## C/S-4-DECENOIC ACID ALTERS RESPIRATORY PARAMETERS IN RAT BRAIN

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Medium-chain acyl-CoA dehydrogenase (MCAD) deficiency is the most frequent disorder of fatty acid oxidation. Biochemically, MCAD-deficient patients present tissue accumulation of the medium-chain fatty acids octanoic, decanoic and *cis*-4-decenoic (cDA) acids. Clinical presentation of MCAD deficiency is related to fasting and increased metabolic stress, which precipitate acute symptoms such as drowsiness or lethargy that may develop into coma or even death. In the present work, we investigated the in vitro effect of cDA on respiratory parameters in mitochondrial preparations from rat brain, namely states III and IV, as well as the respiratory control ratio (RCR), using glutamate/malate and succinate as substrates. It was observed that cDA diminished state II and RCR, while increased state IV in the presence of glutamate/malate and succinate as substrate. Taken together, these data strongly suggest that cDA may lead to impairment of ATP production acting as an uncoupler of oxidative phosphorilation, which could explain, at least in part, the characteristic brain dysfunction observed in MCAD deficient patients

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