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Pathology

A STUDY ON HAEMOGLOBINO PATHIC DISORDERS WITH A SPECIAL REFERENCE TO THALASSAEMIA

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ABSTRACT Thalassaremia is one kind of haemeoglobino pathic disorder. In Thalassaemia, rise in the level of Haemoglobin F is observed in blood. The main reason behind this disease is the unyielding of Haemoglobon A. It is observed that a hetero-zygote situation is emerged due to the presence of Haemoglobin E leading to the restrain in Haemoglobin A and hence, the quantity of Haemoglobin E and F is increased.

It is believed that about 50-90% of total amount of Haemoglobin is found in Haemoglobin E. Furthermore, Haemoglobin C,D and S may be found as abnormal haemoglobins. Thalassaremia disease is the mixture of all these haemeoglobino pathic disorder. In this case, the production of Haemoglobin is affected due to some genetic mutations. The current paper highlights Thalassaremia as haemeoglobino pathic disorder.

KEYWORDS : Haemoglobin, Disorder, Thalassaremia

INTRODUCTION

Basically, Thalassaremia *is the disease which is caused due to some genetic problems. Generally, there are two forms of* Thalassaremia disorder i.e. Thalassaremia major and Thalassaremia minor. In former case, the problem of not producing haemoglobin inherits from both the parents whereas in latter case, the problem of haemoglobin inherits from one of the parent.

In the current study, we examined 63 patients suffering from Thalassaremia disorder. Out of these 63 patients, the tests related to the liver functioning were done in 36 patients. There were 43 males and 20 females participated in the research work. The age of the patients was between 4 and 34 years. Here, Haemolysis was investigated in Thalassaremia. The haemostatic status in this disease was found out in this work.

Here, Cyan-Methaemoglobin method was used to perform the process of estimating haemoglobin. Normally, the value of haemoglobin in males should be between 14 to 17.49 gram and there should be 11 to 14.65 gram haemoglobin in females.

In this test, Potassium Cyanide was mixed with the blood and cyan-methaemoglobin was obtained from methaemoglobin and potassium ferricyanide.

For the determination of packed cell volume, the centrifugation was performed on the oxalated blood foe about half an hour. This process was continued until a consent volume of corpuscles was attained. For this purpose, Wintrobe tubes having diameter of 3mm and height of about 110 mm were used.

For the purpose of counting Reticulocytes, dye was formed with the mixture of absolute alcohol and 1% solution of cresyl blue. Then, this dye was allowed to drop in little amount of blood and was kept on a slide. To spread the blood uniformly, a gentle pressure was applied over the cover of glass. Here, molten paraffin was used to seal the upper part of the slippery cover. A lens immersed in oil was used to examine the film.

Out of 63 patients, 39 were investigated to estimate the plasma haemoglobin. These patients were suffering from homozygous thalassaemia and E- thalassaemia. The Thromboplastin generation test was performed here. The TGT group of 20 cases out of 39 was observed to be normal whereas there were 19 cases whom TGT group was reported to be defective. A range between 3 to 78 mg% was observed for plasma haemoglobin.

DATA ANALYSIS AND INTERPRETATION

The following table shows the haemoglobin variants.

Table 1 The variants in Haemoglobin

	Male	Female	Total
E- Thalassaremia	38	18	56
Homogygous Thalassaremia	3	2	5
S- Thalassaremia	1	0	1
D- Thalassaremia	1	0	1
	43	20	63

Data Interpretation:

It is clear from table1 that out of 43 males, 38 were found suffering from E- Thalassaremia whereas out of 20 females, 18 were found suffering from the same disease.

Clinical Observation: The following table shows the clinical observations of all 63 patients who participated in the study.

It is clear from Table 2 that in the current study done on 63 patients, 45 patients were feeling weakness and 30 were suffering from fever.

Table 2 Clinical Observation

Symptoms	No. of cases
Weakness	45
Fever	30
Pallor	22
Jaundice	15
Swelling	14
Retarded growth	10

Data Interpretation:

On the other hand, the complaint of swelling was reported frequently and was observed in 14 patients and Pallor was found in 22 patients and was fluctuated timely according to the level of anaemia present in the patient body.

Similarly, the problem of Jaundice was noticed in 15 patients while retarded growth was reported in 10 patients.

Besides these symptoms, other symptoms like Diarrhoea, Menstrual irregularities and pain in legs etc. were observed in the participated patients.

Haemoglobin Level:

The following table shows the level of Haemoglobin among all 63 patients.

Table 3 Level of Haemoglobin

Haemoglobin in grams	No. of cases
3.18 – 5	3
5 – 7	35
7 – 9	20
above 9	5

Data Interpretation:

It is clear from Table 3 that out of 63 patients, the quantity of Haemoglobin in 35 patients was observed to be between 5 to 7 grams. On the other hand, there were 20 patients whom haemoglobin level was found to be between 7 to 9 grams.

There were only 5 patients having the level of Haemoglobin more than 9 grams whereas in 3 patients, the haemoglobin range was reported to be between 3.15 to 5 grams.

Packed Cell Volume Range:

The following table shows the range of packed cell volume among all

participated patients.

Table 4 Packed Cell Volume Range

PCV	No. of Patients
8 – 15	4
15 – 25	33
25 – 34	26

Data Interpretation:
It is clear from table 4 that the level of PCV was reported to be between 25 to 34 in 26 patients and there were 33 patients whom PCV was found to be between 15 and 25. On the other hand, the PCV was estimated to be within the range of 8 to 15 in 4 patients. The number of reticulocytes per 1000 red blood cells was counted and shown in table 5.

Reticulocytes count:
The following table shows the percentage of reticulocytes.

Table 5 Reticulocytes count

Reticulocytes count (in %)	No. of cases
Below 5%	6
5 – 15	45
Above 15%	12

Data Interpretation:
It is clear from table 5 that the percentage of reticulocytes was observed to be between 5 and 15 in 45 cases whereas there were 12 cases, where the reticulocytes were reported to be above 15%. On the other hand, in 6 cases, the reticulocytes count was found to be below 5%.

Estimation of Plasma Haemoglobin:
The following table shows the values of estimated plasma haemoglobin.

Table 6 Estimation of Plasma Haemoglobin

	No. of Cases	Plasma Haemoglobin	
		Range in mg%	Mean (mg%)
Normal TGT	20	3 – 40	7.73
Abnormal TGT	19	3.5 - 78	26.07

Data Interpretation:
It is clear from Table 6 that among all 39 patients, the range of plasma haemoglobin was estimated to be between 3 to 40 mg% in 20 patients having normal TGT whereas the range of plasma haemoglobin was found to be between 3.5 and 78 mg% in 19 cases with abnormal TGT.

Also, the mean of plasma haemoglobin was 7.73 mg% in 20 normal TGT patients and the mean of plasma haemoglobin in 19 abnormal TGT cases was found to be 26.07 mg%.

DISCUSSION

It is observed that in thalassaemia disorder, the level of circulating haemoglobin in blood tends to rise. There are many corpuscular factors which may lead to anaemia in case of thalassaemia disorder. The methods related to auto-immune system are used in order to mediate these factors.

There was lower splenectomy and hence, there was no compatible perception of red blood cells in thalassaemia disorder. Therefore, the level of haemolysis in intra-vascular blood cells was enhanced due to plasma haemoglobin.

Also, the thalassaemic red cells are easy to break in nature so they are decomposed into many components on applying external pressure due to friction.

An abnormal coagulation was observed in the patients suffering from thalassaemia disorder having variable haemolysis. This study helps in finding any co-relation between circulating haemoglobin level and the haemo-static abnormalities.

The reason behind the rising level of circulating haemoglobin in plasma is the presence of haemolytic components in haemoglobino pathic disorders like thalassaemia syndrome. It is also observed that the level of damage in red cells of intra-vascular type tends to rise with the reflection of plasma haemoglobin.

In some cases, phospholipids are produced from the platelets and injured red blood cells. The highest level of plasma haemoglobin i.e. 78 mg% was found in patient suffering from E-thalassaemia syndrome and serum was defected during TGT test.

Also, it was observed that the level of plasma haemoglobin was higher in case of defective Thromboplastin generation test as compared to the normal generation test.

It ha3 been shown that the haemoglobin released intravascularly cannot exist in free form. It is bound up primarily by a protein called hepsftoglobln which has been shown to separate with alphag globulin. In instances when excessive haemolysis takes place and the heptoglobin is saturated, any further release of haemoglobin could be bound up by beta globulin, gamma globulin and albumin.

CONCLUSION

Plasma haemoglobin level in circulation, which in turn reflects the degree of haemolysis occurring in haemoglobinopathic disorders, appeared to have a co-relation with the prevailing haemostatic defect. This haemostatic abnormality is possibly a part of the genetically transmitted haemoglobinopathic disease expressing itself in fluctuating degrees in a significant, proportion of patients suffering from this malady.

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