# SUMMARY AND CONCLUSIONS

Our study included (42) B thalassaemia major patients which were divided into two groups splenectomized (14) and non splenectomized (28) patients, age ranged (1-17 years). And also (14) age matched normal healthy children, age ranged (2-13 years), were included as control group.

The present study is a modest attempt to evaluate the possible changes which may occur in the thalassaemic red cell membrane lipids (phospholipids, triglycerides and cholesterol), ATP, ATPase and protein kinase. Moreover, plasma and intra-corpascular $Ca^{++}$ that might account for their rapid lysis, and might have a clinical and scientific value.

The results obtained in this study revealed significant decrease in red cell membrane phospholipids and triglycerides and a non significant decrease in red cell membrane cholesterol in both B-thalassaemia major (splenectomized and non splenectomized) as compared to normal control one.

However there were a significant increase and a significant decrease in red cell membrane phospholipids and

triglycerides respectively in B thalassaemic non splenectomized patients as compared to splenectomized one.

Our finding concerning the significant decrease of red cell membrane phospholipids and triglycerides, and the non significant decrease of red cell membrane cholesterol in B thalassaemic patients could be explained by :

* The possible decrease of erythrocyte membrane ATP formation, which could affect fatty acid acylation pathway to renew native phospho-lipids.
* Decreased activity of the acylase enzyme which is needed to prevent accumulation of deleterious lysophospholipids within the membrane.
* Abnormalities in plasma lipids and lipoprotein metabolism which were associated with the possible changes in composition and fluidity of erythrocyte membrane.
* Accelerated erythropoiesis and histiocytes of the reticulo endothelial system may be one of the factors which may lead to low plasma cholesterol levels.
* And, or, the possibility of decreased synthesis and activity of acylase enzyme due to disturbance in nucleotides precursors and ATP production caused by the disturbed metabolism in the thalassaemic red cell

membrane, together with the formation of abnormal active sites of acylase enzyme on the cell membrane due to the abnormal precipitation and disorganization of $\alpha$ globin chain.

All these factors, together with the possible decrease of acylase activity on the cell membrane lysophospholipids leading to decrease phospho-lipids and concomitant increase in lysophospholipids with its powerful haemolytic activity.

Also in this work, we found a significant decrease in red cell membrane ATP, ATPase and protein kinase in the thalassaemic patients as compared to normal control one, a findings could be explained by :

* Impaired 5 phosphoribosy -1- pyrophosphate (PRPP) synthesis which is a precursor for adenine nucleotides.
* Decreased ATP synthesis due to defective metabolism in the glycolytic pathway.
* Defective in $Ca^{++}$, haemostasis and its absorption may lead to decrease activity of ATPase and protein kinase.
* And or, the decrease of red cell membrane ATP synthesis may lead to the decrease of ATPase and protein kinase which is needed for their synthesis.

As regards plasma and intracorpascular $Ca^{++}$ a non significant changes in the thalassaemic patients were found as compared to normal control one.

Finally, from this study, we could conclude that, a significant decrease in red cell membrane phospholipids, triglycerides, ATP, ATPase and protein kinase in thalassaemic patients.

Together with a non significant changes was found in red cell membrane cholesterol, plasma and intracorpascular $Ca^{++}$ in the thalassaemic patients.

All, these finding might be the cause of decreased flexibility and deformability of thalassaemic red cells leading to decrease in red cell survival and its rapid lysis.

We recommend future studies on plasma and red cell membrane in thalassaemic patients especially the following parameters.

* Lipids and phospholipids fractionation especially fatty acids.
* Proteins and lipoproteins fractionation.

* Evaluation of the acylase enzyme activity.

* Study of $Ca^{++}$ haemostasis.