

## Clinico-pathological features of Congenital biliary malformations with emphasis on Choledochal cysts

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### Abstract:

**Introduction :** Congenital disorders of biliary tract are broadly classified into biliary atresia, fibropolycystic diseases, aberrant biliary ducts, aberrant cystic duct insertions. Choledochal cysts (CCs) are rare and are uncommon congenital anomalies of bile ducts, which are cystic dilatations of any portion of the bile ducts, mostly in the main portion. The aim is to study clinical and histopathological features correlating with the imaging findings and to Classify based on radiological findings and assess complications if present.

**Materials and methods:** This is an observational study which was conducted, in the Department of pathology at Rangaraya Medical College, Kakinada, Andhra Pradesh from July 2009 to June 2018. A total of 33subjects (both male and females) were included in this study.

**Conclusion:** Clinical suspicion of Choledochal cysts should be followed by early diagnosis and management as later the diagnosis, worst is the prognosis. Most lesions warrant resection and histopathological examination to avoid future malignancy.

**Keywords:** choledochal cysts, complications, imaging techniques, todani classification.

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### I. Introduction

Congenital disorders of biliary tract are broadly classified into biliary atresia, fibropolycystic diseases, aberrant biliary ducts, aberrant cystic duct insertions<sup>(1)(4)</sup>. The fibropolycystic diseases are further classified into choledochal cysts, polycystic liver disease and congenital hepatic fibrosis among which choledochal cysts (CCs) are rare and are uncommon congenital anomalies of bile ducts, which are cystic dilatations of any portion of the bile ducts, mostly in the main portion<sup>(2)</sup>. Although they are seen mostly in childhood and infancy, the affected age groups range from newly born to 80 years old; however 60% of such cysts are diagnosed in patients less than 10 years old<sup>(1)(8)</sup>.

### II. Materials and Methods

This is an observational study which was conducted, in the Department of Pathology at Rangaraya Medical College, Kakinada, Andhra Pradesh from July 2009 to June 2018. A total of 33subjects (both male and females) were included in this study.

**Study Design:** Retrospective observational study.

**Study Location:** This was a tertiary care teaching hospital based study done in Department of Pathology at Rangaraya Medical College, Kakinada, Andhra Pradesh.

**Study Duration:** July 2009 to June 2018.

**Sample size:** 33 patients.

### Procedure methodology

Clinical details like age, sex, biochemical & radiological investigations and clinical diagnosis were obtained from the hospital records. Types of specimens obtained were resected CBD specimen, Cholecystectomy specimen, liver biopsy specimen. Specimens were grossed strictly according to Royal College of Pathology protocols and processed. The paraffin embedded samples were cut to 4µ thick sections, and were stained with regular haematoxylin and eosin(H&E) stain.

### III. Results

Total number of cases with biliary atresia = 17

Total number of cases with choledochal cysts= 16

Total females affected with choledochal cysts=12

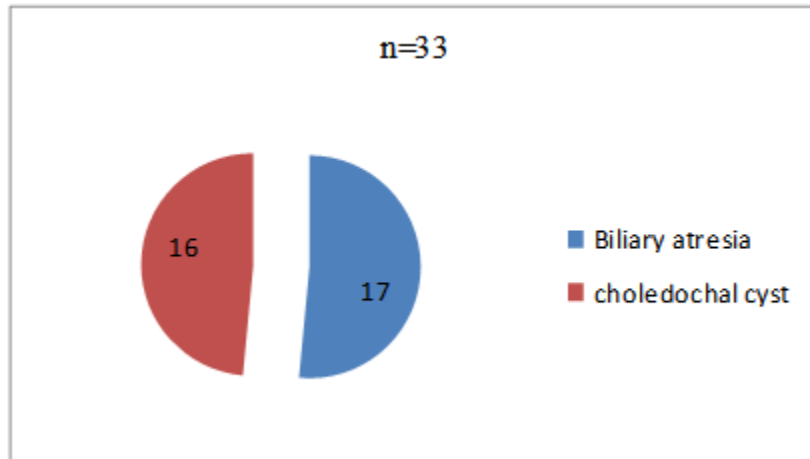
Total males affected with choledochal cysts=4

Most common age group affected = 4to 10 years

Most common subtypes encountered =TypeIA>IB>IC>I-unclassified >Type 4A

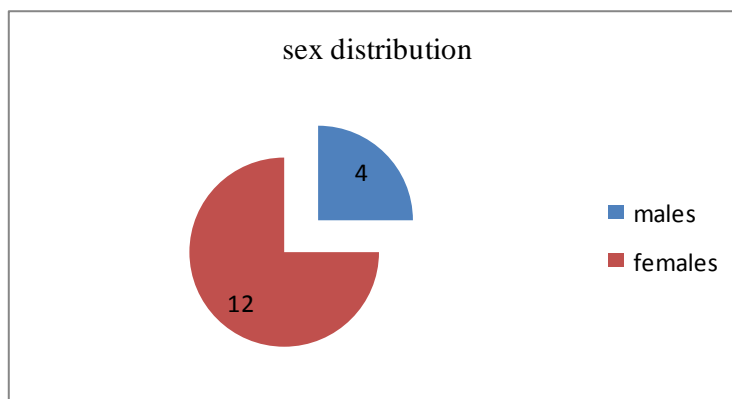
### IV. Figures and Tables

**Figure no 1:** shows sex distribution of choledochal cysts



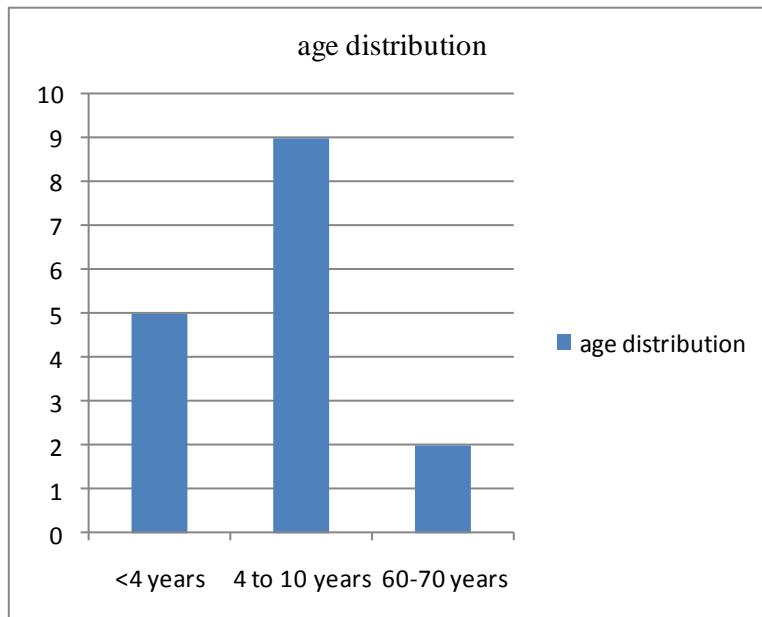
The total number of cases were 33 out of which 17 (51.5%) were biliary atresia and 16(48.5%) were Choledochal cysts

**Figure no 2 :** shows sex distribution



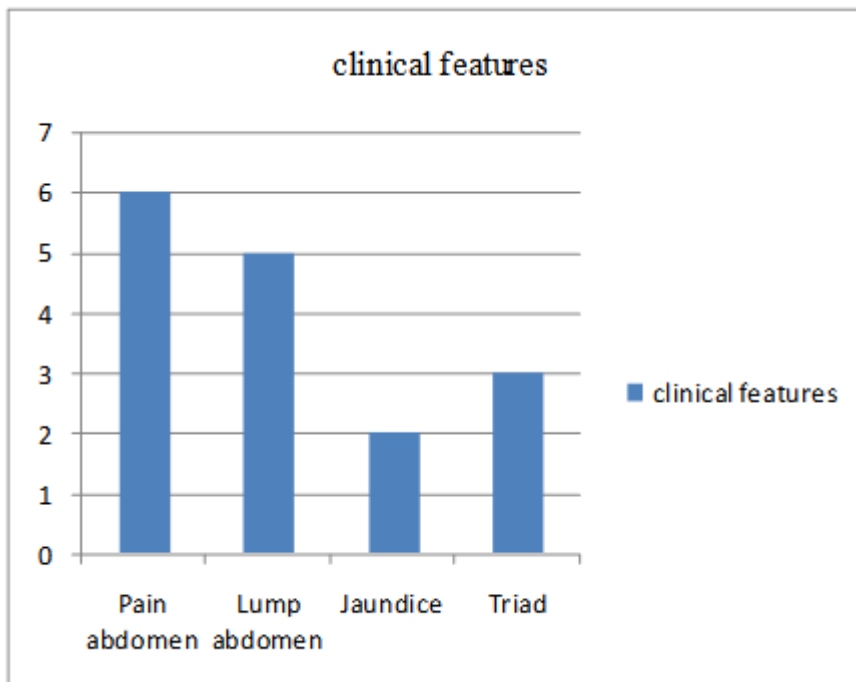
In the present study ,among 16 choledochal cysts 12 (75% ) were females and 4 (25%) were males.

**Figure no 3: Age distribution**



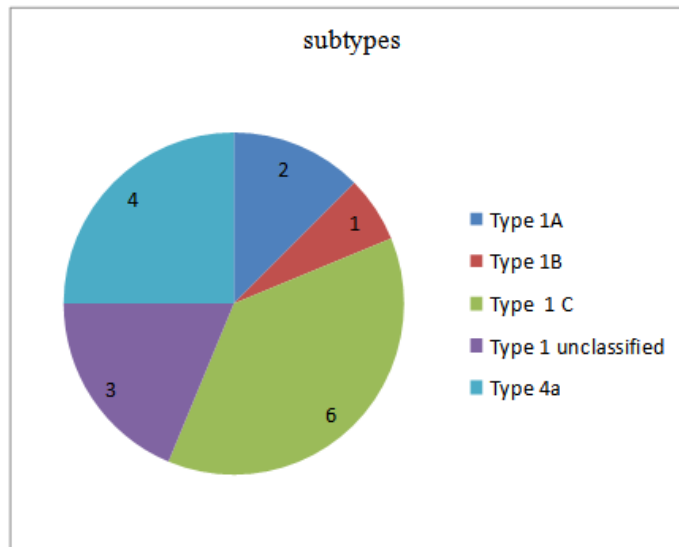
In our present study most of the lesions were diagnosed in the age group of 4 to 10 years. Only two( 12.5%) cases were diagnosed in adults

**Figure no 4: Clinical Features**



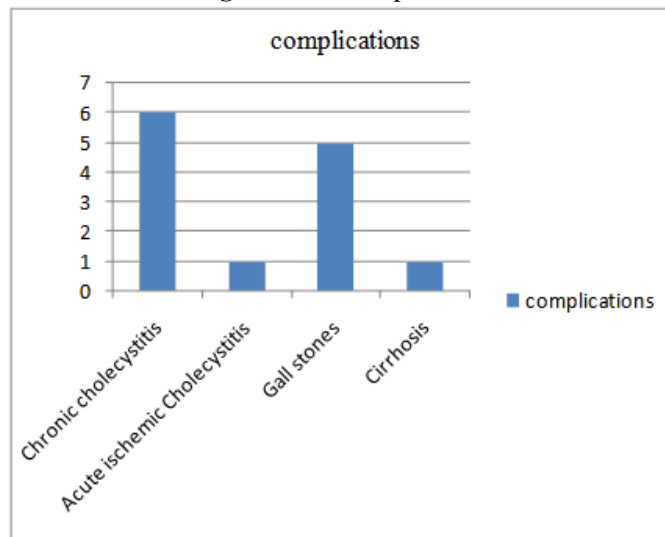
In the present study ,most of the patients (68.5%) had symptoms of pain abdomen and lump abdomen whereas, 18.75% of patients had the clinical triad of symptoms. Jaundice was observed in 12.5%.

**Figure 5: Subtypes of Choledochal Cysts**



In the present study majority of subtypes were of Type IC constituting 37.5% followed by Type VI A . The other subtypes were of Type 1 unclassified constituting 18.75%, Type 1A 12.5% and Type 1B 6.25%.

**Figure no 6: Complications**



In the present study most common complications encountered were chronic cholecystitis and gall stone formation.

#### IV. Discussion

**Epidemiology:** The incidence of CCs is 1 in 100,000–150,000 live births in the western population, but reported to be as high as 1 in 13,500 live births in the United States and 1 in 15,000 in Australia.[1] The incidence is higher in asian population<sup>(3)</sup>. CCs have an female:male preponderance, commonly reported as 4:1 to 3:1<sup>(6)</sup>.

**Classification:** Initially Alonso-Lej classified choledochal cysts into three types (I – III) of dilatations in 1959. Later Todani et al expanded it adding two more groups to it (IV-V) which is widely accepted by most of the surgeons<sup>(7)</sup>.

Todani's classification

Type I A - Cystic dilatation of extra hepatic bile ducts

Type I B – focal/segmental dilatation of CBD

Type I C – Fusiform dilatation of entire CBD

Type II – Supra duodenal diverticulum

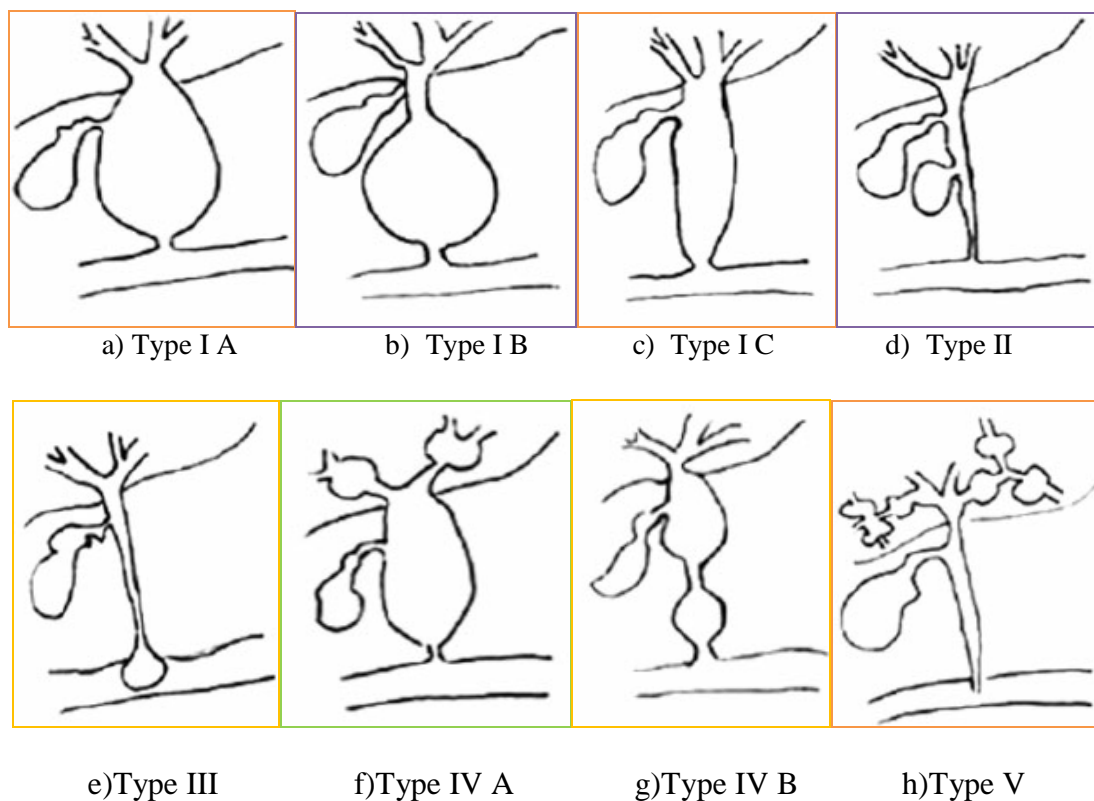
Type III – Dilatation of intra duodenal CBD

Type IV A – Dilatation of both intrahepatic and extrahepatic bile ducts

Type IV B – Multiple dilatations in extrahepatic bile ducts

Type V – Dilatations in intrahepatic bile ducts only

90 to 95% of choledochal cysts are classified as type I this involves a fusiform or saccular dilatation of common bile duct .infants commonly have a complete obstruction of the distal common bile duct<sup>(4)</sup> whereas in adults ,the distal common bile duct is mostly patent . rarely the lesion may be entirely intra duodenal or consist of multiple intrahepatic cysts (caroli's disease)<sup>(8)</sup>.



**Etiopathogenesis:** The most commonly proposed theory is Babbitt's theory, where CCs are supposed to be caused by an abnormal pancreatico-biliary duct junction (APBDJ) in which the pancreatic duct joins the bile duct 1–2 cm proximal to the sphincter of oddi<sup>(5)</sup>. This Abnormal pancreatic and biliary duct union causes reflux of pancreatic secretions & transmission of high pressure into CBD causing dilatation of the duct. The increased pancreatic secretions cause epithelial damage, inflammation and fibrosis .Over longstanding period it causes epithelial hyperplasia & dysplasia, few cases may turn into malignancy<sup>(6)</sup>. This theory is supported by finding of high amylase levels in CCs bile. Few other proposed theories are Obstruction of distal CBD and Sphincter of oddi dysfunction which also also results in pancreatic juice reflux into bile ducts predisposing to formation of choledochal cysts<sup>(3)</sup>.

**Clinical presentation:** The classic triad of symptoms,are pain abdomen, palpable abdominal mass, and jaundice.85% of children have at least 2 features of classic triad<sup>(8)</sup>. whereas only 25% of adults present with at least 2 features of the classic triad.

Chronic obstruction leads to secondary biliary cirrhosis,and pancreatitis . At times the cyst may get infected, and gets ruptured resulting in secondary bacterial peritonitis<sup>(1)</sup>.Recurrent pancreatic reflux causes chronic inflammation, biliary stasis and stone formation<sup>(2)</sup>. Chronic inflammation also causes cell regeneration, impaired DNA breaks & repair leading to dysplasia and malignancy.

**Risk of malignancy:** The Incidence of malignancy is 2.5-17.5% and the risk of malignant change Increases with age and incomplete resection .The histological types are adenocarcinoma (80%) > anaplastic carcinoma (10%) > undifferentiated carcinoma (5-7%) > squamous cell carcinoma

The risk for Type 1 is (50-62%) > Type IV (25%) > Type III&IV (6%) > Type II(3%). Location of carcinoma is extrahepatic bile duct > Gall bladder > intrahepatic bile duct > liver<sup>(2)</sup>.

**Diagnosis:** Blood investigations and imaging should be done in patients with clinical suspicion of CCs. Blood investigation may reveal altered liver function tests and leukocytosis in cholangitis due to CCs. Raised serum

amylase and lipase indicates pancreatitis<sup>(6)</sup>. Raised CA 19-9 should raise the suspicion of malignancy in adults with CCs<sup>(2)</sup>.

**Role of imaging:** Imaging techniques confirm the diagnosis of CCs. Improved imaging techniques has made possible the diagnosis antenatally and also incidentally in adults. Radiographic visualization of both biliary system and pancreatic duct prior to surgery helps in complete excision of CCs<sup>(7)</sup>. Abdominal ultrasound (US) scan is the first step toward confirmation of diagnosis. Sensitivity of US is about 71– 97%<sup>(2,6)</sup>. It is also the preferred investigation in postoperation surveillance. Hepatobiliary scintigraphy using technitium-99 hepatobiliary iminodiacetic acid (HIDA) has a sensitivity of 100% for type I CCs<sup>(3,7)</sup>. It is useful to differentiate biliary atresia from CCs in the newborns<sup>(4)</sup>. Computed tomography (CT) is highly accurate and also help in planning surgical approaches. It delineates well the intrahepatic biliary dilation in type IVA and Caroli's disease and helps in surgical planning.

MRCP is regarded as the “gold standard” for the diagnosis of CCs Sensitivity has been reported to be as high as 90–100%<sup>(6,7)</sup>. MRCP avoid ionizing radiation and is also noninvasive when compared with ERCP.

**Pathological features :** Fibrosis and lymphocytic infiltration of the cyst wall is typical in pediatric CC; adult CC includes evidence of inflammation and hyperplasia. Adjacent liver shows portal fibrosis, central venous distention, parenchymal inflammation, and bile duct proliferation. Certain differences seen in the histologic appearances of the different CC subtypes. Type I (and sometimes type IV) CC lack biliary mucosa; type II CC closely resemble gallbladder duplication. Type III cysts are lined by duodenal mucosa, while type V cysts can have extensive hepatic fibrosis<sup>(2,7,8)</sup>.

In the present study few of the follow up cases with clinical radiological and pathological features were discussed

Case1

A 14 month old baby presented with abdominal pain and jaundice ,serum bilirubin was elevated 15gm/dl and the ultra sound report showed dilatation of CBD as Choledochal cyst.after resection, histopathological findings of the cyst showed cystic dilatation and inflammation, adjacent liver tissue showed features of biliary cirrhosis

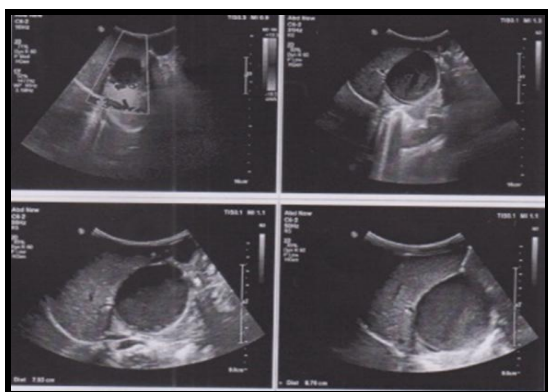


Figure 7: USG dilated choledochal cyst



Figure 8: Bi 1720/18,Gross

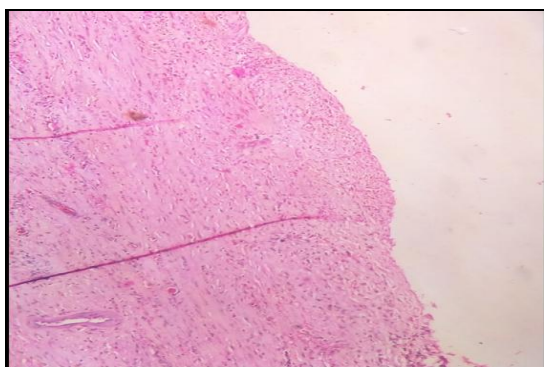


Figure 9: Bi 1720/18 H&E 10X liver  
Liver biopsy showing features of biliary cirrhosis

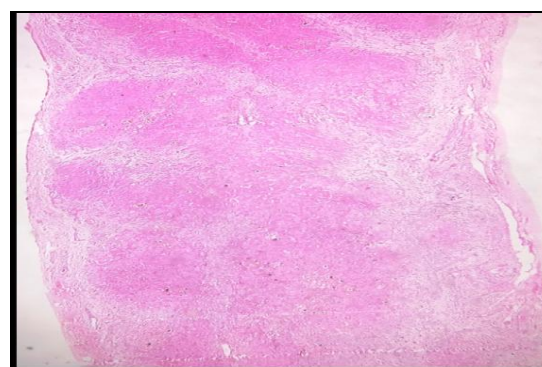


Figure10: Bi1720/18 H&E 4X , liver



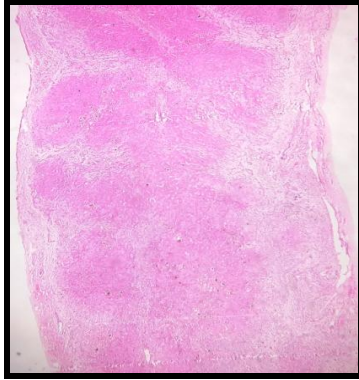


Figure 11: Bi:1720/18 H&E 4X

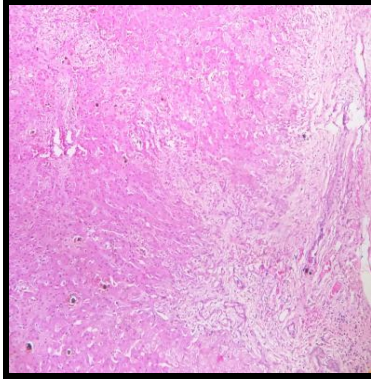


Figure 12: H&E 10X

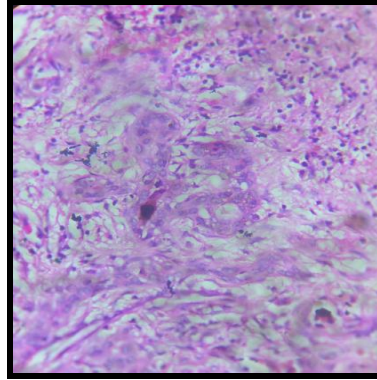


Figure 13: H&E 40X

**Case 2**

A 3 year female child presented with recurrent episodes of pain abdomen since 2 weeks

On MRI reported as Choledochal cyst type IV A with cholelithiasis and choledocholithiasis

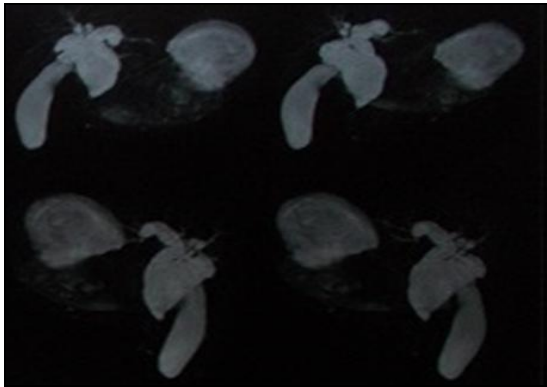


Figure 14: MRI



Figure 15: Bi:3482/17, gross

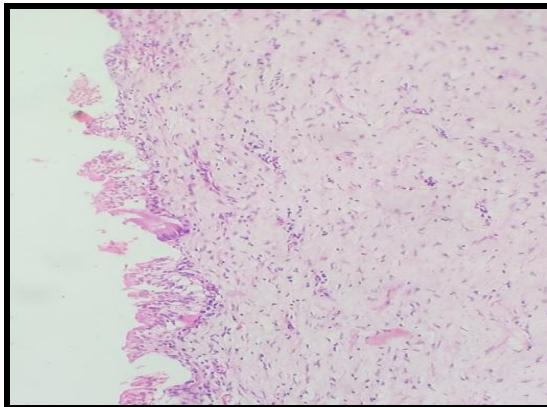


Figure 16: Bi 3482/17, H&E, 10x, choledochal cyst

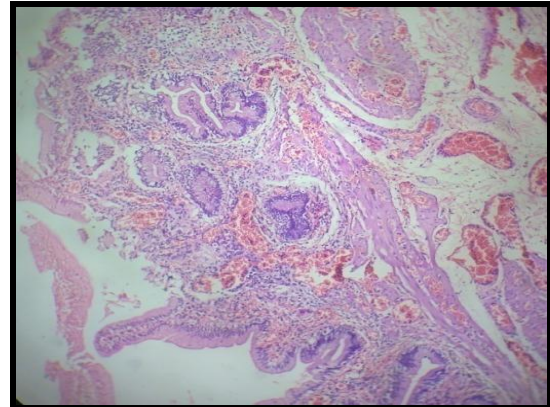


Figure 17: Bi 3482/17, H&E, 10x, Gall bladder

**Case 3**

A 5yr old female -recurrent episodes of pain abdomen since o/e : right hypochondrial mass is seen

USG showed features of Choledochal cyst. MRCP showed fusiform dilatation of CBD, Choledochal cyst Type IC

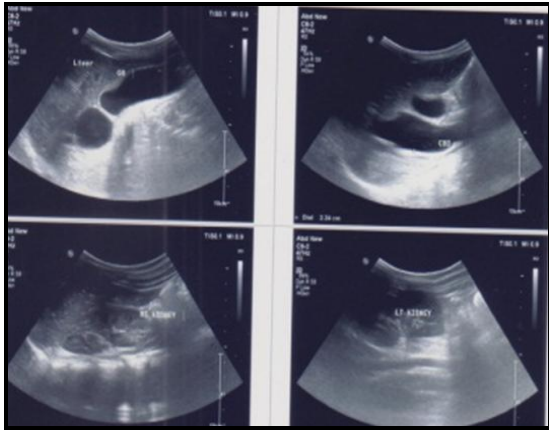


Figure 18:USG:choledochal cyst

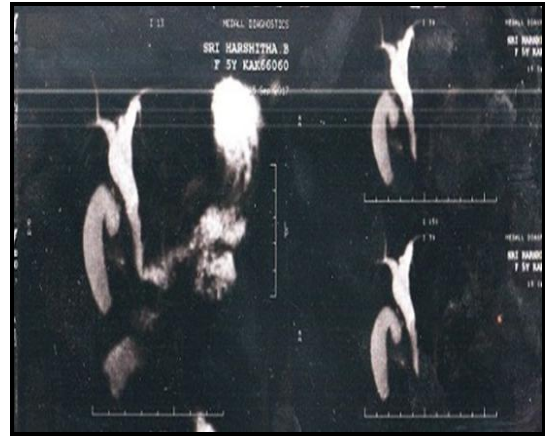


Figure19: MRCP: fusiform dilatation of CBD



Figure 20:Bi : 4555/17 gross

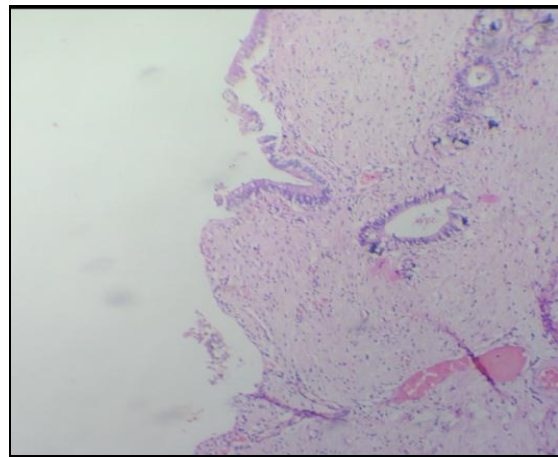


Figure 21:Bi 4555/17 H&E 10x

**Table no 1: Comparison with other studies**

VARIABLES	PRESENT STUDY	SHAKIR RASHID et al	MUKHOPADYAY et al	J.RAWAT et al	PRADEEP CHOPRA et al
Male: Female	1 : 4	1 : 2	1 : 2	1:1.2	1:4
Abdominal Pain	87.5%	78.5%	86%	85%	90%
Triad	18.75%	--	21.5%	20%	10%
Type 1 Choledochal cyst	75%	71%	79.5%	65%	80%
Type IV Choledochal cyst	25%	26%	20.3%	20%	--
Gall Stones	31.25%	40%	--	--	--

## VI. Conclusion

Clinical suspicion of Choledochal cysts should be followed by early diagnosis and management as later the diagnosis , worst is the prognosis. Most lesions warrant resection and histopathological examination to avoid future malignancy

## Acknowledgements

An acknowledgement section may be presented after the conclusion, if de I would like to thank Dr V. Rajani MD, Assistant Professor, of Pathology, Rangaraya Medical college, Kakinada, for their continuous support and guidance.



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