

## Case report

### **INTRAHEPATIC GALL BLADDER WITH CHOLEDOCHAL CYST WITH MALROTATION OF GUT – A RARE ASSOCIATION**

**Abstract:** The Choledochal Cysts are rare cystic dilatory conditions of biliary tree with malignant tendency. These are more frequently reported in children. Surgical removal of these cysts can significantly reduce the risk of malignancy and also reduce the associated complications [1]. However, Choledochal Cysts has been paid lesser attention in adults. Choledochal Cysts associated with Intrahepatic gall bladder and Malrotation of gut has rarely been reported. We herein report a case of 15 years female patient with Type 4a Choledochal cyst associated with intrahepatic gall bladder and mal-rotation of intestine which was incidentally found at surgery. The patient underwent Excision of Choledochal cyst with Cholecystectomy with Appendicectomy with Hepatico-jejunostomy with Jejunio-jejunostomy, and Ladd's procedure was simultaneously performed for asymptomatic Intestinal Malrotation.

**Keywords:** Choledochal cyst, Intrahepatic gall bladder, Malrotation of gut, Hepatico-jejunostomy, Jejunio-jejunostomy, Ladd's procedure

**Introduction:** Choledochal cyst is a relatively rare bile duct abnormality, which is predominantly characterized by cystic dilatation of the biliary tract. There are five different types of CC as described in the Todani classification system [2], among which type 1 and 4 cysts have been found to be more prevalent in Asian populations [3]. There is high rate of incidence of malignancy in choledochal cysts. The cancer risk increases significantly with age and has been found to be higher in those with type 1 and 4 cysts [3,4]. Intestinal Malrotation(IM) is an important surgical disease and is well known to be incidentally associated with other surgical abdominal diseases [5]. However, choledochal cyst associated with Intrahepatic gallbladder and IM have rarely been reported. We here present a case of 15 years female patient with type 4a choledochal cyst, with incidentally found malrotation of intestine and intrahepatic gallbladder, intraoperatively.

**Case Report:** A 15 years female patient presented with pain in abdomen with on and off episodes of vomitings since 6 months. Pain was aggravated with heavy fatty meals. There was history of similar complaints in the past, and patient was managed conservatively at local hospital. On examination, there was mild tenderness in the right hypochondrium, with no guarding, rigidity or distension. On admission, patient was stable vitally, with normal blood counts, normal liver enzymes and serum electrolytes. Ultrasonography was suggestive of fusiform dilatation of CBD, left and right hepatic ducts. CECT abdomen and MRCP was done which was suggestive of "Fusiform dilatation of entire CBD with normal distal most CBD. Significant dilatation of right and left hepatic ducts with dilatation of intrahepatic biliary channels. Gall bladder well distended with 6mm. sized non obstructive calculi with no changes of cholecystitis. (Fig 1,2,3)

Fig 1: CECT transverse section showing choledochal cyst

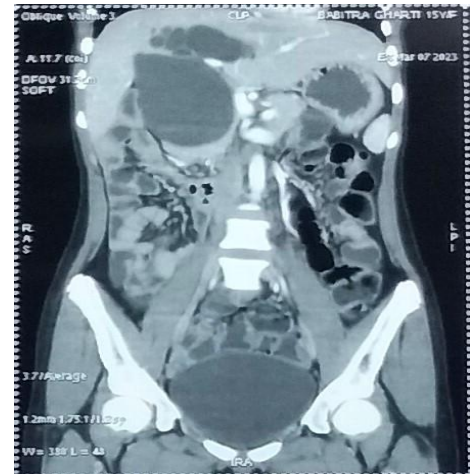
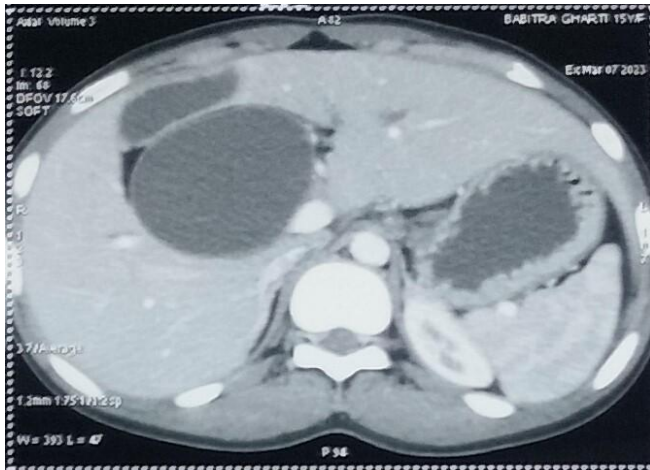


Fig 2: CECT sagittal section showing dilated intrahepatic and extrahepatic bile ducts

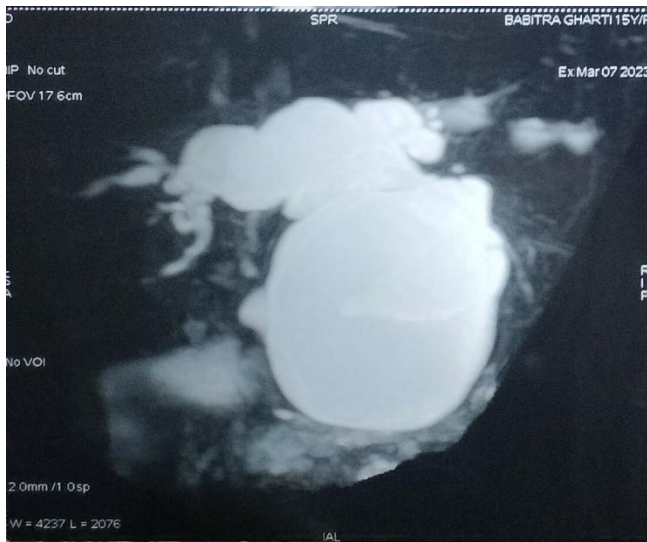


Fig 3: MRCP showing dilated intrahepatic and extrahepatic bile ducts

Patient was planned for OT and with Right subcostal incision "Excision of Choledochal cyst with Cholecystectomy with Appendicectomy with Hepatico-jejunostomy with Jejun-jejunostomy with Ladd's procedure for malrotation of gut was done". Post operative period was uneventful, and Ryle's tube was removed on POD-3, Sips orally was started on POD-7, and patient was discharged with stable vitals on POD-10. On follow-up in OPD after 1month, patient was stable with no any complaints.



Fig 4: Intraoperative picture showing "Intrahepatic gall bladder"

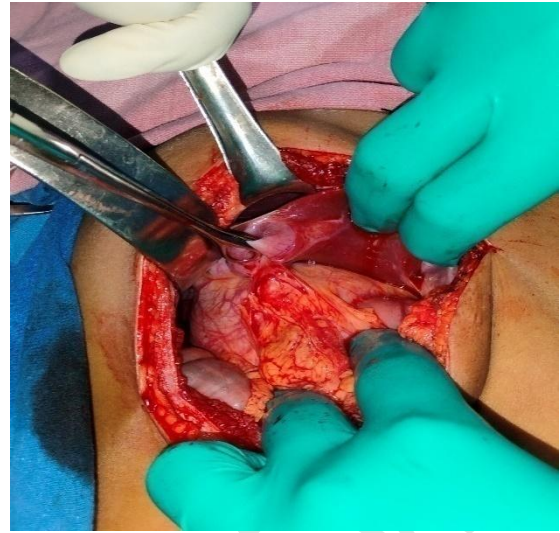


Fig 5: Intraoperative picture showing "Intrahepatic gall bladder"

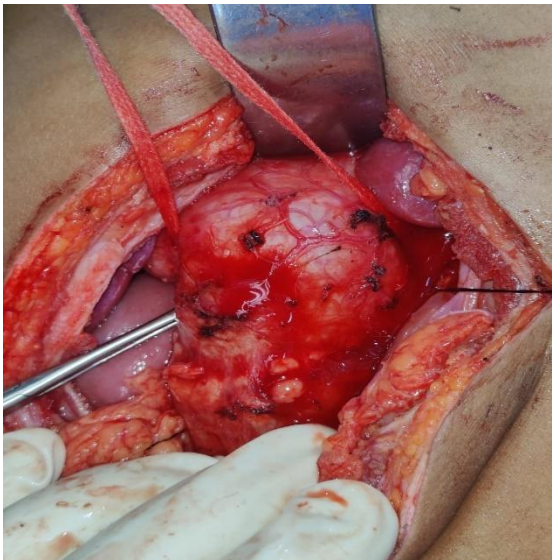


Fig 6: Choledochal cyst exposed

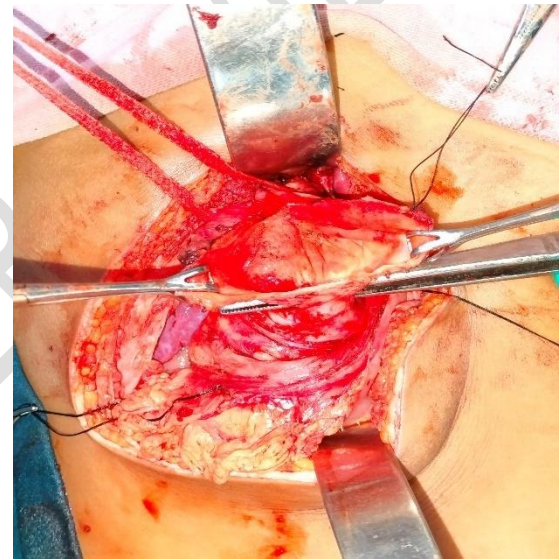


Fig 7: Opening of choledochal cyst

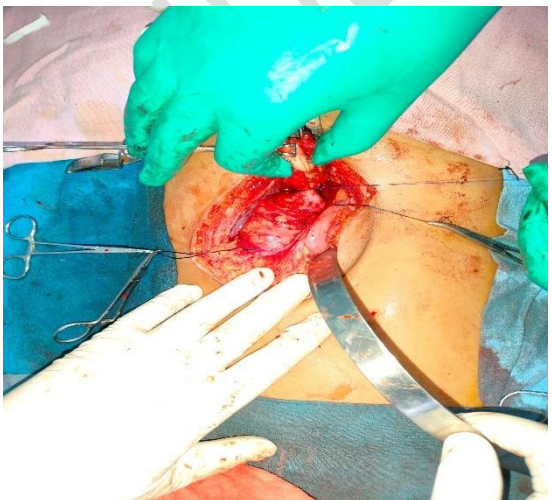


Fig 8: Excision of Choledochal cyst

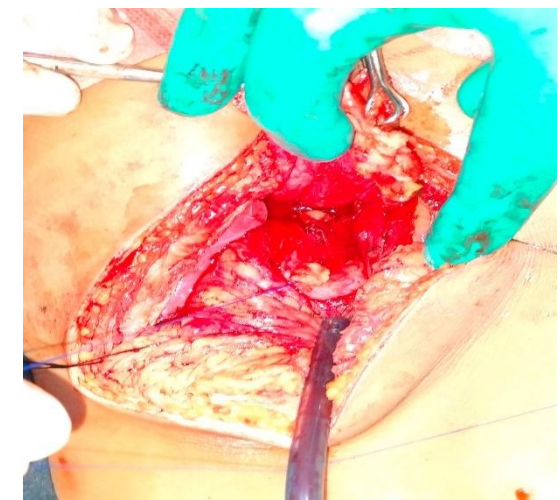


Fig 9: Intraoperative picture after excision of choledochal cyst

**Discussion:** Choledochal cysts are focal or diffuse dilations of the biliary tree. Choledochal cysts can occur as single or multiple cysts throughout the extrahepatic or intrahepatic bile ducts. The cysts can predispose patients to recurrent cholangitis or pancreatitis, choledocholithiasis, secondary biliary cirrhosis, biliary stricture, and malignancy. Approximately 80% of choledochal cysts are diagnosed in children, and 20% of cases present in adults. There are a few case reports of choledochal cysts occurring within families, but generally, they do not have a recognized hereditary pattern.

Todani and colleagues has classified the choledochal cysts into 5 categories based on anatomy. Traditionally, the classic and most common choledochal cyst is type I disease with cystic, saccular, or fusiform dilatation of the extrahepatic biliary tree. Type II cysts are simple diverticula of the common bile duct, which are usually extrahepatic, supraduodenal, and saccular. A type III cyst, also known as *choledochoceles*, is a focal cystic dilatation of the most distal segment of the bile duct. Multiple dilations of the intrahepatic and extrahepatic biliary tree are known as type IV cysts. Type V cyst, Caroli disease, is confined to the entire liver or a solitary lobe, usually on the left.

The most common proposed theory for choledochal cyst formation is related to pancreaticobiliary maljunction. Pancreaticobiliary maljunction is defined as an extramural junction of the pancreatic and biliary ducts in the duodenum beyond the intramural sphincter function and is characterized by a long common channel. Pancreaticobiliary maljunction is also thought to be a significant risk factor for the development of cholangiocarcinoma in the biliary cyst, as well as the development of gallbladder cancer [6].

The association of Intrahepatic gall bladder, Choledochal cyst and Malrotation of gut is very rare, and high index of suspicion is needed while encountering any such congenital conditions, both intraoperatively and while preoperatively evaluating the patients.

#### References:

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