Objective: To study the clinicopathological status, role of various diagnostic and treatment modalities and outcome of patients with choledochal cyst in Kashmir valley.

Materials and Methods: The study was both retrospective and prospective in nature. After a thorough history and examination, the investigations included all baselines like haemogram, LFT, KFT, in addition to the specific investigations like USG, CECT, ERCP, MRCP, bile analysis for amylase and lipase levels and bile culture. The operative findings including the indication and type of procedure were recorded.

Results: Out of total 60 patients, children were 20 and adults 40 with a Male: Female ratio of (1:4.5). The most common feature in both groups was pain (77–80%), followed by jaundice. Triad of pain, jaundice and mass was seen more in children than in adults. Complications before surgery for choledochal cyst including cholangitis, pancreatitis, cholecystitis, stricture and stone formation were more common in adults except biliary peritonitis and ascites which were exclusively seen in children. Only one patient among the children had raised CEA level (>10ng/ml) Vs adults (25%). The main organisms responsible for infection were Klebsiella pneumoniae and E. coli. Histologically fibrosis was a feature of childhood choledochal cyst whereas hyperplasia and inflammation was a feature of choledochal cyst in adults. MRCP was done in 47 patients with a diagnostic accuracy of 100% for diagnosis.

Conclusion: The presentation is often vague and non-specific impending prompt diagnosis. However the diagnosis is facilitated by modern imaging techniques and may be made at any time from antenatal period to adulthood. There appears to be a justification for treating the disease in children and adults as separate entities in view of varied clinical presentation. The classical triad is now known to apply to only minority of patients. MRCP was found to be 100% sensitive for diagnosis.

Key words: choledochal cyst, MRCP, Diagnostic tools, Triad PJM
INTRODUCTION
Choledochal Cysts are congenital anomalies which present as either isolated or combined dilatations of the extra and intra-hepatic biliary tree. [1] As the condition is not confined to the extrahepatic bile ducts, the term ‘choledochal cyst’ is in fact a misnomer and ‘bile duct cyst’ or ‘biliary cyst’ is more appropriate. Nevertheless, due to the longstanding usage and familiarity, the term ‘choledochal cyst’ is generally accepted. It occurs in approximately 1 to 103 live births. [2] The condition typically presents in infancy and childhood. [3] Twenty five percent of the patients are diagnosed within the first year of life and 60% before the age of 10 years and the adult presentation is common. [4] In patients with previous surgery, there is increased likelihood of association of hepatobiliary disease that makes management difficult in adults. [5] Female to male ratio as high as 8:1 has been observed. [6] The classic triad of jaundice, pain and abdominal mass is encountered more often in children than in adults (85 versus 25 percent respectively. [7] The time from development of symptoms to diagnosis and treatment is significantly longer in adults than in children, with a mean duration of about 6 years in the former. [8] Adults usually present with vague symptoms, generally in the right upper abdominal quadrant. Jaundice is a less frequent feature in adults (reported in 10-15 percent) whereas cholecystitis, cholangitis and pancreatitis tend to occur at a somewhat higher rate (20-60 percent) than in children. In neonates or children, choledochal cyst usually presents as abdominal mass or abdominal pain. [9] Biliary tract malignancy has been reported to occur in 2.5% to 28% of patients with choledochal cyst, representing a risk at least 20 times greater than that of normal population. [10] Bile duct cysts are classified according to site, extent and shape of the cystic anomaly. Alanso-Lej et al proposed a classification for extrahepatic bile duct cysts in 1959, which was later modified by Todoni et al. [11] Todani incorporated the intrahepatic dilatations described by Caroli et al (known by the eponym Caroli’s disease). [12] Different types of bile duct cysts, according to Todani’s Classification are.. Type I : Solitary extrahepatic cyst Type II: Extrahepatic supraduodenal diverticulum Type III: Intraduodenal diverticulum (choledochocele) Type IV: Extrahepatic intrahepatic dilatation (IV a) Fusiform extra and intrahepatic dilatation (IV b) Multiple extrahepatic cysts Type V: Multiple intrahepatic cysts (Caroli’s disease)

The distribution of cyst types is similar in adults and children, with the exception of type IV cysts, which are seen more frequently in adults. Type I cysts are most common (79%) and type II (3%). Least common are type V cysts (Caroli’s disease) which are found in less than 1% of all affected patients. [13] There are many proposed theories about etiology of choledochal cyst but none of them is applicable in all such cases. The most favored hypothesis about the formation of choledochal cyst is the “long common channel” (LCC) theory, first described by Babitt. [14] Anomalous pancreatobiliary union is detected in 57% to 96% cases of choledochal cyst leading to reflux of pancreatic secretions into bile duct is also attributed to development of choledochal cyst. [15] Oligoganglionosis of the distal CBD may result in inadequate autonomic innervations, producing a functional obstruction and proximal dilatation. [16] Abnormal function or dysmotility of the
The sphincter of Oddi has been noted in several patients with choledochal cysts.\textsuperscript{[17]} The possibility of viral etiology has been considered after the detection of retroviral RNA in tissues from patients with choledochal cyst.\textsuperscript{[18]} There are few reports of familial cases of choledochal cysts and associated anomalies. A pair of monozygotic twins was discordant for the occurrence of anomalous pancreatobiliary junction and choledochal cyst, suggesting that there is not a strong genetic basis for these malformations.\textsuperscript{[19]} Diagnosing the disease entity has become simpler in the recent years because of various radiological aids. USG can diagnose choledochal cysts with a specificity of 97\% in children.\textsuperscript{[20]} It is an excellent first line investigation of neonatal jaundice persisting for >2 weeks after birth,\textsuperscript{[21]} and may help to differentiate CDC from biliary atresia.\textsuperscript{[22]} Radionuclide scintigraphy is safe and non-invasive, and has been in use for a long time in the diagnosis of choledochal cysts.\textsuperscript{[23]} Following the progress of an isotope from the biliary tract into the small intestine is reported to distinguish with 100\% accuracy between CDC and biliary atresia.\textsuperscript{[24]} Although there are reports of computed tomographic (CT) scans diagnosing choledochal cyst,\textsuperscript{[25]} but it has been found that cysts are missed on CT scans and picked up on MRCP.\textsuperscript{[26]} A better role for CT scanning may be postoperative period where it was shown to be superior to MRCP in locating the biliary-enteric anastomosis and in defining any stenosis thereof. Endoscopic retrograde cholangiopancreatography (ERCP) is an excellent tool for defining biliary anatomy, and as such has been used to diagnose choledochal cyst.\textsuperscript{[27]} (MRCP) represents the current ‘gold standard’ in the imaging of CDC.\textsuperscript{[28]}

Pancreatitits is found in 30-70\% of adults with bile duct cysts. The risk of pancreatitis increases with increased cyst size and those associated with anomalous pancreatic biliary duct. Other complications include stones (cystolithiasis, hepaticolithiasis), inflammation and infections (calcus cholecystitis, cholangitis, intrahepatic abscess), secondary changes (hepatic cirrhosis with portal hypertension).\textsuperscript{[29]} Mechanical obstruction of the pancreatic duct and development of cholangiocarcinoma are associated problems that occurs significantly more often in adults than in children. Hepaticolithiasis is most often noted in patients with type IV bile duct cysts. Cyst rupture on rare occasions in adults may present with an acute abdomen due to cyst rupture and biliary peritonitis.\textsuperscript{[30]} Although this is more frequently encountered in children, situations associated with associated increased intra-abdominal pressure in adulthood, such as pregnancy may predispose to cyst rupture. It is well established that 10-30\% of adults with bile duct cysts develop cholangiocarcinoma.\textsuperscript{[31]} The thickness of the wall of cyst varies from a few millimeters to 1cm. it consists mainly of dense, collagenous connective tissue with occasional elastic fibers and smooth muscle bundles. An inflammatory reaction is often present. Generally, a complete epithelial lining is absent, but scattered islets of cylindrical or columnar epithelium may be found.\textsuperscript{[32]}

The only acceptable method of treatment of choledochal cyst is surgical.\textsuperscript{[33]} Conservative management is generally attended by high mortality. Similarly aspiration of the contents of the cyst as the sole method of treatment is condemned, as it fails to deal with the underlying abnormality and predisposes to bile leak with resultant biliary peritonitis. It has been suggested that cystoenterostomy, by exposing the mucosa of abnormal duct to pancreatic juice may hasten the development of cancer in choledochal cyst. Thus the drainage
procedure may render the patient asymptomatic until cancer appears many years later. Despite near universal recognition of importance of excision, the specific surgical technique remains controversial, particularly with regard to proximal and distal extent of resection and conduit for reconstruction (hepaticojejunostomy vs hepaticojejunoduodenostomy vs appendicoduodenostomy). The appropriate treatment for benign type I and type IV cysts is resection of extra hepatic biliary tree and Roux-en-Y hepaticojejunostomy. This procedure removes the mucosa at highest risk of malignant degeneration. Resection should extend from bifurcation of lobar hepatic portion of the distal bile duct, taking care not to injure the pancreatic duct or the long common channel. Malignancy after surgical resection is rare (<1%). Type II cysts can be treated by simple cyst excision. The defect in the CBD should be closed transversely rather than longitudinally to avoid narrowing of the duct lumen. Cholecystectomy is recommended owing to the potential risk of malignancy. Type III cysts (choledococeal) can be managed in a minimally invasive manner or by means of an open transduodenal approach in the second or third portion of the duodenum. The pancreatic and bile ducts should be identified individually. The pancreatic duct is intubated with a small silastic tube before cyst excision. The mucosa of the bile duct and pancreatic duct is individually sutured to the duodenal mucosa with interrupted sutures. The need for additional sphincteroplasty must be assessed on an individual basis. A small temporary stent (5- or 8) for plastic tube left in the pancreatic duct is recommended to prevent postoperative pancreatitis. Type IVa and IVb cysts require complete resection of the extrahepatic biliary tree whenever possible. With respect to the intrahepatic ducts, surgery should be individualized depending on whether both lobes are affected, strictures and stones are present, cirrhosis has developed or whether an associated malignancy is localized or metastatic. The placement of large silastic transhepatic stents may facilitate postoperative stone extraction and prevent complications; these patients have the greatest likelihood of recurrent cholangitis, stricture formation and hepatolithiasis. The management of type V cysts (Caroli’s disease) is controversial.

MATERIALS AND METHODS

The study entitled “Clinicopathological Status of Choledochal Cyst” was conducted in the Department of Surgical Gastroenterology, Medical Gastroenterology, Radiodiagnosis and Pathology, Sher-I-Kashmir Institute Medical Sciences, Srinagar. The study was both retrospective and prospective in nature. Retrospectively, all those patients who had been admitted as cases of choledochal cyst during the last eight years were undertaken. The data was collected from their case files and analyzed as per proforma. Prospectively all patients admitted as choledochal cysts were included in the study. After a thorough history and examination, the investigations included all baseline like haemogram, liver function test (LFT), kidney function test (KFT), in addition to the specific investigations like USG, CECT, endoscopic retrograde cholangiography (ERCP), magnetic resonance cholangiopancreatography (MRCP), bile analysis for amylase and lipase levels and bile culture, for confirmation of diagnosis as per proforma. The operative findings included the indication and type of procedure was recorded as per the proforma. Follow-up was achieved by reviewing medical records on personal visits.

“Written informed consent was taken from all the patients. We used SPSS
software version 16 for the calculation of p-value in our study. SPSS software was used for statistical analysis in our study.”

**OBSERVATIONS AND RESULTS**

Out of total 60 patients, children up to 15 years of age were 20 (33%) and adults 40 (67%). Majority of patients in our study were females (82%) with a male: female ratio of (1:4.5). The most common feature in both groups was pain (77–80%), followed by jaundice. Abdominal mass was present in 35% of children and 15% adults (p<0.05 statistically significant). Triad of pain, jaundice and mass was seen more in children than in adults (20% children Vs 12.5% adults). Abdominal distension was more in children. Two adult patients were incidentally diagnosed as choledochal cysts. Some patients had presented with complications before surgery for choledochal cyst, these include cholangitis, pancreatitis, rupture, ascites and cholecystitis, all these were more common in adults except biliary peritonitis and ascites which were exclusively seen in children. No patient in our study had malignancy. Previous history of cholecystectomy was present in 10% of adult patients, Choledoco-duodenostomy in 5% of children and adults each. History of CBD exploration was present in four adult patients. 43 (71%) patients had type I, 15 (26%) type IV, 2 (3%) had type V choledochal cyst. No patient had type II and III choledochal cysts. In children 80% were type I and 20% type IV choledochal cysts. Where as in adults 68% were type I, 27% type IV. Type V choledochal cyst was found in adults only (5%). No significant difference between the groups was seen with respect to serum bilirubin, SGOT and SGPT. Serum amylase was raised in 28% children and 19% adults. However levels of ALP were significantly higher in children (50% Vs 27%) (p<0.05).

30/60 (50%) Patients with choledochal cyst were complicated by stone formation in the liver, cyst itself and in gall bladder. Four adult patients had stone in the liver.

One child and nine adult patients had stone in the cyst itself. Gall bladder calculi were present in 14/40 (35%) adults and only in two children (p<0.05). Abnormal pancreato-biliary duct union was detected in 5/20 (25%) children and 12/40 (30%) adults. One child and 9 (22%) adults had strictures present at lower end of CBD, pancreatic duct or hepatic ducts. Bile analysis for amylase was done in 39 patients. 8/39 (20%) patients had raised bile amylase (28% children and 16% adults). One child and one adult patient had raised bile lipase. Only one patient (16%) among the children had raised CEA level (>10ng/ml) Vs adults in which 5/20(25%) had raised CEA level. Around 80% of bile cultures were sterile. The main organisms responsible for infection were Klebsiella pneumoniae and E. coli. 75% of children had grade 0-1 (mild) glandular hyperplasia where as 92% of adults had moderate to severe hyperplasia. 60% of children had infiltration of inflammatory cells from grade 0-1 while (77%) of adults had moderate to severe infiltration. Most of children (75%) had moderate to severe fibrosis compared to only 20% of adults with (grade3-4) fibrosis. Ultrasonography was done in all patients. 54/60 patients were diagnosed by USG. Therefore it had diagnostic accuracy of 90%. ERCP was done in 22 patients and in 20 patients it had picked up choledochal cyst. MRCP was done in 47 patients with a diagnostic accuracy of 100%. CECT was done in 16 patients and among these 15 were correctly diagnosed by CT. The standard operative procedure used in our patients was complete resection of extrahepatic biliary duct with cholecystectomy and Roux-en-Y hepaticojejunostomy, done in 55 patients.
One patient had cyst rupture therefore T-Tube drainage followed by definitive procedure was done. Lillýs technique was used in one pediatric patient due to dense inflammatory adhesions. Two patients with type IVa cyst had resection of left hepatic segment as well. Most of patients had uneventful course in the postoperative period. Incidence of bile leak, wound infection and ascending cholangitis in children was 5% each while it was 5%, 7% and 2.5% in adults respectively.

Table 1: Demographic profile of studied subjects

<table>
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<tr>
<th>Age</th>
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</thead>
<tbody>
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<td>0-15 year.</td>
<td>20</td>
<td>33%</td>
</tr>
<tr>
<td>&gt;15 year.</td>
<td>40</td>
<td>67%</td>
</tr>
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<thead>
<tr>
<th>Gender</th>
<th>number</th>
<th>percent</th>
</tr>
</thead>
<tbody>
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<td>Male</td>
<td>11</td>
<td>18 (%)</td>
</tr>
<tr>
<td>Female</td>
<td>49</td>
<td>82%</td>
</tr>
</tbody>
</table>

<table>
<thead>
<tr>
<th>District</th>
<th>number</th>
<th>percent</th>
</tr>
</thead>
<tbody>
<tr>
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<td>14</td>
<td>24%</td>
</tr>
<tr>
<td>Baramulla</td>
<td>13</td>
<td>22%</td>
</tr>
<tr>
<td>Budgam</td>
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<td>6%</td>
</tr>
<tr>
<td>Kupwara</td>
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<td>15%</td>
</tr>
<tr>
<td>Pulwama</td>
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<td>13%</td>
</tr>
<tr>
<td>Anantnag</td>
<td>9</td>
<td>15%</td>
</tr>
<tr>
<td>Leh</td>
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<td>3%</td>
</tr>
<tr>
<td>Banihal</td>
<td>1</td>
<td>2%</td>
</tr>
</tbody>
</table>

### DISCUSSION

Described initially in 1852 congenital choledochal cysts were separated into three types by Alanzo-Lej et al in 1959. Todani and colleagues modified this classification in 1977 adding types IV and V. The present study emphasizes on the clinicopathological presentation, complications and management of choledochal cysts. Most patients (75%) with choledochal cysts are diagnosed during the first decade of life, with only 20% going undiagnosed until adulthood. However with improved hepatobiliary imaging, adult patients with the disease are being increasingly seen. However the belief that choledochal cysts present mainly in childhood still persists. Usually pediatric patients are concentrated in pediatric surgical centers, whereas adult cases present to various general hospitals. Adult cases are therefore under-reported and revives of literature have probably overestimated the presentation in childhood. This trend might...
reflect an institutional referral bias, but series reporting both children and adults seem to suggest that the increased incidence in adults is real. In our series of 60 patients, 40 (66.3%) were adults. In our study the proportion of female patients was higher (1:4.5). Almost all other studies are showing similar picture. The presentation is often vague and non-specific impending prompt diagnosis. However the diagnosis is facilitated by modern imaging techniques and may be made at any time from antenatal period to adulthood. The classical triad of presentation of choledochal cyst with jaundice, pain and swelling is now known to apply to only minority of patients. [36] In our series this triad was present only in 20% of pediatric patients and 12% of adult patients. Similar reports of triad, seen more in children have been reported by other studies as well. [37] The most common presenting symptom in our patients was abdominal pain (77% -80%), this was followed by vomiting (60% children & 62% adults) and jaundice (35% each). Similar figures were reported by Iliker et al, [38] (45%) in his study. There appears to be a justification for treating the disease in children and adults as separate entities. The classic triad of jaundice, abdominal mass, and pain, originally described as a feature in most patients, was rarely seen in adults. The incidence of abdominal distension and mass was seen higher in children than in adults. However two adult patients in our study had incidental diagnosis of choledochal cyst as part of evaluation of other disease. Shi et al had also reported that adult patients tend to have non-specific symptoms resulting in delayed diagnosis. In 15% of children acholic stools were present compared to 5% patients only. This included one, two month old baby in which acholic stools and jaundice was the only presentation. Ujjal et al, [39] had similarly reported the high incidence of cholestasis in children. Close differential diagnosis is extra hepatic biliary atresia. These patients need urgent treatment to prevent irreversible cirrhosis of liver. As far as the preoperative complications are concerned, rupture (10%) and ascites (15%) were seen in only children. The cause of rupture is unknown; under perfusion of an inflamed dilated duct and mural weakness caused by reflux of pancreatic juice are two possible factors. Trauma may also be one of the causes for rupture. Pancreatitis and cholecystitis were also seen more in adults compared to children (22% Vs 5%) and (15% Vs 0%) respectively. Similar reports of high in incidence of pancreatitis and cholangitis were seen by Shah et al. Activation of pancreatic enzymes by bile reflux in association with APBDJU or stone obstruction at the common channel is thought to be involved in the pathogenesis of pancreatitis in such patients. The previous history of cholecystectomy and CBD exploration was seen only in adults. [40] 28% children and 16.5% adults in our study had raised biliary amylase levels. The average amylase level in our pediatric patients was 8500 IU/L and in adult patients 7000 IU/L. Similar reports of biliary amylase being more in children were published by Jhong et al. [41] Some studies report that 57% of the patients with choledochal cyst and pancreatitis have abnormalities in the biliopancreatic junction. [42] The previous history of cholecystectomy and CBD exploration was seen only in adults. The types of choledochal cysts seen in our patients were type-I 70 (most common), type-IV (25%). Type I & III were not seen in any patient. Similar figures have been shown by other studies as well. Cystolithiasis and gallstones are a frequent accompanying condition occurring in over 70% of adults with bile duct cyst. [43] Choledochal cysts in our study were complicated by stone formation in the liver, cyst itself, or gall bladder in 30 patients.
67% of adults had stone formation compared to 15% of children only (p<0.05), similarly Shah et al had reported high incidence of stone formation (45%) in their series. Choledochal cyst is a premalignant condition and cholangiocarcinoma develops in and around 20% of patients. Carcinoembryonic antigen is a tumor marker and levels are significantly raised in gut malignancy including cholangiocarcinoma. [44] The levels of carcinoembryonic antigen were monitored in 26/60 patients in our study. 26% of adult patients in our study had raised CEA levels (>10ng/ml) compared to only one patient in pediatric age group. This finding may be an indirect evidence of progression towards dysplastic changes in choledochal cyst, although in the follow up period of 18 months no patient developed cholangiocarcinoma. cyst may be responsible for the fibrosis. In 1959 preoperative diagnosis of choledochal cyst was possible in only 30% of cases but now it is possible in almost all of cases with the help of imaging studies. Ultrasonography is a useful screening test and is considered to be the first investigation for diagnosis of choledochal cyst. It is safe and noninvasive investigation with a high degree of reliability. However other investigations like MRCP, CECT, cholangiopancreatography and ERCP are required to demonstrate anatomy and coexistent pathology such as cholilithiasis, cystolithiasis, and post operative biliary stricture with greater clarity. [45] In the presence of cystolithiasis these cysts can be mistaken for simple cholidocolithiasis. Ultrasound can diagnose the choledochal cysts with high specificity although this drops if the width criteria are reduced to increase sensitivity, with normal variants and secondary causes of biliary dilatation can be misdiagnosed as choledochal cysts. We had compared the diagnostic accuracy of various investigations like USG, ERCP, MRCP and CECT. Ultrasound had accurately diagnosed choledochal cyst in 54 (90%) patients. Iliker also supports the use of ultrasound with 70% accuracy. The diagnostic accuracy of ERCP for exactly defining the anatomy of choledochal cyst was 91% and that of CECT 94% in our study. However MRCP was done in 47 patients and could define the exact anatomy in all of them, favoring the concept of MRCP being the gold standard for diagnosis of choledochal cyst. Kobayashi et al [46] pointed out that the incidence of bile duct carcinoma is still high, even after excision of extrahepatic bile ducts in APBDJU patients. Vigilant long-term follow-up is therefore advocated especially in patients with type IVa cysts. Therefore cystic type choledochus should be excised during childhood before it develops into a precancerous stage. In spindle-like and nondilated choledochus, cholecystectomy is mandatory before the age of 40 years. No patient in our study group developed cholangiocarcinoma as a complication. The reasons may be early diagnosis, avoiding drainage procedures like cystoduodenostomy which constantly exposes the mucosa to carcinogenic potentials. One of the reasons may also be the short follow up period of 18 months only and therefore long term follow up of our patients is needed. Surgical management of patients with choledochal cyst is complex. The clinical symptoms often do not reflect accurately the underlying hepatobiliary disease. Pancreatobiliary ductal abnormalities, pancreatitis, and malignancy are frequently associated with choledochal cysts and are major factors affecting outcome. The variety and extent of associated disease, cyst type, and prior cyst drainage procedures also influenced the operative approach to these patients. 55 patients in our study underwent cyst excision with cholecystectomy with hepaticojejunostomy. Complete excision of
type I and II cysts is widely accepted, whereas sphincteroplasty endoscopically or surgically is recommended for type III cysts. For type IV cysts involving both intra- and extrahepatic ducts, resection of the extrahepatic component with Roux-en-Y hepaticojejunostomy at the hepatic hilum is the mainstay of treatment. In cases in which the intrahepatic disease is limited to one lobe, hepatic resection can be considered. Two patients in our study with type-IVa cysts were treated with resection of hepatic segment as well. Type V cysts involving a segment or one lobe of the liver are curable by resection. The diffuse form is associated with a grave prognosis. Nevertheless, liver transplantation appears to be an effective option. The presence of extensive fibrosis, cholangitis, biliary cirrhosis, or portal hypertension and a history of previous biliary surgery make cyst removal technically challenging in some adult patients. The application of Lilly’s technique, in which the cyst is completely excised leaving intact the muscular and posterior fibrous wall, proves useful in such circumstances. Lilly’s technique was used in one pediatric patient in our study. In children, the fate of the bilioenteric anastomosis may be decided by the skill of the operating surgeon. Common hepatic ducts are larger in adults, so anastomosis may be performed relatively easily and safely; cyst excision, however, is comparatively difficult owing to the presence of severe pericystic inflammation. The absence of a normal epithelial lining in most adults prevents a mucosa-to-mucosa anastomosis with the intestine; subsequent anastomotic stricture development is therefore not infrequent. Three of our adult patients developed anastomotic strictures during follow-up. To prevent this complication, most investigators recommend a high-level anastomosis, beyond the relative stenosis in the CBD or a plastic operation at the junction of the intrahepatic ducts with the CBD. Achieving wide, patent anastomosis should be the main goal. In case the dilatation extends to include the hepatic ducts, it is advisable to carry out the transection at the junction of the right and left hepatic ducts and perform the anastomosis at this level. Nevertheless, the possibility of recurrent strictures and intrahepatic stone formation with malignant change cannot be excluded. Careful long-term follow-up is therefore mandatory.

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Abbreviations:
CDC - Choledochal cyst
USG – Ultrasonography
CECT – Contrast Enhanced Computed Tomography
MRCP- Magnetic Resonance Cholangiopancreatography
ERCP- Endoscopic Retrograde Cholangiopancreatography

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