SUMMARY

Thalassemia is among the most common genetic disorders worldwide, particularly in the Mediterranean region, Africa, the Middle East and Southeast Asia. Thalassemia is a hereditary anemia resulting from defects in haemoglobin production. β- Thalassemia, which is caused by a decrease in the production of β globin chains, affects multiple organs and is associated with considerable morbidity and mortality. Accordingly, lifelong care is required and financial expenditures for proper treatment are substantial. The recommended treatment for thalassemia major involves regular blood transfusions, usually administered every 2 to 5 weeks to maintain a pre-transfusion haemoglobin level between 9-10.5 g/dl.

Thalassemic children experience various problems if the transfusion is inadequate but at the same time repeated blood transfusions are associated with hazards like iron overload and risk of acquiring transfusion-transmitted infections (TTIs). Iron overload can lead to endocrinal dysfunction in the form of growth retardation and diabetes mellitus. Transfusion-transmitted infections such as HIV (with risk of progression to AIDS), HBsAg, and HCV (with high risk of developing chronic hepatitis, liver cirrhosis and hepatocellular carcinoma) can also occur. Thus, chronic blood transfusion in thalassemic patients is a double-edged sword. Ultimately thalassemic patients die either due to transfusion complications or due to lack of it, with the result that they seldom survive beyond the age of 25 years.

Packed RBCs were usually used for thalassemic transfusion until recently when little attention had been paid to the contaminating leukocytes present in various blood components and thought to be responsible for many immunological complications as HLA alloimmunization which is considered the main cause of FNHTRs and platelet refractoriness observed in multi-transfused patients, but nowadays those adverse reactions reduced dramatically after the usage of the leucoreduced blood products.

The aim of this study was to detect the rate of HLA alloimmunization in pediatric chronically transfused thalassemic patients in the hematology clinic of Alexandria University Children's Hospital who were transfused by non leucoreduced PRBCs, to determine the relation between HLA alloimmunization and the occurrence of FNHTRs and to study the effect of splenectomy on these adverse reactions and their relation to HLA alloimmunization.

We also compared between transfusing washed and filtered RBCs regarding their ability to prevent the occurrence of FNHTRs taking into consideration the Hb rise after transfusion by both products.

This study was conducted on sixty five thalassemic patients, selected from the hematology clinic of Alexandria University Children's Hospital and twenty healthy volunteers who served as a control group. The patients were further subdivided into forty five non splenectomised chronically transfused β thalassemia major children and twenty splenectomised ones. All patients were maintained on regular blood transfusion program according to their needs and most of them were suffering from FNHTRs.

All patients (n=65) included in the present study were subjected to detailed history of blood transfusions regarding the age of starting transfusion, frequency of transfusions and frequency of febrile reactions then 2 ml of venous blood were drawn from every patient for qualitative estimation of human leucocytic antibodies by anti- lymphocytotoxic antibody (ALA/LCA) ELISA kit.

All thalassemic patients with positive HLA antibody were subjected to 2 sessions of blood transfusion; one by bed-side leucofiltration and the other by washed RBCs, then observing the occurrence of FNHTRs and estimating the hemoglobin rise after transfusion by both products.

Results showed low rate of HLA alloimmunization among studied thalassemic patients (13%), and there was no clear association between HLA alloimmunization and the occurrence of FNHTRs (p=0.887). No relation between frequency of transfusions and the rate of HLA alloimmunization (p=0.362) was noticed.

Regarding splenectomy, we noticed that splenectomised thalassemic patients had fewer need for blood transfusion than non splenectomised patients (p=0.036), there was also a significant relation between splenectomy and the absence of HLA alloimmunization (p=0.048), but there was no relation between splenectomy and the Frequency of FNHTRs (p=0.463).

The comparison between the washed RBCs and filtered RBCs showed no difference regarding their efficiency in prevention of the FNHTRs (p=1.000), there was also no difference in the Hb rise after transfusion by both blood products (p=0.409).