Special Issue

Coenzyme Q10 in Mitochondria and Lysosomal Disorders

Message from the Guest Editor

Coenzyme Q10 (CoQ10) is a lipophilic molecule that serves as an electron carrier within the mitochondrial respiratory chain (MRC) as well as acting as a potent lipid soluble antioxidant. A deficit in CoQ10 status has been associated with a number of MRC disorders, although the cause of this deficiency has yet to be established in the majority of cases. The potential underlying CoQ10 deficiency may explain the therapeutic benefit reported for CoQ10 supplementation in the treatment of a subset of patients with MRC disorders. In recent years there has been a growing interest in the role that CoQ10 plays in lysosomal function, as, together with mitochondria, lysosomes are also a major site of CoQ10 localisation within the cell. Interestingly, reports are now emerging of a CoQ10 deficiency in association with certain lysosomal disorders, including mucopolysaccharidosis type III (MPS III), although the cause of the CoQ10 deficiency has yet to be elucidated.

Guest Editor

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