

JUNE
1ST 2017

VENUE / ILLKIRCH
> IGBMC
1 Rue Laurent Fries

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CONTROVERSY IN NEUROSCIENCE

ARE THE MOLECULAR
MECHANISMS OF TOXICITY
OF C9orf72 CAUSED
BY A LOSS-OF-FUNCTION?

Organizers: **José-Luis Gonzalez De Aguilar** (University of Strasbourg, France)
Luc Dupuis (University of Strasbourg, France)
Pascale Piguet (Neurex, Basel, Switzerland)

YES
NO

DEBATE INTRODUCED BY
JOSE-LUIS GONZALEZ DE AGUILAR

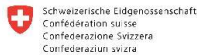
MODERATED BY
LUC DUPUIS
(UNIVERSITY OF STRASBOURG, FRANCE)

SPEAKERS :

LUDO VAN DEN BOSCH
(LEUVEN, BELGIUM)

EDOR KABASHI
(PARIS, FRANCE)

CLOTILDE LAGIER-TOURENNE
(CHARLESTOWN, MA, USA)



SUMMARY

The identification of a hexanucleotide repeat expansion mutation in a non-coding region of the gene on human chromosome 9 open reading frame 72 (*C9orf72*) has recently revolutionized the research on neurodegenerative diseases. *C9orf72* mutation is the most common cause of hereditary amyotrophic lateral sclerosis (ALS), which is also the most common motor neuron disease in the adult. Abnormal *C9orf72* expansion is also present in familial cases of frontotemporal dementia (FTD), which is frequently associated with ALS. At present, the way by which *C9orf72* mutation triggers neurodegeneration remains unclear, and several mechanisms have been proposed based on either a loss or a gain of function. These mechanisms include the decrease in the amount of functional protein, the accumulation in RNA foci of aberrant forms of *C9orf72* RNA, and the aggregation of dipeptide repeat proteins derived from an unusual translation process.

This controversy debate will expose arguments in support or in opposition to each of the postulated pathways. It will provide insight into the cellular and molecular aspects of such intriguing mechanisms of cell death. Discussions will draw the attention of not only experts on ALS/FTD research but also researchers in the field of neurodegeneration and beyond.

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NO

PROGRAM

- 09.00 - 09.10 **PRESENTATION OF THE SPEAKERS**
JOSE-LUIS GONZALEZ DE AGUILAR
(UMR_S INSERM 1118, Strasbourg, F)
- 09.10 - 09.25 ***C9orf72* IN NEURODEGENERATIVE DISEASES**
LUC DUPUIS (UMR_S INSERM 1118, Strasbourg, F)
- 09.25 - 10.10 **ARGUMENTS IN FAVOUR / AGAINST
A LOSS-OF-FUNCTION MECHANISM**
LUDO VAN DEN BOSCH (Leuven, B),
EDOR KABASHI (Paris, F)
& **CLOTILDE LAGIER-TOURENNE** (Charlestown, MA, US)
15 min each
- 10.10-10.25 **COFFEE BREAK**
- 10.25-11.45 **MOLECULAR MECHANISMS OF TOXICITY
OF *C9orf72*: THE DEBATE**
LUDO VAN DEN BOSCH (Leuven, B),
EDOR KABASHI (Paris, F)
& **CLOTILDE LAGIER-TOURENNE** (Charlestown, MA, US)
- 11.45 - 12.00 **CONCLUSION**
LUC DUPUIS (UMR_S INSERM 1118, Strasbourg, F)
& **NICOLAS CHARLET-BERGUERAND** (IGBMC, Illkirch, F)

