Paediatric Choledochal Cysts- Management of Thirty Seven Patients in Paediatric Surgery Department of Guntur Medical College

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ABSTRACT

Introduction: Paediatric choledochal cyst is an uncommon congenital malformation of the biliary tract, involving cystic dilatation of extrahepatic bile duct or intrahepatic bile duct systems or may be both. The aim of this study was to study the pattern of presentation, confirmation of diagnosis, treatment and results of Choledochal cyst.

Material and methods: We collected data of thirty seven patients. Thirty two were female patients and five were male patients. Type I cyst (saccular or fusiform dilatation of CBD) was the most common type. Their mean age at Operation was 39.84 months, youngest patient was12months and oldest patient was 133 months. Female preponderance is noted, with male to female ratio 1:6.4 .Thirty one patients underwent hepatico-jejunostomy. Five patients underwent Hepatico-dueodenostomy. One patient underwent porto-jejunostomy as hepatic ducts were very minute that they can admit only number 22 gauze cannula. Thirty three patients had above procedures as primary procedure. Four patients initially underwent cysto-jejunostomy, later hepatico-jejunostomy as definitive procedure.

Results: We analyzed data of thirty seven patients of Choledochal cysts. Postoperative complications developed in five patients (13.51%).Fortunately Hepatico-dueodenostomy patients did not develop any complications

Conclusion: No death reported in these thirty seven operated patients in our institute. No portal hypertension or biliary cirrhosis noted. No carcinogenesis detected during postoperative follow-up in the study group till now. Operated five Hepatico-dueodenostomy patients fortunately did not develop any complications. Overall complication rates are low, result are comparable to many centers. Good quality of life achieved.

Keywords: Paediatric Choledochal Cysts- Management, Paediatric Surgery

INTRODUCTION:

Choledochal cyst defined as cystic dilatation of the extra-hepatic or intra-hepatic bile ducts or both the ducts. Incidence is very rare in European and American population, one patient per one lakh-1.5 lakh population and more common in people of Asian origin ,with an incidence as high as of 1:13000. Female preponderance is seen (three to eight times more in female children than male children).The most accepted explanation of development of choledochal cyst is presence of anomalous pancreaticobiliary duct junction (APBDJ) (Babbitt's theory)[1]. Cholangiograms studies shows a long common pancreaticobiliary channel (2cms-3cms) in which an abnormal junction between the CBD and the pancreatic duct (normal common channel is 5mm or less). Long common channel may lead to reflux of pancreatic secretions in to CBD and results in proteolytic enzymes activation, damaging and weakening of the duct wall[2]. Another theory is Defective recanalization and epithelialization of bile ducts during the developing stage and leading to congenital weakness of the CBD. Recently, spasm and abnormal function of the sphincter of Oddi are implicated. The abnormal behavior of the sphincter of Oddi due to long common channel, which may lead to high pressure(30-50 cms) in pancreatic duct than CBD pressure(25-30 cms), causing reflux and cause choledochal cysts[3]. This functional obstruction at the level of sphincter of Oddi also predisposes to of pancreatic secretions into the biliary tree. Another theory is that hypoganglionosis in the narrow part of the common bile duct (CBD) leading to a proximal dilatation like Hirschsprung's disease and achalasia cardia[4]. Familial occurrence is rare[5]. It could be concluded from various evidences available that some anatomical or functional obstruction in distal CBD and increased intra-ductal pressure are the two most important factors. Komi et divided abnormal union of the pancreatico-bile ducts (AUPBD) into 3 types[6], type 1 is joining of the ducts at a right angle to each other, with or without dilatation of common channel, type2 is acute angle junction with or without dilatation, type3 is complex types with accessory pancreatic ducts Alonzo-Lej F et al divided the Choledochal cysts into three categories in (1959). Todani T et al added two more types . At present Todani classification is widely used[7]. Todani type I cysts are the most common and incidence is around 85%of choledochal cysts. They consist of fusiform or saccular dilatations of the common bile duct(CBD), which involve the entire duct or a segment of the duct. Type1A is saccular shape and may involves either the entire extra-hepatic bile duct or the most of the duct, Type1B is saccular shape and segmental involvement of the bile duct, Type 1C is more of fusiform shape and involving most or all of the extra-hepatic bile duct. Type2(2% incidence) choledochal

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cysts is a diverticulum from common bile duct. Cyst may be join to the CBD by a narrow stalk or wide attachment. Type 3 (4-5%incidence) choledochal cysts from the intraduodenal part of the common bile duct (also called as choledochocoele). Type 4 A cysts consist of multiple dilatations of the extrahepatic bile ducts and intrahepatic ducts. Type 4B choledochal cysts are with multiple dilatations of only the extra-hepatic bile ducts. Type 5 (Caroli disease) cysts are multiple dilatations confined to intrahepatic bile ducts only. Recently Type 6 added with cystic duct dilatation isolated or associated with other type cysts. The patients presentation varies depending on the age of the child . Most children with choledochal cysts are diagnosed during early childhood, although the condition may be detected at any age. During Infancy presentation may be with jaundice and acholic white colour stools. In early infancy, this may lead to a workup for biliary atresia. Child may present with a palpable mass in the right upper abdomen and along with hepatomegaly. Older children and adolescents may present with intermittent biliary colic or recurrent attacks of pancreatitis or mass abdomen with or without elevated amylase and lipase concentrations. A classic triad of abdominal pain, palpable abdominal mass, and jaundice has been found in only 10-20% of patients. Differential diagnosis includes acute pancreatitis and biliary obstruction. No laboratory test can diagnosis a choledochal cyst. Abdominal ultrasound is initial imaging of choice in the diagnosing of a choledochal cyst. Abdominal ultrasound also useful in the antenatal detection choledochal cyst in a fetus and may detect the Caroli disease as well. Abdominal contrast enhancing CT scanning can show the presence of the disease and intrahepatic ductal involvement extent. Magnetic resonance cholangiopancreatography (MRCP)[8, 9] was especially useful in defining abnormal pancreaticobiliary junctions and diagnostic reliability of magnetic resonance cholangiopancreatography (MRCP) is nearly cent percent. Endoscopic ultrasonography (EUS) is useful to differentiate among pancreatic cysts and choledochal cysts, especially in patients with type 2(diverticulum type) choledochal cysts. Percutaneous transhepatic cholangiography (PTC) useful in delineating extrahepatic or intrahepatic strictures and stones. Endoscopic retrograde cholangiopancreatography (ERCP) useful in detecting abnormal pancreatobiliary junction and detecting the extrahepatic as well as intrahepatic strictures and stones. Intraoperative cholangiography is very useful in cases emergency operations like perforation of cysts. Histologic findings[10] shows evidence of cyst wall chronic inflammation. The cyst wall will be thin, fibrous, and frequently without a true epithelial surface, may be lined by a columnar epithelium in some areas. Infants can develop complete obstructions of the distal common bile duct due to inflammatory changes. Liver also affected by ductal fibrosis and portal triad edema. The most feared histologic finding is the presence of cholangiocarcinoma on histologic examination. Biliary cirrhosis changes may be observed in older children and young adults with long-standing disease. Management Choledochal cysts treatment is one of the challenging tasks in paediatric departments which needs sound anatomy of blood vessels, biliary duct system and presence of innumerable variations as well as awareness of possibilities of life threatening vascular and ductal injuries during operative procedure. The operative treatment for choledochal cysts is complete excision. Antibiotic therapy (broad-spectrum) against common biliary pathogens and supportive treatment should be given to these patients with cholangitis. Patients with choledochal cysts must be on lifelong follow-up because of the high risk of cholangiocarcinoma in 2nd and 3rd decade[11], even after complete excision of the cyst. The operation for choledochal cysts must be complete excision of the cyst, without any remnant of cyst with formation of a biliary-enteric anastomosis to keep continuity with the gastrointestinal tract. Incomplete excision of the cyst as well as internal drainage procedures can lead to increased risks of cholangitis, pancreatitis, calculi and cholangiocarcinoma in long term. Various operative procedures are advocated are resection and Roux-en-Y hepaticojejunostomy[12] or hepaticoduodenostomy and operations by different modes reported like Laparoscopic choledochal cyst excision[13] or Robotically-assisted laparoscopic resection and open method. There are some studies comparing hepaticoduodenostomy with Roux-en-Y hepaticojejunostomy and proposed that hepaticojejunostomy was a preferred because of an high incidence of duodenogastric reflux into biliary tract as high as 33% in the hepaticoduodenostomy group[14]. Choledochal cyst excision by Laparoscopic method and Roux-en-Y reconstruction in children, is a safe, feasible and valid alternative to open excision. Recent addition is robotically assisted laparoscopic excision of choledochal cysts. Majority centers overall complication rate is around ten percent. Post-operative patients who developed cholangitis, may have strictures which may require some intervention for anastomotic stricture. Two percent of patients needed reexploration and postoperative deaths are around one percent according to literature. Reports of Choledochal cyst excision in the neonatal period was with lesser complications and minimal hepatic fibrosis, especially in neonates who underwent resection of a choledochal cyst in first one month of life. Treatment should be given cyst-related complications, like pancreatitis and sepsis, before investigating to define the cysts anatomy with ERCP or MRCP to prevent flare up of these conditions. Surgical procedures:

Surgical procedure

For choledochal cysts devised according to subtype of cyst. Type-1cysts -The treatment goal is complete excision of the dilated portion of the extrahepatic bile duct along with a Roux-en-Y hepaticojejunostomy to restore biliary-enteric continuity or hepaticoduodenostomy . Type 2(diverticulum type cysts)- The dilated diverticulum is completely excised ,defect is closed primarily or excision of entire duct system if wide attachment or associated with saccular or fusiform type of cyst Type3 (choledochocoele)- The option of therapy

depends upon size of the Choledochoceles. If the size of cyst is 3 cm or less can be treated with endoscopic sphincterotomy. If the Cyst is more than 3 cm usually produces some degree of duodenal obstruction and this type of cysts are removed operatively by a transduodenal approach. If the pancreatic duct joining the choledochocele it needs to be reimplanted into the duodenum after excision of the cyst. Type 4-Both subtype 4A and subtype 4B are treated by same procedure. The abnormal extrahepatic duct should be completely excised and restore the continuity by Roux-en-Y hepaticojejunostomy. Regarding Intrahepatic ductal disease in type 4A , it does not require specific therapy unless hepatic abscesses, intrahepatic ductal strictures and hepatolithiasis are present. In such cases, the affected segment liver or lobe of the liver should be resected. Type V (Caroli s disease)-Prognosis of this type of disease is poor .If the Caroli s disease is confined to left lobe it may be amenable to resection by left hepatic lobectomy, if Hepatic functional reserve Is acceptable. Patients with both lobes involvement, who begin to manifest signs of cirrhosis (biliary),with portal hypertension or with liver failure ,may require liver transplantation**.** Rarely Lilly technique may be used , the cyst adherent densely to the portal vein due to chronic inflammatory reaction. In such conditions, complete resection of the cyst may not be possible. In the Lilly technique, adventitia or serosal surface of the duct is left in situ, adhering to the portal vein, while the mucosa of the cyst wall is removed by cautery or curettage. Posterior and medial walls are dissected in the submucosal plane to avoid injury to the portal vein and hepatic artery respectively. Lilly technique used in two females children during hepatico jejunostomy. This may removes the risk of malignant change in that segment of the duct. If choledochal cysts are not excised completely, a high incidence of (9 to 28%) cholangiocarcinoma has been reported, mainly after the second decade of life, because this reason, complete excision is mandatory and surgical treatment is preferably by expertise. This is further supported by a study that found increasing rate of premalignant changes in resected cysts with advancing age. The aim of this study was to study the pattern of presentation, confirmation of diagnosis, treatment and results of Choledochal cyst.

MATERIAL AND METHODS

We collected data of patients with choledochal cysts admitted and operated in our paediatric surgery department of Government general hospital and Guntur Medical College, Guntur, India during May 2006 to November 2017. Commonest presenting symptom was pain abdomen in 29 patients (78.37%) followed by jaundice in six patients (16.21%). Classical triad of abdominal pain, jaundice, and mass abdomen was seen in three cases (8.10%).Age of presentation - youngest patient age was 12 months, oldest patient age was 133 months (mean 39.84months).one patient was diagnosed by prenatal ultrasonography and on follow up and operated at one year age for gaining 10 kg weight as our preference to wait for around 10 kg weight unless there are indications for early intervention, to withstand the prolonged anesthesia time. We collected data of thirty seven patients. Thirty two (86.48%) were female patients and five (13.51%) were male patients. Thirty one(83.78%) patients underwent hepatico-jejunostomy. Five (13.51%) patients underwent Hepatico-dueodenostomy. One (2.70%) patient underwent porto-jejunostomy as hepatic ducts were very minute that they can admit only number 22 gauze cannula. Thirty three (89.18%) patients had above procedures as primary procedure. Four (10.81%) patients initially underwent cysto-jejunostomy, later hepatico-jejunostomy as definitive procedure. Two females children with Choledochal cyst perforation, one female child due to large cyst with poor general condition underwent cysto-jejunostomy initially, later hepatico-jejunostomy as definitive procedure .One male child with Choledochal cyst perforation undergone initially cysto-jejunostomy, later hepatico-jejunostomy as definitive procedure. Total three (8.10%) spontaneous perforations of Choledochal cyst with biliary peritonitis[15]. One male patient of nine years with choledochocele of 26mm diameter presented to us and referred to gastroenterology for transduodenal endoscopic sphincterotomy. Another female patient eleven years old, with multiple intrahepatic cysts in both lobes (Caroli's disease) who requires initial medical management and transplantation referred to Transplantation center. Both these patients not included in our study. Lillys technique used (posterior and medial walls are dissected in the submucosal plane) to avoid injury to portal vein and the hepatic artery respectively in two females children during hepatico jejunostomy.

STATISTICAL ANALYSIS

The obtained quantitative results were expressed as mean ± standard deviation, while qualitative variables were expressed as numbers and percentages

RESULTS

Choledochal cyst patients admitted three days before operative procedure for complete work up, bowl preparation done with cefataxime amikacin and metronidazole. Vit.k given to prevent excess oozing. We did open operative procedures in all patients by Right subcostal incision. Blood transfusion given to all patients per operatively (100%). Mean operative time was four hours and forty five minutes (mean285.85 minutes). Recovery of bowel function with passing stool on fourth day (mean 4.10 days). Duration of hospital stay was10 days (mean 9.88days). Bile leak was noted in one female patient of hepaticojejunostomy which was treated conservatively and within 5 days resolved (2.70%). Post-operative bleeding noted in two patients and resolved with conservative treatment with blood transfusions (5.40%). one patient developed local calcific pancreatitis(2.70%) around the insertion of ampulla of vater after one year of postoperative hepaticojejunostomy, ultrasound and CT scan reported as a retained calculi. On exploration no stone found, only indurated calcific pancreas was the cause of U/S and CT finding. Adhesive intestinal obstruction (2.70%) noted in one female hepaticojejunostomy patient, Adhesionolysis

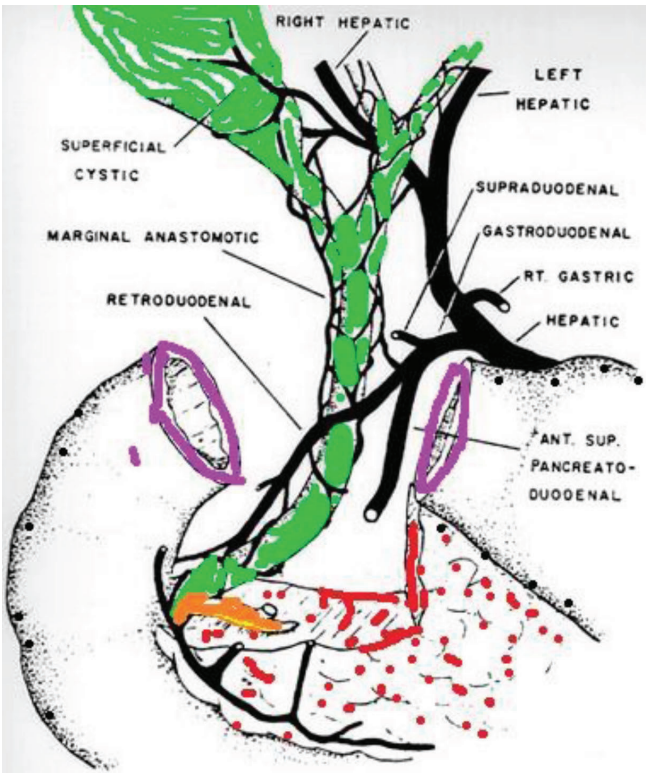

Figure-1: Blood supply of common bile duct

done. No death reported in these thirty seven operated patients. No portal hypertension or biliary cirrhosis noted. No malignancy developed during postoperative follow-up in the study group. However one male patient aged 37 year, who undergone excision of Choledochal cyst in 1980 in our department developed cholangiocarcinoma taking treatment in oncology department. Total postoperative complications five patients (13.51%). Fortunately Hepatico-dueodenostomy patients did not develop any complications like leaks , reflux related cholangitis or pancreatitis which we fearsome. Overall complication rates are low, comparable to many centers. Good quality of life and without any symptom related to biliary system, normal growth pattern achieved.

DISCUSSION

Choledochal cyst are uncommon disease in population. Female children has higher incidence ranging from male to female ratio 1:3 to1:8. Most common presentation is pain abdomen. Jaundice less frequent, mass abdomen rare. Classical triad presentation is very rare. Most of the children present in infancy and early childhood. Diagnosis is confirmed commonly by ultrasound and CECT. MRCP (magnetic resonance cholangiopancreatography) excellent non-invasive imaging for intrahepatic ducts .ERCP, PTC, transduodenal ultrasound and intraoperative cholangiograms are useful in selected cases. Most common type of cyst is Saccular or fusiform dilatation of extrahepatic CBD.[10-14] Choledochal cysts management is challenging task. Treatment modalities in our hospital are-Thirty one (83.78%) patients underwent excision and hepatico-jejunostomy. Five (13.51%) patients underwent excision and Hepatico-dueodenostomy. One (2.70%) patient underwent porto-jejunostomy as hepatic ducts were very minute. we did not have laparoscopic excision of the cyst. Most commonly done operation in the world for choledochal cysts is excision and hepatico jejunostomy, followed by excision and Hepatico-dueodenostomy. Neonatal excision of choledochal cysts with hepatico-jejunostomy reported in literature. Choledochal cysts patients need to follow up till adult hood 3rd -4th decades or more. Postoperative complications are like usual abdominal surgical procedure or cholangitis, anastomotic strictures, stones or development of cholangiocarcinoma mostly 2nd or 3rd decades. Malignancy developing risk in incomplete excision is 9 to 28% over cumulative years. Complications rate is around ten percent.[15] Our institute has 13.51% complication rate, no mortality and overall satisfactory results achieved.

CONCLUSION

No death reported in these thirty seven operated patients. No portal hypertension or biliary cirrhosis noted. No malignancy developed during follow-up in the study group. Fortunately Hepatico-dueodenostomy patients did not develop any complications. Overall complication rates are low, result are comparable to many centers. Good quality of life and symptom free normal growth achieved

REFERENCES

1. Song HK, Kim MH, Myung SJ, et al. Choledochal cyst associated with anomalous union of pancreaticobiliary duct (AUPBD) has a more grave clinical course than choledochal cyst alone. Korean J Intern Med. 1999;14:1–8
2. Schweizer P, Schweizer M. Pancreaticobiliary long common channel syndrome and congenital anomalous dilatation of the choledochal duct: study of 46 patients. Eur J Pediatr Surg. 1993;3:15–21.
3. Nakamura T, Okada A, Higaki J, Tojo H, Okamoto M. Pancreaticobiliary maljunction-associated pancreatitis: an experimental study on the activation of pancreatic phospholipase A2. World J Surg. 1996;20:543–550.
4. Kusunoki M, Saitoh N, Yamamura T, Fujita S, Takahashi T, Utsunomiya J. Choledochal cysts: oligoganglionosis in the narrow portion of the choledochus. Arch Surg. 1988;123:984–986.
5. Iwata F, Uchida A, Miyaki T, et al. Familial occurrence of congenital bile duct cysts. J Gastroenterol Hepatol. 1998;13:316–319.
6. Komi N, Takehara H, Kunitomo K, Miyoshi Y, Yagi T. Does the type of anomalous arrangement of pancreaticobiliary ducts influence the surgery and prognosis of choledochal cyst? J Pediatr Surg. 1992 Jun;27:728-31.
7. 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. .
8. Yamataka A, Kuwatsuru R, Shima H, et al. Initial experience with non-breath-hold magnetic resonance cholangiopancreatography: a new noninvasively technique for the diagnosis of choledochal cyst in children. J Pediatr Surg. 1997;32:1560-2.

9. Park DH, Kim MH, Lee SK, et al. Can MRCP replace the diagnostic role of ERCP for patients with choledochal cysts?. Gastrointest Endosc. 2005;62:360-6.
10. Oguchi Y, Okada A, Nakamura T, 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;103:168-73.
11. Joseph VT. Surgical techniques and long-term results in the treatment of choledochal cyst. J Pediatr Surg. 1990;25:782-7
12. Miyano T, Yamataka A, Kato Y, et al. Hepaticoenterostomy after excision of choledochal cyst in children: a 30-year experience with 180 cases. J Pediatr Surg. 1996;31:1417-21.
13. Li L, Feng W, Jing-Bo F, et al. Laparoscopic-assisted total cyst excision of choledochal cyst and Roux-en-Y hepatoenterostomy. J Pediatr Surg. 2004;39:1663-6.
14. 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;21:5-7.
15. Seema SA, Seth A, Taluja V, Bagga D, Aneja S. Spontaneous rupture of choledochal cyst. Indian J Pediatr. 2000;67:155–156.

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