

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

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Abstract- Thalassaemia 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 thalassaemic 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 serological parameter like SGPT related to iron overburden was studied. A very low hemoglobin level with very high SGPT level along with spleenomegaly is suggestive of poor prognosis in these patients.

Index Terms- Thalassaemia, 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. Thalassaemias are autosomal recessive disorders. In thalassaemia, one of the globin chains syntheses may be defective either due to mutation or deletion resulting in excess production of the other chain which damages the red cell membrane. On the basis of the affected globin chain, the thalassaemias are classified into α or β types. The β thalassaemia 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 thalassaemic patients vary with the type and severity of anemia. However, varying degrees of microcytosis is invariably a common feature of almost all types of thalassaemias. Thalassaemias are classified according to the globin that is affected, hence the names *alpha* and *beta* thalassaemia. Beta thalassaemia is the most well-known type of thalassaemia and is also called Cooley's anemia. Beta thalassaemia 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 thalassaemia 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 thalassaemic 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 thalassaemic patients at K.T. Children Hospital, Rajkot 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 since 2003 till the end of the study period. Patients who had received at least 2 previous blood transfusions were included for serological follow up for 5 successive years (2003, 2004, 2005, 2006 and 2007). 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 Thalassaemic 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

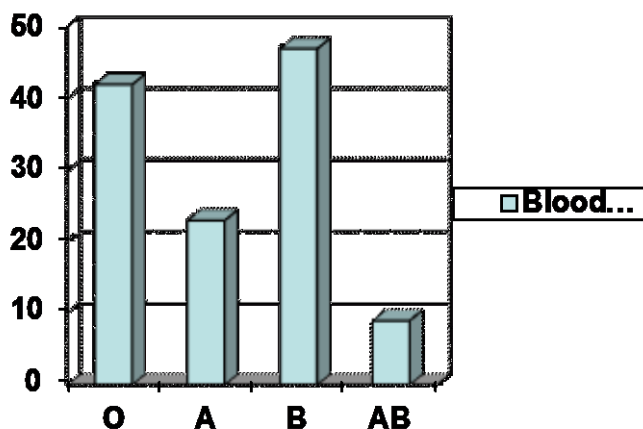


Figure shows in our studies 42 patients out 121 possess Blood group O (34.71%), 23 out of 121 are of blood group A (19%), 47 out of 121 are of blood group B (38.84%) and 9 out of 121 having blood group AB (7.45%). This pattern is similar to the pattern of blood group observed in general population.

Repeated blood transfusion results in Splenomegaly in Thalassemic patients. However some patients undergo splenectomy to reduce amount of blood transfusion.

Spleen status in Thalassemic patients
Figure 2

SPLEEN STATUS

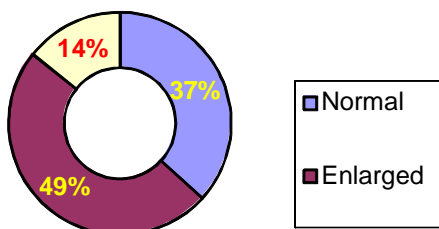
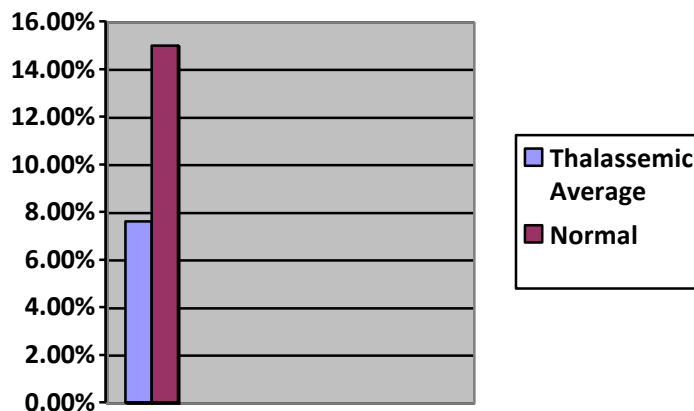


Figure 2 shows that 49% of the patients had splenomegaly and 37% patients had normal size of the spleen. 14% of patients were splenectomized which make them susceptible to microbial infection. However splenectomy procedure is always followed by vaccination to the patients. The estimation of Hemoglobin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

Estimation of Hemoglobin in Thalassemia Patients
Figure 3



		Hemoglobin %
1	Thalassemic Average	7.6%
2	Normal	15%

Above table shows patients with Thalassemia have lower Hemoglobin (Average 7.6%) than Normal (Average 15.0%).

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 microgm/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%).

Above table shows that thalassemia patients show 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 10³ cells/cmm) than Normal people (4.5-11.00 X 10³ 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 10⁶ cmm) than Normal people (Male: 4.5-6.5 X 10⁶ cmm and Female: 3.9-5.6 X 10⁶ 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],[14],[15],[16],[17] 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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