**Table I. Pancreatic neuroendocrine tumors (PNETs)**

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| --- | --- | --- | --- | --- | --- |
| **Name of tumor** | **Name of syndrome** | **Main signs or symptoms** | **Location**  **(%)** | **Malignancy (%)** | **Hormone causing syndrome** |
| I. Functional PNET | | | | |  |
| Gastrinoma | Zollinger-Ellison syndrome | Abdominal pain, diarrhea, GERD symptoms | Pancreas – 30%  Duodenum – 60  Other – 10 | 60-90% | Gastrin |
| Insulinoma | Insulinoma | Hypoglycemic symptoms | Pancreas – 100 | 5-15 | Insulin |
| Glucagonoma | Glucagonoma | Dermatitis, diabetes/glucose intolerance, weight loss | Pancreas – 100 | 60 | Glucagon |
| VIPoma | Verner‑Morrison Pancreatic cholera  WDHA | Severe water diarrhea, hypokalemia | Pancreas – 90  Other – 10 (neural, adrenal, periganglionic tissue) | 80 | Vasoactive intestinal peptide (VIP) |
| Somatostatinoma | Somatostatinoma | Diabetes mellitus, cholelithiasis, diarrhea | Pancreas – 56  Duodenum/jejunum − 44 | 60 | Somatostatin |
| GRFoma | GRFoma | Acromegaly | Pancreas – 30  Lung – 54  Jejunum – 7  Other – 13 (adrenal, foregut, retroperitoneum) | 30 | Growth hormone-releasing factor |
| ACTHoma | ACTHoma | Cushing’s syndrome | Pancreas 4-16 of all, ectopic Cushing’s | >95 | Adrenocorticotropic hormone (ACTH) |
| PNET causing carcinoid syndrome | PNET causing carcinoid syndrome | Diarrhea, flushing | Pancreas − <1% of all carcinoids | 60-90 | Serotonin, Tachykinins |
| PNET causing hypercalcemia | PNET causing hypercalcemia | Signs/symptoms of hypercalcemia | Pancreas – (rare cause of hypercalcemia) | >85 | PTHrP, other unknown |
| II. Nonfunctional PNET | | | | | |
| Nonfunctioning | Nonfunctional  PPoma | Weight loss, abdominal mass, hepatomegaly, chromogranin released but no symptoms due to their hypersecretion | Pancreas – 100 | 60-90 | None: pancreatic polypeptide (PPoma) |

*Abbreviations:* PNET = pancreatic neuroendocrine tumor; WDHA = Watery diarrhea, hypokalemia, and achlorhydria; PP = pancreatic polypeptide; PTHrP = parathormone-related peptide; GRF = growth hormone-releasing factor.