Choledochal Cysts in Adults: The Clinicopathological Features and Surgical Outcomes in a Single Institute.

Ahmed El-Gendi[1], Mohamed El-Shafei[2], Essam Bedewy[3]
[1]Department of Surgery, Faculty of Medicine, Alexandria University, Egypt
[2]Department of Diagnostic and Interventional Radiology, Faculty of Medicine, Alexandria University, Egypt; [3]Department of Hepatology and Tropical Medicine, Faculty of Medicine, Alexandria University, Egypt

ABSTRACT

Introduction: *Choledochal cysts are believed to be a congenital disease however around 20% presents in adulthood. Direct comparison between them is seldom reported. The aim was to compare adult and pediatric choledochal cysts in patients of the same race treated in a single institution to highlight the difference in clinicopathological features, clinical presentation, management and surgical outcomes.* ***Patients and Methods:*** *From January 2015 to December 2018, 42 patients presented with diagnosis of choledochal cyst they were divided into two groups: pediatric group (below 18 years) and adult group (above 18 years). The following data was recorded and compared: demographic characteristics, presenting symptoms, diagnostic images, type of surgery, amylase level in the cyst, final pathology, perioperative complications, and follow-up data.* ***Results:*** *24 pediatric patients (mean 4.5 years) and 18 adult patients (mean 32.4 years) were compared. There was significantly higher female predominance in pediatric group (p=0.03). The main complain at presentation was abdominal pain in adult group (77.8%) compared to abdominal mass in pediatric group (75%). The median amylase level in bile was 220 U/L (range = 15–3580 U/L) in the pediatric group and 390 U/L (range = 37–12000 U/L) in the adult group with P=0.046. Pediatric group had a significant higher predominance for Type Ia cyst. There was no significant difference between the two groups as regards the incidence of gallstones or intrahepatic stones. Pediatric patients were statistically more frequently associated with APBDJ than adult patients (79.2% vs 44.4%, P=0.005). Excision of the cyst and hepaticojejunostomy was the most commonly performed procedure in both groups. Additional hepatic resection was needed for type V patients in two patients in both groups. Liver transplantation was performed for one type V patient in pediatric group. There was no 30 days mortality and no case of 30 days readmission. There was no statistical significant difference between adult and pediatric groups in 30 days morbidity and mortality (p=0.06). The median postoperative length of hospital stay was 7 days in adult group versus 8 days in pediatric group (p =0.65). The median follow-up duration was 24 months (range 15–48 months) in adult group and 26 months (range 14–45 months) in the pediatric group. Till the date of last follow up, all patients are still alive with no development of malignancy in either group.* ***Conclusion:*** *Choledochal cyst in Pediatrics are more likely to be associated with female predominance, present by abdominal mass, Type 1a cyst, and stones inside the choledochal cyst whereas adult patients commonly present by abdominal pain, and are more often associated with APBDJ. Excision of the entire extra hepatic biliary tree with hepaticojejunostomy is the most commonly performed surgical procedure in adults and pediatrics to treat extra hepatic cystic involvement with chronic abdominal pain with recurrent mild cholangitis is the most common long-term complication in both groups.*

INTRODUCTION

Choledochal cysts are uncommon, with incidence is approximately 1 in 100,000–150,000 live births in western world[1]. It is often diagnosed in the first decade of life as a disease of childhood, however, around 20% presents in adulthood[1-2]. Prevalence is higher in Asian than in western countries; most cases are reported in Japan, where they occur in one of every 1,000 live births[3].

The etiology of choledochal cyst is unknown

Correspondence: Ahmed M. El-Gendi, Department of Surgery, Faculty of Medicine, University of Alexandria. El Sultan Hussein Street, El-Azarita, Khartom Square, Alexandria, Egypt. Postcode: 21131. Tel: +2034847426, +201222408223, Fax: +2035426676. Email: draelgendi@gmail.com, ahmed.elgendi@alexmed.edu.eg

and the classical described theory is the anomalous pancreaticobiliary ductal junction (APBDJ) theory with associated reflux of pancreatic juices into the biliary tract resulting in destruction and damage to the bile duct wall with consequent dilatation and cystic formation[4-7]. Other theories are being proposed to explain pathogenesis of all subtypes include partial distal bile duct obstruction, sphincter of oddi dysfunction, and abnormal few ganglion cells in distal bile ducts[8-11].

Todani's classification using an alpha-numeric system to describe different types of choledochal cyst is still currently in use despite being challenged in published literature as being confusing, unsupported by evidence, misleading, and serves no purpose[12]. New subtypes of choledochal cysts have been added to the widely accepted classification system for choledochal cysts[11].

Data from published literature revealed a difference between pediatric and adult patients with choledochal disease in their presenting symptoms and clinical outcomes[13-14]. However, studies comparing pediatric and adult choldochal cyst in patients of the same race treated at single institution during the same time period has been rarely reported[15-16].

The aim of our study is to highlight the difference in clinicopathological features, clinical presentation, management and outcomes of pediatric versus adult patients with choledochal cysts treated at our surgery department in a single tertiary referral center.

PATIENTS AND METHODS

This study was conducted at the surgery department, Faculty of Medicine, Alexandria University. The ethical committee and the review board approved the study and treatment protocol. All patients who agreed to participate in this study had signed a written informed consent. From January 2015 to December 2018, all patients with diagnosis of choledochal cyst were discussed in multidisciplinary team meeting and were enrolled in the study. Patients with previous intervention or surgery were excluded. According to patients' age of presentation, they were divided into two groups: pediatric group (below 18 years) and adult group (above 18 years).

Data collection included: demographic characteristics, presenting symptoms, diagnostic images, type of surgery, amylase level in the cyst, final pathology, perioperative complications, and follow-up data. Diagnosis of choledochal cyst was based upon radiologic imaging including abdominal ultrasonography, magnetic resonance cholangiopancreatography (MRCP) and triphasic abdominal computed tomography. Preoperative MRCP confirmed by data from intraoperative cholangiogram were used to classify the type of choledochal cyst. Choledochal cysts were classified according to Todani's classification. Biliary fluid sample was aspirated and analyzed for amylase level. Choledochal cyst fluid was obtained at the beginning of cyst dissection before any surgical manipulation. Perioperative morbidity and mortality were recorded. Perioperative complications were defined as those occurring within 30 days of surgery or within the same hospital admission.

Statistical Analysis: The raw data were coded and entered into SPSS system files (SPSS package version 18). Analysis and data interpretation was conducted. Categorical data are presented as numbers and percentages. The following statistical measures were used: descriptive statistics including frequency, distribution, mean, median, standard deviation and inter-quartile range were used to describe different characteristics. Kolmogorov – Smirnov test was used to examine the normality of data distribution. Univariate analyses including: Student t-test and Mann Whitney test were used to test the significance of results of quantitative variables. Chi-Square test, Monte Carlo test and Fisher's Exact test were used to test the significance of results of qualitative variables. Statistical significance was set at $P<0.05$.

RESULTS

During the study period, 42 patients presented with diagnosis of choledochal cyst. There were 24 pediatric patients (range from 72 days to 16 years, mean 4.2 years) and 18 adult patients (22 to 59 years, mean 32 years). Female to male ratio was 3.5:1 in pediatric age and 1.4:1 in adults. The female predominance was significantly higher in pediatric age compared to adults ($p=0.03$).

The main presenting symptom, Todani classification, biliary amylase level, presence or absences of stones are listed in Table 1.

Abdominal pain was the main complaint in 77.8% (14/18) in adult group vs. 16.7% (4/24) in pediatric group (P=0.001) whereas abdominal mass was the main presenting feature in pediatric group (75% vs. 5.5%, P<0.0001). There was no statistical significant difference between the two groups as regards the presence of jaundice at time of presentation despite of the significantly higher incidence of stone presence in choledochal cysts in pediatric group (70.8%) vs 22.2% in adult group with P=0.001. The median amylase level in bile was 220 U/L (range = 15–3580 U/L) in the pediatric group and 390 U/L (range = 37–12000 U/L) in the adult group with P=0.72.

Table 1: Demographic, clincopathological features and classification of choledochal cyst in studied population

	Pediatric group 24 patients	**Adult group 18 patients**	**P value**
Age range	72days-16 years	22-59 years	-
Mean	4.2	32	
Sex (M: F)	1:3.5	1:1.4	0.03
Presenting symptom			
• **Abdominal mass**	18/24 (75%)	1/18 (5.5%)	<0.001
• **Abdominal Pain**	4/24 (16.7%)	14/18 (77.8%)	0.001
• **Jaundice**	2/24 (8.3%)	0 (0%)	0.17
• **Incidental**	0 (0%)	3/18 (16.7%)	0.6
APBDJ	19/24 (79.2%)	8/18 (44.4%)	P=0.005
Todani Classification			
Type Ia	14 (58.3%)	3 (16.7%)	0.0039
Type Ib	0	1 (5.6%)	
Type Ic	3 (12.5%)	3 (16.7%)	
Type II	0	1 (5.6%)	
Type III	0	2 (11%)	
Type IVa	4 (16.7%)	3 (16.7%)	
Type IVb	0	0	
Type V	3 (12.5%)	2 (11%)	
Others unclassified	0	3 (16.7%)	
Amylase level in bile	220 U/L	390U/L	0.72
Range	15–3580 U/L	37–12000 U/L	
Presence of stones			
• **Stones in choledochal cyst**	17/24 (70.8%)	4/18 (22.2%)	0.001
• **Gall stones**	4/24 (16.7%)	4/18 (22.2%)	0.764
• **Intrahepatic stones**	3/24 (12.5%)	1/18 (5.6%)	0.842

*P<0.05

As illustrated in table 1, there was a statistical significant difference in the types of choledochal cyst according to Todani's classification. Pediatric group had a significant higher predominance for Type Ia cyst compared to adult group (58.3% vs. 16.7% P=0.0039). In adult group, three patients couldn't be fitted in Todani's classification; 2 cystic duct cysts (figure 1) and one cystic dilatation of right hepatic duct (figure 2). The latter three patients had no symptoms and diagnosis was obtained during routine ultrasound imaging.

Table 2: APBDJ and type of surgery performed in relation to Todani's classification in the two groups

	Pediatric group 24 patients			**Adult group 18 patients**		
	Number	**APBDJ**	**Treatment**	**Number**	**APBDJ**	**Treatment**
Type Ia	14	13/14	Excision+HJ	3	3/3	Excision+HJ
Type Ib	0	0	-	1	0	Excision+HJ
Type Ic	3	3/3	Excision+HJ	3	3/3	Excision+HJ
Type II	0	0	-	1	0/1	Excision
Type III	0	0	-	2	0/2	Excision+HJ
Type IVa	4	3/4	Excision+HJ	3	2/3	Excision+HJ
Type IVb	0	0	-	0	0	-
Type V	2 1	0/3	-Partial Hepatectomy+CBD excision+HJ -LDLT	2	0/2	Partial Hepatectomy+CBD excision+HJ
Others unclassified						
Cystic duct	0	0		2	0	Cholecystectomy
RHD cyst	0	0		1	0	Right hepatetcomy

Stones in choledochal cyst were more common in the cysts of pediatric group patients than those of adult group (70.8% vs. 22.2%, P=0.001). There was no significant difference between the two groups as regards the incidence of gallstones or intrahepatic stones. Anomalous pancreaticobiliary ductal junction (APBDJ) was found in 8/18 patient of the adult group compared to 19/24 patient of pediatric group. Pediatric patients were statistically more frequently associated with APBDJ than adult patients (79.2% vs. 44.4%, P=0.005).

All patients in both groups underwent surgery; type of surgery in relation to Todani's classification is listed in Table 2. Excision of the cyst and hepaticojejunostomy was the most commonly performed procedure in pediatric and adult groups (87.5% and 66.7% respectively). Partial hepatic resection in addition to excision of the extra hepatic biliary system and hepatico-jejunostomy was performed in two patients with type V cysts in pediatric group compared to three patients in adult group as illustrated in figure (two patients with type V as seen in figure 3,4, and one patient with an unclassified type with cyst involving only right hepatic duct as illustrated in figure 5). Living donor liver transplantation was performed for only one patient with type V choledochal cyst (Caroli disease) in pediatric group. Excision of choledochal cyst only was performed to treat type II as illustrated in figure 6.

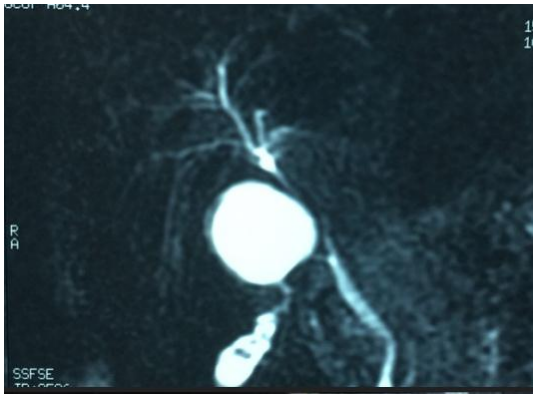


Fig. (1): MRCP showing cystic duct choledochal cyst in adult.

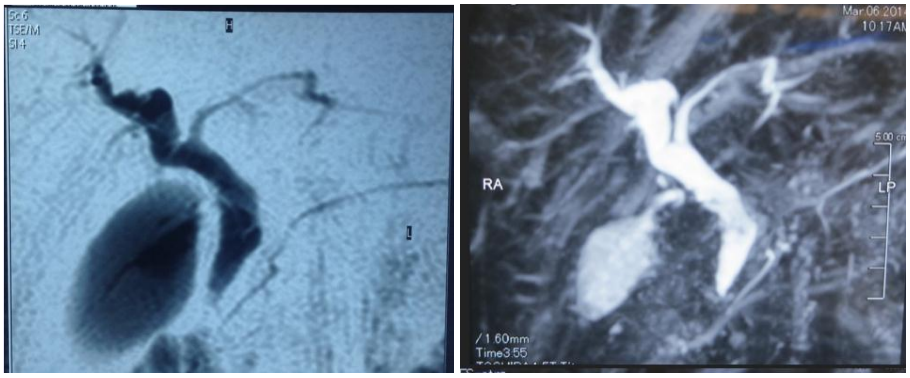
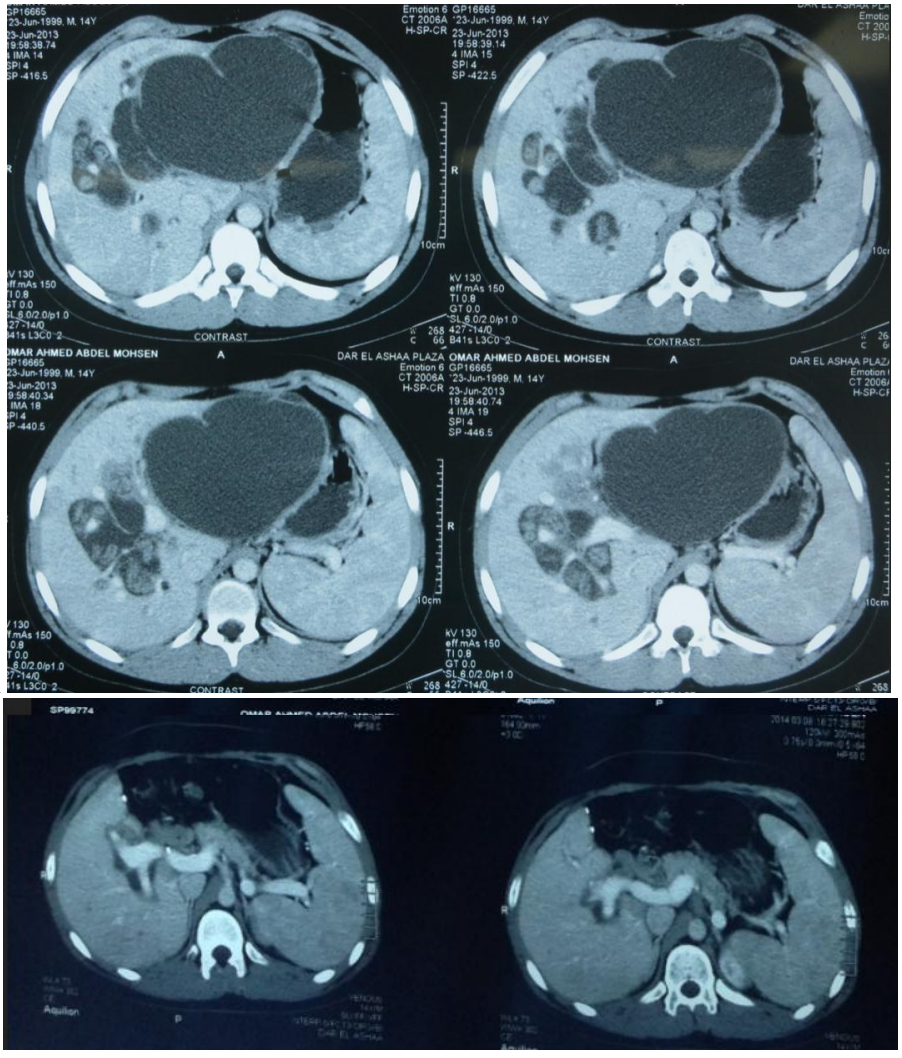


Fig. (2): MRCP for choledochal cyst of the right hepatic duct in adult.



a

b

Figure (3): a) Abdominal CT scan showing Caroli disease with central dot sign with intrahepatic stones. b) Postoperative abdominal CT scan with disappearance of the central dot sign and intrahepatic stone after surgical treatment with left hepatectomy CBD excision and hepaticojejunostomy.

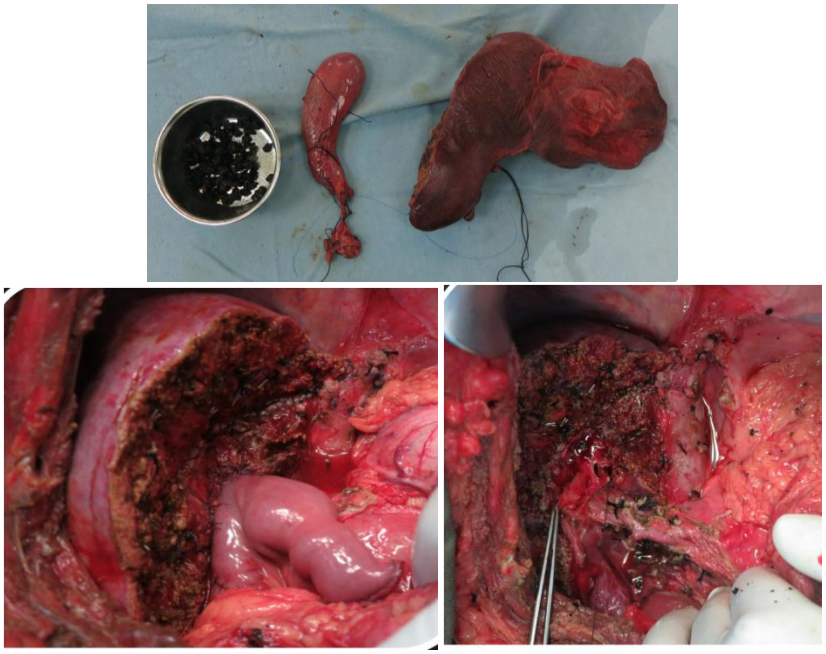


Fig. (4): Operative pictures of the patient with Caroli disease treated by left hepatectomy, stone extraction, CBD excision and hepaticojejunostomy

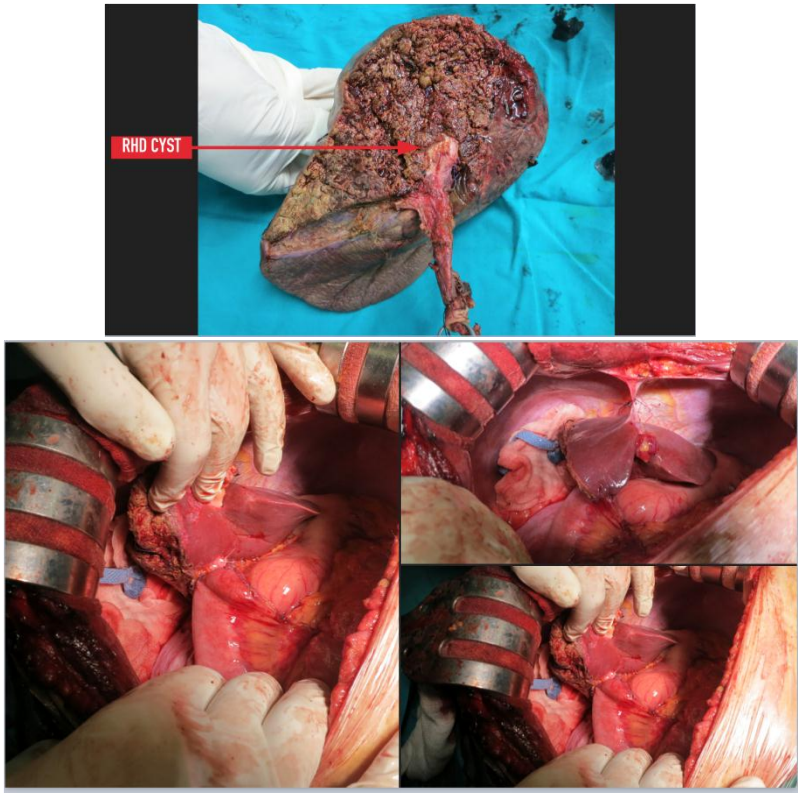


Fig. (5): Showing right hepatetcomy with CBD excision and hepatico-jejunostomy performed to treat adult patient with right hepatic duct cholecochal cyst.

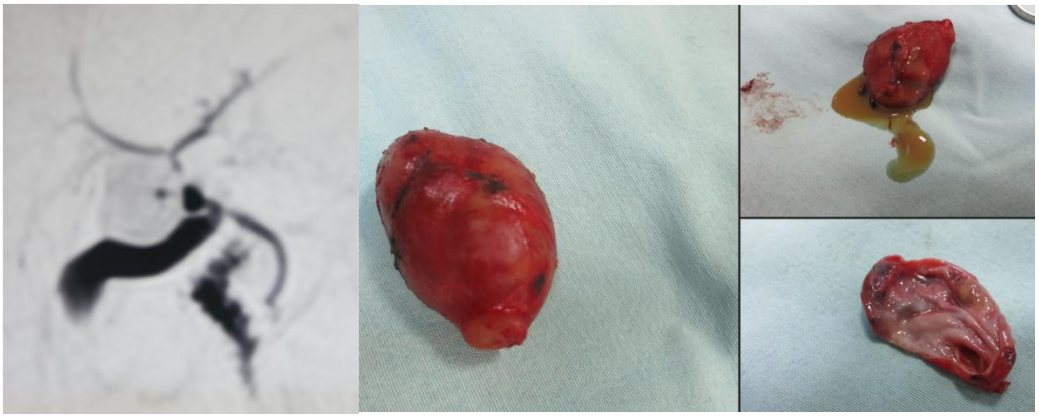


Figure (6): Showing type II choledochal cyst in adult treated by cyst excision only.

No patient had malignant disease at time of choledochal cyst surgery. There was no statistical significant difference between adult and pediatric groups in 30 days morbidity and mortality (p=0.06). 21% (5 patients) of pediatric and 16.7% (3 patients) of adult patients had early postoperative complications. In pediatric group, 2 patients had bile leak that settled conservatively, 1 had wound infection, 1 had chest infection and the transplanted child had hepatic artery thrombosis that was successfully re-vascularized. In adult group, one patient had bile leak that required drainage, and two cases had wound infection. There was no 30 days mortality and no case of 30 days readmission. The median postoperative length of hospital stay was 7 days in adult group versus 8 days in pediatric group (p =0.65).

The median follow-up duration was 24 months (range 15–48 months) in adult group and 26 months (range 14–45 months) in the pediatric group. Till the date of last follow up, all patients are still alive with no development of malignancy in either group. 3 patients in the pediatric groups required re-admission due to repeated attacks of ascending cholangitis, which were treated conservatively by antibiotics. Chronic abdominal pain with recurrent mild cholangitis was the most common long-term complication in both groups (8 in pediatric group and 6 in adult group).

DISCUSSION

In this study, we highlight the difference between pediatric and adult choledochal cyst treated in a single institution with stress upon differences in clinical presentation, APBDJ, Todani's types, biliary amylase level and management. Choledochal cyst is famous for being a congenital disease that is often diagnosed in the first decade of life, however a number of published series have reported cholecochal cyst in adulthood[12, 15-17]. Choledochal cyst in adults if explained by the congenital theory suggests that the cyst can remain asymptomatic and silent for years before the diagnosis[18] or in some patients the disease process pursuit a protracted clinical course leading to delay in time of presentation[19]. The latter may hints towards an acquired nature for some cholodochal cysts. In our series 18 patients presented during their adult life compared to 24 in pediatric age, this number is the highest reported in literature however it may reflect a referral bias. Many recent published series have reported an increasing number of adults with choledochal cyst disease, to the extent that adults make up the majority of patients in one of them. This tend to suggest that choledochal cyst disease is more common in adults than it was initially thought, and its clinical presentation in adults seems to be different from that in children[20]. Although female predominance was commonly reported in previous published series especially in adult age group[12, 15-16, 21-22], in our series, female predominance was not a predominant feature might be reflecting ethnic and racial issues.

Authors have previously reported that abdominal mass and jaundice are more commonly reported as the main presentation in pediatric with choledochal cyst than in adults[3, 16, 23]. Adult patients usually presents with abdominal pain

while pediatric patients presents with jaundice and abdominal mass[15]. In this study, palpable abdominal mass was the main presenting symptom in pediatrics whereas adult patients tend to mainly complaint about abdominal pains. Incidental finding of choledochal cyst was only present in adult age group (16.7%). None of adult group patients experienced obstructive jaundice as the main presentation. The latter could reflect a difference in etiology and pathogenesis of choldochal cyst among adults than in pediatrics. We found a significant difference in the distribution of various types of choledochal cysts between pediatric and adult patients. Type 1 cyst had a significant predominance in pediatric group. Some authors reported no difference in distribution among adult and pediatric population[16] while other have reported that type IVa was predominant in adults[12, 15, 24]. The difference in distribution may be because the intrahepatic dilatation of IVa is poorly defined and the distinction between type Ic and IVa is difficult that has led some authors to believe that Types I and IVa, are variations of a single disease.

Our results showed that type IV, or V cysts in both pediatric and adult age group are at higher risk of stone formation, whereas type Ia in pediatric group is associated with higher frequency for stone formation compared to same type adult group. The later might reflect the time and degree of biliary obstruction leading to stasis. The intrapancreatic convergence of both the main bile duct and the main pancreatic duct into a common pancreaticobiliary channel that flows into the duodenum via the major papilla is found in 80-90% of cases and constitutes the classic anatomical picture of pancreaticobiliary junction[25-26]. Several definitions have been proposed for the anomalous pancreaticobiliary maljunctions; of which the anatomical definition defined as a markedly long common channel with convergence of the main bile duct and main pancreatic duct outside the duodenal wall. There is no definite minimal length of the common channel need for the diagnosis of APBDJ, and the criteria utmost importance is the convergence of the pancreatic and bile ducts outside the duodenal wall[27]. The mean length of the common channel is 4.6 mm (±2.6 mm) in normal adult[28]. However, some patients with a relatively long common channel OF 6mm and more are not classified as having APBDJ because the sphincter of Oddi includes the pancreaticobiliary ductal junction[27].

The APBDJ as an etiology for choldochal disease was proposed by Babbitt, however not all cases of choldochal disease can be explained by this theory[4]. APBDJ can occur in some patients in the absence of choldochal disease and not all cases of choldochal cysts have APBDJ. Many authors reported the finding of APBDJ in their series of choldochal cysts with frequency ranging from 29% to 96%[14, 29-30]. In our study, APBDJ was present more frequently in pediatric age group (58.3%) than in adult group (44.4%). The difference in Todani types and the frequency of APBDJ reported in our study may raise the hypothesis that the etiology and pathogenesis in adult patients is somewhat different from that in pediatrics. Authors have reported that severe narrowing of distal choldochal cyst is more commonly present among pediatric age group compared to adults. The combined effect of APBDJ and distal narrowing cause a severe pancreaticobiliary reflux and stasis, leading to rapid proximal choledochal dilatation, which might explain its early onset[8-10, 31].

The famous and most commonly used classification for choledochal cyst is the modified Todani system, which is a complex, alphanumerical system that classifies choldochal cysts into categories I –V. Several additional subtypes have been proposed to the Todani system. The cystic duct cyst needs to be added to Todani classification as type ID[32-33] (dilation of the cystic duct in addition to dilated CBD and CHD) and type VI CCs (isolated dilation of the cystic duct without CBD or CHD involvement). The latter is extremely rare with one few cases reported in literature[34-35]. Todani classification is being criticized for its complex alphanumeric system, and surgeons needs a simplified classification that directly correlated to a management plan. Visser et al. have challenged the Todani's classification, claiming that it combines several disease entities with different etiologies, pathogenesis, surgical management plan, and long term complication[12]. They proposed that type I and type IVa cysts are simply variations of the same disease, with some variation in the degree of intrahepatic involvement. This was backed up by their reporting in their series that all type I cysts had some degree on biliary dilation at its intrahepatic

pancreatico-biliary ductal system. Journal of pediatric surgery. 1979;14(1):9-15.
6. Kimura K, Ohto M, Saisho H, Unozawa T, Tsuchiya Y, Morita M, et al. Association of gallbladder carcinoma and anomalous pancreaticobiliary ductal union. Gastroenterology. 1985;89(6):1258-65.
7. Ono J, Sakoda K, Akita H. Surgical aspect ot cystic dilatation of the bile duct. An anomalous junction of the pancreaticobiliary tract in adults. Annals of surgery. 1982;195(2):203-8.
8. Craig AG, Chen LD, Saccone GT, Chen J, Padbury RT, Toouli J. Sphincter of Oddi dysfunction associated with choledochal cyst. Journal of gastroenterology and hepatology. 2001;16(2):230-4.
9. Davenport M, Basu R. Under pressure: choledochal malformation manometry. Journal of pediatric surgery. 2005;40(2):331-5.
10. Matsufuji H, Araki Y, Nakamura A, Ohigashi S, Watanabe F. Dynamic study of pancreaticobiliary reflux using secretin-stimulated magnetic resonance cholangiopancreatography in patients with choledochal cysts. Journal of pediatric surgery. 2006;41(10):1652-6.
11. Jablonska B. Biliary cysts: etiology, diagnosis and management. World journal of gastroenterology. 2012;18(35):4801-10.
12. Visser BC, Suh I, Way LW, Kang SM. Congenital choledochal cysts in adults. Archives of surgery (Chicago, Ill : 1960). 2004;139(8):855-60; discussion 60-2.
13. de Vries JS, de Vries S, Aronson DC, Bosman DK, Rauws EA, Bosma A, et al. Choledochal cysts: age of presentation, symptoms, and late complications related to Todani's classification. Journal of pediatric surgery. 2002;37(11):1568-73.
14. Chaudhary A, Dhar P, Sachdev A, Kumar N, Vij JC, Sarin SK, et al. Choledochal cysts--differences in children and adults. The British journal of surgery. 1996;83(2):186-8.
15. Edil BH, Cameron JL, Reddy S, Lum Y, Lipsett PA, Nathan H, et al. Choledochal cyst disease in children and adults: a 30-year single-institution experience. Journal of the American College of Surgeons. 2008;206(5):1000-5; discussion 5-8.
16. Huang CS, Huang CC, Chen DF. Choledochal cysts: differences between pediatric and adult patients. Journal of gastrointestinal surgery : official journal of the Society for Surgery of the Alimentary Tract. 2010;14(7):1105-10.
17. Soreide K, Korner H, Havnen J, Soreide JA. Bile duct cysts in adults. The British journal of surgery. 2004;91(12):1538-48.
18. Ramage AA, Tedesco FJ, Schuman BM. Asymptomatic choledochal cyst. The American journal of gastroenterology. 1985;80(10):816-8.
19. Han SJ, Hwang EH, Chung KS, Kim MJ, Kim H. Acquired choledochal cyst from anomalous pancreatobiliary duct union. Journal of pediatric surgery. 1997;32(12):1735-8.
20. Tan SS, Tan NC, Ibrahim S, Tay KH. Management of adult choledochal cyst. Singapore medical journal. 2007;48(6):524-7.
21. Lenriot JP, Gigot JF, Segol P, Fagniez PL, Fingerhut A, Adloff M. Bile duct cysts in adults: a multi-institutional retrospective study. French Associations for Surgical Research. Annals of surgery. 1998;228(2):159-66.
22. Lee KF, Lai EC, Lai PB. Adult choledochal cyst. Asian journal of surgery. 2005;28(1):29-33.
23. Lipsett PA, Pitt HA. Surgical treatment of choledochal cysts. Journal of hepato-biliary-pancreatic surgery. 2003;10(5):352-9.
24. Hewitt PM, Krige JE, Bornman PC, Terblanche J. Choledochal cysts in adults. The British journal of surgery. 1995;82(3):382-5.
25. Hand BH. An anatomical study of the choledochoduodenal area. The British journal of surgery. 1963;50:486-94.
26. Sterling JA. The common channel for bile and pancreatic ducts. Surgery, gynecology & obstetrics. 1954;98(4):420-4.
27. Kamisawa T, Ando H, Shimada M, Hamada Y, Itoi T, Takayashiki T, et al. Recent advances and problems in the management of pancreaticobiliary maljunction: feedback from the guidelines committee. Journal of hepato-biliary-pancreatic sciences. 2014;21(2):87-92.
28. Dowdy GS, Jr., Waldron GW, Brown WG. Surgical anatomy of the pancreatobiliary

portion and it has been shown that preoperative imaging was unable to accurately predict true intrahepatic involvement in choldochal cysts, and in some circumstances the clear distinction between type I and IVa cysts was confirmed after the cyst was excised and postoperative imaging showed no residual intrahepatic dilatation[36]. In addition, type II cysts was argued as being a variant of gallbladder duplication rather than choldochal cysts also type III (choledochocele) as a variant of duodenal duplication and both should not be included in Todani classification together with the Caroli disease which resemble choldochal cyst in morphology but with completely different etiology and pathogenesis. Visser et al. reserved the term ''congenital choledochal cyst'' to be used exclusively for describing congenital dilation of the extra hepatic and intrahepatic bile ducts (apart from Caroli disease) and that type II (CBD diverticula), type III (choledochoceles), and type V (Caroli disease) should no longer be thought of as subtypes of CCs[12]. Another simplified classification, which is based on management approaches was recently published[37].

The Todani classification does not cover all kinds of cystic biliary disease; isolated dilatation of the cystic duct is not included. Two patients in adult group had isolated cystic dilatation of the cystic duct with gall stone disease and absence of APBDJ. Serradel et al[38] described a similar case and named it ''type VI choledochal cyst'', an addition to the Todani classification. This type is extremely rare, with only nine patients reported in the literature[39-40].

Several authors reported that high biliary amylase level is commonly observed in choledochal cyst with APBDJ[41-43]. Pancreatic amylase is low in neonates due to incomplete exocrine pancreatic development. It increases gradually within 2-3 years to reach the adult level[44]. However, authors have reported high levels of biliary amylase in neonates with choledochal cysts and low levels of biliary amylase in older pediatric patients with CC[45]. In newborns, the mean pancreatic amylase activity is 3% of that of adults, it begins to increase at seven month till reaching adult levels by five years[46].

In our study we measured and compared the levels of biliary amylase in adult and pediatric patents with choldochal cysts. We also found high levels of biliary amylase in pediatric group and low levels of biliary amylase in adult patients with choledocal cyst however no statistical difference were found between the two groups (P=0.72)

In conclusion, Pediatrics with choledochal cysts are more likely to be associated with female predominance, present by abdominal mass and Type 1a cyst whereas adult patients commonly present by abdominal pain, and are more often associated with APBDJ. Despite of the significantly higher incidence of stones presence in choledochal cysts in pediatric group, there was no significant difference in the incidence of gallstones or intrahepatic stones between both groups. Excision of the entire extra hepatic biliary tree and biliary enteric anastomosis with hepaticojejunostomy was the most commonly performed surgical procedure to treat extra hepatic cystic involvement. Chronic abdominal pain with recurrent mild cholangitis is the most common long-term complication in both groups.

Authors' declare no conflict of interest.
This research did not receive any specific grant from funding agencies in the public, commercial, or not-for-profit sectors.

REFERENCES

1. Singham J, Yoshida EM, Scudamore CH. Choledochal cysts: part 1 of 3: classification and pathogenesis. Canadian journal of surgery Journal canadien de chirurgie. 2009;52(5):434-40.
2. Miyano T, Yamataka A, Kato Y, Segawa O, Lane G, Takamizawa S, et al. Hepaticoenterostomy after excision of choledochal cyst in children: a 30-year experience with 180 cases. Journal of pediatric surgery. 1996;31(10):1417-21.
3. Yamaguchi M. Congenital choledochal cyst. Analysis of 1,433 patients in the Japanese literature. American journal of surgery. 1980;140(5):653-7.
4. Babbitt DP, Starshak RJ, Clemett AR. Choledochal cyst: a concept of etiology. The American journal of roentgenology, radium therapy, and nuclear medicine. 1973;119(1):57-62.
5. Arima E, Akita H. Congenital biliary tract dilatation and anomalous junction of the

ductal system. Observations. Archives of surgery (Chicago, Ill : 1960). 1962;84:229-46.
29. Shi LB, Peng SY, Meng XK, Peng CH, Liu YB, Chen XP, et al. Diagnosis and treatment of congenital choledochal cyst: 20 years' experience in China. World journal of gastroenterology. 2001;7(5):732-4.
30. Metcalfe MS, Wemyss-Holden SA, Maddern GJ. Management dilemmas with choledochal cysts. Archives of surgery (Chicago, Ill : 1960). 2003;138(3):333-9.
31. Nomura T, Shirai Y, Wakai T, Yokoyama N, Sakata J, Hatakeyama K. Narrow portion of the terminal choledochus is a cause of upstream biliary dilatation in patients with anomalous union of the pancreatic and biliary ducts. World journal of gastroenterology. 2005;11(41):6503-7.
32. Michaelides M, Dimarelos V, Kostantinou D, Bintoudi A, Tzikos F, Kyriakou V, et al. A new variant of Todani type I choledochal cyst. Imaging evaluation. Hippokratia. 2011;15(2):174-7.
33. Yoon JH. Magnetic resonance cholangiopancreatography diagnosis of choledochal cyst involving the cystic duct: report of three cases. The British journal of radiology. 2011;84(997):e18-22.
34. Conway WC, Telian SH, Wasif N, Gagandeep S. Type VI biliary cyst: report of a case. Surgery today. 2009;39(1):77-9.
35. De U, Das S, Sarkar S. Type VI choledochal cyst revisited. Singapore medical journal. 2011;52(5):e91-3.
36. Acker SN, Bruny JL, Narkewicz MR, Roach JP, Rogers A, Karrer FM. Preoperative imaging does not predict intrahepatic involvement in choledochal cysts. Journal of pediatric surgery. 2013;48(12):2378-82.
37. Martin RF. Biliary cysts: a review and simplified classification scheme. The Surgical clinics of North America. 2014;94(2):219-32.
38. Serena Serradel AF, Santamaria Linares E, Herrera Goepfert R. Cystic dilatation of the cystic duct: a new type of biliary cyst. Surgery. 1991;109(3 Pt 1):320-2.
39. Weiler H, Grandel A, Fruhmorgen P. [Congenital cystic dilatation of the cystic duct associated with an anomalous pancreaticobiliary ductal junction]. Ultraschall in der Medizin (Stuttgart, Germany : 1980). 2003;24(3):197-201.
40. Sethi S, Upreti L, Verma AK, Puri SK. Choledochal cyst of the cystic duct: Report of imaging findings in three cases and review of literature. The Indian journal of radiology & imaging. 2015;25(3):315-20.
41. Oguchi Y, Okada A, Nakamura T, Okumura K, Miyata M, Nakao K, et al. Histopathologic studies of congenital dilatation of the bile duct as related to an anomalous junction of the pancreaticobiliary ductal system: clinical and experimental studies. Surgery. 1988;103(2):168-73.
42. Schweizer P, Schweizer M. Pancreaticobiliary long common channel syndrome and congenital anomalous dilatation of the choledochal duct--study of 46 patients. European journal of pediatric surgery : official journal of Austrian Association of Pediatric Surgery [et al] = Zeitschrift fur Kinderchirurgie. 1993;3(1):15-21.
43. Imazu M, Iwai N, Tokiwa K, Shimotake T, Kimura O, Ono S. Factors of biliary carcinogenesis in choledochal cysts. European journal of pediatric surgery : official journal of Austrian Association of Pediatric Surgery [et al] = Zeitschrift fur Kinderchirurgie. 2001;11(1):24-7.
44. Lebenthal E, Lee PC. Development of functional responses in human exocrine pancreas. Pediatrics. 1980;66(4):556-60.
45. Jung SM, Seo JM, Lee SK. The relationship between biliary amylase and the clinical features of choledochal cysts in pediatric patients. World journal of surgery. 2012;36(9):2098-101.
46. Gillard BK, Simbala JA, Goodglick L. Reference intervals for amylase isoenzymes in serum and plasma of infants and children. Clinical chemistry. 1983;29(6):1119-23.