Original Article

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Late Presentation, Diagnostic Delay with no Facility for Liver Transplantation of Biliary Atresia Cases in Bangladesh: Experience of 103 Cases in a Tertiary Care Hospital

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Abstract

Background: Biliary atresia is an important cause of neonatal cholestasis, which needs timely diagnosis for a satisfactory outcome after the Kasai procedure. It has been reported that bile flow has been re-established in more than 80% of infants who were referred for surgery within 60 days of birth, the success rate drops dramatically to under 20% in those older than 90 days at the time of operation. The study aims to observe age at presentation, at diagnosis after hospital evaluation,the number of cases referred for Kasai before 60 days of age,and the clinico-biochemical findings of biliary atresia cases attending a tertiary care hospital in Bangladesh.

Materials and methods: This was a retrospective cross-sectional observational study from hospital records, approved by the ethical committee of the Pediatric Gastroenterology Department of Bangladesh Medical University (BMU) Dhaka, Bangladesh. The study was conducted from January 2018 to January 2022, after approval from the departmental ethical board. A totalof 184 cases of neonatal cholestasis were admitted, among them, 103 were diagnosed as BA

Results: We found that only 21.45% of biliary cases were reported before 60 days of age, while 79% of cases were reported after 60 days of birth. The mean age at presentation was 92±41 days, with an additional 11.8±3.7 days needed for hospital evaluation. Only 32% of cases underwent the Kasai procedure, including both age groups.

Results: As there is no facility for liver transplantation currently in Bangladesh, patients of biliary atresia with failed/no Kasai procedure, especially from low-income groups faces a worse prognosis in late infancy. Late diagnosis from various factors, including late referral with long hospital evaluation time and no facility for liver transplantation, are the main obstacles forthe management of biliary atresia cases in Bangladesh. Raising awareness for early diagnosis and the establishment of a liver transplantation center at the government level would be the priorities for a better outcome of biliary atresia patients in Bangladesh.

Key words: Biliary atresia; late diagnosis; liver transplantation.

Introduction

Cholestasis is defined as reduced bile formation or flow, resulting in the retention of biliary substances within the liver normally excreted into bile and destined for elimination into the intestinal lumen. The process occurs as a result of impaired bile formation by the hepatocyte or from obstruction to the flow of bile through the intrahepatic and extrahepatic biliary tree.[1,2] One of the most important causes of neonatal cholestasis is biliary atresia. Biliary atresia is the end result of a destructive, idiopathic, inflammatory process that affects intra- and extrahepatic bile ducts, leading to fibrosis and obliteration of the biliary tract and eventual development of biliary cirrhosis.[3] Biliary atresia occurs worldwide, affecting an estimated 1 in 8000–15,000 live births. There is a slight female predominance.[4] Two different forms of biliary atresia are recognized, with disparate pathogenesis: a fetal or embryonic form and apostnatal form. Affected infants present with jaundice (Conjugated hyperbilirubinemia) and acholic stools. The presence of hepatomegaly, failure to thrive, pruritus and coagulopathy depends on the level of progression of the disease. Affected infants are usually born at term, are of normal birth weight and their weight gain is appropriate early in the course. Patients with biliary atresia occasionally present with bleeding as a result of vitamin K deficiency. Examination may reveal hepatomegaly and splenomegaly. Ascites and wasting may be

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seen as late manifestations if biliary cirrhosis has supervened.[5] Increased awareness to ensure early diagnosis and development of methods to prevent progressive hepatic fibrosis are currently needed. Diagnosisof biliary atresia still remains a big challenge for physicians. Distinguishing Biliary Atresia (BA) from other causes of cholestasis is challenging due to the lack of disease-specific clinical signs and laboratory tests. Timely diagnosis of BA is a high priority because the success of portoenterostomy rapidly declines with age.[6] A proportion of patients with biliary atresia will derive long-term benefit from hepatoportoenterostomy. The success rate for establishing good bile flow after the Kasai operation is much higher (90%) if performed before 8 weeks of life.[7,8] The short-term benefitof hepatoportoenterostomy is decompression and drainage sufficient to forestall the onset of cirrhosis and sustain growth until a successful liver transplantation can be done. Therefore, early referral and diagnosis of infants with suspected biliary atresia is urgent. Although late referral of BA cases is a problem all over the world, aim of this study is to observe age at diagnosis, clinical and biochemical characteristicsof BA cases in a tertiary care hospital (With no existing facility for liver transplantation in Bangladesh) to raise clinicians' awareness for early referral and timely diagnosis of BA for a better outcome.

Materials and methods

This was a retrospective cross-sectional observational study from hospital records, approved by the ethical committee of the Pediatric Gastroenterology Department of Bangladesh Medical University (BMU) Dhaka, Bangladesh. The study was conducted from January 2018 to January 2022, after approval from the departmental ethical board. A totalof 184 cases of neonatal cholestasis were admitted, among them, 103 were diagnosed as BA. All these BA patients underwent ophthalmic evaluation, CBC, Liver function test, including serum ALT (SGOT) AST (SGOT) Gamma GT, Prothrombin Time (PT) septic screening and TORCH screening, abdominal ultrasound, hepatobiliary scintigraphy, and liver biopsy as per departmental protocol. Biliary atresia cases were diagnosed based on clinical (Full-term with good birth-weight, persistent pale stool), biochemical (Moderate elevation of direct serum bilirubin) ultrasonographic (Non-visualized or contracted gall bladder, gall bladder contraction absent after meal) scintigraphic (Absent tracer excretion) and finally, characteristic histological findings in liver biopsy. Percutaneous liver biopsy (Using True- cut biopsy needle by same person with ensured normal vitals, coagulation profiles, sonographic exclusion of hepatic cystic lesion) was done after informed consent from parents, hepatic tissue obtained from patients were sliced, stained with massion's trichrome, examined under microscope by same person at the department of Pathology at BMU using BA histologic scoring system.[9] Clinical profile, liver function tests, imaging study results and liver biopsy findings were evaluated in 103 biliary atresia infants who presented with cholestatic jaundice.

Data were collected in a pre-formatted questionnaire. Data were expressed as mean with Standard Deviation (SD) or median (Range) or number or percentage as appropriate. Chi-square and Fisher's exact tests were used for categorical variables. For all statistical tests, a p-value of less than 0.05 was considered statistically significant.

Results

Among 187 cases of neonatal cholestasis, one hundred three (103) patients with biopsy-proven biliary atresia were managed (45 females and 58 males) in the Pediatric Gastroenterology department, BMU, from January2018 to January 2022. Studied BA cases were divided into 3 age groups in days: below 60, 60 to 90, and above 90 days. Only 22 cases (21.4%) were below 60 days, 46(44.7%) were within 60-90 days, and 35(34%) were above 90 days. Their mean age at presentation was 92±41 days (Table I). The age range of the studied BA caseswas 20 days to 7 months, and 58 (56.3%) were male and 45(43.7%) were female (Table II). Hepatomegaly was found in all the cases. Splenomegaly was found more in patients above 60 days of age, a total of 64patients (62%) edema and ascites were not found in any cases at presentation (Table II). During hospital evaluationgrand mean value of serum ALT, GGT, PT/INR was 157±95.3. U/L, 673.2±581 U/L and 36±129/1.8±1.5 respectively (Table III). Ultrasound evaluation of the hepatobiliary system showed features suggestive of BA in 88 (85.4%) cases (18 cases in <60 days, 70 cases in >60 days

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age group) while not suggestive of BA in 15 (14.5%) infants (Table III). Hepatobiliary scintigraphy showed absent tracer excretion in 98 (95%) cases (19 cases in <60 days of age, 79 cases in >60 days age group), even 24 hours after 3 mCi of mebrofenin (BrIDA) administration, except 3 cases in <60 days of age and 2 cases in >60 days age group (Table III). Characteristic histological features of biliary atresia (Bile ductular proliferation, portal tract fibrosis, and bile plugs in portal triads) were found in 93 (90%) infants. Out of 103 cases of BA, 33(32%) patients underwent Kasai PE, 22 patients (66.6%) were from the 0-60 days age group and 11 (13.85%) were from >60 days age group (Table III). A significantly large number of lately diagnosed cases 70(68%) had an age at diagnosis that was not in favourof the Kasai procedure. The Mean Hospital stay in the Paediatric Gastroenterology Department was 11.8±3.7 days (Table III).

Table I Distribution of the studied patients (n=103) according to number, age at presentation, Gender

Age group	No (%)	Age at presentationin days
< 60 days	22(21.4%)	42.3±9.3 days
60-90 days	46(44.7%)	67.3 ±13.2 days
>90 days	35(34%)	128.9 ±37.2 days
Mean age at presentation	92±41 days	
Gender distribution of studied patients	No(%)	
Male	58 (56.3%)	
Female	45 (43.7%)	

Table II Distribution of clinical finding's the studied patients b (n = 103)

Physical findings	< 60 days	60-90 days	>90 days	Total
Ascites	0	0	0	0
Edema	0	0	0	0
Hepatomegaly	22(100%)	46(100)	35(100%)	103(100%)
Splenomegaly	7(31.8%)	22(47.8%)	35(100%)	64(62%)
Hospital stay	11.8± 3.7 days			

Table III Distribution of the studied patients (n=103) according tobiochemical, imaging, biopsy findings, KASAI procedure status (n=103)

Biochemical Findings	Mean Below 60 days	Mean 60 to 90 days	Mean Above 90 days	Grand mean
Serum ALT	161.5±91.1	147.4±94.3	166±95.7	157±95.3
Serum Gamma GT	761±614	696±649	605.5±462.5	673.2±581
PT (Seconds) /INR	23.9±24.8	47±180	23.6±19.11	36±129
	2.01±2.2	1.73±1.4	1.8±1.18	1.8±1.5
Radio-Imaging				
Age range	< 60 days (no)	>60 days (no)	Total (no %)	p value
USG suggestive of Biliary atresia				
Yes	18	70	88 (85.4%)	0.840*
No	04	11	15 (14.5%)	
HB Scintigraphy Features of Biliary atresia				
Yes	19	79	98 (95%)	0.109*
No	03	02	5 (5%)	
Liver biopsy suggestive of BA				
Yes	20	73	93 (90.3%)	1.00*
No	02	08	10 (9.7%)	
BA cases undergone Kasai procedure				
Done	22	11	33 (32%)	p value*
Not done	0	70	70 (68%)	0.0001

*Fisher's exact test.

Discussion

The study showed that among 103 BA cases, there is a slight male preponderance of 56.3%. Though biliary atresia has been reported to occur commonly in females, similar findings were not obtained in our study.[3] There is an important finding in this study, that is the age at which the patients were reported to this tertiary center, only 21.4% reported before 60 days, 44.7% reported between 60 to 90 days and 34% after 90 days of life. We found near 79% of BA cases were reported after 60 days of life, where the mean age at presentation was 92±41 days. Which is66 ± 29.2 days in another study, Study from Taiwan show's mean age at testing was 22 ± 15.7 days).[10,11] Similar findings have been reported from India, showinga mean age of presentation of 118 153 days.[12] Late referral of BA cases is a management problem worldwide, it is more prevalent in Bangladesh. These studies have identified several factors for the delay at referral: i) Families and attending physicians not well informed about the importance of an early diagnosis ii) Family physicians thinking as breast milk jaundice which is very common in this age; fails to recognize the importance of investigating prolonged jaundice in neonates greater than 2 weeks of age iii) Families living in the peripheries

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with limited access to healthcare system. Surprisingly, all the above-mentioned observations exist in Bangladesh, especially confusing biliary atresia cases with breast milk jaundice. The current study showed hepatomegaly was foundin all cases (100%) of BA, which was 72% in our neighboring country.[13] Splenomegaly was present in all patients (100%) above 90 days of BA cases, which was 32% in the below 60 days age group and in total it was 62%, which was 66.7% in another study from the same geographic region.[13] Edema and ascites were not found in any cases at presentation in this study (Table II). 11.8±3.7 days (Mean value) were needed for hospital evaluation, ultimately, nearly at 100 days (Including mean age at presentation) diagnosisof BA was established due to late referral. It's well known that after 90 days success rate of the Kasai procedure drops below 20% (7,8) so at diagnosis,our success rate of the Kasai procedure is theoretically below 20%, which is very frustrating, as at present, we have no/less facility for liver transplantation.Themean value of serum ALT and GGT was 157±95.3. U/L and 673.2±581 U/L,respectively. another study from India showed a mean value of GGT was 604.5 ± 717.1, which is close to our value.[13] Ultrasound evaluation of the hepatobiliary system showed features suggestive of BApresent in 88(85.4%) cases, in 15(14.5%) cases, sonographic evaluation was not in favor of BA; the difference was significant. Choopa et al found that ultrasonography was only able to identify biliary atresia in 69.7% of the 66 patients who had features of biliary atresia on histology, result of the current study is higher and this variation might be due to operator dependency of the imaging modality.[13] Hepatobiliary scintigraphy showed absent tracer excretion in 98 (95%) cases. 3 cases in <60 days of age, 2 cases in >60 days age group result was not in favor of BA, explanation might be due to incomplete obliteration of the common bile duct in the 5 cases. Characteristic histological features of biliary atresia (Bile ductular proliferation, portal tract fibrosis, and bile plugs in portal triads) were found in 93 (90.3%) infants. while in 10(9.7%) cases, histological features were not suggestive of biliary atresia, which might be due to a technical fault, mimics of biliary atresia, etc. The Mean Hospital stay in the Paediatric Gastroenterology ward was around 12 days, which contributes to the already delayed diagnosis, as the majority of cases are reported after 60 days of age. This might be shortened if one stop diagnostic service with a dedicated team could be provided.

The study showed that only 33 (32%) out of 103 biliary atresia cases were referred for the Kasai procedure, [22 patients (66.6%) from the below 60 days age group and 11 (13.85%) from the above 60 days age group]. It is very disappointing that a statistically significant number of cases, 70(68%), in this studydid not underwent the Kasai proceduredue to diagnostic delay from late presentation, along with long hospital evaluation time.

Long gap from initial consultation for neonatal jaundice to final diagnosis of biliary atresia has to be minimized in countries like Bangladesh for a successful Kasai procedure, to save lives up to liver transplantation, though liver transplantation facility is hardly available in this country. Only a few economically well-off families moved abroad for liver transplantation, others from the low-income group without liver transplantation bear a bad prognosis from biliary cirrhosis and its complications.

Limitation

Small sample, single-center cross-sectional study.

Conclusion

The study showed that only 21.45% biliary atresia cases were referred to tertiary level hospital before 60 days of age in Bangladesh. Only 32% biliary atresia cases were referred for the Kasai procedure. Late referraldue to the above-mentioned factors, with prolonged hospital evaluation time, was responsible for the diagnostic delay of biliary atresia cases. Without a facility for liver transplant, biliary atresia cases from the low-income group bear a heartbreaking prognosis within infancy.

Recommendation

Raising awareness at the family, physician's and national level about the importance of timely diagnosis, paying attention to the attending physicians not to missing neonatal cholestasis /biliary atresia cases as breastmilk/physiologic jaundice, improving government facilities to reduce hospital diagnostic evaluation time and

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finally establishing a modern center for liver transplantation is of utmost importance for a better prognosis of biliary atresia cases in Bangladesh. Finally,a large sample multicenter study is recommended for further strengthening of the above-mentioned recommendations.

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Contribution of authors
WM-Conception, design, acquisition of data, drafting & final approval.
MSH-Conception, design, acquisition of data, drafting & final approval.
KF-Data analysis, interpretation of data, critical revision & final approval.
JM-Interpretation of data, critical revision & final approval.
RI-Interpretation of data, critical revision & final approval.
RT-Design, acquisition of data, drafting & final approval.
CSHK-Data analysis, critical revision & final approval.
SP-Interpretation of data, critical revision & final approval.

Disclosure
All the authors declared no conflict of interest.

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