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Coenzyme Q10 as a therapy for mitochondrial disease.

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Abstract

Treatment of mitochondrial respiratory chain (MRC) disorders is extremely difficult, however, coenzyme Q10 (CoQ10) and its synthetic analogues are the only agents which have shown some therapeutic benefit to patients. CoQ10 serves as an electron carrier in the MRC as well as functioning as a potent lipid soluble antioxidant. CoQ10 supplementation is fundamental to the treatment of patients with primary defects in the CoQ10 biosynthetic pathway. The efficacy of CoQ10 and its analogues in the treatment of patients with MRC disorders not associated with a CoQ10 deficiency indicates their ability to restore electron flow in the MRC and/or increase mitochondrial antioxidant capacity may also be important contributory factors to their therapeutic potential.

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KEYWORDS: Coenzyme Q10; EPI-743; Idebenone; Mitochondrial respiratory chain; Oxidative stress

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