



## Hepatobiliary Surgery

## CHOLEDOCHAL CYST IN CHILDREN AND SIGNIFICANCE OF LIVER BIOPSY

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**ABSTRACT** Choledochal cyst is one of the conditions that typically presents in infancy and childhood. 25% of the patients are diagnosed within the first year of life and 60% before the age of 10 years. The classic triad of jaundice, abdominal mass, and pain occurs less commonly in adults with choledochal cysts than in children. Liver histology has not been given due importance in the course of treatment of choledochal cyst. Liver biopsies are not routinely taken during resection of choledochal cysts and histological changes are seldom reported. In the present study 40 cases are included, age at presentation, clinical presentation, type of cyst, surgical procedure and histopathological grading of liver are documented. Histopathological grading of liver is correlated with age, clinical presentation and severity of liver damage. The present study is undertaken to assess the histopathology of liver in choledochal cysts and to grade the hepatocellular damage and fibrosis and correlate with age and clinical presentation of disease and to emphasise the importance of liver biopsy in choledochal cysts.

**KEYWORDS :** Choledochal Cyst, Liver Biopsy, Children, Histopathology

## INTRODUCTION

Congenital dilatation of the extrahepatic biliary tract with or without dilatation of the intrahepatic biliary tract, first described in 1852 by Douglas is known as the choledochal cyst (CC) <sup>1</sup>. It occurs in approximately 1 to 10<sup>3</sup> live births <sup>2</sup>. The condition typically presents in infancy and childhood. 25% of the patients are diagnosed within the first year of life and 60% before the age of 10 years but 20% are diagnosed after the age of 20 years. <sup>3</sup>Todani and his associates have classified the anomaly into five main types and additional subtypes, based on analysis of cholangiogram. <sup>5,6</sup> Choledochal cysts are more common in female than in male patients, and the female-to-male ratio ranges from 3:1 to 4:1. <sup>7,8</sup> The classic triad of jaundice, abdominal mass, and pain occurs less commonly in adults with choledochal cysts than in children. Liver histology has not been given due importance in the course of treatment of choledochal cyst. Liver biopsies are not routinely taken during resection of choledochal cysts and histological changes are seldom reported. These changes correlate with the clinical symptoms at presentation. The present study is undertaken to assess the histopathology of liver in choledochal cysts and to grade the hepatocellular damage and fibrosis and correlate with age and clinical presentation of disease.

## METHODOLOGY

The study is a prospective study done for two years from November 2017 to November 2019 in the Department Of Pediatric surgery, Niloufer hospital for women and children, Red hills, Hyderabad, Telangana. Niloufer hospital is a tertiary care center for pediatric and maternal cases. A total of 40 cases are taken up for the study, children >12 years are excluded from the study. All children <12 years diagnosed with choledochal cyst were evaluated clinically, radiologically and biochemically at presentation. After preoperative evaluation and surgical consent, case is taken for surgery, cyst excision with hepatico-enterostomy was done. Intra-operatively liver biopsy was taken in all cases.

## RESULTS

**AGE DISTRIBUTION:** Age of patients ranged from 4 months to 11 years.

**Table 1 : Age Wise Distribution Of Choledochal Cyst.**

AGE	NUMBER	PERCENTAGE
<1 YR	10	25%
1-5 YRS	12	30%
5-12 YRS	18	45%

**SEX DISTRIBUTION:** Of the 40 cases, 15 cases (38%) were male patients and 25 cases (62%) were female. The Male: Female ratio being 1:1.6.

**Table 2: Sex Distribution**

SEX	NUMBER OF CASES	PERCENTAGE
Male	15	38%
Female	25	62%

**CLINICAL PRESENTATION:** Most common presentation of choledochal cyst in present study is pain abdomen seen in 23 cases

(57.5%). Vomiting is seen in 20 cases (50%), fever in 7 cases (17.5%), jaundice in 19 cases (47.5%), abdominal lump in 5 cases (12.5%), and 2 patients presented with pale stools (5%).

**Table 3: Distribution Of Clinical Presentation**

CLINICAL PRESENTATION	CASES	PERCENTAGE
Pain abdomen	23/40	57.5%
Vomiting	20/40	50%
Jaundice	19/40	47.5%
Lump abdomen	5/40	12.5%
Fever	7/40	17.5%
Pale stools	2/40	5%
Classic triad	3/40	7.5%

**SURGICAL INTERVENTION:** In 28 cases (70%), Roux en Y hepaticojejunostomy was done and 12 cases (30%) hepaticoduodenostomy was done.

**CLASSIFICATION OF CHOLEDOCHAL CYST:** 31 cases had type I and 9 cases had type IV. Majority of cases in the present study belonged to type I cyst.

**HISTOPATHOLOGY OF LIVER BIOPSIES:** Histological changes of higher grades were observed in infants (<1yr) and of lower grades were observed in older children (>5yr). Out of 10 infants, 7 showed grade 2 hepatocellular damage and 3 showed grade 3 hepatocellular damage. Most of the older children showed grade 1 hepatocellular damage. All cases showing grade 3 cholestasis were also infants. All the infants showed liver fibrosis of varying degrees. Most of the infants showed grade 3 and grade 4 liver fibrosis. All cases showing cirrhotic changes were infants.

**Table 4: Histopathological Grading Of Liver Changes**

Age	CHS			HCD			PI			BDP			BDI			CVD			PF			
	1	2	3	1	2	3	1	2	3	1	2	3	1	2	3	1	2	3	1	2	3	4
Infants(10)	3	3	4	0	7	3	2	4	4	3	4	3	3	3	4	4	2	1	2	5	2	
1-5yrs(12)	5	2	5	5	5	2	4	5	2	3	5	2	5	3	3	6	1	5	4	2	0	
>5yrs(18)	3	0	0	10	3	0	5	6	2	5	5	2	7	5	0	3	8	0	5	6	1	0

CHS- cholestasis; HCD- hepatocellular damage; PI- portal inflammation; BDP- bile duct proliferation; BDI- bile duct inflammation; CVD- central venous distension; PF- portal fibrosis.

## DISCUSSION

In the present study, none of the patients were diagnosed antenatally. The age of patients ranged from 4 months to 11 years. The youngest patient was 4 month old male infant. In our setup, we found that choledochal cyst was more commonly diagnosed in children more than one year of age with female preponderance which was comparable to other studies.

**Table 5: Comparative Studies Of Age Of Presentation And Gender Distribution.**

Studies	Age		Gender	
	Infantile	Paediatric	Male	Female

Man Chin Hua et.al(2007)	54.2%	45.8%	2:3
M Germani et al (2011)	41.6%	58.3%	1:5
N.Sugandhi et. al (2014)	8.7%	91.3%	1:1.7
Chatura et al (2016)	31.25%	68.75%	-
PRESENT STUDY	25%	75%	1:1.6

In **Present study** the patients were admitted with jaundice in 19 cases (47.5%), vomiting was seen in 20 cases (50%), fever in 7 cases (17.5%), pain abdomen in 23 cases(57.5%), abdominal lump in 5 cases (12.5%), and classical triad of pain, lump and jaundice was seen in 3 cases which was comparable to other studies.

The most common presentation was pain abdomen overall. Infants mostly presented with jaundice, vomiting and fever as pain abdomen is not evident in infants. Older children mostly presented with pain abdomen and jaundice.

**Table 6: Comparative Studies Of Clinical Presentation**

	Omar et al (2009)	Mukhopadhyay et al (2011)	Arah et al (2015)	Chatura et al (2016)	Nabila Talat et. al (2016)	PRESENT STUDY
<b>Jaundice</b>	31.2%	42%	35%	90%	79.24%	<b>47.5%</b>
<b>Vomiting</b>	-	-	60%	18.75%	30.18%	<b>50%</b>
<b>Fever</b>	-	-	-	80%	54.7%	<b>17.5%</b>
<b>Abdominal pain</b>	81.2%	86%	77%	18.75%	62.26%	<b>57.5%</b>
<b>Abdominal lump</b>	-	30%	-	12.5%	67.9%	<b>12.5%</b>

In the present study, hepatocellular damage correlated significantly with younger age at presentation, damage of higher grade being present in younger patients. It was also observed the liver fibrosis of higher grades was found in younger patients and all the infants showed severe expansive fibrosis and cirrhotic changes. This is in agreement with the fact that infants have more severe disease at presentation.<sup>9,10</sup> This means that these patients have more severe disease and hence they present early, which is reflected by the higher grades of hepatocellular damage seen at the time of presentation.

**Table 7 : Comparison Of Histological Findings With Study By N.Sugandhi et.al**

Histological Parameter	Grade	Present study (% of cases)	Sugandhi et.al AIIMS, DELHI (%)
Cholestasis	0	45	28.25
	1	27.5	37
	2	17.5	28.25
	3	10	6.5
Hepatocellular Damage	0	10	0
	1	37.5	35
	2	37.5	50
	3	15	15
Parenchymal Inflammation	0	10	6.5
	1	27.5	37
	2	42.5	48
	3	20	8.5
Bile Duct Proliferation	0	15	13
	1	27.5	37
	2	40	32.5
	3	17.5	17.5
Bile Duct Inflammation	0	12.5	9
	1	40	39
	2	32.5	41
	3	15	11
Central Venous Distension	0	17.5	35
	1	25	54
	2	50	11
	3	7.5	0
Liver Fibrosis	0	12.5	21
	1	27.5	35
	2	30	35
	3	20	9
	4	10	0

## CONCLUSION:

Choledochal cyst is a disease of bile duct as well as liver. All patients with choledochal cyst have histopathological changes of varying degrees in the liver. More severe symptoms are associated with higher degree of liver damage. These cases demand early intervention for symptomatic relief and also to prevent permanent hepatic damage in the form of fibrosis. Presentation less than 1yr is associated with more parenchymal damage and fibrosis. The hepatocellular damage and fibrosis correlated significantly with younger age at presentation and with severe clinical symptoms. These cases need early intervention and follow up to prevent progressive liver damage.

Liver biopsy is not routinely done in all cases of choledochal cyst. In the present study the severity of liver pathology has impact on the age of presentation and clinical severity of symptoms, hence liver biopsy should be done in all cases of choledochal cysts.

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