS:** Each patient had MEN-1/ZES. Each patient had innumerable gastric carcinoid tumors with symptoms. The fasting gastrin level was 47,000 pg/mL (normal, <200 pg/mL); the basal acid output was 79 mEq/hr (n = 3), and the age at surgical exploration was 47 +/- 6 years, with a duration of MEN-1 of 21 +/- 3 years and of ZES of 15 +/- 2 years. All patients had elevated 5-HIAA or serotonin levels. Somatostatin receptor scintigraphy showed increased stomach uptake in 4 patients (80%). Four patients had a total gastrectomy; 4 patients had lymph node metastases removed, and 3 patients had liver metastases resected. One patient who did not have a total gastrectomy had liver carcinoid metastases. **CONCLUSIONS:** These results demonstrate that gastric carcinoid tumors in patients with longstanding MEN-1/ZES may be symptomatic, aggressive, and metastasize to the liver. With increased long-term medical treatment and life expectancy, these tumors will become an important determinant of survival.