CHAPTER 36

# Surgery for Persistent Hyperinsulinaemic Hypoglycaemia of Infancy

Lewis Spitz

## INTRODUCTION

Hyperinsulinism as a cause of persistent hypoglycaemia in infancy and childhood is very uncommon (1:50,000). The importance of preventing hypoglycaemia (and reducing the likelihood of neurological damage) by the administration of adequate quantities of carbohydrate cannot be over stressed. The diagnosis of persistent hyperinsulinaemic hypoglycaemia of infancy (PHHI) is based on:

- Inappropriately raised plasma insulin levels for blood glucose concentration
- Glucose infusion rate greater than 10 mg/kg per minute to maintain a blood glucose level above 2.6 mmol/l
- Low free fatty acid and blood ketone bodies during hypoglycaemia
- Glycaemic response to glucagons despite hypoglycaemia

In essence, the diagnosis is established by measuring insulin in a blood sample taken during hypoglycaemia (blood glucose <2.6 mmol/l). Among lesions, 40–50% are focal and it is important to identify these in order to avoid unnecessarily performing a near-total pancreatectomy. Pre-operative methods of distinguishing focal from diffuse disease include:

- Pancreatic venous sampling. This involves transhepatic catheterization of the pancreatic venous system and sampling blood at various levels while maintaining blood glucose levels below 3 mmol/l. The results may show suppression of insulin secretion throughout most of the pancreas with one "hot spot" indicative of a focal lesion or generalised dysregulation (diffuse disease).
- Intra-atrial stimulation test where calcium is injected into the gastroduodenal, superior mesenteric and splenic arteries to stimulate insulin secretion.
- Magnetic resonance imaging, computed tomography and positron emission tomography scans have not been proven to be of diagnostic value.

It is essential to insert a central venous catheter to monitor blood glucose levels and to provide a reliable route for intravenous glucose administration. Drug treatment consists of diazoxide with chlorothiazide, somatostatin and nifedepine; indication for surgery is the failure to respond to intensive medical treatment.

**Figure 36.1**

The pancreas is exposed by dividing the vessels in the gastro-colic omentum to expose the head, body and tail of the pancreas.



**Figure 36.2**

Any suspicious nodules, particularly if the pre-operative studies have indicated focal disease, should be excised and sent for frozen-section histopathological examination.

Figure 36.1

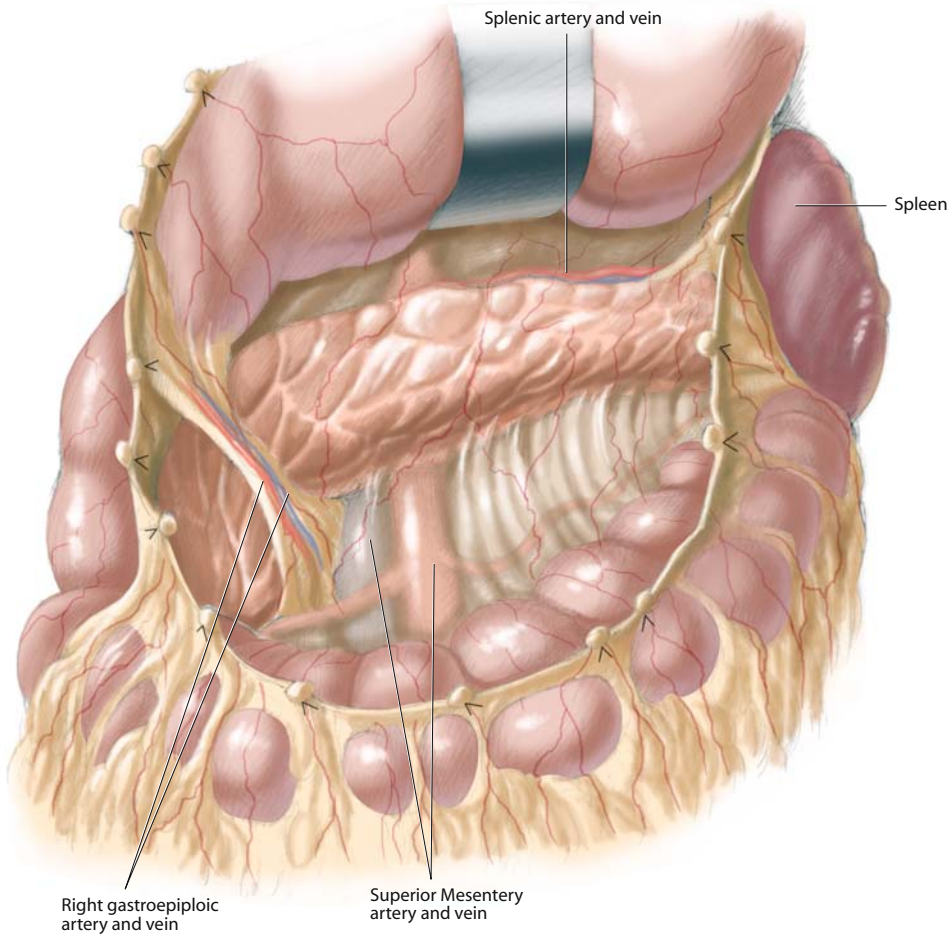



Figure 36.2

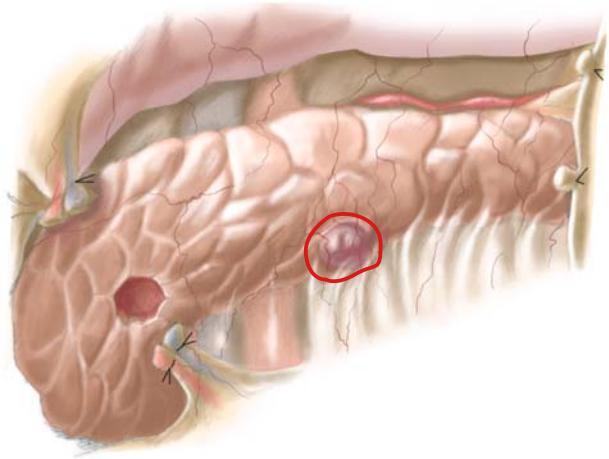


**Figure 36.3**

Near-total pancreatectomy for diffuse disease starts with mobilizing the tail and body of the pancreas. Bipolar coagulation of short pancreatic vessels is carried out, following which the vessels are divided. Starting at the tail of the pancreas in the hilum of the spleen, the dissection proceeds towards the neck of the pancreas. The distal part of the tail of the pancreas is divided and the end sent for frozen-section histopathology to confirm the diffuse nature of the condition.

**Figure 36.4**

In order to perform the resection of the uncinate processus, the superior mesenteric vein is retracted to the left and the uncinate processus is carefully dissected out from behind the vein until it is completely free from any attachments.

Figure 36.3

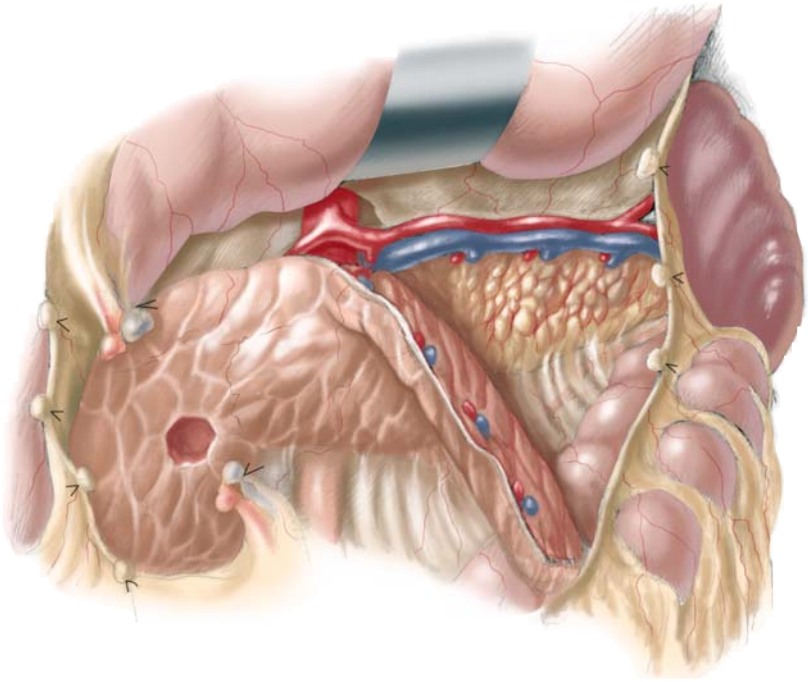


Figure 36.4

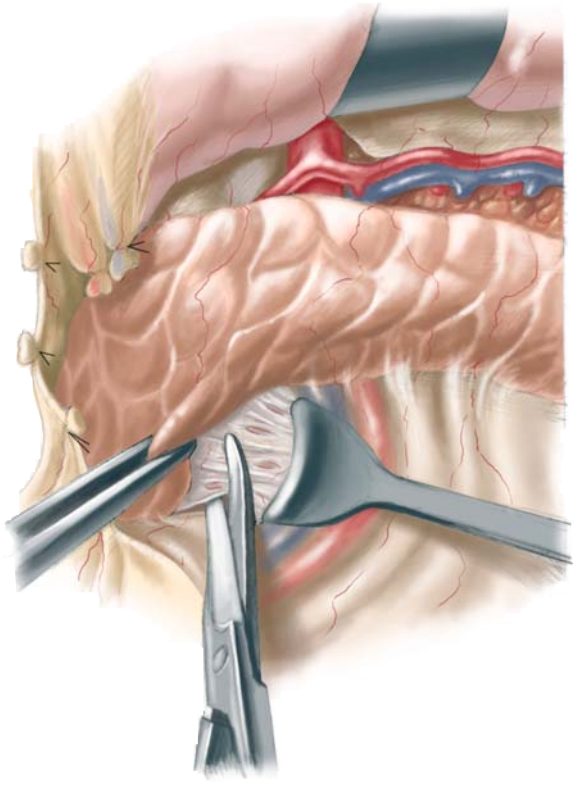


**Figure 36.5**

The common bile duct is defined above the first part of the duodenum and a soft rubber sling is passed around the duct at this point. From within the C-loop of the duodenum a passage is created behind the first part of the duodenum and the rubber sling is transposed behind the duodenum to appear within the C-loop. The aim of this manoeuvre is to identify the distal course of the common bile duct in or posterior to the head of the pancreas and to preserve its integrity during resection of the head of the pancreas.

**Figure 36.6**

The resection includes the head, body and tail of the pancreas including the uncinate process, leaving a small sliver of pancreatic tissue within the C-loop of the duodenum and that part of the pancreas that lies around the common bile duct and between the duct and the duodenum. Before closing the abdominal incision it is important to check the integrity of the common bile duct by applying gentle pressure on the gallbladder and observing for any bile leak. A suction drain is left in the pancreatic bed for 24–48 h postoperatively.

Figure 36.5

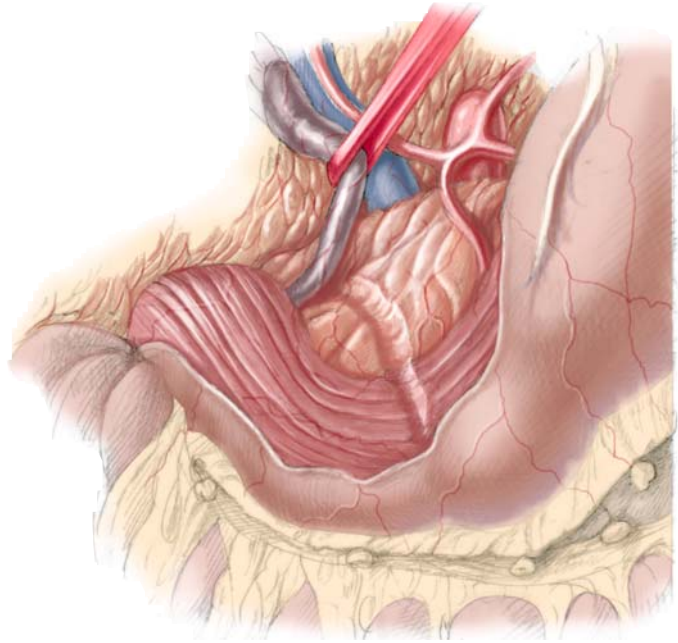
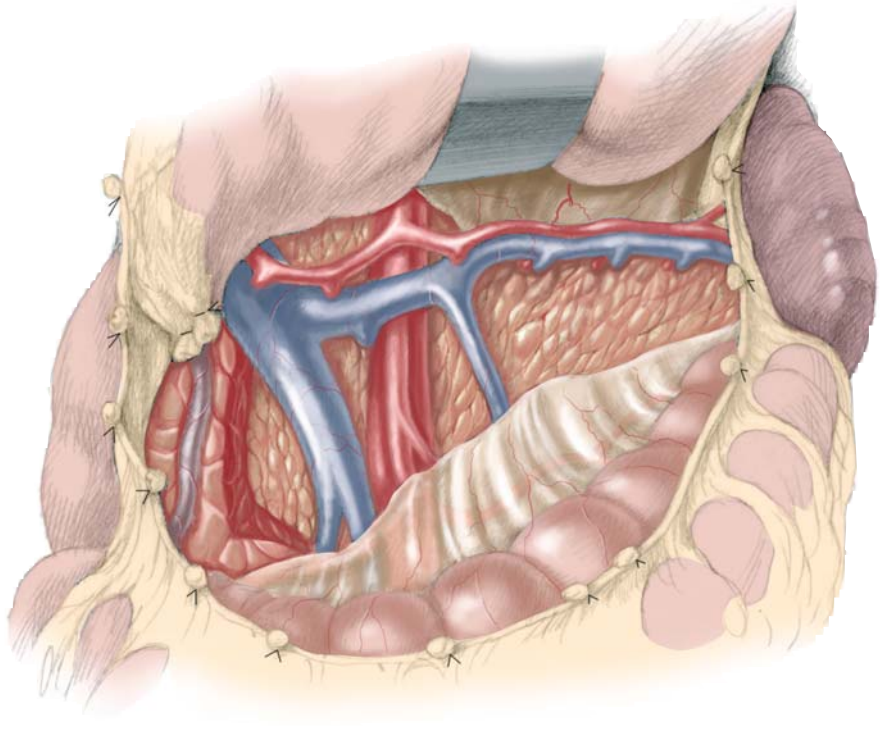


Figure 36.6



## CONCLUSION

Intra-operative haemorrhage should not occur and the most frequently encountered problem is trauma to the bile duct. The duct may be injured only in the near-total resections – incidence of 12%. The injury can occur intra-operatively and, if detected, repaired immediately by direct suture or by choledochoduodenostomy. Late stricture from ischaemia can occur weeks to months post-operatively and these too require drainage by choledocho-enterostomy. Other complications include wound sepsis, adhesion intestinal obstruction and prolonged ileus. The long-term requirements for insulin therapy and exocrine pancreatic replacement need to be carefully assessed.

## SELECTED BIBLIOGRAPHY

Adzick NS, Thornton PS, Stanley CA, Kaye RD, Ruchelli E (2004) A multidisciplinary approach to the focal form of congenital hyperinsulinism leads to successful treatment by partial pancreatectomy. J Pediatr Surg 39:270–275

Aynsley-Green A, Hussain K, Hall J, Saudubray JM, Nihoul-Fekete C, Lonlay- Debeney P, Brunelle F, Otonkoski T, Thornton P, Lindley KJ (2000) Practical management of hyperinsulinism in infancy. Arch Dis Child Fetal Neonatal Ed 82:F98–F107

de Lonlay P, Fournet JC, Touati G, Groos MS, Martin D, Sevin C, Delagne V, Mayaud C, Chigot V, Sepouz C, Brusset MC, Laborde K, Bellane-Chantelot C, Vassault A, Rahier J, Junien C, Brunelle F, Nihoul-Fekete C, Saudubray JM, Robert JJ (2002) Heterogeneity of persistent hyperinsulinaemic hypoglycaemia. A Series of 175 cases. Eur J Pediatr 161:3–48

Lonlay-Debeney P, Poggi-Travert F, Fournet JC, Sempoux C, Vici CD, Brunelle F Touati G, Rahier J, Junien C, Nihoul-Fekete C, Robert JJ, Saudubray JM (1999) Clinical features of 52 neonates with hyperinsulinism. N Engl J Med 340:1169–1175

McAndrew HF, Smith V, Spitz L (2003) Surgical complications of pancreatectomy for persistent hyperinsulinaemic hypoglycaemia of infancy. J Pediatr Surg 38:13–16