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**Diagnosis and management of choledochal cyst: 20 years of single center experience**

Gadelhak N *et al.* Choledochal cyst: Diagnosis and management

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**Abstract**

We introduce the first case series report from Africa and Middle East on choledochal cyst, a disease which shows significant geographical distribution with high incidence in Asian population. In this study, the epidemiological data of the patients were presented and analyzed. Much focus was paid to diagnostic imaging and its accuracy in diagnosis and classification of choledochal cyst. Most cases of choledochal cyst disease have type I and IVA cysts according to todani classification, that support etiological theories of the choledochal cyst, especially Babbitt’s theory of the anomalous pancreaticobiliary duct junction, were clearly stated. Difficulties and hazards of surgical management and methods to avoid operative complications were clarified. Early and late postoperative complications were also noted. This study needs to be followed by multicenter studies all over Egypt to help in assessment of the incidence of choledochal cysts in one of the largest populations in Africa and Middle East.

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**Key words**: Choledochal cyst; Hepatic cyst; Hepaticojejunostomy; Caroli disease; Hepatectomy

**Core tip:** This research reported in this manuscript presents 20 years of single Egyptian center experience in high volume center experience with 50 cases of choledochal cyst. This is the first case series report to come from Africa and Middle East.

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**INTRODUCTION**

Choledochal cysts are disproportionate dilatations of the biliary system[1]. Its incidence shows significant geographic variations, being higher in Asian population reaching up to 1 in 1000[2]. Complete excision of the cyst is the best treatment strategy to avoid long term complications especially malignant transformation, recurrent cholangitis and gall stones[2,3]. To our knowledge, there are no studies on choledochal cyst coming from Africa or Middle East region to assess the local prevalence of the disease. In this study, we introduce 20 years of single Egyptian tertiary center experience with 50 cases of choledochal cyst with highlights on the etiological, clinical and surgical implications according to the findings in this case series report.

**CASE REPORT**

This is a retrospective study on all patients admitted to Mansoura Gastrointestinal Surgical Center in the period from January 1991 to November 2012. Data for this study were retrieved from the internal web-based Ibn Sina registry system supplemented by paper-based records. Data were collected and rearranged in a standardized manner. Choledochal cysts were classified according to the Todani modification of the Alonoso-Lej classification[4]. Early and late complications were noted.

Shapiro-Wilk test is used to assess normality of data. Numerical data are presented as means and standard deviations or as medians with ranges. A *P* < 0.05 was considered statistically significant. Statistical analysis was done with the help of IBM SPSS v20.

A total number of 50 patients (39 females, 11 males, ratio 3.5:1) were admitted to our center during the study period. Data of 2 female patients were lost from the medical records and one female patient refused to undergo surgery. The mean age of presentation was 265 ± 207.7 mo ranging from 3 mo to 65 years. Right hypochondrial pain was the most common presenting symptom (*n* = 45%-93.8%) followed by jaundice (*n* = 28%-58.3%), vomiting (*n* = 23%-47.9%), recurrent fever (*n* = 21%-43.8%) and abdominal mass (*n* = 4%-8.3%). The classic triad of abdominal pain, jaundice and palpable right upper quadrant mass was identified in one patient.

Five patients underwent previous biliary surgery during which choledochal cysts were not detected. Three of the five cases underwent cholecystectomy, and two cases underwent exploration for abdominal cysts that were not operated upon. Moreover, one case was explored for acute abdomen that was mostly attributed to perforated duodenal ulcer but the exploration was negative and the patient improved under conservative treatment. Choledochal cysts were associated with congenital anomalies in 5 cases (10.4%); ventricular septal defect (one case), medullary sponge kidney (one case), multiple bilateral renal cortical cysts (one case), congenital megacolon (one case), and intestinal malrotation (one case).

In our series, abdominal ultrasound (US) was done for 38 cases; diagnosed 19 cases (50%) and accurately classified the cyst type in 11 cases (28.8%) (Figure 1). Magnetic resonance cholangiopancreatography (MRCP) was done for 41 cases; diagnosed 38 cases (92.7%) and accurately classified the cyst type in 36 cases (87.8%) (Figure 2). Anomalous pancreaticobiliary duct junction (APBDJ) was detected by preoperative cholangiography in 6 cases (14.6%), 5 cases by MRCP and one case by percutaneous transhepatic cholangiography (Figure 3). According to the Todani modification of the Alonso-Lej classification, we identified patients with type Ia (*n* = 29%-60.4%), type Ib (*n* = 2%-4.2%), type Ic (*n* = 4%-8.3%), type IV-A (*n* = 12%-25%) and type V (*n* = 1%-2.1%) choledochal cyst. Table 1 shows comparison between the results of our study and other studies from countries in South East Asia regarding patient demographic data, clinical presentation and cyst classification.

Thirty eight patients underwent cyst excision and hepatico-jejunostomy Roux-en-Y (Figure 4), one case underwent pancreaticoduodenectomy due to intrapancreatic extension of the cyst, 3 cases of type IV-A underwent left hepatectomy, extrahepatic biliary resection and right hepatico-jejunostomy Roux-en-Y. Five cases underwent internal drainage procedures via cysto-duodenostomy in 3 cases and cysto-jejunostomy in 2 cases. Of the surgical cases, a mass was detected in the cyst wall in one case and its malignant nature was confirmed by intraoperative frozen section.

Early postoperative complications included postoperative wound disruption (*n* = 1) that was managed surgically; collections (*n* = 4) 3 managed conservatively, and 1 with ultrasound guided tube drainage; biliary leakage (*n* = 3) that was managed conservatively, pancreatic leakage (*n* = 1) that was managed conservatively, internal hemorrhage on top of acute hemorrhagic pancreatitis that was managed surgically (*n* = 1), and air embolism (*n* = 1). Overall early complication rate is 23.4%. There is no early postoperative mortality.

The median follow-up period was 55 ± 38.3 mo mo (mean ± SE). Late postoperative complications included intrahepatic ducts stones (*n* = 2 -4.3%), anastomotic stricture (*n* = 1%-2.1%), liver abscess (*n* = 2%-4.3%) and hepatic malignancy (*n* = 1%-2.1%). Overall late complication rate was 12.8%. There were 2 late postoperative mortalities. One of them died 3 years after surgery due to bilobar liver abscesses. The other, with confirmed malignant transformation by intraoperative frozen section, died 7 mo after surgery with recurrent tumor in segment IV of liver.

**DISCUSSION**

In our experience, most cases of choledochal cyst (64.6%) were diagnosed after the first decade of life. This increased incidence in adults may be attributed to institutional referral bias. However increased incidence in adults is reported by many case series reports of both children and adults[5,9]. This increase is justified according to some authors by the advance in the hepatobiliary imaging techniques[7]. Possibility of choledochal cyst should be kept in mind during surgical exploration in all patients with biliary tract related symptoms. In our series, 5 cases (10.4%) underwent previous biliary surgery and choledochal cysts went unnoticed during the surgery.

The most accepted theory in explaining the pathogenesis of choledochal cyst is Babbitt’s theory of the APBDJ precluding normal sphincter development at the PBDJ. This anomalous junction leads to reflux of pancreatic secretions into the common bile duct because of smaller diameter and higher pressure of the pancreatic duct. This theory is supported by radiological detection of APBDJ or by high level of amylase in the cyst fluid[10]. In our series, APBDJ was detected in 6 cases (14.6%) (Figure 3) but unfortunately cyst fluid amylase was not routinely done in our center. Also, presence of five cases of choledochal cyst (10.4%) with associated congenital anomalies supports other etiological theories of congenital background[11]. These associations give rise to the necessity of thorough evaluation of patients with choledochal cysts to exclude associated congenital diseases for safe surgical and anaesthetic considerations[12].

The so-called classic triad of intermittent jaundice, abdominal mass, and pain was found in a minority of cases according to most case series reports[13]. The most frequently seen presentation was abdominal pain (93.8%) which is a nonspecific symptom and usually associated with a relatively late diagnosis. On the other hand, jaundice was the second most common presentation (58.3%), and it was usually associated with early diagnosis. These clinical findings were corroborated by other studies[6,14].

For any case with biliary symptoms, abdominal ultrasound scan is the initial imaging modality of choice. Precise and accurate delineation of the biliary system mandates cholangiography with the advantage of non-invasive MRCP over endoscopic retrograde cholangiopancreatography [1,11]. In our experience, Abdominal US diagnosed 19 cases (50%) and accurately classified the cyst type in 11 cases (28.8%), while MRCP diagnosed 38 cases (92.7%) and accurately classified the cyst type in 36 cases (87.8%). These findings cope with other studies[2,14,15].

Based on the Todani modification of the Alonso-Lej classification, we found 35 cases of type I (72.9%), and 12 cases of type IVA (25%) making a total of 97.9% of our experience with choledochal cysts. This supports the criticism of this standard classification scheme describing it to be misleading, purposeless and thus it should be abandoned to reserve the term congenital choledochal cyst for congenital extrahepatic or intrahepatic biliary duct dilatation apart from Caroli´s disease, choledochocele and diverticulum of the common bile duct[16].

Consensus is established that the best treatment of choledochal cyst is surgical excision whenever possible. This helps to avoid long term complications of the cyst including pancreatitis, cholangitis, choledocholithiasis, biliary cirrhosis and malignant transformation. To achieve complete cyst excision, accurate recognition of the begining and the termination of the cyst is mandatory[17].

Dissection towards the lower end of the cyst may require pancreaticodoudenectomy in favor of complete cyst excision; otherwise inevitable unplanned pancreatic duct injury would occur. In our experience, one case was complicated by postoperative pancreatic leakage that was managed conservatively, one case was planned for pancreaticodoudenectomy, and one case had an accidental pancreatic duct injury that was managed by pancreaticoduodenostomy over external stent but later on this case was readmitted by acute hemorrhagic pancreatitis and was explored and managed by external pancreatic duct tube reposition. Diao *et al*[18] performed choledochal cyst excision without ligation of the distal stenotic stump in 207 patients and there was no significant difference in the results in comparison to the ligated group of patients. This technique helped to decrease the incidence of pancreatic injury which is reported to be 2%-6% in previous reports from China. A probe inserted into the pancreatic duct through a doudenotomy may help to avoid injury of the pancreatic duct in difficult cases[19].

On the other side, dissection towards the upper end of the cyst should be done considering measures to avoid postoperative anastomotic stricture. The best strategy is to resect at the level of the carina with left duct spatulation for obtaining wide stoma for the anastomosis. However, great therapeutic challenge is present with dilated biliary system above the carina. Complete excision in such cases may put the surgeon in the situation of performing two to four duct anastomoses with normal caliber ducts. Although all portions of choledochal cysts should be removed, residual proximal cyst walls may be left to facilitate biliary anastomosis[20]. Some cases may require hepatectomy or liver transplantation.

Limitations of this study were absence of institutional referral bias to our center making it difficult to rely on this study in comparing incidence of the disease in adult in comparison to children. Also, the absence of the emergency referral to our center omits the study of emergent presentation of the disease as spontaneous perforation and acute pancreatitis. However, this study needs to be followed by multicenter studies all over Egypt to help in assessment of the incidence of choledochal cysts in one of the largest populations in Africa and Middle East.

Choledochal cysts are disproportionate dilatations of the biliary system[1]. It can present at different age with variable biliary symptoms. Thus, a high sense of suspicion for the disease is required. Most cases of choledochal cyst disease have type I and IVA cysts. If left untreated, choledochal cysts have an increased risk of malignant transformation. Early surgical excision and restoration of biliary tract continuity is mandatory whatever the symptom severity to avoid long term complications whenever possible[20].

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**L-Editor E-Editor**

**Figure 1 Ultrasound imaging and** **abdominal computed tomography of type I choledochal cyst.** A: Ultrasound imaging; B: Abdominal CT. CC: Choledochal cyst; GB: Gall bladder; CBD: Common bile duct; CT: Computed tomography.

**Figure 2 Magnetic resonance cholangiopancreatography images of choledochal cyst.** A: Type IVA choledochal cyst with Anomalous pancreaticobiliary duct junction (white arrow); B: Type I choledochal cyst with multiple stones inside (white arrow). IHB: Intrahepatic biliary radicals; CHD: Common hepatic duct; GB: Gall bladder; CBD: common bile duct.

**Figure 3 Anomalous pancreaticobiliary duct junction detected by cholangiography.** A: Percutaneous transhepatic cholangiography of type IVA choledochal cyst; B: MRCP of type IVA choledochal cyst. MRCP: Magnetic resonance cholangiopancreatography.

**Figure 4 Surgical excision of choledochal cyst.** A: Exposure of type IA choledochal cyst; B: Division of biliary sytem at the carina proximal to the cyst; C: The biliary system after excision of the cyst; D, E: Restoration of biliary continuity by hepaticojejunostomy; F: The specimen after complete excision of the cyst consisting of gall bladder (GB) and choledochal cyst (CC).

**Table 1 comparison between our study and other studies from South East Asia**

|  |  |  |  |  |  |
| --- | --- | --- | --- | --- | --- |
|  | **Shi *et al*[5]**  **(2001)** | **She *et al*[6] (2009)** | **Shah *et al*[7] (2009)** | **Woon *et al*[8] (2006)** | **Our study**  **(2013)** |
| Country | China | Hong Kong | Kashmir | Singapore | Egypt |
| Total number | 108 | 83 | 79 | 32 | 50 |
| Sex  (F:M) | 85:23  Ratio 3.7:1 | 60:23  Ratio 2.6:1 | 67:22  Ratio 3:1 | 25:7  Ratio 3.5:1 | 39:11  Ratio 3.5:1 |
| Age  Mean  Range | 27.8 yr  3:68 yr | 45 mo  0 :16 yr | NR | 41 yr  18:74 yr | 265 d  3 mo:65 yr |
| Presentation  Pain  jaundice  Vomiting  Fever  Mass.  Triad  Incidental | (61%-56.5 %)  (77%-71.3%)  NO  (61%-56.5%)  NO  NO  NO | (39%-46.9%)  (35%-42.2%)  (26%-31.3%)  NO  NO  (2%-2.4%)  NO | (58%-73.4%)  (26%-32.9%)  NO  NO  (20%-25.3%)  (17%-21.5%)  (7%-8.9%) | (29%-91%)  (13%-41%)  (12%-38%)  (11%-34%)  (8%-25%)  (4%-13%)  (3%-9%) | (45%-93.8%)  (28%-58.3%)  (23%-47.9%)  (21%-43.8%)  (4%-8.3%)  (1-2%)  NO |
| Classification  I  II  III  IV  V  Unclassified | (75%-69.4%)  NO  (1%-0.9%)  (24%-22.2%)  (6%-5.6%)  (2%-1.8%) | (53%-67.9%)  (4%-5.1%)  (2%-2.6%)  (15%-19.2%)  (4%-5.1%)  (5%-6.1%) | (54%-68.3%)  NO  NO  (21%-26.6%)  (4%-5%)  NO | (27%-84.4%)  (2%-6.2%)  NO  (2%-6.2%)  (1%-3.1%)  NO | (35%-72.9%)  NO  NO  (12%-25%)  (1%-2.1%)  NO |
| APBDJ | NR | NR | (21%-26.6%) | NR | (6%-14.6%) |

NR: Not reported.