P-045 - OXIDATIVE STRESS IN CUBAN PATIENTS WITH MUCOPOLYSACCHARIDOSIS TYPES I, II, III AND IV WITHOUT ENZYME REPLACEMENT THERAPY.

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INTRODUCTION: Mucopolysaccharidoses (MPSs) are caused by deficiency of lisosomal enzyme activities needed to degrade glycosaminoglycans. These syndromes share many clinical features although to variable degrees. MPSs have a chronic and progressive disease course and involve multiple organs systems. Some of them have enzyme replacement therapy but Cuban patient have not this possibility. The unbranched polysaccharides accumulation in fluid and lysosome can explain some of the metabolic and clinical phenotypes. Besides inflammation, mitochondrial dysfunction, affections in autophagy and endoplasmic reticulum stress have been demonstrated in those disorders. Many researches reports oxidative stress presence in MPS patient. This condition has not been studied in Cuban patients diagnosed with different types of MPSs yet. Purpose: Considering these facts the aim of this study was evaluated oxidative stress parameters in Cuban Patients. METHODOLOGY: A case and controls study was carried that included patients with MPS I, II, III and IV and matched controls in age and sex. Plasma levels of malonyldialdehyde (MDA), advanced products of protein oxidation (APOP), free thiol groups, uric acid, creatinine, iron, ferritin, trasferrin, ceruloplasmin and calcium were measured. Also, the intraerythrocytic activities of superoxide dismutase (SOD1), catalese (CAT) and glutathione peroxidase were quantified. All the techniques used were spectrophotometric. RESULTS: Patients showed an increase in MDA, APOP and calcium levels compared to controls and the Cuban reference values. There was a decrease in the SDO1 activity and thiol groups concentrations in patients. No differences were found for the rest of the parameters measured. CONCLUSIONS: The increase of the oxidative damage and the decrease in the antioxidant capacity suggest that the oxidative stress process is presence in those Cuban patients. These results suggest a beneficial effect if we use an antioxidative therapy while acceding an enzyme replacement therapy is beyond our budget.