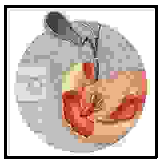

ORIGINAL PROF-657

CHOLEDOCHAL CYST;
RECENT EXPERIENCE

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ABSTRACT

OBJECTIVES: To evaluate clinical presentation, morphology, associated hepatobiliary problems and results of surgical excision of choledochal cyst. **PATIENTS AND METHODS**: Eight patients underwent surgery for choledochal cysts between Nov 2000 to Nov 2001 in Bangabandhu Sheikh Mujib Medical University Hospital, Shahbag, Dhaka Bangladesh. The diagnosis of choledochal cyst was based on clinical presentation, USG, radiological and operative findings. **RESULTS**: Total of 8 patients with choledochal cyst were identified. 7 patients were female and 1 was male. Age range was between 12 to 55 years. Presenting symptoms were episodic abdominal pain in 8 (100%), intermittent jaundice in 4 (50%), jaundice and fever in 3 (37.5%) and abdominal lump in 2 (37.5%) patients. LFT and USG were the initial diagnostic procedures in all patients, followed by ERCP in 4 (50%) and CT in 5 (62%) cases, MRCP in 1 (12.5%) case. Based on USG and Radiological imaging, cysts were classified according to Rodani's classification. Out of eight, 7 (87.5%) were type I and 1 (12.5%) was type IVA. 5 (62.5%) patient had surgical excision and Roux-en-y hepatico-jejunostomy, 1 (12.5%) had cysto-duodenostomy, 1 (12.5%) per-cutaneous drainage of cyst and one patient (12.5%) with type IVA under went excision of extra hepatic cyst and hepaticojejunostomy and one patient died before definitive operation due to liver failure. **CONCLUSIONS**: Choledochal cyst is uncommon cause of bile duct obstruction. Type I is the commonest presentation and recognized treatment is the total excision of cysts with Roux-en-y hepatico-jejunostomy.

INTRODUCTION

Cysts of the biliary duct system are uncommon, and the understanding of them is incomplete. About 80% are diagnosed during child hood, and the remainder became apparent in adult hood[1]. There have been several

descriptions and classifications based upon the location and anatomy of the choledochal cyst. Among most useful is Todani's modification of the classification proposed[2,3] (Table II).

Choledochal cyst are unusual cause of biliary obstruction with up to 85% of reported case being the type I variety, that is fusiform dilatation of the common bile duct. Choledochal cysts are more common in females than males (4:1)[4]. The usual presentation is that of episodic abdominal pain, often recurrent over months or years, generally associated with only minimal jaundice that may escape detection. If the condition persist unrecognized, sequella including cholangitis, cirrhosis and portal hypertension are almost inevitable[5].

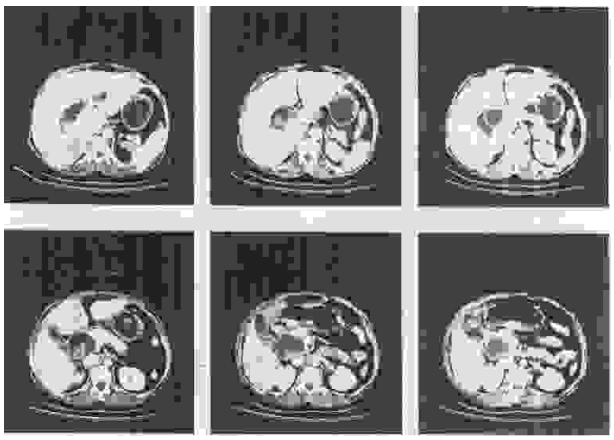
This is a small but well-studied series addressing the unusual cause of biliary obstruction. This study is presenting our recent experience with seven cases of choledochal cysts. Aims of the study were to evaluate clinical presentation, morphology, associated hepatobiliary problems and results of surgical excisions.

PATIENTS & METHODS

Eight patients underwent surgery for choledochal cyst between Nov 2000 to Nov 2001 in Bangabandhu Sheikh Mujib Medical University Hospital, Shahbag, Dhaka Bangladesh. The diagnosis of choledochal cyst was based on clinical symptoms, characteristics of USG, Radiological and operative findings. Cysts were grouped according to Todani's modification of the Alonso-Lej classification(table III).

RESULTS

A total of 8 patients with choledochal cyst were identified. 7 (87.5%) patient were female and 1 (12.5%) was male. Age range was between 12 to 55 years. Presenting symptoms were episodic abdominal pain in 8 (100%), intermittent, Jaundice in 4 (50%), episodic abdominal pain, jaundice and fever in 3 (37.50) and abdominal lump in 3 (37.5%) patients (table I).

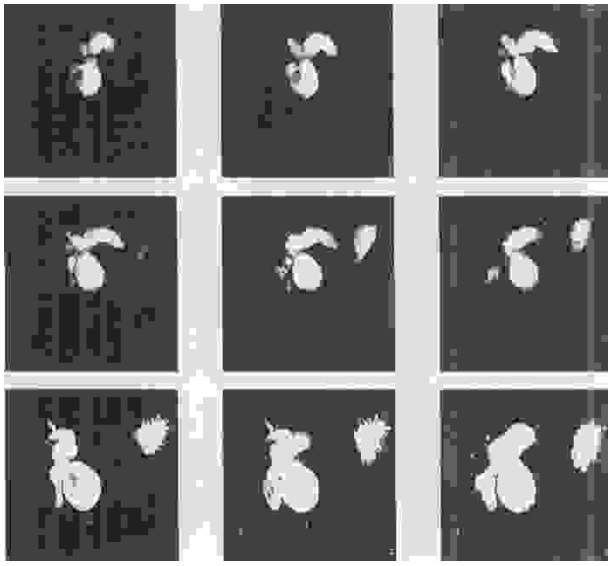


Patient Name = Maleka
Hosp. Reg No. = YC 0094286
Imaging Film = Selected CT scans of the abdomen showing a choledochal cyst with a CBD stone within the cyst.

Table-I. Clinical features (n=8)

	No of patients	%age
Age (13-55 years)	-	-
Male	1	12.5%
Female	7	87.5%
Symptoms & Signs		
Episodic abdominal pain	8	100%
Intermittent jaundice	4	50%
Abdominal lump	3	37.5%
Intermittent abdominal pain, jaundice & fever	3	37.5%

LFT and USG were the initial diagnostic procedures in all patients, followed by ERCP in 4 (50%) & CT in 5 (62.5%) cases & MRCP in 1 (12.5%) case. Based on USG and Radiological imaging, cysts were classified according to Todani's classification (table III)



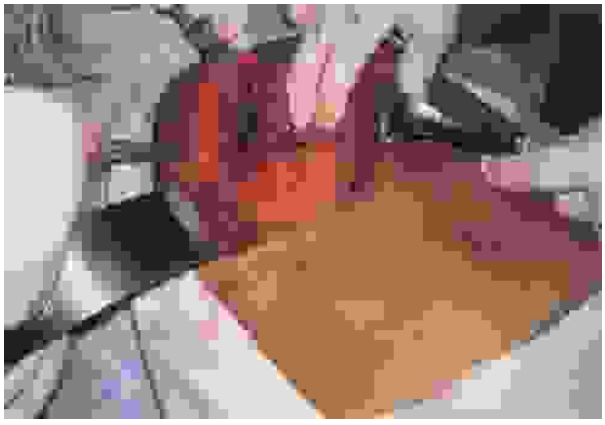
Patient Name = Maleka
Hosp. Reg No = YC 0094286
Imaging Film = Selected MRCP images showing the choledochal cyst with a CBD stone within the cyst. The proximal limit of the cyst appears to be at the level of the bifurcation of the hepatic ducts, with secondary dilatation of the right and left hepatic ducts.

Table-II. Todani's classification

Types	Classification
Type I	Fusiform dilatation of extrahepatic bile duct
Type II	Single secular dilatation or diverticulum of extrahepatic bile duct
Type III	Dilatation of intraduodenal portion of bile duct
Type IVa	Combined intra and extra-hepatic dilatation of bile duct
Type IVb	Multiple dilatations of the extra-hepatic bile duct
Type V	Isolated or defuse intra-hepatic biliary

Out of eight, 7(87.5%) were type I and 1 (12.5%) was type IVA. 5(62.5%) patients underwent surgical excision and Roux-en-Y hepatico-jejunostomy, 1 (12.5%) patient cystoduodenostomy, 1(12.5%) patient with type IVA underwent excision of extraheptic cyst and hepatico-jejunostomy and 1(12.5%) per-cutaneous drainage of cyst (table IV).

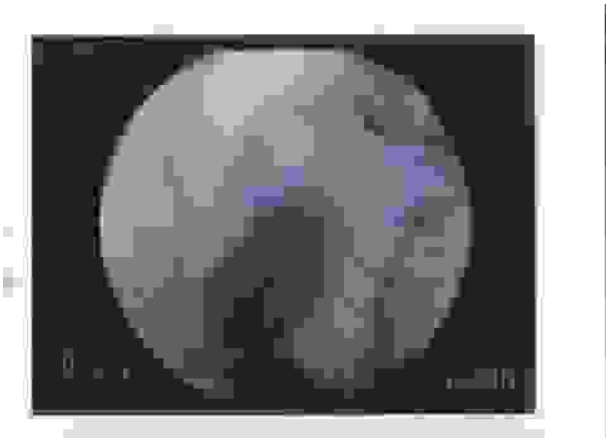
Antwa Choledochal Cyst



Antwa Choledochal Cyst

One patient who had per-cutaneous drainage of cyst died before definitive operation due to liver failure(table V).

Histopathology of cyst wall was done in 7 cases. None of these cases revealed malignant change.



ERCF finding of Choledochal Cyst

Table -III. Type of cysts (n=8)

Type	No of patients	%age
Type I	7	87.5%
Type II	-	-
Type III	-	-
Type IVa	1	12.5%
Type IVb	-	-
Type V	-	-

Table-IV. Surgical procedures (n=8)

	No of patients	%age
Excision of cyst and Roux-en-Y hepatico-jejunostomy	5	62.5%
Excision of cyst and hepaticoduodenostomy	1	12.5%
Cystoduodenostomy	1	12.5%
Per-cutaneous drainage of cyst	1	12.5%

Table-V. Morbidity and mortality (n=8)

	No of patients	%age
Death	1	12.5%
Biliary leakage	2	25%
Intra-abdominal bleeding	1	12..5%
Minor wound infection	1	12.5%

DISCUSSION

Choledochal cysts, originally recognized by Douglas in 1852 is rare anomaly of the biliary tree, characterized by abnormal dilatation of part of the extra-hepatic and or intra-hepatic biliary tree[6]. Incidence is in the order of 1:100,000-150,000 live births, three to four times more frequently in females[7].Although the majority of these patients present in infancy and childhood, about 20-30 percent of patients are diagnosed in adult[1]. The aetiology of choledochal cysts is controversial; One tenable accepted explanation is that proposed by Babbitt in 1969[8]. He incriminated an abnormal pancreatic biliary duct junction with the formation of a "common channel" into which pancreatic enzyme secretion are discharged with resultant weakening of the bile duct wall by gradual enzymatic destruction leading to dilatation, inflammation, and finally cysts formation[4].Seven patients in this series are female and 1 male with age range from 12 to 55 years. Symptoms of choledochal cysts are characteristically intermittent but progressive. Both children and adults present with abdominal pain and jaundice. In present series patient presented with episodic abdominal pain in 8 (100%) cases, intermittent jaundice in 4 (50%) cases and abdominal lump in 3 cases. The classical triad of abdominal pain, jaundice and fever was seen in 3 (37.5%) cases. Kabra V et al reported fever patients presenting with the classical triad[9]. Similar study by Tan and Howward[10] reported that classical triad presentation as in 24% of their patients. Present study showed that type I is common lesion (87.5%). Scudamore CH & Hemming AW[11]. reported similar findings and suggesting upto 85% of reported cases being of type I variety, that is fusiform dilatation of the common bile duct in their study.

This anomaly may be associated with anomalies f the pancreatic duct suggesting that choledochal cysts in this group represent an anomaly of entire pancreaticobiliary system rather than just the biliary tract[12].

Hepatobiliary problems such as cystolithiasis, cholelithiasis, secondary biliary cirrhosis and cholangio-

carcinoma are seen mainly in adults[13,14].

One patient in our series presented with secondary biliary cirrhosis which lead to liver failure and subsequently patient died before definitive surgical management and another patient had cholelithiasis and cystolithiasis.

In previous decades, it was thought that internal drainage of choledochal cysts was adequate. It became evident however that after internal drainage, late morbidity of 30% to 50% resulted with complications such as cholangitis, secondary biliary cirrhosis and development of cholangiocarcinoma[15]. The treatment of choice is now surgical excision and biliary reconstruction with several exceptions[16,17,17]. In our series with type I cysts, five patients underwent total cyst excision with Roux-en-Y hepatico-jejunostomy and one patient underwent cystoduodenostomy. One patient with type IVA cyst under went complete excision of the extra hepatic portion of the cysts with hepatico-jejunostomy. In another we could not do any definitive surgery because patient was very toxic due to severe cholangitis, secondary liver failure and huge abdominal distension. Only percutanecous drainage was performed. Thambi et al (1991)[19] reported five cases with types IVA who underwent extra hepatic cysts excision and Roux-en-Y hepatico-jejunostomy. They reported that the intra hepatic dilatation regressed after excision of extra hepatic cyst just below the hilum of the liver. Out of eight, one patient died before operation and 2 patients developed biliary leakage which was treated conservatively (table V). One patient had postoperative internal bleeding and abdomen was re-explored.

CONCLUSIONS

Choledochal cysts are unusual cause of biliary duct obstruction. Both intra hepatic and extra hepatic biliary duct systems are affected. But type I, that is fusiform dilatation of extra hepatic duct is the commonest lesion. Females are affected more that male. Recognized treatment is the total excision of cysts with Roux-en-Y - hepatico-jejunostomy.

REFERENCES

1. David L, Nahrworld MD. he biliary system. Text book of surgery,, Vol I 5th ed WB Saunders company, Philadelphia, 1997: 1117.

2. Alonso-Lej F, FeVer WB Jr, Passagno DJ. Congenital choledochal cysts, with report of 2 and analysis of 94 cases. Surg. Gynaecol-Obstet 1959.; 108: 1-130.

3. Todani's T, Watanabe Y, Narusue M et al. Congenital bile duct cyst. Am J Surg, 1977; 134: 263-269.

4. Schwartz SI, Shires GT, Spencer FC, Husser WC. Editors. Choledochal cyst-chapter 29 Gall and Extra-hepatic biliary system. Principles of surgery 6th ed. Mcgraw. Hill, Inc. Newyork, 1994; 1387-1388.

5. Oweida SW, Ricketts RR. Hepatico-Jejunoduodenostomy reconstuction following excision of chaledochal cysts in children. Am surg, 1989; 55(1): 2-6.

6. Edward GJ, Bairibre LC. Paediatric hepatobiliary disorders-Surgery aspect. Oxford Text Book of Surgery 1st edn. Oxford Medical Publication, Newyork, 1994; 2:2053-55.

7. A Chaudhary, Dhar P et al. Choledochal cysts.Difference in children and Audlts, Br J. Surgery, 1996; 83: 186-188.

8. Babbit DP. Congenital cysts. New etiological concept based on anomalous relationships of the common bile duct and pancreatic duct bulb. Ann Radiol, 1969; 12: 231-240.

9. Kabra V, Agarwal M et al. Choledochal Cyst. a changing patern of presentation. AZN J Surg, 2001; 71(3): 159-61.

10. Tan KC and Howard ER. Choledochal cysts. a 14 years surgical experience with 36 patients. Br. J. Surg 1988; 75: 892-5.

11. Charles H. Scudamore MD,Alan W. Hemming MD, Julian P. Teare, P, Fache JS, Erb SR, Watkinson AF. Surgical management of choledochal cysts. Am J. Surgeyr, 1994; 167(S): 197-200.

12. Rattner DW, Schapiro RH, Warshaw AL et al. Abnormalities of the pancreatic and biliary ducts in adult patients with choledochal cysts. Arch surg, 1983; 118: 68-73.

13. Orenstein SR, Whitington PF. Choledochal cyst

resulting in congenital cirrhosis. American journal of diseases of child, Mar 1982; 136: 1025-26,

14. Martvin LW, Rowe GA, Portal hypertension secondary to choledochal cysts. Ann surg, 1979; 190: 636-9.

15. O' Ncil JA. Choledachol cysts. Curr. Probl. Surg, 1992; 29: 65-410.

16. Joseph VT. Surgical techniques and long term results in the treatment of choledochal cysts, J Pediatric surg, 1990; 25: 782-787.

17. Nunez-Hoye M, Lee CD, Herman RF et al. Bile duct cysts experience with 15 patients. Am J surg, 1982; 144: 295-299.

18. Lopez RR, Pinson CW, Campbell SR et al. Variations in management based on type of choledochal cysts. Am J. Surg, 1991; 161: 61-612.

19. Thambi Dorai CR, Vsvanathan R, Mcall GL. Type Iva choledochal cysts. Surgical management and review. Aust NZJ Surg, 1991; 61(7): 505-10.