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Original Research Article

Clinical profile, management, and outcomes of choledochal cysts in adults: experience from a tertiary care center in the Sub-Himalayan region

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ABSTRACT

Background: Choledochal cysts (CDCs) are rare congenital anomalies of biliary system, and adult presentations are uncommon and pose diagnostic and surgical challenges, particularly in distinguishing type VI CDC, a rare variant involving isolated cystic dilation of the cystic duct. This retrospective observational study evaluates the clinical profile, imaging, surgical management, and outcomes of adult CDC cases.
Methods: A retrospective review of 31 adult patients with CDCs from July 2021 to December 2024 was conducted. Data on demographics, clinical presentation, imaging findings, surgical approach, and postoperative outcomes were analyzed.
Results: The mean age was 37.55 years (range: 16-73), with 83.87% female predominance. There were 20 (64.52%) type I, 1 (3.22%) type II, 9 (29%) type IV, and 1 (3.22%) type VI CDC, initially misclassified as type IVA. Roux-en-Y hepaticojejunostomy (RYHJ) was performed in 83.87% of cases, with 8% requiring Lilly's technique. Postoperative complications included biochemical leaks (23.08%), pancreatic fistulas (7.69%), and bile leaks (7.69%), all managed conservatively.
Conclusions: Complete cyst excision remains gold standard, preventing complications such as cholangitis and malignancy. Type VI CDCs are frequently misclassified, necessitating intraoperative reassessment for accurate diagnosis. MRCP though gold standard, has limitations, reinforcing need for surgeon awareness and intraoperative decision-making.

Keywords: Choledochal cysts, Type VI Choledochal cysts, Biliary anomalies

INTRODUCTION

Choledochal cysts (CDCs) are rare congenital anomalies characterized by cystic dilatation of the bile ducts. The incidence of CDCs varies geographically, being higher in Asia (1 in 5000 live births) than in Western countries (1 in 100,000-150,000).[1]

The majority of CDCs are diagnosed in infants and children within the first decade of life, presentation occurs in adulthood in approximately 25% of patients.[2] Although predominantly diagnosed in childhood, adult presentations are not uncommon and are often associated with delayed diagnosis and complications such as cholangitis, pancreatitis, and malignancy.

The classification of CDCs has evolved over time, with the Todani classification being the most widely used, it categorises CDCs into five types based on the location and morphology of the cysts.[3]

Magnetic resonance cholangiopancreatography (MRCP) is currently the most reliable pre-operative imaging modality for evaluating cyst anatomy and the classifying the disease based on the standard Todani classification.[4,5]

Type I cysts, characterised by fusiform or cystic dilation of the extrahepatic bile duct, are the most common, followed by type IV, which involves multiple cysts with intrahepatic involvement.[6] Type VI CDC is the most recent addition to the classification system by Serradel et al.[7]

This study aims to evaluate the clinical profile, management strategies, and outcomes of adult CDCs patients treated at a tertiary care centre in the Sub-Himalayan region of North India, highlighting the challenges and lessons learned from this cohort.

METHODS

Study design and setting

This retrospective observational study was conducted at Indira Gandhi Medical College, Shimla (HP), analysing adult CDCs cases treated from July 2021 to Dec 2024. The institution is a tertiary care centre catering to a predominantly rural and high-altitude population in the Sub-Himalayan region.

Study population

A total of 39 cases were initially identified, of which eight cases were excluded due to incomplete records, leaving thirty-one cases for analysis. The inclusion criteria required patients to be aged sixteen years or older, have a radiological diagnosis of CDCs, and possess complete medical records along with follow-up data. Patients were excluded if they had incomplete records, inadequate follow-up data, or were below sixteen years of the age.

Data collection

Data were collected from medical records, imaging reports, and operative notes. The parameters analyzed included demographics, presenting symptoms, imaging findings, surgical strategies, complications, and outcomes. Patients with incomplete records were excluded.

Statistical analysis

Descriptive statistics were used to summarize the data. Continuous variables were expressed as means and standard deviations, while categorical variables were presented as frequencies and percentages. Outcomes were compared based on CDCs type and the management strategy.

RESULTS

Demographics and clinical presentation

The mean age of the cohort was 37.55 years (range: 16-73), with a significant female predominance (83.87%; female-to-male ratio of 5:2) (Table 1).

Most patients (61.29%) were residents of high-altitude areas. The predominant presenting symptoms were abdominal pain (81.81%) and biliary symptoms such as nausea and vomiting (64.51%). Six patients in this cohort (19.35%) had already undergone cholecystectomy: two laparoscopic cholecystectomies, two open cholecystectomies, and 2 laparoscopic converted to open sub-total cholecystectomies for cholelithiasis. Five other patients had concomitant cholelithiasis on imaging. Three (9.67%) patients had preoperative evidence of cystolithiasis in this study cohort.

One patient with cystolithiasis and jaundice had iatrogenic cyst perforation and acute pancreatitis following endoscopic retrograde cholangiopancreatography (ERCP), which was successfully managed with conservative treatment.

None of the adult patients demonstrated classic triad of pain, jaundice, and a palpable mass, nor did they present with cholangitis or pancreatitis. Preoperative ERCP was not performed in any patient to diagnose CDCs or to demonstrate an anomalous pancreaticobiliary junction.

Imaging and diagnosis

Ultrasound was the initial diagnostic modality for all patients, with MRCP providing a confirmatory diagnosis in all cases. The mean size of the CDCs in the study cohort was 24.94±10.97 mm (Range 14-50 mm) (Table 1). One patient was diagnosed during ERCP for cystolithiasis, with a pre-ERCP diagnosis of choledocholithiasis.

There was total of 20 (64.52%) type I, 1 (3.22%) type II, 9 (29%) type IV and 1 (3.22%) type VI CDCs in this study cohort (Figure 1 and 2). The type VI CDC was reported as type IVA in the preoperative imaging.

Management strategies

Among the 31 patients, 26 (83.87%) underwent surgical excision with RYHJ (Figure 2). Two patients with dense adhesions between cyst wall and the portal vein required Lilly's technique with RYHJ.[8] Non-op management was employed for 5 patients (16.12%) due to advanced age, multiple comorbidities/refusal of surgery.

Outcomes

The mean surgical duration was 210±40 minutes, and the mean hospital stay was 7 days, with a range of 5 to 25

days. Eight patients (30.77%) in the operative cohort had drain fluid amylase levels exceeding three times the serum amylase levels. Among them, six (23.08%) were noted to have biochemical leaks, while two (7.69%) developed grade-B postoperative pancreatic fistulas as defined by the international study group on pancreatic surgery guidelines.[9]

Additionally, 2 (7.69%) experienced bile leaks during postop period, which were managed conservatively and resolved within 10-25 days. wound complications observed in 3 patients, classified as Clavien-Dindo grade I-II.[10] There was no mortality in study cohort. Mean follow-up period 26.36±6.90 months, and all operated patients remained asymptomatic during this time.

Table 1: Summary of demography, types of cysts, operative parameters, and postoperative complications.

Parameters	Details	
Demography	Mean age: 37.55 years (range: 16-73 years)	
	Female predominance: 26/31(83.87%), female: Male=5:2	
Types of CDCs	Type I: 20 (64.52%)	
	Type II: 1 (3.22%)	
	Type IV: 9 (29%)	
	Type VI: 1 (3.22%)	
Cyst size (Mean±SD, mm)	24.94±10.97 mm (Range 14-50 mm)	
Associated cholelithiasis and hepatolithiasis	Cholelithiasis-11/31 (35.48%) patients	
	Hepatolithiasis-3/31 (9.68%) patients	
Previous surgery	06/31 (19.35%),	02 laparoscopic cholecystectomies
		02 open cholecystectomies
		02 laparoscopic converted to open subtotal cholecystectomies
Operative procedure	CDC excision with RYHJ: 24 (92%)	
	Lilly's technique with RYHJ: 2 (8%)	
Operative parameters	Mean surgical duration: 210±40 minutes	
	Mean hospital stay: 7 days (range: 5-25 days)	
Postoperative complications	Wound complication, Clavien-Dindo grades I-II: 3/26 (11.54%)	
	Biochemical leak/grade-A pancreatic fistula: 6/26 (23.08%)	
	Grade-B pancreatic fistula: 2/26 (7.69%)	
	Bile leaks: 2/26 (7.69%)	

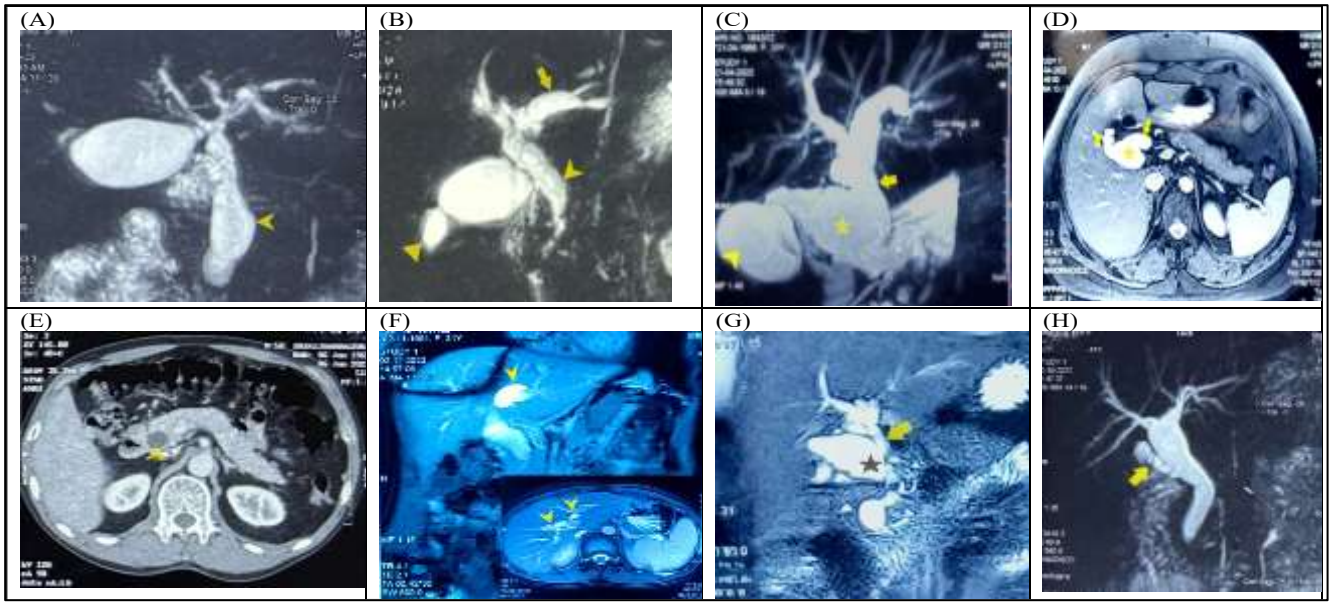

Figure 1 (A-H): Radiological variants of CDCs.

(A) Type I CDC: Fusiform dilation of the common bile duct (CBD) (arrowhead), characteristic of a Type I CDC. (B) Type IV CDC: Dilated CBD (arrowhead) along with dilated right and left hepatic ducts (arrows), indicating intrahepatic involvement. An associated Phrygian cap (triangle) is present in the gallbladder. (C and D) Type VI CDC: Cystic dilation of the cystic duct (star) arising between the undilated CBD (arrow) and the gallbladder (arrowhead). The gallbladder remains anatomically positioned, with an apparent close association with the cyst. This anomaly is also confirmed intraoperatively (refer to Figure 2 D). (E) Type I CDC with Intrapancreatic Involvement: Fusiform dilation extending into the intrapancreatic portion of the bile duct (arrowhead). (F) Type IV CDC: Sagittal and axial images (inset) showing a dilated right hepatic duct (arrowhead) and sectoral ducts (arrows), confirming intrahepatic involvement. (G) Type II CDC: Diverticulum-like outpouching (star) arising from the CBD (arrow). (H) Cystic duct remnant dilation, type I CDC: Post-cholecystectomy patient with prominent cystic duct remnant.

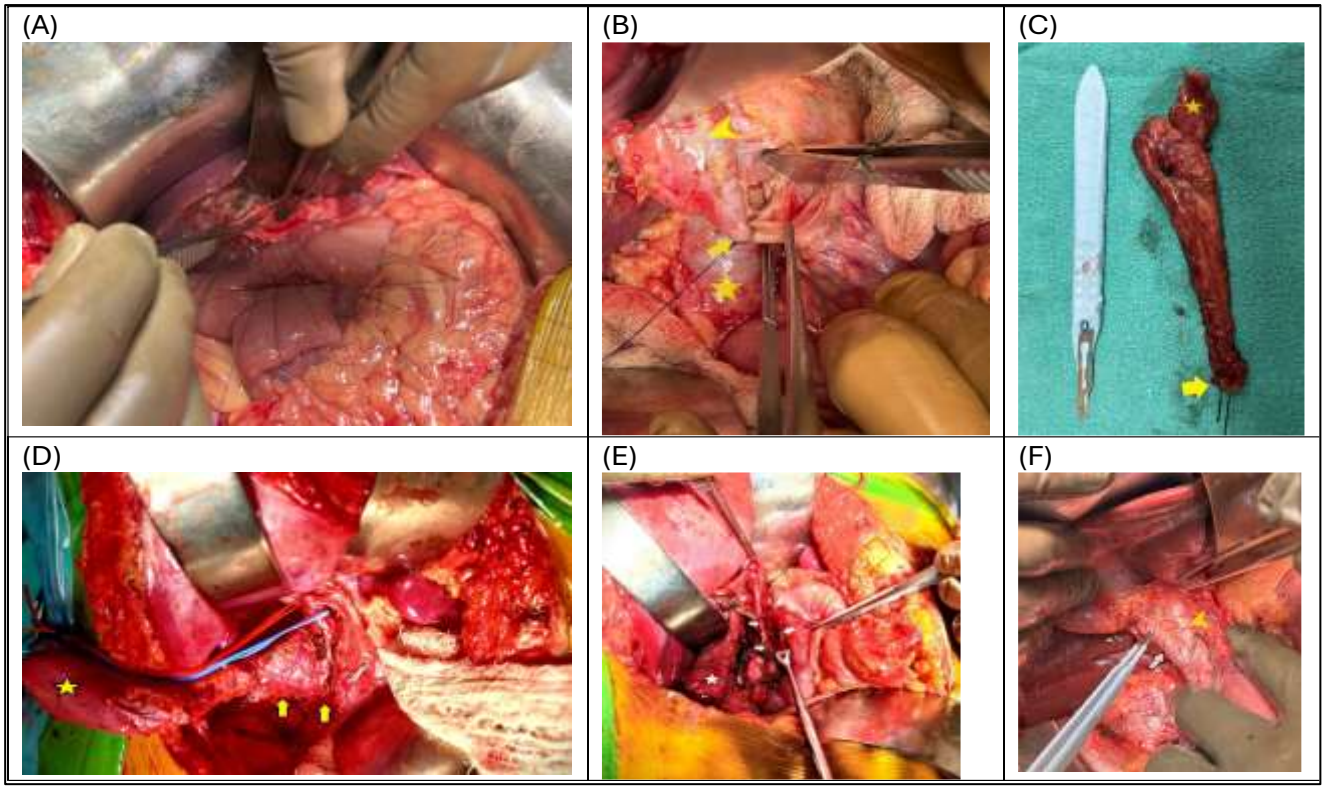

Figure 2 (A-F): Intraoperative images.

(A) Type IV-A CDC-post excision: Intraoperative image showing RYHJ in progress. The right and left hepatic ducts (pointed by forceps) are prominent. The common hepatic duct is divided close to the hilum, with dilated intrahepatic ducts left intact (Refer to Figure 1 B for MRI correlation). (B) Type I CDC-Intrapancreatic CBD dissection: dissection of the intrapancreatic segment of the dilated CBD, with ligation at the waist (arrow). The duodenum and pancreas are retracted manually. The inferior vena cava is marked with a star (Refer to Figure 1E for MRI correlation). (C) Excised specimen of type IV CDC: Gross specimen showing the excised cyst along with a contracted gallbladder (star). A suture at the lower end (arrow) marks the division site. D) Type VI CDC-Cystic dilation of the cystic duct (arrows) arising between the undilated common bile duct (looped in blue tape) and the gallbladder (star). The right hepatic artery looped in red tape, coursing anterior to the common hepatic duct (Refer to Figure 1 C and D for MRI correlation). (E) Lilly's technique in dense adhesions: Cyst wall laid open as part of Lilly's technique, used in cases of dense adhesions between the choledochal cyst and the portal vein. (F) Type I CDC-low insertion of prominent cystic duct: Prominent cystic duct (arrow) inserting low into the common bile duct (arrowhead), seen in type I CDC.

DISCUSSION

The aetiology of type I and IVA CDCs is believed to stem from an abnormal pancreaticobiliary duct junction. This anomaly creates a long common channel, facilitating the reflux of pancreatic juice into the bile ducts, which leads to mucosal damage, inflammation, and subsequent cystic dilatation.[11,12]

Other theories propose a congenital origin, such as distal aganglionosis or aberrations in embryological recannulation, resulting in proximal bile duct dilation.[13] Type V cysts remain unique due to their restriction to intrahepatic ducts and lack of extrahepatic involvement.

The diagnosis of CDCs in adults remains challenging due to the absence of classical symptoms, reflecting the often-subtle presentation in adults.[14,15] The classic triad of pain, jaundice, and a palpable mass was not observed in any patient in this cohort, leading to delayed recognition and misdiagnosis. Instead, most patients presented with non-specific biliary symptoms, such as abdominal pain (81.81%) and nausea/vomiting (64.51%), which were often attributed to cholelithiasis or functional biliary disorders rather than CDCs.

A key diagnostic challenge in this cohort was misclassification of CDCs types on preoperative imaging. One case, initially classified as Type IVA CDC on MRCP, was later confirmed intraoperatively as Type VI CDC. Type VI CDC, a rare entity involving cystic dilation of the cystic duct, was first described by Bode and Aust in 1983, and officially recognized in the CDC classification by Serradel et al.[7,16]

Given its rarity, preoperative imaging often fails to differentiate it from type I or IV CDCs. This underscores the importance of intraoperative reassessment, as well as the need for standardized radiological criteria for CDCs subtypes.

Complete surgical excision remains the gold standard treatment for CDCs to prevent complications such as cholangitis, pancreatitis, and malignancy.[17] Internal drainage procedures, such as cyst-enterostomy, are contraindicated due to their association with recurrent cholangitis and stricture formation.[17,18]

In this study, 83.87% of patients underwent RYHJ, the preferred surgical approach for definitive cyst excision and biliary reconstruction.

One of the key surgical challenges encountered was the presence of dense adhesions between the cyst and surrounding structures, particularly in patients with a history of biliary inflammation or previous surgery. In two cases (8% of the surgical cohort), Lilly's technique was required due to strong adhesions between the cyst and the portal vein, making standard excision unsafe.[8]

In patients with previous cholecystectomy (19.35%), the CDCs diagnosis was missed at the time of surgery, leading to delayed intervention. This highlights a major gap in awareness among general surgeons and radiologists.

The overall surgical outcomes were favorable, with no mortality and a mean hospital stay of 7 days (range: 5-25 days). However, postoperative complications included; biochemical leaks/grade-A pancreatic fistula (23.08%), grade-B pancreatic fistulas (7.69%), requiring prolonged drainage but resolving conservatively. Bile leaks occurred in 2 (7.69%) patients, which were successfully managed nonoperatively within 10 to 25 days.[8,9]

These findings suggest that biochemical leaks are relatively common but self-limiting in most cases. The use of intraoperative drain placement allowed for early detection and conservative management, avoiding the need for additional surgical interventions.

The risk of long-term complications, including anastomotic stricture and malignancy, remains a major concern. Literature suggests a 10% risk of biliary anastomotic stricture following RYHJ reconstruction, but no strictures were observed during the mean follow-up period of 26.36±6.90 months in this cohort.

While this is encouraging, long-term follow-up beyond two years is needed to assess the true incidence of strictures and late complications.[19]

Malignancy risk in CDCs is well documented, with literature citing a 15-28% risk of cholangiocarcinoma if left untreated.[20,21]

Although no malignancies were identified in this cohort, the need for complete excision is reinforced by the potential risk of malignancy and long-term oncologic surveillance remains essential.

This study provides several key takeaways that can enhance the diagnosis and management of CDCs. Early MRCP based diagnosis is crucial, particularly in patients undergoing cholecystectomy with atypical biliary symptoms, as misdiagnosed cases may lead to unnecessary procedures and delayed intervention. However, preoperative imaging alone may not always be conclusive, as demonstrated by the misclassification of type VI CDC as type IVA on MRCP. This underscores the importance of intraoperative reassessment in confirming cyst type and planning definitive management.

Surgical challenges, including dense adhesions, require specialized techniques such as Lilly's technique, which is invaluable in ensuring safe excision.

Additionally, routine postoperative monitoring of drain amylase levels is essential, as it allows for early detection and conservative management of biochemical leaks and pancreatic fistulas, minimizing the need for reintervention.

Given the potential for late complications, including anastomotic strictures and malignancy, extended follow-up remains necessary, especially in high-risk patients.

Moreover, a major concern highlighted in this study is the high rate of unrecognized CDCs in patients who had previously undergone cholecystectomy (19.35%). This emphasizes the need for increased awareness and training for surgeons performing cholecystectomy, enabling them to identify CDCs intraoperatively and prevent missed diagnoses.

Future research should focus on developing standardized MRCP criteria for CDCs subtypes, particularly type VI, and exploring minimally invasive approaches such as robotic-assisted cyst excision to optimize surgical outcomes.

The study's limitations include its retrospective design, single-centre data collection, and relatively small sample size. Moreover, certain parameters, such as the length of the common channel and the presence of an abnormal pancreaticobiliary junction, were not consistently documented, limiting the ability to fully analyse these factors.

CONCLUSION

This study highlights the diagnostic, surgical, and postoperative challenges in managing adult CDCs. The misclassification of type VI CDC as type IVA on preoperative imaging demonstrates the limitations of MRCP alone and the importance of intraoperative evaluation. Surgical outcomes were favorable, with no mortality and manageable complications, but long-term surveillance remains critical. The lessons learned emphasize the need for improved preoperative imaging

protocols, intraoperative decision-making, and structured postoperative follow-up to optimize patient outcomes.

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