

CASE REPORT

Recurrent Pancreatitis in a Preschool Child

Biggs Saravanan Ramachandran, Shana Baby

Department of Gastroenterology and Interventional Endoscopy, Bharath Hospital, Kerala, India

ABSTRACT

Anomalous pancreatobiliary union in association with choledochal cyst can cause recurrent pancreatitis. Anomalous pancreatobiliary union is a rare condition that can cause a diagnostic challenge and some types can cause recurrent acute pancreatitis. Here we describe a case of re-current pancreatitis with choledochal cyst and choledocholithiasis referred to our hospital for endoscopic retrograde cholangiopancreatography which lead to the incidental finding of anomalous pancreatobiliary union type IA and choledocholithiasis.

INTRODUCTION

Anomalous pancreatobiliary union in association with choledochal cyst can cause recurrent pancreatitis. APBU is a rare condition that can cause a diagnostic challenge and some types can cause recurrent acute pancreatitis.

CASE REPORT

A five-year-old girl was referred to our hospital for ERCP after second episode of pancreatitis in two months period. Patient was evaluated at the referring hospital with USG abdomen, serum amylase and lipase. USG showed bulky pancreas with dilated CBD 12 mm. Serum amylase was 1560 U and Serum lipase was 6800 U. Her Liver function test reports showed total bilirubin of 1.8 mg/dL, indirect bilirubin was 0.4 mg/dL, alkaline phosphatase was 412 IU/L and aspartate amino transferase and alanine amino-transferase were 38 and 64 respectively. Patient was evaluated with MRCP which again showed a dilated CBD with calculi suggestive of choledochal cyst Type IV A (**Figure 1**). Patient was taken up for ERCP under general anaesthesia. On ERCP, CBD showed choledochal cyst with intrahepatic dilatation as well, suggestive of choledochal cyst type IVA. Interestingly patient had abnormal pancreaticobiliary union with long common channel, Komi type IA (**Figure 2**). Small sphincterotomy

balloon sweeping and stone removals were done. Then 5F 5 cm straight biliary stenting was done (**Figure 3ab**). Patient recovered uneventfully after the ERCP and she was referred for pediatric surgical management.

DISCUSSION

Choledochal cysts are rare congenital anomalies which are principally diagnosed by dis-proportional dilatation of extrahepatic bile ducts, after excluding possibility of tumor, stone or inflammation as these can cause the dilatation. Choledochal cysts are believed to arise from the anomalous union of the common bile duct (CBD) and pancreatic duct (PD) outside the duodenal wall which is also proximal to the sphincter of Oddi. The multidetector CT with reformatted imaging, MRCP, and ERCP represent the important techniques providing the anatomic resolution [1].

The classification system for choledochal cyst was devised by Todani *et al.* [2] which is based on the cholangiographic morphology, location and number of intrahepatic and extrahepatic bile duct cysts (**Figure 4**). Type I cysts are confined to the extrahepatic bile duct (EHD) and can be further subdivided into Type I A. Alternatively, a type I cyst involving the entire EHD can further subdivided into IB which involves only a focal segment of the EHD, and IC involving only the CBD. Type II cysts are true diverticula of the EHD. Type III cysts are also referred to as choledochocoele and are confined to the EHD within the duodenal wall. Type IV cysts have multiple features which include both an extrahepatic and intrahepatic component. Type IV cysts can be further subdivided into Type IV A involving both EHD and intrahepatic bile duct (IHD), as well as type IV B involving multiple segmental dilatations of EHD. Finally, Type V (Caroli's disease) cysts are confined to the IHD.

The incidence is higher in Asian people and it is more common in females (male: female – 1:4) [3]. Though the diagnosis of the choledochal cyst is most often made

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Abbreviations APBU Anomalous pancreatobiliary union; ERCP Endoscopic retrograde cholangiopancreatography
Correspondence Biggs Saravanan Ramachandran
Department Of Gastroenterology and Interventional Endoscopy
Bharath Hospital
Azad Lane, Kottayam
Kerala, India
Tel +91 8281527251
Fax +91 8281527251
E-mail rbiggs21@yahoo.com

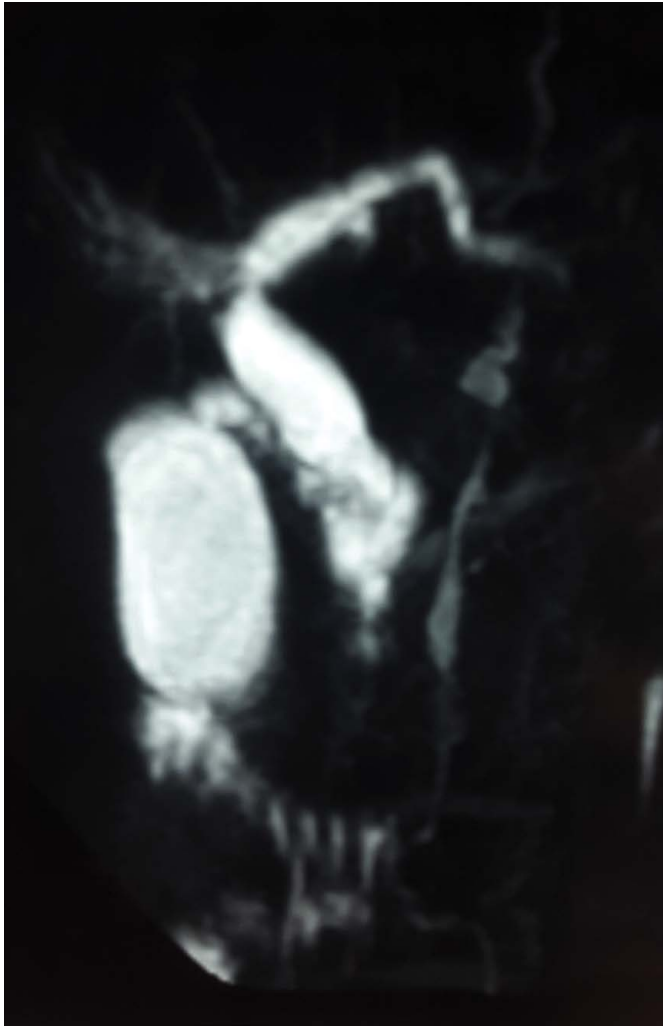


Figure 1. This MRCP picture shows dilated CBD with filling defects with segmental IHBR dilatations suggestive of choledochal cyst type IV.

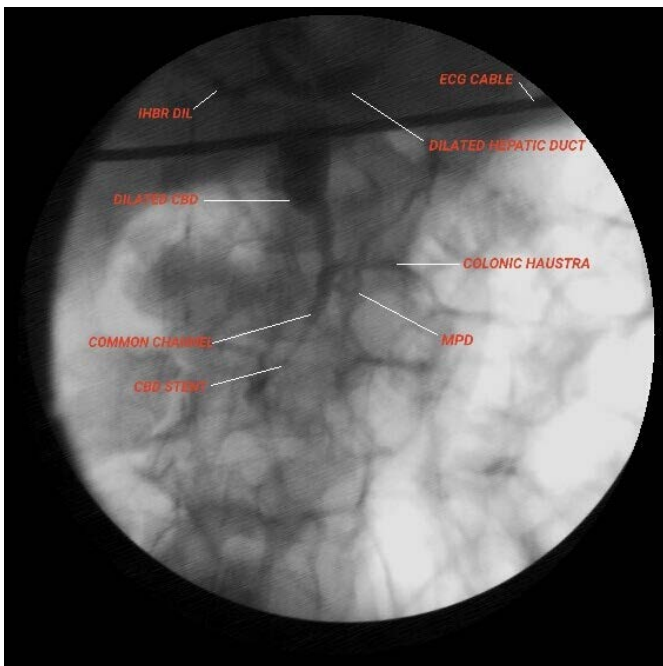


Figure 2. This fluoroscopic picture shows dilated CBD, segmental IHBR dilatation and long common channel with small biliary stent in it. The union of CBD and PD are at right angles and without dilatation (Komi type IA). There are few artifacts like ECG cable and colonic haustral shadow in the picture provided.

during the childhood, 25% of patients are usually seen as adults. Developmentally CBD and PD normally unite in, sphincter of Oddi to form common channel. Normal length of common channel is 0.2-1.0 cm [1].

The maximum normal length of the common channel in neonates and infants less than one year is 3 mm. It increases with age to a maximum of 5 mm in children and adolescents between 13 and 15 years of age [4]. A common channel longer than 4mm is considered abnormal, in adults it is considered abnormal if longer than 6 mm [5]. In our case, length of common channel was 8mm. It is postulated that the anomalous union of the CBD and PD outside the duodenal wall and proximal to the sphincter of Oddi is responsible for the choledochal cyst formation [6]. Reported frequency of APBU on ERCP in series varies from 1.5-8.7% [7].

Komi *et al.* classified APBU [8] into 3 types according to the angle of this ductal union (**Figure 5**). Type I union are defined by right angle between the ducts along with sub classification type IA and type IB based on the association without dilatation (IA) or with dilatation (IB) of the common channel, type II unions are defined by acute angle between ducts with sub classification type IIA and IIB. Type III unions are defined by more complex patterns of accessory pancreatic ducts with or without intricate duct networks. The sub classification includes type IIIA which is equivalent to the classic pancreas divisum with biliary dilatation, Type IIIB is characterised by absence of Wirsung duct. Type IIC 1 contains tiny communicating ducts between the main duct and accessory ducts. Type IIC 2 is characterised by common channel made up of common and accessory ducts of equal calibre finally type IIC 3 is characterised by total or partial dilatation of ductal system.

Only Type IB, IIB and IIC3 are associated with recurrent pancreatitis and are treated surgically. ERCP has been regarded as the gold standard for the diagnosis of choledochal cysts and other associated anomalies, and is superior to MRCP in evaluation of minor ductal anomalies [9]. MRCP is a useful noninvasive tool which shows good overall accuracy in the detection and classification of choledochal cyst [9]. In our case, APBU was missed in MRCP. ERCP guided interventions in patients with complicated choledochal cysts combined with APBU have been reported to be helpful in optimizing the patient condition prior to definitive surgery [10].

The predisposing factors causing recurrent acute pancreatitis in this case were APBU, Choledochal cyst and Choledocholithiasis.

CONCLUSION

Anomalous pancreatobiliary union is a rare developmental abnormality of pancreas which can cause recurrent pancreatitis in children with or without other

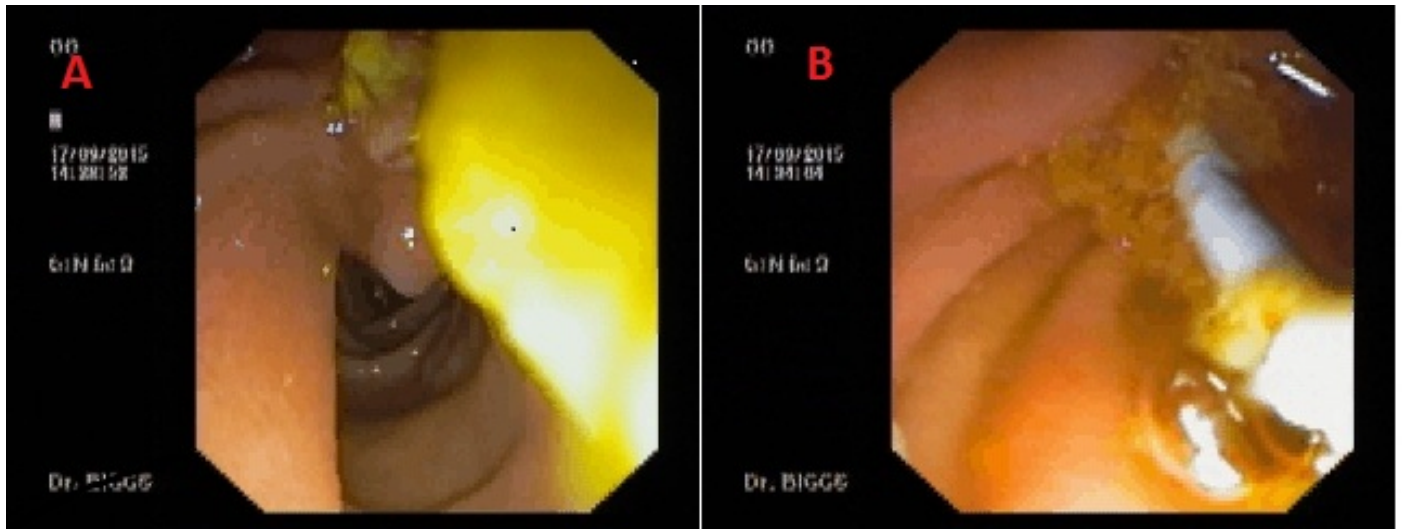


Figure 3. (a) Shows the stone being removed during balloon sweeping of CBD. (b). Shows the ampulla after stent placement.

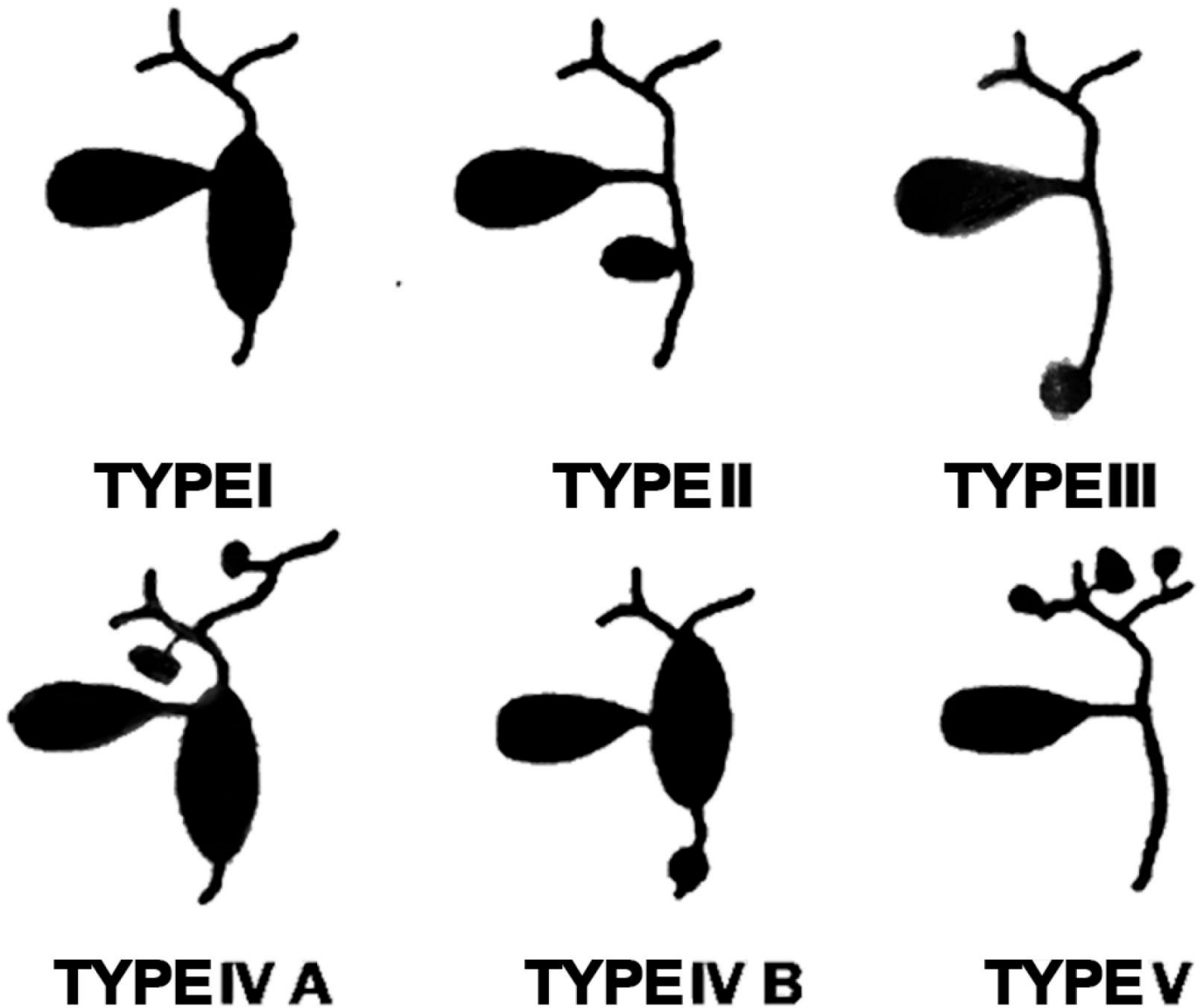


Figure 4. Showing the Todani classification.

KOMI CLASSIFICATION

TYPES		DESCRIPTION
IA	<p>Type I</p>	Have single papilla & The Common Hepatic and pancreatic ducts join each other at a right angle with a non dilated common channel.
IB		Have single papilla & The Common Hepatic and pancreatic ducts join each other at a right angle with a dilated common channel.
IIA	<p>Type II</p>	Have single papilla & The Common Hepatic and pancreatic ducts join each other at An acute angle with a non dilated common channel.
IIB		Have single papilla & The Common Hepatic and pancreatic ducts join each other at An acute angle with a dilated common channel.
IIIA	<p>Type III</p>	Have two papilla & are equivalent to the classic pancreas divisum with biliary dilatation.
IIIB		Have two papilla & are characterized by the absence of the Wirsung's duct.
IIIC1		Have two papilla & contain a tiny communicating duct between the main duct and the accessory ducts.
IIIC2		Have two papilla with & characterized by a common channel made up of common and accessory ducts of equal caliber.
IIIC3		Have two papilla with intricate network of dilated ducts that join each other by total or partial dilatation of the ductal system.

Figure 5. Showing the Komi classification of various types of APBU.

structural abnormalities. ERCP is the gold standard for diagnosis and optimising the patient's condition before being taken up for definite surgical management.

Conflict of Interest

The authors declare that there is no conflict of interests regarding the publication of this paper.

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