Abstract
Congenital Muscular Dystrophy type 1D (CMD1D) is characterized by an abnormal glycosylation of α-DG (α-dystroglycan) and associate to central nervous system (CNS) abnormalities such cognitive impairment. The purpose of the research were evaluate the blood-brain barrier permeability (BBB) permeability and matrix metalloproteinases (MMP) -2 and -9 in adult Largemyd-/- mice in order to understand the physiopathology of brain involvement during the CMD1D process. To this aim, we used adult homozygous Largemyd-/- (mutation in Large), heterozygous Largemyd+/- as well as wild-type (C57BL/6) mice. The animals were submitted to evaluation of BBB permeability and MMP-2 and MMP-9 in striatum, hippocampus and cerebral cortex. There were an increase of BBB permeability in hippocampus and striatum associated with increase of protein levels of MMP-2 in cerebral cortex and striatum and MMP-9 in hippocampus in adult Largemyd-/- mice. Our results suggest that the pathophysiologic processes can be associated to the action of MMPs and BBB disruption and that the BBB breakdown is relevant to the perpetuation of brain inflammation and can be related to brain dysfunction observed in CMD1D patients.

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