Purpose: Pancreaticobiliary anomalies represent uncommon variants of pancreatic ductal development. Patients affected by these anomalies often present with cryptic signs and symptoms which can be missed or poorly appreciated, thus delaying or even preventing appropriate treatment.

Methods: Endoscopic retrograde cholangiopancreatography (ERCP) was used to define pancreaticobiliary anomalies. A retrospective review was performed of 5522 ERCPs done at a tertiary care center by a single surgical endoscopist from 1972 to 2015.

Results: There were 297 (5%) patients with pancreaticobiliary anomalies including 179 patients with pancreas divisum (PD), 31 patients with congenital biliary dilatation (CBD) (Todani’s type I cyst), 20 patients with anomalous pancreaticobiliary ductal union (APDU), 9 patients with choledochocele (Todani’s type III cyst), 4 patients with Caroli’s disease (Todani’s type V cyst), 5 patients with other abnormalities.

Among 179 patients with pancreas divisum, 59 patients (33%) had pancreatitis, and 8 (4.5%) required minor sphincterotomies for multiple unexplained acute pancreatitis.

Among patients with CBD, 7 had an associated APDU, and 4 had an associated PD. 15 (48%) patients required an operation, including 4 who underwent a choledochoectomy and hepatico-jejunostomy, 3 who had a cholecystectomy with choledochoduodenostomy, 3 who had major sphincterotomies, and 5 who had a cholecystectomy alone.

Of the 20 patients with APDU, 7 (35%) had associated CBD, and 2 (10%) had associated PD. 8 (40%) required operative intervention, which was comprised of 3 cholecystectomies, 1 cholecystectomy with lymphadenectomy, 2 endoscopic sphincterotomies, and 2 choledochoectomies with hepatico-jejunostomy.

20 patients were found to have other anomalies, including 9 patients with choledochocele, 4 with Caroli’s disease, 3 with pancreas bifidum, and 1 patient each with a duplication of the gallbladder, a cystic duct diverticulum, an annular pancreas, and an abnormal cystic duct origin.

Conclusion: Pancreaticobiliary anomalies are rare, and can be defined using ERCP. When symptomatic, these anomalies require surgical or endoscopic therapies. Minor sphincterotomy for pancreas divisum is performed for selected patients with recurrent bouts of acute pancreatitis. Asymptomatic APDU and CBD do pose increased risk of malignancy, and when identified, benefit prophylactic interventions. The appreciation of these abnormalities is important for the proper diagnosis and treatment of these rare biliary and pancreatic disorders.
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2013 - SAGES Jeffery L. Ponsky Master Educator in Endoscopy Award