Pancreaticobiliary maljunction and choledochal cysts: from embryogenesis to therapeutics aspects.

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**Résumé:**  
Pancreaticobiliary maljunction (PBM) and choledochal cysts (CC) are rare and little-known diseases. Several definitions have been proposed for the PBM, but the most widely accepted is an excessive length of the common pancreaticobiliary duct due to the abnormal convergence of the pancreatic and biliary ducts out of the duodenal wall. This anomaly, thought to develop during embryogenesis, is associated with a loss of regulation of the Oddi’s sphincter leading to a pancreaticobiliary or biliopancreatic backflow. This reflux could be responsible, or associated with cystic dilatation of the bile ducts and biliary tract cancers, to various biliary or pancreatic events such as cholangitis or pancreatitis. For the diagnosis of PBM, magnetic resonance cholangiopancreatography has now become the gold standard as a noninvasive imaging tool. However, the main risk of PBM is the development of bile duct cancer, most often on a distended area. PBM without CC increase the occurrence of gallbladder cancer and require a preventive cholecystectomy. Surgical treatment of PBM with concomitant CC is more complex and depends on localization of the dilatation(s) as reported in the Todani's classification. This review describes the pathogenesis, embryogenesis, clinical features, investigation and management of PBM and CC.

**MeSH:** Bile Duct Neoplasms/etiology|Bile Ducts, Extrahepatic/abnormalities|Cholangiopancreatography, Magnetic Resonance|Choledochal Cyst/diagnostic imaging/embryology/surgery|Humans  

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