

ORIGINAL ARTICLE

Endoscopic Retrograde Cholangiopancreatography in Patients with Choledochal Cyst, A Tertiary Care ExperienceERUM KAZIM¹, MUHAMMAD ALI TAJ², AFTAB AHMED LEGHARI³, RABIA FERAZ⁴, RAHEEL AHMED⁵, SHAHRIYAR GHAZANFAR⁶¹Assistant Professor Surgery, Dow University of Health Sciences and Civil Hospital Karachi.²Consultant gastroenterologist, Civil hospital Karachi³Chief Medical officer surgical unit IV, Dr. Ruth K. M. Pfau, Civil Hospital Karachi⁴Assistant Professor, DMC, DUHS, Karachi⁵Assistant Professor, DMC, DUHS, Karachi⁶Professor of Surgery, Dow University of Health Sciences and Civil Hospital KarachiCorresponding author: Erum Kazim, Email: erumkazim@duhs.edu.pk, Cell: 0332-2323808**ABSTRACT****Objective:** Our aim of this analysis was to evaluate the clinical features, laboratory values, radiological findings, endoscopic management, and technical success in patients with Choledochal cyst (CC).**Materials & Methods:** This retrospective study was carried out at our Endoscopy suite, between January 2010 to January 2022. All patients of choledochal cyst who underwent ERCPs in last 12 years were analyzed. Technical success was defined as achieving a biliary cannulation and successfully acquiring the desired goal. Descriptive and Frequency analysis were incorporated in Statistical analysis with SPSS.**Results:** Procedures performed in 100 patients with suspected choledochal cyst either for a diagnostic or therapeutic purpose were evaluated. 48 ERCPs (48%) were performed for therapeutic purpose and 52 (52%) for diagnostic purpose. Diagnostic findings after successful ERCP were CC in 67 (67%), CC with stone in 28 (28%), CC with cholangiocarcinoma in four (4%) and CC with biliary stricture in one (1%). No mortality was documented in our patients who underwent Procedure. Patients referred for surgery were 92 (92%) and ERCP affected management in 100% (100/100 procedures).**Practical Implications:** This study will determine the clinical features, laboratory values, radiological findings, endoscopic management, and technical success in patients with Choledochal cyst.**Conclusion:** ERCP is a contemporary procedure, its utility in cases with CCs in a developing country with scarce resources and limited availability of gold standard modality (MRCP), is astounding and safe.**Keywords:** Endoscopic retrograde cholangiopancreatography; Choledochal cyst; Classification; Endoscopic management.**INTRODUCTION**

Choledochal cysts (CCs) are defined as cystic dilatation of different parts of bile duct and can be either intrahepatic or extra-hepatic or both. They account for about 1% of all benign biliary tract diseases and are most common in Asian population^{1,2}. Their exact pathogenesis remains unclear; however an abnormal pancreaticobiliary union (APBDU) and subsequent biliary contents reflux of into the biliary tree is observed to play a role³. The initial classification of CC was modified by Todani and colleagues in 1977^{4,5}. They classified choledochal cyst in to five basic types⁶.

Abdominal pain, jaundice and a palpable mass in right upper quadrant is the classic symptom triad but the majority of patients present with complications^{7,8}. Bile stasis causes a variety of complications in patients with choledochal cyst including cholangitis, pancreatitis, stone formation, portal hypertension, liver fibrosis, secondary liver cirrhosis and spontaneous cyst perforation⁹. There is an association of choledochal cyst with malignancy. It is a premalignant condition in which cancer develops 10–15 years earlier than in the general population^{9,10}.

Choledochal Cyst should be diagnosed early and managed appropriately as mortality and morbidity associated with CC increases as the age of the patient increases. Ultrasonography, computed tomography, endoscopic retrograde cholangiopancreatography, and magnetic resonance cholangiopancreatography are all important in the diagnosis of CC. There have been technological advances in noninvasive cross-sectional imaging modalities during the last four decades, including the development of magnetic resonance cholangiopancreatography (MRCP)¹¹. MRCP is not readily available in this part of the world and there are cost issues. Although ERCP is invasive, it is still the most precise diagnostic method for CCs. ERCP completely delineates biliary anatomy, assisting in the detection of an aberrant pancreaticobiliary duct junction or CBD filling abnormalities, which can range from simple stones to malignancies^{12,13}.

Our purpose of this research was to evaluate the clinical features, laboratory values, radiological findings, endoscopic management and technical success in CC patients.

METHODS

This study was carried out at our Department of Gastroenterology; between January 2010 to January 2022. Medical data of patients procedure done was assessed and analyzed retrospectively. For the purpose of this evaluation, demographic and clinical records were assessed including age, sex, medications used, signs and symptoms, co-morbid conditions, previous surgery, laboratory tests and imaging modalities used, indications for ERCP, complications, cannulation attempts, cannulation time, total procedure time, mode of therapy and referrals to surgical team. All the procedures were performed by a team of expert endoscopists, performing ERCPs for the last many years. Consent was taken from all patients and it was a part. Approval was taken from the institutional review board of DUHS. Procedures were performed under conscious sedation by a combination of nalbuphine (Agorid; Standpharm) and midazolam (Dormicum; Roche), all under the supervision of a senior anesthesia nurse. Deep sedation with propofol (Diprivan; AstraZeneca) by anesthesia specialist was used in selected cases, at the discretion of the endoscopy specialist. Cephalosporin was used as a pre requisite antibiotic prophylaxis during the procedure which was continued in an oral form of the same group for 5 days. ERCP was performed by employing the standard technique, using adult therapeutic duodenoscope (TJF 180; Olympus America Inc.). The ionic contrast medium Urografin (A mixture of salts of diatrizoic acid) was used to opacify the bile and pancreatic duct. Pulse oximeter was used to monitor oxygen saturation continuously during the procedure. In cases of an overactive duodenum, we also administered intravenous atropine 0.5 mg as required in increments, not to exceed a total of 3 mg or 0.04 mg/kg. A triple-lumen sphincterotome (CleverCut3™; Olympus Medical) was used, which allows the injection of contrast without removing the wire. A wire-guided cannulation technique (WGC) was used in all, with an intention to reduce the rate of complication notably post-ERCP pancreatitis (PEP). We attempted biliary cannulation with the sphincterotome for a maximum of eight times or no more than ten minutes, if unable to cannulate, we shifted to the other alternative cannulation techniques (needle knife sphincterotomy). Sphincterotomy and stent was placed, if required and bile duct stones were removed with the help of Dormia basket

and/or balloon catheters; all at the discretion of the endoscopist. If there was a suspicion of a tumor or mass biopsy specimens were taken. Technical success was defined as achieving a biliary cannulation and successfully acquiring the desired goal. Procedural complications were identified and their severity graded according to the Cottons classification¹⁴. CCs were classified according to the Todani et al⁵ and type VI by Maheshwari¹⁵. Serum amylase and lipase were checked when clinically indicated.

Statistical analysis included descriptive statistics and frequency analysis. Data was analyzed using SPSS version 17 (SPSS Inc., Chicago, IL, USA).

RESULTS

From January 2010 to January 2022, procedures performed in 100 patients with suspected choledochal cyst either for a diagnostic or therapeutic purpose were evaluated. There were 33 (33%) men and 67 (67%) women with a mean age of 34.4 ± 1.75 , range of 4 years to 65 years and a median of 34 years. 48 ERCPs (48%) were performed for therapeutic purpose and 52 (52%) for diagnostic purpose. Clinical features were abdominal pain in 76 (76%), jaundice in 20 (20%), nausea in 20 (20%), fever in 11 (11%), vomiting in 7 (7%) and abdominal mass in 1 (1%). Laboratory investigations were; median alanine aminotransferase (ALT) of 24 U/L, median serum alkaline phosphatase of 455 U/L, median bilirubin of 4 mg/dl and a median leukocyte count of $7,000/\text{mm}^3$. Tran-sabdominal ultrasound was abnormal in 100 (100%) and computed tomography (CT) scan was performed in 59 (59%) patients and was abnormal in all. Magnetic resonance cholangiopancreatography (MRCP) was performed in only seven (7%) patients. Main indications for ERCP were deranged liver function tests with dilated common bile duct (CBD) (LFTs) in 68 (68%), suspected choledocholithiasis with dilated CBD in 28 (28%) and suspected mass with dilated CBD in 4 (4%). Technical success was achieved in all the cases (100%); first procedure success rate was 97% in 97 patients, with a repeat procedure and success in the remaining 3 (3%). Successful cannulation of the native papilla was achieved in first attempt in 97 (97%), second attempt in 2 (2%) and third attempt in 1 (1%). Total number of ERCPs performed in all the patients were 117; three (3%) had four, six (6%) had three, three (3%) had two and 81 (81%) had one. Median time from referral time to ERCP was 11 days with a range of 4 to 32 days. Biliary access was achieved after mean attempts of 3.34 (range: 1-8); mean time of successful cannulation was 4.8 min (range: 1 min to 12 min) and median total time of a single procedure was 11 min with a range of 7 min to 25 min. Techniques used to cannulate the desired duct were sphincterotomy in 91 (91%), sphincterotomy with guidewire impacted first in 6 (6%) and needle knife pre-cut sphincterotomy in 3 (3%). Assumed factors related to a difficult cannulation were small ampulla in twelve (12%), unstable position in 8 (8%), long and floppy ampulla in 6 (6%); cannulation was effortless and straightforward in 74(74%). Type and associated features of choledochal cysts on imaging techniques are shown in table 1. Diagnostic findings after successful ERCP were CC in 67 (67%), CC with stone in 28 (28%), CC with cholangiocarcinoma in 4 (4%) and CC with biliary stricture in 1 (1%). Total number of therapeutic interventions performed in all the patients were 110; endoscopic biliary sphincterotomy in 32 (29%), CBD balloon traction in 31 (28.1%), biliary stent placement in 19 (17.2%), biliary stent exchange in 11 (10%), biliary balloon sphincteroplasty in 7 (6.3%), endoscopically guided biopsy in 4 (3.6%), basket stone extraction in 3 (2.7%) and needle knife pre-cut sphincterotomy in three (2.7%). Varying sizes of 30 single plastic biliary stents in different sessions were placed in 19 (20.4%) patients; 10F 10-cm in fifteen (50%), 10F 9-cm in six (20%), 10F 7-cm in three (10%) and 10F 12-cm in six (20%). PEP occurred in two (2%) patients; both the patients had mild PEP with an asymptomatic recovery. Immediate bleeding after sphincterotomy was noted in 3 (3%) patients which was stopped with balloon-tamponade by using standard occlusion balloons and spraying with adrenaline (epinephrine) (1/10000), followed by injection of adrenaline

(epinephrine) into the papilla between 10 and 12 o'clock. One (1%) patient was admitted with delayed bleed, he was discharged with a smooth recovery after being monitored for 72hrs. There was no procedure-related mortality in our patients. Patients referred for surgery were 89 (89%) and ERCP affected management in 100% (100/100 procedures).

Table 1:

	Types of choledochal cysts n/100	Frequency
1	IA 57	61.2%
2	IC 22	23.7%
3	III 3	3.2%
4	IVA 9	9.7%
5	IVB 1	1.1%
6	VI 1	3.2%
7	APBDJU* 12	12.9%

Table 1

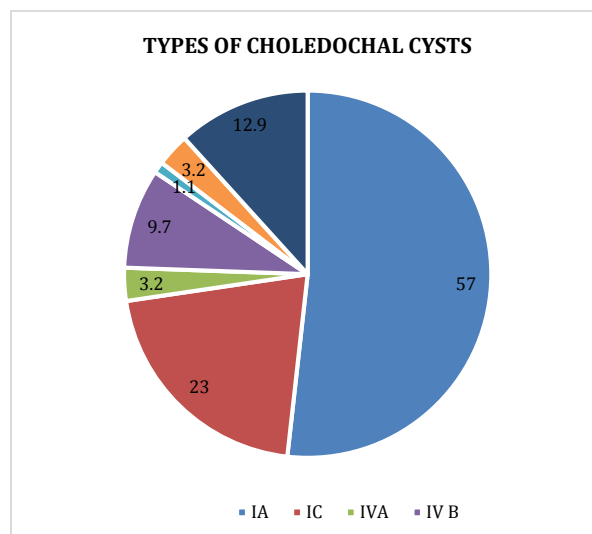


Figure 1:

Table 2: ERCP

Indications	Findings	Frequency
Diagnostic 52	<ul style="list-style-type: none"> CC in 67 (67%), CC with stone in 28 (28%), CC with cholangiocarcinoma in 4 (4%) CC with biliary stricture in 1 (1%). 	
Therapeutic 48	<ul style="list-style-type: none"> endoscopic biliary sphincterotomy in 32 (29%), CBD balloon traction in 31 (28.1%), biliary stent placement in 19 (17.2%), biliary stent exchange in 11 (10%), biliary balloon sphincteroplasty in 7 (6.3%), endoscopically guided biopsy in 4 (3.6%), basket stone extraction in 3 (2.7%) needle knife pre-cut sphincterotomy in three (2.7%). 	

DISCUSSION

Endoscopic retrograde cholangiopancreatography (ERCP) is a dynamic procedure, with an evolution from a diagnostic to a leading-edge therapeutic procedure for a variety of pancreatobiliary disorders. CC are uncommon congenital defects of the bile ducts; abnormal and disproportionate cystic dilatation of the bile ducts are the hallmarks. The incidence of CC is lower in

western population but higher in the Asian population with two third cases are reported from Japan and the reason is still unknown¹⁶. Type I are the most common with a distribution of 50% -80%, type II are 2 %, type III are 1.4%-4.5%, type IV are 15%-35% and type V are 20 %¹⁷. In our series, most common presentation was of type I CC and one patient had a rare type VI CC.

In 1959, Alonso-Lej and colleagues devised the initial classification system for CC, which included three types of CC⁴. In 1977, Todani and colleagues expanded this classification system, which is most commonly used by clinicians⁵. Another classification based on the cyst's relation to the ampulla of Vater and the pancreatic duct was coined by Sarris and colleagues; they subdivided CC into five types. Another entity of CC described by Lilly and colleagues¹⁸ was "form fruste" CCs with a spectrum of clinical features including abdominal pain, obstructive jaundice and abnormal pancreaticobiliary duct junction (APBDJU) but with normal bile ducts. There are few case reports where different type of cysts can be combined together in one patient as described by Kaneyama¹⁹. Visser and colleagues recently proposed a new classification system, grouping together different diseases, marking different causes, natural courses, surgically viable options and ensued complications^{20,21}. We used Todani classification system⁵ in our series for type I to type V CC and Maheshwari¹⁵ for type VI CC.

The etiology of CCs is still not known and many notions have been proposed, among them a much acclaimed one is the Babbitt's theory²². According to this theory an abnormal pancreaticobiliary duct junction (APBDJ) is identified as etiological factor contributing in development of choledochal cyst. This results in mixing of the pancreatic and biliary juice activating the pancreatic enzymes causing inflammation and degradation of the biliary duct wall, leading to ductal expansion²². There are other theories with lesser impact, Davenport and Basu²³ proposed that CCs are purely congenital anomalies and Ohkawa and colleagues suggested deficiency of elastin is the major factor behind the etiology²⁴. Variety of developmental anomalies can occur with CCs¹⁷. We did not encounter any of the above mentioned anomalies in our series.

CCs are considered to be a premalignant condition; there is predisposition for an early cancer, manifesting 10–15 years earlier²⁵. An estimated 10%-15% overall risk of cancer has been reported in patients with CC and this risk as well as histopathological dysplasia enhances as the patients age increases^{26,27}. Spectrum of cancers found in patients with CC and adenocarcinoma is the most common with a percentage of 73%–84%²⁸. The extrahepatic bile duct was the most common site in 50%–62%, followed by gallbladder in 38%–46%, intrahepatic bile ducts in 2.5%, and liver and pancreas in 0.7% each²⁹. In our series, 4% of the patients presented with carcinoma and surgical referral was deferred in all of them.

Timeline of clinical presentation is diverse and can occur at any time but in the majority of patients occurs earlier, coinciding with our series. Abdominal pain, jaundice and a palpable abdominal mass is the most common clinical triad with which the patient presents but occurs in less than 20% of patients³⁰. Spectrum of diversifying complications occurs due to bile stasis leading to recurrent super infection, inflammation and stone formation in patients with CC³⁰. Ultrasonography is generally the first modality used for the diagnosis of CCs due to noninvasiveness, easy availability and economic viability, with a sensitivity of 71%–97% and is best for follow-up surveillance³¹. In our series, it was abnormal in all the cases. Hepatobiliary iminodiacetic acid (HIDA) scans can be used but is limited by its decreased ability to visualize the intrahepatic ducts, its sensitivity is variable i.e. 100% for type I but only 67% for type IV cysts³¹. HIDA scan was not done in any of our patients. Computed tomography (CT) scans have many advantages as it shows complete biliary system, surrounding structures are well delineated with a good depiction of malignancy³². Magnetic resonance imaging (MRI) and MRCP, being noninvasive is the gold standard modality for CC.

MRI has some limitations as its sensitivity and specificity decreases in the presence of intraductal air, blood, debris, stones or protein plugs, which leads to the interference with signal intensity and modifies visualization. Situation is quite different in this part of the world as MRI is not readily available and is expensive, only seven patients in our series were able to perform it. In patients with CCs, MRI has some disadvantages: intraductal air, blood, debris, stones or protein plugs, can interfere with the signal and alter visualization³². In spite of all these limitations the sensitivity of MRI is above 90%. In post-operative anastomosis the sensitivity of MRI is only 84% and for pancreaticobiliary junction abnormalities decreases to as low as 46%–60%³².

Current treatment of CCs is essentially surgery. Historically, cystenterostomy was considered the method of choice but due to increased incidence of malignancy (30%) in the remnant tissue, the procedure has been abandoned³³. Complete removal of the cyst and biliary diversion is the modality of choice either through hepaticoduodenostomy, or hepaticojejunostomy³³. ERCP has been invasive but it is still the most precise diagnostic modality for CCs. ERCP completely delineates biliary anatomy preoperatively, helps in revealing an abnormal pancreaticobiliary duct junction or filling defects in the CBD, which can be from simple stones to cancers³³. The sensitivity of ERCP decreases in cases of recurrent inflammation and scarring where the procedure becomes difficult³⁴. Being an invasive procedure, ERCP has been associated with an increase in the rate of cholangitis and post-ERCP pancreatitis³⁴. Despite some of these drawbacks we performed ERCP because our surgeons required a precise knowledge of the pancreaticobiliary ductal systems along with that most of our patients are from lower socioeconomic class and MRCP is not conveniently available as well as the imaging modality is expensive. In comparison, ERCP is easily available, technicians are skilled and endoscopists are expert at our institution as well as it is superior to MRCP in delineating minor ductal anomalies of the pancreaticobiliary ductal systems. We performed ERCP-guided interventions in complicated CCs, where required, all at the discretion of the endoscopist. Jang et al.³ (27) has concluded an effective role of ERCP in complicated cases of CCs prior to surgery and inferred that preoperative ERCP provide surgeons a more precise anatomical knowledge of the pancreaticobiliary ductal system thus making situation easier during total surgical excision of the CC. We had three patients of choledochoceles (type III CC) endoscopic sphincterotomy was performed in all the patients. Endoscopic sphincterotomy is the prime treatment for choledochoceles, it allows the drainage of bile and stones, even if the patients are asymptomatic, and some authors recommend prophylactic endoscopic sphincterotomy³⁶. Complication rate in our patients was low, two patients had mild PEP and four had a bleeding episode with an uneventful recovery.

CONCLUSION

ERCP is a contemporary procedure, its utility in cases with CCs in a developing country with scarce resources and limited availability of gold standard modality (MRCP), is astounding and safe. Its role in delineating the pancreaticobiliary junction anatomy and associated obstructive pathologies is considerable and is of great help in planning the surgery.

Conflict of Interest: No conflict of interest

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