

New results published in the peer-reviewed journal *Glia* further support masitinib's potential mode of action in ALS

AB Science SA (NYSE Euronext - FR0010557264 - AB) today announced the publication by an international team of researchers of a previously unknown mechanism linked to the progression of Amyotrophic Lateral Sclerosis (ALS) that further reinforces the rationale for masitinib's potential neuroprotective effect in ALS.

The publication, led by researchers from the Institut Pasteur de Montevideo, the University of Alabama at Birmingham (UAB), the Oregon State University (OSU) and the IMAGINE Institute of Paris, is entitled, 'Schwann cells orchestrate peripheral nerve inflammation through the expression of CSF1, IL-34, and SCF in amyotrophic lateral sclerosis'. This article is accessible online from the peer-reviewed scientific journal Glia https://onlinelibrary.wiley.com/doi/abs/10.1002/glia.23768.

This research provides strong evidence for a previously unknown inflammatory mechanism triggered by Schwann cells in ALS that can potentially be therapeutically targeted by masitinib.

"These findings provide a new and complementary mechanism of action for masitinib in ALS", said Professor Luis Barbeito, Head of the Neurodegeneration Laboratory (Institut Pasteur in Montevideo, Uruguay) and senior author of the paper.

Schwann cells are associated with neuronal maintenance and survival in the peripheral nervous system. Findings have revealed, for the first time, specific Schwann cell phenotypes in ALS that have the potential to trigger local inflammation through two signaling pathways (CSF-1R and c-Kit) targeted by masitinib. Remarkably, these Schwann cell phenotypes and their accompanying inflammatory effectors were present in both sporadic human ALS subjects and a paralytic animal model (SOD1G93A) of ALS. Pharmacological inhibition of CSF-1R and c-Kit with masitinib in the animal model of ALS sharply decreased Schwann cell reactivity, immune cell infiltration and proliferation along the peripheral motor pathways.

Dr Emiliano Trias, senior researcher (Institut Pasteur in Montevideo, Uruguay) and lead author of the paper said, "We showed for the first time that Schwann cells can orchestrate a harmful chronic inflammation along the peripheral motor pathway, contributing to ALS progression. This uncontrolled neuroinflammation can be inhibited by masitinib, even after onset of paralysis, by targeting cells implicated in injuring the peripheral nerves".

Overall, these data suggest a mechanism by which masitinib exerts a neuroprotective effect on peripheral nerve pathology and may further explain the multifaceted therapeutic effects of masitinib noted previously in ALS patients [1] and animal models [2–4].

- [1] Mora J, et al. Masitinib as an Add-on Therapy to Riluzole in Patients with Amyotrophic Lateral Sclerosis: A Randomised Clinical Trial. Amyotroph Lateral Scler Frontotemporal Degener. 2019 Jul 7:1-10. https://doi.org/10.1080/21678421.2019.1632346.
- [2] Trias E, et al. Mast cells and neutrophils mediate peripheral motor pathway degeneration in ALS. JCI Insight. JCI Insight. 2018;3(19):e123249. https://doi.org/10.1172/jci.insight.123249.
- [3] Trias E, et al. Evidence for mast cells contributing to neuromuscular pathology in an inherited model of ALS. JCI Insight. 2017;2(20):e95934. https://doi.org/10.1172/jci.insight.95934.

[4] Trias E, et al. Post-paralysis tyrosine kinase inhibition with masitinib abrogates neuroinflammation and slows disease progression in inherited amyotrophic lateral sclerosis. J Neuroinflammation. 2016;13(1):177.

About masitinib

Masitinib is a new orally administered tyrosine kinase inhibitor that targets mast cells and macrophages, important cells for immunity, through inhibiting a limited number of kinases. Based on its unique mechanism of action, masitinib can be developed in a large number of conditions in oncology, in inflammatory diseases, and in certain diseases of the central nervous system. In oncology due to its immunotherapy effect, masitinib can have an effect on survival, alone or in combination with chemotherapy. Through its activity on mast cells and microglia and consequently the inhibition of the activation of the inflammatory process, masitinib can have an effect on the symptoms associated with some inflammatory and central nervous system diseases and the degeneration of these diseases.

About AB Science

Founded in 2001, AB Science is a pharmaceutical company specializing in the research, development and commercialization of protein kinase inhibitors (PKIs), a class of targeted proteins whose action are key in signaling pathways within cells. Our programs target only diseases with high unmet medical needs, often lethal with short term survival or rare or refractory to previous line of treatment.

AB Science has developed a proprietary portfolio of molecules and the Company's lead compound, masitinib, has already been registered for veterinary medicine and is developed in human medicine in oncology, neurological diseases, and inflammatory diseases. The company is headquartered in Paris, France, and listed on Euronext Paris (ticker: AB).

Further information is available on AB Science's website: www.ab-science.com.

Forward-looking Statements - AB Science

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These forward-looking statements can often be identified by the words "expect", "anticipate", "believe", "intend", "estimate" or "plan" as well as other similar terms. While AB Science believes these forward-looking statements are reasonable, investors are cautioned that these forward-looking statements are subject to numerous risks and uncertainties that are difficult to predict and generally beyond the control of AB Science and which may imply that results and actual events significantly differ from those expressed, induced or anticipated in the forward-looking information and statements. These risks and uncertainties include the uncertainties related to product development of the Company which may not be successful or to the marketing authorizations granted by competent authorities or, more generally, any factors that may affect marketing capacity of the products developed by AB Science, as well as those developed or identified in the public documents filed by AB Science with the Autorité des Marchés Financiers (AMF), including those listed in the Chapter 4 "Risk Factors" of AB Science reference document filed with the AMF on November 22, 2016, under the number R. 16-078. AB Science disclaims any obligation or undertaking to update the forward-looking information and statements, subject to the applicable regulations, in particular articles 223-1 et seq. of the AMF General Regulations.

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