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A Study Of Clinical Presentation And Management Of Infantile Choledochal Cyst: Case Series

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Abstract

Introduction: Choledochal cysts (CC) are a rare congenital cystic dilation of the biliary tract, first described by Vater and Ezler in 1723. They present primarily in female infants and young children. If not identified and managed early, it may result in significant morbidity and mortality. It has an overall incidence of 1 in 100/150 000 live births in the western population, with female to male ratio of 3-4:1, 75% of which are diagnosed in childhood. The aim of the study was to analyse the clinicopathological features, outcomes of surgical management and challenges faced in the management of infantile choledochal cyst.

Material and methods: All cases of infantile choledochal cyst, admitted and treated in the Department of General Surgery, JNIMS, Imphal during January 2017 to December 2019, were included. Patients were subjected to radiological investigations which are Ultrasonography and MRCP. Clinical presentations, investigations, types, surgical management and outcomes recorded and analysed.

Results: A total of 15 patients studied, with female:male ratio of 4:1. MRCP showed type 1 choledochal cyst in 13 patients and type IV in 2 patients. 13 patients underwent Excision & Roux-en-Y Hepatico-jejunostomy, one had External Drainage of cyst, other one had Excision & Hepatico-duodenostomy. all the survived patients were jaundice free. Stool colour came to normal. Liver parenchyma was reversed to normal.

Conclusion: More cases are being diagnosed in infancy with MRCP providing excellent delineation of the cysts. Early surgery can reverse biliary cirrhosis and portal hypertension. If diagnosed in antenatal or neonatal period surgery should be done within 8 weeks to prevent cirrhosis.

Keywords: choledochal cysts, classification, Roux-en-Y Hepatico-jejunostomy

Introduction

Choledochal cysts (CC) are a rare congenital cystic dilation of the biliary tract, first described by Vater and Ezler in 1723 [1]. Overall incidence of CC is 1 in 100/150 000 live births in the western population, with a 3-4:1 female predominance, 75% of which are diagnosed in childhood [1,2]. Approximately 80% of CC are diagnosed in infants and young children within the first decade of life [2,3]. Alonso-Lej and colleagues[4] proposed the first CC classification in 1959. Komi and associates[5] later proposed a new CC classification according to the type of APBDU(anomalous pancreaticobiliary ductal union) based on 2 unique features: a long common channel and the angle of the junction between the pancreatic duct and distal CBD as they converge on the sphincter of Oddi.[6] However, the most widely accepted classification was reported by Todani and colleagues[7] in 1977, derived from the original Alonso-Lej classification and based on the site of

cystic change. Five types of CC are described and classified as type I (80% to 90% of all CC), type II, type III, type IV (15% to 20% of all CC) and type V or Caroli's disease.

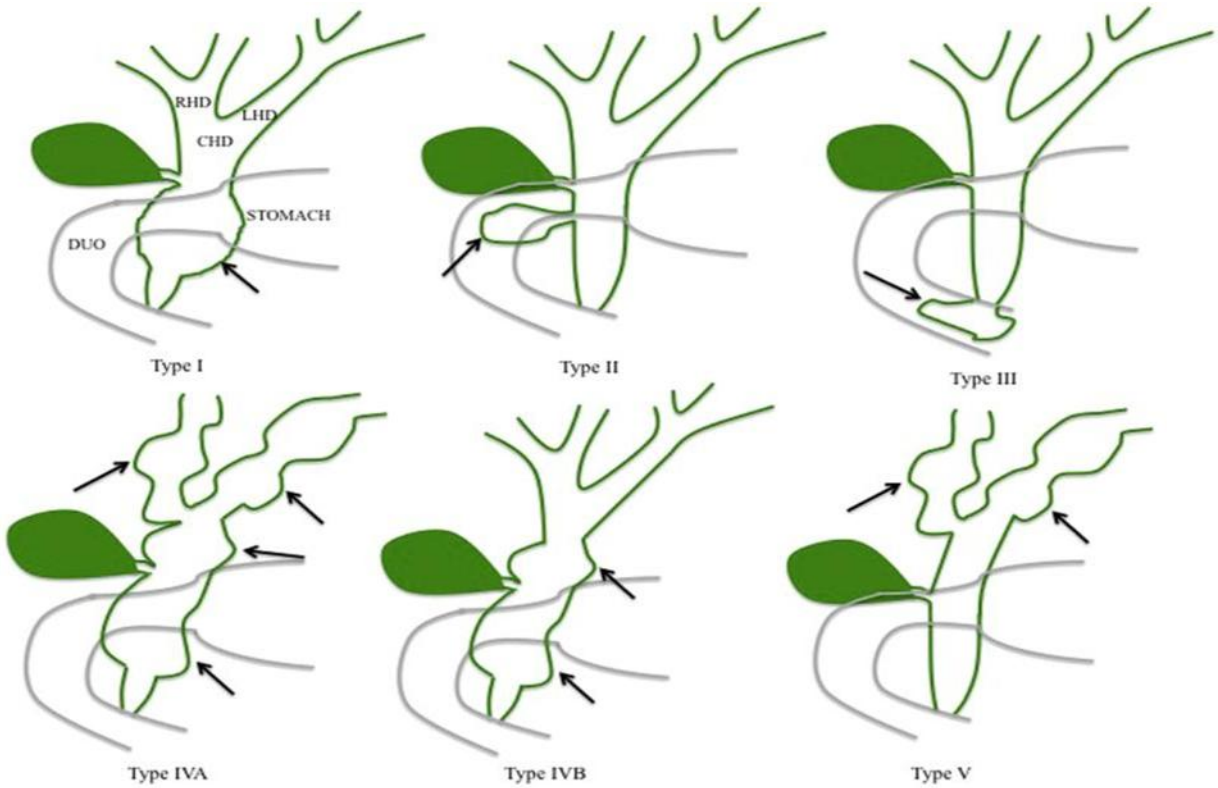

Figure: Types of choledochal cysts (CC). Type I cysts are fusiform dilatations of the common bile duct (CBD). Type II cysts are true diverticula of the CBD and type III CC (choledochoceles) are intraduodenal dilations of the common channel. Type IVA CC consist of multiple intrahepatic and extrahepatic biliary dilatations, while type IVB CC have extra-hepatic biliary dilatation with a normal intrahepatic biliary tree. Type V CC, or Caroli's disease, consist of cystic dilation of the intrahepatic biliary tree, right hepatic duct, left hepatic duct, common hepatic duct, duodenum.

Although diagnosis of Choledochal Cyst (CC) in utero and adult is also common, they are usually diagnosed in childhood. Some of the common presentations include abdominal pain, jaundice and right upper quadrant mass and are most commonly seen in pediatric patients. CC is diagnosed using multimodality imaging including ultrasound, CT, MRI and MRCP. Ultrasound is the most frequently used imaging modality given its low cost and accessibility and has been shown to be reliable and cost effective as single modality imaging in the pediatric population. MRCP technology has removed the need for exaggerated breath holding techniques, increasing its utility and accuracy in pediatric patients. The MRCP is highly sensitive (70% to 100%) and specific (90% to 100%) in CC diagnosis and classification.[8]

If left un-treated Choledochal Cyst (CC) has complications that range from bile duct obstruction to cholangiocarcinoma. The treatment of the CC will depend on the types of CC, accordingly to the Todani's classification. For Type I and IV CC (the commonest of all), the treatment is surgery, although, the reconstruction technique of the bile duct has no gold standard. The surgical approach has evolved from drainage procedures to the excision of the cyst and reconstruction of the bile duct as the treatment of choice. There are two reconstruction techniques, hepaticojejunal anastomosis in Roux-en-Y and hepaticoduodenal anastomosis.[9]

Methods:

All cases of infantile choledochal cyst, admitted and treated in the Department of General Surgery, JNIMS, Imphal during January 2017 to December

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2019, were included in this study. All the patients were subjected to radiological investigations which are Ultrasonography (USG) and MRCP .Liver function tests were done to confirm jaundice. Their clinical presentation, investigations, types, surgical management and outcomes were recorded and analysed.

Results:

A total of 15 patients studied during the time period. There were 12 females and 3 males, female:male ratio of 4:1. MRCP performed in all patients which showed type 1 choledochal cyst in 13 patients and type IV in 2 patients. 13 patients underwent Excision & Roux-en-Y Hepatico-jejunostomy, one had External drainage of cyst, other one had Excision & Hepatico-duodenostomy. At the time of follow up, all the survived patients were jaundice free. Stool colour came to normal. Liver parenchyma was reversed to normal. Splenomegaly was subsided with good catch up growth. Liver enzymes came to normal level

Table 1: Types of choledochal cysts

Type of cyst	No. of cases
Type I	13
Type II	0
Type III	0
Type IV	2
Type V	0

Figure: Type 1 choledochal cyst.

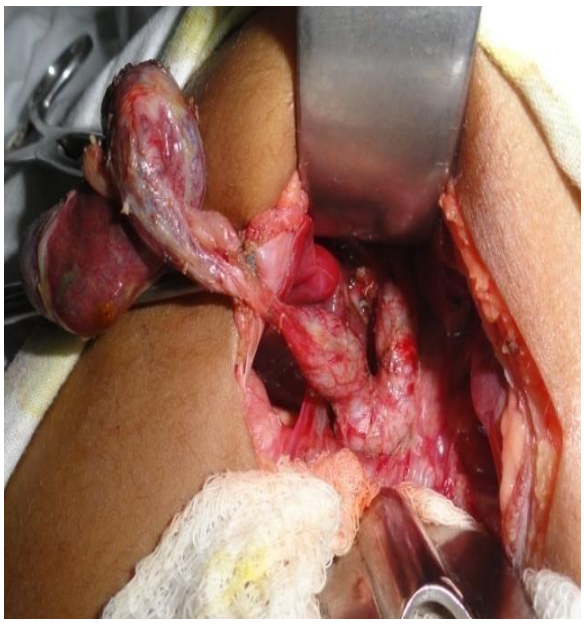

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Figure: Ultrasound finding in a 4 month old child

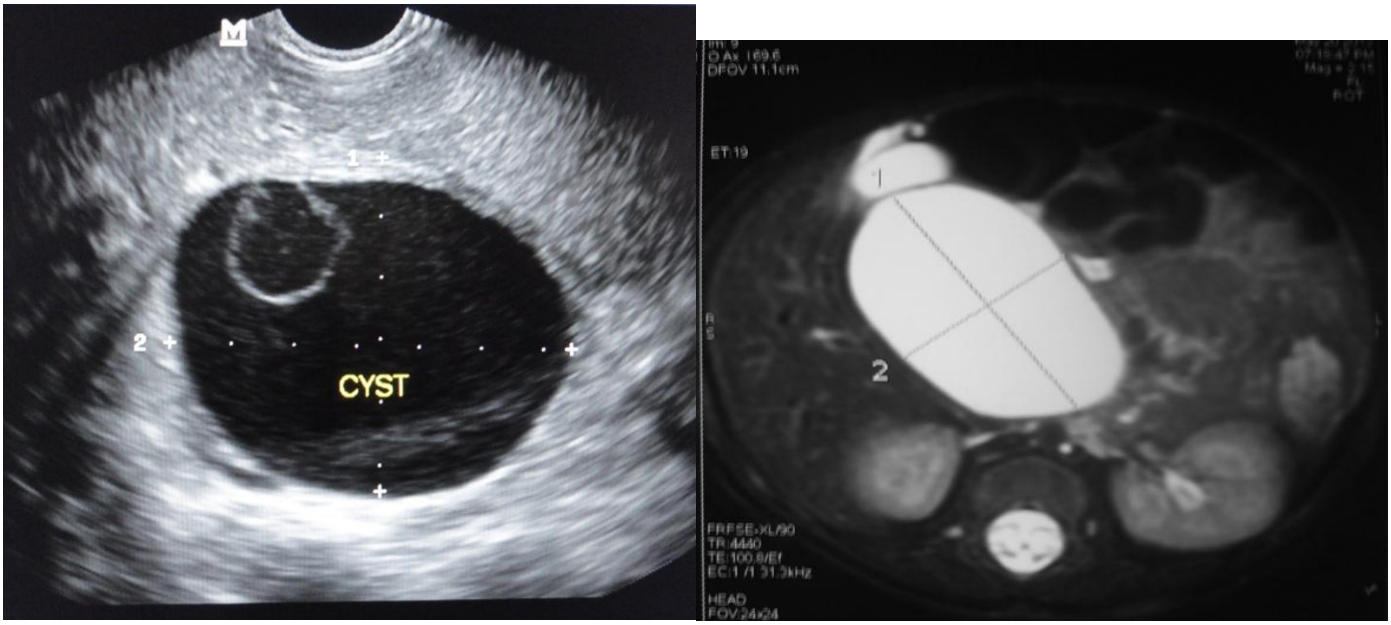

Figure: MRCP of 2 months old female child showing type 1 choledochal cyst

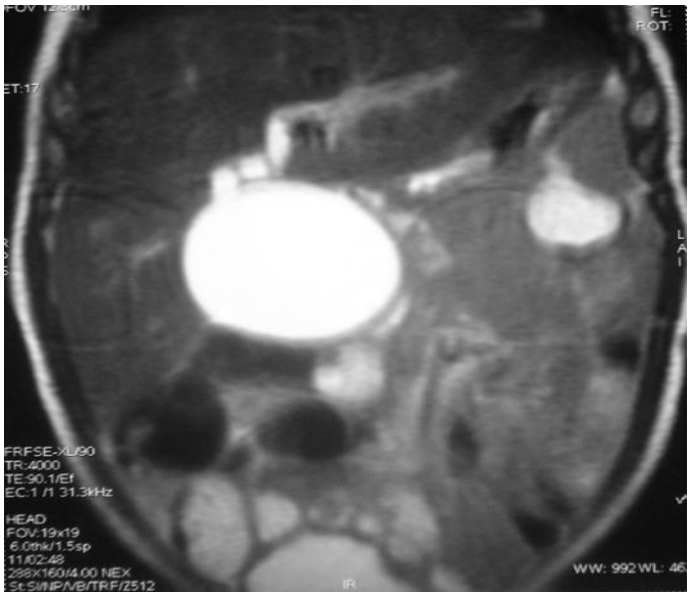

Table 2: Liver parenchyma

Liver parenchyma	No. of patients
Normal	6
Early cirrhosis	5

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Cirrhosis	2
Advanced cirrhosis	2

Table 3: Types of procedure done

Procedure	No. of patients
Excision & Roux-en-Y Hepatico- jejunostomy	**13**
External drainage of cyst	**1**
Excision & hepatico-duodenostomy	**1**

Figure: Hepatico-jejunostomy procedure.

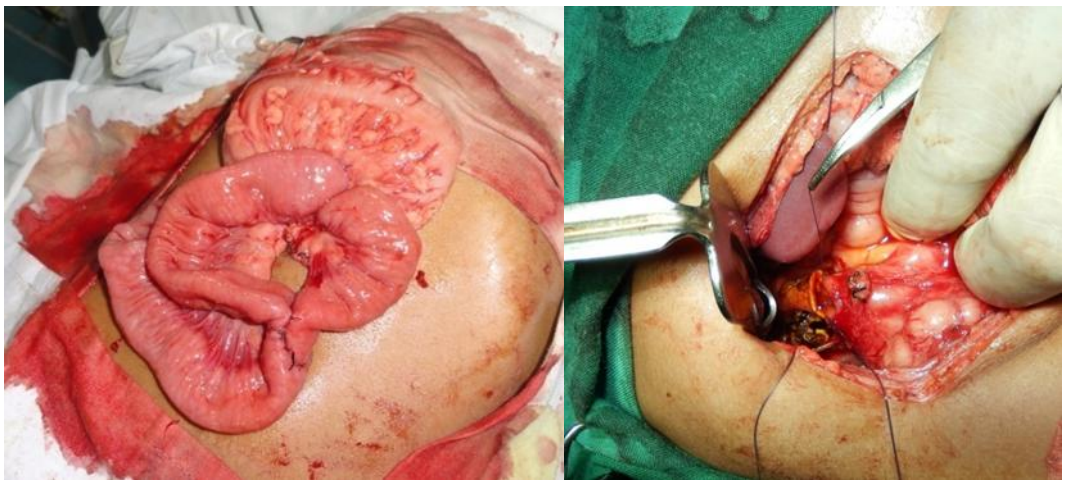

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Figure: operative finding (type 1 CC in 2 months old female).

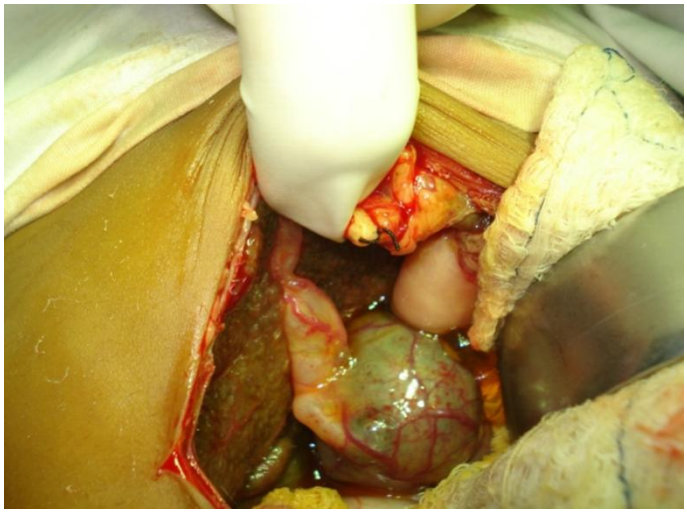

Table 4: Outcome of the patients.

	No. of patients
Recovered uneventfully	**11**
Post op intestinal obstruction(recovered)	**1**
Expired	**3**

Discussion:

Choledochal cysts present primarily in female infants and young children and are more prevalent in East Asian populations[8]. International literature reports a 4:1 female to male ratio with predominance in Asian population, however, in Mexican literature this is variable with different authors reporting different proportions. Orozco-Sanchez et al., 1997 reported 6:1 ratio, Gallardo-Meza et al., 2010 reported 2:1 ratio. In our study male to female to male ratio is 4:1. The classical clinical triad of jaundice, pain, and a palpable mass can be found in about 20% of cases, with predominance in childhood[9]. In this study, there are 11 patients in this group having significant jaundice and 5 cases with hepato-splenomegaly.

Abdominal ultrasound (US) is the first choice of imaging for diagnostic approach, whose certainty, can reach a correct diagnosis in 50%,with an appropriate CC classification in 30%. Although it is operator dependant. All our patients had an abdominal US. The MRI has the advantage of simultaneously and effortlessly delineatimg both the biliary and pancreatic duct and therefore accordingly to the cholangiography morphology classify the images as per Todani's classification. MRCP and ERCP superceded the use of Computed Tomography for preoperative anatomical delineation of the pancreatico-biliary tract. Type-I choledochal cyst was the most common type (87%) and type-IV was seen in 13% as per Todani's classification[9]. We performed MRCP for all the patients, out of which 13 patients had type 1 Choledochal Cyst. Early surgery can reverse biliary cirrhosis and portal hypertension. Most surgeons advocated total excision with hepaticojejunostomy as the procedure of choice. We performed the same in 13 out of 15 patients. At the time of follow-up all the survived patients were

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jaundice free and liver parenchyma returned to normal. Some of the difficulties faced during diagnosis are physiological jaundice, features similar to neonatal hepatitis and biliary atresia and parental delay in seeking medical advice.

Conclusions:

More cases of choledochal cyst are being diagnosed in infancy. Early ultrasonography should be done for prolonged neonatal hyperbilirubinemia. MRCP provides excellent delineation of the cysts. Early surgery can reverse biliary cirrhosis and portal hypertension. If diagnosed in antenatal or neonatal period surgery should be done within 8 weeks to prevent cirrhosis. In prolonged neonatal hyperbilirubinemia choledochal cyst should be kept in mind. Small babies also tolerate the surgery provided liver function is good. Roux-en-Y hepatico-jejunostomy is the standard procedure, however Hepatico-duodenostomy in resurging as an viable alternative.

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