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SPECIAL SUPPLEMENT WITH THE ABSTRACTS

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increasing interest has arisen in the neonatal screening of X-ALD, which is possible through the analysis of C26:0-lysophosphatidylcholine (C26:0-LPC). Although a considerable number of studies has demonstrated the importance of this new biomarker for the diagnosis of X-ALD, its role in the pathophysiology of this disease has not been investigated.

OBJECTIVES: Considering that oxidative stress is a well described mechanism of damage in X-ALD, our objective was to investigate if this mechanism could be related with C26:0-LPC accumulation in X-ALD patients.

MATERIALS AND METHODS: We measured blood C26:0-LPC concentrations in five patients with X-ALD (2 children, one with CCER and the other with AMN; and 3 adult heterozygous women) by liquid chromatography tandem mass spectrometry. Oxidative stress was investigated in these patients through the measurement of the reactive species formation by the 2',7'-dichlorofluorescin oxidation assay (DCF) in plasma and by determination of plasma sulphydryl groups, whose reduction reflects protein oxidation.

RESULTS: Our results showed a significant increase of C26:0-LPC in blood of X-ALD patients when compared with healthy controls of similar ages, being higher in the male X-ALD patients in relation to the X-ALD female carriers. We also verified a strong inverse correlation between plasma sulphydryl groups and C26:0-LPC (r=−0.817, p=0.091) and a positive correlation between C26:0-LPC and DCF (r=0.611, p=0.274).

CONCLUSIONS: The correlations verified in this study between oxidative stress parameters and C26:0-LPC probably could be significant whether the number of analyzed patients was higher, which would make it possible to separate the patients according their phenotypes. Even so, preliminary data from this study suggest that C26:0-LPC may be involved in the induction of oxidative imbalance in X-ALD, deserving further investigation.

P-146 - N-ACETYL-L-CYSTEINE, TROLOX, AND ROSUVAstatIN PROTECT GLIAL CELLS EXPOSED TO HEXACOSANOIC ACID AGAINST INFLAMMATION, LIPID PEROXIDATION AND NITRATIVE STRESS

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INTRODUCTION: X-linked adrenoleukodystrophy (X-ALD) is a peroxisomal disorder caused by disfunction of the ABCD1 gene, which encodes a peroxisomal protein responsible for the transport of the very long-chain fatty acids from the cytosol into the peroxisome, to undergo β-oxidation. The major accumulated saturated fatty acids are hexacosanoic acid (C26:0) and tetracosanoic acid (C24:0) in tissues and body fluids. Recent evidence shows that oxidative and nitrative stress seems to be related with pathophysiology of X-ALD and many studies are associating antioxidants as an adjuvant therapy, since there is no completely satisfactory treatment for this neurogenetic disorder.

OBJECTIVES: Considering that glial cells are widely used in studies of protective mechanisms against neuronal oxidative stress, we investigated whether C26:0, incorporated in a lecithin vesicle, was capable to induce oxidative/nitrative damages and inflammation to glial cells and if the compounds N-acetyl-l-cysteine (NAC), trolox (TRO), and rosuvastatin (RSV) were able to protect cells against C26:0-induced damages.

MATERIALS AND METHODS: C26:0 was incorporated in lecithin vesicle by sonication. Glial cells were cultured in DMEM and at confluence, the vesicles containing lecithin and C26:0 were added. A pre-treatment was performed for 2h at 37°C with NAC (100 μM), RSV (5 μM), and TRO (75 μM). Supernatants were collected for analysis. IL-1β was measured by an Invitrogen ELISA kit, NO equivalents and isoprostanes was detected by a Cayman kit.

RESULTS: It was observed that glial cells exposed to C26:0 presented increased NO levels, high IL-1β levels, and increased isoprostane levels, compared to native glial cells without C26:0 exposures. Furthermore, NAC, TRO, and RSV were capable to mitigate these damages caused by the C26:0 in glial cells.

DISCUSSION AND CONCLUSION: Our data demonstrate, for the first time in literature, that C26:0, by itself, induced in glial cells culture: lipid peroxidation, nitrative stress and inflammation. Furthermore, we verified that NAC, TRO, and RSV were capable to attenuate damages caused by C26:0 in glial cells. The ability of these compounds to exert protective effects in glial cell culture might be of relevance as an adjuvant treatment for X-ALD, since there is still no completely satisfactory therapy for this disorder.

P-147 - SITOSTEROLEMIA IN COSTA RICA: REPORT OF THE FIRST CASE

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INTRODUCTION: Sitosterolemia is a rare autosomal recessive disorder of lipid metabolism characterized by increased intestinal absorption and a decrease in the biliary