Key protein in cellular respiration discovered

April 8 2009

Many diseases derive from problems with cellular respiration, the process through which cells extract energy from nutrients. Researchers at the Swedish medical university Karolinska Institutet have now discovered a new function for a protein in the mitochondrion - popularly called the cell's power station - that plays a key part in cell respiration.

Every time we take a breath, our blood transports oxygen to the mitochondria, where it is used to convert the nutrients in our food to a form of energy that the body can use. Problems with this process, which is called cellular respiration, have been linked to a number of morbid conditions, from unusual genetic diseases to diabetes, cancer and Parkinson's, as well as to the normal ageing process. Despite the fact that cellular respiration is so basic, there is much scientists have yet to understand about how it is regulated.

Cellular respiration depends on proteins synthesised outside the mitochondrion and imported into it, and on proteins synthesised inside the mitochondrion from its own DNA. Researchers at Karolinska Institutet have now shown that a specific gene (Tfb1m) in the cell's nucleus codes for a protein (TFB1M) that is essential to mitochondrial protein synthesis. If TFB1M is missing, mitochondria are unable to produce any proteins at all and cellular respiration cannot take place.

"Mice completely lacking in TFB1M die early in the foetal stage as they are unable to develop cellular respiration," says Medodi Metodiev, one of the researchers involved in the study, which is presented in Cell Metabolism. "Mice without TFB1M in the heart suffer from progressive

heart failure and increase mitochondrial mass, which is similar to what we find in patients with mitochondrial diseases."

The scientists believe that the study represents a breakthrough in the understanding of how mitochondrial protein synthesis is regulated, and thus increases the chances of one day finding a treatment for mitochondrial disease, something which is currently unavailable.

More information: 'Methylation of 12S rRNA Is Necessary for In Vivo Stability of the Small Subunit of the Mammalian Mitochondrial Ribosome', Metodi D. Metodiev, Nicole Lesko, Chan Bae Park, Yolanda Cámara, Yonghong Shi, Rolf Wibom, Kjell Hultenby, Claes M. Gustafsson and Nils-Göran Larsson, Cell Metabolism, 8 April 2009, doi:10.1016/j.cmet.2009.03.001.

Source: Karolinska Institutet (news : web)