

The Clinical Features and Strategy for Diagnosis of Choledochal Cyst Using Improved Diagnostic Techniques

Dr. Muhtarima Haque^{1*}, Prof. Dr. Mohammad Ali²

¹MBBS, FCPS (Surgery), Dept. of Surgery, Birdem General Hospital Shahbag Dhaka, Bangladesh

²MBBS, FCPS, FRCS (Ed), FACS, Rtd. Head of Dept. of Hepato Biliary and Pancreatic Surgery and Liver Transplant, Birdem General Hospital Shahbag, Bangladesh

DOI: <https://doi.org/10.36348/sjimps.2024.v10i12.012>

| Received: 18.11.2024 | Accepted: 24.12.2024 | Published: 26.12.2024

*Corresponding author: Dr. Muhtarima Haque

MBBS, FCPS (Surgery), Dept. of Surgery, Birdem General Hospital Shahbag Dhaka, Bangladesh

Abstract

Background: Choledochal cysts are rare congenital anomalies of the biliary system, presenting variably across age groups. Early diagnosis is crucial to avoid complications such as cholangitis, pancreatitis, and malignancy. Advancements in diagnostic imaging have significantly improved the detection and management of this condition. **Objective:** To assess the clinical features and diagnostic strategies for choledochal cysts using improved diagnostic techniques. **Methodology:** A retrospective observational study was conducted in the Hepato-Biliary-Pancreatic Surgery (HBPS) department of BIRDEM Hospital, Dhaka, spanning five years from January 2003 to January 2008. Thirty patients diagnosed with choledochal cysts were included based on clinical history, examination, and imaging. Data analysis was performed using SPSS version 22, with descriptive statistics and chi-square tests employed to explore associations between variables. **Results:** The study cohort comprised 21 females (70%) and 9 males (30%), with the majority of cases (49.5%) occurring in the 0–15 years age group. Jaundice (36%), abdominal mass (32%), and right upper quadrant pain (32%) were the most common clinical features. Complications such as cystolithiasis (24%) and pancreatitis (29%) were observed, while cirrhosis and malignancy were rare. Ultrasonography was utilized in all patients as an initial diagnostic tool, while MRCP (33%) and CT scans provided additional insights. ERCP was performed in 42% of cases, but its use has since declined due to associated risks. **Conclusion:** This study highlights the demographic and clinical features of choledochal cysts, with a focus on the utility of advanced imaging modalities like MRCP for non-invasive and accurate diagnosis. Early detection and intervention are key to reducing complications and improving outcomes.

Keywords: Choledochal cyst, biliary anomaly, jaundice, abdominal mass, MRCP, diagnostic imaging.

Copyright © 2024 The Author(s): This is an open-access article distributed under the terms of the Creative Commons Attribution 4.0 International License (CC BY-NC 4.0) which permits unrestricted use, distribution, and reproduction in any medium for non-commercial use provided the original author and source are credited.

INTRODUCTION

Choledochal cysts are rare congenital anomalies of the biliary tree characterized by abnormal dilatation of the bile ducts. These anomalies, often presenting in early childhood but sometimes diagnosed in adulthood, can lead to serious complications such as cholangitis, pancreatitis, or biliary malignancy if untreated. Understanding their clinical presentation and leveraging advanced diagnostic techniques are essential for timely diagnosis and appropriate management [1-3].

The clinical presentation of choledochal cysts is highly variable, ranging from asymptomatic cases to severe symptoms, including jaundice, abdominal pain, and recurrent episodes of cholangitis. In pediatric populations, the classic triad of jaundice, right upper

quadrant pain, and a palpable abdominal mass is a hallmark, though it is less frequently observed in adults. In adults, nonspecific symptoms like dyspepsia or intermittent abdominal discomfort may delay diagnosis, increasing the risk of complications [4-6].

Traditional diagnostic approaches for choledochal cysts relied heavily on clinical examination and basic imaging modalities, which often lacked the precision needed for accurate characterization. With advancements in imaging techniques, the ability to diagnose and classify these cysts has significantly improved [7]. Modern modalities such as ultrasonography (USG), magnetic resonance cholangiopancreatography (MRCP), and endoscopic retrograde cholangiopancreatography (ERCP) allow detailed visualization of the biliary anatomy, enabling

clinicians to distinguish choledochal cysts from other hepatobiliary conditions [8, 9].

Ultrasonography remains a first-line investigation due to its non-invasive nature and widespread availability. It provides crucial initial insights into the presence of biliary dilatation. MRCP, on the other hand, offers a non-invasive and highly detailed evaluation of the biliary and pancreatic ducts, making it the diagnostic modality of choice [10]. ERCP, while invasive, is instrumental in cases requiring therapeutic intervention, such as biliary drainage or stent placement [12].

Classification of choledochal cysts, as per Todani's system, further aids in guiding clinical management. This system categorizes cysts into five main types based on their location and morphology, influencing both the diagnostic approach and surgical planning. Enhanced diagnostic precision has not only facilitated early detection but also enabled minimally invasive surgical strategies, improving patient outcomes [13, 14].

Given the potential for severe complications, the timely diagnosis of choledochal cysts is crucial. This underscores the need for clinicians to remain vigilant about its clinical features and utilize improved diagnostic techniques effectively. Future advancements in imaging and molecular diagnostics may further refine our understanding and management of this rare condition.

Objective

To assess the clinical features and strategy for diagnosis of choledochal cyst using improved diagnostic techniques.

METHODOLOGY

Type of Study

This study was a retrospective observational analysis conducted to evaluate the clinical presentation, diagnostic strategies, and outcomes associated with choledochal cysts.

Place of Study

The study was carried out in the Hepato-Biliary-Pancreatic Surgery (HBPS) department of BIRDEM Hospital, Dhaka, a specialized tertiary care

center renowned for treating complex hepatobiliary conditions.

Period of Study

The data for this study were collected over a five-year period, spanning from January 2003 to January 2008.

Sample Size

A total of 30 patients diagnosed with choledochal cysts were included in the study.

Criteria of Selection

Patients were selected based on a confirmed diagnosis of choledochal cysts. This diagnosis was established through a combination of patient history, clinical examination, and diagnostic imaging investigations conducted at BIRDEM. Additionally, patients previously diagnosed at other centers and referred to the HBPS unit in BIRDEM for further management were also included. Inclusion was random, irrespective of the patients' age or sex, ensuring a diverse representation of cases.

Statistical Analysis

Data collected from the patients were systematically recorded, cleaned, and analyzed using **SPSS software (version 22)**. Descriptive statistics were used to summarize the demographic and clinical characteristics of the study population. Continuous variables such as age were expressed as mean \pm standard deviation (SD), while categorical variables, including gender distribution and clinical presentation, were presented as frequencies and percentages. Relationships between clinical parameters and diagnostic modalities were assessed using chi-square tests or Fisher's exact tests where appropriate. A p-value of <0.05 was considered statistically significant.

RESULTS

The study included 30 patients with choledochal cysts, comprising 21 females (70%) and 9 males (30%). The age distribution revealed that the majority of cases (49.5%) occurred in the 0–15 years age group, with 10 females and 5 males. The 16–35 years age group accounted for 26.4% of cases, including 6 females and 2 males, while 23.1% were in the 36–50 years age group, consisting of 5 females and 2 males. Notably, no cases were observed in patients above 50 years.

Table 1: Age and sex distribution of the patients

Age group	Female	Male	%
0-15	10	5	49.5
16-35	6	2	26.4
36-50	5	2	23.1
>50	0	0	00

In children, presence of abdominal mass is the most common clinical feature. It presented in 36% of patients. Jaundice is common in all age group, about

32%. On the otherhand abdominal pain is more common in elderly patient.

Table 2: Classical triad of clinical presentation of choledochal cyst

Clinical feature	Age group(yrs)			%
	0-15	15-35	35-50	
jaundice	6	2	1	36
Abdominal mass	8	-	-	32
Right upper quadrant pain	2	5	1	32

Cystolithiasis are present in 24% of patient. Another common presentation is pancreatitis, present in 29% of patients. Cirrhosis with portal hypertension and malignancy are less common. Two patients were

previously operated. In both Cases Cysto-duodenostomy was done. One patient presented with feature of cholangitis and the other presented with obstructive jaundice due to stricture of the anastomosis.

Table 3: Clinical presentation of associated Hepato-Biliary pathology

Associated Hepato-Biliary pathology				
Clinical feature	Age group(yrs)			%
i. Cystolithiasis	-	4	2	24
ii. Fever with features of cholangitis	4	3	2	29.7
iii. Pancreatitis	-	3	4	28
iv Hepatolithiasis	-	-	-	0
v. cirrhosis of liver			1	3.3
vi. Malignancy	-	-	-	0
Previously operated	1	1	-	8

All the jaundiced patients had hyperbilirubinaemia and raised liver enzymes. However

some non-icteric patients had raised alkaline phosphatase and prolonged prothrombin time.

Table 4: Results of liver function tests of the patients

Parameters	No. of Patients	% of patients
Hyperbilirubinaemia	7	21
Raised ALT	7	21
Raised AST	7	21
Raised Alkaline phosphatase	10	30
Reduced Albumin	7	21
Prolonged Prothrombin Time	8	24

The most common investigation done is USG, almost in all patients. ERCP done in 42% of patients. But now It is avoided because of post ERCP complications.

The most sensitive and non invasive is MRCP and done in 33% of patients. But CT scan can describe the status of surrounding structure of cyst.

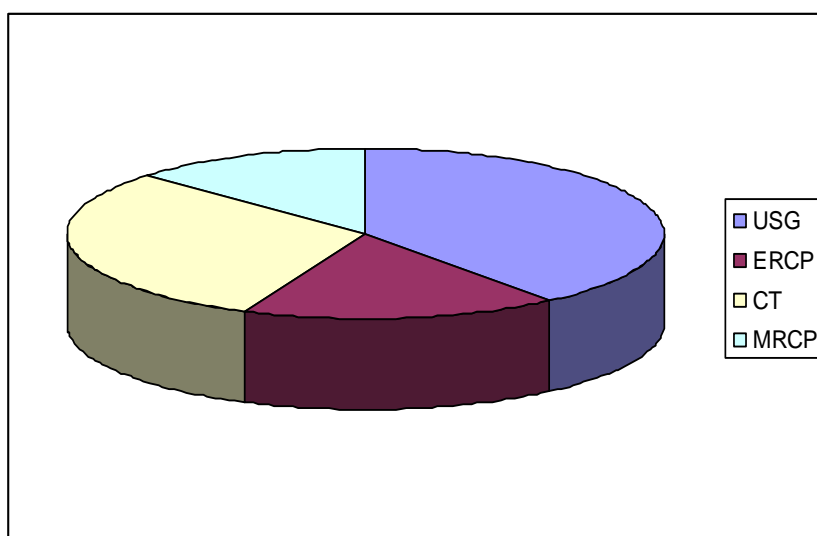


Fig 1: Invesgations done in choledochal cyst

DISCUSSION

This study highlights the demographic and clinical profile of patients with choledochal cysts and compares findings with previously published data. The predominance of females (70%) aligns with existing literature, which also reports a higher prevalence of choledochal cysts in females. The age distribution in our study shows that nearly half (49.5%) of cases occur in children aged 0–15 years, consistent with studies that emphasize the congenital nature of this condition [11]. Notably, no cases were observed in individuals older than 50 years, a finding also supported by other reports that show a sharp decline in prevalence with increasing age.

The clinical features in our study align with classical presentations. The triad of jaundice, abdominal pain, and abdominal mass is frequently cited in other research. In our study, jaundice was observed in 36% of patients, with a predominance in younger age groups. This is comparable to studies where jaundice is reported as the most common symptom across all age groups [12]. Abdominal mass, observed in 32% of patients, was particularly common in children, echoing findings that this symptom is more pronounced in younger individuals. Conversely, right upper quadrant pain was more prevalent in older patients, consistent with literature indicating that abdominal pain often accompanies complications like cholangitis or pancreatitis in adults.

Associated hepatobiliary pathologies were notable in our cohort. Cystolithiasis was found in 24% of patients, and pancreatitis in 29%, findings that are comparable to similar studies reporting these as common complications [14]. Cirrhosis and malignancy were rare in our study, aligning with the relatively lower prevalence of these severe complications in other reports. Two previously operated cases with complications, including cholangitis and obstructive jaundice due to anastomotic stricture, underscore the importance of careful post-operative monitoring.

Biochemical investigations revealed that hyperbilirubinemia, raised liver enzymes, and prolonged prothrombin time were common, particularly in jaundiced patients. These findings are consistent with other studies, which similarly report elevated liver function parameters in choledochal cyst cases [15, 16]. However, non-icteric patients with elevated alkaline phosphatase levels highlight the variability in clinical and biochemical presentations.

Ultrasound (USG) was the most frequently used diagnostic modality, performed in nearly all cases. This reflects its widespread availability and utility as an initial screening tool. While endoscopic retrograde cholangiopancreatography (ERCP) was used in 42% of patients, its use is now declining due to the risk of complications, as noted in contemporary guidelines.

Magnetic resonance cholangiopancreatography (MRCP), performed in 33% of cases, is emerging as the most sensitive and non-invasive diagnostic tool, corroborating trends observed in other studies [17]. Computed tomography (CT) remains valuable for delineating surrounding structures and complications.

Overall, our findings are consistent with existing research, while underscoring the importance of early diagnosis and intervention to prevent complications. Improved imaging modalities like MRCP and multidisciplinary management strategies offer better outcomes, especially in cases requiring complex surgical correction.

CONCLUSION

This study highlights the demographic, clinical, and diagnostic profile of choledochal cysts, emphasizing a higher prevalence in females and younger age groups, particularly children under 15 years. Jaundice, abdominal mass, and abdominal pain were the most common clinical features, with associated complications such as cystolithiasis and pancreatitis observed in a significant number of cases. Biochemical investigations confirmed liver dysfunction in many patients, while imaging modalities, particularly ultrasound and MRCP, proved invaluable for diagnosis. These findings underscore the importance of early detection and the adoption of advanced diagnostic techniques to guide timely and effective management, minimizing complications and improving patient outcomes.

REFERENCE

- O'Neill Jr, J. A. (1992). Choledochal cyst. *Current problems in surgery*, 29(6), 371-410. doi: 10.1016/0011-3840(92)90025-x.
- Liu, H., & Lu, X. H. (1996). The diagnosis of choledochal cyst (a report of 50 cases). *Xin Xiaohuabingxue Zazhi*, 4, 259.
- Zhang, Z., & Wei, H. L. (1996). Congenital choledochal dilatation in adult (a report of 3 cases). *Xin Xiaohuabingxue Zazhi*, 4(Suppl 5), 32.
- Qiao, Q., Sun, Z., & Huang, Y. (1997). Diagnosis and treatment of congenital choledochal cysts in adults. *Zhonghua wai ke za zhi [Chinese Journal of Surgery]*, 35(10), 610-612.
- Wang, L., Wang, S. F., & Li, Y. G. Choledochal cyst (A report of 2 cases). *Xin Xiaohuabingxue Zazhi*, 4(Suppl 5), 210.
- Stain, S. C., Guthrie, C. R., Yellin, A. E., & Donovan, A. J. (1995). Choledochal cyst in the adult. *Annals of surgery*, 222(2), 128-133. doi: 10.1097/00000658-199508000-00004.
- Weyant, M. J., Maluccio, M. A., Bertagnolli, M. M., & Daly, J. M. (1998). Choledochal cysts in adults: a report of two cases and review of the literature. *Official journal of the American College of Gastroenterology/ ACG*, 93(12), 2580-2583. doi: 10.1111/j.1572-0241.1998.00633.x.

8. Kaneko, K., Ando, H., Watanabe, Y., Seo, T., Harada, T., Ito, F., ... & Sugito, T. (1999). Secondary excision of choledochal cysts after previous cyst-enterostomies. *Hepato-gastroenterology*, 46(29), 2772-2775.
9. Lipsett, P. A., Pitt, H. A., Colombani, P. M., Boitnott, J. K., & Cameron, J. L. (1994). Choledochal cyst disease. A changing pattern of presentation. *Annals of surgery*, 220(5), 644-652. doi: 10.1097/00000658-199411000-00007.
10. Hewitt, P. M., Krige, J. E. J., Bornman, P. C., & Terblanche, J. (1995). Choledochal cysts in adults. *Journal of British Surgery*, 82(3), 382-385. doi: 10.1002/bjs.1800820333.
11. Chijiwa, K., Komura, M., & Kameoka, N. (1994). Postoperative follow-up of patients with type IVA choledochal cysts after excision of extrahepatic cyst. *Journal of the American College of Surgeons*, 179(6), 641-645.
12. Chijiwa, K., & Tanaka, M. (1994). Late complications after excisional operation in patients with choledochal cyst. *Journal of the American College of Surgeons*, 179(2), 139-144.
13. Chaudhary, A., Dhar, P., & Sachdev, A. (1997). Reoperative surgery for choledochal cysts. *British journal of surgery*, 84(6), 781-784.
14. Akhan, O., Demirkazik, F. B., Özmen, M. N., & Ariyürek, M. (1994). Choledochal cysts: ultrasonographic findings and correlation with other imaging modalities. *Abdominal imaging*, 19, 243-247. doi: 10.1007/BF00203517.
15. Guo, Y. H., & Zhang, X. (1993). Role of endoscopic retrograde cholangiopancreatography in the diagnosis and treatment of choledochoceles (A report of 14 cases). *Xin Xiaohuabingxue Zazhi*, 1, 92-93.
16. Okada, A., Oguchi, Y., Kamata, S., Ikeda, Y., Kawashima, Y., & Saito, R. (1983). Common channel syndrome—diagnosis with endoscopic retrograde cholangiopancreatography and surgical management. *Surgery*, 93(5), 634-642.
17. Shimada, K., Yanagisawa, J., & Nakayama, F. (1991). Increased lysophosphatidylcholine and pancreatic enzyme content in bile of patients with anomalous pancreaticobiliary ductal junction. *Hepatology*, 13(3), 438-444.