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# A pediatric case of pancreaticobiliary maljunction demonstrated by endoscopic ultrasonography



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## ABSTRACT

Pancreaticobiliary maljunction (PBM) is a congenital anomaly defined as a junction of the pancreatic and bile ducts located outside the duodenal wall, usually forming a markedly long common channel. Diagnosis of PBM in children is usually made by magnetic resonance cholangiopancreatography (MRCP) or endoscopic retrograde cholangiopancreatography (ERCP). Endoscopic ultrasonography (EUS) is a high-resolution imaging modality and a well-established diagnostic tool in adults with pancreaticobiliary (PB) disease. There are several articles which report the value of EUS in the diagnosis and evaluation of PBM in adults, but there are no reports of PBM demonstrated by EUS in children. Here, we present a case of PBM in a pediatric patient that was clearly demonstrated by EUS. EUS can be used as a second line examination modality for demonstrating PBM in pediatric patients in whom first line examinations, such as MRCP or ERCP, cannot be carried out or have not been conclusive.

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Pancreaticobiliary maljunction (PBM) is a congenital anomaly defined as a junction of the pancreatic and bile ducts located outside the duodenal wall. Diagnosis of PBM is usually made by magnetic resonance cholangiopancreatography (MRCP) or endoscopic retrograde cholangiopancreatography (ERCP) in children. Endoscopic ultrasonography (EUS) is a well-established diagnostic tool in adults with pancreaticobiliary (PB) disease. There are several articles which report the value of EUS in the diagnosis and evaluation of PBM in adults, but there are no reports of PBM demonstrated by EUS in children. We present a case of PBM in a pediatric patient that was clearly demonstrated by EUS.

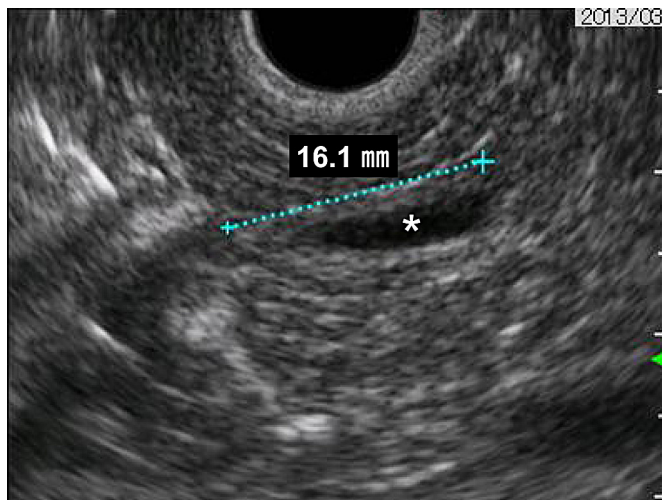
## 1. Case report

A 13-year-old Japanese girl was first diagnosed with acute pancreatitis at the age of 4 years. She initially presented with epigastric pain with serum pancreatic enzyme elevations (amylase 356 IU/L, trypsin 1987 ng/mL, lipase 480 U/dL). After initial hospitalization and recovery of symptoms, she was followed as an out-patient. Although PBM had been suspected as the etiology of the

pancreatitis, definitive diagnosis had not been made. Follow-up review of systems revealed no complaints of abdominal pain, back pain, or fever. Her family history was unremarkable. On follow-up examination, vital signs were normal. Height was 149 cm and weight was 65 kg. She was obese with a body mass index of 29. Her abdomen was soft and there was no tenderness. The liver and spleen were not palpable. Laboratory studies were all within normal range. Conventional abdominal ultrasonography showed dilatation of the common bile duct (CBD) to 5.9 mm in diameter. MRCP was not able to demonstrate a connection between the CBD and the pancreatic duct. ERCP was not carried out due to the potential risk of complications in obese patients. EUS was then carried out in a further attempt to visualize a connection between the CBD and the pancreatic duct. EUS was performed with an echoendoscope (GF-UE260, GF-UCT260, Olympus, Tokyo, Japan). The procedure was done with the patient in the left-sided position under intravenous sedation with midazolam and ketamine. Continuous cardiac and capillary oxygen saturation monitoring were carried out during the procedure and the postprocedural period until full recovery was achieved. EUS clearly demonstrated the PB junction to be located outside the duodenal wall (Fig. 1). The common channel measured 16 mm in length (Fig. 2). The common bile duct measured 4.7 mm in diameter. The gallbladder wall was thickened and there were hyperechoic polypoid structures. PBM was highly suspected. No complications related to the EUS occurred. We concluded that further confirmation of PBM with ERCP was not indicated because of the potential for complications.

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**Fig. 1.** EUS showed a long (16 mm) common channel (asterisk), with the PB junction situated outside the duodenal wall. (For interpretation of the references to color in this figure legend, the reader is referred to the web version of this article.)

## 2. Discussion

In PBM, EUS can demonstrate the confluence of the pancreatic duct and the bile duct outside the duodenal wall. It is also useful for diagnosing other associated biliary diseases such as malignancies. Sugiyama et al. prospectively assessed the diagnostic value of EUS for PBM and compared it with ERCP in 188 adult patients with PB diseases. On ERCP, the length of the common channel measured  $23.4 \pm 5.4$  mm in 25 patients with PBM and  $4.1 \pm 2.0$  mm in 163 patients with normal PB junction. EUS demonstrated the length of the common channel in 25 patients with PBM to be  $\geq 12$  mm in 88%. In contrast, in patients with a normal junction, the channel was  $<6$  mm on EUS. Sugiyama et al. also noted that EUS was valuable for detecting PB diseases associated with PBM such as PB carcinoma, gallbladder lesions, and congenital choledochal cysts. They concluded that EUS plays an adjunctive role in the evaluation of patients with PBM [1]. ERCP depicts PBM more reliably than EUS but is more invasive with a greater potential risk of morbidity. The rate of complications secondary to ERCP is 2.5%–10% in children [2–4]. Cotton et al. reported that obesity is one of the risk factors for severe



**Fig. 2.** Black arrowhead common bile duct; white arrowhead main pancreatic duct. (For interpretation of the references to color in this figure legend, the reader is referred to the web version of this article.)

or fatal complications with ERCP [5]. We felt that ERCP was not indicated in our patient because of its potential for complications and because ERCP would likely not provide more information than had already been obtained by EUS.

There are six reports regarding the use of EUS in gastrointestinal disease in children [6–11]. These reports demonstrate that EUS exams can be carried out without complications, even in small children. These authors concluded that EUS is a safe and useful examination modality in children. According to these reports, EUS was used mainly for the evaluation of pancreatic diseases (e.g., recurrent pancreatitis and cysts/tumors of the pancreas) or submucosal lesions of the gastrointestinal tract. There were no cases of PBM in those reports. To our knowledge there are no previously published case reports of the use of EUS in diagnosing PBM in children.

## 3. Conclusion

We suggest that EUS can be used as a second line examination modality for demonstrating PBM in pediatric patients in whom first line examinations, such as MRCP or ERCP, have not been conclusive or carry high risk.

## Consent

Written informed consent was obtained from the patient for publication of this case report and accompanying images. A copy of the written consent is available for review by the Editor-in-Chief of this journal on request.

## Conflict of interest

On behalf of all authors, the corresponding author states that there are no conflicts of interest.

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