Agenesis of the dorsal pancreas (ADP) is a rare congenital anomaly. We describe four patients with ADP presenting with acute pancreatitis. One patient had diabetes mellitus and another had malrotation of duodenum as an additional feature. All patients improved on conservative management. [Indian J Gastroenterol 2006;25:35-36]

Agenesis of the dorsal pancreas (ADP) is a rare congenital defect. It may remain asymptomatic, or present as an incidental finding or with clinical symptoms. We report four patients with ADP presenting with pancreatitis, seen over a period of 5 years.

**Case Reports**

**Case 1:** A 25-year-old man presented with epigastric pain associated with vomiting over a three-month period. The pain was continuous and non-radiating in nature. He had a history of diabetes mellitus requiring insulin therapy for 4 years. On examination, there was mild epigastric tenderness. Investigations: raised total white count, and amylase and lipase levels (1800 U/L and 1480 U/L; normal values 0-200 and 0-190, respectively) consistent with pancreatitis. Ultrasonography revealed mild peripancreatic edema; the body and tail of the pancreas could not be visualized. CT scan three weeks later revealed normal head of pancreas; the body and tail of pancreas were absent. ERCP demonstrated filling of the normal ventral duct only. The minor papilla was not visualized despite careful examination. The common bile duct was normal. The patient was treated conservatively and is well at 1 year.

**Case 2:** A 19-year-old man presented with non-radiating epigastric pain and vomiting. He had experienced similar attacks over the past five years. There was no history of diabetes mellitus. Physical examination revealed mild epigastric tenderness. Investigations: elevated serum amylase and lipase levels (1200 U/L and 860 U/L, respectively). Ultrasonography revealed only partial visualization of the pancreas. CT scan showed absence of the tail and body of the pancreas; in addition, the superior mesenteric artery and vein were located anterior to the pylorus and ventral pancreas. Magnetic resonance cholangio-pancreatography (MRCP) revealed hypoplasia of the distal body and tail of the pancreas and absent main pancreatic duct. There was malrotation of the duodenum; the “C-loop” was directed cephalad and anteriorly. ERCP demonstrated ectatic ducts limited to the head. The minor papilla was absent. The patient was treated conservatively and is well at 3 years.

**Case 3:** A 35-year-old woman presented with recurrent episodes of epigastric pain. The pain was continuous and radiated to the back. Physical examination revealed epigastric tenderness. Investigations: high amylase and lipase levels (750 U/L and 920 U/L, respectively). Ultrasonography revealed mild peripancreatic edema. CT abdomen (Fig) and MRCP performed earlier at other hospitals revealed hypoplasia of the body and tail of the pancreas. ERCP confirmed the presence of normal ventral pancreas. The minor papilla was absent. The patient was treated conservatively; she is well at 4 years.

**Case 4:** A 38-year-old man presented with recurrent epigastric pain, continuous and radiating to the back. Physical examination revealed epigastric tenderness. Investigations: high amylase and lipase levels (840 U/L and 475 U/L, respectively). Ultrasonography showed partial visualization of pancreas. CT abdomen revealed the pancreatic duct only till the head region. ERCP showed filling of only the ventral duct of pancreas. The minor papilla was absent. The patient was treated conservatively; he is well at 2½ years.

In all our cases, absence of the dorsal pancreatic ductal system, accessory duct and minor papilla was documented. Lipid profile, serum calcium and renal function tests were normal. There was no family history of stillbirths or early infant death, or history of recurrent infections. The patients did not have skeletal or dental defects. Echocardiography in all patients did not show significant cardiac defects.

**Discussion**

Pancreatic structural abnormalities have been grouped into partial hypoplasia, malfusion and malrotation.1
Complete agenesis of the pancreas is incompatible with life. Partial hypoplasia would result in congenital short pancreas or short pancreas divisum. Agenesis of the dorsal pancreatic bud results in complete absence of the dorsal ductal system. Pancreas divisum is the commonest pancreatic developmental anomaly encountered. A combination of developmental abnormalities may occur, as exemplified by our second patient, who had malrotation of the duodenum and agenesis of the dorsal pancreas.

Accurate diagnosis of ADP requires exclusion of pancreas divisum, congenital short pancreas and congenital short pancreas divisum. Less common mimics include pseudo-agenesis of the pancreas, pancreatic pseudolipodystrophy and obstructing pancreatic tumors. ERCP and radiological imaging are complementary. ERCP is necessary to differentiate ADP from “partial agenesis”. A diagnostic triad is required, documenting absence of the dorsal ductal system, the accessory duct (Santorini) and the minor papilla. The absence of the body and tail of the pancreas is best demonstrated on CT scan, MRI or MRCP. With increasing availability of MRCP, awareness of ADP is required.

ADP results from developmental failure of the dorsal pancreas. This condition is very rare; a total of 15 cases have been reported since 1913 till date. Abdominal pain and diabetes mellitus are commonly reported. At week 12 of embryogenesis, discrete islets of Langerhans form primarily within the tail of the pancreas and the dorsal pancreas. The number of insulin cells increases with age. Diabetes mellitus is thus associated with reduced islet cell mass secondary to absence of the body and tail of pancreas.

The association of ADP and pancreatitis is less well defined. Three prior cases were reported to have pancreatitis. One case presented with obstructive jaundice and was found histologically to have chronic pancreatitis; the second had symptomatic pancreatitis. All our patients presented with symptomatic pancreatitis.

Two possible mechanisms contributing to pancreatitis are proposed. Sphincter of Oddi dysfunction may play a role in the pathophysiology of dorsal pancreatic hypoplasia and pancreaticobiliary diseases associated with it. Alternatively, compensatory hyper-secretion would result in hypertrophy of the remnant ventral gland and higher intrapancreatic duct pressures. In isolation, these changes may not result in pancreatitis. However, the role of genes in the pathogenesis of acute and chronic pancreatitis is increasingly recognized. In southern India, genetic predisposition results in a high incidence of pancreatitis. The combination of increased genetic susceptibility and congenital anomaly would explain the occurrence of pancreatitis in our patients.

In summary, the interactive roles of genetic and environmental factors in the pathogenesis of chronic pancreatitis have become increasingly recognized. In our patients, congenital agenesis of the dorsal pancreas contributes as a rare inherited cause of pancreatitis.

References

Correspondence to: Dr Reddy. Fax: (40) 2332 4255. E-mail: aigindia@yahoo.co.in
Received June 8, 2005. Accepted September 16, 2005