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Researchers seek tools to modulate the synthesis of CoQ10 in human cells

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Marta Luna, a researcher at the lab of the University of Granada Centre for Biomedical Research.

A study which includes the participation of University of Granada scientists has provided new data on the Q10 coenzyme (CoQ10), a molecule which is synthesized within the cells of the organism itself and which has essential functions for cellular metabolism. This study opens

the door for the development, in the not too distant future, of tools to modulate the synthesis of CoQ10 in human cells according to metabolic needs. This is particularly important for the treatment of diseases caused by primary and secondary deficiencies in CoQ10.

Its role in the production of energy required by the cell and its antioxidant capacity are among the best known functions of this coenzyme. Human cases have been described in which the deficiency in CoQ10 can be attributed to defects in the biosynthetic pathway, which causes a syndrome with a very heterogeneous clinical picture.

CoQ10 deficiency is a rare mitochondrial disease which affects mostly children. The details of this biosynthetic pathway are not known in their totality, since there are steps whose catalysing enzymes remain unknown, or proteins in the pathway whose specific function is either unknown or has not yet been fully demonstrated.

One of those proteins is Coq9, which the UGR research group demonstrated in 2013 is an essential protein in the biosynthesis of CoQ, and which specifically regulates the Coq7 protein, an enzyme with a hydroxylase activity that catalyses one of the intermediate steps for the synthesis of CoQ10.

This study is now led by Dr. David J. Pagliarini (U. of Wisconsin-Madison) in collaboration with Dr Liang Tong's team (Columbia U.) and the U. of Granada researchers Marta Luna Sánchez and Luis Carlos López García. It has been recently published in the journal *PNAS*. This research conclusively proves that protein CoQ9 regulates enzyme CoQ7.

Through the crystallization of the human protein and experiments conducted on mice, the study proves that Coq9 has a lipid-binding structure, which would give it the capacity to provide enzyme Coq7 with the intermediary metabolite that it uses as a substrate in the reaction it

catalyses. The results of the study suggest, besides, that the biosynthetic machinery of CoQ10 is organized as a multiprotein complex in mammals, with the purpose of increasing the efficiency of its synthesis and enable its regulation.

More information: "Mitochondrial COQ9 is a lipid-binding protein that associates with COQ7 to enable coenzyme Q biosynthesis." *Proc Natl Acad Sci* U S A. 2014, 111(44):E4697-705. DOI: 10.1073/pnas.1413128111. PMID: 25339443

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