Paediatric pancreatic problems: A five-year experience

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ABSTRACT

Background: To analyse a tertiary care experience with various pancreatic problems in the paediatric age group and clarify the frequency of various pancreatic diseases in the same patient population as well as the role of surgical or conservative management in their treatment. **Patients and Methods:** All patients with clinical suspicion of disease pertaining to the pancreas, substantiated by laboratory investigations or imaging modalities over a period of five years are included in our study. **Results:** Of the total 228 patients with various pancreatic problems, who presented to us, mostly with acute pancreatitis and pancreatic trauma, 64 needed surgical interventions. The total mortality rate encountered was a meagre 3.5%. **Conclusion:** Paediatric pancreatic disorders are not so infrequent; and a high index of suspicion aided by the newer modalities of investigation greatly helps in the appropriate and timely management.

Key words: Annular pancreas, pancreatic pseudo cysts, pancreatic problems

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INTRODUCTION

Congenital malformations like annular pancreas may present early but pancreatic divisum is difficult to diagnose without a deep suspicion of the condition.[1-3] Meanwhile, the clinical picture in acute pancreatitis may range from mild transient self limiting abdominal discomfort to fulminant, rapidly deteriorating discomfort complicated by multi organ failure with a fatal outcome within hours or days.[17] Pancreatic malignancies are even rarer to be encountered in childhood. Any child with unexplained recurrent abdominal pain should be investigated for diseases of the pancreas and moreover because of better imaging facility more and more cases are now being detected. Diagnostic imaging thus plays a major role in evaluation of the pancreas in infants and children. Familiarity with the range of normal appearances and the diseases that commonly affect this gland is important for the accurate and timely diagnosis as well as prompt surgical intervention in cases of pancreatic disorders in the paediatric population. We put forth our experience, in this regard, of five years.

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PATIENTS AND METHODS

All patients aged 17 years or below, who presented to our institute over a span of five years (2004-2009) with a clinical suspicion of pancreatic involvement, substantiated by various laboratory investigations like, serum amylase, serum lipase and other imaging modalities like ultrasonography, plain and contrast enhanced computed tomography (CT) and magnetic resonance imaging (MRI).

The criteria of suspicion was not rigid but included upper abdominal pain, multiple episodes of vomiting, abdominal lump, bleeding manifestations, failure to thrive, raised white cell counts with low platelets counts, not attributable to any obvious cause. Various laboratory investigations were performed based on requirements of a particular case vis-à-vis serum amylase; lipase was performed in almost all cases, and insulin levels in cases of nesidioblastosis. Ultrasonography (USG) and CT was performed in almost all cases while specific investigations like MRI, Magnetic resonance cholangiopancreatography (MRCP) and endoscopic retrograde cholangiopancreatography (ERCP) were performed in difficult diagnostic conditions like pancreatic divisum and pancreatic neoplasms.

All pancreatic disorders encountered, were included in our study, viz. congenital pancreatic disorders, pancreatitis and its complications, pancreatic trauma and pancreatic tumours

RESULTS

We encountered a total of 228 patients suffering from

various pancreatic disorders [Table 1]. The list is tabularly depicted. Out of these 228 patients, 64 were operated upon; the list is given in Table 2.

The overall mortality was 3.5%, of which only two patients of acute pancreatitis and pancreatic trauma died postoperatively. The mean age of study was 9.5 years with 108 females and 120 males.

DISCUSSION

With the exception of pancreas divisum, other congenital abnormalities of pancreas are extremely rare. Pancreatic anomalies are suspected early in life if an infant presents with high intestinal obstruction, neonatal hyperglycaemia, failure to thrive and maldigestion.

Annular pancreas is a rare anomaly, consisting of a band of tissue encircling the second part of duodenum,[1] 50% of them present in paediatric age group of which 85% are neonates.[2] Newborns present with symptoms of acute or sub acute duodenal obstruction and demonstrate a classical "double bubble" appearance on plain radiograph. Definitive diagnosis can only be made on laparotomy. Surgical approach is a bypass operation preferably duodeno-duodenostomy or a duodeno-jejunostomy [Figure 1]. Division of annular pancreas is not recommended due to the risk of damage to a major duct.

Table 1: Patients with pancreatic problems encountered

Disease	No. of cases
Acute pancreatitis	56
Chronic pancreatitis	28
Pseudocysts	39
Pancreatic ascites with fistula	3
Pancreatic trauma	87
Pancreatic head adenoma	5
Pancreatic divisum	1
Nesidioblastosis	1
Pancreatic head teratoma	3
Annular pancreas	1
Lymphoma	4
Total no. of events	228

Table 2: List of operated cases

Disease	No. of operations
Acute haemorrhagic pancreatitis (pancreatic divisum)	3 (1)
Chronic pancreatitis with calculi needing LPJ	2
Recurrent pain in abdomen , pancreatitis	4
Pancreatitis with biliary stone	2
Pancreatic head adenoma needing Whipple's surgery	1
Hyperinsulism needing pancreatectomy	1
Pseudo cysts	25
Pancreatic trauma	20
Pancreatic head teratoma	1
Pancreatic ascites with fistula	3
Annular pancreas	1
Lymphoma	1

Pancreas Divisum is the commonest congenital anomaly of the pancreas with an overall incidence of 4%-11% and is a common cause of acute pancreatitis in children. The associated anatomic or functional stenosis, at the minor papilla may account for increased incidence of acute or recurrent pancreatitis in these patients who lack alternate pancreatic outflow. They are diagnosed while investigating a case of unexplained recurrent abdominal pain or pancreatitis. It can be considered whenever a patient has recurrent attacks of abdominal pain associated with elevated serum amylase.[3-6] Pancreas divisum is suspected on ERCP when the duct of Wirsung fails to be visualised or is rudimentary after injection of the major papilla of Vater. A successful attempt to cannulate the minor papilla demonstrates that duct of santorini is the dominant duct and it extends to the entire length of body and tail of the pancreas. Endoscopic minor papilla sphincterotomy, balloon dilatation and stenting of duct of Santorini have been tried with variable success.[14] But if chronic pancreatitis has developed and in the presence of dilated duct with multiple stenoses, a longitudinal pancreatico-jejunostomy should be done as done in our case.[7,8] The pancreas are usually edematous and swollen [Figure 2].

Basic pathology in nesidioblastosis is diffuse B cell and islet hyperplasia.[9] It is the most common cause of persistent neonatal hypoglycaemia and 50% of neonates with persistent hypoglycaemia have hyperinsulinism.[10] Signs and symptoms include hypotonia, irritability, abnormal reflexes and convulsions and coma. Diagnosis is based on:

(a) Inappropriately raised insulin levels in relation to blood glucose levels taken at various times (insulin levels greater than 10 mU/ml and associated with simultaneous glucose level less than 50 mg/dl).
(b) Glucose infusion rate which is greater than 10 mg/kg/min required to maintain a blood glucose level above 35 mg/dl in absence of glucosuria.
(c) Low levels of plasma free fatty acids and ketone bodies during hypoglycaemia.
(d) Glycaemic response to glucagon despite hypoglycaemia.

Though frequent feeds provide a constant input of glucose, diazoxide is mainstay in medical therapy.[11] It inhibits glucose-stimulated insulin secretion. Octreotide

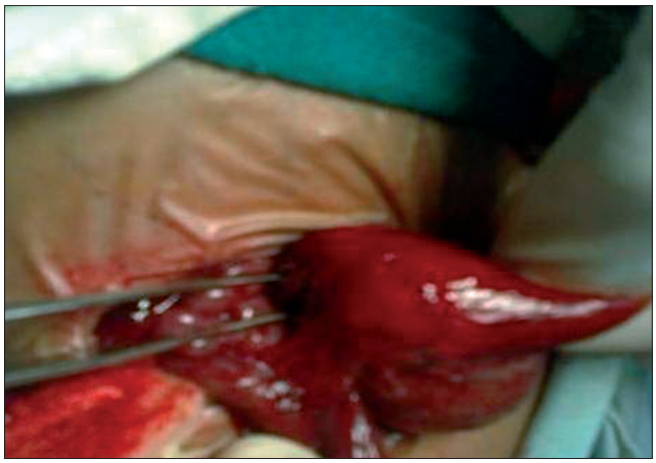


Figure 1: Duodeno-duodenostomy for annular pancreas

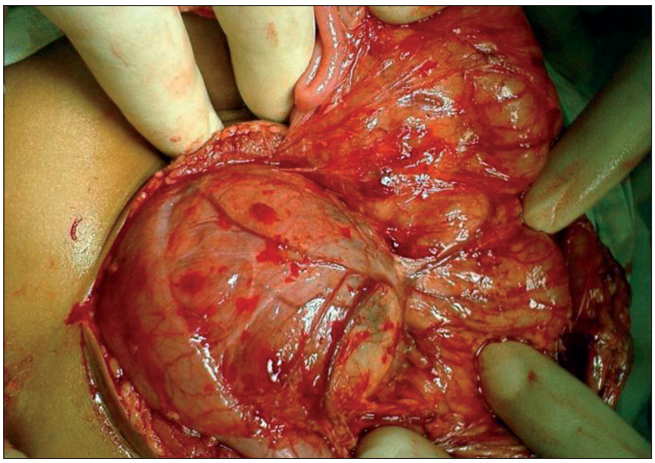


Figure 2: Edematous and swollen pancreas in a case of pancreatic divisum.

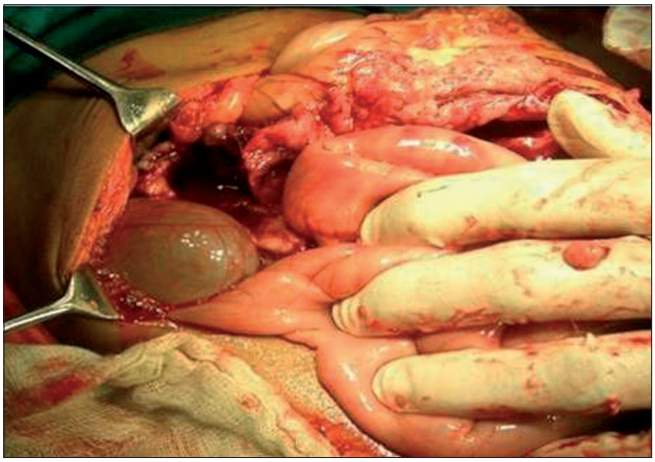


Figure 3: Pancreatico duodenectomy specimen.

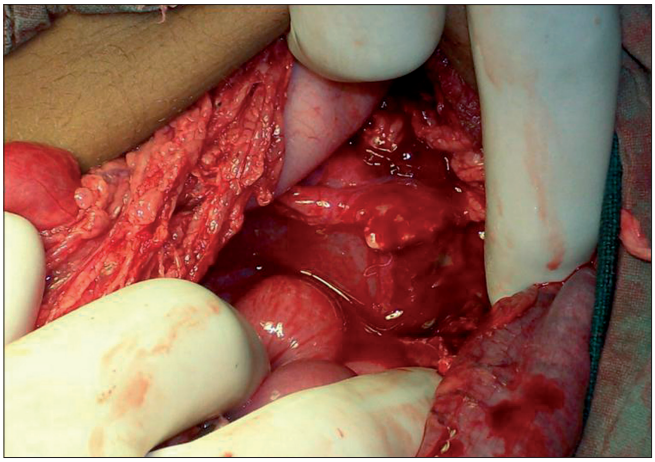


Figure 4: Solid and cystic components in a pancreatic teratoma.

inhibits insulin release and is useful for short time therapy before surgery.[12,13] Surgical treatment is indicated when infant continues to depend on intravenous glucose in spite of diazoxide and chlorthiazide, which potentiates the action of diazoxide.[14] A near total pancreatectomy is done leaving a thin rim of pancreas along the duodenum and common bile duct.

Pancreatic neoplasm cases are rare among infants and children. They can be cystic or solid and benign or malignant and may or may not be hormonally active. Cystic tumours are cystadenoma, cystadenocarcinoma, mucin cystadenoma and carcinoma or teratoma. Most endocrinologically active ones originate in the islet cells. Glucagonomas and somatostatinomas are not seen in children. USG, CT scan, arteriography and transhepatic portal vein sampling help in localising the tumour. Patients with pancreatic head adenomas or teratomas usually present with non remitting jaundice, moderate grade fever, clay coloured stools as well as with Raynaud's pentad. A CT scan, MRCP and an intra-operative cholangiogram form the imaging modalities of choice. A standard pancreatico-duodenectomy (Whipple's procedure) forms the mainstay of surgical therapy in cases of adenomas [Figure 3] whereas an enucleation procedure is mandatory in cases of teratomas with both solid and cystic components [Figure 4]. Non-Hodgkins lymphomas, being equally

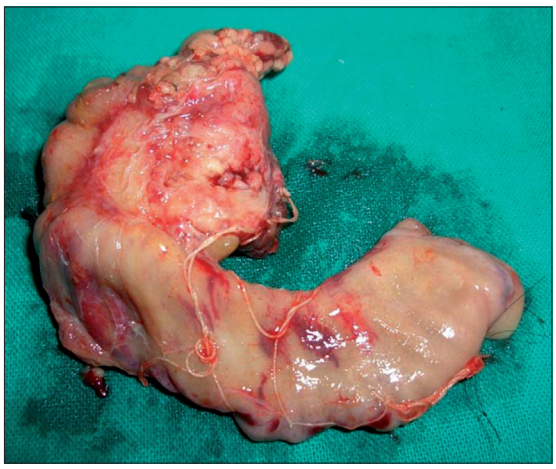


Figure 5: Non hodgkins lymphoma.

rare in this age group, have a similar presentation and require a triple bypass procedure in their management [Figure 5].

The most common causes of acute pancreatitis in children are systemic infection, trauma, congenital malformation of pancreatic or bile ducts or drugs,[15,16] less commonly, metabolic disorders and familial pancreatitis. About a third of cases of pancreatitis are related to disorders of the biliary or pancreatic ducts with choledochal cyst being most common. Anomalous pancreatico-biliary duct junction (ABPDJ), with its long common channel, permits reflux of bile into the pancreatic duct.[17,18] Several viral infections are known to cause acute pancreatitis.[19-22] The typical radiation of the pain to the back observed in adults is missing in 60-90% of children.[23,24] Pain is associated with anorexia, nausea and vomiting. In severe disease, the child is irritable and assumes a flexed position. The abdomen may be distended with guarding and rigidity present. In very severe cases child may present with shock, respiratory distress or multi organ failure or unexplained jaundice or ascites. When necrosis, abscess or pseudo cyst develop, the presentation may be with a mass in the epigastrium.

Serum amylase is the widely used test in acute pancreatitis. It may be normal in the first 24 hours; peak between 24 and 72 hours and in uncomplicated cases, remain elevated for two to five days. Serum lipase levels are usually elevated in pancreatitis and remain elevated for longer time than amylase. USG is the most useful investigation and may show increased pancreatic size and decreased echogenicity. It may also show peripancreatic fluid collection. Contrast enhanced dynamic CT scan is the imaging modality of choice; it shows the size, texture of inflamed pancreas, peripancreatic fluid collection, abscess, pseudo cyst, ductal dilatation, calcification and perfusion defects of the pancreas to differentiate interstitial pancreatitis from necrotising pancreatitis. Treatment of acute pancreatitis is largely supportive in the form of fluid resuscitation and child is deprived of oral intake with nasogastric suction. Full monitoring of central venous pressure (CVP), intake and output, blood gases, serum electrolytes, creatinine, calcium, glucose and lipid profile were done. Parenteral analgesics using either narcotic or NSAID should be started even in mild pancreatitis because pain can be extreme. Measures to put the pancreas to rest such as H2 blockers, atropine, calcitonin, glucagons and stomatostatin have not shown to change course of disease. Recent studies have shown that some antibiotics like cefuroxime, ciprofloxacin and imipenem cross blood pancreatic barrier and are found in high level in pancreas and within the necrotic pancreatic tissue.[25] It has been shown that in severe acute pancreatitis, it can reduce the incidence of septic complications.[26,27] Total parenteral nutrition is indicated if extended period of fasting is anticipated. Re feeding can be started when pain and tenderness is reduced, initially with carbohydrates. Complications include respiratory failure, shock, renal failure, disseminated intravascular coagulation, which indicate that the patient has severe pancreatitis. Sterile or infected pancreatic and peripancreatic necrosis can be distinguished by USG or CT guided fine needle aspiration.[28] Late complications include pseudo cyst and abscess formation.[29] These inflammatory masses develop in about 15% of children. Surgery is indicated if diagnosis is in doubt, if patient develops infected necrosis or in sterile necrotising pancreatitis; if patient deteriorates in spite of adequate medical management or if there is pancreatic abscess or the patient develops a pseudo cyst. Infected pancreatic necrosis needs debridement and multiple drainage of lesser sac.

Pancreatic pseudo cysts are a contained collection of fluid, rich in pancreatic enzymes, not lined by epithelium. The most common site is in lesser sac behind the stomach. They occur in 20% of cases of acute or chronic pancreatitis. Most common cause of pseudo cyst in children is after trauma or infection.[30] It may or may not communicate with the pancreatic duct system. The fluid is usually clear with some debris and amylase level is always > 50,000 somogyi units. Most pseudo cysts resolve over a course of six weeks, particularly associated with acute pancreatitis. Pseudo cysts that persist beyond six weeks, those larger than six centimetres and those associated with chronic pancreatitis are less likely to resolve spontaneously and require some degree of surgical intervention. Rupture, infection and haemorrhage are the complications that can occur in a pseudo cyst. For many years the standard treatment has been laparotomy and internal drainage of the cyst into the gastrointestinal tract. CT-guided percutaneous aspiration and endoscopic cysto gastrostomies are the newer methods of drainage.

Diagnosis of pancreatic trauma can be difficult in a stable blunt trauma patient. The value of serially rising amylase level is stressed. Serial CT scan is usually necessary to reveal parenchymal disruption. Prudent approach is to operate early as there is higher mortality of delayed surgical intervention after unsuccessful conservative management.[31] Major injuries at the body and tail of pancreas are best managed with low

complication rate by distal pancreatectomy with or without splenectomy. Debridement, wide drainage and local control of haemorrhage are all needed in unstable patient. Pancreatic head disruptions requiring pancreaticoduodenectomy are extremely rare, probably due to concomitant major vessel disruption causing death.[32]

CONCLUSION

Diseased pancreas is no more a domain of adults. High index of suspicion is needed for prompt diagnosis and subsequent timely intervention in children. Conditions like those enumerated and discussed above must be borne in mind whilst dealing with unexplained or unremitting abdominal pain in a child. Though pancreatic tumours are rare in children but these can also be found. Thus a high index of suspicion is a must for an accurate and timely management.

As the saying goes- '***what the mind knows is what the eyes see'***.

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