

Creating a new model of MND using stem cell technology

<https://neurodegenerationresearch.eu/survey/title-of-picreating-a-new-model-of-mnd-using-stem-cell-technology/>

Title of project or programme

Title of PI Creating a new model of MND using stem cell technology

Principal Investigators of project/programme grant

Title	Forname	Surname	Institution	Country
Professor Siddharthan	Chandran		University of Edinburgh	UK
Professor Ian		Wilmut	University of Edinburgh	UK
Professor Tom		Maniatis	Columbia University	USA
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Address of institution of lead PI

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- United Kingdom

Total sum awarded (Euro)

960000

Start date of award

01-06-2010

Total duration of award in months

36

The project/programme is most relevant to

- Motor neurone diseases

Keywords

fibroblasts, induced pluripotential stem cells, iPS cells, motor neurons, astrocytes

Research abstract in English

The discovery that TDP-43 is the major protein within ubiquitinated cytoplasmic inclusions – the hallmark pathology in 90% of familial and sporadic cases – and the recent identification of causative TDP-43 mutations in 1-4% of familial and sporadic cases, highlights the need for cellular models to

address the consequences of mutant TDP-43 in both motor neurons and astrocytes. Recent discoveries in reprogramming and resulting derivation of human iPS cell lines permit human-based models to study the consequences of disease-causing mutations. Against this background, our derivation and validation of TDP-43 mutant and control iPS lines offers new opportunities for the development of an in vitro human disease model of MND. Specifically, this collaborative project will exploit the unique opportunity of deriving patient-specific motor neurons and astrocytes carrying disease-causing TDP-43 mutations in order to address key questions concerning autonomous and non-cell autonomous mechanisms of motor neuron degeneration.

Lay summary

In which category does this research fall?

- Basic research