



InFlectis BioScience

Innovative therapeutics to treat protein misfolding diseases

Press release

InFlectis BioScience raises €6 million in a Series A round to perform clinical trial in humans

Nantes, France - June 1st, 2016. InFlectis BioScience SAS, a biopharmaceutical company specializing in the discovery and development of drugs for the treatment of protein misfolding diseases, today announced the completion of a Series A financing round amounting to $\in 6$ million. The financing was co-led by CM-CIC INNOVATION and REMIGES VENTURES with the participation of its historic shareholders, GO CAPITAL and PARTICIPATIONS BESANÇON. According to the financial terms of the agreement, the investors pay $\in 4.5$ million and have an investment option of an additional $\in 1.5$ million under certain pre-negotiated terms.

This fundraising will allow InFlectis BioScience to accelerate the development of its drug candidate IFB-088 by financing the regulatory preclinical development and the first clinical study in healthy volunteers. These studies are a prerequisite for the realization of a Phase 2 study currently planned in 2018 in patients affected by Charcot-Marie-Tooth (CMT), one of the most frequent peripheral neuropathies.

The IFB-088 drug candidate is an orally available small chemical molecule having a validated mechanism of action and a promising pharmacokinetic profile to target the central and peripheral nervous systems. IFB-088 is a selective inhibitor of PPP1R15A (GADD34), a regulatory subunit of PP1 phosphatase, induced by endoplasmic reticulum stress and involved in the response to misfolded proteins. The inhibition of PPP1R15A by IFB-088 extends the natural attenuation of the translation of proteins in stressed cells, allowing "chaperone" proteins that control the folding of proteins to restore the protein balance of the cells (i.e. proteostasis). Proof-of-concept studies in two CMT animal models have demonstrated IFB-088 efficacy on motor function of animals and the safety of this new therapeutic approach, foreshadowing a significant clinical potential.

Finally, the funds raised will also allow the selection of a second drug candidate, a selective inhibitor of PPP1R15A, as part of a separate program for the treatment of undisclosed degenerative diseases related to protein misfolding.

"InFlectis BioScience has developed a breakthrough technological platform around PPP1R15A inhibitors to address large unmet therapeutic needs, foremost of which is the disease, Charcot-Marie-Tooth. Its potential has immediately captivated us. Financing companies is our expertise, and we are delighted to support InFlectis BioScience in this stage of development of its first drug candidate IFB-088", together state Emilie LIDOME, Board Member of CM-CIC INNOVATION and Taro Inaba, Managing Partner of REMIGES VENTURES, co-leads of this Series A financing round. "We have supported InFlectis BioScience's research since its inception and are excited by the prospect of launching a clinical trial of its first drug candidate IFB-088" jointly declare Jérome Guéret, Managing Partner of GO CAPITAL and Pierre Besançon, Managing Partner of PARTICIPATION BESANÇON.

Philippe Guédat, President and CEO of InFlectis BioScience SAS, said: "The strong financial commitment of REMIGES VENTURES, a US fund with a strong connection to the Japanese pharmaceutical industry, and CM-CIC INNOVATION, alongside our historical shareholders GO CAPITAL and PARTICIPATIONS BESANÇON, demonstrates their confidence in the company's strategy as well as its potential; we thank them".

Notes for editors:

ABOUT CHARCOT-MARIE-TOOTH DISEASE

Charcot-Marie-Tooth disease (CMT), named for the three doctors who first described it, is one of the commonest inherited neurological disorders. Also known as hereditary motor and sensory neuropathy (HMSN) or peroneal muscular atrophy (PMA), the disease comprises a group of disorders that affect both motor and sensory peripheral nerves. The age of onset and associated disability vary widely, from a mild impairment of gait and balance in adulthood to a childhood requirement for a wheelchair. Symptoms usually begin before the age of 20 years, and include clumsiness, leg weakness, fatigue, and foot drop together with typical deformities that include unusually high-arched feet, hammer toes, and wasting of the lower legs. Nerve and muscle pain, decreased sensation, difficulty with mobility and balance, and wasting of hand muscles commonly occur.

CMT is classically divided into two major types, a demyelinating form (CMT1 and CMT4) and an axonal form (CMT2). CMT3, also known as Dejerine-Sottas syndrome, is a severe type of CMT in which symptoms begin in infancy or early childhood. An X-linked variant also occurs (CMTX). Approximately 60% of all CMT patients have CMT1, which is predominantly demyelinating. About 70% of these patients have CMT1A, which is associated with an autosomal dominant 1.4 MB duplication on chromosome 17p11.2 that includes the peripheral myelin protein 22 gene (PMP22) expressed predominantly in the compact myelin of Schwann cells. Another 5-10% of CMT1 cases have CMT1B, which is associated with mutations in the major myelin protein zero gene (MPZ). The CMT1A subtype is by far the most common form of CMT, followed by CMT1X, CMT1B and CMT2A. Together these four subtypes account for more than 85% of all genetic diagnoses in CMT.

ABOUT CM-CIC INNOVATION (www.cmcic-investissement.com):

CM-CIC Innovation is a subsidiary of CM-CIC Investissement specializing in venture capital investments. Its objective is to invest in companies developing promising technologies. CM-CIC Innovation selects companies with strong growth potential in dynamic sectors such as information technologies, telecommunications, electronics, life sciences, new materials or the environment. The policy of CM-CIC Innovation is to provide support with long-term capital to innovative startups to streamline their chances of success.

ABOUT REMIGES VENTURES (www.remigesventures.com)

Remiges Ventures is a US-based cross-border venture capital firm with close connections to the Japanese pharmaceutical industry having two major pharmaceutical companies in Japan as limited partners. Remiges has a strong focus on drug discovery and development and invests in early stage companies having promising drug candidates based on disruptive science. Remiges' goal is to support entrepreneurs to develop innovative therapeutics that help patients suffering from diseases.

ABOUT GO CAPITAL (www.gocapital.fr)

GO CAPITAL is a venture capital firm managing more than €120 million, mainly invested in highly innovative technology companies in the west of France. The fund GO CAPITAL Amorçage was made with the support of the French State-owned seed Fund (Fonds National d'Amorçage, FNA), managed by bpifrance's Future Investments Programme, the

European Investment Fund, the regions of Brittany, Pays de la Loire and Normandy, and banking partners Crédit Mutuel Arkea and Crédit Agricole. GO CAPITAL, through its funds GO CAPITAL Amorçage and Ouest Ventures, has made investments in about thirty companies in the areas of Digital, Health and Energy Transition

ABOUT PARTICIPATIONS BESANÇON

Participations Besançon is a family company headed by Mr. and Mrs. Pierre Besançon, head-quartered in Paris, France. Its primary investments are in listed and unlisted French pharmaceutical and biotech companies.

ABOUT INFLECTIS BIOSCIENCE (www.inflectisbioscience.com)

InFlectis BioScience aims to discover and develop new molecules for the treatment of protein misfolding diseases. The company plans to demonstrate the clinical effectiveness of its candidate IFB-088 in humans, then partner with a pharmaceutical company for its subsequent development and commercialization. Meanwhile the company continues to develop new chemical series for the treatment of non-orphan diseases whose etiology also lies in the accumulation of misfolded proteins. The Medical Research Council (MRC) in the UK is one of InFlectis BioScience historical shareholders with which it co-owns patents protecting the use of IFB-088 for the treatment of various neurodegenerative diseases, including CMT.

Based in Nantes in Western France, InFlectis BioScience is supported by Atlanpole (<u>www.atlanpole.com</u>), the science park of the economic area of Nantes Atlantique.

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