Hemorheological profiles in Thalassemia: a new approach for the study of complications

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Several studies demonstrated the presence of high incidence of thromboembolic events in β-thalassemia, more common in thalassemia intermedia than in thalassemia major. In these patients a chronic hypercoagulable state is evident partially due to the impairment of the natural endothelial anticoagulant system, where the thrombomodulin is increased and the coagulation inhibitors are diminished. Red blood cells of thalassemic patients exhibit impaired flow properties that facilitate microcirculatory disorders: enhanced aggregability, reduced deformability, as well as a marked elevation of adherence to endothelial cells. Levels of pro-coagulant microparticles derived from endothelium, platelets, RBC and leukocytes are also elevated in thalassemia intermedia patients. The increase in adhesion proteins and vascular cell adhesion molecules suggest that endothelial injury or activation may be a feature of β-thalassemia, and may play an important role in the recruitment of leukocytes and erythrocytes and promote thrombosis at vascular inflammation sites. The hemorheological profiles of patients affected by β-thalassemia major and intermedia have been evaluated and significant differences have been observed between the groups, mainly in relationship with transfusion therapy. These results could give reason of the high incidence of thromboembolic events in b-thalassemia, more common in thalassemia intermedia than in regularly transfused thalassemia major.