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Paediatric Acute Liver Failure, aetiology & outcome: Experience of fifty five cases at a tertiary care hospital.

Md. Wahiduzzaman Mazumder*, ASM Bazlul Karim, Md. Rukunuzzaman*****

*Associate Professor,**Professor ***Associate Professor.
Department of Paediatric Gastroenterology & Nutrition, Bangabandhu Sheikh Mujib Medical University (BSMMU), Dhaka, Bangladesh.
Corresponding author: Md. WahiduzzamanMazumder, Associate Professor ,Department of Paediatric Gastroenterology & Nutrition, Bangabandhu Sheikh Mujib Medical University (BSMMU), Dhaka, Bangladesh. E-mail: mazumdermw@gmail.com Phone: 01718601459
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Abstract:

Background: Acute liver failure (ALF) is a rapidly progressive, potentially fatal syndrome resulting from rapid death or injury to a large proportion of hepatocytes, caused by a variety of insult, leaving insufficient hepatic parenchymal mass to sustain liver function. The aetiology of ALF varies according to the age of patient and development of the country. The outcome of ALF also varies according to aetiology: survival is better in paracetamol poisoning whereas it is poor in metabolic diseases.
Objective: The present study was undertaken to observe the underlying aetiology and outcome of ALF in children under 16 years of age admitted at the department of Paediatric Gastroenterology & Nutrition, Bangabandhu Sheikh Mujib Medical University, Dhaka, Bangladesh.
Methods: It was a retrospective review of medical records from November 2013 through February 2016.
Results: During this period a total of 55 patients were diagnosed to have ALF. Aetiology was established in 42 (76.4%) cases. Wilson's Disease (WD) was found in 23 (41.8%) cases, Viral hepatitis in 17 (30.9%), where as no identifiable cause was found in 13(23.6%) cases. After treatment 21 (38.2%) ALF patients survived, 17 (30.9%) left hospital with risk bond (DORB), and 17 (30.9%) patients died. The study showed that among the 17 death patients, 8 (29.4%) had viral hepatitis, 7 (41.2%) Wilson's disease, and in 5 (29.4 %) no cause could be identified.
Conclusion: Wilson's disease was found as the most common causes of Acute Liver Failure, around 38% cases of acute liver failure survived with supportive management in this study.Future studies with larger sample size are required to know the actual causes&outcome of acute liver failure in children.

Key words: Paediatric Acute liver failure, aetiology, children, out come.

Introduction:

Acute liver failure (ALF) is a rapidly progressive, potentially fatal syndrome resulting from rapid death or injury to a large proportion of hepatocytes, caused by a variety of insults, leading to insufficient hepatic paranchymal mass to sustain liver function. Paediatric ALF is defined as presence of biochemical evidence of liver injury (deranged transaminases) and coagulopathy not corrected by one dose of parenteral vitamin K administration with International Normalized Ratio (INR) of >1.5 in the presence of encephalopathy or an INR of >2 with no evidence of encephalopathy within 8 weeks of onset of liver injury without prior known existing liver disease[1]. The etiology of ALF varies according to the age of patient and development of the country[2-4]. The outcome of ALF also varies according to etiology: survival is better in few aetiologies like paracetamol poisoning whereas it is poor in metabolic diseases[4,5]. The study was undertaken to observe the underlying aetiology and outcome of ALF in children under 16 years of age admitted at the department of Paediatric Gastroenterology &

Nutrition, Bangabandhu Sheikh Mujib Medical University (BSMMU), Dhaka, Bangladesh.

Methods:

It was a retrospective review of medical records. All cases of ALF aged below 16 years of age diagnosed at the department of Paediatric Gastroenterology & Nutrition, BSMMU, from November 2013 to February2016 were reviewed in this study. Diagnosis of ALF was based on the revised definition given by paediatric ALF study group 1. All patients with past history of liver disease or had stigmata of chronic liver disease found during physical examinations were excluded from the study. All patients were tested for viral markers. Screening for Wilson disease (WD) & autoimmune hepatitis were done whenever it was necessary. All children below one year of age were investigated for galactosemia, neonatal iron storage disease and a1-AT deficiency according to our limited resource & laboratory setting, as confirmatory tests were not available, we have to depend on clinical ground and existing laboratory facilities for these cases. Liver biopsy was not done.All the ALF patients were managed according to the standard departmental protocol. Severity of hepatic encephalopathy was graded as per standard definition[5]. Aetiology of ALF was divided into two groups: Group1 (where underlying cause was identified) and Group 2 (where no cause was found). Group 1 was subdivided according to identified causes. Outcome of ALF was grouped into 3 categories: survived, left hospital on risk bond (DORB), and died during management at hospital.

Results:

During the study period a total of 55 patients were diagnosed to have ALF. Twenty one (38.2%) of 35 cases were below 5 years of age, 24 (43.6%) between 5-10 years, and 10 (18.2%) above 10 years of age (Table 1) and mean age was 7.8 years with range of 5 months to15 years.

Table 1: Age distribution of studied patients (n=55)

Age	No (%)
<5 yrs	21 (38.2)
5-10 yrs	24 (43.6)
>10 yrs	10 (18.2)

Of the 55 cases, 60 % were male. In this study, aetiology was identified in 42 (76.4%) cases, whereas in 13 (23.6%) cases no identifiable cause was found. Wilson's Disease (WD) was found in 23 (41.8 %) cases, and Viral hepatitis was found to be the underlying cause of ALF in 17 (30.9%) cases. HAV & HBV were found in 13 (23.6%) and 4 (7.3%) cases respectively.

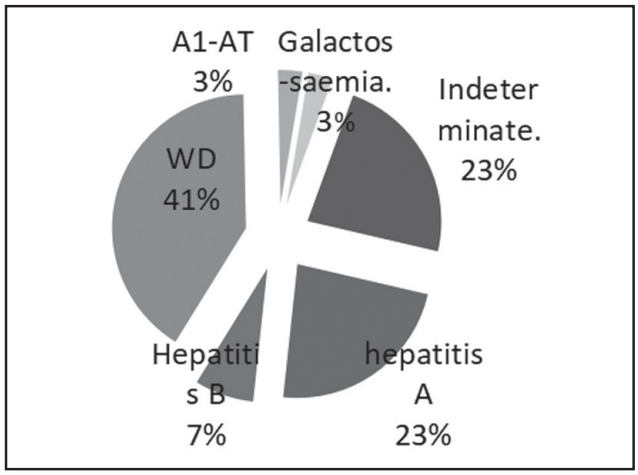

Figure 1: Aetiology of studied patients (n=55).

Among the metabolic causes, WD was found in 23 (41.8%) cases, galactosemia in 1 (3%) case and, a1-AT deficiency in 1 (3%) case (Figure 1). After treatment 21(38.2%) ALF patients survived and discharged with advice, 17 (30.9%) left hospital with risk bond (DORB), and 17 (30.9%) patients died (Figure 2). The study showed that out of 17 death, 5 (29.4%) had viral aetiology, 7 (41.2%) had WD, and in the remaining 5(29.4%) no cause was identified.

Discussion

In the present study an attempt was made to find out the aetiology of acute live failure in children and after doing available investigation it was

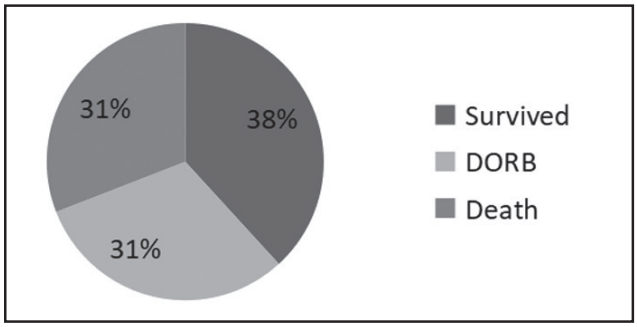

Figure 2:Outcome of studied patients (n=55)

identified in 76.4% of cases. In recent studies no cause (indeterminate) of ALF was found in up to 22% cases6-9,which is similar (23%) to the finding of present study. Results of present study high lights the fact that Wilson's disease remains the most common cause (41%) of ALF, where as viral hepatitis was the next common cause(30%) which is around 1% in developed countries[10]. Hepatitis A virus was found in 23% of ALF cases in the present study which is around 40% as shown in studies done in other developing countries[11]. Hepatitis A virus was found three times more common than Hepatitis B virus, this may be explained by the fact that prevalence of hepatitis B virus infection has markedly come down after launching of universal vaccination program against HBV. Regarding the metabolic causes, WD remains in top position (41%). Among the other metabolic causes, a1AT deficiency was found in 3% and galactosemia also in 3% cases only. This study also showed that with limited treatment facilities and without any facilities for liver transplantation, nearly 38% patients recovered fully. Death from ALF was found to be 31%, of which viral cause was 29.4% which is preventable. In 41.2% cases death occurred due to WD, which could also be minimised by early screening, treatment, and avoiding consanguineous marriage. Number of patients who left hospital (31%) on risk bond (DORB) were much higher, majority of them was in terminal stage. Causes behind discharge on risk bond (DORB)could be due to frustration from higher treatment cost, an anticipation of bad prognosis, also few of them were referredto specialised centre outside the country for liver transplantation.

Conclusion :

Wilson's disease was found to be the most common cause and viral hepatitis (Hepatitis A) found to be second common cause of Paediatric acute liver failure in this study. The study also showed around one third patients with acute liver failure survived, in the remaining two third cases outcome were poor. Future multi centre study with large sample size may show the exact aetiology & outcome of paediatric acute liver failure in our country.

Limitations:

The limitationsof the study includes: single centre study, small sample size and retrospective design.

What this study adds:

Simple interventions like improving hygienic practices, immunization against hepatitis A,B, and screening with early diagnosis of Wilson's disease cases could substantially decrease paediatric acute liver failure cases. By providing modern treatment facilities (Paediatric ICU, artificial liver support system and liver transplantation) death rate and rate of discharge on risk bond (DORB) could be further minimized to a much lower level.

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