Adult-onset Nesidoblastosis

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Adult-onset Nesidoblastosis

- Objective:
  1. Discuss how to diagnose Insulinoma
  2. Understand the localization studies for Insulinoma
  3. Adult-onset Nesidioblastosis

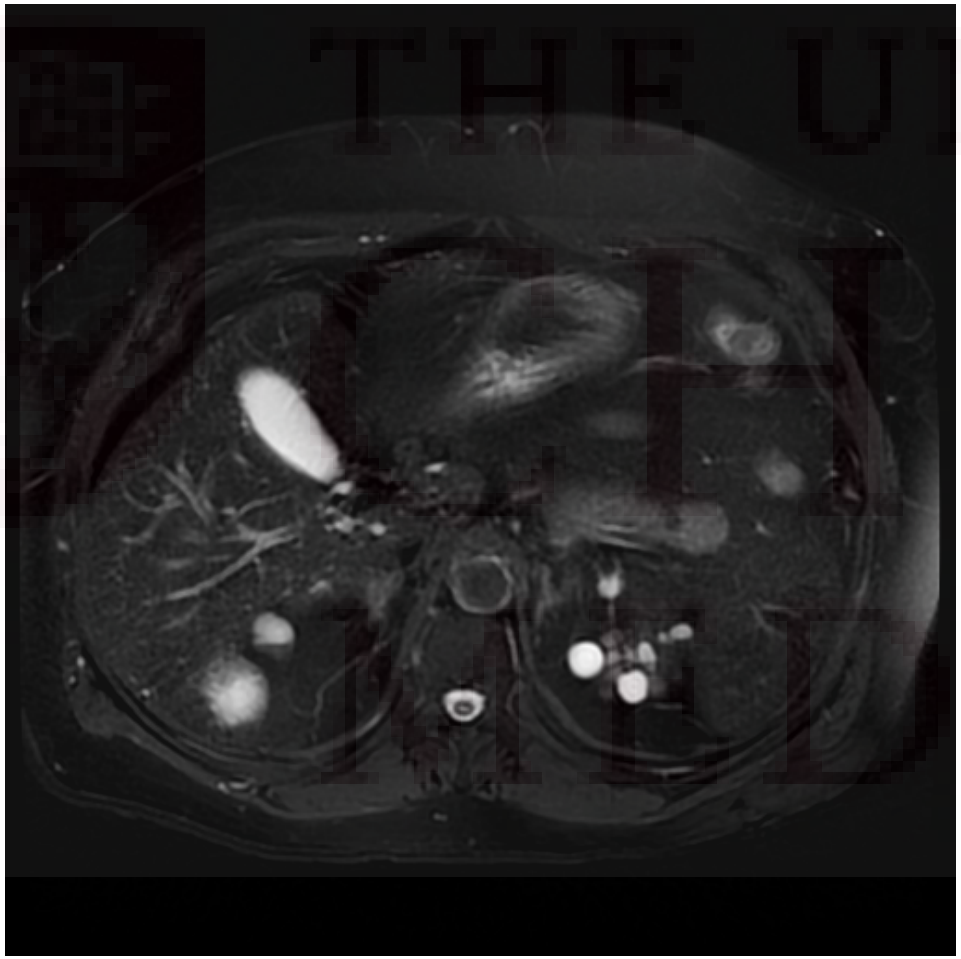
GA

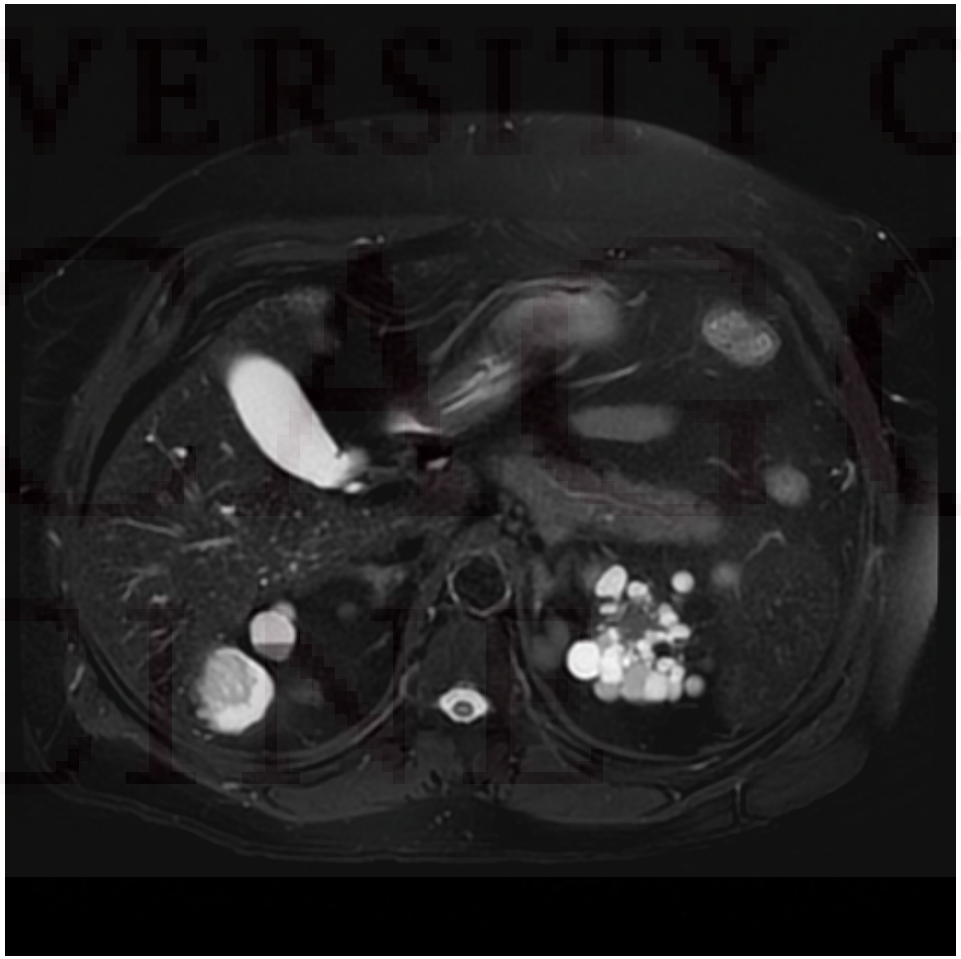
- **Chief complain:** light headed/sweating/palpitation with fasting
- **HPI:**
  - 70 yrs old M with neuroglycopenic sx for 5 years.
    - W/U found to have Low serum glucose/Elevated C-peptide/Insulin.
  - PMH:
    - HL, HTN, PCKD, 2nd hyperparathyroidism, OSA.
  - PSH:
    - Tonsillectomy, AV fistula.
  - Meds:
    - Sevelamer, Octreotide, Simvastatin, Cinacalcet, Alprazolam.

GA

- Physical Exam:
  - Unremarkable
- Lab Ix:
  - C-peptide: >13.pmol/ml (Ref: 0.3- 2.35)
  - Pro-insulin: 310 (Ref: 3-20)
  - Insulin: 110 mcu/ml (Ref: 2.6-24.9)
  - Glucagon: 84
  - Sulfonylurea panel negative

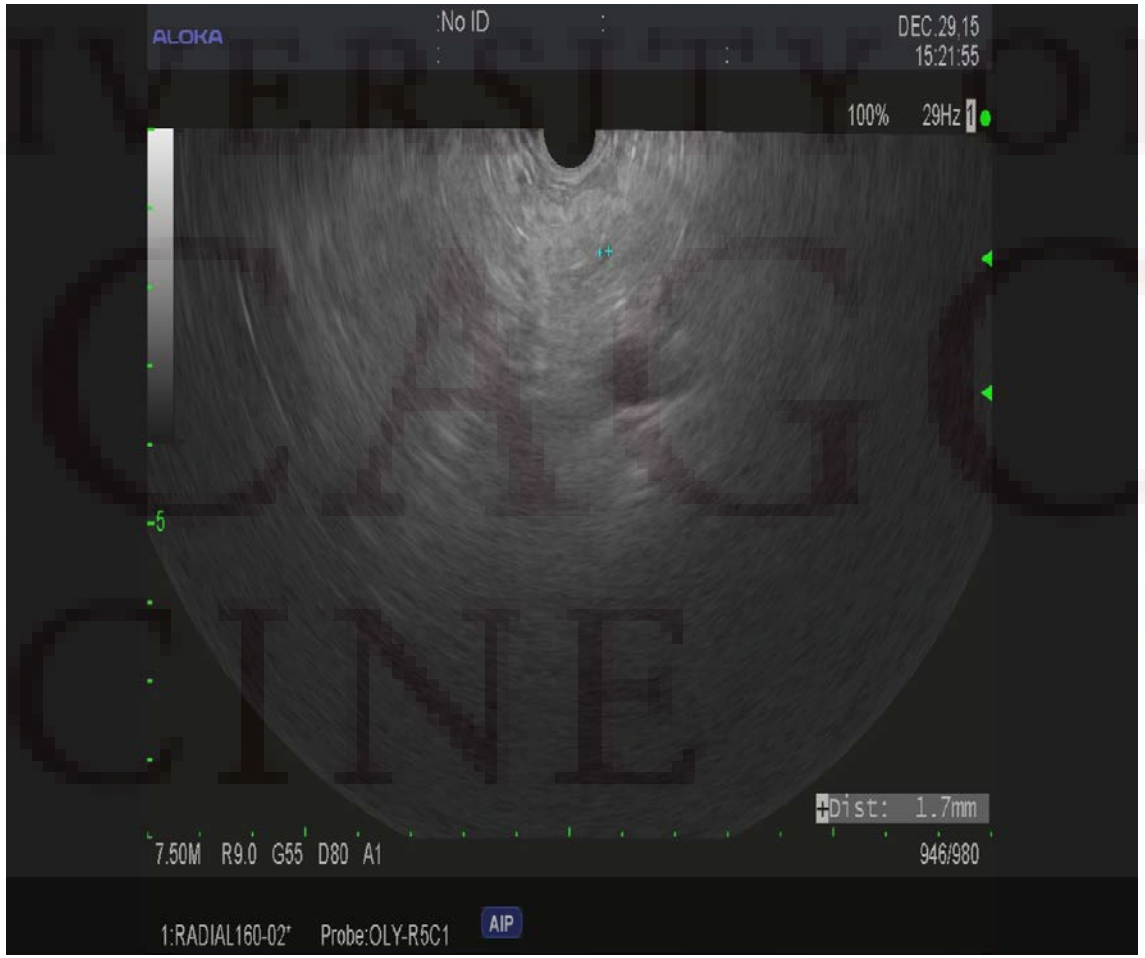
Imaging



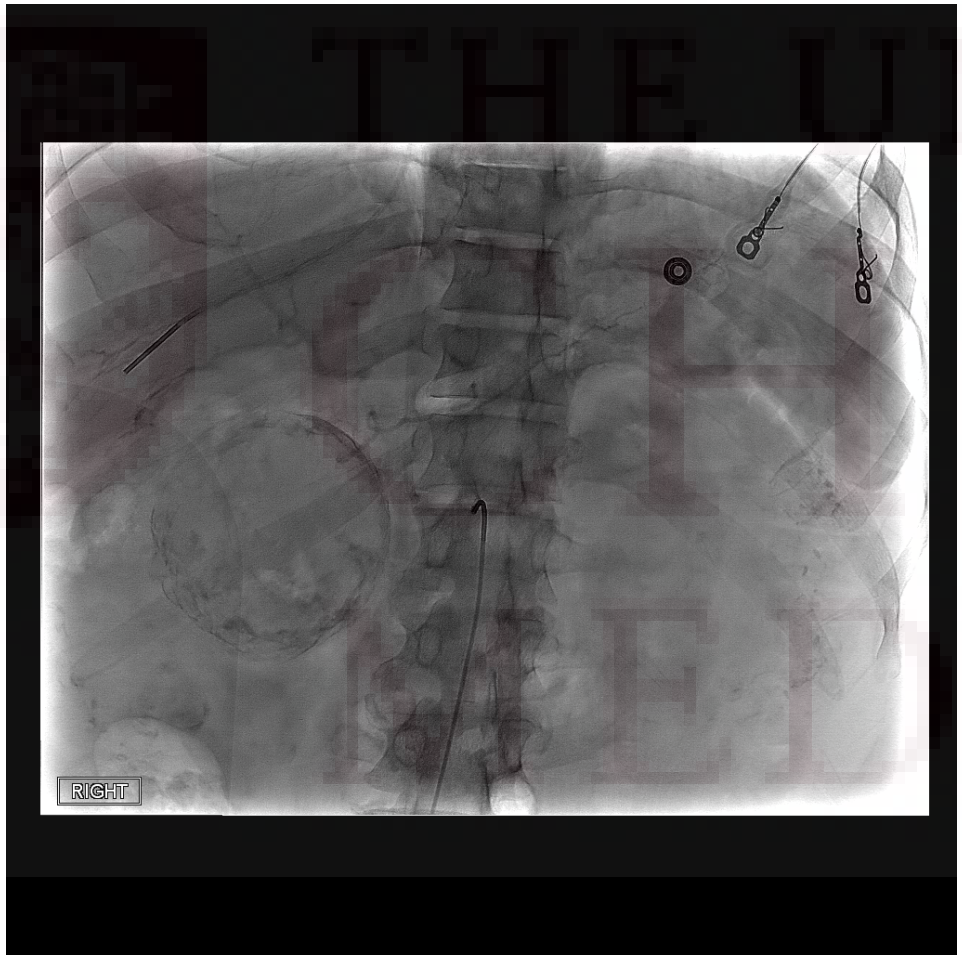


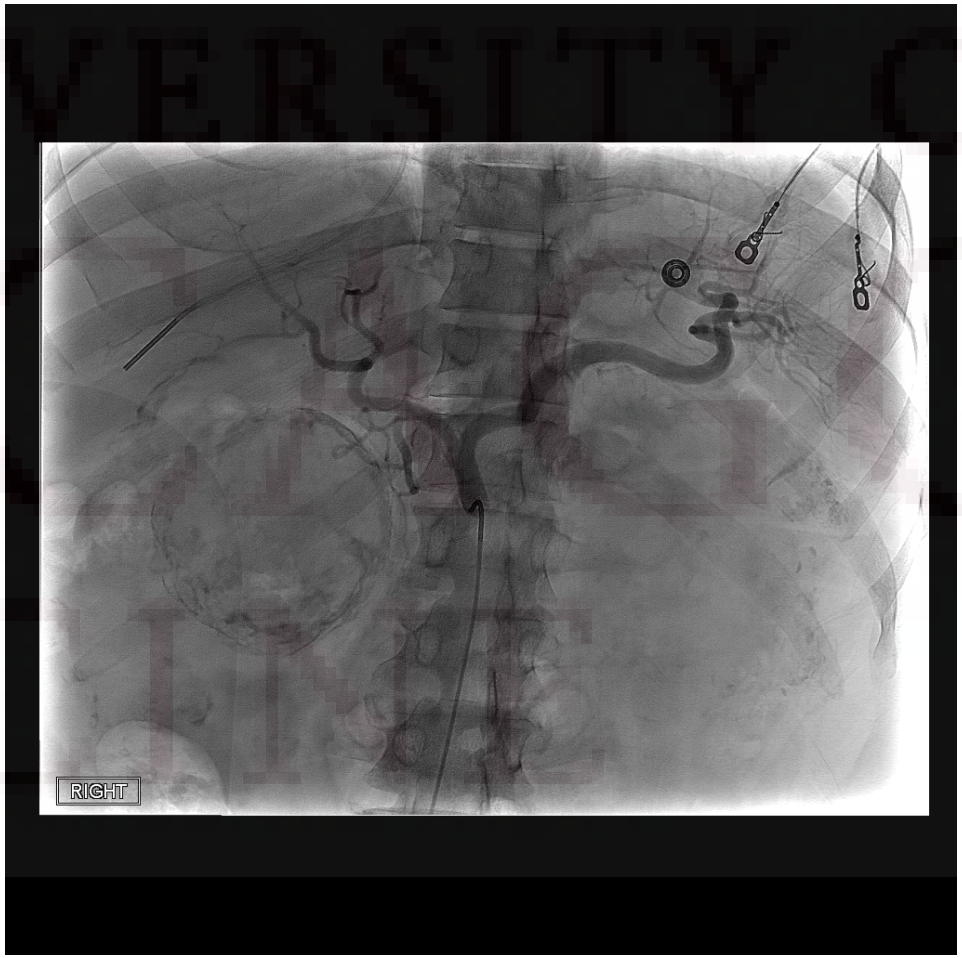
EUS

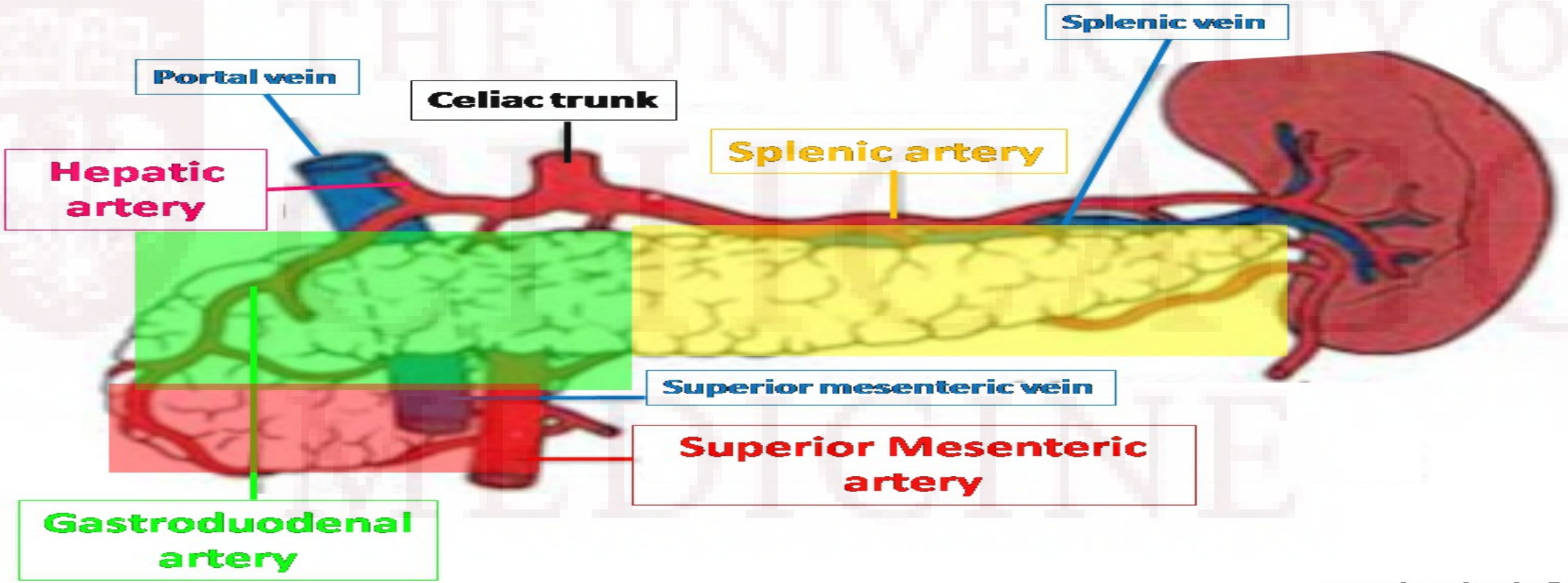
- No endosonographic evidence of pancreatic mass



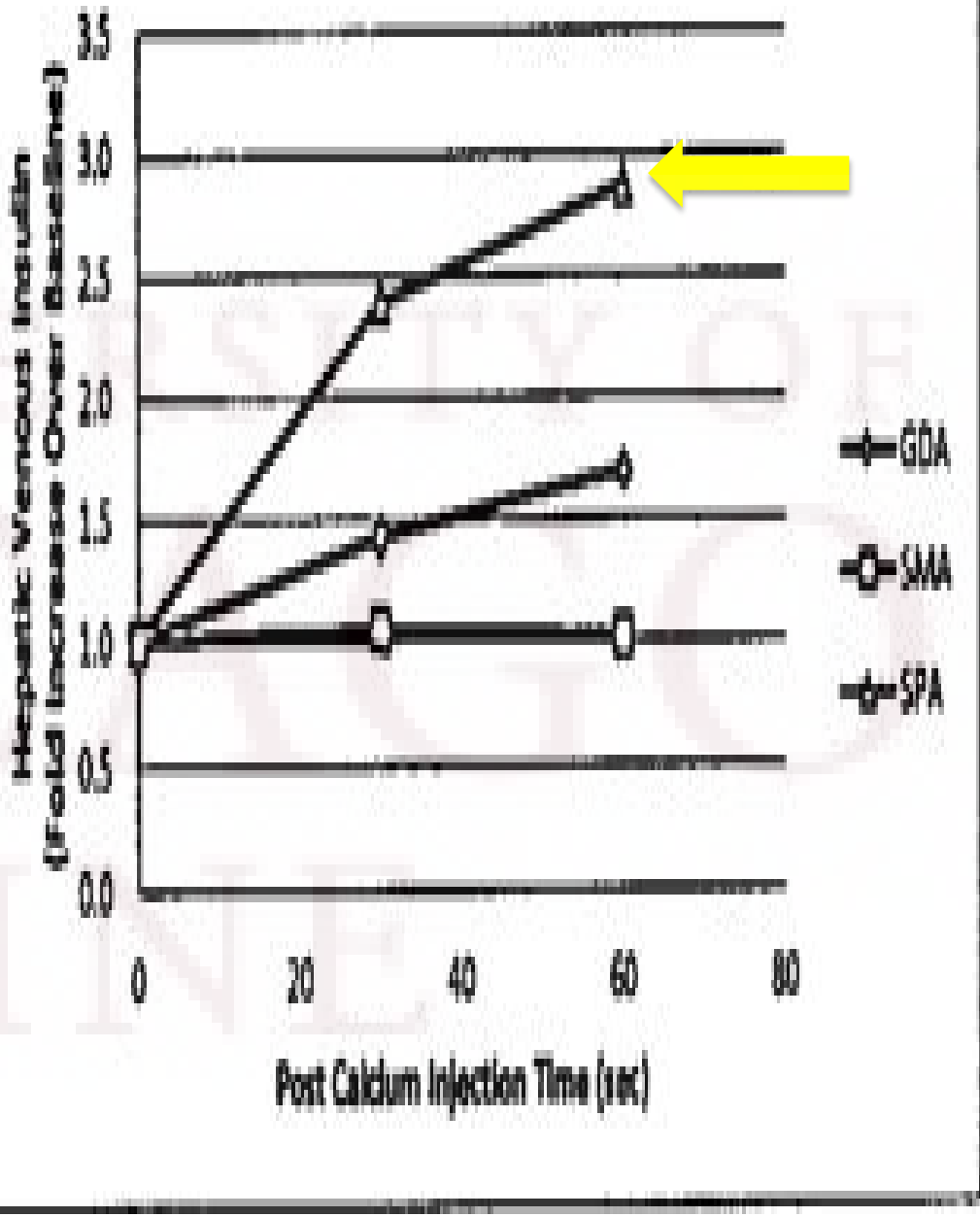
Calcium Stimulation test






Splenic vein
Portal vein
Celiac trunk
Hepatic artery
Splenic artery
Superior mesenteric vein
Superior Mesenteric artery
Gastroduodenal artery
www.bondysalud)

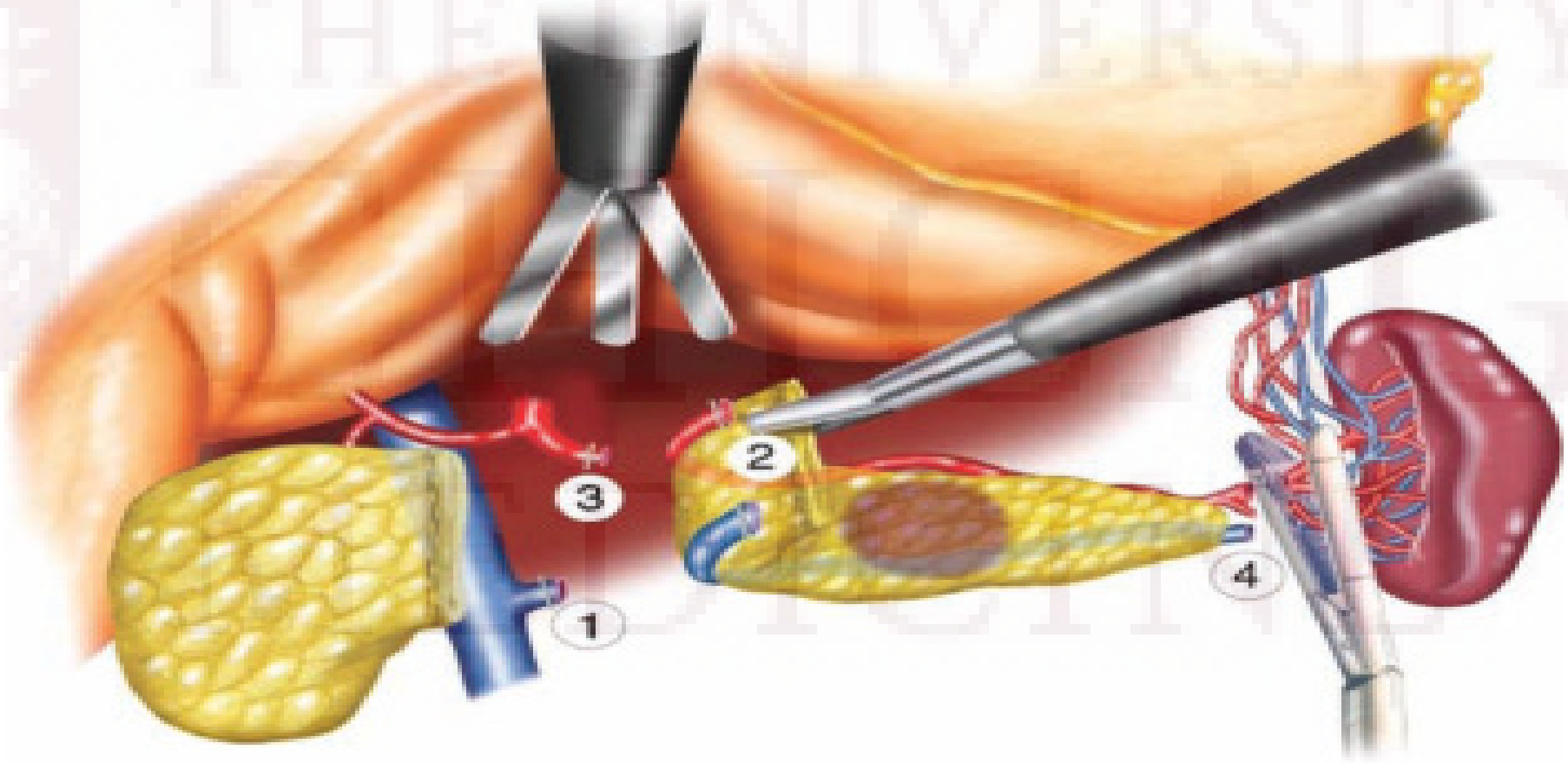
				MEASURED VALUES	CALCULATED VALUES
Time Point	Draw Time	Accession #	Location	Insulin (μIU/mL)	Fold Increase over baseline
0	12:25:00	H14285	Gastroduodenal Artery (GDA1)	40.2	1.0
30	12:26:00	H14298	Gastroduodenal Artery (GDA2)	57.5	1.4
60	12:27:00	H14314	Gastroduodenal Artery (GDA3)	68.3	1.7
			Gastroduodenal Artery (GDA4)		
0	12:30:00	H14291	Superior Mesenteric Artery (SMA1)	51.9	1.0
30	12:31:00	H14307	Superior Mesenteric Artery (SMA2)	54.6	1.1
60	12:32:00	H14321	Superior Mesenteric Artery (SMA3)	52.7	1.0
			Superior Mesenteric Artery (SMA4)		
0	12:17:00	H14296	Splenic Artery (SPA1)	26.9	1.0
30	12:18:00	H14310	Splenic Artery (SPA2)	64.3	2.4
60	12:19:00	H14330	Splenic Artery (SPA3)	77.3	2.9
			Splenic Artery (SPA4)		



GA

- Operation 10/18/17
  - Robotic splenic sparing distal pancreatectomy.
- Post-operative course:
  - Uneventful d/c to rehab HD # 9
  - Lab: C-peptide **2.25** pmol/ml

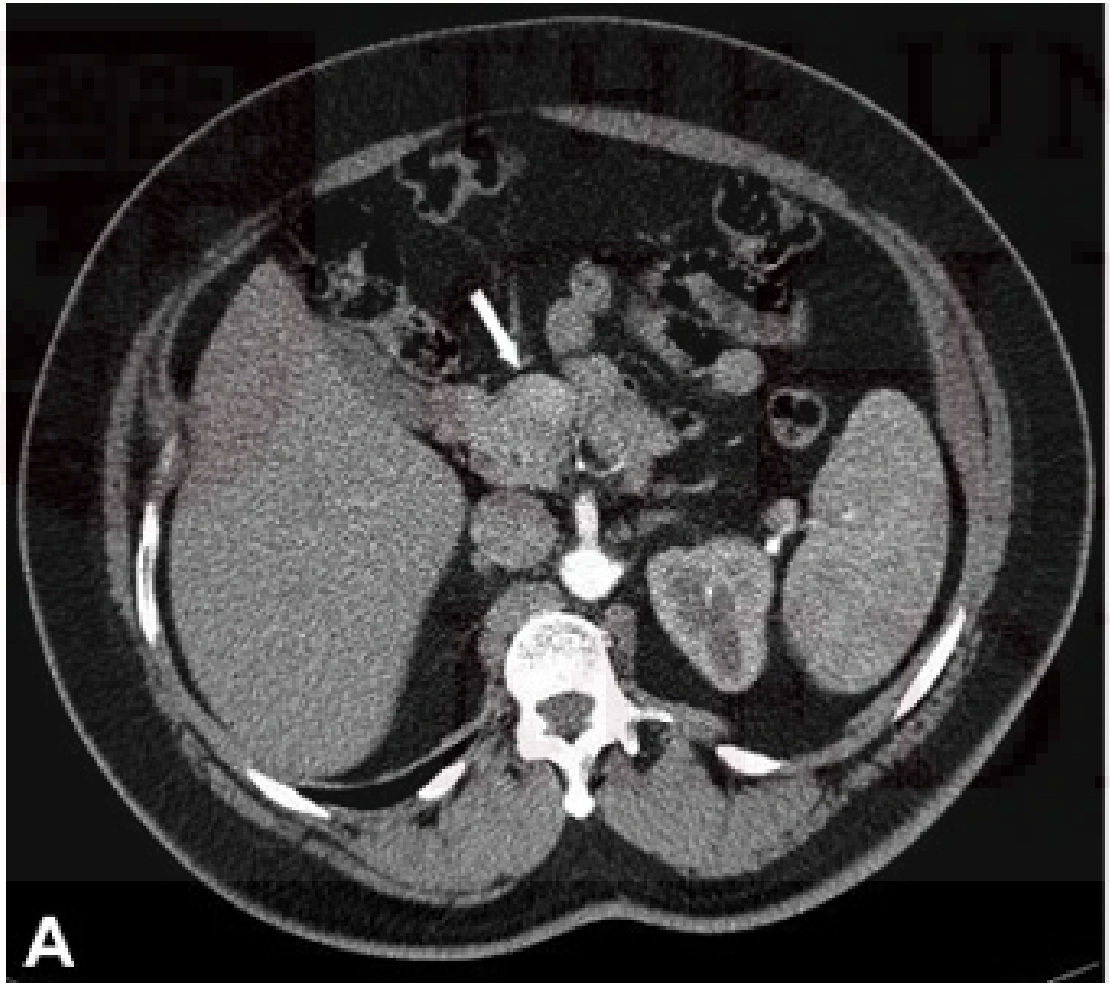
GA

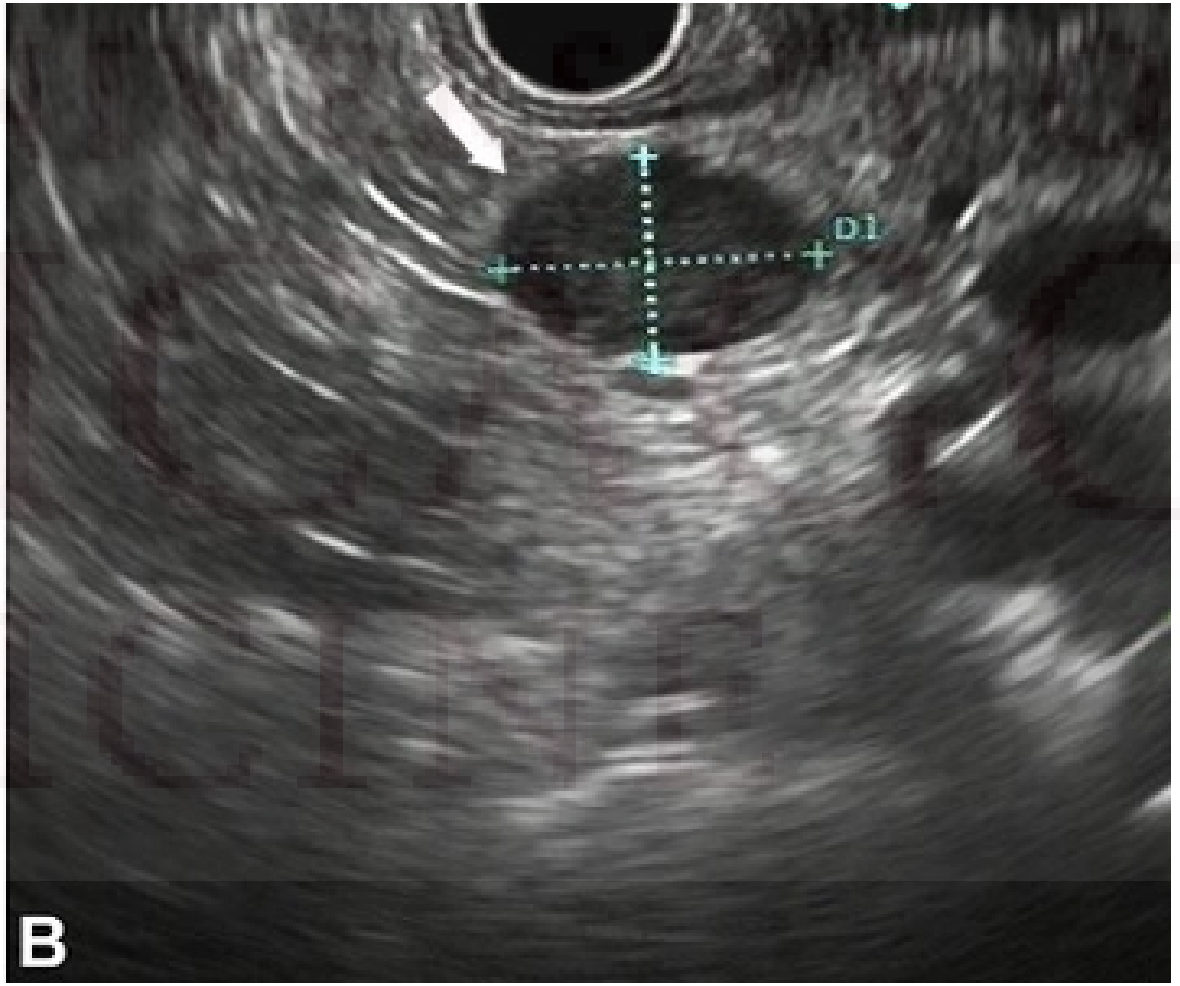


INSULINOMA

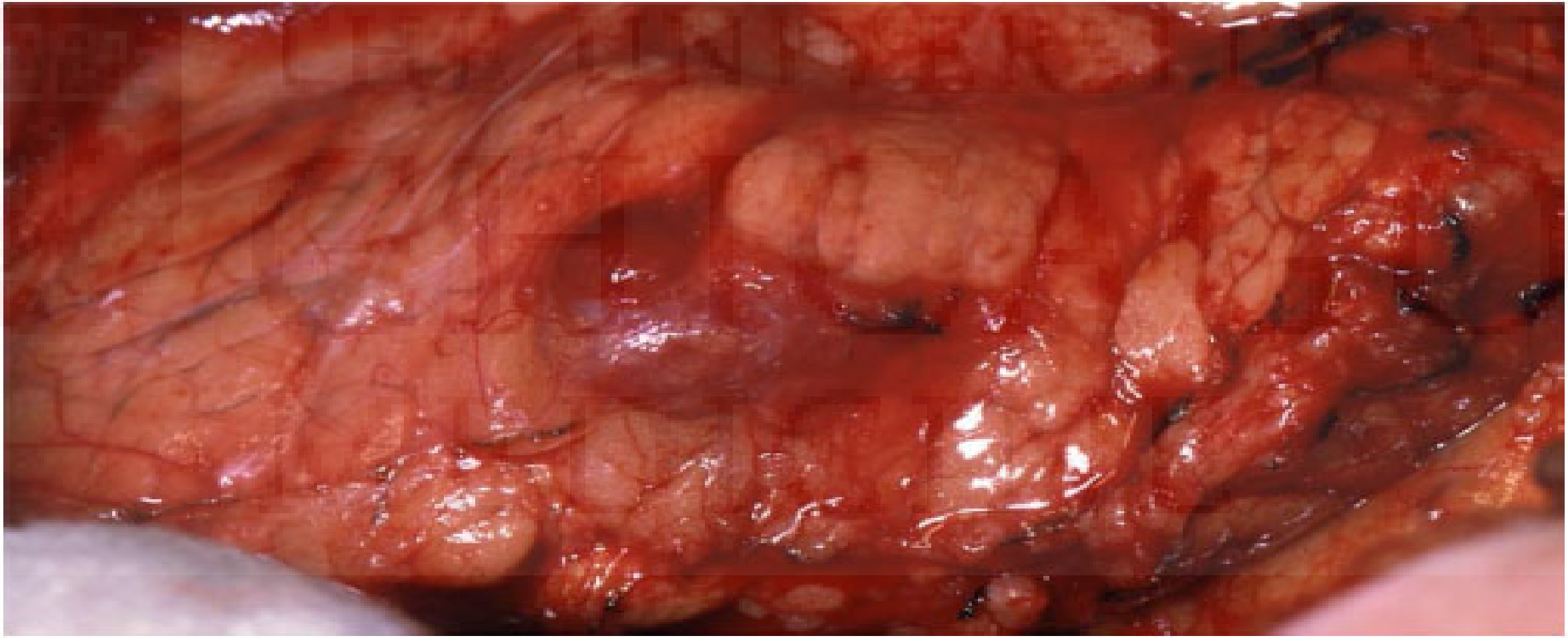
- Neuroendocrine tumor
  - Excessive secretion of insulin
  - Sporadic vs syndromic (MEN)
  - Sx Whipple's triad
    - Hypoglycemia during fastin/excercis
    - Blood sugar < 45 mg/dl
    - Sx improved with administration of glucose
  - Diagnosis:
    - Low serum glucose
    - Elevated Serum Insulin/pro-insulin/c-peptide
    - Negative sulfonyl panel test

INSULINOMA





INSULINOMA



NESIDIOBLASTOSIS

- Hyperinsulinoma hypoglycemia
- First described by George Laidlaw in 1938
  - Greek words “nesidion” islet and “blastos” builder
    - Neoformation of Langerhans islets from pancreatic duct epithelium
  - Most common cause of neonatal hyperinsulinemia

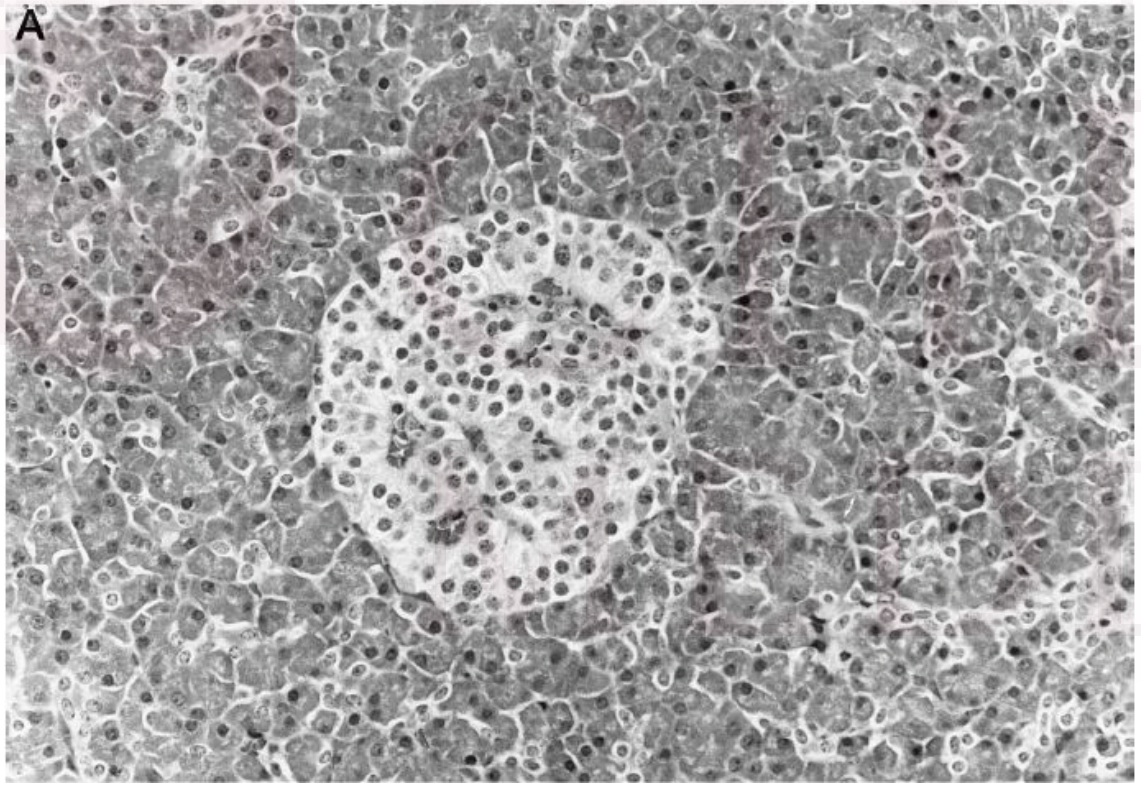
- Adult-onset Nesidoblastosis
  - First described in 1975 by Sandler
  - 0.5% to 7% of all cases of hyperinsulinemic hypoglycemia
  - 16% at the University of Chicago

NESIDIOBLASTOSIS

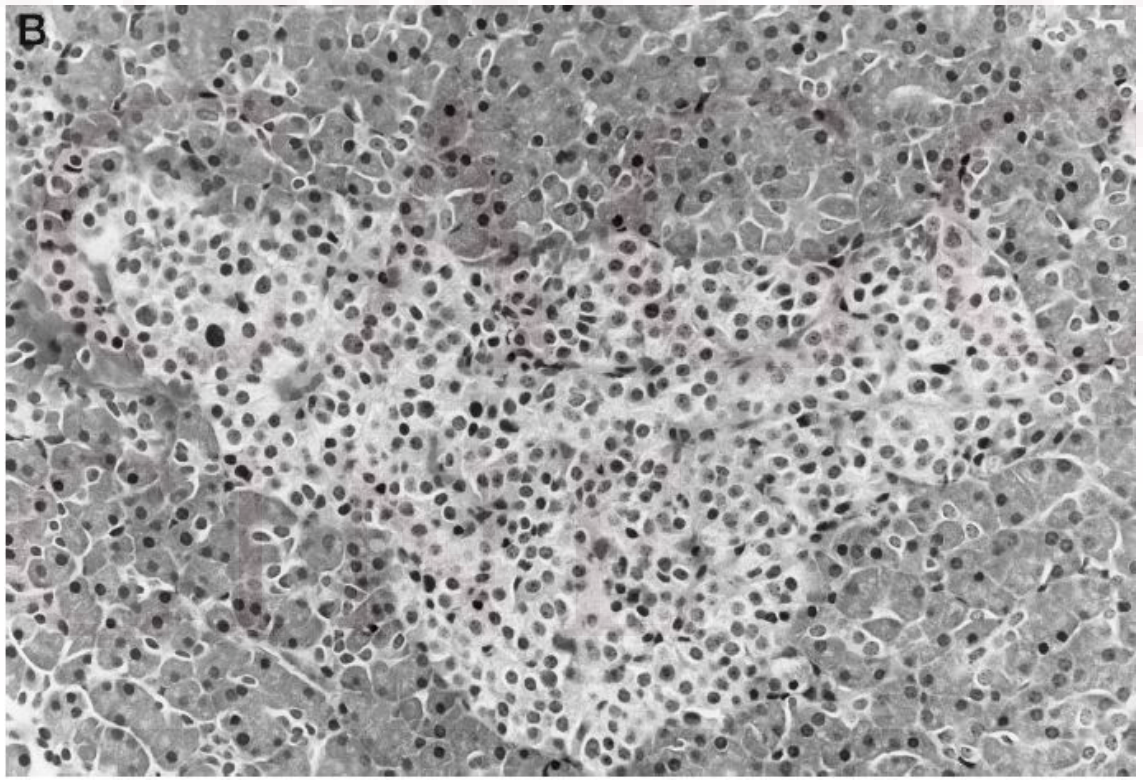
- Aetiology:
  - Unknown
  - 2 genetic alteration SUR1 and Kir 6.2 on Chromosome 11
    - Code fro Potassium channel on Beta cell membrane
  - Bariatric population after “Roux-en-Y gastric bypass”
- Symptoms:
  - Similar to Insulinoma; neuroglycopenic/adrenergic sx with fasting.
- Diagnosis of adult-onset Nesidioblastosis:
  - Hyperinsulinoma
  - Exclusion of insulinoma (Imaging, Macroscopic, Microscopic, IHC)
  - Pathological features of Nesidioblastosis

NESIDIOBLASTOSIS

Normal pancreatic islet

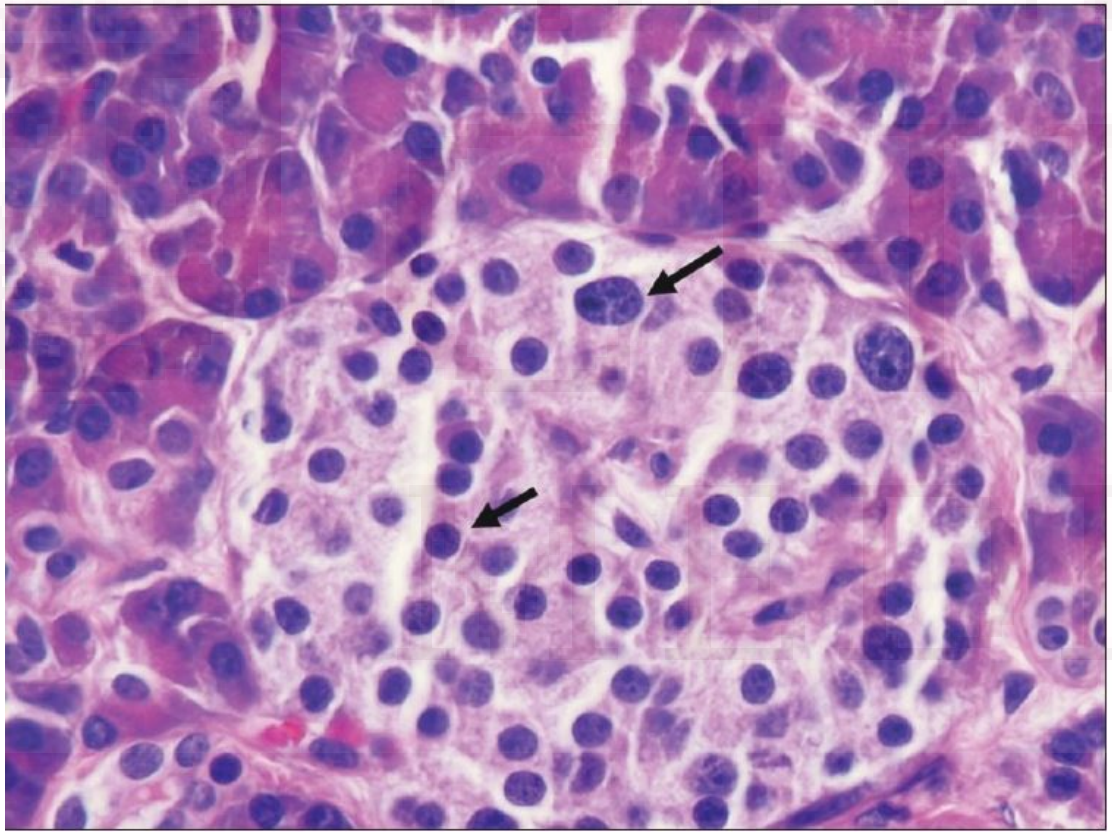


Hypertrophic islet

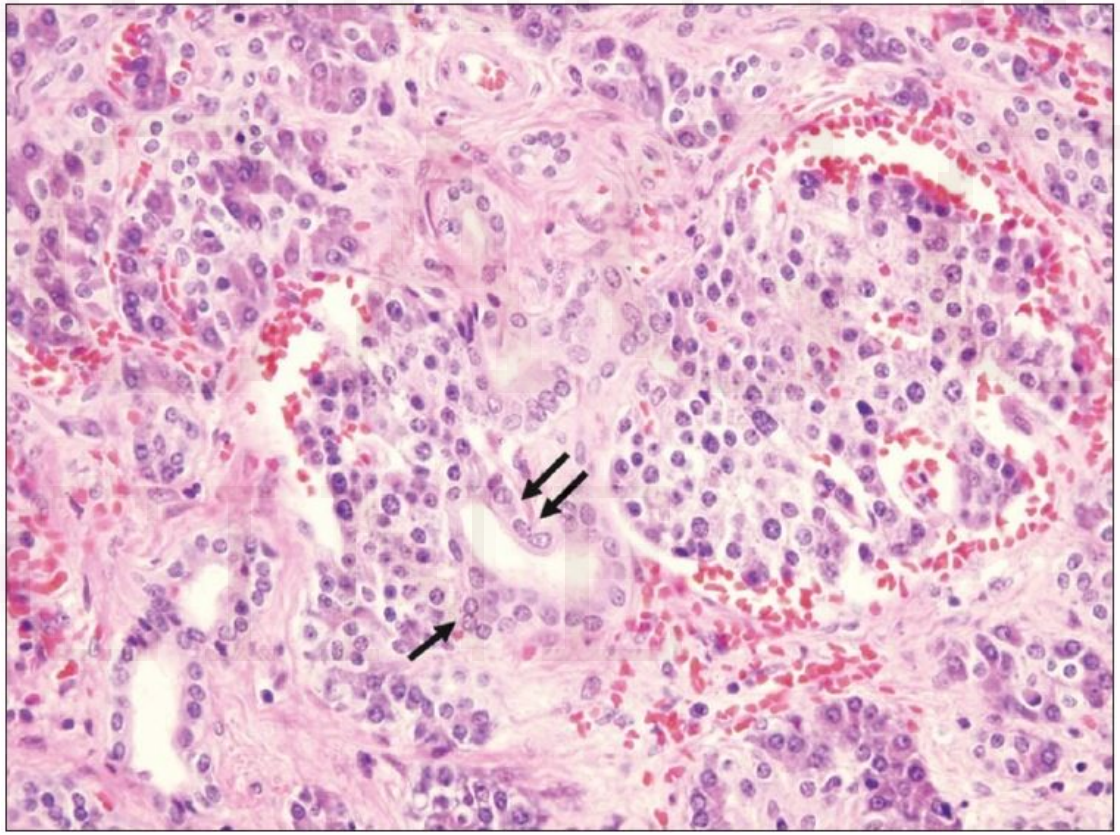


NESIDIOBLASTOSIS

Islet cell pleomorphism and hyperchromatic islet cell nuclei



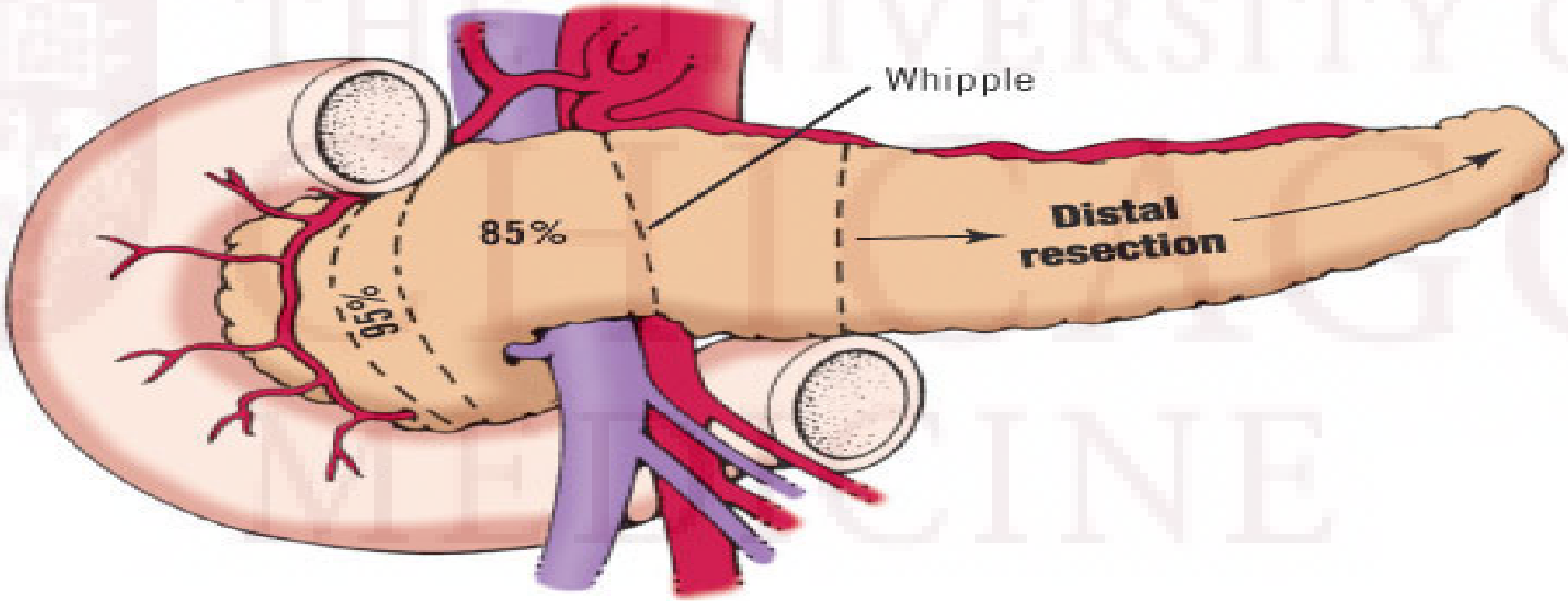
Ductal Insular complex



NESIDIOBLASTOSIS

- Management
  - Surgical:
    - Near total pancreatectomy (90-95%)
      - Insulin dependent DM 40%.
    - Distal pancreatectomy (60-80%)
  - Medical treatment
    - Diazoxide
    - Somatostatin
    - Calcium channel-blockers

NESIDIOBLASTOSIS



NESIDIOBLASTOSIS

Questions ?