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A friedreich's ataxia neuronal cell model with evidence of oxidative stress mediated neurodegeneration

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Friedreich's ataxia (FRDA) is caused by severely reduced levels of frataxin as a result of a large GAA triplet-repeat expansion within the first intron of the frataxin gene. The prominent feature of this pathology is the neurodegeneration interesting both central and peripheral nervous systems. A distal length-related axonal degeneration affects upper motor neurons of the corticospinal tract, the posterior columns of the spinal cord, the spinocerebellar tract and the large sensory fibers of the peripheral nerves. To date, there is still no known pharmacological treatment that cures, or even slows down, FRDA neuropathology. Indeed, the study of the selective vulnerability of specific neurons to frataxin deficiency is make it difficult by the scarce accessibility of the nervous tissue and by the lack of an appropriate neuronal cell model which reproduces the pathophysiology associated to FRDA.

We developed an in vitro system of FRDA neuronal degeneration by using the NSC34 motoneuronal cell line in which frataxin deficiency was obtained via siRNA gene silencing . We demonstrated that frataxin deficiency specifically reduce the activity of mitochondrial respiratory chain complex I and determine an oxidative stress condition showed by glutathione imbalance. Frataxin silencing does not affect neuronal differentiation and axon generation but make motoneuronal cells more sensitive to neuritis degeneration after treatment with several oxidant.

Our neuronal cell model has the potential to provide an experimentally accessible cell population to allow a more detailed examination, not only of nervous tissue specific FRDA pathophysiology, but also of oxidative stress-mediated neurodegeneration.

Biography

Barbara Carletti began her scientific career studying the development of neural progenitor cells by using the cerebellum as model system. She joined Dr. Ferdinando Rossi's research group at the University of Turin in 2000 after getting her Master's Degree in Biology at the University of Siena and she was enrolled in the PhD program in Neuroscience. At the beginning of 2007 she left Dr. Rossi's Department and joined the laboratory of Dr Carol Ann Mason at Columbia University, New York. She is currently working as junior research biologist at the "Bambino Gesù" Children's Hospital in Rome. She has published 12 papers in peer-reviewed journals.

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