

CHOLEDOCHALCYST IN CHILDHOOD

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ABSTRACT

OBJECTIVE.TO STUDY CLINICAL PRESENTATION, MANAGEMENT AND RESULT OF TREATMENT OF 11 CONSECUTIVE CASE OF CHOLEDOCHAL CYST MANAGED FROM 2012 TO 2019.

METHOD.11 CHILDREN PRESENTED WITH UPPER ABDOMINAL PAIN(72.72%),JAUNDICE(27.27%),MASS(18.18%).ULTRASONOGRAPHY DIAGNOSED/SUUGESTED IN ALL 11 CHILDREN AND MAGNETIC RESONANCE CHOLANGIOPANCREATIGRAM SUGGSTED IN 11 PATIENT.

RESULT.9 PATIENT FROM 11 CHILDREN HAVE TYPE 1A (CYSTIC DILATION).2 PATIENT HAVE TYPE 1C(FUSIFORM DILATATION). OPERTIVE MANAGEMENT MAGEMENT INCLUDED PRIMARY COMPLETE EXCISION OF CHOLEDOCHAL CYST WITH ROUX-EN-Y-HEPATICODOCHOJEJUNOSTOMY(N=9),AND CYST EXCISION WITH HEPATICODOCHODUEDENOSTOMY(N=2).THERE ARE 1 POST OPERATIVE DEATH DUE TO DEEP ICTERUS AND POOR GENERAL CONDITION PRIOR TO SURGERY.SHORT TERM FOLLOW UP WAS SATISFACTORY IN ALL 10 PATIENT.

CONCLUSION.CHOLEDOCHAL CYST KEPT IN MIND WHILE EVALAUTING CHILDREN WITH RECCURENT UPPER ABDOMINAL PAIN AND JAUNDICE.PRIMARY EXCISION OF CHOLEDOCHAL CYST WITH ROUX-EN-Y-HEPATICODOCHOJEJUNOSTOMY PROVIDING SATISFACTORY RESULT IN MANAGEMENT OF VAST MAJORATITY OF CASE OF CHOLEDOCHAL CYST IN CHILDHOOD.

CHOLEDOCHAL CYST IS A CYSTIC OR FUSIFORM DILATATION OF BILIARY TREE AND RARE MALFORMATION INCIDANCE 1 IN 100000 TO 190000 LIVE BIRTH IN WESTERN POPULATION.IT HAS HIGHER INCIDANCE IN ASIA PARTICULAR IN JAPAN(1 IN 1000 LIVE BIRTH).

CONGENITAL BILIARY DILATION ASSOCIATED WITH ANOMALOUS PANCREATOBILIARY JUNCTION (90% PATIENT WITH CHOLEDOCHAL CYST) AND OBSTRUCTIVE FACTOR LOCALIZED AT JUNCTION OF CHOLEDOCHUS AND DUODENUM AS ABNORMAL ANGULARITY OR CONGENITAL STENSOSIS OF COMMON BILE DUCT.

THIS PAPER DESCRIBE THE PRESENTATION ,INVESTIGATE AND MANAGEMENT AND OUTCOME IN 11 CONSECUTIVE CASE OF CHOLEDOCHAL CYST MANAGED AT OUR INSTITUTE, A TIRTIARY LEVEL CARE CENTRE OVER PEROIUD OF LAST 6 YEAR.

MATERIAL AND METHOD

THE MEDICAL RECORD OF 11 CONSECUTIVE PATIENTS, BETWEEN 2012 TO 2019 WERE REVIEWED. AMONG THESE CASES THERE WERE 9 FEMALE AND 2 MALE. MALE TO FEMALE RATIO 8.1 TO 1.8.

THE PRESENTING SYMPTOMS AND SIGNS SHOWN IN TABLE 1 AND TABLE 2.

NO. CASE CASE IN THIS SERIES SHOWED CLASSICAL TRIAD OF PAIN, ABDOMINAL LUMP, JAUNDICE.

TABLE 1. PRESENTING SYMPTOMS.

SYMPTOMS	NO. PATIENT(%)
UPPER ABDOMINAL PAIN	8(72.72%)
JAUNDICE	3(27.27%)
MASS	2(18.18%)
CLAY COLOURED STOOL	1(9.09%)
FEVER	2(18.18%)
VOMITING	2(18.18%)

TABLE 2. PRESENTING SIGN

SIGN	NO. PATIENT(%)
ICTERUS	3 (27.27%)
MASS	2(18.18%)
ASCITIS	0
FEATURE OF PERITONITIS	0

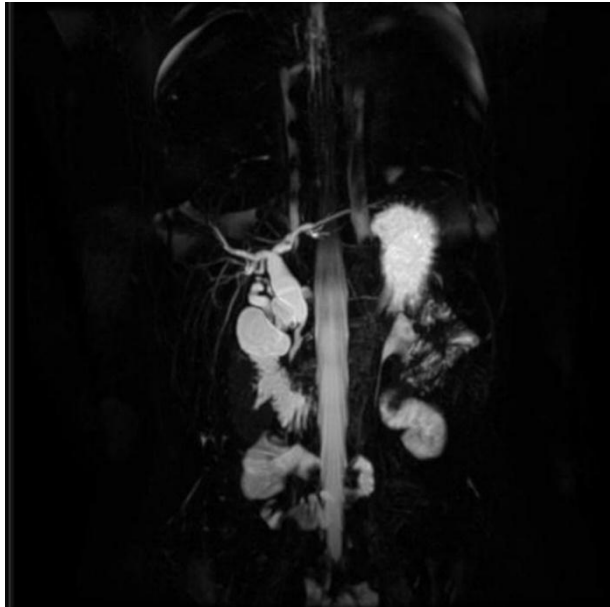
TABLE 3. HEMATOLOGICAL INVESTIGATIONS

ANEMIA(LESS THAN 10%)	3(27.27%)
JAUNDICE LESS THAN 5 MG%	2(18.18%)
MORE THAN 5 MG%	1(9.09%)
ELEVATED ALKALINE PHOSPHATASE	5(45.45%)
DERANGED LFT	6(54.54%)

IMPORTANCE PREOPERATIVE HAEMATOLOGICAL AND BIOCHEMICAL FINDING IN ALL 11 PATIENTS ARE SHOWN IN TABLE 3.

AMONG RADIOLOGICAL INVESTIGATION ABDOMINAL USG DIAGNOSED /SUGGESTED CHOLEDOCHAL CYST IN ALL PATIENTS. MRCP DONE IN ALL PATIENTS TO SUBCATEGARIZE TYPE OF CHOLEDOCHAL CYST AND BILIARY TREE ANATOMY.

ALL 11 PATIENTS UNDERWENT SURGERY WHREEVER NECESSARY, PRE OPERATIVE STABILIZATION OF PATIENTS WITH PARTICULAR REGARD TO CORRECTION OF ANEMIA,ALTERED ELECTROLYTE IMBALANCE,COAGULIZATION PROFILE AND DEHYDRATION. SURGICAL EXPLORATION PERFORMED IN ALL PATIENTS WITH RIGHT SUBCOSTAL INCISION.



MAGNETIC RRESONANCE IMAGING CHOLANGIOPANCREATOGRAM SHOWING CHOLEDOCHAL CYST.

RESULT

ON BASIS OF RADIOLOGICAL FINDING,11 PATIENTS INCLUDING ,ALL THEM HAVE EXTRAHEPATIC CHOLEDOCHAL CYST. 9 OF THEM HAVE CYSTIC DILATION AND 2 OF THEM FUSIFORM DILATION OF COMMON BILE DUCT.

SUBTYPE OF CHOLEDOCHAL CYST IN ALL 11 PATIENTS CONFORMED BY OPERATIVE FINDING IS SHOWN IN TABLE 4.

TABLE 4 SUBTYPE OF CHOLEDOCHAL CYST(N=2)

TYPE 1A	TYPE 1C
9	2

AMONG 11 PATIENTS, 8 PATIENTS ARE ADMITED BETWEEN 2 TO 3 YEAR AND 3 PATIENTS ADMITTED BETWEEN 6 TO 8 YEAR.

OPERATIVE MANAGEMENT AND OUTCOME

CHOLECYSTECTOMY WITH PRIMARY EXCISION OF CHOLEDOCHAL CYST WAS PERFORMED IN ALL 11 PATIENTS.AFTER ISOLATION OF CHOLEDOCHAL CYST FROM SURROUNDING

STRUCTURE LOWER END WAS DISSECTED TILL IT TAPPER TO NARROW CALIBRE.SUTURE LIGATED AT THIS LEVEL.PROXIMAL DISSECTION CARRIED OUT TILL LEVEL OF PROXIMAL COMMON HEPATIC DUCT ENSURING THAT CHOLEDOCHAL CYST WAS EXCISED COMPLETELY. A ROUX-EN-Y-HEPATICODOCHOJEJUNOSTOMY WITH LONG ASCENDING (40-60 CM LONG DEPENDING ON ALL AGE OF CHILD) WAS PERFORMED IN 9 PATIENTS AND HEPATICODOCHODUENOSTOMY WAS PERFORMED IN 1 PATIENTS AFTER COMPLETE EXCISION OF CHOLEDOCHAL CYST.

ALL PATIENTS HAD TUBE DRAIN INSERTED WHICH WAS REMOVED 24 HOUR AFTER ANY DRAINAGE STOPPED.THE MEAN PERIOUD FOR WHICH THE DRAINED WAS INSETERED WAS 5 DAYS.

POSTOPERATIVE COMPLICATION INCLUDING MINOR WOUND INFECTION IN 2 PATIENTS.1 POST OPERATIVE DEATH DUE TO DEEP ICTERUS AND POOR GENERAL CONDITION PRIOR TO SURGERY.

ALL PATIENT OTHER 10 PATIENTS WERE DISCHARGED AFTER RECOVERY FROM SURGERY.IN 2 SURVING PATIENTS WHO HAVE SIGNIFICANT PRE OPERATIVE HYPERBILURUBENEMIA ,SERUM BILURUBINE NORMAL OR ELSE MILDLY ELEVATED LESS THAN 2 MG% AT TIME OF DISCHARGE.

ONLY 2 OF 11 PATIENTS RETURNED FOR FOLLOW UP .ONE PATIENT CAME 1 YEAR AFTER SURGERY,ONE AFTER 2 YEAR,BOTH HAVE COMPLAIN OF RECCURENT UPPER ABDOMINAL PAIN .ABDOMINAL USG AND BIOCHEMICAL LIVER FUNCTION TEST NORMAL IN THESE 2 PATIENTS.

DISCUSSION

CHILDHOOD CHOLEDOCHAL CYST,SHOWING CLASSICAL TRAIID OF CLINICAL PRESENATATION ABDOMINAL PAIN,JAUNDICE,MASS. FOR DIAGNOSIS OF CHILDHOOD CHOLEDOCHAL CYST THAT PROPERLY PERFORMED HIGH RESOLUTION ADDOMINAL USG IS BEST SCREENING TEST. IN RECENT YEAR MRCP ADVOCATED AS IDEAL NON INVASIVE DIAGNOSTIC MODALITY FOR CHOLEDOCHAL CYST AS IT ENSURE ACCURATE VISUALIZATION OF ENTIRE PANCREATICOBILIARY SYSTEM.INTRAOPERATIVE ENDOSCOPY OF COMMON CHANALE PANCREATIC DUCTAND INTRAHEPATIC DUCT USING FREE CHOLANGIOCATHETER DURING SURGERY , IT HELP TO DETECT AND MANAGE PROTEINE PLUG,STONE AND DEBRIS,IN THIS STRUCTURE,DETECT INTRAHEPATIC DUCTAL STENOSIS, CYSTIC INTRAHEPATIC DILATION ,STRICTURE,AT JUNCTION OF INTRAHEPATIC DUCT AND COMMON HEPATIC DUCT AT HILUM.

IN ANATOMICAL VARIANT OF CHOLEDOCHAL CYST IN THIS SERIES IT IS SIGNIFICANT THAT TYPE 1A CYSTIC DILATION OF EXTRAHEPATICBILIARY SYSTEM IS MOST COMMON. REGARING SURGICAL CHOICE PROCEDURE, TOTAL CYST EXCISION WITH BILIOENETERIC DRAINAGE IS IDEAL.WHILE MORE OF SURGEONS ADVOCATE A ROUX-EN-Y-HEAPATICODOCHOJEJUNOSTOMY WITH LONG LENTH OF ASCENDING LOOP (40 TO 60 CM) FOR DRAINAGE AFTER COMPLETE EXCISION OF CHOLEDOCHAL CYST .SOME SURGEONS PREFER HEPATICODOCHODUEDENOSTOMY.HOWEVER SOME RECENT REPORT TESTIFY TO

HIGH INCIDANCE OF BILIARY REFLUX INDUCED GASTRITIS DUE TO HEPATICODOCHODUEDENOSTOMY.OUR EXPERIENCE SUGGEST COMPLETE EXCISION WITH ROUX-EN-Y-HEPATICODOCHOJEJUNOSTOMY GIVE GOOD RESULT AND USUALLY SAFE AND FEASIBLE AS PRIMARY PROCEDURE.

CONCLUSION

CHOLEDOCHAL CYST IS IMPORTANT ENTITY IN CHILD SHOULD BE KEPT IN MIND WHILE EVALUATING CHILDREN WITH RECURRENT ABDOMINAL PAIN AND JAUNDICE.ABDOMINAL USG IS BEST SCREENING TEST.INCREASING AVAILABILITY AND USE OF MRCP WILL ELIMINATE NECESSITY OF INTRAOPERATIVE CHOLANGIOGRAPHY AND MINIMIZE THE NEED FOR INTRAOPERATIVE ENDOSCOPY SO IDEAL NON INVASIVE TOOL FOR DIAGNOSIS AND STUDY CONFORM THAT PRIMARY EXCISION CHOLEDOCHAL CYST WITH HEPATICODOCHODUEDENOSTOMY PROVIDE SATISFACTORY RESULT IN MANAGEMENT OF VAST MAJORITY OF CASE OF CHOLEDOCHAL CYST.

REFERENCES

- 1.Howard ER. Choledochal cysts. In Howard ER, ed. Surgery of liver disease in children. Oxford; Butterworth- Hienemann, 1991; 78-90.
2. Miyano T, Yamataka A, Congenital biliary dilatation.Semin Pediatric Surgery 2000; 9: 187-195.
- 3.Mukhopadhyay B,Bommayya N,Mukhopadhyay .Choledochal cyst-experience in five years. Journal Indian Association,Pediatric Surgery 1999; 4: 118-122.
4. Stringer MD, Dhawan A, Davenport M, Mieli-Vergani G,Mowat AP, Howard ER. Choledochal cysts: lessons from a 20year experience. Archives Diseases Childhood 1995; 73: 528-531.
5. Todani T, Watanabe Y, Narusue M, Tabuchi K, Okajima K.Classification, operative procedures and review of thirtyseven cases including cancer arising from choledochal cyst. 134: 263-269.
6. Lilly JR. Total excision of choledochal cyst. Surg Gynaecology\Obstetric 1978; 146: 254-256.
7. O'Neill, Jr. JA. Choledochal cyst. In O'Neill, Jr. JA, Rowe MI,Grosfeld JL, Fonkalsrud EW, Coran AG. eds. Pediatric Surgery,5th ed Vol. 2., St. Louis, Missouri; Mosby-Year Book, Inc, 1998;1483-1494.
8. Vijayaraghavan P, Lal R, Sikora SS, Poddar U, Yachha SK.Experience with choledochal cysts in infants. Pediatric Surgery Int 2006; 22 : 803-807.
9. Joseph VT. Surgical techniques and long-term results in the treatment of choledochal cyst. J Pediatric Surgery 1990; 25: 782-787.

10. Le L, Pham AV, Dessanti A. Congenital dilatation of extrahepatic bile ducts in children. Experience in the central hospital of Hue, Vietnam. *Eur J Pediatric Surgery* 2006; 16: 24-27.
11. Suzuki M, Shimizu T, Kudo T et al. Usefulness of nonbreath-hold 1-shot magnetic resonance cholangiopancreatography for the evaluation of choledochal cyst in children. *J Pediatric Gastroenterology Nutrition* 2006; 42: 539-544.
12. Sarin YK. Biliary ductal and vascular anomalies associated with choledochal cyst. *J Indian Ass Pediatric Surgeons* 2005; 10:86-88.
13. Narasimhan KL, Chaudhury SK, Rao KLN. Management of accessory hepatic ducts in choledochal cyst. *J Pediatric Surgery* 2001; 7: 1092-1093.
14. Muise AM, Turner D, Wine E, Kim P, Marcon M, Ling SC. Biliary atresia with choledochal cyst: implications for classification. *Clinical Gastroenterology Hepatology* 2006; 4 : 1411-1414.
15. Koshinaga T, Wakabayashi K, Inoue M et al. Pancreatitis after primary and secondary excision of choledochal cysts. *Surgery Today* 2006; 36 : 686-691.
16. Martinez-Ferro M, Esteves E, Laje P. Laparoscopic treatment of biliary atresia and choledochal cyst. *Semin Pediatric Surgery* 2005;14 : 206-215.
17. Srimurthy KR, Ramesh S, Babu N. Laparoscopic correction of choledochal cyst in 10 children. Paper presented at the 30th Annual Conference of the Indian Association of Pediatric Surgeons, 28th – 31st October, 2004, at Jabalpur, India.
18. Li L, Feng W, Jing-Bo F et al. Laparoscopic-assisted total cyst excision of choledochal cyst and Roux-en-Y hepatoenterostomy. *J Pediatric Surgery* 2004; 39: 1663-1666.
19. Todani T, Watanabe Y, Mizuguchi T et al. Hepaticoduodenostomy at the hepatic hilum after excision of choledochal cyst. *Am J Surgery* 1981; 142: 584-587.
20. 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? *Pediatric Surgery Int* 2005; 21: 5-7.
21. Takada K, Hamada Y, Watanabe K, Tanano A, Tokuhara K, Kamiyama Y. Duodenogastric reflux following biliary reconstruction after excision of choledochal cyst. *Pediatric Surgery Int* 2004; 21: 1-4.

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