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Effect of Coenzyme Q10 supplementation on mitochondrial electron transport chain activity and mitochondrial oxidative stress in Coenzyme Q10 deficient human neuronal cells.

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Abstract

Primary Coenzyme Q10 (CoQ10) deficiency is an autosomal recessive disorder with a heterogeneous clinical presentation. Common presenting features include both muscle and neurological dysfunction. Muscle abnormalities can improve, both clinically and biochemically following CoQ10 supplementation, however neurological symptoms are only partially ameliorated. At present, the reasons for the refractory nature of the neurological dysfunction remain unknown. In order to investigate this at the biochemical level we evaluated the effect of CoQ10 treatment upon a previously established neuronal cell model of CoQ10 deficiency. This model was established by treatment of human SH-SY5Y neuronal cells with 1 mM para-aminobenzoic acid (PABA) which induced a 54% decrease in cellular CoQ10 status. CoQ10 treatment (2.5 μM) for 5 days significantly ($p<0.0005$) decreased the level of mitochondrial superoxide in the CoQ10 deficient neurons. In addition, CoQ10 treatment (5 μM) restored mitochondrial membrane potential to 90% of the control level. However, CoQ10 treatment (10 μM) was only partially effective at restoring mitochondrial electron transport chain (ETC) enzyme activities. ETC complexes II/III activity was significantly ($p<0.05$) increased to 82.5% of control levels. ETC complexes I and IV activities were restored to 71.1% and 77.7%, respectively of control levels. In conclusion, the results of this study have indicated that although mitochondrial oxidative stress can be attenuated in CoQ10 deficient neurons following CoQ10 supplementation, ETC enzyme activities appear partially refractory to treatment. Accordingly, treatment with >10 μM CoQ10 may be required to restore ETC enzyme activities to control level. Accordingly, these results have important implication for the treatment of the neurological presentations of CoQ10 deficiency and indicate that high doses of CoQ10 may be required to elicit therapeutic efficacy.

KEYWORDS: Coenzyme Q(10); Mitochondrial electron transport chain; Mitochondrial membrane potential; Neuronal; Reactive oxygen species

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