

## Aetiology and Clinical Profile of Children with Hepatomegaly in Pediatric Age Group

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### Abstract

*Introduction:* Liver disease in pediatric age group is one of the most significant causes of morbidity and mortality and includes a broad spectrum of disorders such as infections, developmental abnormalities, metabolic and neoplastic disorders that finally results in hepatic dysfunction and cirrhosis [1] and may recover fully.

*Objective:* 1)To study the clinical profile of hepatomegaly in pediatric age group. 2)To identify etiological spectrum hepatomegaly in pediatric age group. *Material and Methods:* A prospective study will be carried out during 01.12.2014 to 31.05.2016 in 100 patients of pediatric age group presenting with hepatomegaly admitted in Navodaya Medical College Hospital Raichur. All the pediatric patients (birth - 18 years) admitted with hepatomegaly, (defined clinically) with span more than normal for age. The patients will be evaluated clearly based on history, clinical examination and requisite laboratory parameters to find out the cause of hepatomegaly. This study will be conducted as a prospective Clinical study, wherein written informed consent will be taken prior to the investigation after detailed information given to the guardians regarding the study. Children with hepatomegaly will be screened through history, detailed examination and correlated through lab investigations. *Results:* Male predominance was seen in our study. Maximum incidence of hepatomegaly was seen between 1 to 4 year age group (34%). Majority of the cases had mild hepatomegaly (64%). Commonest cause of hepatomegaly was infections. Fever was the most common symptom associated with hepatomegaly. Splenomegaly was the most common sign associated with hepatomegaly. *Conclusion:* Infection is the most common cause of hepatomegaly in our study. Male preponderance was seen in majority of the cases who had hepatomegaly and belonged to pre school age group.

**Keywords:** Hepatomegaly; Anemia.

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### Introduction

Liver disease in pediatric age group is one of the most significant causes of morbidity and mortality and includes a broad spectrum of disorders such as infections, developmental abnormalities, metabolic and neoplastic disorders that finally results in hepatic dysfunction and cirrhosis [1] and may recover fully.

In developing tropic and sub-tropic countries hepatomegaly in pediatric age group is a common problem in medical practice.

*Hepatomegaly Generally Occurs via Five Mechanisms:*

1. Inflammation
2. Excessive storage
3. Infiltration
4. Congestion
5. Obstruction [2]

Early diagnosis and treatment of children who have liver disease is important because specific treatments are available for some diseases that can prevent disease progression or hepatic failure.

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Received on 03.08.2017, Accepted on 01.09.2017

As many number of pathological conditions and few secondary to cardiac conditions, pulmonary conditions are also often being seen associated with hepatomegaly the study is relevant to existing situation.

### Materials and Methods

Hundred consecutive cases, from birth to 18 years of age with varying grades of hepatomegaly of different etiologies, admitted in pediatric wards of NMCH&RC were studied from Jan2015 to Dec2015.

On admission, detailed study of each case including history, through physical examination and necessary investigations within the limitations of available laboratory facilities were done depending upon the history and clinical findings.

The examination of cases was done according to the given proforma. Enlargement of the liver less than 4cm below right subcostal margin was graded as mild, between 4-7 cm or upto the umbilicus is graded as moderate and more than 7cm or beyond the umbilicus as massive [3].

Routine investigations like complete blood count, peripheral smear, urine, and stool analysis, chest x-ray, mantoux test were done initially.

Relevant investigations like WIDAL, HIV, HbSAG, HAV, LFT, Bone marrow examination, bleeding time, clotting time, Hb electrophoresis, Liver biopsy, blood group, blood culture was done in relevant cases, depending upon the provisional diagnosis made on history and clinical examination.

### Observation and Results

#### Sex Distribution

**Table 1:** Shows sex incidence

Sex	No. of Cases
Male	62
Female	38

#### Age Distribution

**Table 2:** Shows the age distribution

Age in years	No. of Cases	Percentage
< 1year	16	16
1-4	34	34
5-8	29	29
9-12	12	12
13-18	09	09

**Table 3:** Shows Symptoms and Signs Associated with hepatomegaly:

Sl. No.	Symptoms	Percentage
1	Fever	73
2	Facial puffiness/edema of feet	19
3	Jaundice	37
4	Breathlessness/Hurried breathing	11
5	Pain in abdomen	10
6	Distension of abdomen	09
7	Vomiting	26
8	Mass in abdomen	08
9	Failure to thrive	04
10	Neonatal Jaundice	03
11	Altered level of conciousness	06
12	Convulsions	04
	<b>SIGNS</b>	
13	Splenomegaly	78
14	Anemia	48
15	Lymphadenopathy	52

**Table 4:** Shows grades of hepatomegaly

Grades of hepatomegaly	Percentage
Mild(below 4cms)	64
Moderate 4-7cms	32
Massive >7cms	04

**Table 5:** Shows etiological groups of 100 cases

Etiological Analysis	Percentage
Infectious	48%
Heamatologic	12%
Miscellaneous	14%
Congestive	18%
Malignancy	04%
Cholestatic	03%
Metabolic	01%

**Table 6:** Shows etiological analysis of 100 cases

<b>Infections</b>	
<b>A. Acute</b>	48
Infective Hepatitis	18
Enteric fever	09
Malaria	08
Dengue	04
Pyogenic meningitis	02
Infective endocarditis	01
<b>B. Chronic</b>	
Tuberculosis	04
Chronic hepatitis	02
HIV	02
<b>Hematological :</b>	
Thalassemia	08
Sickle cell disease	03
Hereditary spherocytosis	01
<b>Miscellaneous:</b>	<b>14</b>
PEM	08
Rheumatic fever	04
Juvenile rheumatoid arthritis	02
<b>Malignancy</b>	<b>04</b>
Leukemia	02
Lymphoma	01
Neuroblastoma	01
<b>Congestive</b>	<b>18</b>
CHD	14
Constrictive pericarditis	03
Budd-chiari syndrome	01
<b>Cholestatic</b>	<b>03</b>
Biliary atresia	02
Gall stones	01
<b>Metabolic</b>	<b>01</b>
Glycogen storage disease	01
<b>Hematological :</b>	
Thalassemia	08
Sickle cell disease	03
Hereditary spherocytosis	01
<b>Miscellaneous:</b>	<b>14</b>
PEM	08
Rheumatic fever	04
Juvenile rheumatoid arthritis	02
<b>Malignancy</b>	<b>04</b>
Leukemia	02
Lymphoma	01
Neuroblastoma	01

<b>Congestive</b>	<b>18</b>
CHD	14
Constrictive pericarditis	03
Budd-chiari syndrome	01
<b>Cholestatic</b>	<b>03</b>
Biliary atresia	02
Gall stones	01
<b>Metabolic</b>	<b>01</b>
Glycogen storage disease	01

Table 7: Shows age incidence in each etiological group

Age	Infection	Hematologic	Congestive	Malignancy	Cholestasis	Miscellaneous	Storage
<1yr	01	04	08	00	02	01	0
1-4y	14	06	05	02	01	05	01
5-8y	19	02	03	01	0	04	0
9-12	07	00	02	01	0	02	-
13-18	07	00	-	-	-	02	-

Table 8: Shows incidence of each grade of splenomegaly in each etiological group

Grades of hepatomegaly	Infections	Hematological	Miscellaneous	Congestive	Malignancy	Storage Disorders	Cholestasis
Mild(2-4) cms	37	06	10	10	00	00	01
Moderate(4-7)cm	11	05	04	07	02	01	02
Massive >7cm	00	01	00	01	02	00	00
Total	48	12	14	18	04	01	03

Table 9: Shows analysis of consistency of 100 cases

Consistency	Percentage
Soft	88
Firm	12

## Discussion

### Age Incidence

In this study 100 cases were studied with ages ranging from birth to 18 years of age. 16% of cases belonged to the age group of birth to 1 year, 34% from one to 4 years, 29% from 5 to 9 years of age, 12% from 9 to 12 years of age and 9% from 13-18 years of age. The data suggests that incidence of hepatomegaly is higher in children below 9 years of age. It may be due to common occurrences of infections in this age group like viral hepatitis.

### Sex Incidence

Males were affected more than females. In our study 62% of the affected children were males and 38% were female. Studies on various diseases which cause hepatomegaly like dengue fever, visceral leishmaniasis, chronic myeloid leukaemia also show male preponderance [4,5,6].

### Presenting Symptoms

Children in our study presented with various

symptoms depending upon the etiology some symptoms were specific whereas others were vague.

1. *Fever*: Fever was the most common presenting symptom seen in 67% of children of our study. It was seen in infections, infestations and malignancies.
2. *Vomiting*: Vomiting is seen in 26% of the cases was associated with various infections like TB meningitis due to raised ICT, in enteric fever, in viral fever associated with gastritis and mechanical discomfort as in ascites.
3. *Jaundice*: Jaundice was the presenting symptom in 37% of cases and was associated with cirrhosis of liver, infective hepatitis, typhoid due to hepatocellular dysfunction, It was seen in haemolytic anemias due to degradation of abnormal haemoglobin.
4. *Swelling of the face or feet*: Swelling of the face or feet was seen in 19% of cases and associated with severe anemias, PEM and congestive cardiac failure.
5. *Breathlessness*: Breathlessness/hurried breathing was seen in 11% of cases was due to associated

lung infections or congestive heart failure.

6. *Pain in abdomen*: Pain in abdomen was associated with 10% of cases due to tumours and acute hepatitis.
7. *Distension of abdomen*: Distension of abdomen was seen in 9% of cases may be due to organomegaly or free fluid like ascites.

#### Signs

The commonest sign associated with hepatomegaly was splenomegaly followed by anemia and lymphadenopathy.

1. *Splenomegaly*: Splenomegaly was seen in 78% of cases was associated with infections, haemolytic anemias, leukemias and congestive disorders.
2. *Anemia*: Anemia seen in 48% of cases was associated with haematological disorders, malignancy, congestive disorders and storage diseases.
3. *Lymphadenopathy*: Lymphadenopathy seen in 52% of cases was associated with leukemias, HIV, viral fever, enteric fever.

In a study from Brazil fever, pallor, weight loss and jaundice were the most common presenting symptoms [6]. Most studies show that fever is the most common presenting symptom associated with hepatomegaly [6,7,8]. A study on hepatosplenomegaly and anemia also observed that fever was the most common presenting symptom followed by abdominal distension, pallor, failure to thrive, oedema, dyspnea and jaundice [9].

#### Etiological Analysis of Hepatomegaly

1. *Infections Group*: In the present study, 48% of cases shows infectious etiology forming the most common cause of hepatomegaly in children. Out of those 48% cases 40% are acute infections and other 8% are chronic infections.

Among the 48% of acute infections group infective hepatitis 18%, enteric fever 9%, malaria 8%, dengue 4%, pyogenic meningitis 2%, infective endocarditis 1%. Among the 8% cases of chronic infectious group tuberculosis 4%, chronic hepatitis 2%, HIV 2%.

According to WHO infectious diseases accounted for 41.5% of total global DALY (Disability Adjusted Life Years). Communicable diseases like malaria, tuberculosis, HIV are highly prevalent. A study done in 2011 in Guntur shows infectious group as the most common cause of hepatomegaly in children. In a study done in Uttaranchal shows infectious group as the most common cause of hepatomegaly in children [10].

2. *Congestive Group*: It comprised of 18% of cases in our study out of which 14% were due to congenital heart diseases, 3 were due to acquired constrictive pericarditis, and 1% due to Budd-Chiari syndrome. As compared to a similar study done in Guntur congestive causes in our study are low [10].

3. *Hematological causes*: This constituted 12% cases in our study. Out of which 8 were thalassemia major, 2 were sickle cell disease and one was hereditary spherocytosis.

A study from Mumbai, on hepatosplenomegaly with anemia found thalassemia major as most common hemoglobinopathy on HB electrophoresis [9].

4. *Miscellaneous*: 14% of cases belonged to miscellaneous group. Out of which 8 were due to PEM, 4 were due to rheumatic fever, 2 were due to juvenile rheumatoid arthritis.
5. *Malignancy*: 4 cases belonged to malignancy group out of which 2 were due to acute lymphoid leukemia, one was due to lymphoma, and the other neuroblastoma.
6. *Cholestatic*: 3 cases were due to cholestasis out of which 2 were due to biliary atresia, and one was due to gall stones.
7. *Storage disorders*: Storage disorders comprised of only 1% in our study.

#### Analysis by Consistency

In our study consistency was evaluated based on palpation, 88% of livers are soft in consistency which is because of infections as predominant etiology, 12% are firm in consistency.

#### Analysis by Area

In our study majority of the are from rural areas 72% compared to urban as most of the cases admitted in our wards are from rural background and infection susceptibility is more in lower socio economic status.

#### Etiological Analysis of 100 Cases

Among the 9 cases of enteric fever only 3 had blood culture positive reports, others were diagnosed based on history, clinical examination, and positive Widal test. There were 8 cases of malaria 5 had positive peripheral smear for *Pl. falciparum*, 2 were positive for *Pl. vivax*, 1 had mixed *Pl. falciparum* and *vivax*. 18 cases were due to infective hepatitis Hepatitis A virus (HAV) positive in 7 cases, IgM positive in 4

cases and 1 case having HbsAg positive. One case had infective endocarditis, and had positive blood culture reports. Two cases had pyogenic meningitis which was confirmed by CSF study, 4 cases had dengue fever which was confirmed by dengue IgG and IgM, NS1Ag. Ten cases had chronic infections out of which 2 were positive for HIV, all were confirmed by positive HIV elisatest. Both of them had lymphadenopathy with diarrhoea and pyoderma. Four cases had tuberculosis out of which two had abdominal tuberculosis confirmed by ascitic tap, and two had pulmonary tuberculosis.

In a study done on typhoid fever, 71 children presented with fever and gastro intestinal symptoms including abdominal pain, diarrhea, nausea, vomiting and constipation. Hepatosplenomegaly was most common physical sign followed by abdominal tenderness [46]. Another study on HIV from Mumbai showed that hepatosplenomegaly lymphadenopathy and opportunistic infections together in a child may suggest of HIV infection [49].

Fever, sweating, and hepatomegaly were common clinical findings in children with brucellosis [50]. Hepatomegaly was in 72% of cases in dengue fever [43]. In a study from central India, 70% of fever cases had malaria, out of which, 87%, were caused by pl. falciparum [51].

#### *Hematological Disorders*

These constituted 12% of patients of our study, 8 were thalassemia major, 3 were sickle cell disease, one due to hereditary spherocytosis.

Hemoglobin levels ranged from 3 to 12.2gms/dl. Hemoglobin levels, retic count and peripheral smear gave clue to diagnosis in many cases. Hemolytic diseases like thalassemia, were confirmed by Hb electrophoresis. 50 cases with hepatosplenomegaly with anemia were studied in Mumbai Hb electrophoresis showed 10 out of 12 cases with abnormal electrophoresis pattern, whereas one each had sickle cell anemia and Hbs/â thalassemia. The presenting features were fever followed by abdominal distension, failure to thrive, oedema, dyspnoea, jaundice. Hepatosplenomegaly and pallor were the main clinical signs [9].

#### *Miscellaneous*

14% belonged to the miscellaneous group. Out of which 8 were due to PEM, 4 were due to rheumatic fever, 2 were due to juvenile rheumatoid arthritis.

Two cases were juvenile rheumatoid arthritis diagnosed by positive RA factor. Eight cases were

PEM diagnosed by clinical features and examination. 4 cases of rheumatic fever diagnosed after fulfilling modified Jones criteria.

#### *Congestive Group*

18% belonged to the congestive group out of which 14 were of CHD 6 had VSD, 3 had PDA, 3 had TGA, one had trisuspid atresia and one had ebsteinsanomaly. They were diagnosed by chest x-ray, 2-D Echo and clinical findings. Y. Shivaramakrishna study showed 32% belonged to congestive group [10].

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#### *Malignancy*

4 cases belonged to malignancy group, out of which 2 were acute myeloid leukemia, one was acute lymphoblastic leukemia and one was neuroblastoma. The cases were confirmed by bone marrow by bone marrow examination. All cases of malignancy had significant lymphadenopathy. In a study in 530 patients with acute lymphoblastic leukemia from India, the authors found that the most common features were lymphadenopathy and hepatosplenomegaly [11].

#### *Cholestatic*

3 cases belonged to cholestatic group out of which 2 were due to biliary atresia diagnosed by triangular cord seen on USG and one was due to gall stones diagnosed by x-ray and USG.

#### *Storage Disorders*

One case belonged to storage disorder and it is Glycogen storage disease diagnosed by clinical features and liver biopsy.

#### **Conclusions**

- Maximum incidence of hepatomegaly was seen in 1-4 years age group and with Male preponderance.

- Mild hepatomegaly (2-4cm) was the most common grade of hepatomegaly. Commonest causes were infectious causes.
- Fever was the most common presenting feature in our study associated with hepatomegaly.
- Among infections, infective hepatitis 18% is the most common cause followed by enteric fever 9%.

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