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A study of Serological and Hematological Parameters in Thalassaemic Patients of Gujarat, Western India

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Absract: *Thalassemia describes a group of inherited disorders characterized by reduced amount of hemoglobin, the oxygen-carrying protein inside the red blood cells. Various haematological and serological parameters were studied in a cohort of thalasaemic patients coming for routine blood transfusion regime in a government run children hospital of Rajkot. Due to poor socio-economic condition, none of the patients were taking chelation therapy needed for better management of thalassaemia. To get insight into effect of multiple blood transfusions on these patients, age, spleen status, distribution of blood group, hemoglobin level and other hematological indices and serolological parameter like SGPT related to iron overburden was studied. A very low hemoglobin level with very high SGPT level along with sleenomegaly is suggestive of poor prognosis in these patients.*
Key Words: *Thalassemia, Hematology, Serology, Spleenomegaly*

I. Introduction

Normal hemoglobin A consists of two α and two β chains. The globin gene clusters are present on chromosome 16 while the β gene clusters are present on chromosome 11. Thalassemias are autosomal recessive disorders. In thalassemia, one of the globin chains syntheses may be defective either due to mutation or deletion resulting in excess production ofthe other chain which damages the red cell membrane. On the basis of the affected globin chain, the thalassemias are classified into α or β types. The β thalassemia is prevalent throughout the world while α is found more in the Mediterranean region, [1] Middle East, [2],[3],[4] South Asia [5],[6] and South East Asia. [7],[8],[9]

The hematological parameters in the thalassemic patients vary with the type and severity of anemia. However, varying degrees of microcytosis is invariably a common feature of almost all types of thalassemias. Thalassemias are classified according to the globin that is affected, hence the names *alpha* and *beta* thalassemia. Beta thalassemia is the most well-known type of thalassemia and is also called Cooley's anemia. Beta thalassemia major usually causes severe anemia that can occur within months after birth. If left untreated, severe anemia can result in insufficient growth and development, as well as other common physical complications that can lead to a dramatically decreased life-expectancy. To avoid that repeated blood transfusions are required. Individuals with beta thalassemia major receive regular blood transfusions, usually on a monthly basis. This helps prevent severe anemia and allows for normal growth and development. However, in poor socio economic set up this regime is not strictly adhered to and result in a grim scenario in terms of maintenance of satisfactory hematological and serological parameters for targeted aim of well being of such patients. Life sustaining transfusion regime also has the flip side in terms of spleenomegaly and increased SGPT levels due to iron overload. An attempt was made to study hematological and serological parameters in thalassemic patients on regular blood transfusion regime without chelation therapy. This along with other complications of blood transfusion can decrease life expectancy of such patients dramatically.

II. Materials and Methods:

A group of 130 thalassemic patients at K.T. Children Hospital, Rajkot, Gujrat were initially included in the study in the year of 2003. At the end of the study we had registered 218 patients. These patients had been receiving blood transfusions regularly at K.T. Children Hospital. Transfusion and clinical records of all patients were maintained. About 3 ml blood sample was collected and samples were preserved.

Analysis of 121 patients was done at the end of the study and the others were excluded due to various reasons (05 patients moved, sample was inadequate in 02, one patient died and one was lost to follow up).

-Screening of Hematological parameters**:** All the samples were screened by poCFH-100i Automated Hematology Analyzer by Sysmax, Transasia. It shows parameters like WBC, RBC, HGB, HCT, MCV, MCH, MCHC, PLT, LYM#, LYM%, MXD#, MXD%, NEUT#, NEUT%, RDW-SD, RDW-CV, PWD*, MPV, P-LCR.It shows histogram of WBC, RBC and PLT.

-Screening of blood groups (ABO)**:** Monoclonal agglutinating sera were used for the determination of human blood groups (ABO) by Span Diagnostic kit.
-Screening for S.G.P.T. detection specially in Thalassemic patients: All the samples were screened by S.G.P.T. IFCC method, Kinetic by ERBA S.G.P.T. Test kit.
-Screening of Iron & TIBC Level by Crest Biosystems: Test is based on Ferrozine Method for the determination of Iron and Total Iron Binding Capacity in serum.

III. Result and Discussion

Present study was conducted to observe haematological and serological parameters in thalassemia patients who received multiple blood transfusion without chelation therapy in and around Rajkot city. Figure 1 shows prevalence of blood group in Thalassemia patients to study the preference of any particular blood group.

Blood group wise distribution of Thalassemia patients
Figure

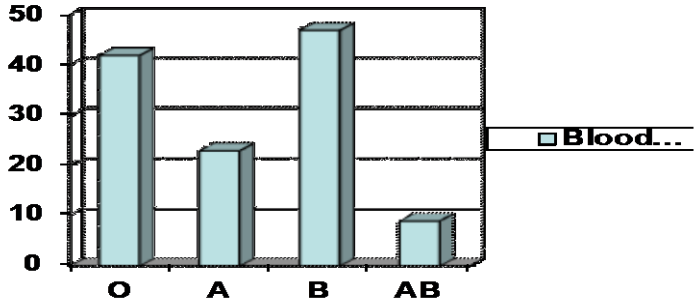


Figure shows in our studies 42 patients out 121 possess Blood group O (34.71%), 23 out of 121 are of blood group A (19%), 47 out of 121 are of blood group B (38.84%) and 9 out of 121 having blood group AB (7.45%). This pattern is similar to the pattern of blood group observed in general population.
Repeated blood transfusion results in Spleenomegaly in Thalassemic patients. However some patients undergo spleenactomy to reduce amount of blood transfusion.

Spleen status in Thalassemic patients
Figure 2

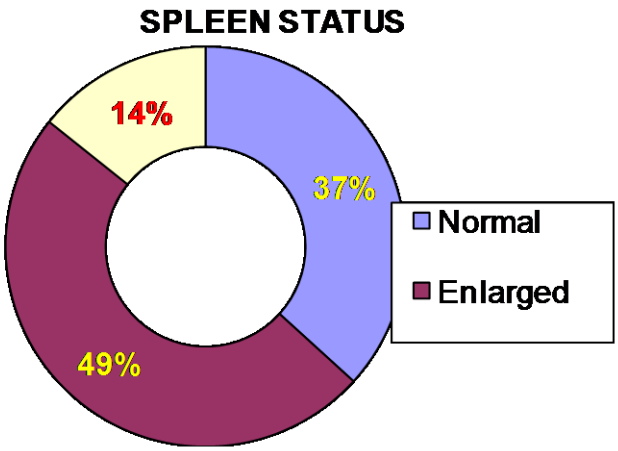
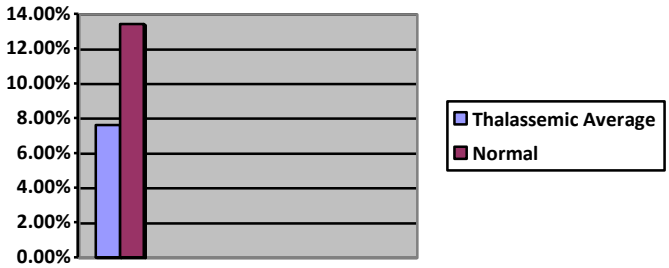


Figure 2 shows that 49% of the patients had spleenomegaly and 37% patients had normal size of the spleen. 14% of patients were spleenectomized which make them susceptible to microbial infection. However spleenectomy procedure is always followed by vaccination to the patients. The estimation of Hemoglobin always reveals the deviation of Hb level from the normal. Anything below 13.8 gm% in males and 12.0 gm% in females in considered as anemia.

Estimation of Hemoglobin in Thalassemia Patients
Figure 3



		Hemoglobin gm %
1	Thalassemic Average	7.6 gm%
2	Normal	13.2 gm%

Above table shows patients with Thalassemia have lower Hemoglobin (Average 7.6 gm.%) than Normal (Average 13.2 gm.%).
Above Figure shows patients with thalassemia shows lower Hemoglobin % in our study, Thalassemic patients have average 7.6% of Hemoglobin as compare to normal patients have 15.0 % of Hemoglobin.

Table 1
Serological Parameters

	Normal values	Thalassemic Patient's Average value
SGPT	6-21 IU/ml	55.1 IU/ml
TIBC	228-428 mcg/ml	506.5 mcg/dl
S. Iron	80-175 microgm/100ml	206.18 mcg/100ml

Above table shows Thalassemia patients show higher level of S.G.P.T. enzyme (55.1IU/ml) than Normal people (6-21 IU/ml), higher T.I.B.C. levels (506.49 mcg/dl) than Normal people (228-428 mcg/dl) and higher level of S. Iron. (206.18 µg/100 ml) than Normal people (Male: 80-175 µg/100 ml and Female: 60-160 µg/100 ml)

Heamatological Parameters
Table 2

	Normal Values	Thalassemia Patients Values
HCT	37-47%	21.6%
RDW-CV	11.5-14%	21.44%
RBC	4.5-6.5 X 10^6 cmm	2.82 X 10^6 cmm
WBC	4.5-11.00 X 10^3 cells/microliter	12.5 X 10^3 cells/microliter

Above table shows Thalassemic patients have lower levels of H.C.T. (21.6%) than Normal people (37-47%). They also have higher level of R.D.W.–C.V. (21.44%) than Normal people (11.5-14.0%). Also total count of leucocytes is always increased during thalassemia. Above table shows Thalassemic patients show high levels of W.B.C. (12.5 X 10^3 cells/cmm) than Normal people (4.5-11.00 X 10^3 cells/cmm. The number of white blood cells may appear raised due to the presence of a large number of immature (nucleated) red blood cells, which the cell counter may mistakenly identify as white blood cells. RBC count is also low. Microcytic anemia is observed. Above table shows Thalassemic patients have lower levels of Red Blood Cells (2.82 X 10^6 cmm) than Normal people (Male: 4.5-6.5 X 10^6 cmm and Female: 3.9-5.6 X 10^6 cmm)

Statistical Correlation analysis of various parameters:
Correlation analysis of various parameters in hemogram as well as incidence of microbial infections with the amount of blood transfused was carried out.

	Hb	S.G.P.T.	S.Iron	RBC	HCT	MCV	RDW-CV	WBC
Hb	1							
S.G.P.T.	0.034	1						
S.Iron	-0.228	0.001	1					
RBC	**0.769**	0.011	-0.183	1				
HCT	**0.774**	-0.019	-0.180	**0.959**	1			
MCV	0.134	-0.077	-0.033	0.118	**0.378**	1		
RDW-CV	-0.152	-0.226	0.083	-0.023	0.050	0.175	1	
WBC	0.094	0.137	-0.057	0.078	0.115	0.116	0.152	1

As shown in the table there is a significant correlation between hemoglobin value, RBC count and hematocrit value. There was also a significant correlation of moderate degree between hematocrit value and mean corpuscles value.

Thalassemia is a worldwide disorder. α- and β -thalassemia are the most common single-gene hemoglobin disorders in the world. It is more prevalent in areas endemic for malaria. [18] South East Asia, [7],[8],[9] India, [5],[6] Mediterranean region [2],[3],[4] and Middle East including Saudi Arabia [2],[3],[4] are the regions from where large number of cases are reported. The change in hematological parameters depends on the type of thalassemia. The clue for thalassemia is low mean corpuscular volume (MCV) < 78 fl or low mean corpuscular hemoglobin (MCH) <27 pg. Although iron deficiency is the most common cause of a low MCV or a low MCH, it is likely that this finding will point to thalassemia in regions of countries with thalassemia-prone ethnic populations. There are several causes of the anemia produced by different abnormal hemoglobins. Microcytic hypochromic anemia is a common hematological abnormality in clinical practice and usually is caused by iron deficiency and thalassemia trait.

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