suggest that the regulation of both genes may be coordinated through a bi-directional mechanism. However the STOPB gene is not transcriptionally induced by spermatids but is upregulated with spermatogonia and with the PML gene. Its homologs to the EP170 gene, which has been implicated in spermatid differentiation, as well as its overexpression in ES352 cell lines raise the questions of its possible role in red cell disorders or function.

**Abstract 2996**

**HEREDITARY SPHEROCYTOSIS DEFICIENT IN PROTEIN 4.2, ASSOCIATED TO MULTIPLE MYELOMA, ONE CASE REPORT.** E. Greco, M.L. Robme, A. Novoa, J. Cordoves, M.M. Ribera, Hospital S. Joao, Centro de Catequiza Experimental, Porto, Centro Hospitalar de Coimbra, Coimbra, Portugal.

Hereditary spherocytosis (HS) is an inherited hereditary anemia characterized by anemia, jaundice, splenomegaly and splenectomy. This may be due to mutations in the genes encoding ankyrin (ANK), the sodium exchanger, spectrin, protein 4.2 and protein 4.2. Hemolytic crises are frequent, are secondary to the reticulocytoclastic hyperplasia that accompanies protein 4.1 deficiency and other oxidative stress. HS patients have a mild, polyclonal hypergammaglobulinemia. The authors report a 60 year old Caucasian male referred to hematology out-patients clinic of hospital de S. Joao in May 1994, because of long standing history of anemia, severely aggravated by pregnancies and infections. One month before he had an episode of fever and headache. Physical examination revealed anemia, splenomegaly and a slight polychromatophilia. The hematological and biochemical parameters were compatible with sphero-eritic hemolytic anemia. The diagnosis of HS was confirmed by an increased osmotic fragility test and the finding of 12% decrease in protein 4.2 in the red cell membrane protein electrophoresis by Laemmli and Peutzmann. HS patients may have patients with protein 4.1 deficiency and splenomegaly.

**Abstract 2997**


Hereditary spherocytosis is an oxidative stress on account of its limited biosynthetic capacity, which predicts the repair or replacement of damaged proteins. Therefore, any normal RBC during its life span undergoes physical and chemical changes, which become more pronounced with age. The RBC membrane undergoes modifications, namely a reduction in the reticulocytoclastic hyperplasia that accompanies protein 4.1 deficiency and the increase of ankyrin. The pathway for removal of abnormal or damaged RBC involves the development of a leukocyte cell antigen (LCA) related to band 3 protein, which marks the RBC for death, by triggering the binding of specific auto-antibodies. The antibody accumulation activites. In the case of spherocytosis, presenting a disordered membrane structure, with loss of membrane components and with an abnormal reduction in life span, the modifications in band 3 profile are probably enhanced. In the present study, we attempted to study the oxidative stress from a biomolecular profile of band 3. A total of 12 RBC samples were studied, 6 normal RBCs and 6 HS RBCs. We studied the determination of a new biomarker concentration of BCR and GSH in the RBCs and in the HS RBCs. We studied the biomarkers' concentrations of BCR and GSH in the RBCs and in the HS RBCs. The results showed a significant increased oxidative stress in spherocytosis patients who did not undergo spleenectomy. The results showed a significant increased oxidative stress in spherocytosis patients who did not undergo spleenectomy. The changes in band 3 profile, hMBBM and in GSH activity are enhanced in spherocytosis patients who did not undergo spleenectomy when compared to the control, showing a higher oxidative and proteolytic membrane damage and mitochondrial degradation. These results provide evidence of premature senescence and removal of RBC in these patients. The removal of the spleen improved the oxidative stress in the heterozygous patients. However, the changes in band 3 profile, hMBBM and in GSH activity are even more pronounced, so it is easier to observe the effect of the organ removal. In the HS RBCs in these patients can circulate for longer periods of time although presenting more oxidative and proteolytic lesions. One data suggests that spherocytosis from patients who underwent splenectomy are more sensitive to an oxidative and proteolytic stress and, therefore, the development of chronic oxidative stress may trigger an hemolytic event. Currently these patients often present an hemolytic event during infections or inflammatory processes.