ORIGINAL PROF-615

CHOLEDOCHAL CYST;
RESULTS OF MANAGEMENT OF 13 CASES IN CHILDREN

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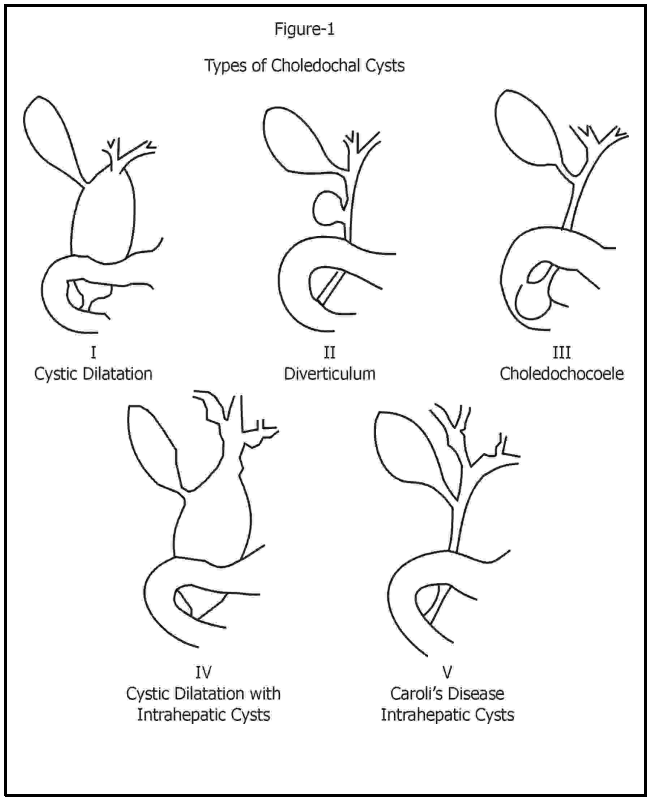
ABSTRACT

OBJECTIVES: To assess the mode of presentation and results of surgical management of choledochal cysts in children. **DESIGN;** Retrospective study **PERIOD:** 1995 to 2000. **SETTING:** Department of Paediatric Surgery, Allied Hospital Faisalabad and Department of Surgery Ghafoor Bashir Children Hospital Faisalabad: **PATIENTS & METHODS:** The case records of 13 patients with choledochal cyst were reviewed with regard to their mode of presentation, anatomical type, operative procedure performed and post-operative course. **RESULTS:** Male to Female ratio was 1.00:1.17. Nearly one third of the cases presented under the age of six months. Infants presented with jaundice and acholic stools. Episodic jaundice and pain in the RHC were the common symptoms in children. Two cases presented with acute cholangitis. All cases were operated upon and excision of the cyst with a roux-en-Y hepatico jejunostomy was done in 10, roux en Y cysto-jejunostomy in 2 and tube cystostomy in 1 case. There was one death. **CONCLUSION:** The treatment of choledochal cyst is excision which is a safe option in our setting provided the diagnosis is made in time.

KEY WORD: Choledochal Cyst.

INTRODUCTION

Choledochal cyst dilatation of the common bile duct is a relatively rare condition. It was first reported by Vater and Ezlar in 1923[1].In 1959 Alonzo Laj proposed a classification of the lesion and divided them into three types[2]. Later this classification was modified by Todania et al in 1977[3] and two more types were added (Fig 1). Type I is the commonest and constitutes 85% to 90% of all cases in the reported series[4,5,6]. There is a very high incidence of this condition in Orientals especially Japanese[7].A number of theories have been put forward to explain the etiology of the condition. Among these the pancreatic-biliary reflux theory is most popular due to the commonly observed mal-positioning of the pancreatic duct in the majority of these cases. This leads to reflux of the pancreatic secretions into the biliary tree[8] .The treatment of this condition has undergone gradual evolution over the years. Initially cystoenterostomy was practiced leaving the cyst intact but the incidence of carcinoma in this led to methods in which the cyst was excised in toto[9].



Continuity was restored by a Roux en Y hepatico jejunostomy. This gained wide acceptance[10,11]. Jejunal interposition with an anti reflux valve between the stump of the hepatic duct on one side and the lateral wall of the descending loop of the duodenum, is also being practiced in some centres[12].

PATIENTS & METHODS

The record of 13 cases of choledochal cyst managed over a six year period (1995-2000) at two hospitals of Faisalabad viz. Department of Paediatric surgery, Allied Hospital (8 cases) and department of surgery, Ghafoor Bashir Children hospital (5 cases) was reviewed . The relevant features in the history and physical examination were noted. In addition to the base line investigations special investigations done included liver function tests, ultrasonography and HIDA scan. Patients with signs of cholangitis were treated with antibiotics and operated later on. Laparotomy was performed in every case through a right upper transverse incision. The types of choledochal cyst were recorded. In the majority, the cyst was excised and hepatico jejunostomy done to a Roux loop. Any postoperative complications were also recorded.

RESULTS

During the past six years a total of 13 patients presented for management of choledochal cyst. The youngest patient was 2 months old and the oldest was 7 years. Mean age was 3.1 years. There were 6 boys and 7 girls in this series (M : F 1:1.17 (table I).

Two distinct forms of presentation were noted. The "infantile form" seen in patients under the age of 1 year was seen in 3 infants. The fourth infant presented with biliary peritonitis due to a perforated choledochal cyst. They were presented with a history of jaundice since birth and acholic stools in all cases. The other "adult form" of presentation was seen in children who presented with episodic or continuous pain, jaundice and a palpable mass in the right hypochondrium.

The classical triad of pain, jaundice and mass was seen in only 4 (30.7%) cases. Two presented with acute cholangitis, were treated conservatively and later recalled for the definitive procedure. On physical examination a swelling was visible in the RHC in 4 cases and was palpable in 6 (table II).

Ultrasonography of the abdomen was done in all except 2 cases. These cases had presented with an acute abdomen at a time when ultrasonography was not available. Liver function tests were done in all cases. HIDA scan was done in 8 cases and confirmed the diagnosis in all cases (table III).

All cases underwent exploration through a right upper transverse incision. The anatomic findings were recorded and the lesion classified (table IV).

The operative procedures done in all the cases are given in table V. Cystojejunostomy though not the procedure of choice, was done in 2 cases in which excision of the cyst was not possible.

Table-I. Age and sex distribution

Age	Male	Female	Total
2/12	-	2	2
4/12	1	-	1
6/12	-	1	1

2 yrs	1	1	2
4 yrs	3	1	4
5 yrs	1	-	1
7 yrs	-	2	2
Total	6	7	13

Table-II. Symptoms and signs

Disease	No of patients
JAUNDICE	
Since birth	3
Episodic	3
Acute onset	2
Absent	5
PAIN	
Episodic	4
Continuous (Acute)	3
Absent	6
MASS IN RHC	
Visible & palpable	3
Palpable	4

Table-III. Investigations

	Performed	Abnormal
Ultrasonography	11	11
Liver function tests	11	6
HIDA scan	8	8

Tube cystostomy was done in a 4 month old infant who had presented with jaundice and acholic stools since birth. He was in liver failure with a deranged clotting profile. He was given multiple infusions of FFP and then explored. In view of his serious condition we decided to drain the cyst as a temporizing measure. This was done but the patient did not improve and expired on the 3rd post-operative day. This was the only fatality in the series.

Table-I. Age and sex distribution

Age	Male	Female	Total

Table-IV. Anatomic type

Types	No of pts	%age
Type I	10	76.9%
Type II	1	7.7%
Type IV	2	15.4%

Table-V. Operation procedure

	No of pts	%age
Cysto-jeunostomy Roux en Y	2	7.7%
Excision of cyst with Roux en Y hepatico jejunostomy	10	76.9%
Tube cystostomy	1	15.4%

Post operatively, wound infection was seen in 3 (23.1%) cases. This was mild in 2 and severe in 1 which had disruption of the hepatico jejunal anastomosis on the 5th post-operative day. She was re-explored and anastomosis redone. She then developed bile leakage from the wound which was treated conservatively with IV antibiotics and TPN. She settled on this regimen and was discharged but later on developed an incisional hernia and is presently awaiting surgical repair of the same.

Out of the 12 cases discharged, only 7 (53.8%) have been visiting the OPD regularly. All of these were asymptomatic. One case is awaiting repair of an incisional hernia.

DISCUSSION

In our study the mean age of the children is 3:1 years. This conforms with the findings of Poddar et al and Rha et al[13,14]. The sex ratio is nearly equal in our series. This is at variance with most of the published data which ranges from M:F ration of 1:5 as reported by Ming et al from Bejing and Watanatittan from Bangkok to 1:8 reported from Japan by Chijiiwa[12,15,16]. Poddar from

Chandigarh in India has observed the same sex distribution[13]. As both of our cities fall in the same area this sex distribution may be peculiar to our region.

Jaundice was seen in 8 (61.5%) cases, pain in 7 (53.8%) and a palpable mass in 6 (46.15%). The "classical Triad" of jaundice, pain and a palpable mass was seen in 4 (30.7%) of our cases. This is at variance with the reported series and again may be a regional variation[13,15,16,17]. Of the 4 infants in our series, 3 presented with acholic stools and a visible mass in the RHC. All 4 had a type I cyst. The same presentation in infants has been described by Todani and Hsu[18,19].

Diagnosis was confirmed by ultrasonographic examination of the abdomen. This identified the cyst in all 11 cases in which it was done. In one infant the cyst was very large and caused some confusion. Biliary scintigraphy HIDA was done in 8 cases to further confirm the diagnosis. Other methods of diagnosis namely ERCP & MRCP are not available at our centre.

Anatomically type I was in 10 (76.9%) cases, type IV in 2 (15.4%) and type II in 1 (7.7%). This incidence compares with the reported series which have roughly the same findings[4,6,13,16].

As has been mentioned earlier the treatment of this condition has evolved over the years. Initially simple drainage of the cyst into a loop of the intestine was done[9,20]. With time, the occurrence of malignancy in the retained cyst led to the development of the current practice of excision of the cyst in toto[19,21]. This is followed by diversion of the bile into the intestine using a loop of jejunum. To reduce the incidence of ascending cholangitis a Roux en Y loop is used. Lately diversion of bile into the duodenum using an isolated loop of the jejunum is being practiced. We have done excision of the cyst with a hepatico-jejunostomy in 76.9% of our cases. Except for one case in whom anastomotic breakdown followed by bile leakage subsequent to a re-do, we did not have any major complications. Same from Hong Kong reported 3 anastomotic leaks in a series of 84 patients which settled on conservative treatment which is comparable[22].

CONCLUSION

Surgical excision of the cyst with a Roux en Y Hepatico Jejunostomy is the procedure of choice for this condition. It is a relatively safe procedure with low morbidity and minimal mortality if done at the proper time.

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