

Clinical and Etiological Profile of Chronic Liver Diseases in Children Attending a Tertiary Care Centre

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Abstract

Introduction: Chronic liver disease (CLD) in children is a progressive condition characterized by persistent liver injury lasting more than three months, leading to fibrosis, cirrhosis, and impaired liver function. The etiological spectrum in children differs from adults and includes metabolic disorders, autoimmune diseases, viral infections, and biliary abnormalities, with varied clinical presentations such as jaundice, hepatomegaly, and growth failure.

Aim: To study clinical and etiological profile of chronic liver diseases in children <12 years attending a tertiary care centre.

Methods: This was a cross sectional study conducted in Paediatrics Department SAT hospital Thiruvananthapuram from January 2018 to July 2019. 90 children who were diagnosed to have CLD based on clinical, biochemical and ultrasound abdomen findings and/or liver biopsy and/or transient elastography, were included in the study. All patients were subjected to detailed history and thorough physical examination. Results of routine investigations and etiology directed investigations were recorded using structured proforma.

Results: Of 90 children included in the study mean age of patients was 4.1± 4.45yrs. Male to female ratio for the study population was 1.2:1. Jaundice (77%), ascites (27%), bleeding manifestations (17%), encephalopathy(5%) were presenting symptoms in chronic liver disease patients. 13% of patients were asymptomatic at presentation. 22.3% of patients presented with features of hepatic decompensation at presentation. Neonatal cholestasis syndromes (NCS), and metabolic liver disorders (MLD) were the leading causes of CLD (56.6% and 22.1%, respectively). NCS comprised extra hepatic biliary atresia (46.7%), neonatal hepatitis (3.3%), and paucity of interlobular bile ducts (2.2%). MLD included Wilson disease (16.7%) glycogen storage disease (2.2%) Niemann Pick disease (2.2%) and Tyrosinemia (1.1%). Other causes of CLD comprised Pediatric non alcoholic fatty liver disease (NAFLD) (8.9%), pediatric autoimmune liver disease (PAILD) (3.3%), congenital hepatic fibrosis (1.1%) and sclerosing cholangitis (1.1%). Aetiology could not be detected in 7.7% of patients. Chronic viral hepatitis B and C were rare cause CLD in study group (1.1%). Among children with EHBA 78% underwent Kasai's portoenterostomy with mean age at surgery being 66 days.

Conclusion: Common etiology of CLD in children was constituted by neonatal cholestasis syndrome (NCS) and metabolic liver disease (MLD) in the study population. EHBA was the commonest cause of CLD in children < 6 years of age whereas Wilson disease is the most common cause of CLD in older children (> 6 years). Pediatric NAFLD constitute 3rd most common cause of chronic liver disease which is important from a public health perspective. Jaundice, ascites and bleeding manifestation were the most common presentation of CLD in children. 22.3% of children presented with decompensated chronic liver disease in the study population.

Keywords: Chronic liver disease, children, neonatal cholestasis syndrome, metabolic liver disease, paediatric NAFLD.

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Introduction

A variety of liver diseases are encountered in children and they represent a growing health problem in children with significant morbidity and mortality.[1] Chronic Liver Disease (CLD) refers to wide spectrum of disorders characterized by ongoing liver damage with a potential for progression to cirrhosis / end stage liver disease. Unlike in adults long duration of disease is no longer considered mandatory aspect of definition of CLD in children, as the progressive irreversible changes can occur in children even with symptoms as short as 1 week [2]. It is also not unusual to see children with established cirrhosis presenting for the first time. CLD contributes 1-5% of ward admissions [3] one third of referral to paediatric gastroenterology OP from various centres in North India from the published studies. Children with CLD can have a varied clinical presentation or can remain asymptomatic till late in natural history of the disease. This protean presentation should be recognized early and diagnosis should be made to prevent progression to end stage liver disease (ESLD). The aetiology of CLD comprises a wide spectrum of disorders which vary according to the age of presentation. Indian childhood cirrhosis (ICC) which was the commonest cause of chronic liver diseases in India until 1970s, has become a rare entity. On the other hand, due to advances in the diagnostic techniques and availability of such facility, metabolic disease are being increasingly diagnosed in our nation. The two most important causes of chronic liver disease in Indian infants are biliary atresia and neonatal hepatitis. Wilson disease and autoimmune liver diseases are the most common causes in older children. Pediatric Non Alcoholic Fatty Liver Disease (NAFLD) is also being increasingly diagnosed in our children in recent years.

There have been major advancements available today, like development of vaccines which are highly effective against Hepatitis B, specific dietary therapy for metabolic diseases which when started at the appropriate age can completely prevent the disease progression. Enzyme replacement therapy for various storage disorders like glucocerebrosidase for Gauchers disease which can completely cure the disease. Now, liver transplantation, mostly Live donor liver transplantation (LDLT) has become a lifesaving procedure for children with end stage liver disease. Thus, considering these factors, it is important to have the knowledge of current spectrum of liver diseases in children. The present study tried to identify and describe clinical profile and aetiology of chronic liver diseases in children. Our centre is the only tertiary care hospital under public sector in the state of Kerala with a paediatric gastroenterology services. Also, there is paucity of published literature regarding the chronic liver diseases in children from South India. In this

context we undertook this study on the clinical and etiological profile of CLD in children.

Materials and Methods

This was a cross sectional study conducted at Paediatrics Department, SAT Hospital Thiruvananthapuram, a tertiary care referral centre in South India from January 2018 to July 2019. The study protocol was approved by Institutional Ethics Committee. All consecutive children below 12 years with chronic liver disease as evidenced by Persistent elevation of liver enzymes more than twice the normal, for a period of 3-6 months or more and /or other biochemical evidence and / or radiological evidence (ultrasound and /or transient Elastography /CT /MRI) and/or histological evidence of chronic liver disease obtained from liver biopsy, attending SAT Hospital Thiruvananthapuram were included in the study. Children with malignancy such as hepatoblastoma, hepatocellular carcinoma /secondaries were excluded. After taking written informed consent from parent, data was collected using structured questionnaire including detailed history, clinical examination, blood investigations, radiological investigations, liver biopsy and genetic studies. Data was collected from case files for already diagnosed patients and by direct examination for new cases who underwent diagnostic evaluation according to the hospital protocol. Data entry and statistical analysis were performed using Microsoft excel and SPSS software. Descriptive statistics were used to summarize the data. Continuous variables were expressed as median and interquartile range while categorical data were summarized as percentages.

Results

A total of 90 children with CLD presented to Paediatric Department, SAT hospital Thiruvananthapuram were included in the study. Table 1 describes the demographic and clinical features. 44% children with CLD were in <1 year of age, 29% between 1-7 years and 27% were 7-12 years of age. 53% of children in the study group were males. More than half of them were (n=48, 53%) newly diagnosed cases of CLD during study period, while others were known cases of CLD with re admissions. The most common feature on presentation was jaundice (n=69, 77%). Abdominal distension (33%), oedema (20%), and upper GI bleeding were other common manifestations. 4% of children had encephalopathy and abnormal behaviour and tremors were present 1 child (1%). 12 (13%) children were asymptomatic at presentation. On physical examination two third (78%) of children had icterus. Pallor (42%), ascites (38%), oedema (20%), palmar erythema (13%), clubbing (6%), KF ring (6%) were other physical examination findings. About two third

(75%) of children had hepatomegaly. splenomegaly was present in 41%.

Table 1: Demographic and clinical characteristics of children with CLD

Characteristics	No. of patients (N=90)
Age	
≤1 year	40(44%)
1-6 years	26(29%)
7-12 years	24(27%)
Sex	
Male	48(53%)
Female	42(47%)
Newly diagnosed	48(53%)
Old case	42(47%)
Symptoms	
Jaundice	69(77%)
Abdominal distension	30(33%)
Oedema	18(20%)
Upper GI bleeding	15(17%)
Encephalopathy	4(4%)
Abnormal behaviour	1(1%)
Tremor	1(1%)
Asymptomatic	12(13%)
Physical examination	
Icterus	70(78%)
Pallor	42(49%)
Ascites	38(42%)
Oedema	20(24%)
Palmar erythema	12(13%)
Clubbing	6(7%)
KF ring	5(6%)
Hepatomegaly	64(75%)
Splenomegaly	37(41%)

The laboratory investigations of the children in the study group showed elevated bilirubin >2 mg/dl in 68% of children. SGPT was raised 3 times the normal (<120) in 79% of children whereas in 76% children SGOT was raised. PT INR was prolonged in 30% of children. Out of 90 patients in the study group 21(23%) underwent liver biopsy and 7 (8%) underwent genetic study for etiological diagnosis.

Table 2: Laboratory investigations in children with CLD

Investigations	No. of patients (N=90)
Serum bilirubin (mg/dl)	
<2 mg/dl	29(32%)
>2 mg/dl	61(68%)
SGPT	
<120 meq/L	15(17%)
>120 meq/L	75(83%)
SGOT	
<120 meq/l	12(13%)
>120meq/l	78(87%)
PTINR	
Prolonged	26(27%)
Not prolonged	64(73%)
Liver biopsy	21(23%)
Genetic study	7(8%)

The etiological profile of the children in the study showed Neonatal Cholestasis syndrome 56.6% (51) and Metabolic Liver diseases 22.2% (20) as the leading cause of paediatric CLDs. NAFLD

constituted 8.9%(8) of patients with CLD. Cryptogenic causes of CLD was found in 7 (7.7%) and Autoimmune hepatitis was encountered in 3 (3.3%). We had 2 (2.2%) patients with chronic

viral hepatitis and 1 patient each with congenital hepatic fibrosis and Sclerosing cholangitis. Among the patients with NCS, Extra hepatic biliary atresia constituted the vast majority (46.6%, n=42) followed by Neonatal hepatitis (8%, n=7), among which 4(4%) patients had congenital CMV hepatitis while 3(3%) were idiopathic neonatal

hepatitis.2 (2.2%) patients with Alagelle syndrome were also present in study group. Among the patients with metabolic liver diseases, Wilson disease (15, n=17%) was the most common cause. Glycogen storage disorders 2(2%), Niemann pick disease2(2%), tyrosinemia 1 (1%)were the other causes found.

Table 3: Etiological profile of chronic liver disease in children

Etiology	No. of patients
	(N=90)
Neonatal Cholestasis syndrome	51(57%)
EHBA	42(47%)
CMV hepatitis	4(4%)
Idiopathic neonatal hepatitis	3(3%)
Alagelle syndrome	2(2%)
Metabolic Liver diseases	20 (22%)
Wilson disease	15(17%)
Glycogen storage disorder	2(2%)
Neimann pick disease	2(2%)
Tyrosinaemia	1(1%)
Pediatric NAFLD	8 (9%)
Cryptogenic	4 (4%)
Autoimmune hepatitis	3 (3%)
Chronic viral hepatitis	2 (2%)
Hepatitis B	1(1%)
Hepatitis C	1(1%)
Congenital hepatic fibrosis	1 (1%)
Sclerosing cholangitis	1 (1%)

Table 4: Clinical profile of EHBA (n=42)

Mean age at presentation	1.4 months
Male: female ratio	1.2:1
Underwent surgery(Kasai's portoenterostomy)	79%
Mean age at Surgery(KPE)	66 days
Post liver transplant	1 case
Presentation in decompensated stage	12(28.5)
Hepatomegaly	38(90.4%)
Splenomegaly	35(83.3%)
Ascites	20 (47.6%)
HIDA scan	33 (78.5%)
Preoperative Liver biopsy	6 (14.2%)

Table 5: Clinical profile of Wilson disease (n=15)

Clinical features	Number	Percentage
Male :female	1.14:1	
Mean age at presentation of hepatic Wilson	5.4years	
Mean age of presentation of neuro Wilson	9.5 years	
Mean duration from diagnosis	2.5 years	
Presentation as a/c hepatitis	5	30
Presentation as C/c hepatitis	3	18
Presentation as Fulminant hepatic failure	2	13.3
Mixed neuro +hepatic Wilson	2	12
Hepatic +renal	1	6
Asymptomatic patients	3	18

Discussion

In this study an attempt was made to elucidate the spectrum of chronic liver disease with regard to

their varying aetiologies and clinical presentation in children who presented to a tertiary level teaching hospital.90 children with CLD were included in the study. The mean age of chronic liver disease

patients was 4.1 years (range 1 month to 12 years), among which 44.4% < 1 year. This is lower than in a study in North India where the mean age of presentation was 8.5 years and 10.4 years in a study by Dar et al [5]. This younger mean age of presentation in our study may be attributed to the greater health awareness and treatment seeking behaviour of parents and easier access to health care facilities in this part of the country. Male to female ratio for the study population was 1.2:1; a finding similar to most previous studies [5,6,7] that reported male predominance from 58% to 69% in children with CLD. 22% of patients of CLD presented late with features of advanced liver disease and had a grim prognosis. Among them 45% were infants with EHBA. Jaundice (76.7%), ascites (27%), bleeding manifestations (17%) and encephalopathy (5%) were the common presenting symptoms at presentation. This is similar with the observations in a study among a cohort from northern India [4]. 13% of children had no symptoms of liver diseases at presentation. Majority of them were evaluated for other symptoms and found to have altered liver function tests and diagnosed to have CLD like NAFLD and Wilson disease as a part of family screening. 75% of the patients with CLD had hepatomegaly at presentation. This finding corroborates well with the observations made in many previous studies [4,7,8] that majority of children (63-85% in various studies) with CLD have enlarged liver at presentation. Most patients with chronic liver disease had elevated serum bilirubin (70%); 27% had coagulopathy and 20% of patients had features of portal hypertension. Neonatal cholestasis Syndromes were the most common (56.7%) etiology of chronic liver disease in the study group. Extra hepatic biliary atresia was the commonest cause of neonatal cholestasis identified. The mean age at presentation of EHBA was 44 days with a male to female ratio of 1.2:1. 79% patients with EHBA underwent Kasai's portoenterostomy with mean age at surgery being 66 days, which is primary treatment with expected five year native liver survival [10]. Neonatal hepatitis was second most common in NCS. Alagille syndrome was the 3rd cause of NCS. Similar pattern was reported in a study [1] in Egypt where they demonstrated that neonatal cholestasis constituted 41.05% of CLD. Karim B et al in his study has reported a mean age of presentation of EHBA as 3.5 months though mean age of presentation was 40 days in his study group [9]. Metabolic liver diseases constituted the second largest subset of children (22%) with chronic liver disease. Wilson disease was the commonest cause of metabolic disease. Most patients with Wilson disease presented with hepatic manifestations only, with mean age at presentation being 5.4 years in hepatic Wilson while 9.5 years in neuro Wilson. KF ring was a feature in 33%. GSD,

Neimann Pick disease and tyrosinaemia constituted other causes of metabolic liver disease. These findings were in agreement with other studies [11,12,13]. Pediatric NAFLD was seen in 7.8% of patients which constituted 3rd most common cause of CLD. Mean age at presentation was 10 years and all children were asymptomatic at presentation. 38% with pediatric NAFLD children were overweight and 38% were obese. Hepatomegaly was present in 75% and all had fatty liver in ultrasound. Several recent studies have demonstrated the frequency of NAFLD is rising in children [14-16]. The increasing incidence can be attributed to change in socioeconomic status, dietary habits and lifestyle changes. Pediatric autoimmune liver disease was seen in 3.5% patients with female predominance. This is in agreement with Rajeshwari and Gogia [4] who reported the incidence of AIH as 4%.

Chronic hepatitis B & C virus infection was a rare cause of chronic liver disease in children (only 1 patient each; 1.2%). Dangwal et al [16] also in a similar series observed that the prevalence of chronic viral hepatitis was rather low in children. Etiology of liver disease could not be found in 7.7% of children. This is contradictory to what was observed by Dar et al [18] as no etiology was found in 52% of cases. This large difference might be attributable to increasing awareness among treating doctors and improved facilities for diagnostic tests such as liver biopsy, metabolic work up, genetic study etc.

Conclusion

Common etiology of CLD in children was constituted by neonatal cholestasis syndrome (NCS) and metabolic liver disease (MLD) in the study population. EHBA was the commonest cause of CLD in children < 6 years of age whereas Wilson disease is the most common cause of CLD in older children (> 6 years). Pediatric NAFLD constitutes 3rd most common cause of chronic liver disease which is important from a public health perspective. Jaundice, ascites and bleeding manifestation were the most common presentation of CLD in children. 22.3% of children presented with decompensated chronic liver disease in the study population.

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