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Review

C. elegans as a model organism for human mitochondrial associated disorders

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Abstract: Mitochondria are small cytoplasmic organelles whose most important function is to provide the energy required by our cells and organism to live. To maintain an adequate mitochondrial homeostasis cells possess numerous mitochondrial quality control and protective compensatory pathways, which can be activated to cope with a certain degree of mitochondrial dysfunction. However, when the mitochondrial damage is too severe and these defensive mechanisms are not anymore sufficient to deal with it, pathological signs arise. In the past few decades numerous genetic disorders ascribed to severe mitochondrial defects have been recognized with variable onset and symptomatology ranging from neuromuscular degeneration to cancer syndromes. Unfortunately, to date, only symptomatic and no curative therapies exist for most of these devastating, life-threating disorders. Model organisms, and especially the nematode *Caenorhabditis elegans*, with its sequenced and highly conserved genome, and a simple but well-characterized nervous system, have enormously contributed in the past years to gain insight into the pathogenesis and treatment of different diseases. Here, we will summarize some of the advantages offered by the nematode system to model neurodegenerative diseases associated with mitochondrial electron transport chain defects and screen for therapeutic interventions.

Keywords: mitochondria; mitochondrial diseases; neurodegeneration; *C. elegans*; *in vivo* drug screening; therapy.