Choledochal Cyst: Experience from A Tertiary Level Hospital of Bangladesh

Adnan Bacha1*   Arif Salam Khan2   Arup Kumar Biswas3   Zulfikar Rahman Khan4

Key words: Caroli’s disease; Choledochal cyst; ERCP; Hepaticojejunostomy; MRCP.

Introduction
Choledochal cyst is the cystic dilatation of intra-hepatic or extra-hepatic or both parts of the biliary tract. It is a rare congenital but not familial anomaly and may occur singly or in multiple numbers. The incidence is higher in Asian population 1:1000 compared to 1:100,000 to 1:150,000 in western population1,2. Although congenital, symptoms appear in the later part of life. Choledochal cysts also have an unexplained female: male predominance, commonly reported as 4:1 or 3:12,3. In 1977, Todani classified choledochal cyst into five types and subtypes (Figure 1).

The most common (50 -80%) is Type I with diffuse or segmental fusiform dilatation of the extra-hepatic biliary channels of uncertain origin. It may clinically manifest at any age. Biochemical tests are not diagnostic but imaging modalities help in accurate diagnosis. Surgery in the form of complete excision of the cyst with diversion is the treatment of choice and decreases the risk of dreaded complication of cholangiocarcinoma. The aim of this study is to demonstrate the sex distribution, symptomology, biochemical presentation, radiological findings and outcome of surgery in Bangladeshi population.

Methods and materials: A total of 12 cases were considered for this study which included all the cases that were admitted at Bangabandhu Sheikh Mujib Medical University (BSMMU) during June 2013 to May 2014. The study also covered the incidence of the type of the cyst, post-operative complications and follow up of the patient for 5 years at least.

Results: 9 of the patients were female and the median age of presentation was 17. Todani type I was the most common type. Patients attended hospital with upper abdominal pain (100%) alongwith other symptoms like vomiting, jaundice, etc. S. bilirubin and alkaline phosphatase were raised in most of the patients. Radiological diagnosis was done effectively by ultrasonography, CT (Computer Tomography) scan and MRCP (Megnatic Resonance Cholangiopancreatography). All the patients underwent complete excision of the Choledochal cyst with cholecystectomy with Roux en Y hepaticojejunostomy and post-operative complications include wound infection (n=2) scar pain and recurrent cholangitis (16.67% each).

Conclusion: Surgery is the treatment of choice and decreases the risk of cholangiocarcinoma. Long term follow is required in these patients.
Etiology of choledochal cyst is still unclear, although many theories have been put forth. The most commonly proposed theory is Babbitt’s theory, where choledochal cysts are supposed to be caused by an Abnormal Pancreatico-Biliary Duct Junction (APBDJ)\(^6\). Obstruction of the distal CBD is another theory which is supported by studies on animal models\(^5\). Some authors also suggest pure congenital theory in which normally a few ganglionic cells are seen in the distal CBD in patients with choledochal cyst resulting in proximal dilatation in the same manner as achalasia of oesophagus and Hirschsprung’s disease\(^6\).

Choledochal cysts most commonly present in childhood and about 25% patients present in adulthood. The classical triad of symptoms, which include abdominal pain, palpable abdominal mass and jaundice, are seen in less than 20% of cases. About 85% of children have at least 2 features of classic triad, whereas only 25% of adults present with 2 features of the classic triad\(^10\). Improved imaging techniques have made diagnosis of choledochal cyst possible, and can be diagnosed even antenatally. Abdominal ultrasonography is the first imaging modality generally used and is considered as the test of choice for the diagnosis of choledochal cyst\(^11\). Other useful imaging modalities include CT (Computer Tomography) scan, MRI (Magnetic Resonance Imaging) of the abdomen and Magnetic Resonance Cholangiopancreatography (MRCP). MRCP is regarded as the “gold standard” for the diagnosis of choledochal cyst with sensitivity for diagnosis as high as \(90\%-100\%)\(^12\) (Figure 2).

**Figure 2** CT scan picture showing multiple intrahepatic cystic dilations and dilatation of the CBD (Carol’s disease) & MRCP image showing fusiform dilation of the CBD (Type 1)

Cholangiocarcinoma is the most feared risk, but early diagnosis and appropriate surgical intervention with complete removal of the cyst appears to decrease the risk of development of malignancy. Perforation is another complication of the choledochal cyst and might mimic perforation of hollow viscus\(^13\). Management of Choledochal cyst has undergone a lot of development in past years. Mcwhorter first described cyst excision and hepaticojejunostomy in 1924 but was soon abandoned because of multiple complications. Other strategies that evolved with time include cyst marsupialization with choledochorraphy and cystenterostomy, which where once popular procedures for the management of the disease. High risk of malignancy in the cyst helped regain excision of the cyst with Roux en Y hepaticojejunostomy to become the surgery of choice for choledochal cysts\(^14\). Laparoscopic resection of Choledochal cyst has been widely used recently, but in spite of advantages like less bleeding, shorter fasting time and shorter hospital stay pose disadvantages like technical differences and frequently inevitable complications\(^2\).

**Methods and materials**

A prospective study was carried out at Bangabandhu Sheikh Mujib Medical University (BSMMU) Dhaka, Bangladesh from May 2013 to June 2014. A total of 12 patients were admitted for choledochal cyst and evaluated.

Patients were included if they had been diagnosed with choledochal cyst and underwent surgery, but the patient underwent drainage procedure where not considered for the study. Statistical analyses were performed with Statistical Package for Social Sciences (SPSS 19.0 software).

**Results**

A total of twelve (12) patients were treated for choledochal cyst, of whom 9 (75%) were female with a male female ratio of 1:3 (Figure 3).

**Figure 3** Pyramidal graph representing sex and age distribution of the Choledochal cyst

The mean age of presentation was 16, and median 17. The presenting complaints varied, however all the patients presented with intra-abdominal pain (100%) while half of them presented with cholangitis and half with jaundice. Vomiting was second most common presentation (83.33%). The symptoms are summarized in Figure 4.
In our case series we have considered Ultrasonography (USG) Computer Tomography (CT) and Magnetic Resonance Cholangio-Pancreaticography (MRCP) for all the cases in order to come to a pre-operative diagnosis, classification and hence planning of the surgery. All the three imaging modalities were seen to be effectively able to diagnose and classify Choledochal cyst. Out of 12 cases, 8 (66.67%) were type I, 3 (25%) were type IV and 1 (8.3%) were diagnosed as type II choledochal cyst (Figure 5).

Strikingly it was seen that in all the 12 (100%) cases, some sort of stone disease in the form of choliolithiasis or choledocholithiasis or hepaticolithiasis or combination of two or all were present.

Our approach was through a right subcostal or a roof top incision. Complete excision of the cyst with cholecystectomy and Roux-en Y hepaticojejunostomy was done in all 12 cases (100%). Irrigation and choledoscopic extraction of the stones was done where ever required. Specimen were sent for histopathological examination which revealed chronic cholecystitis and features compatible with choledochal cyst (n=12). None of the specimen was positive for malignancy.

The early post-operative complication was that only two patients developed wound infection, which was managed by regular dressing, broad spectrum antibiotics based on culture sensitivity report and later secondary closure of the wound. The median duration of the hospital stay was 12 days (Range 6 to 36 days). No post-operative mortality was seen within 30 days of follow up. Planned follow up at the interval of 1 month, 6 months, 12 months, 3 years and 5 years was done using tools like physical examination, ultrasonography of whole abdomen and CA19-9. During follow up 2 (16.67%) patients complained of mild scar pain treated effectively by mild to moderate analgesics while as another 2 patients had features of recurrent cholangitis, both of them were treated conservatively by antibiotics. Ultrasonography revealed retained stone in one patient. No evidence of malignancy or anastomotic stricture was seen on follow up.

Figure 4 Presenting symptoms of the patients with Choledochal cyst

Laboratory findings demonstrated neutrophillic leucocytosis in 4 patients (33.33%). Liver function tests were done in which SGPT was elevated in 33.33% (n=4) and SGOT in 8.33% (n=1), while alkaline phosphatase was increased in 66.67% (n=8). S. Albumin was less in (50%) 6 patients but was unexpectedly more in one patient. Serum bilirubin was increased in 11 patients (91.67%). Tumor marker was done to correlate with malignancy, it was normal in all the cases except 1 (8.33%) which too was not significantly high. Prothrombin time and INR was seen to be normal in all the cases. Findings are summarized in Table I.

Table I Biochemical results of patients presenting with Choledochal cyst

<table>
<thead>
<tr>
<th>S. No.</th>
<th>Blood Test</th>
<th>Normal</th>
<th>Levels</th>
<th>Increased</th>
<th>Decreased</th>
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<td>Leucocytosis</td>
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<td>4</td>
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<tr>
<td></td>
<td></td>
<td></td>
<td>Neutrophilia</td>
<td>8</td>
<td>4</td>
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<tr>
<td>2.</td>
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<td></td>
<td>SGPT</td>
<td>8</td>
<td>4</td>
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<td></td>
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<td>SGOT</td>
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<td>1</td>
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<td></td>
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<td>S. Bilirubin</td>
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</tr>
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<td>3.</td>
<td>CA19-9</td>
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<td>11</td>
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<td>-</td>
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<td>4.</td>
<td>PT/INR</td>
<td></td>
<td>12</td>
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Figure 5 Pie chart showing frequency of different Types of Choledochal cysts in Bangladeshi population
Discussion
Choleodochal cyst is an abnormal dilatation of the biliary channel. Worldwide incidence is less than 1:13000 persons but it appears to be a little commoner in Japan with an incidence of about 1:200. No such data is available for Bangladesh. This is much common in female 4:1, which is somewhat similar to the result of our study where female ratio was 3:1.(2, 3) It can affect any age, but is common in children and younger age group. In this study (n=7) the age group was between 10 -14 years. Etiology is still unclear, although many theories have been put forth. The most commonly proposed is the Babbit’s theory of long common channel. The classical triad of symptoms jaundice, abdominal pain and abdominal mass is not always present, however two of the symptoms are present in most of the patients. The most common presentation is intra-abdominal pain. In our study all the patients presented with intra-abdominal pain while none presented with intra-abdominal mass. Type I choleodochal cyst fusiform dilatation of the extra-hepatic biliary channels is the most common variety of the disease (60-80%). Which is similar to our study where 8 (66.67%) patient were diagnosed as Type I disease. Although ultrasonography can detect the cyst in almost all the cases, MRCP and CT Scan should be done to diagnose any intra-hepatic cyst or dilatation and for pre-operative planning for surgery. Relationship between the stone disease jaundice, abdominal pain and abdominal mass is not always present, however two of the symptoms are present in most of the patients. The most common presentation is intra-abdominal pain. In our study all the patients presented with intra-abdominal pain while none presented with intra-abdominal mass. Type I choleodochal cyst fusiform dilatation of the extra-hepatic biliary channels is the most common variety of the disease (60-80%). Which is similar to our study where 8 (66.67%) patient were diagnosed as Type I disease. Although ultrasonography can detect the cyst in almost all the cases, MRCP and CT Scan should be done to diagnose any intra-hepatic cyst or dilatation and for pre-operative planning for surgery. Relationship between the stone disease jaundice, abdominal pain and abdominal mass is not always present, however two of the symptoms are present in most of the patients. The most common presentation is intra-abdominal pain. In our study all the patients presented with intra-abdominal pain while none presented with intra-abdominal mass. Type I choleodochal cyst fusiform dilatation of the extra-hepatic biliary channels is the most common variety of the disease (60-80%). Which is similar to our study where 8 (66.67%) patient were diagnosed as Type I disease. Although ultrasonography can detect the cyst in almost all the cases, MRCP and CT Scan should be done to diagnose any intra-hepatic cyst or dilatation and for pre-operative planning for surgery. Relationship between the stone disease jaundice, abdominal pain and abdominal mass is not always present, however two of the symptoms are present in most of the patients. The most common presentation is intra-abdominal pain. In our study all the patients presented with intra-abdominal pain while none presented with intra-abdominal mass. Type I choleodochal cyst fusiform dilatation of the extra-hepatic biliary channels is the most common variety of the disease (60-80%). Which is similar to our study where 8 (66.67%) patient were diagnosed as Type I disease. Although ultrasonography can detect the cyst in almost all the cases, MRCP and CT Scan should be done to diagnose any intra-hepatic cyst or dilatation and for pre-operative planning for surgery.

Conclusion
Although rare, choleodochal cyst remains an interesting and somewhat challenging clinical problem because of its late and non-specific presentation. Hence it should be considered as a differential diagnosis for obstructive jaundice. Biochemical markers are non-specific but imaging may lead to specific diagnosis and pre-operative planning of surgery. Complete excision of the cyst with Roux-en Y hepaticojejunostomy is the treatment of choice. Specimen should be sent for histopathological examination to exclude malignancy which may warrant further investigations and treatment.

Recommendations
Further long time follow up is required, to prove that complete excision of the cyst removes all the risk of development of cholangiocarcinoma.

Disclosure
All the authors declared no conflict of interest.

References


