Molecular and Cellular Neurobiotechnology

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Descripción de las actividades del grupo.

1) *Role of PrP^C in epilepsy*
Rapid progressive dementia such us fast Alzheimer's disease or prionopathies are characterized by myoclonus and epilepsy. In humans, a decrease in the cellular prion protein PrP^C can be observed in these diseases. In a collaboration between four laboratories (J.M. Torres (INIA), Giuseppe Legname (SISSA), Isidre Ferrer (UB) and Franc Llorens (Germany) we determined the role of PrP^C in epilepsy. We determined that in absence of the protein neural excitability increases and neurons become more sensitive to kainate or glutamatergic insults. These effects can be seen in 4 different models of Prion diseases with different genetic background. Results were published in *Scientific Reports*.

2) Neurodegenerative diseases
We recently determined the role of a natural neural protein PrP^C in the evolution of Alzheimer's disease (published in *Molecular Neurobiology*). Results point to PrP^C as neuroprotective factor in Alzheimer's disease. Further experiments will continue in this direction, and also will be expanded to Parkinson's disease. Our hypothesis is that

PrP^C is a cross-link protein between different neurodegenerative diseases presenting taupathy. In addition, we determined that the N-terminal domain of the protein is the responsible of these neuroprotective effects.

3) Development of new lab on a chip devices for neurobiological research
We recently developed a new device able to reproduce axon lesioning *in vitro* in a single chip (published in *RSC Advances*). Current experiments of our group in collaboration with groups of IBEC and CIBER-BBN aimed at developing new lab on chip devices to mimics and modulate particular neurobiological processes. For example: cortico-spinal chips to develop genetic studies; molecular gradient generation for migrating neurons and *in silico* 3D modeling for neurodegenerative diseases

Publicaciones científicas más relevantes (últimos 6 años)

1. Ansoleaga B, Garcia-Esparcia P, Llorens F, Hernandez-Ortega K, Carmona Tech M, **Del Rio JA**, Zerr I, Ferrer I **(2016)** Altered Mitochondria, Protein Synthesis Machinery, and Purine Metabolism Are Molecular Contributors to the Pathogenesis of Creutzfeldt-Jakob Disease. J Neuropathol Exp Neurol. Jun 12. pii: nlw048.

2. Del Rio JA*, Gavin R **(2016)** Functions of the cellular prion protein, the end of Moore's law, and Ockham's razor theory. Prion.10(1):25-40.

3. Frau-Mendez MA, Fernandez-Vega I, Ansoleaga B, Blanco Tech R, Carmona Tech M, **Del Rio JA**, Zerr I, Llorens F, Jose Zarranz J, Ferrer I **(2016)** Fatal familial insomnia: mitochondrial and protein synthesis machinery decline in the mediodorsal thalamus. Brain Pathol. Jun 24. doi: 10.1111/bpa.12408

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5. Tomas-Roig J, Piscitelli F, Gil V, **Del Rio JA**, Moore TP, Agbemenyah H, Salinas-Riester G, Pommerenke C, Lorenzen S, Beissbarth T, Hoyer-Fender S, Di Marzo V, Havemann-Reinecke U **(2016)** Social defeat leads to changes in the endocannabinoid system: An overexpression of calreticulin and motor impairment in mice. Behav Brain Res.303:34-43.

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11. Llorens F, Zafar S, Ansoleaga B, Shafiq M, Blanco R, Carmona M, Grau-Rivera O, Nos C, Gelpi E, **Del Rio JA**, Zerr I, Ferrer I **(2015)** Subtype and regional regulation of prion biomarkers in sporadic Creutzfeldt-Jakob disease. Neuropathology and applied neurobiology 41 (5):631-645.

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Papel en el proyecto: CO-IP

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Coordinador: José A. Del Río

(2015-2018) Role of the cellular prion protein as “cross-talk” protein between □-syn/LRRK2 and p-Tau in sporadic and familiar Parkinson’s disease.
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