Original Article

Pediatric Choledochal Cyst, Symptoms/Signs, Management and Outcome, Our Experience

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ABSTRACT

Objective: To discuss the symptoms/signs, investigations and management of children presented with choledochal cysts (CC)

Study Design: Retrospective study

Place and Duration of Study: This study was conducted at the Department of Pediatric and Neonatal Surgery Bahawal Victoria Hospital from September 2014 to September 2022.

Materials and Methods: 63 patients under 16 year of age with diagnosis of choledochal cyst were studied. For statistical analysis we divided patients into two groups , <1 year of age and above 1 year to 16 year of age. Mass right hypochondrium, jaundice, painabdomen, nausea /vomiting with raised alkaline phosphatase level , and imaging USG/CT Scan abdomen finding favouring choledochal cyst were the inclusion criteria.

Results: Out og 63 children studied, 17(26.9%) were infants (<1 year) and 46 (73.1%) were older than one year of age. Seventeen months was the mean age with range from 21 days to 16 years. thirty five (55.9%) patients were female and 28 (44.1%) male. Type-I choledochal cyst 59 patients, Type-IV , 3 patients and type V(caroli disease)1 patient.as regards symptoms/signs(S/S) jaundice in 13(76.5%), and acholic stool in 9 (59%) patients among <1 year , while second group(>1 year) presented with pain abdomen 38 (83%), vomiting 35(77%), fever18 (43%) and acute pancreatitis 12(27%). Mass right hypochondrium23 (50%)was another main presenting feature in older age. post operative complications were noted in 7 (11.7%) patients which were managed.

Conclusion: The symptom/signs of Choledochal cysts vary according to the patient age and operative excision is the main stay of treatment. So early detection and management will prevent the rate of complications.

Key Words: Choledochal cyst, symptoms/signs, Surgical outcome

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INTRODUCTION

Transportation of bile from the liver to the gall bladder and duodenum is carried out by bile duct whose cystic dilatation is a rare congenital condition, called as choledochal cyst, Vater and Ezler described it for the first time in 1723 .[1] The incidence of choledochal cyst is recorded as one in 1000 in Asian population with 2/3rd from Japan which is considered as highest. While in western countries its incidence is one in 100,000 to 150,000 live birth.[2] choledochal cyst may be diagnosed at any age but usually presents in older children.[3]

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According to anatomical site, as described by Todani Choledochal cysts is classified into 5 types which may be intrahepatic (within the liver) or extrahepatic(outsie the liver).type 1 Choledochal cysts is the most common and acoounts for ninty percent of all. It is extrahepatic and usually fusiform or sphericle in shape. Type-II CC is diverticulum of the common bile duct. . Type-III is intraduedenal or intrapancreatic dilatation of duct,with normal common bile duct. Type IV CC is multiple cystic dilatations either intrahepatic or extrahepatic or in both locations. Type V choledochal cyst consists as either single or multiple cyst with hepatic fibrosis, also called as Caroli disease.[4,5,6]

The development of choledochal cyst is mainly unknown. However abnormal pancreatic duct opening at the junction of pancreatic duct and common bile duct is reported in aproximaely 70% of cases which occurs extrduedenally and thus pancreatic fluid is refluxed into biliary tree.[7,8,] this refluxed pancreatic fluid containing pancreatic enzymes causes inflammation, weakness and ultimately pancreatic/common bile duct dilatation.[10] The most common age ,about 80%, of presentation is under 10 years of age.[7,9] the clinical features vary a bit according to the age of patient, however classical triad

of jaundice, pain abdomen and mass right hypochondrium or epigastium is present only approximately 1/3rd of patients[7]. the complications associated with delay in diagnosis of choledochal cyst include pancreatitis , cholangitis, cholilithiasis and very rarely malignant transformation.[10]

As local studies related to choledochal cyst are lacking so objective of this study was to explore common symtoms/signs,investigations required and management of choledochal cyst. This study will help clinicians to early diagnose, manage the disease in both infants and older children and thus prevent them from complications associated with it.

MATERIALS AND METHODS

After taking approval from ethical review committee this study was conducted at department of pediatric and neonatal surgery bahawal Victoria hospital. record of all pediatric patients with choledochal cyst sdmitted in pediatric surgery department from September 2014 to September 2022 were collected and reviewed .

For data collection a predesigned Performa was designed. data which noted down included name, age, sex and symptoms and signs; pain abdomen, vomiting ,fever, jaundice, change in color of stool(acholic stool), right upper quadrant abdominal mass, Classic triad(pain abdomen , jaundice, mass abdomen) and pancreatitis . serum bilirubin (total, direct/indirect) alkaline phosphatase, serum amylase and lipase level, alanine transaminase (ALT), aspartate transaminase (AST), and prothrombin time (PT),activated partial thromboplastin tima (APTT), international normalized ratio (INR). Imaging modalities required USG (ultrasononograghy) abdomen, abdominal CT scan, MRI/ MRCP (magnetic resonance cholangiopancreatography) were noted. According to age two groups were made; patients under one year of age and older i.e 1 to 16 year of age . surgical procedures performed and their results were noted. Data collected was compared between two groups.

Chi-Square test was used for Statistical analysis which was done by using SPSS 23 for windows. P-value of <0.05 was considered significant.

RESULTS

Sixty three patients with choledochal cyst were studied, 17(26.9%) were infants (<1 year) and 46 (73.1%) were older than one year of age. Seventeen months was the mean age with range from 21 days to 16 years. thirty nine (61.9%) patients were female and 24(38.1%) male (table 1). Type-I CC was the commonest, 59 (93.6%) patients then Type-IV, 3 (4.7%) patients and type V caroli disease 1(1.5%) patient (table 2). Infants usually presented with yellow discoloration of eyes and body (jaundice) in 13(76.5%), and acholic stool in 9 (53%) patients.the commonest S/S, whereas pain abdomen (83%), vomiting (77%), fever (43%) and acute pancreatitis (27%) were the commonest S/S in older patients. Mass right hypochondrium was another main presenting feature in older age. (table 3) Lab investigations done as shown in table 4.abdominal USG was performed in all the patients as a baseline imaging modality,CT scan abdomen was performed in 53 (84%) patients and MRI/MRCP in 5(7.93%) patients for the confirmation of diagnosis and to identify the types of choledochal cyst. Surgical excision of the cyst was performed in all the patients after optimising. Post-operative complications of excision of CC were observed in 7 (11.7%) patients which includes anastomotic leak in 2 patients , 1 managed consevatively and one patient underwent re exploration, and 5 patients complaint of pain abdomen and ascending cholangitis that were managed conservatively.

Table No. 1: Gender distribution according to age.

Age	Male	Female	Total	Percentage
>1 year	6	11	17	26.9%
>1 year	18	28	46	73.1%
Total	24	39	63	100%

Table No. 2: Types of choledochal cyst. (n; 63)

Type	Number	Percentage
1	59	93.6%
11	0	0%
111	0	0%
1v	3	4.7%
V	1	1.5%

Table No. 3: symptoms/signs according to age.

Clinical feature	Age <1year	Age <1year	Total	P.value
jaundice	13	7	20	0.005
Acholic stool	9	2	11	0.001
Pain abdomen	3	38	41	< 0.001
vomiting	3	35	38	< 0.001
fever	1	18	19	0.001
Mass abdomen	2	23	25	0,001

Table No. 4: investigations

Laboratory	Mean ± SD	No. of patients
Total bilirubin	3.7 ± 3.3	37
Direct bilirubin	2. 5±2.0	37
INR	1.3 ± 1.0	47
ALT	123± 105	56
AST	134 ± 125	53
Alkaline phosphatase (ALP)	433 ± 411	63
Serum amylase	1021± 893	29
Serum lipase	1167 ± 925	27
Prothrombin time	9.5± 6.33	47

DISCUSSION

This retrospective study shows that the ratio of female patients suffering from CC as compared to male is high which is similar to the findings observed by studies conducted by Farooq MA (56%)and Nazir Z (78%) from Pakistan.[3,10] A female preponderance is also reported in literature from other parts of the world.[4]

This study shows that presentation of CC is different in infants and older age pediatric patients. obstructive jaundice (13 patients) and acholic stools (9 patients) were specifically observed in infants whereas in older patients, only seven patients had obstucted jaundice and 2 clay colored stools (p. 0.005 and 0.001). Similar results were observed in a study conducted concluded by Hung MH.[11] Pediatric patients of older age group specifically presented with abdominal pain, nausea/ vomiting and fever were noted more commonly among children with more than 1 year of age as compared to patients under 1 year of age (p <0.001). similar result were mentioned jn the stubies conductsd by Hung et al and Chen et al[10,11]. almost similar results to our study i.e. jaundice and acholic stool in all patients under I year and in only 35% patients above 1 year of age was reported from a study conducted at Taiwan over 25 years.[3]

We observed only 2 patients with Classic triad pain abdomen, vomiting and mass right upper abdomin . this fact is supported by literature that his classic presentation is rare in children4. Fumino et al. in their study reported no patient with classic triad and 7.47% patients were reported by Ohashi et al.which are similar to our study.[7,11]

In our study, raised serum bilirubin and PT/INR were more commonly observed in infants as compared to ALT/ AST whereas older children had elevated levels of serum amylase, lipase and ALT/AST. Similar observations were reported Ohashi T et al in their study.[10]

We observed that Type-I CC is the commonest type followed by Type-IV in both age groups. Same observations were reported from other studies conducted at different parts of the world.[12,13] Khandelwal C, reported type 1in up to 80% of the cases.[14] USG abdomen was done in all the patients in our study whereas in 53 (84%) patients CT abdomen and in 5(7.9%) patients MRI/MRCP was done. as reported in literature USG abdomen can easily diagnose Choledochal cyst and is also cost effective easily accessible however CT and MRI are advanced diagnostic equipments which can even diagnose prenatally.[13]

We performed Cyst excision and Roux-en-Y hepaticojejunostomy in all our patients. This procedure has good results among all age patients with minimal complications. Early diagnosis and surgical management results in good outcome with minimal complications[14,15]. In our study 7 patients deneloped postperative complications 6 were managed conservatively while 1 underwent re exploration. A stydy conducted by Farooq MA reported 4(12%) patients who developed complications and were managed conservatively.[3]

CONCLUSION

The symptom/signs of Choledochal cysts vary according to the patient age and operative excision is the main stay of treatment. This study will facilitate the pediatric surgeons for early detection and management of CC and thus will prevent the rate of complications.

Author's Contribution:

Concept & Design of Study:	Muhammad Ramzan Bhutta
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Conflict of Interest: The study has no conflict of interest to declare by any author.

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