

Choledochal Cysts in Children: A Retrospective, Single Institutional Intermediate - Term Report

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Abstract

Aim:

To study the clinical and management aspects of pediatric choledochal cysts (CC).

Methods:

This is a retrospective study conducted in the department of pediatric surgery at our centre comprising 28 children with choledochal cysts treated surgically from March 2009 to January 2020. We had reviewed the data of patients of CC attending the follow up clinic and documented the information like demographic data, age of presentation, gender, clinical presentation, preoperative diagnosis (type of CC), intraoperative findings, type of surgery done, preoperative and postoperative bilirubin levels, complications and duration of follow up. Study includes all children up to 15 years of age with CC who were diagnosed and managed at our hospital. We had excluded all children with CC operated elsewhere but are being followed at our hospital.

Results:

Our study comprises 28 children with CC with an average age 6.73 years (ranging from 1 month to 15 years), majority (42.8%) were below 4 years of age. Among clinical features, abdominal pain was most common [85.7% (n=24)] followed by jaundice (n=16), clay colored stool (n=11), lump (n=12) and the classic triad (pain, jaundice and lump) was seen only 6 (21.4%) children. 14.2% (n=4) children presented with pancreatitis. Three children were diagnosed antenatally. Cyst excision with Roux-en-y Hepatico-jejunostomy (RYHJ) was performed in 82.1%, (n=23) and cyst excision with Hepatico duodenostomy (HD) was done in 5 (17.8%) children. Mean operative time for HD was 2 hours 17 minutes where as for RYHJ was 3 hours 13 minutes. Cholangitis (n=3), pancreatitis (n=4), gastritis (n=2), wound infection (n=2) and anastomotic leak (n=2) were among the complications. Mean follow up period was 57.9 months ranging from 3 months to 106 months.

Conclusion

Prevalence of CC is relatively low. Occurrence of classic triad of pain, lump and jaundice is rare. With increased usage of USG it is now possible to diagnose CC antenatally. HD is easy to perform and less time consuming than RYHJ and requires long term follow up with endoscopy for bile reflux. Malignancy occurs late in the adulthood hence requires long follow up.

Key words:

Choledochal cyst, Roux-en-y Hepatico-jejunostomy, Hepatico duodenostomy

Introduction

Choledochal cyst (CC) is a congenital cystic dilatation of biliary tract which was first described by Vater and Ezler in 1723 [1]. CC is a rare entity with an incidence of 1 in 100,000-150,000 live births in the West [2] and one in 1,000 live

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births in Asian countries with higher incidence (nearly two third) from Japan [3]. Its primarily a disease of pediatric population and it occurs nearly three times more commonly in females [4]. Exact etiopathogenesis of CC is not known, most accepted theory is reflux of pancreatic enzymes into biliary tract, resulting in inflammation, ductal wall weakness and cystic dilatation due to anomalous pancreaticobiliary duct union (APBDU) proposed by Babbitt in 1969 [5].

We present a case series of CC with the demographic data, different clinical presentations, complications, management in a tertiary care centre with an intermediate-

term follow up.

Methods

This is a retrospective study conducted in the department of pediatric surgery at our centre comprising 28 children with choledochal cysts treated surgically from March 2009 to January 2020. We had reviewed the data of patients of CC attending the follow up clinic and documented the information like demographic data, age of presentation, gender, clinical presentation, preoperative diagnosis (type of CC), intraoperative findings, type of surgery done, preoperative and postoperative bilirubin levels, complications and duration of follow up. Study includes all children up to 15 years of age with CC who were diagnosed and managed at our hospital. We had excluded all children with CC operated elsewhere but are being followed at our hospital.

Results

Our study comprises 28 children with CC with an average age 6.73 years (ranging from 1 month to 15 years), majority (42.8%) were below 4 years of age (Figure 1). Twelve children were females and 16 were males. Among clinical features (Table 1) most common feature was abdominal pain constituting about 85.7% (n=24) followed by jaundice (n=16), Clay colored stool (n=11) lump (n=12) and the classic triad (pain, jaundice and lump) was seen only in 6 (21.4%) children. 14.2% (n=4) of children presented with pancreatitis. Three children were diagnosed antenatally. Type I CC was the most common type (82.1%, n=23) followed by type IVa CC (17.8%, n=5). Cyst excision with Roux-en-y Hepatico-jejunostomy (RYHJ) was performed in 82.1%, (n=23) and cyst excision with Hepatico-duodenostomy (HD) was done in 5 (17.8%) children. Mean operative time for HD was 2 hours 17 minutes where as for RYHJ was 3 hours 13 minutes. Mean preoperative direct bilirubin (DB) level was 2.38 mg/dL (range 0.6 to 9.4mg/dL) and mean post-operative bilirubin level was 0.7 mg/dL ranging from 0.1 to 4.1 mg/dL. Cholangitis (n=3), pancreatitis (n=4), gastritis (n=2), wound infection (n=2) and anastomotic leak (n=2) were among the complications. Mean follow up period was 57.9 months ranging from 3 months to 106 months.

Table 1: Clinical features in this series

Clinical Presentation	No. of patients n=28 (%)
Abdominal pain	24 (85.7)
Jaundice	16 (57.4)
Upper abdominal lump	12 (42.8)
Clay colored stool	11(39.2)
Antenatal detection	3(10.7)
Pancreatitis	4 (14.2)

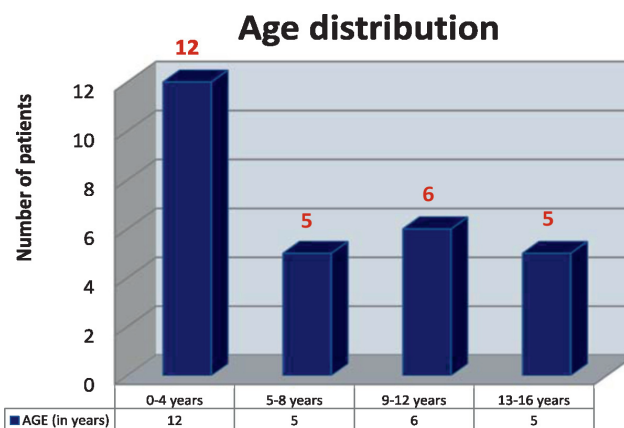


Figure 1: Age Distribution

Discussion

Alonso-Lej and colleagues [6] described three types of bile duct dilatation in 1959, Todani and colleagues [7] expanded this system and classified CC into five types in 1977. Type-I cysts were further divided into 3 types (type IA- cystic dilation of the entire extrahepatic biliary tree, with normal intrahepatic ducts, type IB- focal, segmental dilation of the extrahepatic bile duct, type IC- smooth fusiform dilations of the entire extrahepatic bile duct, extending from the pancreaticobiliary junction to the intrahepatic biliary tree), Type II - diverticulum of the extrahepatic duct connected to the CBD with a narrow stalk, Type III- distal intramural dilation of the common bile duct within the duodenal wall, Type IVA- multiple intrahepatic and extrahepatic dilations, Type IVB - multiple dilations of the extrahepatic biliary tree alone, Type V - (Caroli Disease) multiple cystic dilations of the intrahepatic bile ducts alone [8]. Standard management of CC is cyst excision with hepatico-enterostomy [9].

Mean age of presentation in our study was 6.73 years, *MD Stringer et al* reported that nearly two third of these were below 5 years [10]. In our study, 42.8% (n=12) were under the age of 4 years. 75 % of CCs are diagnosed by first decade of life.[11] In our series 67.8% of children were under 10 years. *Rangsan Niramis MD et al* reported 72.5% of females with CC in his study [12], In our study, only 42.8% (n=12) were females. Most common way of presentation (more than 75%) is abdominal pain.[13] Most common clinical feature in our series was abdominal pain (85.7%, n=24). Reported occurrence of classic triad of pain, lump and jaundice is only about 6- 12%.10, 12 In our series classic triad was seen in about 21.4% (n=6).

Prenatally, CC can be diagnosed by 20 weeks of gestational age, though CBD can be detected by ultrasonogram as early as 15 - 16 weeks [14]. It appears as anechoic fluid filled structure at the porta. We had diagnosed 3 cases prenatally between 28 and 32 weeks. We suspected either a cystic biliary atresia or a choledochal cyst, postnatally evaluated and found to have type I CC in all three cases. Early

intervention in these children had facilitated in easy dissection and excision of cyst as there were no cholangitis episodes (figure 3). Hence prenatal detection and early intervention in infancy can result in better outcome. The complications associated with CC are Cholangitis, pancreatitis, hepatocholelithiasis and spontaneous perforation [15]. In our series 14.2% (n=4) of children presented with pancreatitis preoperatively which were managed conservatively and about 10.7% (n=3) children developed cholangitis postoperatively responded well to antibiotics. Most common type of CC is type I (80-90%) [16]. In this series, we report 82.1%, (n=23) children had type I and (17.8%, n=5) had type IVa. Primary cyst excision with biliary reconstruction can be by means of Roux-en-Y hepaticojejunostomy (RYHJ) or hepaticoduodenostomy (HD) [17] or jejunal Inter position graft [18]. Cyst excision and biliary reconstruction can be performed either by open technique, laparoscopic or robotic assisted techniques [19]. Open cyst excision with Hepatico-duodenostomy (HD) was done in 5 children (17.8%) and excision with Roux-en-y Hepatico-jejunostomy (RHJ) in 23 (82.1%) children. Both were performed by a single surgeon. HD was done initial cases, no special criteria taken into consideration to select cases for HD procedure, it was purely surgeon's choice. Advantages of HD procedure are technically easy to perform, more physiological than HJ and amenable for endoscopic intervention if required [20]. HD also consumes less time to operate, the mean operative time for HD was 2 hours 17 minutes where as for RYHJ was 3 hours 13 minutes.

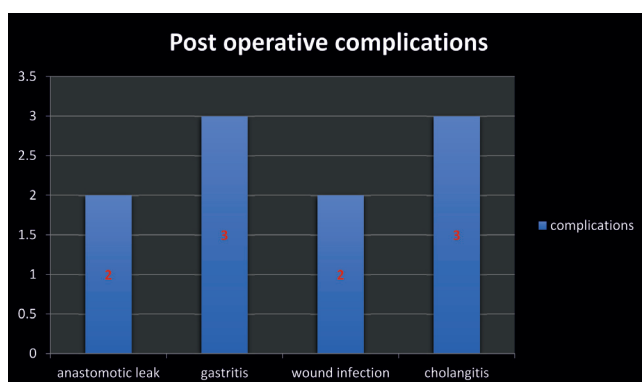


Figure 2: Complications in this series

Three children in our series presented with gastritis following HD. Upper Gastro-intestinal (UGPI) endoscopy revealed pangastritis due to biliary reflux which got subsided by oral Pantoprazole for 2 weeks in both the cases. Repeat endoscopy after 3 months was normal. It has been reported that about 3.8-33% of children present with biliary gastric reflux following HD [21,22]. Tarun John K Jacob et al reported zero cases of biliary gastric reflux in about 35 cases of HD with a mean follow up of 17.5 months (Table 2) [23].



Figure 3 : Choledochal cyst (black arrow) in a 2 months old infant. [note: no adhesions (signs of previous cholangitis) are visible]

Anastomotic leak is an important complication that is associated significant morbidity and mortality following a biliary-enteric anastomosis. Sarath Kumar Narayanan et al performed a systematic review and meta-analytic study and reported the occurrence of anastomotic leak following HD is about 2.1% [24] and it has been reported that a higher incidence of biliary leak following RHJ which was about 2.5 – 10% [25]. Two (7.14%) children in our series had anastomotic leak following HJ, both were initially treated conservatively. One child which did not respond to conservative therapy and required re-exploration, performed thorough wash and kept a transjejunal tube (though roux limb) at the bifurcation of hepatic ducts to drain bile and leak subsided after 7 days. Another child with leak was managed by aspiration of perianastomotic fluid under Ultrasound guidance and antibiotics. The mean follows up period was 57.9 months ranging from 3 months to 106 months.

During the follow up emphasis was given for physical examination, liver function tests and USG abdomen during. The reported incidence of malignancy in CC is about 10% [26]. Srinivasan et al recently reported two cases (5.5%) of malignancy in CC occurring at the age of 46 and 67 years [27]. Malignancy can occur anywhere along the biliary tract usually in the dilated segments of biliary tract before surgery, near the anastomotic site and from residual biliary epithelium in liver and pancreas after resection [28]. The eldest patient of our series being under follow up is 23 years old at present, no of malignancy reported in our series so far, but since the evidence shows malignancy occurs late in the adulthood, we need strict long term follow up for detection of malignancy and early intervention.

Prevalence of CC is relatively low. Compared to literature we report choledochal cysts are slightly higher in males. Occurrence of classic triad of pain, lump and jaundice is

Table 2: Clinical presentation and postoperative complications in relation to HD and RYHJ

Sl. No.	Clinical Parameter	Present Series		Tarun John K Jacob et al ²²	
		Hepatico-duodenostomy (n=5) (%)	Roux en y hepatixojunostomy (n=23) (%)	Hepatico-duodenostomy (n=35) (%)	Roux en y hepatixojunostomy (n=35) (%)
Presenting Complaint					
1.	Cholangitis	0 (0)	0 (0)	4 (11.4)	4 (11.4)
2.	pancreatitis	1(20)	3 (13)	2 (5.7)	3 (8.54)
3.	vomiting	0 (0)	0 (0)	6 (17.1)	3 (8.5)
4.	Abdominal lump	4 (80)	8 (34.7)	3 (8.5)	2 (5.7)
5.	Abdominal pain	4 (80)	20 (86.9)	26 (74.2)	24 (68.5)
6.	Jaundice	3 (60)	13 (56.5)	13 (37.1)	17 (48.5)
7.	Antenatally	1 (20)	2 (8.6)	2 (5.7)	0 (0)
Post-Operative Complications					
1.	Wound infection	2 (40)	0 (0)	1 (2.8)	1 (2.8)
2.	Bile reflux (gastritis)	3 (60)	0 (0)	0 (0)	0 (0)
3.	Anastomotic leak	0 (0)	2 (8.6)	0 (0)	0 (0)
4.	cholangitis	3 (60)	0 (0)	0 (0)	4 (11.4)
5.	Pancreatitis	0 (0)	0 (0)	3 (8.5)	4 (11.4)
6.	Pelvic abscess	0 (0)	0 (0)	0 (0)	1 (2.8)

rare. With increased usage of USG it is now possible to diagnose CC antenatally and early intervention in infancy result in minimal intra-operative and post-operative complications. Type I CC is the most common type. HD is easy to perform and less time consuming but requires long term follow up with endoscopy for bile reflux. Children with CC frequently present with complication like cholangitis and pancreatitis and can be managed conservatively. Malignancy occurs late in the adulthood hence requires long follow up.

Conflict of interest:	All authors declare no COI
Ethics:	There is no ethical violation as it is based on voluntary anonymous interviews
Funding:	No external funding
Guarantor:	Dr. Vijay Kumar Kundal will act as guarantor of this article on behalf of all co-authors.

References

- Vater A. Dissertatio de scirrhis viscerum occasione sectionis viri typanite defunte. Wittenburgae, 4 Pamphlers. 1723; 881:22.
- Lee HK, Park SJ, Yi BH et al. Imaging features of adult choledochal cysts: a pictorial review. Korean J Radiol. 2009;10:71–80.
- Hung MH, Lin LH, Chen DF, Huang CS. Choledochal cysts in infants and children: experiences over a 20-year period at a single institution. Eur J Pediatr 2011; 170:1179-85.
- Lipsett PA, Pitt HA. Surgical treatment of choledochal cyst. J Hepatobiliary Pancreat Surg 2003;10:352–59.
- Babbit DP. Congenital choledochal cyst: new etiological concept based on anomalous relationships of the common bile duct and pancreatic bulb. Ann Radiol. 1969;12:231-40.
- Alonso-Lej F, Rever WB, Pessango DJ. Congenital choledochal cyst, with a report of 2, and analysis of 94 cases. Int Abstr Surg 1959;108:1-30.
- Todani T, Watanabe Y, Narusue M et al. Congenital bile duct cysts: classification, operative procedures, and review of thirty-seven cases including cancer arising from choledochal cyst. Am J Surg 1977;134:263-9.
- Singham J, Yoshida EM, Scudamore CH. Choledochal cysts: part 1 of 3: classification and pathogenesis. Can J Surg. 2009;52(5):434.
- Todani T, Narusue MI, Watanabe Y et al. Management of congenital choledochal cyst with intrahepatic involvement. Ann Surg. 1978;187(3):272.
- Stringer MD, Dhawan A, Davenport M et al. Choledochal cysts: lessons from a 20 year experience Arch Dis Child. 1995;73(6):528-31.
- Shah O, Shera A, Zargar S et al. Choledochal Cysts in Children and Adults with Contrasting Profiles: 11-Year Experience at a Tertiary Care Center in Kashmir. World J Surg. 2009;33(11):2403-2411.
- Niramis R, Narumitsuthon R, Watanatittan S et al. Clinical differences between choledochal cysts in infancy and childhood: an analysis of 160 patients. J Med Assoc Thai. 2014;97(11):122-8.
- De Vries J, de Vries S, Aronson D et al. Choledochal cysts: Age of presentation, symptoms, and late complications related to Todani's classification. J Pediatr Surg. 2002;37(11):1568-73.
- Schroeder D, Smith L, Prain HC: Antenatal diagnosis of choledochal cyst at 15 weeks' gestation: Etiologic implications and management. J

- Pediatr Surg. 1989;24:936-38.
- 15) Lal R, Agarwal S, Shivhare R, Kumar A, Sikora S, Kapoor V et al. Management of Complicated Choledochal Cysts. *Dig Surg.* 2007;24(6):456-462.
 - 16) Soares K, Goldstein S, Ghaseb M, Kamel I, Hackam D, Pawlik T. Pediatric choledochal cysts: diagnosis and current management. *Pediatr Surg Int.* 2017;33(6):637-650.
 - 17) Cosentino CM, Luck SR, Raffensperger JG, Reynolds M. Choledochal duct cyst: resection with physiologic reconstruction. *Surgery.* 1992;112(4):740-747.
 - 18) Okada A, Ohguchi Y, Kamata S, et al. Jejunal interposition hepaticoduodenostomy for congenital dilatation of the bile duct (choledochal cyst). *J Pediatr Surg* 1983;18(5):588-91.
 - 19) Xie, X., Li, K., Wang, J. et al. Comparison of pediatric choledochal cyst excisions with open procedures, laparoscopic procedures and robot-assisted procedures: a retrospective study. *Surg Endosc* 2020;34:3223–3231.
 - 20) Narayanan S, Chen Y, Narasimhan K, Cohen R. Hepaticoduodenostomy versus hepaticojejunostomy after resection of choledochal cyst: A systematic review and meta-analysis. *J Pediatr Surg.* 2013;48(11):2336-2342.
 - 21) Liem N, Pham H, Dung L, Son T, Vu H. Early and Intermediate Outcomes of Laparoscopic Surgery for Choledochal Cysts with 400 Patients. *J Laparoendosc Adv Surg Tech A.* 2012;22(6):599-603.
 - 22) Shimotakahara A, Yamataka A, Yanai T, et al. Roux-en-Y hepaticojejunostomy or hepaticoduodenostomy for biliary reconstruction during the surgical treatment of choledochal cyst: which is better? *Pediatr Surg Int* 2005;21(1):5-7.
 - 23) TJ KJ, Jehangir SH, Sebastian T, Karl IS. Comparison of hepaticojejunostomy with hepaticoduodenostomy techniques for biliary reconstruction after choledochal cyst excision in children. *Trop Gastroenterol.* 2017;38(1):42-6.
 - 24) Narayanan SK, Chen Y, Narasimhan KL, Cohen RC. Hepaticoduodenostomy versus hepaticojejunostomy after resection of choledochal cyst: a systematic review and meta-analysis. *J. Pediatr. Surg* 2013;48(11):2336-42.
 - 25) Santore MT, Behar BJ, Blinman TA, et al. Hepaticoduodenostomy vs hepaticojejunostomy for reconstruction after resection of choledochal cyst. *J Pediatr Surg* 2011;46(1):209-13.
 - 26) Gong L, Qu Q, Xiang X, Wang J. Clinical analysis of 221 cases of adult choledochal cysts. *Am Surg.* 2012;78:414–418.
 - 27) Honnavara Srinivasan P, Anbalagan A, Shanmugasundaram R, Obla Lakshmanamoorthy N. Management of Choledochal Cysts at a Tertiary Care Centre: A Nine-Year Experience from India. *Surg Res Pract.* 2020;2020:1-8.
 - 28) Friedmacher F, Ford KE, Davenport M. Choledochal malformations: global research, scientific advances and key controversies. *Pediatr. Surg. Int.* 2019; 35(3):273-82.

