

# Medical Policy: Pancreaticoduodenctomy (Whipple Resection) for Zollinger Ellison Syndrome

# [Policy Number]
Effective November 8, 2001
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# PANCREATICODUODENECTOMY (WHIPPLE RESECTION) FOR ZOLLINGER-ELLISON SYNDROME

Policy

VCHCP does <u>NOT</u> cover pancreaticoduodenectomy (Whipple resection) for the treatment of patients with Zollinger-Ellison syndrome. The value of pancreaticoduodenectomy in this condition remains to be established. The morbidity and mortality related to this approach may outweigh its potential benefits.

## **Background:**

Zollinger-Ellison syndrome (ZES) is characterized by severe peptic ulcer disease that results from non-beta islet cell tumors, gastrinomas, of the gastrointestinal tract. The mean age at presentation is 45 to 50 years, and men are affected more often than women. Gastrinomas can be subdivided into tumors that are sporadic, constituting about 75 % of patients with ZES, and those that are genetically transmitted and associated with multiple endocrine neoplasia type 1 (MEN 1), constituting about 25 % of patients with ZES. Zollinger-Ellison tumors associated with MEN-1 occur at an earlier age than the sporadic tumors and have been characterized by some researchers to follow a more benign course.

Currently, proton-pump inhibitors such as lansoprazole (Prevacid) and omeprazole (Prilosec) are the treatment of choice for ZES. In patients with sporadic ZES, exploratory surgery with tumor resection is also appropriate. However, the role of pancreaticoduodenectomy (Whipple resection) in patients with sporadic gastrinomas and in patients with MEN-1 is controversial. Furthermore, the effect of aggressive surgery, such as the Whipple resection, on survival is unclear.

#### ICD-9 Codes/CPT Codes:

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251.5 abnormality of secretion of gastrin (e.g., Zollinger-Ellison syndrome)

## CPT Code:

pancreatectomy, proximal subtotal with total duodenectomy, partial gastrectomy, choledochoenterostomy, and gastrojejunostomy (Whipple-type procedure); with pancreatojejunostomy



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#### Place of Service:

Inpatient

A. Attachments: None

### **B.** References:

- 1. Del Valle J & Scheiman JM. Zollinger-Ellison syndrome. In: "Textbook of Gastroenterology", Vol. I, 3<sup>rd</sup> edition, T. Yamada et al (eds.), 1999; Lippincott Williams & Wilkins, Chapter 65, pp. 1445-1462.
- 2. Wolfe MM & Jensen RT. Zollinger-Ellison syndrome: Current concepts in diagnosis and management. N Engl J Med 1987; 317(19): 1200-1209.
- 3. MacFarlane MP et al. Prospective study of surgical resection of duodenal and pancreatic gastrinomas in multiple endocrine neoplasia type 1. Surgery 1995; 118(6): 973-979.
- 4. Meko JB & Norton JA. Management of patients with Zollinger-Ellison syndrome. Ann Rev Med 1995; 46: 395-411.
- 5. Jensen RT. Management of the Zollinger-Ellison syndrome in patients with multiple endocrine neoplasia type 1. J Intern Med 1998; 243(6): 477-488.
- 6. Wells SA, Jr. Surgery for the Zollinger-Ellison syndrome. N Engl J Med 1999; 341(9): 689-690.
- 7. Norton JA et al. Surgery to cure the Zollinger-Ellison syndrome. N Engl J Med 1999; 341(9): 635-644.
- 8. Maton PN. Review article: The management of Zollinger-Ellison syndrome. Aliment Pharmacol Ther 1993; 7(5): 467-475.
- C. Reviewers: Richard O. Ashby MD, John Prichard MD, QA Committee