

LAUSANNE INTEGRATIVE METABOLISM AND NUTRITION ALLIANCE (LIMNA)

Monday January 12, 2015 10.30am
Conference Room: AI 1153 (*) EPFL – Lausanne

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**“Uncovering the role of misfolded SOD1 in the
pathogenesis of Amyotrophic Lateral Sclerosis”**

Hosts: Kristina Schoonjans and Johan Auwerx

Abstract

ALS is a neurodegenerative disorder characterized by the loss of motor neurons resulting in paralysis and death. Some Familial ALS cases are caused by mutations in SOD1, which lead to misfolding of the SOD1 protein, and gain of a toxic function. Several antibodies have been generated that are specific for the misfolded protein, and have been used for therapeutics. We used one antibody, B8H10, to demonstrate that misfolded SOD1 associates with mitochondria and those mitochondria have a significantly larger volume and produce more reactive oxygen species. We have demonstrated that not all misfolded SOD1 specific antibodies detect misfolded SOD1 at mitochondria. This finding suggests that there may be multiple misfolded SOD1 conformers. Using a panel of antibodies specific against misfolded SOD1 we aim to reveal which misfolded SOD1 conformers are relevant to mitochondrial dysfunction versus other mechanisms of disease. These studies will aid in identifying which misfolded SOD1 antibodies will be effective in therapeutics, and which SOD1 antibodies will be useful as markers of pathology.

(*) IMPORTANT NOTICE: All external participants have to pass through SV Reception/Welcome Desk to be able to access to AI 1153.
Contact person to call at arrival at SV Reception Desk: Johan Auwerx 30951/Administrative Assistant: 39522.

