



University of Michigan
Medical School

Gastrinomas and Rare Functioning Islet Cell Tumors

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Pancreatic ICTs

- General approach
- Gastrinoma
- Glucogonoma
- VIPoma
- Somatostatinoma

General principles

- Biochemistry before radiology or surgery
- Separate control of syndrome from control of malignancy as much as possible
- Tailor the risk of the operative approach to the severity of the disease



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Step 1: Establish diagnosis biochemically

- **Gastrinoma** Elevated fasting gastrin (>130) with elevated BAO; abnormal secretin test
- **VIPoma** Secretory diarrhea (>3 l per day) and elevated serum VIP
- **Glucagonoma** Rash, hypoaminoacidemia, type 2 DM, elevated fasting serum glucagon (> 1000 pg/ml)
- **Somatostatinoma** Steatorrhea, diabetes, gallstones and elevated somatostatin levels



Step 2: Familial versus Sporadic

- Careful family history with specific attention to evidence for MEN-1
- Check serum levels of calcium and prolactin to screen for MEN-1
- If in doubt, may investigate suspicious first degree relatives



Step 3: Treat symptoms of hormone excess

- **Gastrinoma** H2-blockers or proton pump inhibitors
- **VIPoma** Octreotide and potassium supplements
- **Glucagonoma** Octreotide, anticoagulation and IVC filter



Step 4: Imaging studies

- **Gastrinoma** Preoperative CT and octreoscan; rarely secretin angiogram; ? endoscopic ultrasound
- **Glucagonoma, VIPoma, Somatostatinoma** Preoperative CT scan and octreoscan

Occasionally use MR scan to characterize liver lesions

Islet Cell Tumor Localization

- Transdermal ultrasound
- Endoscopic ultrasound
- CT scan
- MR scan
- Somatostatin receptor scintigraphy
- Angiography \pm provocative testing
- Portal venous sampling
- Intraoperative ultrasound

Islet Cell Tumor

- CT scan always necessary
- Assess the pancreatic anatomy
- Correlate with somatostatin receptor scintigraphy

Locally Advanced
and Metastatic
VIPoma

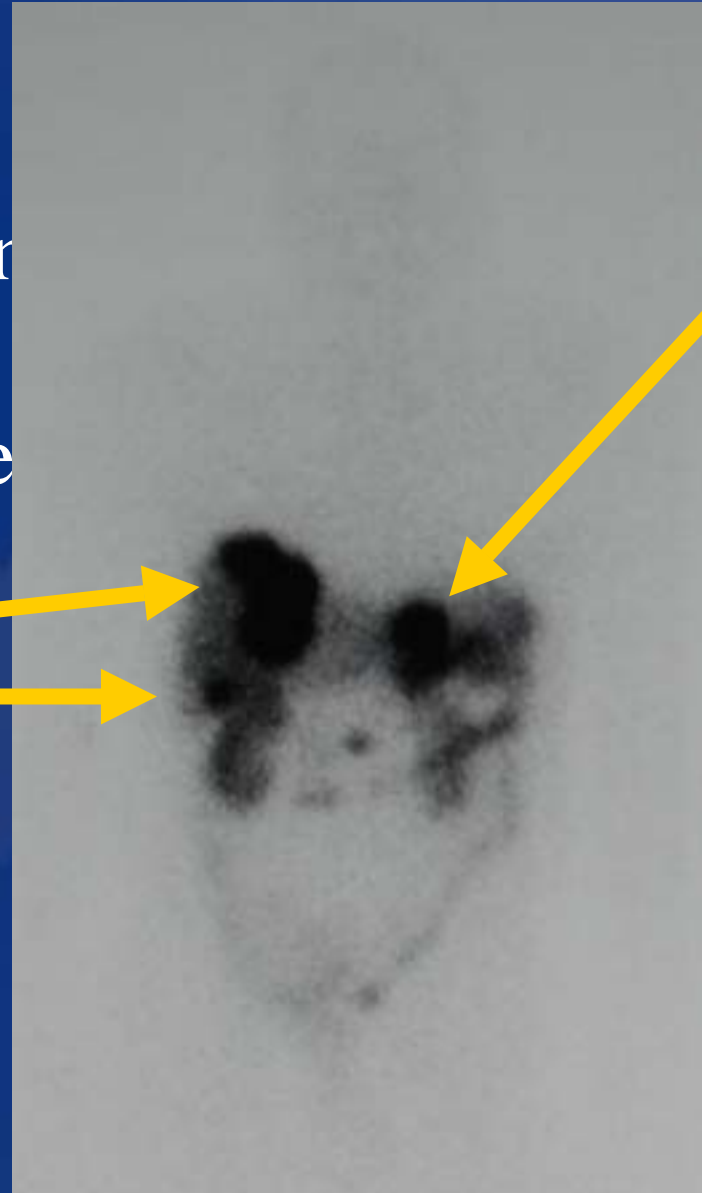


Islet Cell Tumor

- Octreoscan
- Useful for identifying other wise unsuspected disease

Liver

Pancreas



Step 5: Immediate preoperative preparation

- Pneumococcal vaccine (if splenectomy potentially necessary)
- Mechanical bowel preparation (optional)
 - Insulinoma patients must be on IV glucose during bowel prep
 - Zollinger-Ellison syndrome patients must be admitted for IV acid-blockade during bowel prep
- Octreotide (?) and antibiotics beginning immediately preoperatively

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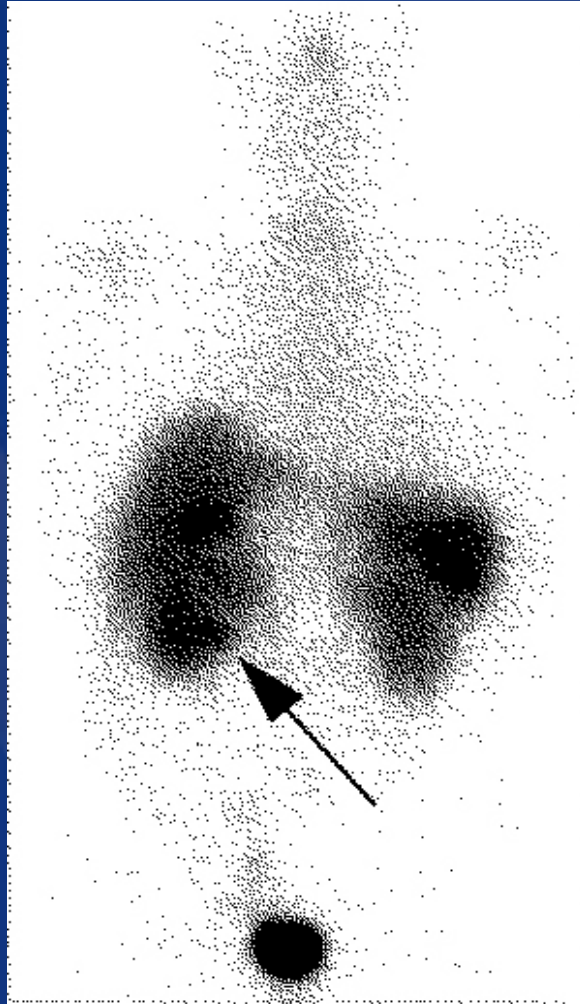
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TUMOR

TUMOR	SYNDROME	PANCREATIC TUMOR (%)	DUODENAL TUMOR (%)	MALIGNANT (%)	MEN-1 (%)
Insulinoma	Hypoglycemia	100	0	10	6
Gastrinoma	Zollinger-Ellison	38	38 (20% other)	70	25
VIPoma	Watery diarrhea Hypokalemia Achlorhydria(WDHA)	85	15	40	1
Glucagonoma	Rash Diabetes Thrombophlebitis	100	0	70	3
Somatostatinoma	Steatorrhea Diabetes Gallstones	50	50	78	1
GRFoma	Acromegaly	90	10	30	30
ACTHoma or CRFoma	Cushings syndrome	100	0	100	0

- Tenacious though indolent malignancy for most
- Diagnosed by elevated acid output and simultaneous elevated serum gastrin, or secretin provocation
- Symptom management excellent
- More aggressive, but still low-morbidity resection warranted, to include nodal dissection

Duodenal gastrinoma



Gastrinoma

- Metastases to regional lymph nodes are frequent
- Local recurrences can be large and locally invasive
- Liver and bone metastases are the life-threatening features
- Thorough, but low-morbidity resection warranted, including nodal dissection

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