# ERA-Net NEURON

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## iPSoALS: Modeling sporadic ALS in motor neurons by genetic reprogramming of patient skin fibroblasts

## Project Description

Amyotrophic lateral sclerosis (ALS) is a severe and incurable neurodegenerative disease. In ALS motor neurons in spinal cord, brainstem and cerebral cortex are progressively lost and disconnected from their targets. As a consequence patients lose control of voluntary movement and invariably die, most often from respiratory problems. ALS is also a major socio-economic issue. In Europe ALS is now more common than multiple sclerosis, Jacob-Creutzfeld disease and AIDS together and expenses for individual supportive care can exceed 150.000 Euros per year. Almost nothing is known about the disease mechanisms in sporadic ALS, the most common form. This is due to the difficulty to obtain human motor neurons and to study their degeneration in relevant model systems.

To overcome this limitation we propose to generate human motor neurons by a recently described technique of cellular reprogramming. Skin fibroblasts will be obtained from ALS patients and healthy volunteers and will be genetically re-programmed into motor neurons. We will analyze whether patient's motor neurons show changes in survival, growth or metabolic function, try to understand the underlying degenerative mechanisms and investigate whether these latter originate within motor neurons or in cells of their environment. To this purpose we gathered leading teams in ALS genetics, stem cell biology, motor neuron biology and life imaging working at renowned institutes in France, Germany, Israel and Sweden.

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