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# An unique clinical presentation of choledochal cyst in middle aged female

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**Abstract**
Choledochal cyst are congenital malformations characterized by bile duct dilatation with Intra and/or extrahepatic localization. About 80%of cases are diagnosed in childhood, so their presentation in adults is rare and repeatedly associated with complications. we report a case of 41 year old female with atypical presentation of choledochal cyst of type 1b diagnosed after Ultrasonography, CT scan and MRCP imaging & managed successfully by open radical choledochal cyst excision with hepaticojejunostomy and jejunojejunostomy. Delay in the diagnosis increases the frequency of associated biliary pathology, malignant alternation and suboptimal surgical therapy.

**Keywords:** Choledochal cyst, todani's classification, hepaticojejunal anastomosis, APBDJ: abnormal pancreatico bile duct junction, Alonso LEJ classification

## Introduction
Bile duct cysts are congenital malformations characterized by biliary Duct dilatation with an intra and/or extrahepatic localization. There are Incidences of 1:100000 to 150000 live births in the West and 1:1.000 in Asia, being most prevailing in female at a ration from 3 to 4:1. Around 80% of cases are diagnosed in childhood, thence, the presentation in adults is rare and it is repeatedly associated with complications such as Cholangitis, stone formation, cyst rupture, secondary biliary cirrhosis, Obstructive jaundice and malignancy (cholangiocarcinoma) [3, 5].
The typical clinical presentation features abdominal pain, jaundice and Palpable mass in the right upper quadrant. However, in adults the Presentation can be difficult with nonspecific abdominal pain, under Consideration the classic triad observed in about 25% of cases for this Group [3].
USG confirms the presence of an abnormal cyst and magnetic Resonance imaging (MRI)/MRCP will reveal the anatomy, in particular the relationship between the lower end of the bile duct and the Pancreatic duct [1].
Radical excision of the cyst is the treatment of choice, with Reconstruction of the biliary tract using a Roux-en-Y loop of jejunum. Complete resection is important because of an association with the Later development of cholangiocarcinoma [1].

## Case Report
A 41year old Female Presented with A History Of Upper Abdominal Pain Radiating To Back And Vomiting Containing Food Particles Since 15 Days.
She doesn't Have Complain of Anorexia, Jaundice and Weight Loss, No History of Similar Complain Previously.
She doesn't have any complain of abdominal pain, abdominal lump or Jaundice in childhood.
On Physical Examination vitals within normal range, No Icterus Found, On Per Abdomen Examination Tenderness Present in Right Hypochondriac and Epigastric Region, However, No Mass Was Palpable.
In Laboratory Investigation billirubin level Was 0.6 mg/dl and Liver Function Test was within normal limit.

## Imaging
USG Abdomen Suggestive of Dilated CBD with One 63.7 X 43.1 Mm Cyst Seen in

Proximal CBD upto Pancreatic Duct S/O Choledochal Cyst. CECT Abdomen Suggestive of Well Defined approx 47 X 46 X 53 Mm Size Cystic Lesion Near Porta Abuting Cystic Duct, Possibilities of Choledochal Cyst/ Pancreatic Pseudocyst/ Hydatid Cyst.

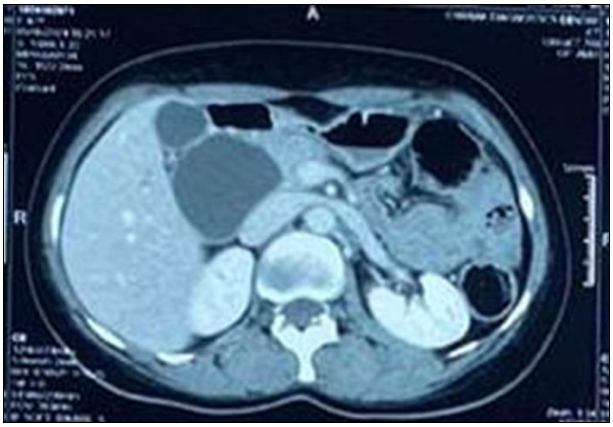

**Image 1:** CECT image showing choledochal cyst

MRCP Suggestive of Approx. 47 X 54 X 40 Mm Size Well Defined Fusiform Dilatation of Proximal CBD – Suggestive of Choledochal Cyst [Type Ib] Likely, Distal CBD Normal and Anomalous Pancreaticobilliary Junction, CHD Appears Dilated with Maximum Diameter 11 Mm, With High Junction of Pancreatic Duct with The Common Bile Duct Resulting in Long Common Channel Measuring 15 Mm.
Out of all three radiological investigation MRCP is the best diagnostic Tool for choledochal cyst, as observed in our case.

**Image 2:** MRCP image showing choledochal cyst

After required preoperative work up and preoperative preaparation Exploration done through right subcostal [kocher incision], Intraoperatively approx. 5 x 4 cm size choledochal cyst type 1b found Involving supraduodenal and retroduodenal part of CBD, cystic duct And CHD directly open into cyst.
Cholecystectomy with cyst excision was done taking utmost care to Safeguard important viral structures adjacent to cyst like Inferior vena Cava; portal vein & Duodenum; there was high junction of main Pancreatic duct with common bile duct so care was taken not to Damage Main pancreatic duct & head of pancreas, To restore the Continuity end to side hepaticojejunostomy and side to side Jejunojejunostomy was done. Postoperative course was uneventfulWithout any complications. Drain was removed on 7th post operative Day after less than 25 cc serous drain output for three consecutive days Patient discharged on 12th day after surgery.

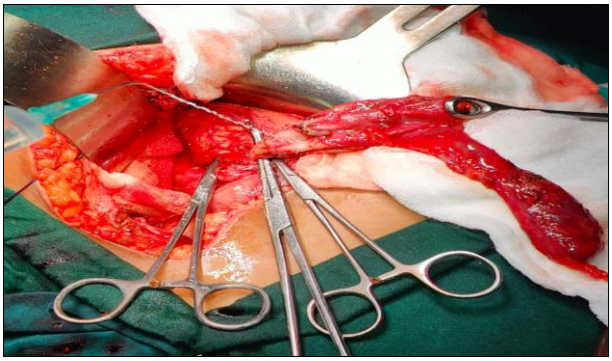

**Image 3:** Intraoperative image of choledochal cyst with gallbladder and cystic duct

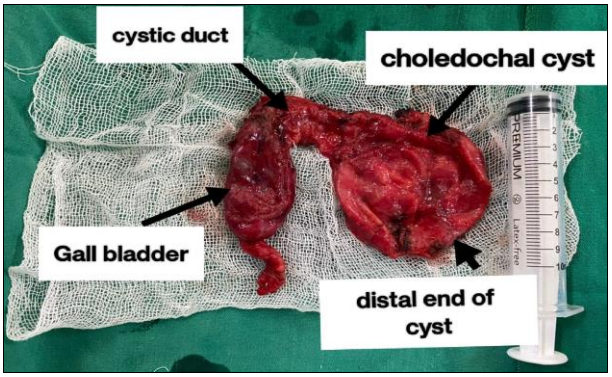


**Image 4:** Specimen of choledochal cyst after surgical resection

**Discussion**
Congenital biliary dilatation may be found at any age, but more than two-thirds of cases are diagnosed in children younger than 10 years of age, and some cases are diagnosed prenatally by ultrasound examinations as early as the 15th week of gestation. But in our case patient was 41 year old middle aged female [2].
Alonso-Lej classified extrahepatic bile duct cystic dilatation into three types. Type I is congenital cystic dilatation of the common bile duct; type II is congenital diverticulum of the common bile duct; and type III is choledochocele. Todani refined the classification of bile duct cysticdisorders into five types and included the concept of pancreaticobiliary maljunction. Type IV-A is congenital biliary dilatation associated with intrahepatic duct dilatation, and type V is multiple intrahepatic bile duct dilatations. The major clinical symptoms are recurrent abdominal pain, nausea and vomiting, mild jaundice, an abdominal mass, and fever, But in our patient only abdominal pain & vomiting was there no any palpable abdominal lump, jaundice. As upper abdominal pain & vomiting are feature of so many other surgical condition it was difficult to diagnose choledochal cyst clinically in our patient. Diagnosis of congenital biliary dilatation requires demonstration of abnormal dilatation of the bile duct and pancreaticobiliary maljunction on the basis of diagnostic imaging or anatomic abnormalities. USG is primary non invasive investigation of choice for diagnosis, but MRCP is best imaging modality for diagnosis of choledochal cyst, to delineate duct anatomy & better plan of surgery as observed in our case. Main complications are biliary stones, pancreatitis, spontaneous perforation of the

bile duct and carcinoma. Accordingly, the definitive treatment of congenital biliary dilatation is complete excision of the extrahepatic bile duct with Roux-en-Y hepaticojejunostomy. Type II diverticulum, type III choledochocele, and type V are rare, and pancreaticobiliary maljunction is not usually associated. Caroli disease is usually autosomal recessive. The clinical onset of Caroli disease usually occurs during childhood, and symptoms include cholangitis, portal hypertension, and abdominal pain. The long-term prognosis for such patients is poor, with a marked predisposition to septicemia, liver abscess, resultant hepatic failure, portal hypertension, or cholangiocarcinoma [2].

## Conclusion

Choledochal cyst is a rare congenital abnormality in adults. Emphasis should be placed on thorough preoperative assessment of cyst anatomy and the necessary limits of resection. This to a large extent is achieved by imaging methods like MRCP and ERCP preoperatively and by on-table cholangiography and endoscopic ultrasound intraoperatively. Abnormal APBDJ is likely to be seen in the paediatric patients more than in adults but in our case it was there in middle aged female also. Abdominal mass and jaundice are common presentation in children, while the adult patients are more likely to present with complications including cholangitis, choledocholithiasis, pancreatitis, and malignant transformation. Complete cyst excision is the treatment of choice for extrahepatic component of the disease, although the optimal treatment of intrahepatic bile duct dilatations remains controversial, especially for type IV-A. Due to the age related risk of synchronous and metachronous cholangiocarcinoma, complete cyst excision should be carried out early. Long term follow-up is required in these patients as they are prone to cholangitis, anastomotic stricture, and malignancy in the residual biliary tree [2, 3, 4].

## Conflict of Interest

Not available.

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