Spectrum of congenital pancreatico-biliary ductal anomalies - Demonstration by MRCP positively impacts the management protocol

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Learning objectives

Congenital pancreatic and biliary ductal anomalies are not infrequently encountered at imaging. Clinically, these conditions are uncommonly symptomatic but may present themselves with associated conditions ranging from benign acute abdominal pain to carcinomas [1]. Recent developments in both imaging techniques and endoscopic manipulations together with increasing awareness have improved the clinical diagnosis and management of congenital pancreatico-biliary ductal anomalies and the associated complications. The purpose of this educational exhibit is to illustrate the spectrum and unusual presentations of congenital pancreatico-biliary ductal anomalies using MRCP and to outline the advantages of the technique which alter the patient management.
Background

Magnetic resonance cholangiopancreatography (MRCP) is a technique that has evolved over the past two decades. It continues to have a fundamental role in the noninvasive investigation of many pancreatico-biliary disorders. Advances in MR imaging techniques and technology have improved MRCP image quality greatly. This has led to an increased recognition of already described and certain hitherto undescribed congenital pancreato-biliary ductal anomalies.

Congenital anomalies and normal variants involving the biliary tract include aberrant or accessory biliary ducts, aberrant cystic duct insertion; bile duct cysts, alterations of the biliary tract associated with situs anomalies, and anomalous junction of the common bile duct with the pancreatic duct. Congenital anomalies of pancreatic duct include pancreas divisum, annular pancreas and variations of pancreatic ducts. Recognition of these entities as anomalies and normal variants may avoid diagnostic errors, aid in surgical planning, and prevent inadvertent ductal injury.

Aberrant Biliary Ducts:

The RPD is by far the most anomalous in terms of insertion site. Drainage of the RPD into the LHD before its confluence with the RAD is the most common anatomic variant of the biliary system and has been reported to occur in 13%-19% of the population. In approximately 12% of the population, the RPD will not pass the RAD posteriorly, but will empty into the right aspect of the RAD (2). The so-called "triple confluence" is another common variant of the main hepatic biliary branches (11% of the population). This anomaly is characterized by simultaneous emptying of the RPD, RAD, and LHD into the CHD to form a trifurcation. These variants at the level of the confluence become important in patients being considered as potential donors for right hepatic lobe transplantation.

Aberrant Cystic Duct Insertion:

There are three common variants in the cystic ductal anatomy: a low cystic duct insertion, characterized by fusion of the cystic duct with the distal third of the EBD (9% of cases); a medial cystic duct insertion, in which the cystic duct drains into the left side of the CHD (10%-17%); and a parallel course of the cystic duct with the CHD (1.5%-25%), which is thought to be present when the cystic duct closely adheres to the CHD and courses parallel to it for at least 2 cm. Familiarity with these variants is important prior to laparoscopic cholecystectomy due to the risk of injury to the cystic and hepatic ducts posed by this procedure.

Bile Duct Cysts:

Choledochal cysts are rare congenital biliary tract anomalies characterized by biliary tree dilatation. The five subtypes as defined by Todani probably have different
pathophysiologic features but are grouped together according to the cystic changes they have in common.

Type 1: Choledochal Cyst

Type 1 bile duct cysts account for 80% to 90% of all bile duct cysts and are characterized by fusiform dilation of the extrahepatic bile duct. It is theorized that choledochal cysts form as the result of reflux of pancreatic secretions into the bile duct via an anomalous junction of the common bile and pancreatic ducts. The classic triad of jaundice, abdominal pain, and mass is not commonly seen in adult patients. Because the cyst should be resected completely to prevent associated complications, such as ascending cholangitis, cystolithiasis, and malignant transformation [3], accurate determination of the length of the extrahepatic bile duct involved by the cyst is crucial in surgical planning. MRCP is an effective imaging technique for diagnosis and preoperative evaluation of bile duct cysts [4]

Type 2: Diverticulum

Type 2 bile duct cysts account for 3% of all bile duct cysts and represent a true diverticulum. At imaging, type 2 bile duct cysts are seen as saccular outpouchings arising from the supraduodenal extrahepatic bile duct or the intrahepatic bile ducts.

Type 3: Choledochocele

Choledochoceles account for 5% of all bile duct cysts and represent protrusion of a focally dilated, intramural segment of the distal common bile duct into the duodenum. Although choledochoceles may manifest in adults with episodic abdominal pain, jaundice, nausea, and vomiting, many are detected incidentally in patients without symptoms referable to the biliary tract. MRCP reveal a bulbous dilation of the intramural portion of the bile duct bulging into the duodenum. Choledochoceles may be successfully managed with endoscopic sphincterotomy, surgical excision, or both, in symptomatic patients.

Type 4: Multiple Communicating Intra and Extrahepatic Duct Cysts

Type 4 biliary cysts represent the second most common type of bile duct cysts (10%) and are subdivided into subtypes A and B. Type 4A is characterized by fusiform dilation of the entire extrahepatic bile duct with extension of dilation of the intrahepatic bile ducts. Type4B cysts are extremely rare and are seen as multiple cystic dilations involving only the extrahepatic bile duct.

Type 5: Caroli’s Disease

Carolí's disease is a rare form of congenital biliary cystic disease manifested by cystic dilations of intrahepatic bile ducts that may diffusely involve the right and left hepatic ducts [5]. There is an association with benign renal tubular ectasia and other forms of renal cystic disease. MRCP feature of Carolí’s disease is cystic dilation of the intrahepatic bile ducts seen in conjunction with a normal extrahepatic bile duct.
Anomalous Junction of the Biliary Ductal System with Pancreatic Duct:

Anomalous junction of the pancreaticobiliary ductal system is usually defined as a union of the distal common bile duct and the pancreatic duct proximal to the duodenum whose length is greater than 1.5 cm [6]. Anomalous junction of the pancreaticobiliary ductal system has been reported in association with cholangiocarcinoma, gallbladder carcinoma, choledocholithiasis, and chronic pancreatitis. In this entity, pancreatic secretions can reflux into the common bile duct and can result in the development of a bile duct cyst (type I).

Pancreas Divisum:

Pancreatic ductal anatomy can be subject to a number of variations with pancreas divisum being the most common congenital pancreatic ductal anatomic variant. The abnormality results from failure of the dorsal and ventral pancreatic anlage to fuse during the sixth to eighth weeks of gestation. In most cases of pancreatic divisum, no communication exists between the dorsal and ventral pancreatic ducts. In some patients, the ventral pancreatic duct may be absent. In all cases, most pancreatic secretions drain through the minor ampulla. The main features of pancreas divisum when using MRCP include the dorsal pancreatic duct in direct continuity with the duct of Santorini, which drains into the minor ampulla, and a ventral duct, which does not communicate with the dorsal duct but joins with the distal bile duct to enter the major ampulla.

Annular Pancreas:

Annular pancreas is a rare anomaly in which a band of pancreatic tissue surrounds the descending duodenum, either completely or incompletely, and is in continuity with the head of the pancreas. The most widely accepted theory of etiopathogenesis is that the ventral pancreatic anlage is responsible for the anomaly by dividing early into two segments. The anomaly may be discovered incidentally in asymptomatic patients [6]. In others, annular pancreas is associated with duodenal stenosis, postbulbar ulcerations, pancreatitis, or biliary obstruction. Before the advent of CT, MRI, and MRCP, the diagnosis of annular pancreas was usually established by ERCP, as an aberrant pancreatic duct communicating with the main pancreatic duct and encircling the duodenum. MR images may show normal pancreatic tissue, with or without a small pancreatic duct, encircling the duodenum. Surgical resection is recommended for symptomatic cases.
Patients: Retrospective analysis of MRCP was done from the imaging archives, in our university based tertiary care center. Investigations done between January 2011 and October 2011 were evaluated, which resulted in viewing of 197 MRCP examination and we came across 20 cases of pancreato-biliary ductal anomalies.

Technique: The routine protocol for the said investigation at our center includes patients fasting for 4 hours prior to the study (in order to reduce fluid secretions within the stomach and duodenum, reduce bowel peristalsis and promote gallbladder distension). MRCP was performed on a 1.5T MRI system (Seimens Magnetom Avanto, Erlangen, Germany) using a phased array body coil. Initially, an axial 2D breath-hold HASTE sequence is performed to include the whole of the liver down to the duodenal ampulla in two breath-hold acquisitions. Following this, we perform two 3D respiratory-triggered heavily T2-weighted FSE sequences in the coronal oblique planes. The imaging plane is selected from the initial axial T2-weighted images, with one acquisition aligned to the common bile duct (CBD) in the head of the pancreas and the second acquisition aligned to the pancreatic duct at approximately 90 degrees to the first imaging plane. Respiratory triggering is achieved with the use of a navigator sequence that employs an MR pre-pulse to monitor respiratory motion. The navigator is placed over the edge of the diaphragm on the coronal and sagittal localizers and image acquisition is triggered when the position of this diaphragm interface with the lung falls within a pre-specified acceptance window. In this way a consistent position of the imaging slice is obtained. The patient is asked to breath regularly throughout this acquisition, which takes between 3-5 min to acquire. A stack of 40 slices are obtained, which are contiguous and each of 1.5 mm in thickness. From this volume of data, a MIP reformat can be generated. We conventionally create 18 MIP reformats at 10-degree intervals to each other over a radial array of 180 degrees. In addition to, or as an alternative to the MIP reformats, a thick collimation slab is obtained in the coronal plane. This involves performing a fat saturated HASTE sequence where a single slab of data 4 cm in thickness is acquired in a 1 to 2-s breath-hold. In order to evaluate the duct walls, and any focal parenchymal pathology, 3D fat suppressed T1-weighted GRE sequences before and after intravenous contrast administration is performed.

Results: Out of 20 patients, we had 6 cases of choledochal cysts, 5 cases of anomalous PB junction, 3 cases of low insertion of cystic duct, 2 cases of pancreatic divisum, 1 case each of annular pancreas, pancreatic ductal duplication and anomalies insertion of RAD to left HD. One unusual case of ectopic opening of CBD into minor duodenal papilla was found.
Fig. 8: Coronal MRCP image showing fusiform dilatation of CBD S/O type 1 choledochal cyst

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Fig. 7: coronal MRCP image showing type 1 choledochal cyst with long common channel
S/O anomalous pancreatico-biliary junction

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**Fig. 2:** MIP MRCP images showing drainage of pancreatic duct through minor papilla S/O pancreas divisum. Also noted is the complete drainage of uncinate process through the minor papilla. (DD: dorsal duct, VD: ventral duct, CBD: common bile duct)

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Fig. 1: An 18-year-old female with a clinical history of recurrent pancreatitis coronal MRCP images showing drainage of pancreatic duct through minor papilla S/O pancreas divisum.

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Fig. 3: A young male presented with long history of recurrent vomiting immediately after food intake with distension of abdomen. Axial T1W mage showing presence of pancreatic tissue (arrow) encircling 2nd part of duodenum.

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Fig. 10: MRCP image shows prominent CBD upto its lower end with its termination at the minor papilla. Also noted was duplication (arrows) of main PD in the pancreatic head region. A diagnosis of ectopic insertion of CBD into minor papilla with duplicated PD was made

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Fig. 9: A 45 year old male patient presented with a history recurrent jaundice. The patient was a known hypertensive and was diagnosed to have chronic renal parenchymal disease. There was H/O failed ERCP. coronal MRCP image shows moderate ascites with prominent CBD upto its lower end with its termination at the minor papilla (arrow).
**Fig. 6:** Post Fentanyl MRCP shows distension of pancreato-biliary ductal system due to sphincter spasm with visualization of part of annular duct (arrow)

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Fig. 5: Pre fentanyl MRCP showing normal pancreatic and biliary ducts

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Fig. 4: A young male presented with long history of recurrent vomiting immediately after food intake with distension of abdomen. Axial T1W mage showing presence of pancreatic tissue (arrow) encircling 2nd part of duodenum.

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Fig. 11: Repeat ERCP through minor papilla was later successful which shows hook shaped CBD confirming our MRCP diagnosis

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Conclusion

Knowledge of embryological and anatomical details of the pancreato-biliary ductal system is important in diagnosis and management of congenital pancreato-biliary ductal anomalies, particularly in unusual cases and can be effectively carried out noninvasively by MRCP.
References


