

# INVESTIGATING THE ROLE OF THE LET-7 MICRORNA IN MACHADO-JOSEPH DISEASE

<https://neurodegenerationresearch.eu/survey/investigating-the-role-of-the-let-7-micrna-in-machado-joseph-disease/>

**Name of Fellow**

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**Institution****Funder**

FCT

**Contact information of fellow****Country**

Portugal

**Title of project/programme**

INVESTIGATING THE ROLE OF THE LET-7 MICRORNA IN MACHADO-JOSEPH DISEASE

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FCT

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€ 116,640

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**The project/programme is most relevant to:**

Spinocerebellar ataxia (SCA)

**Keywords****Research Abstract**

Machado-Joseph disease/spinocerebellar ataxia type 3 (MJD/SCA3) is a genetic neurodegenerative disorder associated with expansion of the number of CAGs within the coding region of the MJD1 gene, which translates into an expanded polyglutamine tract within ataxin-3, a protein involved in deubiquitination. MJD patients have severe clinical manifestations and

premature death and there is no treatment available.

We and others provided evidence that autophagy impairments contribute to MJD pathogenesis with autophagosome accumulation, reduction of autophagy-associated protein levels, accumulation of mutant ataxin-3 and neurodegeneration (Nascimento-Ferreira et al 2011, Brain). Very recently, a collaborator in this proposal (La Spada A., who first discovered polyglutamine diseases) brought evidence that the Let-7 microRNA is a key regulator of autophagy with particular relevance in polyglutamine disorders.

Therefore, in this project we will dissect the role of the Let-7 microRNA in the regulation of the autophagy pathway, investigate its de-regulation in MJD and evaluate its potential as a new therapeutic approach to MJD.

**Types:**

Fellowships

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Portugal

**Diseases:**

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