Pancreatic Disorders

60

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Contents

60.1 Introduction

Although most pancreatic disorders are rare in childhood, there are a number with which the paediatric surgeon must be familiar. These conditions may be congenital or acquired and may affect the exocrine or the endocrine components of the gland. In this chapter the most frequently encountered conditions are discussed including: structural pancreatic abnormalities, ectopic pancreatic tissue, acute and chronic pancreatitis, pancreatic trauma, hyperinsulinism, cystic fibrosis, pancreatic cysts and pseudo cysts, and pancreatic neoplasms.

60.2 Structural Abnormalities

A wide range of developmental structural abnormalities of the pancreas can occur. These include pancreas agenesis, aplasia and hypoplasia, hyperplasia and hypertrophy, and dysplasia. The commonest structural abnormalities are annular pancreas and pancreas divisum. Both these conditions result from abnormal pancreatic development and therefore an understanding of normal pancreatic development is important. The pancreas is formed from two endodermal buds

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(a large dorsal bud and a smaller ventral bud) which both arise from the embryonic foregut during the 5th week of gestation. Each bud has its own duct and by the end of the 6th week the dorsal duct (duct of Santorini) drains into the foregut and slightly later the ventral one (duct of Wirsung) drains into the hepatic diverticulum. The dorsal bud develops faster than the ventral one and becomes larger overall. The rapid growth of the developing duodenum results in the ventral bud rotating behind the duodenum so that it comes to lie adjacent to the inferior and posterior aspects of the dorsal bud. During the 7th week, the two pancreatic buds fuse resulting in a single gland and a combined ductal system. The dorsal bud forms the body and tail of the definitive pancreas, whereas the ventral bud forms the head and uncinate process. The derivation of the definitive pancreatic ductal system is somewhat unexpected in that the minor accessory duct arises from the larger dorsal bud, whereas the main pancreatic duct arises distally from the dorsal bud and proximally including the ampulla from the ventral bud. Acinar cells and islet cells both appear to develop from the cells lining the pancreatic ducts. Whereas the acinar cells appear to remain non-secretory during foetal life, secretion from foetal islets seems to play an important part in foetal homeostasis.

60.3 Annular Pancreas

As its name implies, this condition results from normal pancreatic tissue encircling the duodenum. It is caused by abnormal rotation of the developing ventral pancreatic bud and the prevailing theory of pathogenesis is that the ventral bud splits into two with part rotating posteriorly and the remainder rotating anteriorly forming a complete or incomplete ring of pancreatic tissue.

60.3.1 Diagnosis

Whilst annular pancreas may be completely asymptomatic and be found incidentally at laparotomy or post mortem, the commonest presentation is that of partial or complete duodenal obstruction. The obstruction either results from extrinsic compression of the duodenum by the encircling pancreas or can be due to an associated duodenal atresia (25% of duodenal atresias are associated with annular pancreas). The condition may be diagnosed antenatally by the presence of polyhydramnios and a dilated stomach and first part of duodenum on antenatal ultrasound. Postnatally, cases of complete duodenal obstruction classically present in the first few hours of life with the onset of vomiting. The vomit is usually bile-stained as the obstruction is most commonly in the second part of the duodenum distal to the ampulla of Vater. A plain abdominal x-ray demonstrates the typical ‘double bubble’ of duodenal atresia with absence of distal gas. Cases of incomplete obstruction often present more insidiously and diagnosis is often delayed. An upper gastrointestinal contrast study is useful in these cases.

60.3.2 Management

Once the diagnosis of duodenal obstruction has been confirmed, the stomach is decompressed with a nasogastric tube and fluid and electrolyte disturbances are corrected. After appropriate resuscitation and parental consent, the baby is taken to theatre. Once annular pancreas had been confirmed at laparotomy, the proximal and distal duodenum are carefully mobilised and a duodeno-duodenostomy is performed. The classically described ‘diamond-shaped’ anastamosis can be used, although a simple side-to side anastamosis is usually sufficient. The surgical procedure can be performed as an open or laparoscopic intervention.

60.4 Pancreas Divisum

Pancreas divisum is the commonest congenital abnormality of the pancreas and is found in up to 11% of patients at post mortem. It results from failure of the ventral and dorsal pancreatic ducts to unite during fusion of the ventral and dorsal pancreatic buds (see above). As a consequence, the duct of the dorsal bud (duct of Santorini) becomes the main pancreatic duct but drains into the duodenum through the minor papilla. A number of other

anatomical variants of the ductal system are also encountered.

60.4.1 Diagnosis

Many individuals with pancreas divisum remain asymptomatic and as such it is often considered as a developmental variant rather than a pathological condition. Indeed this abnormal configuration of the pancreatic ducts is found in over 3% of patients under going endoscopic retrograde cholangiopancreatography (ERCP). However, the restricted drainage of the pancreatic secretions through the minor papilla can result in dilatation of the pancreatic duct and pancreatitis. In this group of patients, pancreas divisum can be confirmed by magnetic resonance cholangiopancreatography (MRCP) or ERCP. Although the latter is associated with a higher morbidity, it has the advantage that therapeutic intervention can be performed at the same time.

60.4.2 Management

The principal aim of treatment is to establish adequate drainage of the 'main' pancreatic duct in order to relieve symptoms and to prevent chronic pancreatitis and pancreas insufficiency from developing. Unfortunately, by the time of presentation chronic changes are often evident. Drainage of the pancreas can be achieved by sphincteroplasty or endoscopic sphincterotomy of the minor papilla. If chronic changes of duct dilatation have developed, a pancreaticojejunostomy (Puestow procedure) may be indicated. Pancreatic resection may be required in severe forms of chronic pancreatitis (see below).

60.5 Ectopic Pancreatic Tissue

Ectopic pancreatic tissue is frequently found associated with foregut-derived structures. This is thought to be the result of transition of the embryonic epithelium. These pancreatic 'rests' are usually asymptomatic. However, pancreatic tissue can be found at the base of a Meckel's diverticulum and can cause bleeding and inflammation.

60.6 Acute Pancreatitis

Acute pancreatitis is an inflammatory condition of the pancreas resulting from intrapancreatic activation, secretion and digestion of the pancreas by its own enzymes. It is rare in childhood but must always be considered in children presenting with abdominal pain. There are many different causes of acute pancreatitis and the principle ones are outlined in Table 60.1.

60.6.1 Diagnosis

The diagnosis of acute pancreatitis is based on a careful history and physical examination, and confirmed by the results of both laboratory and radiological investigations. A careful history is essential both for the diagnosis to be made and for determining the underlying cause. Acute pancreatitis classically presents with

Table 60.1 Principle causes of acute pancreatitis

Idiopathic	
Hereditary	
Systemic infections	– Mumps
	– Coxsackie B
	– Rubella
Trauma	
Iatrogenic injury	
Developmental anomalies of pancreatico-biliary system	
	– Pancreas divisum
	– Pancreatcobiliary malunion
	– Choledochal cyst
Gallstones	
Drugs	– Azathiorpine
	– Steroids
	– Valproic acid
	– Tetracycline
	– L-Aspariginase
	– Immunosuppressants
Metabolic Abnormalities	– Hypercalcaemia
	– Hyperglyceridaemia
	– Cystic fibrosis
Miscellaneous	

epigastric pain radiating through to the back and left upper quadrant. However, the pain may be less well localised and particularly in younger children, nausea and vomiting is often the predominant feature. In determining the underlying cause, it is important to elucidate a history of abdominal trauma, gallstones, recent mumps, familial pancreatitis, or pancreatitis-associated medication. Physical examination may elicit a range of signs including low-grade pyrexia, epigastric tenderness, generalised peritonitis, and in severe forms of pancreatitis the child may present with the signs of hypovolaemic shock. In cases of necrotising or haemorrhagic pancreatitis, pathognomonic patterns of bruising may be present on the abdominal wall either around the umbilicus (Cullen's sign) or in the flanks (Grey-Turner sign). Grossly elevated serum amylase levels are usually diagnostic of acute pancreatitis but normal levels do not exclude the diagnosis, and mildly elevated levels can be present in a number of other conditions including salivary inflammation, intestinal perforation, and renal failure. Elevated urinary amylase levels occur only if the serum amylase is greatly elevated. As a result of these discrepancies with amylase, some centres measure serum lipase levels instead, as lipase is specific to the pancreas.

A plain abdominal x-ray is important to exclude perforation. Other features that may be seen include gallstones, a dilated loop of small bowel (sentinel loop), and a dilated ascending colon with 'cut off' of air in the transverse colon. A chest x-ray may demonstrate pulmonary oedema or a left sided pleural effusion. Abdominal ultrasound can be useful for identifying pancreatic oedema and can be helpful in identifying pancreatic anatomy. However, overlying dilated bowel loops frequently obscure the visibility of the pancreas. Computed tomography (CT) with double contrast is therefore often indicated and provides a more accurate picture of the degree of pancreatic damage and better visualisation of the ductal anatomy (Fig. 60.1). Apart from its therapeutic use following pancreatic trauma, ERCP is rarely indicated in the acute phase of pancreatitis. Indeed, non-interventional ERCP is itself associated with a 10% incidence of acute pancreatitis. If duct anatomy needs to be further elucidated, a MRCP is useful.

Other laboratory tests are performed to determine the severity of the disease including a full blood count, liver function tests, serum calcium, blood glucose, and an arterial blood gas (see below).

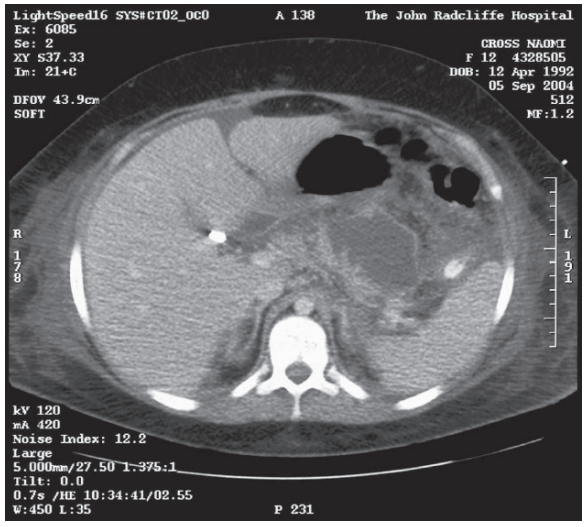

Fig. 60.1 An abdominal CT scan of a 12 year old girl presenting with acute necrotizing pancreatitis secondary to gallstones. The scan demonstrates a diffusely swollen pancreas with 75% necrosis

60.6.2 Management

There are three main aims of management of these patients, namely to support the child during the acute phase of the disease, to help prevent the sequelae of the disease, and to identify and treat any underlying causes.

Children with acute pancreatitis require very close monitoring throughout their admission. Early signs of multi-organ failure must be treated aggressively as the child can deteriorate rapidly. This is often best achieved by the child being located in a high dependency unit or an intensive care unit.

Active resuscitation is essential from the onset in all patients. Oxygen is administered and breathing carefully assessed. Children with severe pancreatitis may require ventilation. Acute pancreatitis is associated with large volume fluid loss and therefore aggressive fluid replacement and close monitoring of urine output are vital. A nasogastric tube is inserted to decompress and 'rest' the bowel. Analgesia is administered as soon as the child has been assessed, but opiates are used selectively due to their spasmodic effects on the sphincter of Oddi and hence their indirect effects on intra-pancreatic pressure. Broad-spectrum antibiotics are often administered and indeed it is the author's preference to do so. However, there is no conclusive evidence-base for this approach. A number of different regimes have been attempted in order to reduce

pancreatic enzymatic activation in the acute phase but it is unclear which have a clear advantage. There does seem to be moderate evidence that somatostatin analogues are beneficial. In addition, many clinicians start the child on H_2 antagonists or proton pump inhibitors in order to prevent gastric ulceration and also potentially prevent gastric acid from triggering pancreatic secretions within the duodenum. Total parenteral nutrition is commenced early in the treatment regime not only because prolonged bowel rest is the norm, but also because a positive nitrogen balance has been shown to be associated with a more favourable outcome. Surgical intervention is rarely required in the acute phase of the disease. Peritoneal lavage has been used widely in the past but is rarely indicated in children. Necrotising pancreatitis may require necrosectomy or pancreatic debridement and this may sometimes need to be done repeatedly. The complications of acute pancreatitis such as pancreatic abscess and pancreatic pseudocyst may also require surgical treatment (see below).

Once the child has been stabilised, the underlying cause of the acute pancreatitis should also be addressed and treated appropriately. This includes correcting conditions such as hyperlipidaemia and hypercalcaemia and also planning surgical treatment for gallstones and correction of any underlying pancreatico-biliary malformations.

60.6.3 Prognosis

A number of different systems for predicting patient outcome have been devised. These include the Ranson and Imrie Criteria, and Apache II. However, none of these systems have been worked out on children and they also tend to make a presumption that all the parameters being measured rise and fall at similar stages in the disease.

The majority of children with acute pancreatitis make a good recovery.

60.6.4 Complications

The main complications of acute pancreatitis are pancreatic abscess, pancreatic pseudocyst, pancreatic fistula, and relapsing pancreatitis.

Pancreatic abscesses require open drainage having first been confirmed by CT-guided aspiration. There is some evidence that delaying abscess drainage beyond the first 2 weeks of the onset of the disease is associated with a better outcome.

A pancreatic pseudocyst results from accumulation of leaked pancreatic enzymes enclosed within an inflammatory non-epithelial lining. They commonly appear 10–14 days after the onset of acute pancreatitis and should be suspected if elevated amylase levels reappear after having settled. About half of acute pancreatic pseudocysts settle spontaneously. If they persist, maturation of the pseudocyst wall is essential before definitive treatment is performed. It is conventional to allow 6–8 weeks for this process to occur. Treatment usually consists of internal drainage of the pseudocyst either by open gastro-cystostomy or by minimally invasive procedures such as endoscopic insertion of gastro-cystic stent or percutaneous drainage. Occasionally if the pseudocyst is confined to the tail of the pancreas, excision can be considered. Complications of pancreatic pseudocysts include bleeding, infection, and rupture.

A pancreatic fistula is usually a post-operative complication and can be associated with a low or high output. Most low output fistulae close spontaneously. Treatment is therefore directed at maintaining adequate nutrition of the child, attempting to reduce secretions with pharmacological agents such as somatostatin analogues, and ensuring that the fistula tract does not become prematurely obstructed. Persistent fistulae may require surgical intervention.

Although the majority of cases of acute pancreatitis are single episodes and self-resolving, some children may develop acute relapsing pancreatitis and others may develop irreversible damage of the pancreas leading to chronic pancreatitis.

60.7 Chronic Pancreatitis

Chronic pancreatitis occurs when the pancreas has been permanently damaged by inflammation. Several classifications exist for this disease and the simplest is an anatomical one sub-dividing the condition into either being calcifying or obstructive. The former is more common and is associated with dense fibrosis and intra-pancreatic stone formation. The commonest aetiology in children is hereditary or familial pancreatitis (Fig. 60.2). Obstructive chronic pancreatitis on the other hand is associated with less aggressive scar formation

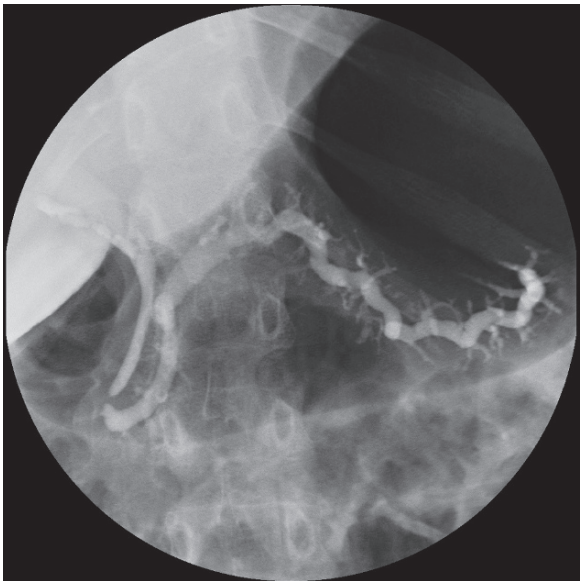

Fig. 60.2 Chronic pancreatitis due to missense mutation N34S (SPINK 1 gene). The pancreatic duct is dilated up to 1 cm. After a Puestow procedure (long side-to-side pancreatic jejunostomy) the further time course was uneventful (by courtesy of M. E. Höllwarth)

and most commonly arises as a result of congenital structural abnormalities such as pancreas divisum.

60.7.1 Diagnosis

High amylase or lipase levels are less of a feature in chronic pancreatitis. CT scans are helpful in determining the extent of damage to the pancreas, to look for calcification, and to determine the anatomy of the ductal system. An ERCP is useful if intervention to the ducts is required in the form of stenting or sphincterotomy at the same time as elucidating ductal anatomy. MRCP is the investigation of choice if a detailed view of the pancreatic ducts is required without intervention.

The main presenting features are chronic intractable pain and weight loss, but the child's first presentation may be with one or more of the complications described below. Although up to 15% of patients with chronic pancreatitis are relatively pain free, for the remainder chronic pain in the epigastrium and back dominates their lives.

This pain can be exacerbated by food or alcohol. The aetiology of the severe pain is unclear, but theories include the effects of pancreatic obstruction and distension, and also the exposure of peri-pancreatic nerves as a result of pancreatic damage.

The cause of weight loss in these patients is multifactorial in that it can be simply due to the reduced food intake of many of these patients due to the associated exacerbation of pain, but can also be due to malabsorption, as well as the fact that many of the adult patients with this disease have a chronic alcohol problem.

60.7.2 Management

The intractable pain associated with chronic pancreatitis is often difficult to control. Whilst patients can get some relief from simple measures such as leaning forward while sitting, the main stay of treatment is strong analgesia. As a result, patients with this condition frequently become opiate-dependent. Pharmacological and surgical nerve blocks can help with pain control. Some surgeons advocate partial or total pancreatectomy for pain control earlier in the disease process than others, but careful patient selection is always critical. If total pancreatectomy is performed and the individual is shown pre-operatively to have maintained endocrine function, there is a strong case for stating that the patient should also be offered a simultaneous islet autotransplant. Whilst this procedure may enable the patient to remain insulin-independent, even if it does not, it should enable the patient to avoid the particularly brittle form of diabetes that often follows pancreatectomy. Drainage procedures may be indicated if ductal dilatation is present. Longitudinal pancreaticojejunostomy (Puestow procedure) works well if the obstruction of the pancreatic duct is distal to the pancreatic neck. However, frequently the head and neck of the pancreas are also fibrosed.

Exocrine insufficiency is treated with long-term oral pancreatic enzyme replacement. The number of enzyme capsules required is titrated by the patient against the appearance of the stool.

When endocrine insufficiency has fully developed, it requires insulin treatment. As stated above, the brittle form of the disease associated with chronic pancreatitis is often difficult to control and needs close input from the diabetes team.

Obstruction of the structures adjacent to the pancreas often requires radiological or surgical intervention in the form of stents or bypass surgery.

60.7.3 Complications

The complications of chronic pancreatitis may present late in the disease process or particularly if the disease has been preceded by acute relapsing pancreatitis, may be one of the presenting features. The principle complications are exocrine insufficiency, endocrine insufficiency, and damage to adjacent structures.

Exocrine insufficiency is usually a late feature and presents with steatorrhea. Stools are bulky with a particularly offensive smell, and are difficult to flush away in the toilet. This can be exacerbated by a high lipid diet.

Despite the islets of Langerhans accounting for only 2% of the pancreatic tissue, the endocrine component of the pancreas is often surprisingly spared until the later stages of the disease. Before frank diabetes develops however, abnormal patterns of glucose-tolerance tests can often be detected.

The pancreas is associated with a number of other structures including the common bile duct, the duodenum, and the splenic vessels. All these structures can become partially or completely obstructed as a result of chronic fibrosis. Colonic obstruction can also occur if the inflammation is extensive.

The precise link between chronic pancreatitis and pancreatic cancer is unclear, but several studies have suggested a direct correlation.

60.8 Pancreatic Trauma

Although pancreatic injury occurs in less than 3% of adults who have suffered blunt or penetrating abdominal trauma, the anatomical position of the pancreas traversing the upper lumbar vertebrae means that this organ is vulnerable to transection and this injury is greatly increased in young children due to their thin abdominal walls. Whilst pancreatic trauma often presents with all the features of acute pancreatitis described above, it has some unique treatment dilemmas that warrant separate discussion.

60.8.1 Diagnosis

A history of blunt abdominal trauma to the epigastric region is frequently given. This may be in the context of a motor vehicle accident or is frequently due to an object such as a bicycle handle being caught in the upper abdomen. Examination may reveal traumatic bruising within the epigastric region. Penetrating abdominal injury is rare in children in the UK.

Non-accidental injury (NAI) must be considered especially if the history is inconsistent, the history is at odds with the clinical findings, or if any other stigmata of NAI are present.

Pancreatic injury may not be an isolated injury and associated injuries to the duodenum, small bowel, and spleen are common. The child may therefore, present with profound shock or signs of peritonitis.

Sometimes the initial pancreatic injury can go unnoticed and instead the child presents later on with a complication of pancreatic injury such as a pseudocyst.

Some degree of hyperamylaseaemia or hyperlipaseaemia is usually present, but this does depend on the severity of injury and the timing of presentation.

Abdominal ultrasound can be useful in cases of minor pancreatic trauma. However, this modality is poor for assessing injuries of the pancreatic duct and therefore CT scan with double contrast is required in most patients (Fig. 60.3). This also enables a full assessment of adjacent structures including the duodenum. MRCP can also be useful for further delineation of the duct.

Based on CT findings pancreatic trauma is classified into 4 grades of severity (see Table 60.2).

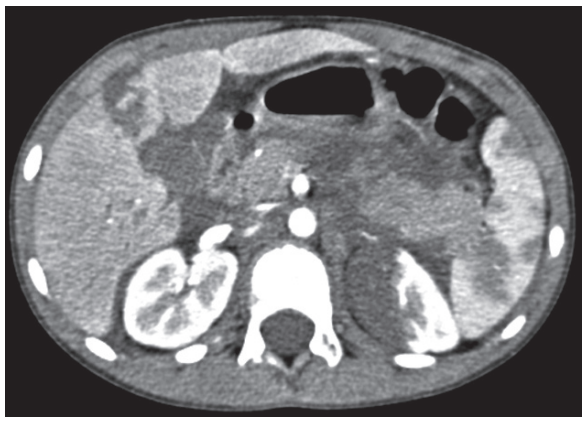

Fig. 60.3 Subtotal traumatic rupture of the pancreas and infarction of the dorsal part of the kidney after an abdominal trauma (by courtesy of the Dept. of Paediatric Radiology, Medical University of Graz)

Table 60.2 Classification of Pancreatic Injury (American Association for the Surgery of Trauma)

Grade I	Minor contusion or superficial laceration without ductal injury
Grade II	Major contusion or laceration without ductal injury or tissue loss
Grade III	Distal transection or parenchymal injury with ductal injury
Grade IV	Proximal transection (to the right of the superior mesenteric vein) or parenchymal injury involving the ampulla
Grade V	Massive disruption of pancreatic head

60.8.2 Management

As with all cases of major trauma in children, the systematic approach of advanced paediatric life support is essential. The child is carefully assessed and resuscitation started. The pancreatic injury may be an isolated injury or multiple injuries may be present and treatment is tailored accordingly.

As far as the pancreatic injury is concerned, the majority of grade I and II injuries can be treated conservatively. Close observation of the child must be continued throughout to detect clinical deterioration and repeated serum amylase or lipase levels taken and repeat imaging performed to determine whether complications such as pseudocyst have developed.

If transection of the distal pancreas has occurred, early distal pancreatectomy with splenic preservation (if possible) is advocated and is associated with good recovery in most patients. However, in cases of delayed presentation, this procedure can be difficult, and a more conservative approach is often preferable in the first instance. Treatment is then targeted at the sequelae of the acute injury such as pancreatic pseudocyst. Major injuries to the pancreatic head provide the greatest challenge. In the acute phase, major bleeding sometimes requires immediate packing and abdominal closure. If the child is haemodynamically stable and the injury to the pancreatic head is major and involves the duodenum, a pancreaticduodenectomy is indicated. Major ductal injuries with preservation of pancreatic tissue can either be managed by endoscopic stenting or by pancreaticojejunostomy (Fig. 60.4).

60.8.3 Complications

The commonest complication of pancreatic trauma is a persistent ductal leak, which can manifest itself as a pancreatic pseudocyst or pancreatic fistula (Fig. 60.5a,b). A pseudocyst is managed as for acute pancreatitis but surgery may be required if the ductal leak persists (see above). Surgery for pancreatic fistulae is only indicated after several months of conservative treatment have been attempted.

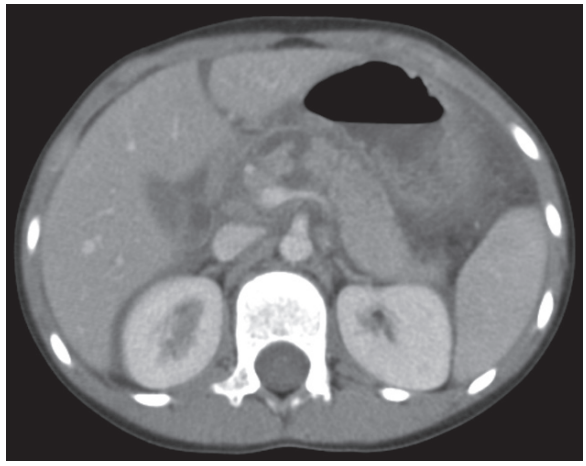

Fig. 60.4 Abdominal trauma with total disruption of the pancreatic head from the corpus. In this case the proximal part of the pancreas was closed with sutures and the distal part was anastomosed to a Roux-en-Y jejunal loop (by courtesy of the Dept. of Paediatric Radiology, Medical University of Graz)

60.9 Persistent Hyperinsulinaemic Hypoglycaemia of Infancy (PHHI)

Hyperinsulism in neonates is relatively common but is usually transient. Persistent hyperinsulinism is a rare condition affecting 1 in 50,000 live European births but can affect up to 1 in 2,800 in familial cases. A number of different terms have been used for this condition including pancreatic endocrine dysregulation syndrome (PEDS) and the more extensively used term nesidioblastosis. However, whilst the latter term was favoured by surgeons it was a pathological definition and did not take into account different forms of the disease, and as a result a few years ago the more general term PHHI was introduced. PHHI is characterised by persistent non-ketotic hypoglycaemia and the baby can present with fits and early brain damage. It is therefore imperative that all neonates are checked for hypoglycaemia immediately after birth.

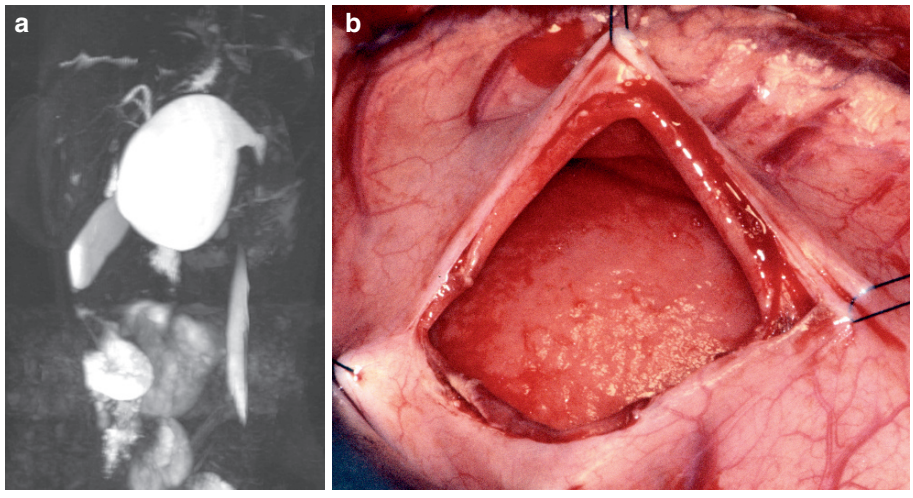

Fig. 60.5 **a** The patient of figure 60.3 developed a pancreatic pseudocyst (by courtesy of the Dept. of Paediatric Radiology, Medical University of Graz) **b** The pseudocyst was treated by a cystogastrostomy. The photograph shows the bulging of the dorsal gastric wall by the pseudocyst (by courtesy of M.E. Höllwarth)

Detailed research over recent years has helped elucidate the pathogenesis of the disease at a molecular level and also identified two subtypes of the disease namely diffuse PHHI and focal PHHI. The former is characterised by abnormal islets with large pleomorphic, hyperchromatic nuclei being present throughout the pancreas (see Fig. 60.2). Focal PHHI on the other hand, involves nodules of adenomatous islet cell hyperplasia with normal islets surrounding these areas. Familial and sporadic forms of PHHI have also been identified. The genetic defect for certain forms of this condition has been located to the gene encoding the sulphonylurea receptor on the short arm of chromosome 11.

60.9.1 Diagnosis

Diagnosis is based on the confirmation of persistent non-ketotic hypoglycaemia in the presence of hyperinsulinism, with a glucose requirement of >12.5 mg/kg/min and a hyperglycaemic response to glucagon. A number of different diagnostic tests have been advocated for imaging the PHHI pancreas and determining whether the disease is diffuse or focal. These have included serial venous sampling of insulin and more recently laparoscopic pancreatic biopsy. However, the current ‘gold standard’ is positive emission tomography (PET), which seems to locate the lesions with greatest accuracy.

60.9.2 Management

Glucose infusions are commenced immediately. The mainstay treatment for these babies is medical with drugs such as diazoxide which acts on the potassium-ATP channels within beta cells, and somatostatin analogues. Both these reduce insulin secretion and may be required for many years. Surgery is indicated for diffuse PHHI that fails to respond to medical management, in which case a 95% pancreatectomy is required, and for focal disease in which limited pancreatic resection can be curative. Surgery can be performed as an open procedure or laparoscopically.

60.9.3 Complications

The complications of delayed diagnosis of hypoglycaemia can be catastrophic. For this reason early diagnosis is vital. Prolonged treatment with diazoxide is associated with profound facial hirsutism, which can be particularly problematic for older children on long-term treatment. For those infants undergoing surgery, incomplete resection of focal disease can result in unresolved hyperinsulinism. Further resection of the remaining ‘cuff’ of pancreatic tissue is required. Persistent symptoms in babies who have undergone surgery for focal disease, is either a result of incomplete

excision of the focal area or multi-focal disease. More accurate pre-operative localisation with PET scanning will potentially minimise this problem. Even in the most experienced hands, pancreatic resection involving the proximal pancreas in neonates is associated with not inconsiderable morbidity of surrounding structures including bile duct and superior mesenteric vein. The majority of patients who have undergone 95% pancreatectomy for PHHI are insulin-dependent by the age of 20. Pancreatic insufficiency is also common.

60.10 Pancreatic Cysts

Pancreatic cysts can be congenital or acquired. Congenital cysts are either single or multiple and can be also be part of gastro-intestinal duplication (Fig. 60.6).

Single congenital cysts are most commonly present in the tail and body of the pancreas and usually do not communicate with the pancreatic duct. There is a female predominance. These developmental cysts can present at any age but the majority are diagnosed before 2 years of age. Multiple congenital cysts can be confined to the pancreas or be part of a polycystic systemic disorder such as von Hippel-Landau syndrome.

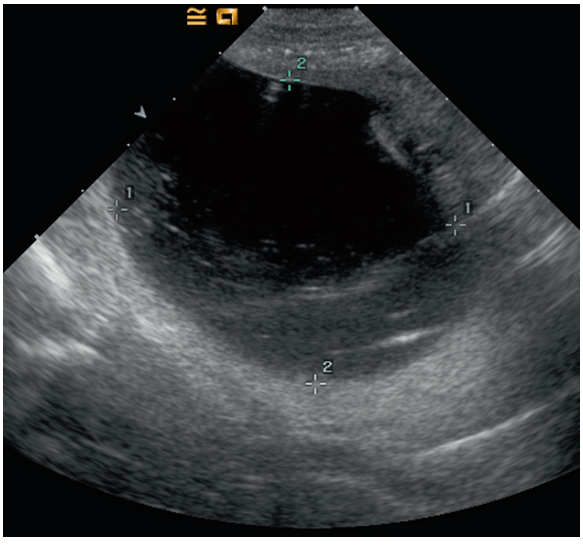

Fig. 60.6 Congenital pancreatic cyst in a newborn baby with sedimentation phenomenon on the dorsal part of the cyst. The cyst was anastomosed to a Roux-en-Y loop (by courtesy of M. E. Höllwarth)

A number of different types of acquired pancreatic cysts occur. The most frequent is a pancreatic pseudocyst (discussed above). Retention cysts secondary to pancreatic duct obstruction and tropical cysts usually associated with hydatid disease also occur.

60.10.1 Diagnosis

Clinically, such cysts may be incidental findings during physical examination or ultrasound or may present with a palpable abdominal mass. In addition, they may present with symptoms resulting from the effects of pressure on surrounding structures such as nausea and vomiting, epigastric pain, and jaundice. Abdominal ultrasound is helpful in confirming the diagnosis and CT or MRI scan help to further delineate the nature of the cyst and its anatomical relations.

60.10.2 Management

The treatment of pancreatic cysts is dependent on the exact nature of the cyst. Single congenital cysts are excised if they occur in the body or tail of the pancreas and internally drained if they are located in the pancreatic head. Small incidental, asymptomatic congenital cysts can be left and observed if neoplasia has been excluded. Symptomatic multiple congenital cysts may require extensive pancreatic resection. The treatment of pancreatic pseudocysts has already been discussed. Duplication cysts are often amenable to resection whereas the priority for hydatid cysts is to treat the underlying cause. Hydatid cysts sometime resolve spontaneously and there are also a range of surgical options including marsupilization, cystectomy, and partial pancreatectomy.

60.11 Pancreatic Tumours

Pancreatic tumours are rare in childhood but a large number of different types of pancreatic tumour can occur. These can be primary or secondary, cystic or solid, involve the exocrine or endocrine component of

the gland, and can be secretory or non-secretory. An extensive discussion of pancreatic tumours is beyond the scope of this chapter but a brief overview of primary tumours of the exocrine and endocrine pancreas will be given.

60.11.1 Tumours of the Exocrine Pancreas

Benign tumours of the exocrine pancreas include serous adenomas (microcystic and oligocystic), mucinous cystadenomas, and mature cystic teratomas. Serous adenomas do not require excision unless symptomatic but mucinous cystadenomas should undergo wide excision because they belong to the same spectrum of tumours that include mucinous cystadenocarcinoma and have definite malignant potential. This malignant tumour spreads locally in the same manner as ductal adenocarcinoma. However, if complete excision is achieved, prognosis is excellent. Ductal adenocarcinoma is rarely encountered in the paediatric age group but has been reported in teenagers. Prognosis for this condition is poor. Acinar adenocarcinoma is also rare but has a slightly better prognosis than ductal adenocarcinoma. The commonest pancreatic neoplasm in children is a pancreatoblastoma. This tumour can present at any age but the mean age for presentation is 4 years of age. The tumour is more common in males (2:1) and is associated with Beckwith-Widemann syndrome. Tumours tend to be large and solitary, and can be located in any region of the pancreas. The tumour has a mixed histology and although a malignant tumour, the presence of a capsule means that it behaves in less aggressive manner than acinar adenocarcinoma. Treatment is by radical surgical excision and metastatic disease often responds favourably to chemotherapy. Radiotherapy is used for local recurrence. Overall 30% of patients with this tumour are tumour-free 5 years after initial treatment.

Papillary-cystic tumour (also termed a Frantz tumour) is classified as being 'low-grade' malignant. Occurring predominantly in young women, these tumours present as large, round, solitary masses and can arise from any region of the pancreas. Tumours may be found incidentally or may present with abdominal discomfort. Complete excision is associated with a 95% cure rate.

60.11.2 Tumours of the Endocrine Pancreas

Both benign and malignant tumours of the endocrine pancreas can be secretory (functioning) or non-secretory. Insulinoma is the commonest functioning pancreatic endocrine tumour. Over 90% of these are benign although malignant forms do exist. Most are solitary but multiple lesions have been reported. Clinical presentation usually consists of intermittent episodes of profound hypoglycaemia. Treatment is by surgical enucleation or excision. The commonest malignant pancreatic endocrine tumour of childhood is a gastrinoma. These may occur in the pancreas itself or in extrapancreatic sites. It is associated with the Zollinger-Ellison syndrome and is also a feature of multiple endocrine neoplasia 1 syndrome (MEN 1). These are well-circumscribed, non-encapsulated lesions. Treatment is by aggressive resection which may require pancreaticoduodenectomy if the tumour is in the pancreatic head. Other endocrine pancreatic tumours encountered in children include VIPomas and islet cell carcinomas.

60.12 Conclusions

Pancreatic disorders present a number of different challenges to the paediatric surgeon. As these cases are infrequent and often complex, there is a strong case for these children to be managed in a small number of specialist centres where the facilities and expertise for investigating and treating the full range of pancreatic disorders is available.

Further Reading

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