

Original Research Article


# A study of symptomatic choledochal cyst in South India

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## Abstract

**Introduction:** Choledochal cyst is a type congenital anomaly in which there is dilatation of the intra or extrahepatic biliary tree. Choledochal cysts are classified into five types based on location or shape of the cysts.

**Materials and methods:** A prospective analytical study was conducted from January 2015 to June 2017 at Madras Medical College, Chennai, India. All patients who underwent ERCP and had finding of choledochal cyst on cholangiogram were included in the study. Patients were assessed on their demographic data, clinical findings, presentation, and complications.

**Results:** The incidence of symptomatic choledochal cyst was 2.5%. Type 1C was the most common type of choledochal cyst with obstructive jaundice due to large CBD stones as the most common presentation in this study. None had 30 day post-operative mortality.

**Conclusion:** Type 1C is the most common type among symptomatic choledochal cyst in south India. As it is a premalignant condition management includes endoscopic retrograde cholangiography followed by surgery with good clinical outcome.

## Key words

Choledochal cyst, South India, Symptoms, Biliary tree.

## **Introduction**

Choledochal cysts were first described by Vater and Ezler in 1723 as congenital cystic dilation of the intra or extrahepatic biliary tree although it is benign, can be associated with serious complications like cholelithiasis, cholangitis, pancreatitis and malignant transformation [1, 2].

## **Aim and objectives**

- To classify patients with symptomatic choledochal cyst into various types as per Todani's classification.
- To assess outcome of endotherapy on follow up.

## **Materials and methods**

The study was designed as prospective analytical study from January 2015 to June 2017 at Madras Medical College. All patients who underwent ERCP and had finding of choledochal cyst on cholangiogram were included in the study. Patients were assessed on their demographic data, clinical findings, presentation, and complications. Outcome of the endotherapy was assessed on the basis of successful cannulation, cholangiographic assessment, choledocholithiasis, size of stones, presence of periampullary diverticulum, post ERCP complications. All patients were referred hepaticojejunostomy as choledochal cyst is a premalignant condition. Mortality at 30 days after procedure was assessed. All results were tabulated and assessed using SPSS programme.

## **Results**

25(2.5%) patients with findings of choledochal cyst on cholangiogram in 990 ERCP's performed during study period were included. Average age of study population was  $43.2 \pm 18.5$  years and M: F ratio was 1: 4 with female preponderance. 11(44%) patients presented with obstructive jaundice, 9(36%) with recurrent biliary pain, 2(8%) with recurrent acute pancreatitis and 3 (12 %) were asymptomatic. 5(10%) patients had cholangitis at presentation. Type IC was the most common type seen in 16(64%) patients followed

by type IB in 4 (16%) patients. One had Type IA with anomalous pancreaticobiliary malunion. 2(8%) had type III. 1 had type V. Successful CBD cannulation was achieved in 21(84 %) patients following a biliary sphincterotomy in 24 (96%) cases. Choledocholithiasis and hepatolithiasis was found in 13(52%) and 2 (8%) patients respectively. 2(8%) had choledochoduodenal fistula and 3(10%) had periampullary diverticulum. Successful stone retrieval was done in 10(40%) patients. All patients were referred for surgery based on the type of choledochal cyst for further management. None had post-operative 30 day mortality.

## **Discussion**

The incidence of choledochal cyst in western population is 1 in 100 000–150 000 live births, whereas in Asian populations it has a higher incidence of 1 in 1000. More than 60% of cases occur in Japan. Incidence shows a clear female preponderance with M: F of 3:1 [3, 4]. Our study showed a higher incidence of 2.5 % as all patients were symptomatic seeking tertiary medical care. However it showed a male predominance.

Choledochal cysts are usually diagnosed in childhood during antenatal second trimester anomaly screening, or an adulthood diagnosis is also common [5]. It can present with a myriad of presentations including asymptomatic, abdominal pain, obstructive jaundice, mass abdomen, spontaneous perforation, cholangitis, pancreatitis, portal hypertension, and abnormal liver function tests [6]. Classic triad of jaundice, abdominal pain, mass abdomen is found only in 20% of patients [7]. Our study showed 11(44%) patients presented with obstructive jaundice, 9(36%) with recurrent biliary pain, 2(8%) with recurrent acute pancreatitis, 3 (12 %) were asymptomatic and 5(10%) patients had cholangitis at presentation.

The cause of choledochal cyst still remains undefined [8]. Congenital weakness of the bile duct wall may be a primary abnormality of

epithelial proliferation during embryologic ductal development, and congenital obstruction of bile ducts has been hypothesized. Choledochal cysts have been associated with congenital anomalies like colonic atresia, duodenal atresia, imperforate anus, pancreatic arteriovenous malformation, multiseptate gallbladder, ventricular septal defect, aortic hypoplasia, pancreatic divisum [9]. Patients in our study did not have any associated congenital anomalies. Our study focused on adulthood presentations. However all patients became symptomatic only in adulthood.

Choledochal cysts are classified into five types based on Todani's classification. Type-IA is cystic dilation of the extrahepatic duct. Type-IB is focal segmental dilation of the extrahepatic duct. Type-IC is fusiform dilation of the entire extrahepatic bile duct. Type-II is simple extra axial diverticula of the common bile duct. Type-III is called choledochoceles in which distal intramural dilation of the common bile duct within the duodenal wall is dilated. It is a close differential for duodenal duplication cyst. Type-IVA is combined intrahepatic and extrahepatic duct dilation. Type-IVB is multiple extrahepatic bile duct dilations. Type-V is also called as Caroli disease in which multiple intrahepatic bile duct dilation was seen [10]. This type is associated with another ductal plate malformation called congenital hepatic fibrosis and cysts in kidney [9]. According to western literature type I is the most common type occurring in 50%–80% followed by type IV, type V, type III, type II with 15%–35%, 20%, 4.5%, 2% occurrence respectively. In our study, Type IC was the most common type seen in 16(64%) patients followed by type IB in 4 (16%) patients. 2(8%) had type III.

Anomalous pancreatico biliary malunion (APBU) is characterized by a long common channel where insertion of the CBD is farther than 15 mm from the ampulla of Vater. It occurs in less than 2% of the general population. It is more commonly seen in pediatric population with choledochal cyst. 80-96% of pediatric choledochal cysts are associated with APBU

[11]. The overall risk of malignancy (cholangiocarcinoma) has been reported to be 10% to 15% in choledochal cyst. The risk of malignancy increases with age. Presence of APBU is a notable risk factor for malignancy. Only one patient had anomalous pancreaticobiliary malunion which was associated with type IA.

Selective biliary cannulation in the presence of periampullary diverticulum is difficult due to altered anatomy of the distal CBD. More so, many patients have a distal CBD stricture making selective biliary cannulation even more difficult. Cannulation in first attempt was noted in 14 of patients, rest needed biliary sphincterotomy for cannulation. Successful CBD cannulation was achieved in patients following a biliary sphincterotomy in 24 (96%) cases. Following cannulation cholangiogram done with biliary contrast showed filling defects in 13 (52%) of patients. Biliary trawling was attempted after adequate sphincterotomy. Successful stone retrieval was done in 10(40%) patients. Hepatolithiasis was found in 2 (8%) patients. 2(8%) had choledochoduodenal fistula and 3(10%) had periampullary diverticulum. All patients were referred for surgery for further management.

All patients were referred for surgery following ERCP and decompression. Cholangiographic appearance and type of choledochal cyst was compared with intraoperative findings. 95% of cases had concordance. Patients with type I choledochal cyst (20 patients) underwent hepaticojejunostomy or choledochojejunostomy with a Roux-en-Y anastomosis as a drainage procedure. Type III patients were managed with biliary sphincterotomy. One patient with Caroli's disease (Type V) was managed with biliary stenting. Patients with hepatolithiasis managed with segmentectomy and respective drainage procedure. All patients were followed up for 30 days. None had 30 day post-operative mortality.

## **Conclusion**

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Type IC is the most common type among symptomatic choledochal cyst in south India. As it is a premalignant condition management includes endoscopic retrograde cholangiography followed by surgery with good clinical outcome.

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