

Research Article**Study of Etiological and Clinical Profile of Hepatosplenomegaly in Children between 1 Month and 15 Years of Age****G Anusha, G Somaiah*, Ashraf Mohinuddin Siddique, B Srikanth, M Suresh Babu, NS Vamsidhar**

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Abstract: Hepatosplenomegaly is the simultaneous enlargement of the liver (Hepatomegaly) and spleen (Spleno-megaly). Hepatosplenomegaly is a common finding in infants and children with various etiological factors. It may be very easy to find the diagnosis or it may require extensive evaluation in order to distinguish benign, self limited disease process from serious life threatening conditions involving liver and spleen. The Present study was conducted to evaluate the possible etiology and clinical features of children presented with Hepatosplenomegaly. A total of 150 cases, from One Month to Fifteen years of age with Hepatosplenomegaly admitted in the Pediatric wards of Mamata General Hospital from January 2012 to October 2013 were included in this study. The incidence of Hepatosplenomegaly was 43.3% in age group of 6-10 years, followed by 42% in age group of 0-5 years and 14.7% in age group of 11-15 years in the present study. Infectious diseases mainly Malaria which is very much prevalent in the surrounding rural agency areas of Khammam and Bhadrachalam is the common cause of hepatosplenomegaly in the present study followed by hematological, storage and congestive disorders.**Keywords:** Hepatosplenomegaly, Liver span, Spleno-megaly.

INTRODUCTION

Hepatosplenomegaly is the simultaneous enlargement of the liver (Hepatomegaly) and spleen (Spleno-megaly). Hepatosplenomegaly is a common finding in infants and children with various etiological factors. Hepatosplenomegaly is a sign seen in various disease processes. Normal liver size estimations are based on age related clinical indices, such as the degree of the clinical extension of the liver edge below the costal margin, the span of dullness to percussion, or the length of the vertical axis of the liver, as estimated from imaging techniques [1]. Hepatomegaly was defined as when the liver span should be more than the expected for the corresponding age. Infants 5-6.5cms; 1-5yrs 6-7cms; 5-10yrs 7-9cms; 10-15yrs 8-10cms [2]. Spleno-megaly was classified into 3 grades on clinical grounds i.e., Mild (1-3cms), Moderate (4-7cms) and Massive (>7cms) [3].

Liver is regarded as chemical factory of the body [4]. Francis Kiernan has introduced a concept that the unit of hepatic structure is a hexagonal lobule, consisting of a terminal tributary of the hepatic vein in the centre and the portal tract containing bile duct, portal vein radical and hepatic artery branch at the periphery [5]. The lining of sinusoids consists of endothelial and phagocytic cells of RES, Kupffer cells [6]. It has to do manifold magic performances of complex nature but

with simple structure. "It is this combination of apparent simplicity with actual complexity which is the despair of all who attempt to correlate to the structure with disorder of function of liver" (William Boyd). The liver gets a dual blood supply from the hepatic portal vein and hepatic arteries. Approximately 80% of the liver's blood supply is through the hepatic portal vein. The remaining 20% of blood supply is by the hepatic arteries [7].

The spleen in infants and children is commonly involved in a variety of pathological process. Some of these processes cause isolated splenic disease whereas other involve the spleen as a part of a systemic illness [8]. Enlargement of the spleen may result from increase in its vascular, lymphoid or reticuloendothelial elements. An attempt is made in the present study to know the various etiological factors and clinical features of Hepatosplenomegaly in the cases admitted in Mamata General Hospital. The aim of the present work was to study the possible Etiology, Clinical features and Evaluation of children presented with Hepatosplenomegaly.

MATERIALS AND METHODS

A total of 150 cases, from One Month to Fifteen years of age with Hepatosplenomegaly admitted in the Pediatric wards of Mamata General Hospital, were

included in this study. Those children presented with pushed down liver due to various causes are excluded by measuring liver span for that particular age. Hepatomegaly was defined as when the liver edge was palpated below the right costal margin at the mid clavicular line for infants and older children and the liver span should be more than the expected for the corresponding age. Infants 5-6cms; 1-5yrs 6-8cms; 5-10yrs 8-9cms; 10-14yrs 8-12cms. Splenomegaly was classified into 3 grades on clinical grounds i.e., Mild (1-3cms), Moderate (4-7cms) and Massive (>7cms)³. A detailed study of the cases including history, thorough physical examination and necessary investigations depending on the history and clinical findings were done. Day to day progress has been observed during the hospital stay and after discharge, the cases have been followed up every month. Routine investigations like complete blood picture, peripheral smear, urine analysis, stool examination, Chest X Ray, Mantoux tests were performed in all cases. Relevant investigations like Widal Test, Pediatric HIV ELISA, Liver function tests, Bleeding time, Clotting time, prothrombin time, Hb Electrophoresis, Blood culture, X ray skull, X ray long bones were done in relevant cases, depending upon the provisional diagnosis made on history and clinical examination. Special investigations like CT scan, Sickling test, Osmotic fragility test were done in few cases wherever indicated.

RESULTS AND DISCUSSION

One hundred and fifty consecutive cases, from one month to fifteen years of age with Hepatosplenomegaly of different Etiologies admitted in Pediatric wards of Mamata Medical College, Khammam, were analysed to evaluate the following data. In the present study 43.3% cases are represented from 6-10 years of age group followed by 42% cases of 0-5 years age group. 14.7% cases are represented from 11-15 years of age group. Male preponderance was seen in the present study accounting for 58.67% while female cases accounts to 41.33%. The most common presenting features in our study are fever (80.7%), anemia (66.67%) and jaundice (37.33%), followed by loss of appetite (34.67%), pain

abdomen (30.67%), lymphadenopathy (20%), abdominal distension (18%), FTT (16%), Rash (10.67%) and breathlessness (8%).

Etiological Analysis (Table 1)

Infectious group

In our study, 68% of cases showed infectious etiology forming the most common cause of hepatosplenomegaly in children. In the infectious group, 29.33% were due to Malaria, 13.33% were of Enteric Fever, 11.33% were due to Viral Hepatitis [9,10] , 6.67% due to Tuberculosis, 4.67% due to Dengue fever, 2.67% are due to Septicemia.

Hematological disorders

These constituted 22.67% of patients of our study, forming the second major group. Out of which 11.33% were Thalassemia [11], 8.67% were due to Sickle cell disease [12-14], 1.33% due to hereditary spherocytosis and 1.33% due to hereditary persistence of fetal Hb [15].

Congestive group

It constitutes about 2% of Hepatosplenomegaly cases of which all 3 cases were congestive cardiac failure.

Storage disorder

These constituted about 2% in our study, consisting of 2 cases of Mucopolysaccharidosis (Morquio's syndrome) [16] and 1 case of Gaucher's disease [17, 18].

Connective tissue disorders

These constitute about 2% of the cases in our study, all these 3 cases were Juvenile Idiopathic Arthritis [19].

Miscellaneous

This group constituted 3.33%, which consisted of 3 cases of Protein Energy Malnutrition, diagnosed based on history and Anthropometry and 2 cases of Osteopetrosis [20, 21], diagnosed based on clinical examination and characteristic X-Ray changes.

Table 1: Shows Etiological Analysis of 150 Cases

Infections	102	68%
Malaria	44	29.33%
Enteric fever	20	13.33%
Viral Hepatitis	17	11.33%
Tuberculosis	10	6.67%
Dengue Fever	7	4.67%
Septicemia	4	2.7%
Hematological	34	22.67%
Thalassemia	17	11.33%
Sickle cell Anemia	13	8.7%
Hereditary Spherocytosis	2	1.33%
Hereditary persistence of Fetal Hb	2	1.33%
Storage Disorders	3	2%
Mucopolysaccharidosis	2	1.33%
Gaucher's Disease	1	0.67%

Congestive	3	2%
Congestive Cardiac Failure with Infective Endocarditis	3	2%
Connective Tissue Disorder	3	2%
Juvenile Idiopathic Arthritis	3	2%
Miscellaneous	5	3.3%
Osteopetrosis	2	1.33%
Protein Energy Malnutrition	3	2%

Table 2: Shows age incidence in each etiological group observation

Age	Infectious	Hematologic	Congestive	Storage Disorder	Connective Tissue Disorder	Misc.	Total
0-5	44(29.33%)	14(9.33%)	1(0.6%)	1(0.6%)		4(2.7%)	64(42.56%)
6-10	45(30%)	12(8%)	2(1.33%)	2(1.33%)	3(2%)	1(0.7%)	65(43.4%)
11-15	13(8.67%)	8(5.33%)					21(14%)
Total	102(68%)	34(22.67%)	3(2%)	3(2%)	3(2%)	5(3.4%)	150(100%)

Age incidence in each etiological group

In our study 42.5% belonged to 0-5 year age group. Out of which 29.33% belonged to infectious group, 9.33% belonged to Hematological, 0.67% Congestive, 0.67% Storage Disorder, 2.67% miscellaneous. 43.4% belonged to 6-10 years age group. Out of which 30% are Infectious group, 8% Hematologic group, 2% Connective tissue disorder, 1.33% congestive, 1.33%

storage disorder and 0.67% Miscellaneous. 14% belonged to 11-15 years age group. Out of which 8.67% are Infectious group and 5.33% Hematologic group (table 2). In all the age groups, the major cause of Hepatosplenomegaly was infectious diseases. Infectious diseases constituted 68.75% (44 out of 64) in 0-5 age group, 69.2% (45 out of 65) in 6-10 age group and 68.42% (13 out of 19) in 11-15 age group.

Table 3: Comparitive studies of hepatosplenomegaly with other studies

	Fever	Infectious	Anemia	Hepatitis	Hematologic	Congestive	Storage disorder	Connective tissue Disorder	malignancy	Jaundice
Bricks LF [22]	44%	39%	79%	7%	-	-	8%	-	6%	16%
Ali N [23]	-	-	-	-	73%	-	9%	-	18%	-
Present study	80.7%	68%	66.7%	11.33%	22.67%	2%	2%	2%	-	37.3%

The relative incidence of the cause of hepatosplenomegaly is subject to geographical variation. In western countries, the leukemias, malignant lymphomas, myeloproliferative disorders, hemolytic anemias and portal hypertension account most of the cases. In Tropical countries, however, the incidence of these hematological causes of hepatosplenomegaly is swamped by the greater preponderance of hepatosplenomegaly caused by parasitic tropical infections like Malaria, tuberculosis (Table 3).

To conclude, hepatosplenomegaly is a common pediatric problem. Hepatosplenomegaly is highest in 6-

10 years of age group. Fever, pallor, jaundice and anorexia are the common presenting complaints. Anemia, jaundice, fever, stunting, wasting and abdominal distension are the common examination findings. Fever is most commonly seen in infectious causes. Pallor is predominantly seen in Hemolytic anemias and infections like Malaria. Jaundice was commonly the presenting feature of infectious causes like viral hepatitis, Malaria. It was seen in Hemolytic anemias due to degradation of abnormal Hemoglobins. Abdominal distension is one of common symptom. Hepatomegaly, splenomegaly and ascites contribute to distension. Rash is associated with Dengue fever, septicemia. Failure to thrive was seen in 16% of the

cases. Lymphadenopathy is associated with Tuberculosis, Enteric fever. Pain abdomen is associated with Enteric fever, Viral Hepatitis, Dengue fever. Breathlessness (8%) is associated with congestive cardiac failure, Septicemia etc.

Lucia F Bricks *et al.* [22] study showed 6.7% cases with breathlessness. Loss of appetite is associated with Tuberculosis, Enteric fever and Viral Hepatitis. The liver enlargement varied from 2.5cm to 6cm and the spleen enlargement varied from 2cm to 9cm. Infections and Hemolytic Anemias are the most common diagnosis detected in the study. Malaria is the most common infection detected in the present study. An investigation required varies with the type of case presentation. The progression of the disease varied in different conditions. Growth Retardation was noticed in majority of storage disorders and Protein energy malnutrition. Infections, Connective tissue disorders, Storage disorders and congestive disorders are found commonly between 6-10yrs of age. Extramedullary hematopoietic disorders (hemolytic anemias, Osteopetrosis) are commonly found between 0-5yrs of age. Hepatosplenomegaly with lymphadenopathy is most commonly associated with Tuberculosis in the present study.

REFERENCES

1. Kliegman RM, Stanton BMD, Geme JS, Schor N, Behrman RE; Nelson textbook of paediatrics. 19th edition, Philadelphia, Saunders, 2011.
2. Paul V, Bagga A; Ghai Essential paediatrics, 8th edition, CBS Publishers, 2013: 310.
3. Lakshmanaswamy A; Clinical Paediatrics History Taking and Case Discussion, 3rd edition, Wolters Kluwer Health/Lippincott Williams & Wilkins (India), 2013: 466.
4. Askenazi S; Mimouni F; Merlob P; Litmanovitz I, Reisner SH; Size of liver edge in full-term, healthy infants. *Amer J Dis Child.*, 1984; 138(4): 337-378.
5. Malarkey DE, Johnson K, Ryan L, Boorman G, Maronpot RR; New Insights into Functional Aspects of liver morphology. *Toxicol Pathol.*, 2005; 33(1): 27-34.
6. Douglas M, Bennet GC; Principles and practice of infectious diseases. 7th edition, Newyork Churchill livingstone.
7. Longo DL, Fauci A, Kasper D, Hauser S, Jameson JJ, Loscalzo J; Harrison's principles of Medicine, 18th edition.
8. Paterson A, Frush DP, Donnelly LF, Foss JN, O Hara SM, Bisset GS; A pattern oriented approach to splenic imaging in infants and children. *Radiographics*, 1999; 19(6): 1465-1485.
9. Podymova SD; Acute hepatitis in infectious disease. *Eksp Klin Gastroenterol.*, 2013; 4: 38-43.
10. Abraham P; Viral hepatitis in India. *Clin Lab Med.*, 2012; 32(2):159-174.
11. Martin A, Thompson AA; Thalassemias. *Pediatr Clin North Am.*, 2013; 60(6):1383-1391.
12. Quinn CT; Sickle cell disease in childhood: from newborn screening through transition to adult medical care. *Pediatr Clin North Am.*, 2013; 60(6):1363-1381.
13. Shafiq M, Ali N; Bone marrow necrosis - initial presentation in sickle cell anemia. *Am J Case Rep.*, 2013; 16;14: 416-418.
14. Aloni MN, Tshimanga BK, Ekulu PM, Ehungu JL, Ngiyulu RM; Malaria, clinical features and acute crisis in children suffering from sickle cell disease in resource-limited settings: a retrospective description of 90 cases. *Pathog Glob Health.*, 2013; 107(4):198-201.
15. Forget BG; Molecular basis of hereditary persistence of fetal hemoglobin. *Ann N Y Acad Sci.*, 1998; 850: 38-44.
16. Pagni L, Bartolozzi L, Giacchetti D; Mucopolysaccharidosis. A case report of Morquio's type-A disease. *Minerva Stomatol.*, 1992; 41(11): 527-533.
17. Jadhav MV, Landge MP, Surana S, Sawaimoon SK; Gaucher's disease: report of 4 cases. *Indian J Pathol Microbiol.*, 2007; 50(4): 766- 768.
18. Kaplan P, Baris H, De Meirleir L, Di Rocco M, El-Beshlawy A, Huemer M *et al.*; Revised recommendations for the management of Gaucher disease in children. *Eur J Pediatr.*, 2013;172(4): 447-458.
19. Correll CK, Binstadt BA; Advances in the pathogenesis and treatment of systemic juvenile idiopathic arthritis. *Pediatr Res.*, *Pediatr Res.*, 2014;75(1-2):176-83.
20. Marks SC Jr.; Osteopetrosis--multiple pathways for the interception of osteoclast function. *Appl Pathol.*, 1987; 5(3): 172-183.
21. Usta M, Gulec SG, Karaman S, Erdem E, Emral H, Urgancı N; A case report of malignant infantile osteopetrosis. *Iran J Pediatr.*, 2012; 22(3): 421-424.
22. Ali N, Anwar M, Ayyub M, Nadeem M, Ejaz A, Qureshi AH *et al.*; Hematological evaluation of splenomegaly. *J Coll physicians Surg Pak.*, 2004; 14(7): 404-406.
23. Bricks LF1, Cocozza AM, Resegue R, Sucupira AC, Rodrigues D, Kobinger ME *et al.*; Experience in the evaluation of children with hepatosplenomegaly at a teaching ambulatory SAO Paulo, Brazil. *Rev Inst Med Trop Saopaulo.*, 1998; 40(5): 269-275.