Alteration of gene expression profile in niemann-pick type C mice correlates with tissue damage and oxidative stress

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Background: Niemann-Pick type C disease (NPC) is a neurovisceral lipid storage disorder mainly characterized by unesterified cholesterol accumulation in lysosomal/late endosomal compartments, although there is also an important storage for several other kind of lipids. The main tissues affected by the disease are the liver and the cerebellum. Oxidative stress has been described in various NPC cells and tissues, such as liver and cerebellum. Although considerable alterations occur in the liver, the pathological mechanisms involved in hepatocyte damage and death have not been clearly defined. Here, we assessed hepatic tissue integrity, biochemical and oxidative stress parameters of wild-type control (Npc1 +/+; WT) and homozygous-mutant (Npc1 -/-; NPC) mice. In addition, the mRNA abundance of genes encoding proteins associated with oxidative stress, copper metabolism, fibrosis, inflammation and cholesterol metabolism were analyzed in livers and cerebella of WT and NPC mice. Methodology/Pr