Original

Biliary Tract Disease in Pediatric Surgery Department: 10 Years Experience in Khouzestan-IRAN

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Abstract

Introduction: Our aim was to evaluate clinical manifestation, and outcome of biliary tract disease in patients referred for treatment to two referral centers of pediatric surgery of Ahvaz.

Materials and Methods: In this retrospective study, patients with biliary tract disease admitted in Imam Khomeini and Abuzar hospitals (two referral centers for pediatric surgery in Ahvaz) during a 10-year period starting from March 2000 were evaluated. Age, sex, clinical manifestation, type of surgery, imaging finding, laboratory finding, duration of hospital stay, blood product infusion, and mortality rates were recorded. Data was analyzed with SPSS Ver 13.0(Chicago, IL, USA). We used Chi-square and t-test for comparison.

Results: Twenty cases (m=13, f=7) of biliary atresia were included in this study. Mean age at the time of diagnosis and operation was 82.11 days (30 days- 6.5 months). Jaundice (100%), acholic stool (55%), and dark brown urine (55%) were the most frequent clinical manifestation in patients with biliary atresia. Of all cases, 17 patients underwent surgery. Eighteen cases (m=11, f=7) of cholecystitis were included in this study. Abdominal pain (72%) was the most frequent sign. Eleven cases underwent surgery. Five cases of choledochal cyst (m=0, f=5) were included in this study. Abdominal pain and vomiting was the most common clinical manifestation in cases with choledocal cyst.

Conclusion: Jaundice, acholic stool, and dark brown urine were the most frequent clinical manifestation in cases with biliary atresia. Mean age at the time of diagnosis and operation for biliary atresia was 82.11 days (30 days-6.5 months). Abdominal pain was the most frequent sign of cholecystitis. Early referral and more experience are needed in order to increase survival of biliary atresia cases in our hospital.

Keywords

- Biliary atresia
- choledochal cyst
- cholecystitis
- Jaundice
- gallstone
- abdominal pain
- acholic stool
- dark brown urine

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Introduction

Biliary tract diseases are rare in children. In cases such as biliary atresia (BA), and choledochal cyst they can be difficult to diagnose and require accurate management.[1] Five year survival following surgery has been reportedto be about 50% in France [2] and UK.[3] Most of studies about BA were from developed countries [2,3] or from Asian courtiers with high socioeconomic status.[4] Choledochal cyst is a rare medical condition, occurring in1 in 100/000 to 150/000 live birth. However there is unexplained high incidence about 1/1000 in Asian population. [5] The aim of our study was to evaluate etiology, complication, and outcome of biliary tract disease in pediatric surgery wards of a developing country.

Materials and Methods

In this retrospective study, patients with biliary tract disease admitted in Imam Khomeini and Abuzar hospitals (two referral centers for pediatric surgery in Ahvaz) during a 10-year period starting from March 2000 were evaluated. Age, sex, clinical manifestation, type of surgery, imaging finding, laboratory finding, duration of hospital stay, blood product infusion, and mortality rates were recorded. Data was analyzed with SPSS Ver 13.0(Chicago, IL, USA). Chi-square and t-test was used for comparison.

Results

Twenty cases (M=13, F=7) with BA, Five cases (M=0, F=5) with choledochal cyst and eighteen cases with cholecystitis (M=11, F=7) were included in this study. All patients underwent surgery. No mortality occurred. **Table 1** shows demographic features, clinical manifestation, blood product infusion, laboratory findings, and outcome of cases.

Biliary atresia: Age at disease onset was 0-124 day(s) with a mean of 12.3 days. Mean age at the time of diagnosis and operation was 82.11 days (30 days- 6.5 months). In this study, 11 cases (55%) underwent Kasai surgery along with liver biopsy.

Choledochal cyst: In this study, 4 (80%) cases had AST and ALT greater than 100 IU/dl **Table2**.

Surgery was done in four cases. Three (60%) of them underwent cyst excision and Roux-en-Y heapticojejunstomy and two of them underwent simultaneous cholecystectomy. One patient underwent cyst resection and Roux-en-Y choledocojejunostomy. Complication of surgery that is fever, vomiting, and bleeding from incision site was seen in two (50%) patients. There was no recurrence after surgery.

Cholecystitis: Abdominal pain was the most frequent presenting symptom in 13 cases (Table-1). Of 18 cases, 7 (38.89%) did not have surgery, from which one had sickle cell anemia and disagreed with surgery. Two cases with major thalassemia visited the hospital for splenectomy. Gallstone was detected by sonography in one case and intra-operatively in another case **Table-3.**

Two cases underwent simultaneous cholecystectomy and splenectomy. Complication of surgery was seen in 6 of the 11patients whom underwent surgery. These complications include fever (3 cases), vomiting (1 case), epigastric pain and tachycardia (one case), and decreased level of consciousness in one case. Of the 18 cases, 10 (55.55%) were without associated anomaly or disease. Sickle cell disease was found in 5 (27.7%) cases. Major thalassemia was found in 3 cases. Table 3 shows imaging findings in our patient.

Table 1: Demographic, clinical manifestation, and laboratory finding among cases with biliary tract disease

	Biliary atresia(n=20)	Choledochal cyst(n=5)	Cholecystitis(n=18)
Age	12.3 days(0-124 days)	4.21 yrs (25 day -9 years)	8.85 yrs (8-12 years)
Clinical manifestation	Jaundice (20,100%) Acholic stool (11,55%) Dark brown urine (11,55%) Hepatomegaly (6,35%) Vomiting (6,35%) Abdominal distention (5,25%) Lethargy (3,15%) Greasy stool (2,10%) GI bleeding (2,10%) Fever (2,10%) Restlessness (2,10%) Nose bleeding (1,5%) Decreased reflexes (1,5%) Poor feeding (1,5%) Ascites(1,5%)	Abdominal pain (4,80%) Vomiting (4,80%) Loss of appetite (3,60%) Constipation (2,40%) Jaundice (1,20%) Pruritus (1,20%) Abdominal distention (1,20%)	Abdominal pain (13,72%) Vomiting (8.44%) Jaundice (6,33%) Hepatomegaly (4,22%) Splenomegaly (4,22%) Loss of appetite (3,16%) Fever (2,11%)
Blood product infusion	Packed Red Blood Cell (8,40%) FFP (14,70%) Vit K (19,95%)	Packed Red Blood Cell(0) FFP(0) Vit K(2)	Pack Red Blood Cell (4,22%) FFP (1,5%) Vit K (3,16%) PLT (1,5%)
Duration of surgery(Minute)	92(30-195)	177.5(75-300)	101(45-200)

Table-2: Laboratory finding in our cases

		Biliary atresia(n=20)	Choledochal cyst(n=5)	Cholecystitis(n=18)
Total Bilirubin(mg/dl)		14.05(8-26.1)	4.62(0.4-17.4)	5(0.2-26)
Direct Bilirubin(mg/dl)		8.09(2.9-17.8)	0.3-8.4(2.35)	3.6(0.1-18)
WBC		12640(8300-25300)	6400-15000(11440)	9610(2700-18900)
Hb		9.78(7.6-13.5)	10.4-13.4(11.88)	10.5(6-13.8)
U/A			Normal	
SGOT	>100	18(90%)	4(80%)	2(11%)
	<100	2(10%)	1(20%)	16(89%)
SGPT	>100	14(70%)	4(80%)	2(11%)
	<100	6(30%)	1(20%)	16(89%)
Mortality		4(20%)	0(0)	0(0)

Table-3: Imaging finding among cases with biliary tract disease

Biliary atresia(20)	Choledochal cyst(5)	Cholecystitis (18)
Normal (3, 15%)	Suggesting choledochal cyst (5,100%)	Gall bladder wall thickness (5, 27.77%)
Invisible or contracted gallbladder (9, 45%)		Gallstone (8, 44.14%)
Collapsed gallbladder (1, 5%)		Biliary Sludge (2, 11.11%)
Splenomegaly (2, 10%)		Hepatomegaly (1, 5.55%)
HIDA scan consistent with BA (15, 75%)		Splenomegaly (2, 11.11%)

Discussion

Biliary atresia: In our study, 13 (65%) of the 20 cases were male. In a study by Tiao et al 46 of the 93 cases were male. [6] In studies by Sasson et al [7] and Liem et al [8], about 50% of cases were male. In contrast to the above studies, 61.9% of cases in a study by Toyosaka et al were female [9] on the other hand in the study from Egypt, 20 (30%) of cases were female. [10]

In the present study, mean age at onset was 12.3 days (0-124). Al- Alawi et al, reported 3-42 days as age at disease onset which is lower than our study. [11]

In our study, mean age at surgery was 82.1 days which is higher than the age reported by Raval et al. [12], Susson et al [4] and Tiao et al.[6] Raval et al reported that the mean age at which porto-enterostomy is performed in the United States is 65.5 days and has

not improved in the past 10 years. [8] In a study by Wildhaber et al, mean age at surgery was 68 days (30-126) which is higher than ours but range of time was similar to our study. [13] Mean of age in our study was similar to Kielin et al. study (80.5, 25-297).[14] In Study by Scheriber et al., median patient age at time of the Kasai operation was 55 days. [15] In the study by Shoen et al., authors concluded that there was no contraindication for performing Kasai for children aged > 75 days [16] but, increasing age at the time of Kasai procedure, progressively worsens the outcome of patients. [17] As shown above, mean age at the time of surgery in our study was more than most studies. This higher mean age may be the reason for the higher mortality rate in our study. In our study, 11(55%) patients underwent Kasai procedure. Liberek et al. performed Kasai in 14 cases out of 15 cases. [18] Oh et al. performed Kasai in 59 of 62 cases.[19] Elsadat performed Kasai procedure in 100% of cases. [10] One approach that can help in early detection of biliary atresia is evaluation of stool color in jaundiced infants. That is why in Japan stool color card is used for screening neonates and has been effective for identifying infants with biliary atresia.[20] Also In Taiwan after implementation of nationalized stool color card screening program, 58.6% of infants with biliary atresia have been detected early and received treatment prior to 60 days of age compared to 23% before implementations of the screening program.[21] The Kasai procedure is useful for infants with BA, yet it is not without complications. Cholangitis, portal hypertension, hepatopulmonary syndrome, and hepatic malignancy have been reported as complications. [22,23]

In our study the mortality rate was 20% which is higher than Elsadat study.[10]

Also gallbladder was not visible in 45% (9/20) of our cases. In the study by Humphrey and Stringer [24], of 30 cases, gallbladder was not detected in 7 (23/33%) cases. Other results of the two studies were similar.

Due to delay in referral of BA cases to pediatric surgeons, we were unable to perform Kasai procedure in most of the cases. This delay may be due to several problems. Some physicians in primary health care centers have limited experience with BA. The field of pediatric gastroenterology in Iran has a limited number of specialists. For example in our province with a population of 4,000,000 people, there is only one pediatric gastoenterologist. Also the traditional belief of some people in the rural area in herbal medicine may result in delay of presenting the infant to a pediatrician. Another reason may be due to economic problems.

Choledochal cyst: All of our cases were female. In another study female was the predominate sex which is consistent with our study. [25,26] Eighty percent of cases had abdominal pain. Only 20% of cases had jaundice. In other studies, abdominal pain and jaundice were the most common manifestation. [24,27,28] Singhavejsakul reported that from 32 cases with choledochal cyst, jaundice was found in 17 cases and abdominal pain in 16 cases. [24] Huang et al reported that in children with choledochal cyst the main presenting symptom is palpable abdominal mass, but in adults it is abdominal pain. [25] In the study on 27 cases with choledochal cyst, Elhalaby et al reported the most common presenting presenting symptoms to be jaundice, jaundice and palpable mass, and abdominal mass without jaundice. [29] Savic et al studied 31 cases with choledochal cyst. Abdominal pain and jaundice were found in 95% of cases, vomiting in 74%,

fever in 56%, and palpable tumor in 48% of cases. [30] In our study, 80% of cases had ALT and AST greater than 100 IU/l. Singhavejsakul showed that children aged < 2 years of age had significantly higher level of AST than those over the age of 2. [17] The low sample size of our study may be the cause of this difference.

Conclusion

Age at the time of Kasai surgery in our cases was higher than other studies. Early referral of each case with 1st day jaundice or prolonged jaundice is recommended to prevent complication resulted from delay in treatment or diagnosis. Implementation of new screening program may help us prevent further complications. Improvement in training of pediatric gastroenterologists may improve quality of care in cases with biliary tract disease.

Limitation

The main limitations of this study were the retrospective method and low sample size.

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Conflict of Interest

There is no conflict of interest.

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