Centre de Recherche Méditerranéen
de Médecine Moléculaire

U 1065

Dr Jean-Ehrland RICCI,
Directeur de l'équipe 3

2 years postdoctoral position at INSERM U1065 (C3M)-team 3, Nice, France on the study of cell death of motor neurons. Starting March 2017.

A **two-year postdoctoral position** starting March 1st 2017, funded by the French National Research Agency is available in the 'Metabolic control of cell death' team (INSERM U1065), located at the Archet Hospital in **Nice, south of France** (http://www.unice.fr/c3m/index.php/research-teams/jean-ehrland-ricci/).

Title: how mitochondrial dysfunction leads to motor neuron disease?

Recently, in close collaboration with Pr. Paquis-Flucklinger, we showed that mitochondrial dysfunction can have a causative effect in motor neuron degeneration. We reported a large family with a mitochondrial myopathy associated with motor neuron disease and cognitive decline looking like frontotemporal dementia (FTD). We identified a missense mutation (p.Ser59Leu) in the CHCHD10 gene coding for a mitochondrial protein whose function was unknown (***Genin EC et al. EMBO Mol Med 2015 Dec 14:58-72***). We and others reported CHCHD10 mutations in patients with dementia-amyotrophic lateral sclerosis (FTD-ALS) and familial or sporadic pure ALS.

Project: Amyotrophic lateral sclerosis is a devastating disease affecting upper and lower motor neurons leading to progressive failure of the neuromuscular system and death from respiratory failure. Among all factors involved in ALS pathogenesis, mitochondrial dysfunction has always been recognized as a candidate major player. However, whether mitochondria have a causative role in ALS has been always debated. Our results open a new field to explore the pathogenesis of motor neuron disease by showing that mitochondrial dysfunction may be at the origin of some of these phenotypes.

Our goals are

(i) to better characterize the role of the CHCHD10 protein on cell death and to compare the effects of different CHCHD10 mutations leading to different clinical phenotypes,

(ii) to understand how CHCHD10 mutations lead to motor neuron cell death by generating specific human cellular (IPS) and characterizing in vivo models,

Candidate profile:

The candidate should hold a PhD in physiology, pharmacology or related disciplines and have previous expertise in induced pluripotent stem cells (IPS) generation/characterization (required) and She/he should be skilled in *in vivo* animal experimentation techniques (required) as well as in cellular and molecular biology techniques. Practice or knowledge of primary motor neuron culture and analysis would be appreciated.

How to apply?

Candidates should send a curriculum vitae with publication list, a short summary of research achievements, and the names and email addresses of at least two references to ricci@unice.fr

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