Unusual Pancreatic Neoplasms

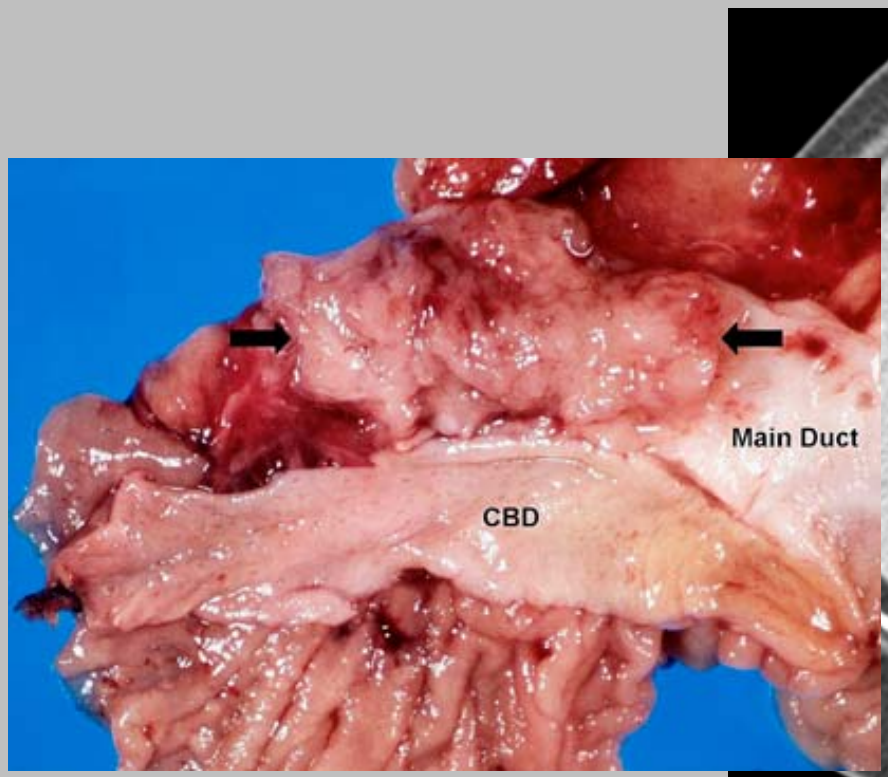
RTC 2/11/2011

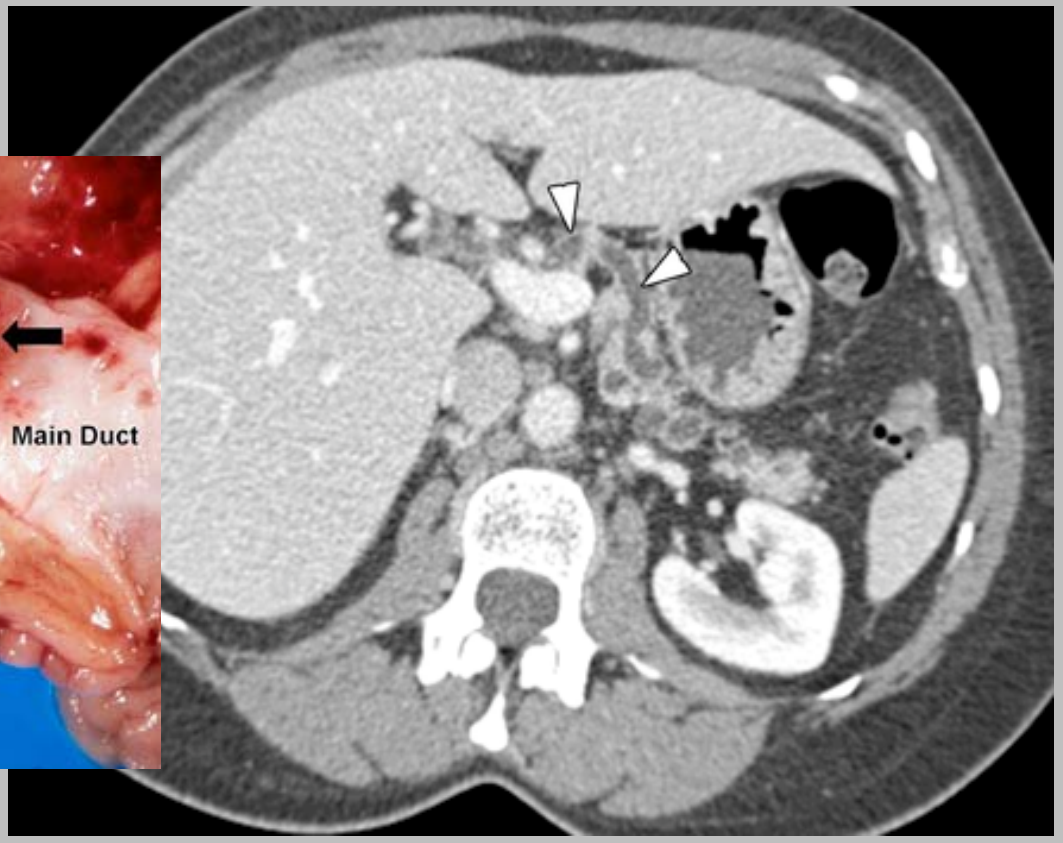
Objectives

- Intraductal Papillary Mucinous Neoplasm (IPMN)
- Mucinous Cystic Neoplasm (MCN)
- Islet Cell Tumors
  - Insulinoma
  - Glucagonoma
  - VIPoma
  - Somatostatinoma
  - Gastrinoma
- Case presentation
- Questions

Intraductal Papillary Mucinous Neoplasm (IPMN)

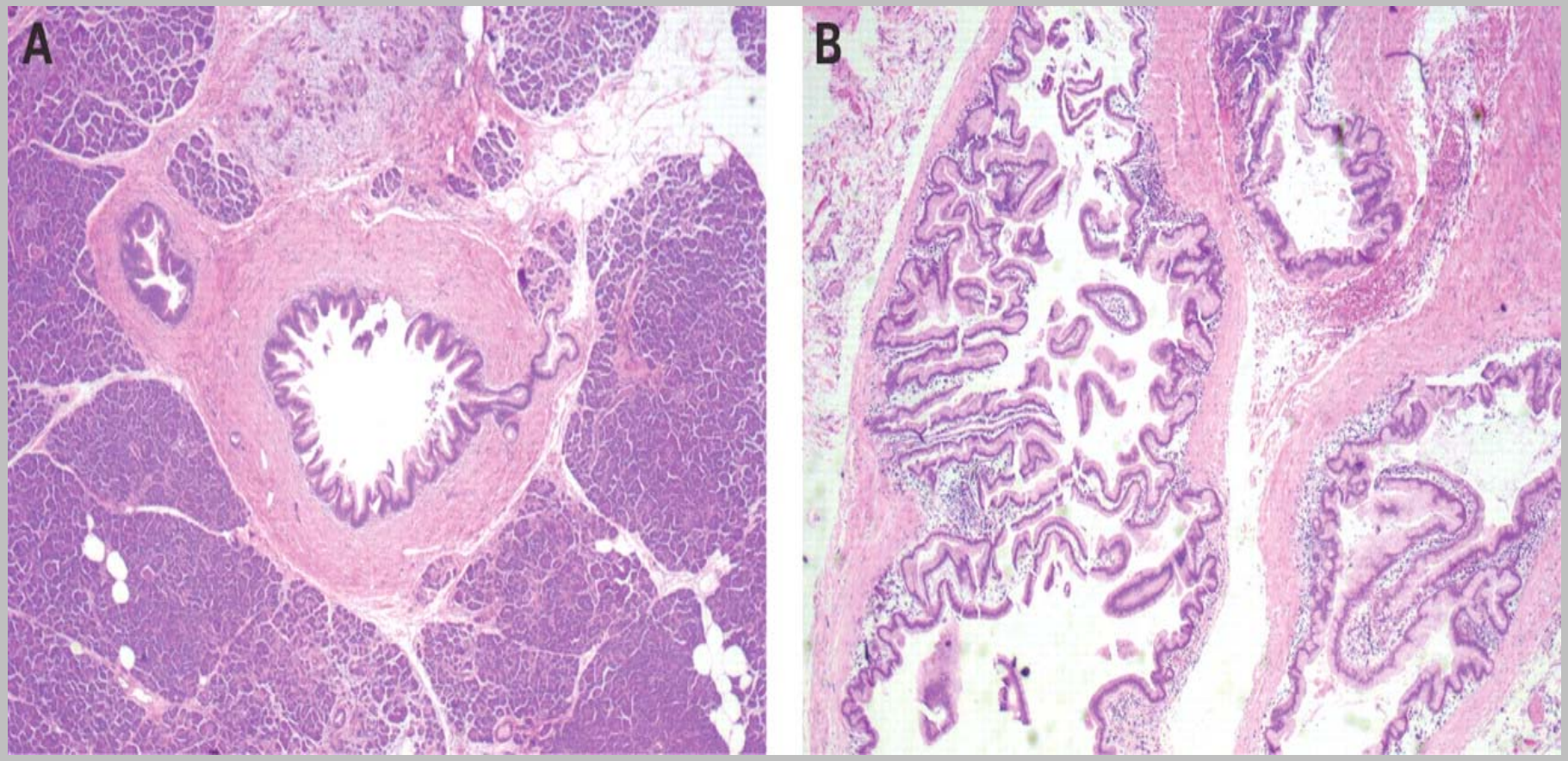
- **Intraductal**: communicates with main pancreatic duct or its major branches





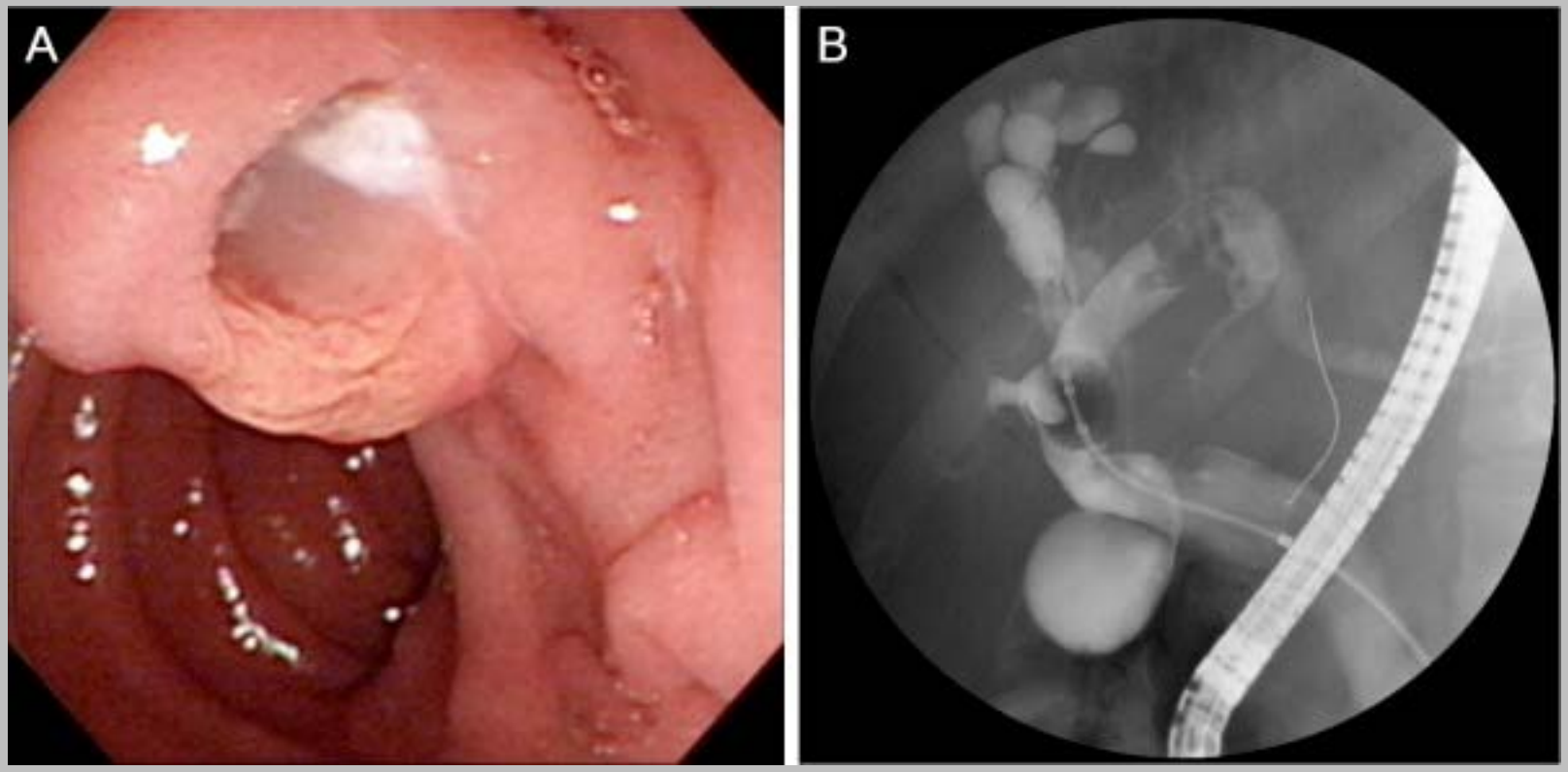
Intraductal Papillary Mucinous Neoplasm (IPMN)

- **Papillary**: intraductal epithelial projections



Intraductal Papillary Mucinous Neoplasm (IPMN)

- **Mucin**: produce thick mucin-rich fluid

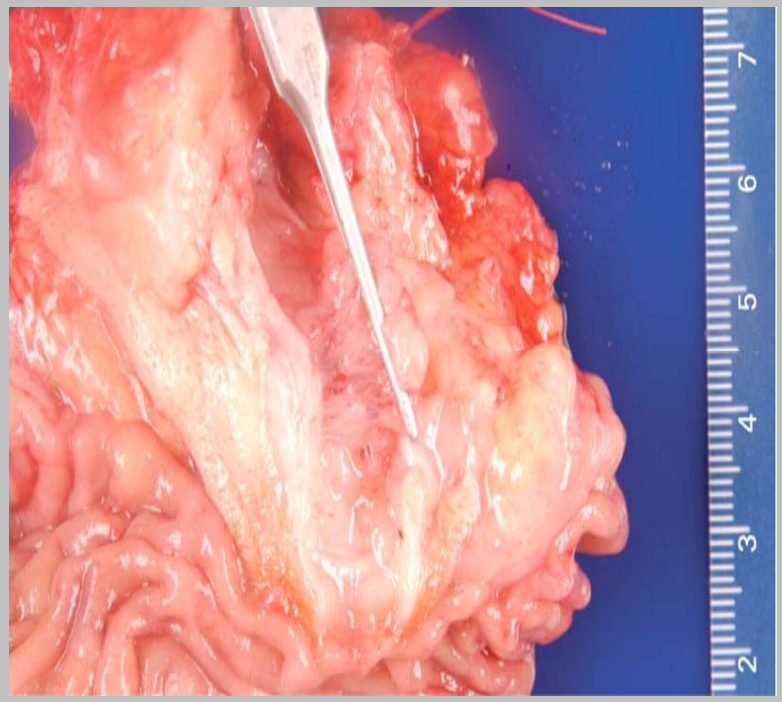


Intraductal Papillary Mucinous Neoplasm (IPMN)

- **Intraductal**: communicates with main pancreatic duct or its major branches
- **Papillary**: intraductal epithelial projections
- **Mucinous**: produce thick mucin-rich fluid
- Associated with duct dilatation (>10mm)
- Proximal in location
- Male predominance
- Occurrence 7th decade

Intraductal Papillary Mucinous Neoplasm (IPMN)

- 1% of all pancreatic neoplasms
- Approx 30-40% invasive
- Malignancy:
  - Tumor size (>3cm)
  - Main duct involvement (>12mm)
  - Proximal location
  - Presence mural nodules
  - Elevated CA 19-9
  - Biliary obstruction, Pancreatitis
- Extra-pancreatic neoplasms - *Gastric, Colorectal*
- EUS-FNA

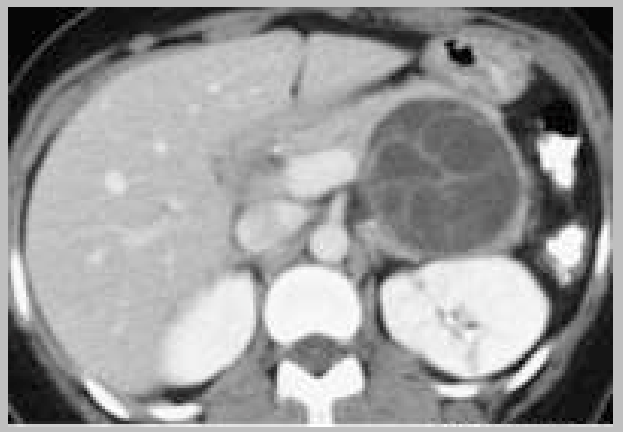


Intraductal Papillary Mucinous Neoplasm (IPMN)

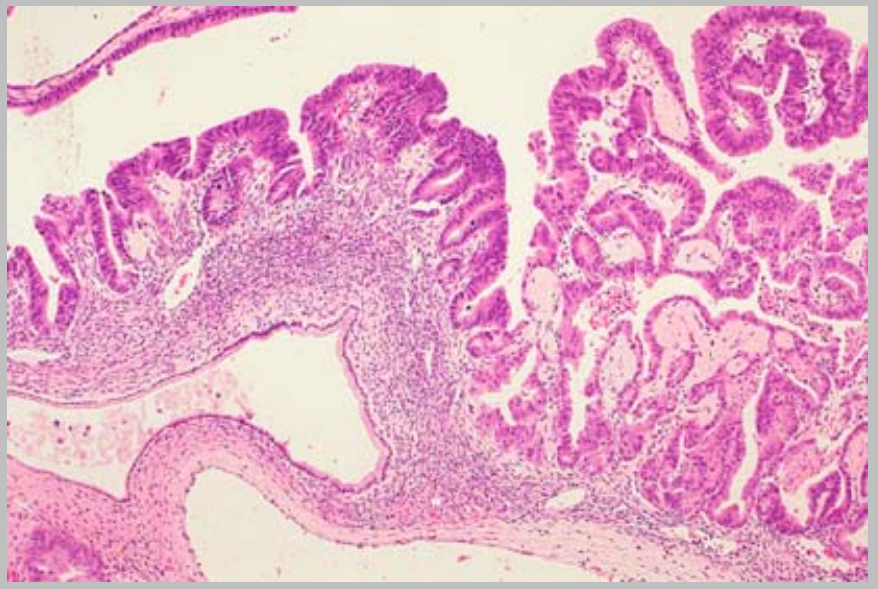
- Indications for Resection:
  - Main duct lesions
  - Branch duct lesions involving main duct
  - Tumors >3cm
  - Presence mural nodules
  - Obstructive jaundice; Pancreatitis
- Indications for Observation:
  - Lesion <3cm
  - Branch-duct involvement only
  - Asymptomatic or ⇧ perioperative risk
- Prognosis invasive IPMN: 40-60% 5-year survival
- Surveillance Imaging – all lesions

Mucinous Cystic Neoplasm (MCN)

- **Unilocular** or **Multilocular**
- Encapsulated by **fibrous tissue**

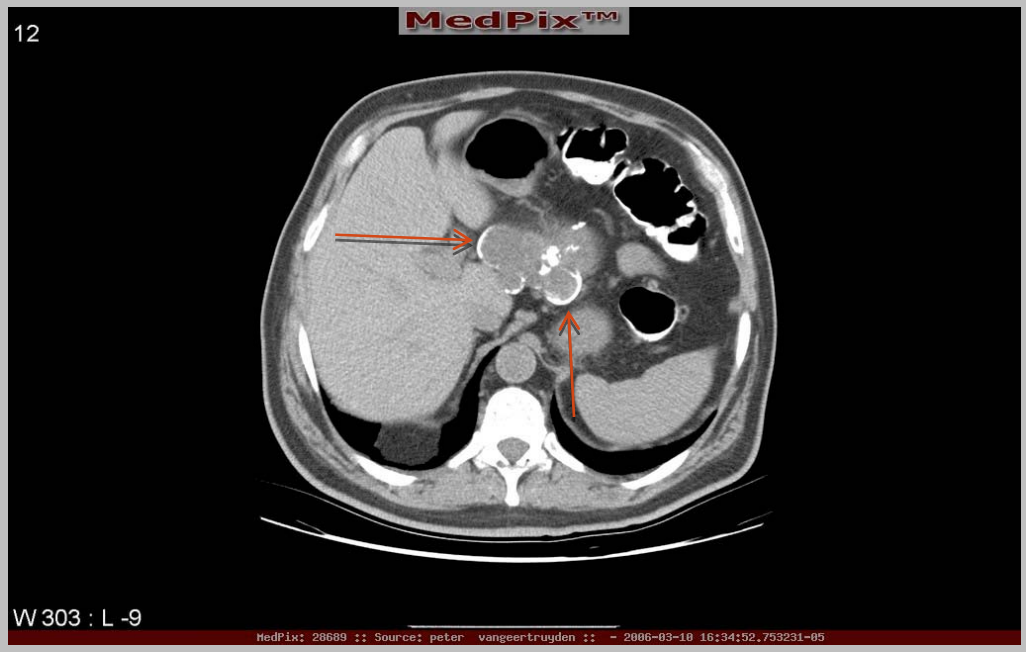


- **Ovarian-like stroma**



Mucinous Cystic Neoplasm (MCN)

- **No communication with pancreatic ducts**
- Presence of **calcification**
- Body/Tail of Pancreas
- Female predominance
- 5-6th decades

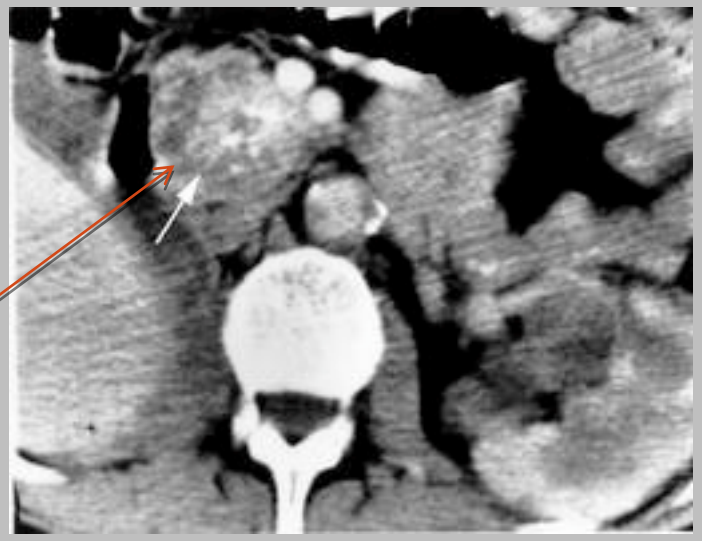


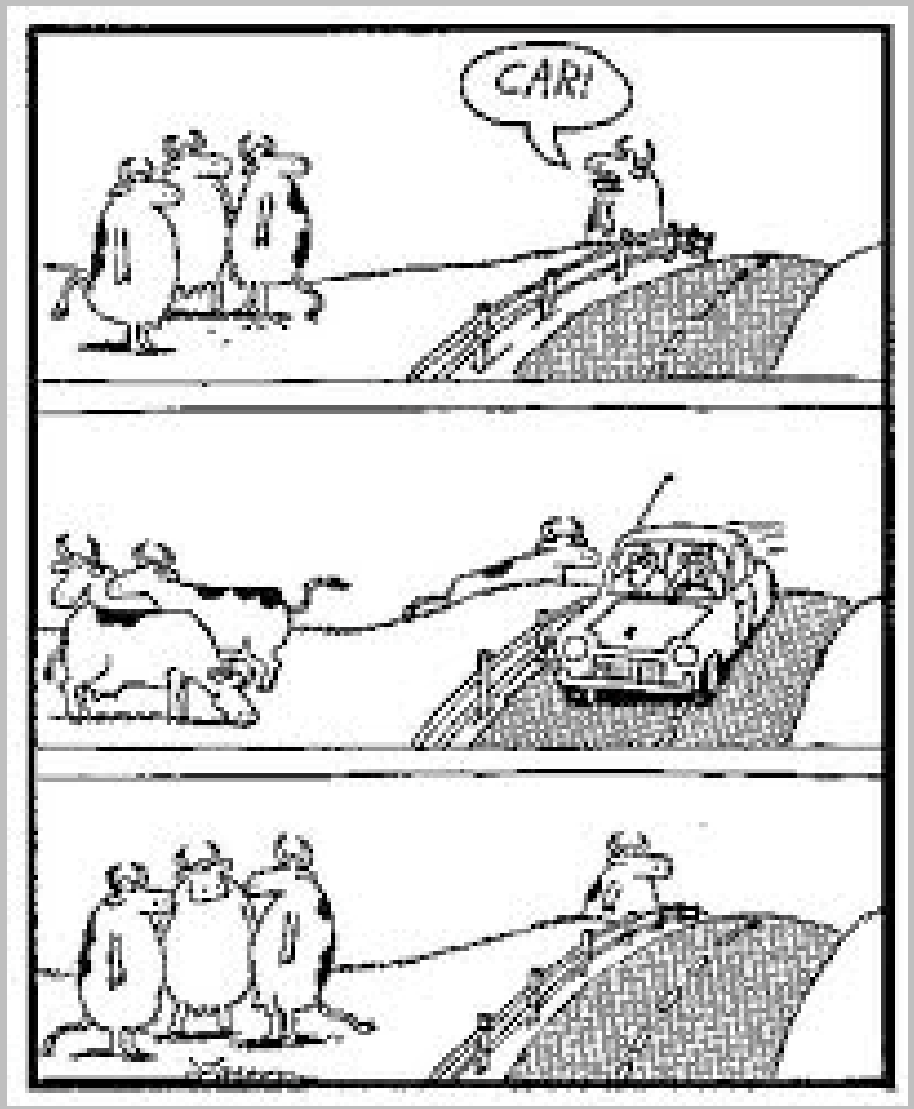
Mucinous Cystic Neoplasm (MCN)

- 2% of all pancreatic neoplasms
- Approx 30% invasive
- Malignancy:
  - Tumor size (>3cm)
  - + calcification
  - + mural nodules
  - Elevated CA 19-9
- Treatment
  - Resection – Distal pancreatectomy
  - Prognosis invasive MCN: 50% 5-year survival
  - Surveillance Imaging – invasive lesions only

Serous Cystadenoma

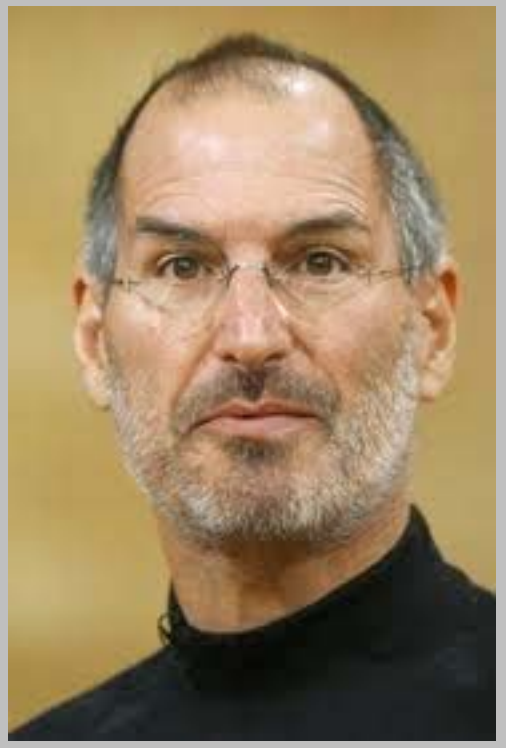
- Multiple serous fluid-filled cysts
- Variable size and location
- **No** presence of **mucin**
- Thin-walled capsule on CT
- *Sun-burst* calcifications
- Thin-walled *honeycombing* septae
- **No malignant potential**
- Resect only if symptomatic




CAR!

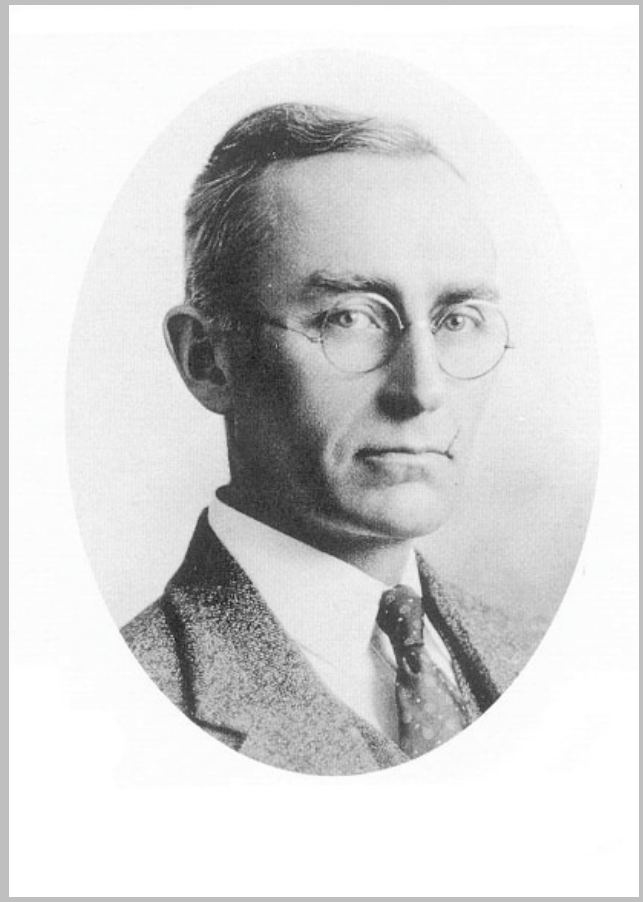
Pancreatic Islet Cell Tumors

- Insulinoma, Glucagonoma, VIPoma, Somatostatinoma, Gastrinoma
- 2500 cases/year
- Often associated with MEN-1 syndrome
- May be functional or non-functional



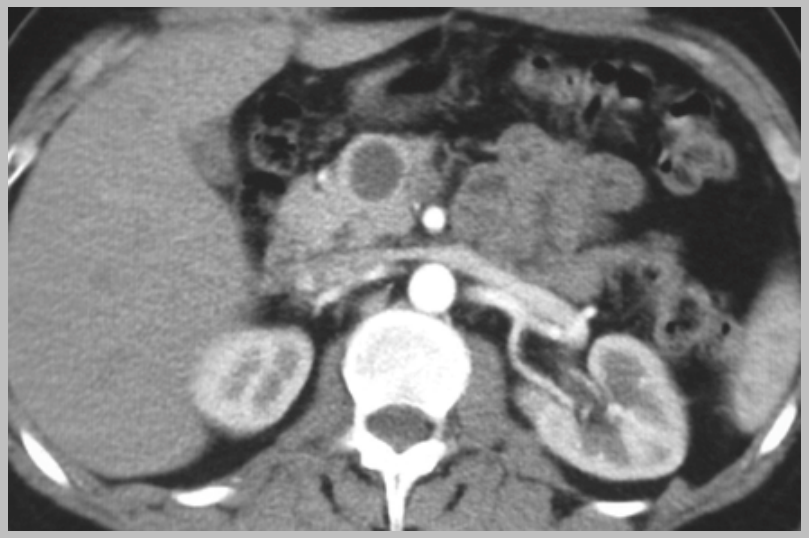
Insulinoma

- Most common islet cell tumor
- Secret Insulin ➔ hypoglycemia
- Diagnosis:
  - Whipple's triad:
    - + symptoms hypoglycemia
    - BG < 40mg/dl
    - Relief with glucose
  - Elevated serum insulin level
    - Insulin/Glucose > 0.3
    - Tolbutamide test
    - Glucagon Stimulation test



Insulinoma

- Imaging
  - CT/MRI (50% sensitivity)
  - EUS (80% sensitivity)
  - Intraoperative u/s (90% sensitivity)
  - **Rare detection** with Octreotide Radionucleotide scan

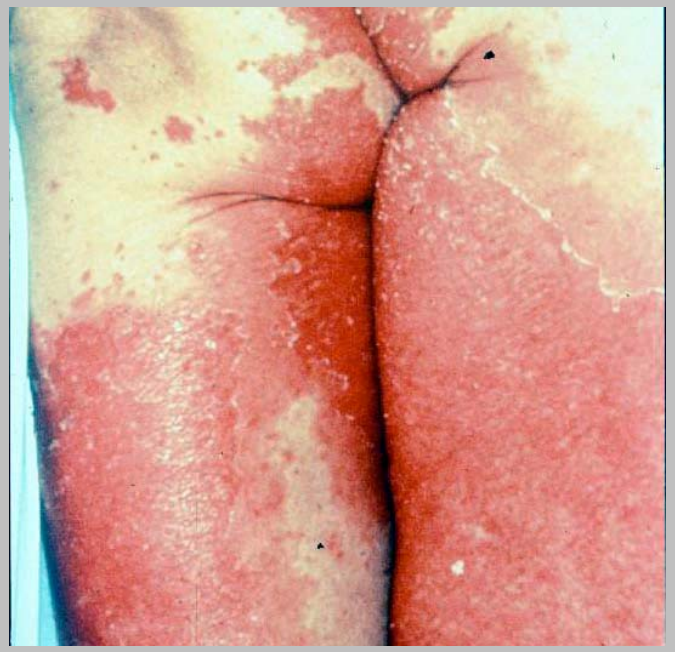


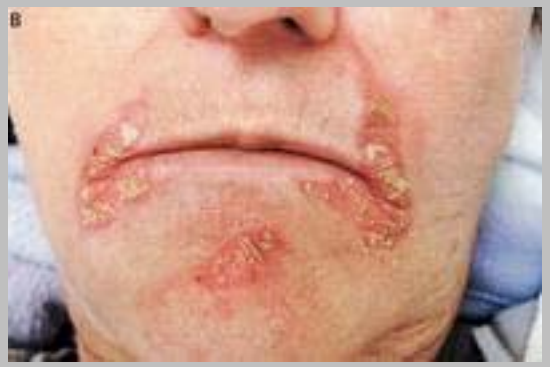
Insulinoma

- Tumor Characteristics
  - Usually < 2cm
  - Solitary; evenly distributed location
  - <3% outside pancreas
  - 90% benign
  - 5% associated with MEN-1
  - 97% 5-Year survival
- Treatment
  - Enucleation
  - Pre-op Diazoxide

Glucagonoma

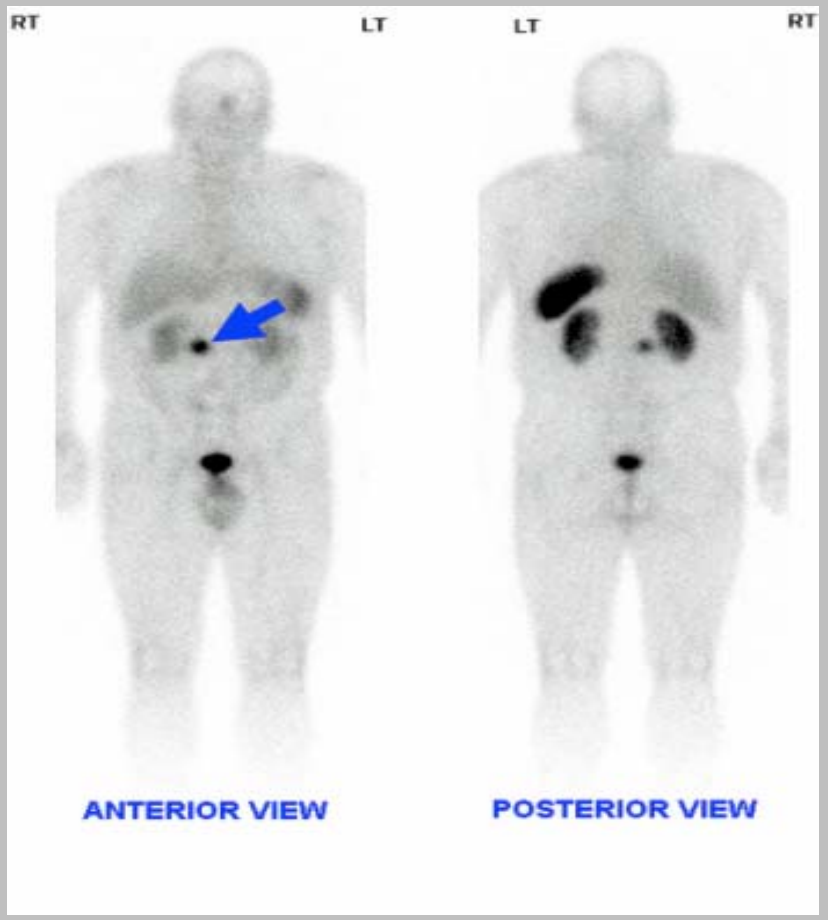
- Diagnosis:
  - 4D's
    - Diabetes (75-95%)
    - Dermatitis
    - DVT (30%)
    - Depression
  - Necrolytic migratory erythema
  - Serum Glucagon >500 pg/ml
  - Anemia, Cheilitis





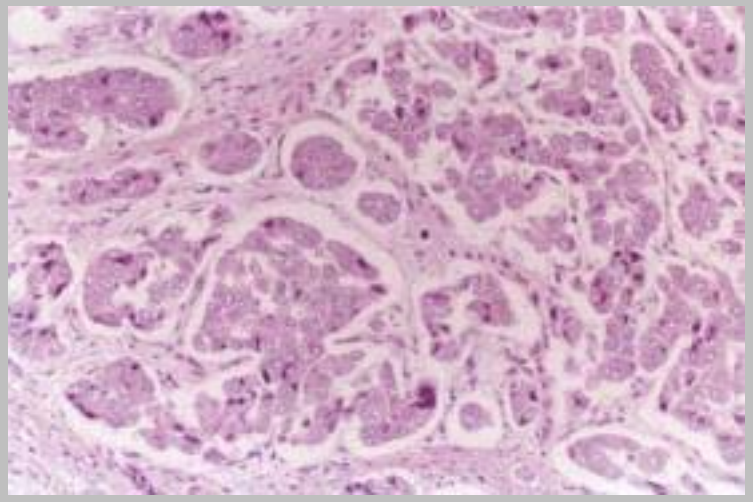
Glucagonoma

- Imaging
  - CT/MRI
  - Octreotide scan
    - ~ 75% sensitivity



Glucagonoma

- Tumor Characteristics
  - Usually >5cm at diagnosis, solitary
  - Location: body/tail
  - 50-80% malignant at diagnosis
  - < 5% multicentric or outside pancreas
- Treatment
  - Aggressive pre-op nutrition
  - Prophylactic heparin
  - Formal resection
  - 50-60% 5-year survival



VIPoma

- Clinical Features:
  - Rare, 1/10 million per year
  - WDHA Syndrome:
    - Watery Diarrhea
    - Hypokalemia
    - Achlorhydria
  - Abdominal pain, flushing, muscle weakness
  - VIP level > 200 pg/ml

VIPoma

- Tumor Characteristics:
  - Usually >3 cm at diagnosis, solitary
  - < 5% multicentric
  - Location: tail (< 10% outside pancreas)
  - 40-70% malignant
  - 60% metastatic at diagnosis
- Imaging: CT/MRI/Octreotide scan
- Treatment
  - Correct metabolic derangements
  - Octreotide, glucocorticoids
  - Resection

On This Day...

- On February 11, 1990 Nelson Mandela was freed after 27 years in prison for resisting apartheid.

Somatostatinoma

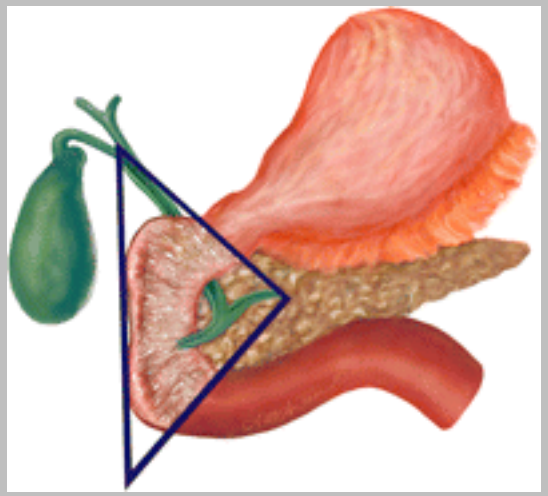
- Clinical Features:
  - Most rare islet cell tumor
  - Diabetes (60%)
  - Cholelithiasis (60%)
  - Steatorrhea
  - Somatostatin level > 10 ng/ml

Somatostatinoma

- Tumor Characteristics:
  - Usually >5cm at diagnosis, solitary
  - 70% located in pancreas (head)
  - 30-40% outside pancreas
  - 75% metastatic at diagnosis
- Imaging: CT/MRI/Octreotide scan
- Treatment:
  - Resection
  - 45-50% associated w/MEN-1
  - 40% 5-year survival

Gastrinoma

- Gastrin-secreting tumor
- Most commonly in duodenal wall, sporadically in pancreas
- 80% arise within triangle, but often multicentric
- > 50% are malignant at diagnosis
- 25% associated with MEN-1
- ZES – triad:
  - Gastrinoma
  - Hypergastinemia
  - Refractory PUD
- Clinical indicators:
  - Refractory/Recurrent ulcers
  - Diarrhea
  - Pernicious Anemia



Gastrinoma

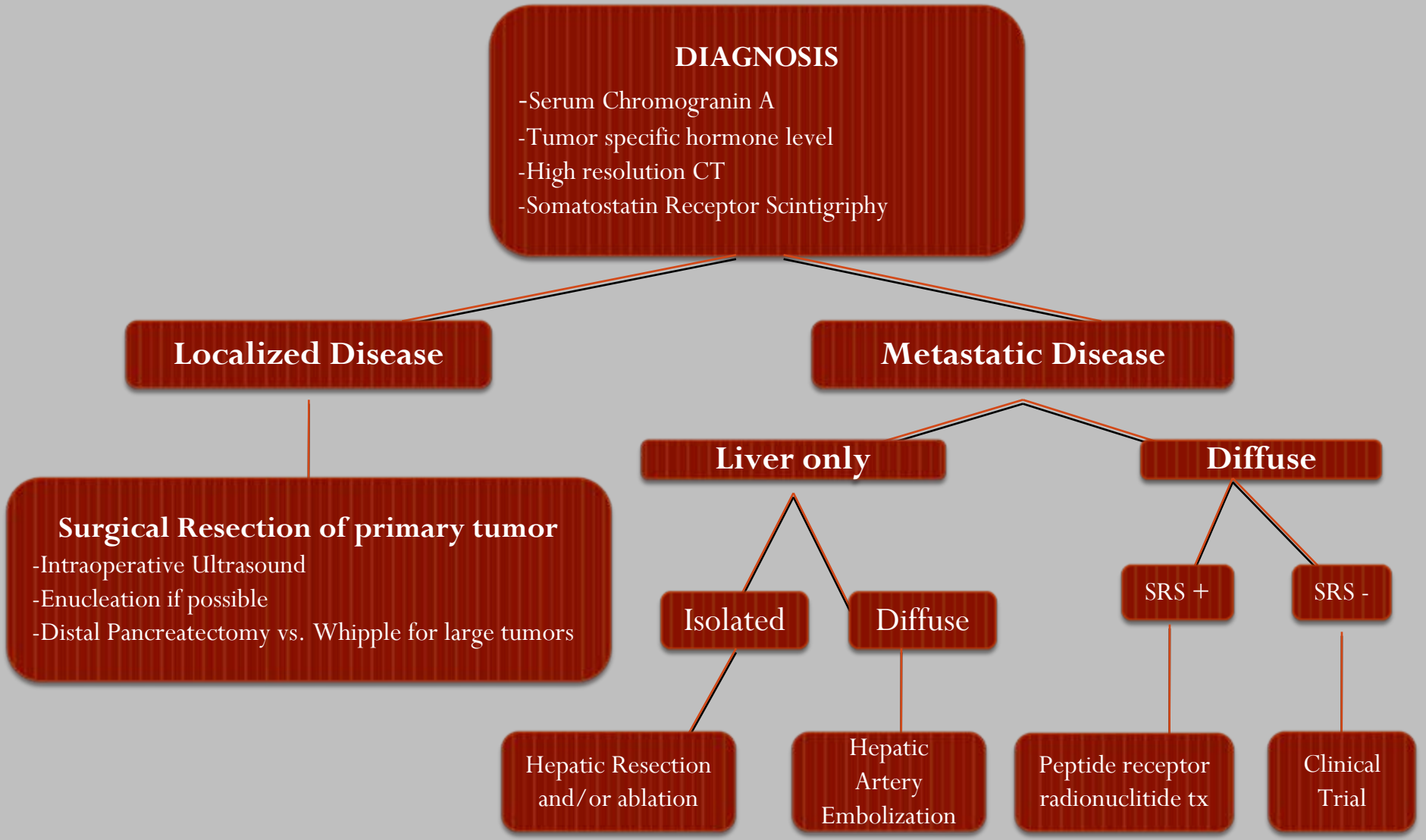
- Diagnosis:
  - Fasting Serum Gastrin > 150 pg/ml
  - Basal Acid Output > 15 meq/hr
  - Gastric pH > 3 excludes Gastrinoma
  - **Secretin Stimulation test**
  - Serum Chromogranin A
- Imaging:
  - Somatostatin Receptor Scintigriphy
  - EUS
  - CT/MRI

Gastrinoma

- Treatment
  - Medical Options
    - PPI, H2 blockers
    - Chemo for metastatic disease
  - Surgical Options
    - Resect localized disease
    - Exploratory Laparotomy (unidentified lesions)

Mr. Osborne, may I be excused? My brain is full!

Surgical Treatment

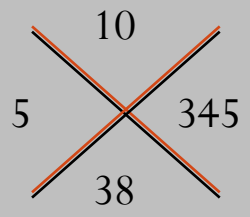


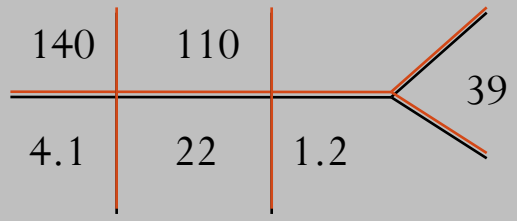
Case Presentation

- 55 yo male with HTN and GERD is admitted to the trauma unit after an MVC. He sustained multiple orthopedic injuries, but otherwise is ok. Prior to his accident he had been experiencing episodes of dizziness with occasional blurred vision.
- He gives no history of prior operations, and no family history of malignancy.
- Social: Occasional alcohol, no tobacco
- Meds: Omeprazole, Lisinopril

Case Presentation

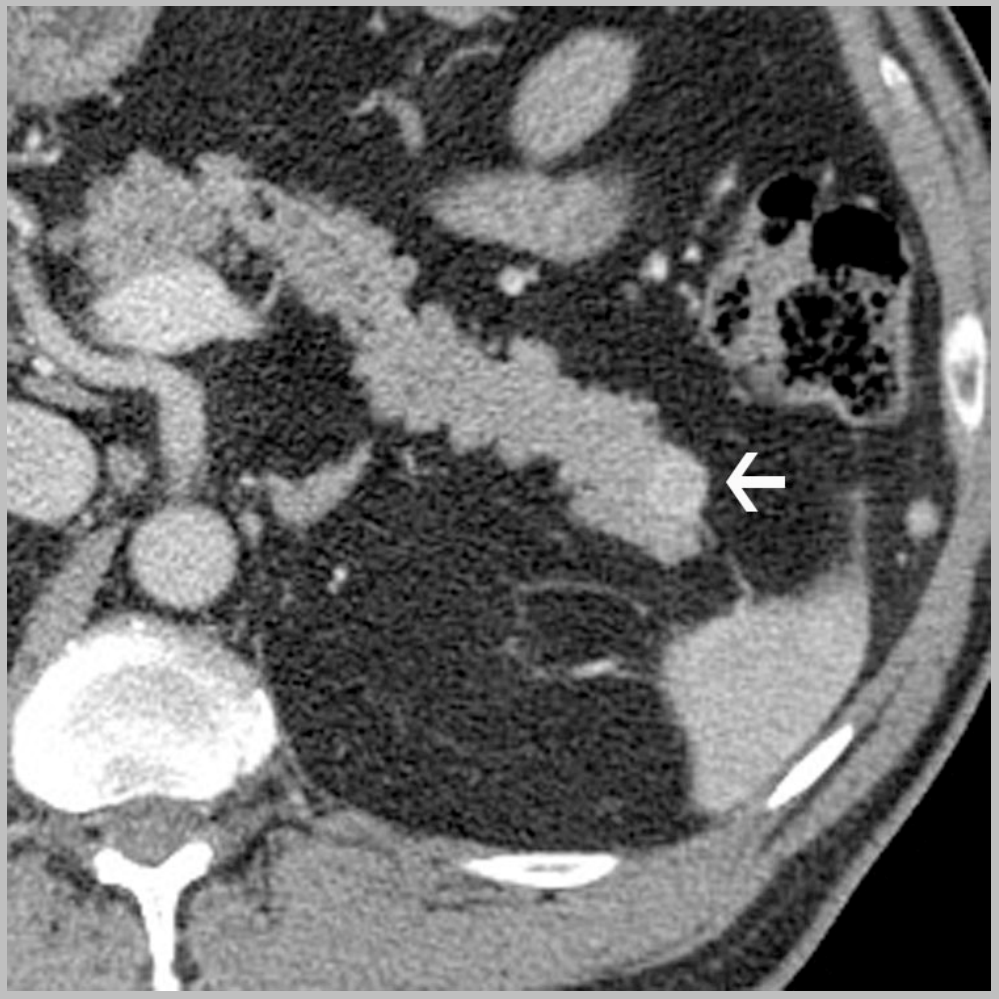
- PE:
  - Alert, no distress
  - Lungs clear, RRR
  - Abdomen soft, nontender, no masses
  - RLE in external fixation
  - No rash
- Labs:





Case Presentation

- Traumagram:



Differential Diagnosis

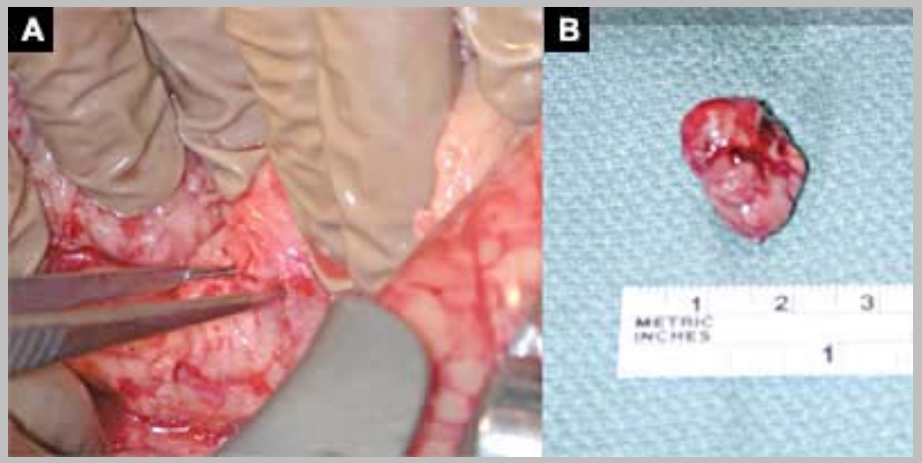
- Pancreatic Pseudocyst
- Islet Cell Tumor
  - Insulinoma
  - Glucagonoma
  - VIPoma
  - Somatostatinoma
  - Gastrinoma
- IPMN
- MCN
- Adenocarcinoma
- Metastatic Disease
- Lymphoma
- Abscess
- Congenital cyst

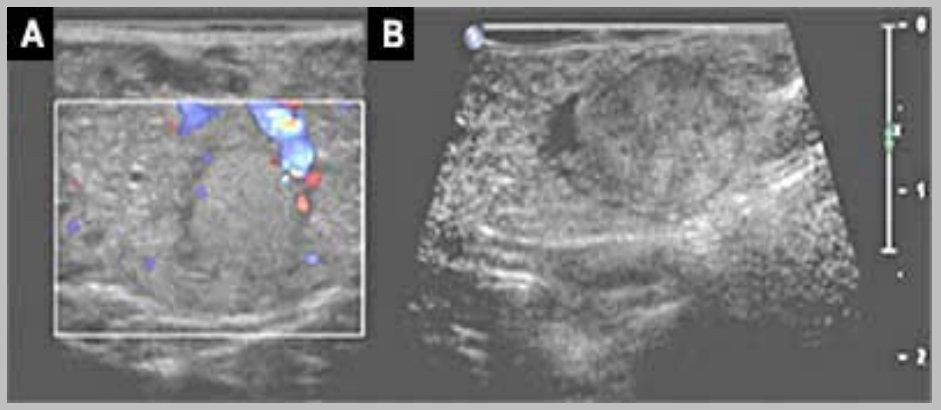
Case Presentation

- Diagnosis
  - Medical Evaluation
    - 72-hour Fast:
      - Serum Insulin
      - Glucose
      - C-peptide
  - Radiographic Evaluation
    - CT demonstrated 1.2cm mass distal pancreas
    - Additional studies?

Case Presentation

- Treatment
  - Preoperative Therapy
    - Diazoxide
    - Octreotide
    - Careful monitoring of glucose level
    - D10 infusion perioperatively
  - Surgical Therapy
    - Solitary vs. Multicentric (MEN-1)
    - Intraoperative ultrasound
    - Enucleation





Question 1

- A 43 yo female undergoes EGD for symptoms of early satiety and GERD. Mucus is seen extruding from the ampulla. CT shows pancreatic mass. All of the following are true regarding this lesion except:
  - A) It communicates with the main pancreatic duct
  - B) Presence of mural nodules increases risk of malignancy
  - C) It contains a fibrous capsule
  - D) If < 3cm the lesion may be observed
  - E) Prognosis is worse with proximal lesions

Question 2

- True or False:
  - Approximately 30-40% of Mucinous Cystic Neoplasms are invasive, while Intraductal Papillary Mucinous lesions are always benign.
    - True
    - False

Question 3

- All of the following characteristics of Insulinomas are true except:
  - A) They are usually solitary, but vary in location
  - B) They are often associated with MEN-1
  - C) Diagnosis is suggested by Whipple's triad
  - D) Preferred treatment is enucleation
  - E) Majority of lesions are benign

Question 4

- A 51 yo male presents with a recurrent gastric ulcer despite PPI therapy and persistent diarrhea. Of the following, which is not associated with his condition?
  - A) Vitamin B12 deficiency is common
  - B) > 50% of these lesions are malignant
  - C) Somatostatin Receptor Scintigriphy localizes lesion
  - D) Distal pancreas is most common location
  - E) A rise in serum gastrin follows secretin challenge
  - F) Don't have a clue

Questions