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A study of hepatic function in multiple transfused thalassaemia patients

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***Abstract*---**Background: Thalassemia syndomes are the most common genetic disorder on worldwide basis. The selective pressures that have made the thalassemias so common are known but are assumed to relate to the geographic distrubution of malaria. Material & Methods: This was Prospective study conducted in the department of pediatrics in a teriary care hospital. The patients who had received more than 50 blood transfusions are considerd as multiple transfusion. Result: A total of 50 patients were enrolled in the study 90 % of patients were under 15 years of age male, female ratio was 1.27:1. The prevelance of thalassemia syndrome was common in sindhis and punjabis pupulation. Conculsion: The blood should be always screened for viral maricers, HIV and other blood born infections all thallsemai patients should be immunized with hepatitis B vaccine if patients are screnegative for HIV infection.

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Introduction

In 1925 Cooley and Lee described five children with anaemia, hepatosplenomegaly, pigmention of the skin, thickening of long bones and skull, decreased osmotic fragility and leukocytosis. The term'thalassemia' devived from the Greek'θ ά λ ά σ σ ά the sea was first used in 1936 by whipple and bradford [1]. All though there are more then 200 mutions for beta-thalassemia, most are rare. About 20 common alleles consitiute 80% of the known thalassemias worldwide. The thalassemia syndromes are heterogeneous group to mendelian disorders, all characterized by lack of or decreased synthesis of either the alpha or the beta globin chain of Hemoglobin A ($ά_2$ $β_2$) β- thalassemia is characterized by deficient synthesis of the beta blobin chain, whereas ά – thalassemia is characteized by deficient synthesis of the alpha chain. The hematologic consequences of diminished synthesis of one globin derive not only from the low intracellular hemoglobin (hypochromia) but also the relative excess of the other chain.[2]
The quantitative defect in globin chain synthesis are defined as thalassemia namely:

1. β – thalassemia – decreased β – chain synthesis
2. ά- thalassemia - decreased ά – chain synthesis
3. ά, β – thalassemia – decreased ά ,β' – chain synthesis

β – thalassemia syndromes can be clasified into two categories:

1. $β^0$ – thalassemia: Total absence of beta – globin chain in homozygous state.
2. $β^+$ – thalassemia: Reduced (but detectable) beta-globin synthesis in homozygous state.

Most of result from point mutation

The abnormality common to all beta- thalassemias is a total lack or a reduction in the synthesis of structurally normal beta- globin with unimpaired synthesis of alpha chanis. In β – thalassemia there is an excessive of alpha chain. As a consequence, free alpha chain trend to aggregate into insoluble inclusions within erythrocytes and their precursors, causing premature destruction of maturing erythroblasts with in the marrow (ineffective erythropoiesis) as well as lysis of mature red cells in the spleen (Hemolysis) in thalassemia.

As per WHO estimates 4.5% of the world population are carriers of hemoglobinopathies. The largest concentration of thalassemia patients is seen in South- East Asia, Shri Lanka, Bangladesh, North West India, Pakistan, Middle East Countires North Greece and Italy [3] Thalassemia itself and the complications or iron overload can damage multiple organ systems. The various pathological changes in heart, liver, endocrine system etc are due to:

- Iron overload
- Chronic, severe hemolytic Anaemia

- Long term effect of hypoxia
- Consequence of therapy

The most importent abnormalities of liver function include hypergammaglobulinemia, hypoalbuminemia and moderate decreases in the coagulation factors the are synthesized in liver as well as increased levels of transaminase. Serum bilirubin level is moderately elevated upto 3mg / dl. For liver function the use of alanine amino transferase (SGPT or ALT) is a rapid screening procedure and an inexpensive technique To study of liver function in multiply transfused thalassaemia patients

Materials and Method

Between January 2018 to January 2020, 50 already diagnosed thalassaemic patients admitted in thalassaemia ward in Mahatama Gandhi Medical College & Hospital for blood transfusion are selected in prospective study. The patients who had received more than 50 blood transfusions are considered as "multiply transfused"

Inclusion criteria

1. Patient recived at least 50 transfusion
2. Only Hb electrophoresis diagnosed case of thalcessemia

Exclusion Criteria:

1. Patients are reciving hepatotoxic drugs
2. Other chronic liver disease in thalassemia patients

Methods

Liver Function: Following laboratory parameters are taken into consideation:
SGPT > 150U/L
SGOT> 88U/L
(The serum transaminase levels are regarded best indicators of liver damage. If either transaminase level is higher than 2.5 times the upper limit of normal range, then it is considered as significant derangement.)
Serum Bilirubin > 3 mg / dL

1. Aminotransferas:

Principle:

$$\text{L-Alanine + 2-oxoglutarate} \xrightarrow{\text{ALAT}} \text{glutamate + Pyruvate}$$

$$\text{Pyruvate + NADH + H}^{+} \xrightarrow{\text{LDH}} \text{lactate + NAD}$$

Addition of Pyrodoxal- 5-phosphate (P-5-P) stabilizes the transminases and avoids falsely low values in samples containing insufficint endogenous P-5-P

Procedure:

Sample 100 ul
Reagent 1 1000ul
Mix, incubate for 5 min then add: Reagent2
Mix, read absorbance after1 min and start stopwatch

Read absorbance again 1,2 and 3 min. thereafter

Reference Range

Vomen <34 U/L
Men <45 U/L
Children 1-30 days < 25 U/L
2-12 month <35U/L
1-3 years <30 U/L
4-6 years <25U/L
7-9 years <25 U/L
10-18 years < 30 U/L

Results

In Present study 28 (56%) were male and 22 (44%) were female (Male: Female = 1.27:1
90 % patients were under age of 15 years age in the study.
In Present study the most common group was sindhis (26%) Closely followed by then Punjabis (22%) and Muslim (16%)

Table 1: Serum Bilirubin levels under study

S. Bilirubin	Total (n=50)
Increased	34 (68%)
Normal	16 (32%)
Total	50 (100%)

In present study serum bilirubin levels were increased in 68% of patients

Table 2: Serum transaminase levels under study

Serum Transaminase Levels	Total (n= 50)
Increased	26 (52%)
Normal	24 (48%)
Total	50 % (100%)

Serum transaminase levels were increased in 52 % of patients

Discussion

Thalassemia syndrome is one of the commonest inherited hematological disorder. In beta-thalassemia syndrom, there is decreased of synthesis of the beta- globin chain. It is characterised by low intracellular hemoglobin. These patients need of regular blood transfusions to maintain hemoglobin levels at 10-12 gm/ dl, they are also more prone to iron over load and blood borne infections.

In thalassemic patients liver is affected in non transfused patients due to extramedullary hematopoiessis in form of hetatomegaly, whereas in transfused patients, the liver in prime organ for deposition of excess iron and become fibrotic and eventually cirrhotic secondary in iron deposition. In thalassemic patients liver

dysfunciton may be due to either iron overload or transfusion induced viral infections. The most important abnormalities of liver functions are hyper-gammaglobulinemia, hypoalbumenia, moderate decrease in coagulation factors and increase in serum transminases levels. Serum bilirubin levels are moderately elevated up to 3 mg /dl.

Age and Sex distribuiton

Age: In the present study, 90% patients were under 15 years of age and remaining 10 % were above 15 years of age. In a similar study conducted by Prashant srivastava et al in 32 patients, 94% patients were under 15 yr of age.[4]

Sex: In the Present study male to female ration was1.27: 1%.

In the study by Khaled et al, in 80 thalassemic patients male to femail ratio was 1.16.:1%.[5]

Family History of thalassemia:

In present study 40% patients had family history of thalassemia. In the study of Khaled et al 43.75 % had positive family history.[5]

Liver functions:

Serum transaminases levels and serum bilirubin levels:

In the present study 68 % cases had increased serum bilirubin levels. In 52 % cases had increased serum alanine aspratate levels, but non of case had viral markers positive.

A similar study conducted by prashant et al. In 32 hyper transfused patients and found around 50 % of cases had increased serum transaminases levels & 78.12 % cases had increased serum bilnbin levels.[4]

A similar study by Saad A. Mustafa conducted in 100 patients of multiple transfused thalassemia, & out of 100 pt., 66% patients had elevated SGOT & SGPT .[6]

Conclusions and Recommendations

Present study was conducted in 50 multiple transfused thalassemic patients. In present study serum transaminase levels and serum Bilubin in leveles were taken as parameter of liver funciton

1. 90% patients were under 15year of age
2. In study male : Female was 1.27:1
3. The Thallesemia Syndrome was common in sindhis,punjabis, population.
4. 52% patients had increased serum transaminases levels.
5. Serum bilirubin levels were increased in 68% patients

The routinely patients should be screening for liver functions like serum transaminase levels,Serum bilirubin levels, prothrombin time, serum iron levels serum ferritin, MRI study of liver biopsies at regular interval.

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