Cortico-Spinal Motor Neurons in Amyotrophic Lateral Sclerosis : Contribution, Mechanisms and Therapy

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Research Abstract
The long-term goal of this proposal is to shed light on the contribution of cortical upper motor neurons to amyotrophic lateral sclerosis, in order to design and test alternative therapies based on the protection and/or the replacement of this exact neuronal population.
Amyotrophic Lateral Sclerosis (ALS) is the most common and the most severe adult-onset neurodegenerative disease of the motor system, leading to death two to five years after diagnosis. ALS is clinically defined as the combined degeneration of two specific neuron types: upper motor neurons (UMN, corticospinal motor neurons), located in the cerebral cortex and that extend axons to the medulla and spinal cord, and lower motor neurons (LMN, spinal motor neurons), located in the medulla and spinal cord and that connect to the skeletal muscles. This dual neuronal impairment distinguishes ALS from other, much less severe, motor diseases affecting either UMN only or LMN only. Despite this clear description, it is striking to note that preclinical studies on ALS have, so far, mostly focused on LMN.
Recent advances in the fields of corticogenesis have provided scientists with new molecular implements allowing now to bypass the cellular complexity of the neocortex, and to study specific cortical neuronal subtypes (e.g. UMN), not only in the embryo but also in the diseased adult mouse. Here, we propose to take advantage of these new tools, to understand the role of specific UMN degeneration in the onset and progression of ALS. More specifically, we will i) determine whether specific UMN degeneration in ALS induces, results, or is independent from LMN degeneration; ii) unravel the molecular mechanisms that trigger specific UMN degeneration in ALS; and iii) induce the generation of new UMN, within the brain of the diseased mouse, and determine their effects on LMN survival, motor behavior and life expectancy. The expected results are meant to instruct the development of alternative therapeutic approaches to ALS.

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