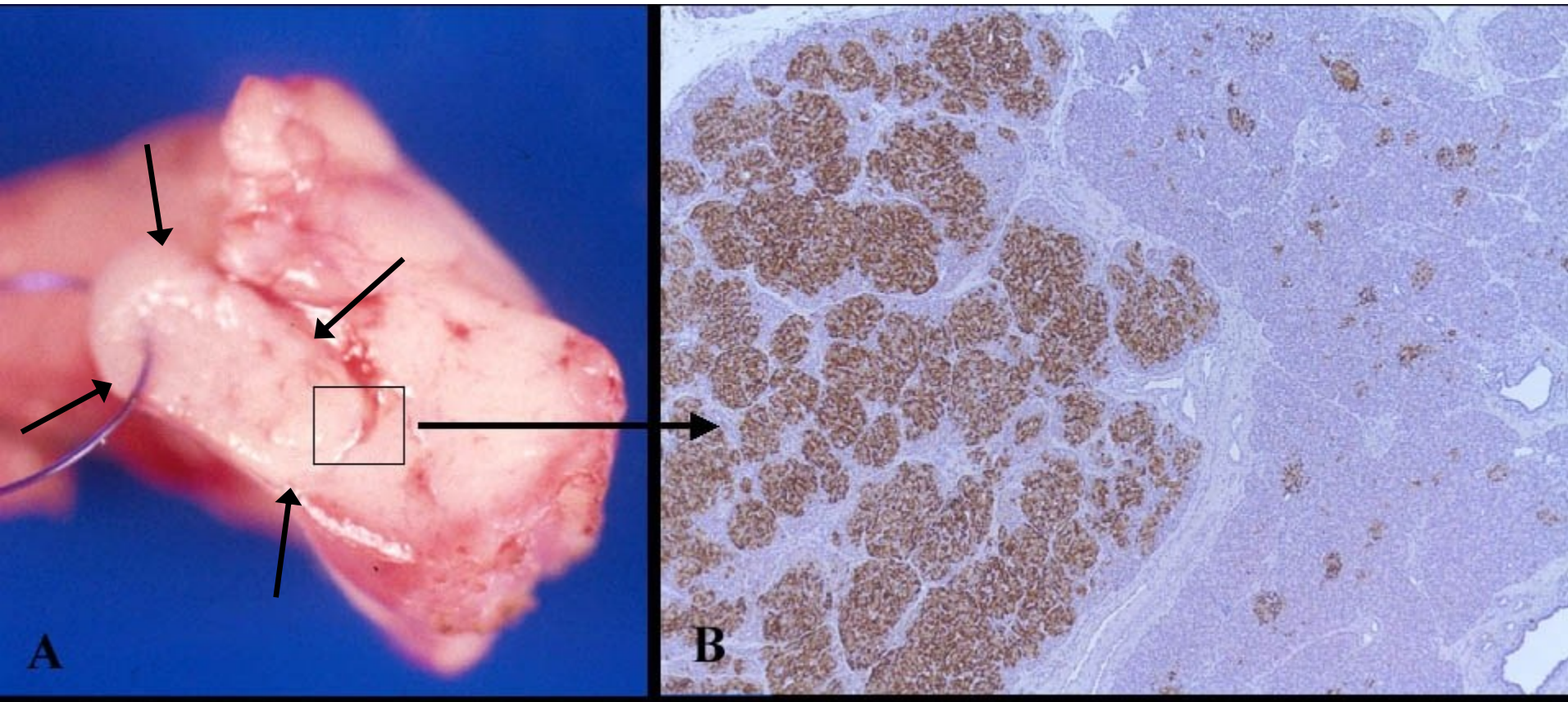
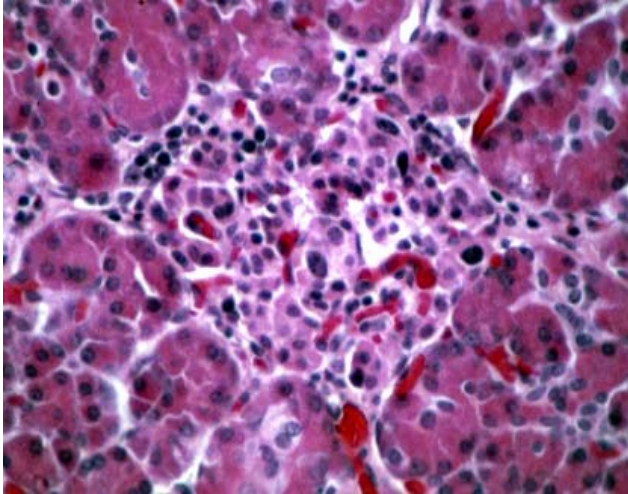


Partial Pancreatectomy for Focal Hyperinsulinism

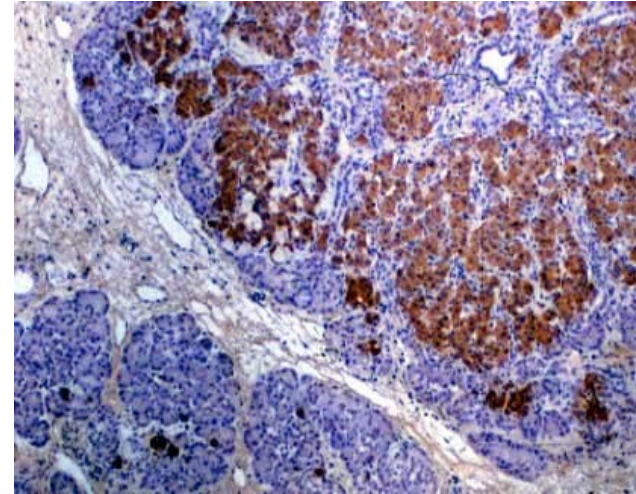


Surgical Approach at CHOP



Diffuse HI:

Intraop biopsies (3-5 mm) taken to confirm diffuse disease, then near-total pancreatectomy

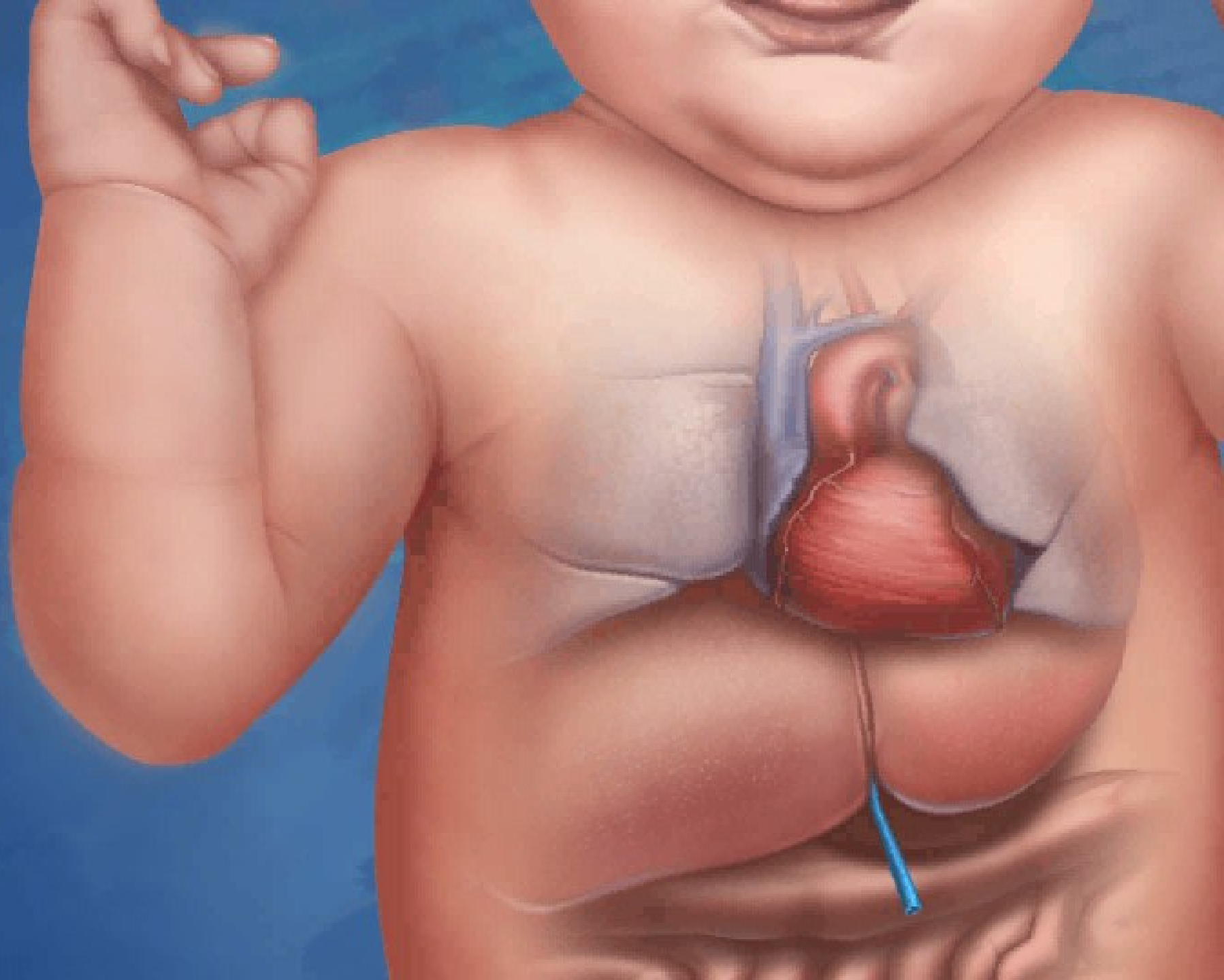


Focal HI:

Intraop biopsies taken to confirm focal disease, localize the focal lesion, then **partial** pancreatectomy to completely remove the focal lesion

PURPOSE

- Review our experience with pancreatectomy for focal and diffuse HI in 327 patients from 12/1998 to 2/2012 using a multidisciplinary diagnostic and therapeutic approach (endocrinology, radiology, pathology, surgery).
- Evaluate the effectiveness of radiologic procedures (particularly the 18-Fluoro-DOPA PET scan) in the localization of focal HI.
- Assess the clinical outcome of partial pancreatectomy in infants with focal HI.



Prevalence of Focal HI

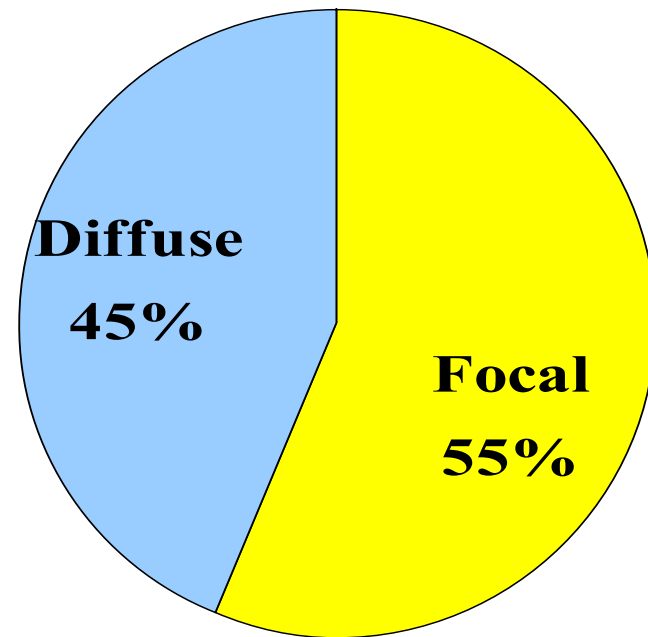
327 pancreatectomy patients, 12/98 – 2/12

169 Focal

5 Focal/Redo

144 Diffuse

9 Insulinoma

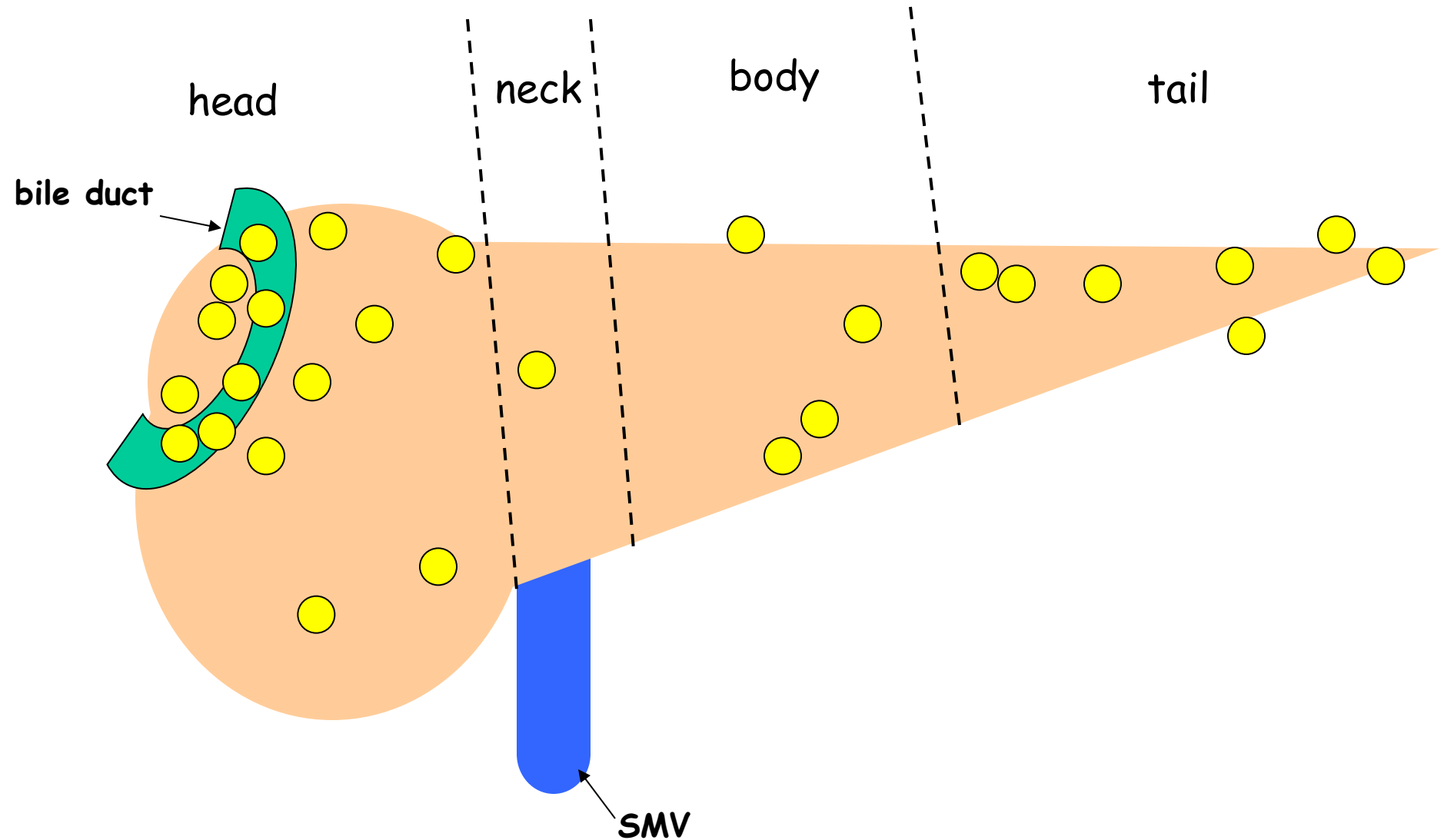




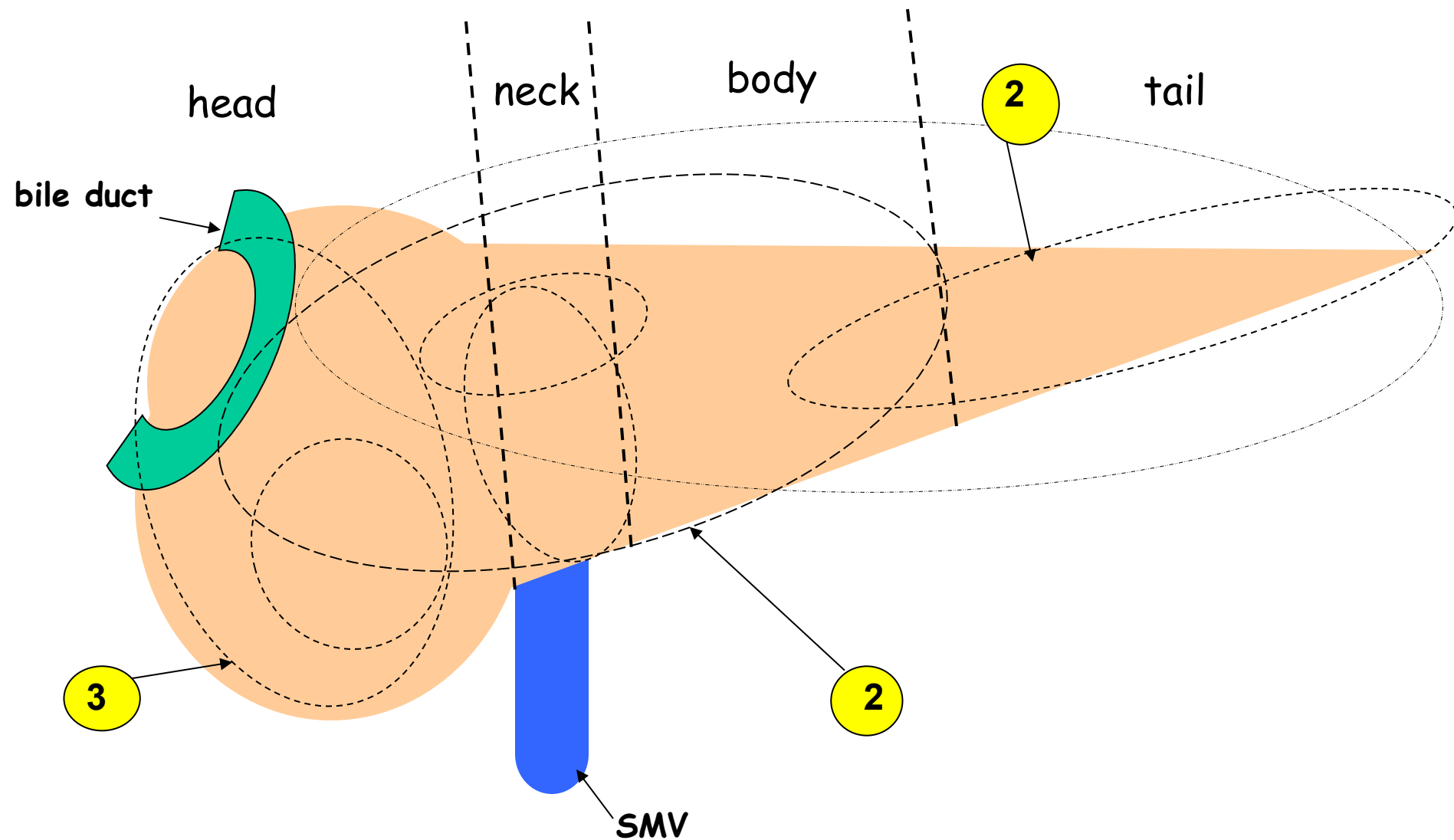
Partial Pancreatectomy in 169 Cases of Focal HI

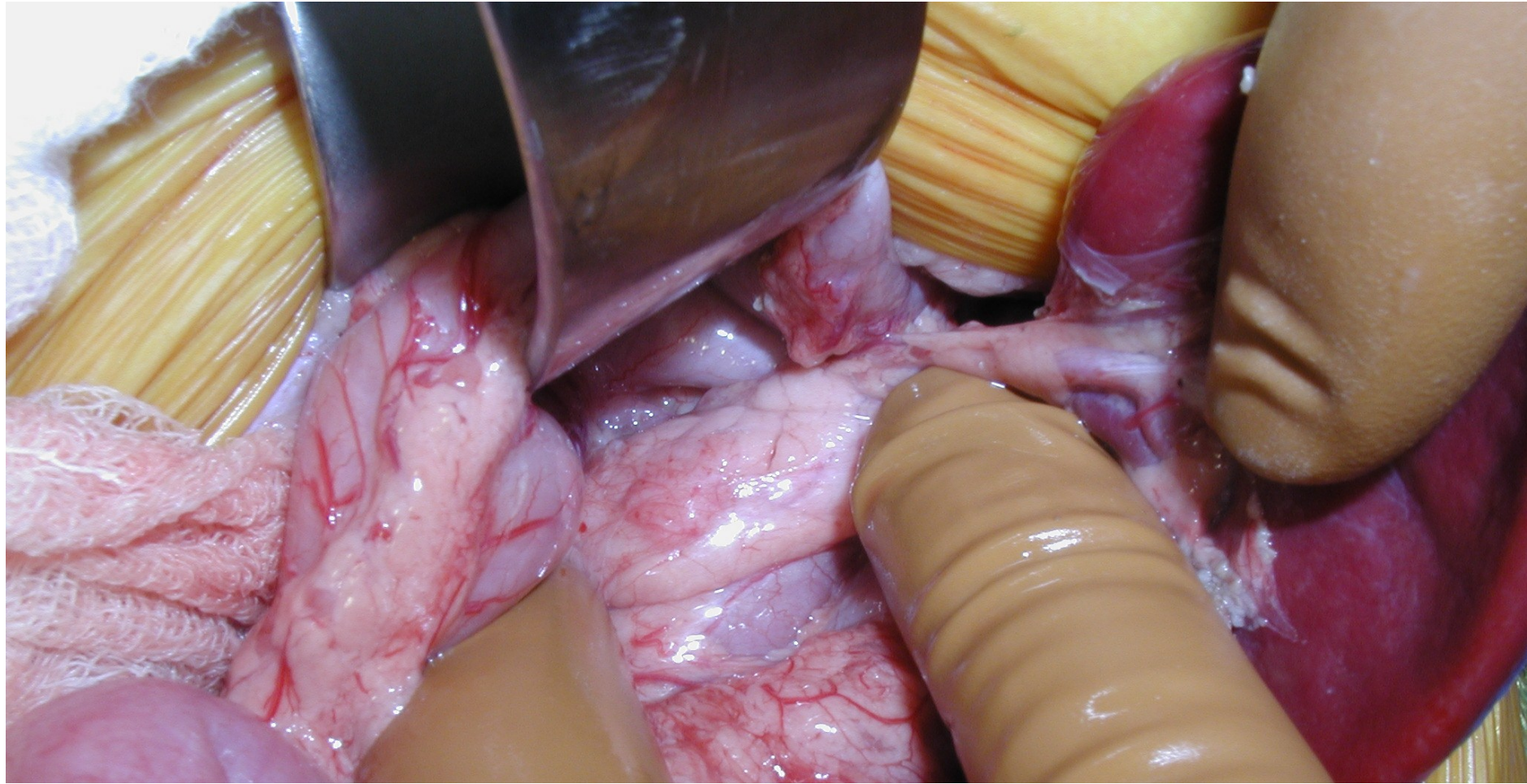
- 89 girls, 80 boys. Age at operation: 1 week to 14 months, median age = 7 weeks.
- ~2/3rds have visible and/or **palpable** clue for location.
- Spleen preserved in all cases; 1 CBD injury.
- Pancreatectomy extent ranged from 2% to 98%. Vast majority of pancreatectomies were less than 50%.
- 56% of focal lesions involve the pancreatic head; 27 Roux-en-Y operations including 2 Whipple procedures.
- 5 laparoscopic resections (tail); 1 inadequate resection.
- > 95% of focal HI pts were cured.
- 7 pts require glycemic meds or tube feedings for persistent hypoglycemia. Early lesson = accurate intraop frozen section confirmation of clear margins is imperative.
- No patient is diabetic.

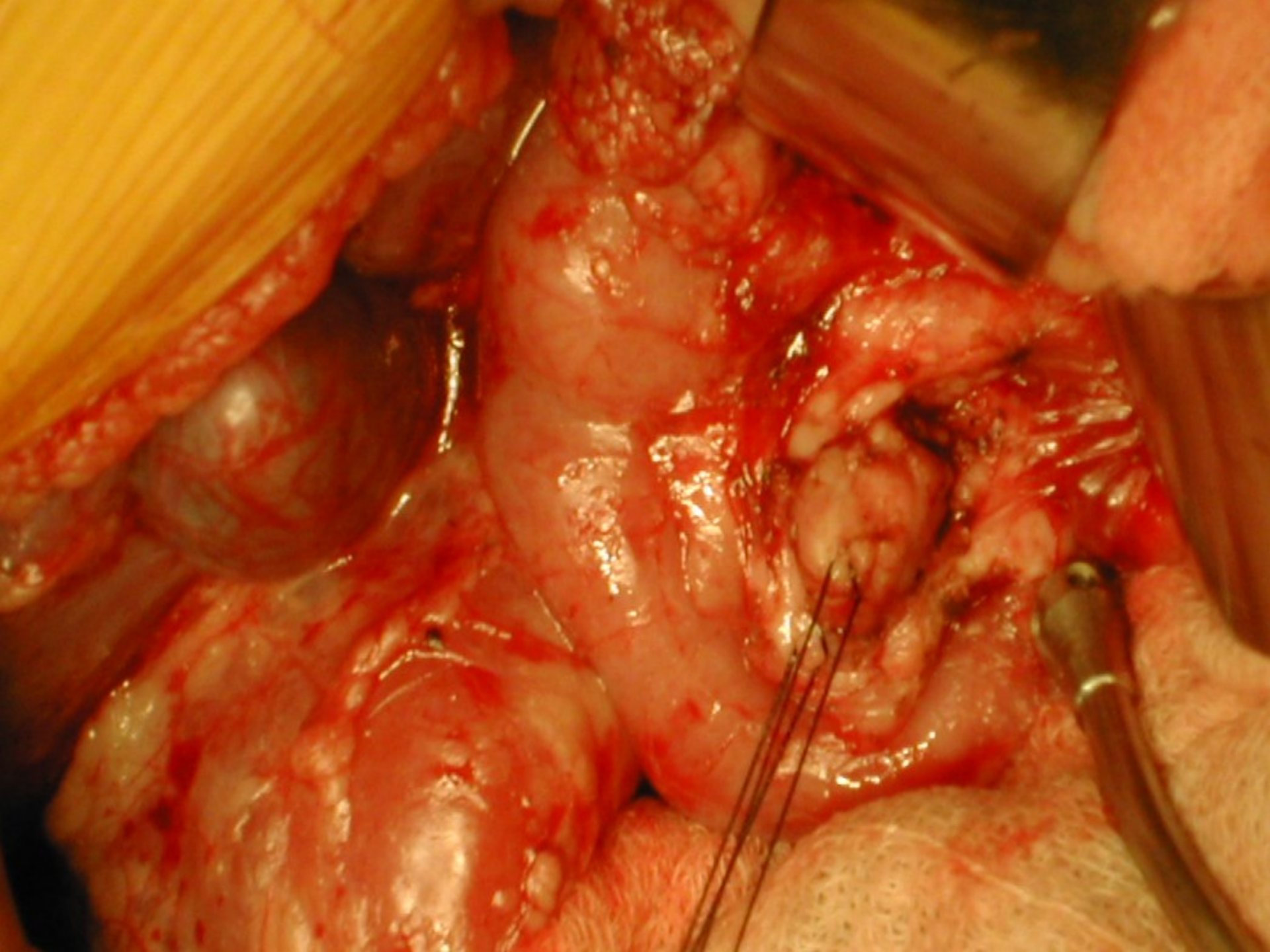
27 of 38 Focal HI Lesions < 1 cm in diameter



11 of 38 Focal Lesions > 1 cm in diameter





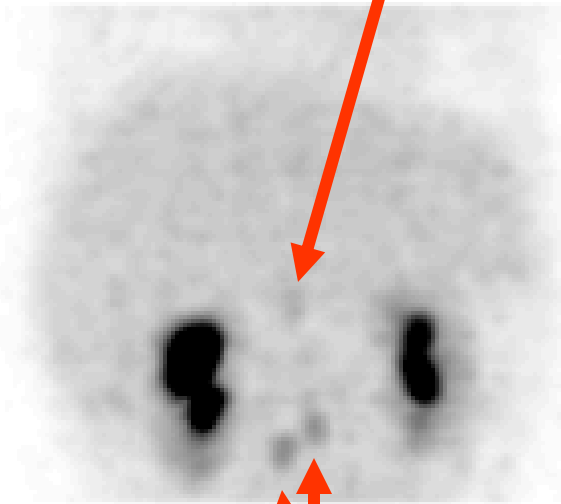


An Important Lesson

- Three additional patients with focal HI were referred after 95-98% pancreatectomy showed normal pancreas.
- A focal lesion was present in the residual pancreatic head tissue and the lesion was resected.
- One patient from 13 years ago has persistent hypoglycemia treated medically despite complete pancreatectomy and choledochoduodenostomy. Subsequent PET scan showed additional unresected focal lesion within the duodenal wall.
- Two patients are insulin-dependent diabetics & require pancreatic enzymes. One of these patients had additional focal lesions in the small bowel.

Ectopic hyperinsulinism lesions

Head of pancreas



Ectopic lesions

0 deg

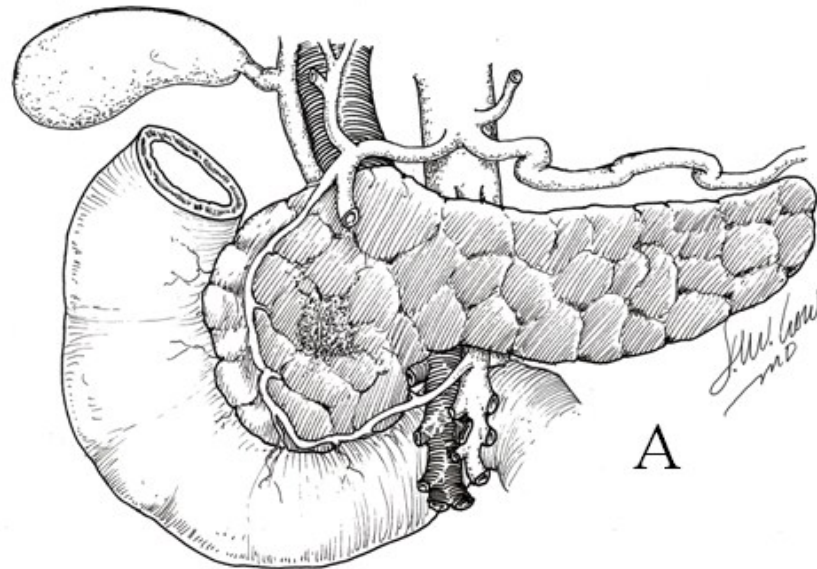


SURGICAL COMPLICATIONS: FOCAL LESIONS

- Additional resection for residual disease (7 of 8 cured; 4 before and 2 after Roux-en-Y)
- 7 small bowel obstructions (including 5 postoperative SB-to-SB intussusceptions - reduced)
- Chylous ascites (3) – resolved with elemental formula. Ligate lymphatics at base of mesentery. No longer leave drain or use fibrin glue in pancreatic bed.

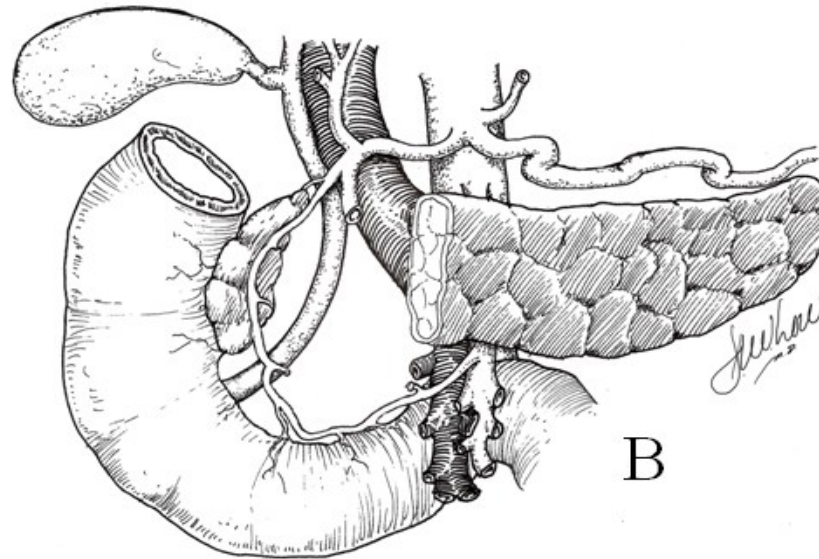
Technique

- Transverse supraumbilical laparotomy, Kocher maneuver, expose entire pancreas.
- Inspection / palpation of the entire pancreas, intraoperative biopsies.
- If a large/deep focal lesion is identified in the head of the pancreas a near-total head resection is performed to ensure clear margins by frozen section analysis



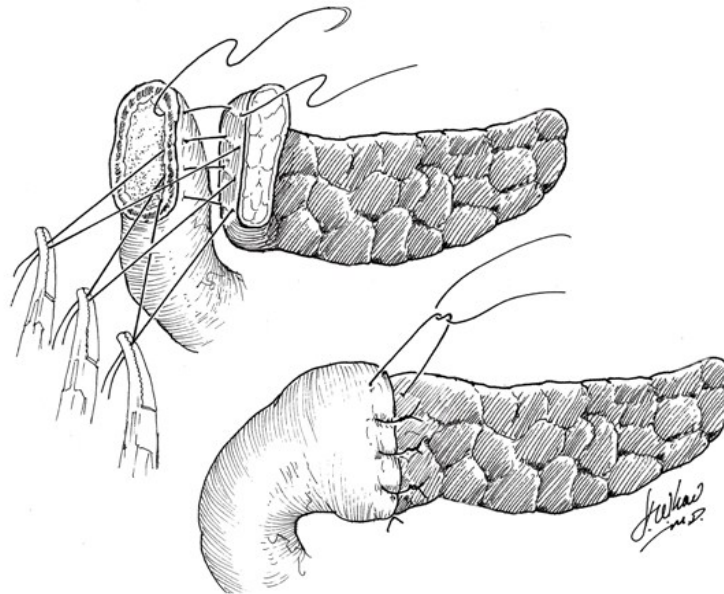
Technique

- Common bile duct carefully dissected and skeletonized.
- *Gastrooduodenal* and *pancreaticoduodenal* arteries and vascular arcade along the duodenum are identified and preserved.
- Pancreatic body transected sharply with no electrocautery.
- A thin strip of pancreatic head is left between the CBD and the duodenal wall.



Technique

- 25-cm-long Roux-en-Y retrocolic jejunal limb.
- Anastomosis to the *capsule* of the pancreas, *beyond* the cut end; 5-0 monofilament suture. Cut pancreatic end is tucked into the jejunal lumen.
- Posterior aspect first (knots *inside*) anterior aspect last (knots *outside*).
- Interrupted stitches; omentum then wrapped around the anastomosis.

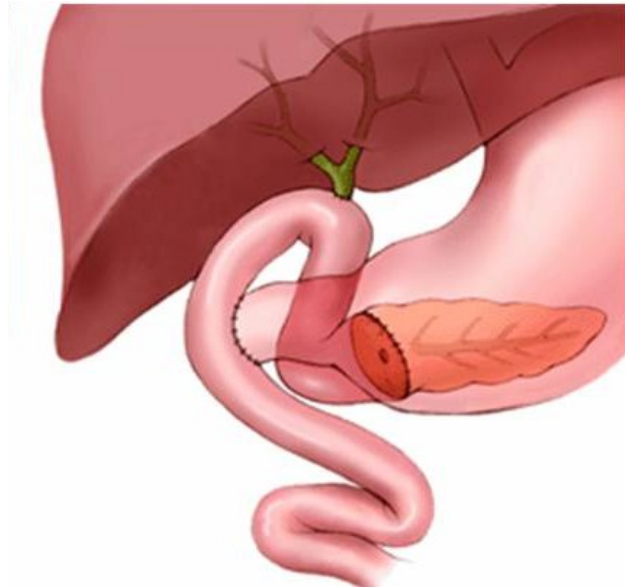


Results

- 27 patients, 14 males, 13 females
- Median *age* at surgery: **8 weeks** (range: 3 to 56)
- Median *weight* at surgery: **5.8 kg** (range: 4.3 to 9.8 kg)
- Mean blood loss: 25 ml (range: 5 to 75 ml)
- One CBD injury: choledochoduodenostomy
- Drains were not used
- Median total OR time: 241 min (*Biopsies, frozen sections)
- Standardized postop protocol for IV Glucose Infusion Rate

Results

- 4 patients had a Roux-en-Y done *after* a previous incomplete focal head resection.
- 2 patients had a lesion involving the duodenal wall and required a pylorus-preserving *Whipple* procedure after lesion resection led to duodenal ischemia.



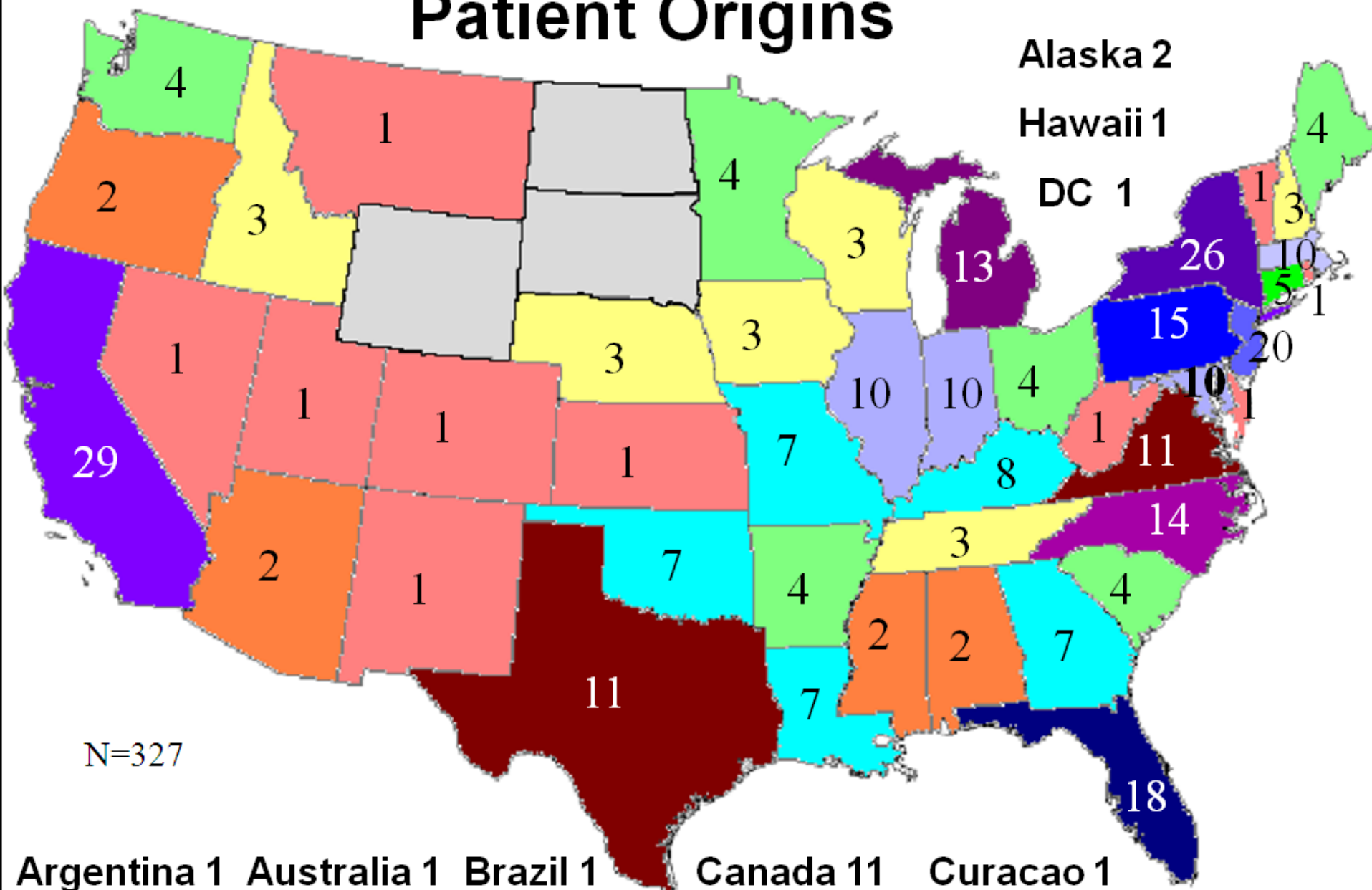
Results

- One patient developed a 15-cc fluid collection (percutaneously drained)
- Same patient developed SBO 4 months later; exploration/lysis of adhesions.
- *2 reoperations* after the Roux-en-Y due to residual disease in the pancreatic head close to the duodenum. Intraoperative ultrasound was helpful.
- Median hospital stay: 22 days (range 13 to 60)
- All patients cured of Hyperinsulinism
- None have diabetes or pancreatic exocrine insufficiency

Diffuse Disease (144)

- Surgery = 95-98% pancreatectomy
- 16% well-controlled; 37% diabetic; 47% required glycemic meds, or 2nd pancreatectomy (9 cases)
- 3 CBD injuries: choledochoduodenostomy; no splenectomies
- 1 required resection of distal duodenum
- 1 required IR drainage of intraabdominal collection
- 2 preop deaths due to NEC; 4 postop small bowel obstructions (3 SB-to-SB intussusceptions - reduced)
- Long-term risk of insulin-dependent diabetes
- Surgery is **not** a cure for diffuse disease but can help prevent hypoglycemia and brain damage

Patient Origins



Argentina 1 Australia 1 Brazil 1 Canada 11 Curacao 1
Ecuador 1 Iran 1 Israel 2 Japan 1 Saudi Arabia 1 Venezuela 1

CONCLUSION

A multidisciplinary approach (specialists in pediatric endocrinology, radiology, pathology, and surgery) for patients with the focal form of congenital hyperinsulinism can distinguish focal from diffuse disease, localize focal lesions, and permit partial pancreatectomy with cure in most patients.

Congenital Hyperinsulinism Paradigm 2012

