Chapter 14
Diagnosis of PBM and CBD by MRCP

Shigehisa Fumino and Tatsuro Tajiri

Abstract A diagnostic work-up of pancreaticobiliary maljunction (PBM) and congenital biliary dilatation (CBD) depends on noninvasive imaging modalities rather than direct cholangiography. Magnetic resonance cholangiopancreatography (MRCP) is widely used for hepatobiliary and pancreatic disease and should be considered the first-line imaging test for PBM and CBD after ultrasonography in current clinical practice. The advantages of MRCP over computed tomography and endoscopic retrograde cholangiopancreatography in such cases include its excellent contrast resolution, low invasiveness, and lack of irradiation. However, it is still challenging to perform high-quality MRCP in children, especially very young children, due to these patients' small-caliber ducts, a poor signal, and unavoidable patient motion, which creates artifacts. MRCP was able to visualize PBM in only 44.4% of cases, and the minimum age for successful visualization of PBM with MRCP was 1 year and 11 months in the authors' series. Recent technical improvements in the image quality may lead to better diagnostic accuracy of MRCP in young patients in the near future.

Keywords Magnetic resonance cholangiopancreatography (MRCP) · Endoscopic retrograde cholangiopancreatography (ERCP) · Pancreaticobiliary maljunction (PBM) · Congenital biliary dilatation (CBD) · Protein plug

14.1 Perspective of MRCP

A diagnostic work-up of pancreaticobiliary maljunction (PBM) and congenital biliary dilatation (CBD) depends on noninvasive imaging modalities rather than direct cholangiography. Magnetic resonance cholangiopancreatography (MRCP) is

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widely used for hepatobiliary and pancreatic disease and should be considered the first-line imaging test for PBM and CBD after ultrasonography in current clinical practice [1].

Magnetic resonance imaging (MRI) depends on the detection of energy released from hydrogen protons after their forcible alignment in a strong field. The technique is safe with certain provisos (i.e., patients with claustrophobia or metal foreign bodies, such as pacemakers and stainless plates). MRI has excellent contrast resolution, better than that of computed tomography (CT), although worse spatial resolution than CT. Multiple planes (axial, coronal, sagittal) can be reconstructed as needed. For MRCP, the bile within the biliary tree is imaged with heavily T2-weighted sequences without contrast medium. The sequences are heavily T2 weighted using long echo times in the range of 300–1000 msec, so that only tissues or fluid with a prolonged transverse relaxation time (T2) retains the signal. These tissues and fluid are seen as hyperintense structures. The background soft tissues with a shorter T2 do not retain a significant signal long enough in a sequence with a prolonged echo time and are, therefore, suppressed. Blood vessels are not seen, since flowing blood does not produce any signal on these images. Therefore, MRCP can depict the overall biliary system, including the intrahepatic and extrahepatic bile ducts as well as PBM (see Fig. 14.1) [2].

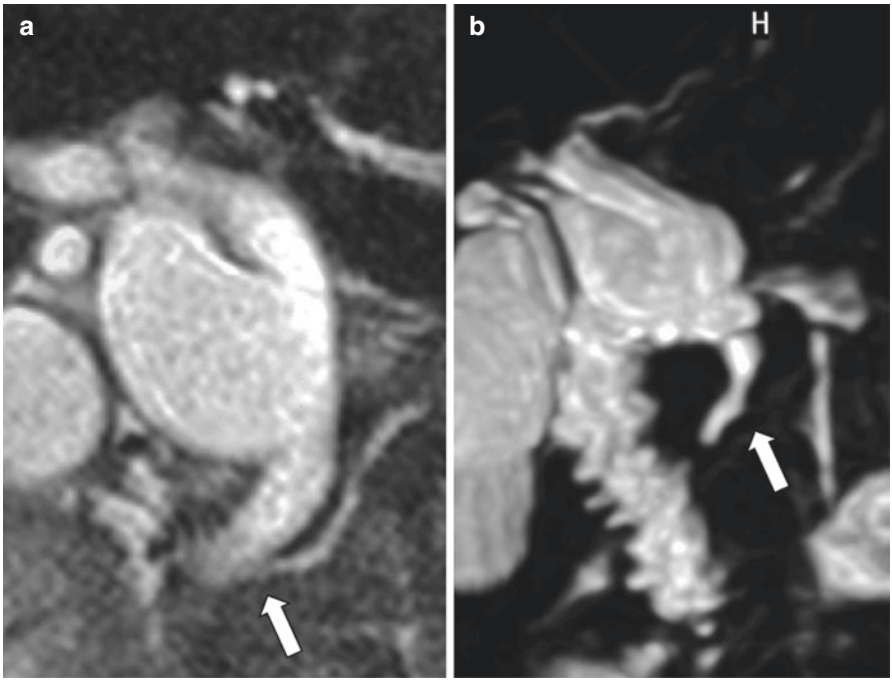

Fig. 14.1 (**a**) A 3-year-old boy with cystic-type choledochal dilatation. MRCP showed clearly PBM (arrow). (**b**) A 23-month-old girl with cystic-type choledochal dilatation. MRCP showed clearly PBM (arrow)

14.2 Diagnostic Purpose and Accuracy of MRCP for CBD and PBM

The purposes of imaging studies for PBM and CBD are principally classified into four items [3].

1. First, the most important role of imaging is the total evaluation of the biliary system. This is important because the presence of a choledochal cyst must be confirmed, and these obtained images provide a road map for surgical planning. Furthermore, the morphological changes in the intrahepatic bile duct, such as stenosis and enlargement, must be assessed simultaneously.
2. Imaging also allows for the evaluation of the pancreatic system. Using imaging, one can visualize the changes of the pancreas parenchyma, the dilatation of the pancreatic duct, and the presence of a protein plug (see Fig. 14.2). This may be accompanied by pancreas divisum or an annular pancreas.
3. Imaging also allows for the demonstration of PBM, which is necessary particularly for the diagnosis of non-dilatation-type PBM to assess surgical indications.
4. Imaging allows for functional evaluations. In PBM, two-way regurgitation can occur with pancreatic juice reflux into the bile duct or bile juice regurgitation into the pancreatic duct. Contrast material-enhanced MRCP and dynamic MRCP with secretin have been reported useful in this regard [4].

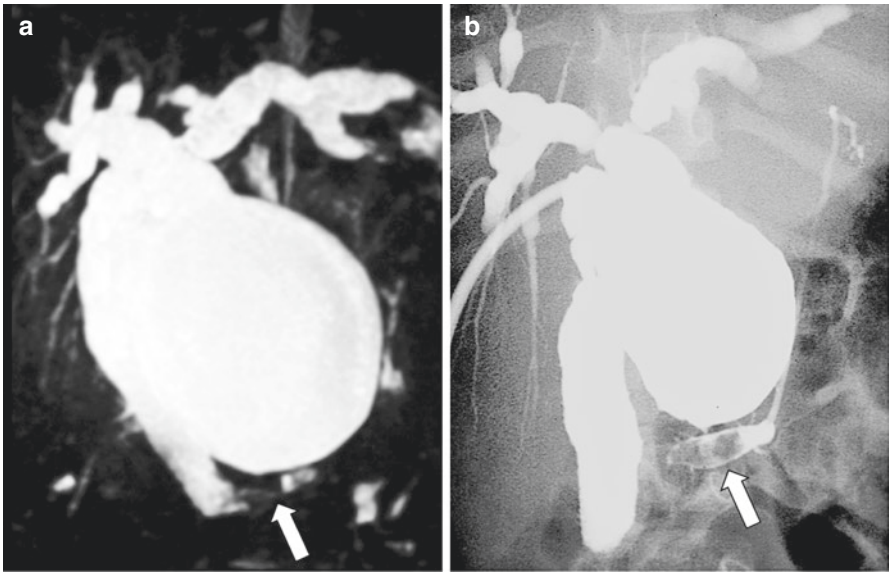

Fig. 14.2 (**a**) Pre-drainage MRCP showed a protein plug (arrow) within a common channel in a cystic-type CBD. (**b**) At percutaneous bile drainage, the protein plug was confirmed by direct cholangiography (arrow)

MRCP is suitable for the abovementioned purposes. Furthermore, a major advantage of MRCP is that it is less invasive and involves no irradiation, unlike CT and endoscopic retrograde cholangiopancreatography (ERCP). MRCP is also superior to ERCP in the depiction of the overall biliary tract including the intrahepatic and extrahepatic bile ducts. Although it is a noninvasive test, which is particularly useful for pediatric patients, the visualization of PBM is often difficult for infants and patients with a short common channel.

The rate of MRCP accurately detecting CBD is reported to be 38–100%. In addition, the diagnostic criteria of MRCP for PBM are equivalent to those with ERCP; however, the definitive detection rate thereof is reported to be 60–100%. The detection rates of PBM for adults and children are reported to be 82–100% and 40–80%, respectively. In cases where the common channel is ≥15 mm, the detection rate is reported to be 82%. The incomplete detection of PBM is often due to the overlap of the dilated bile duct and PBM. As MRCP does not possess as high a spatial resolution as X-ray examinations, it is unclear how precisely it depicts complicated junctions. Therefore, in cases with short or complicated junction, a definitive diagnosis of PBM using direct cholangiography, such as ERCP or intraoperative cholangiography, is required [5].

14.3 Practical Consideration of MRCP

In practical use, MRCP requires some consideration. It is still challenging to perform high-quality MRCP in children, especially very young children, due to these patients' small-caliber ducts, poor signal, and unavoidable patient motion, which creates artifacts.

The need for deep sedation because of the long sequence time is another major drawback of pediatric MRCP. In our institute, infants are sedated with 30–50 mg/kg of body weight oral chlorate hydrate, and children over 1 year of age who cannot tolerate the examination are administered 30–50 mg/kg thiopental sodium rectally.

Although MRCP has been shown to be almost 100% accurate in the evaluation of a choledochal cysts, the visualization rate of PBM ranges from 40% to 83% [6]. It is particularly difficult to visualize PBM in children under 2 years of age and in those with a large choledochal cyst overlapping PBM.

In our institute, routine MRCP imaging is preoperatively performed using the Intera 1.5 T (Philips, Best, Netherlands) with a body array wraparound coil without breath-holding. Patients are studied in the supine position with a thick-slab 2D turbo spin echo (TSE), obtaining coronal and oblique coronal 40 mm thick slices on a 320 × 256 matrix. These image sections are then processed by the standard maximum intensity projection (MIP) algorithm to obtain views of the entire pancreaticobiliary system.

In our series, MRCP was able to demonstrate the extrahepatic bile duct clearly in all patients. The gallbladder was visualized in 92.6%, and the main pancreatic duct was visualized in 81.5%. However, MRCP was able to visualize PBM in only 44.4%

of cases, and the minimum age for successful visualization of PBM with MRCP was 1 year and 11 months. This means that we obtained a diagnostic accuracy of almost 100% in the presence of a choledochal cyst, but the accurate diagnosis rate of PBM was under 50% in the MRCP study. Therefore, routine direct cholangiography is still mandatory, especially in non-dilatation-type PBM [7].

However, MRCP is being used increasingly frequently and has become a viable alternative to ERCP for diagnostic purposes. Furthermore, MRCP can visualize the other surrounding organs outside the pancreaticobiliary luminal structure, including hepatosplenomegaly, hepatic tumors, pancreatic masses, intestinal disease, cystic kidney, and so on. Several recent technological advances have resulted in improvements in coil technology, an increased speed of acquisition, and refinements in respiratory compensation techniques. Therefore, continuous improvements in the image quality are expected to lead to greater diagnostic accuracy of MRCP in the near future.

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