



INTRABIO EXTENSION PHASE FOR NIEMANN-PICK C ACCEPTED IN UNITED STATES

Oxford, UK / IntraBio Inc. is pleased to announce that the Extension Phase for the IB1001-201 Clinical Trial has been accepted by the FDA (US Food and Drug Administration). The Extension Phase has also been previously approved by all European countries in which the trial is being conducted, including Germany, Slovakia, Spain, and the UK.

IB1001-201 is a multinational, clinical trial that investigates IB1001 (N-acetyl-L-leucine) for the treatment of Niemann-Pick disease Type C (NPC). The acceptance of the Extension Phases provides US patients enrolled in the current trial the opportunity to continue treatment with IB1001 if the principal investigator determines it is in the patient's best interest.

The Extension Phase aims to provide further evidence of the symptomatic benefit of IB1001 and demonstrate the disease-modifying, neuroprotective effect of treatment. The potential benefit of long-term treatment has previously been observed in compassionate use studies and is supported by in vitro and in vivo data.

NPC is a rare, debilitating, inherited lysosomal storage disorder that predominately affects pediatric patients. The disease begins in early childhood and is chronic and progressive in nature and severely impacts quality of life. The average age of death for NPC patients is approximately 10 years, with half of the patients dying before the age of 12.5 years.

In addition to Clinical Study IB1001-201, IntraBio is running parallel multinational clinical trials involving IB1001: IB1001-202 for the treatment of GM2 Gangliosidosis (Tay-Sachs and Sandhoff disease), and IB1001-203 for the treatment of Ataxia-Telangiectasia (A-T). To investigate both the symptomatic and neuroprotective, disease-modifying effects of treatment, IB1001 is assessed during two treatment sequences: a 6-week parent study, and a one-year extension phase. The parent studies of IB1001-201 and IB1001-202 are expected to be complete Q2 2020. Extension Studies are being conducted for all trials.

About IntraBio Inc.

IntraBio Inc is a biopharmaceutical company with a late-stage drug pipeline including novel treatments for common and rare neurodegenerative diseases. IntraBio's platform results from decades of research and investment at premier universities and institutions worldwide. Its clinical programs leverage the expertise in lysosomal function of its scientific founders from the University of Oxford and the University of Munich.

IntraBio's management team and consultants have vast commercial experience and a successful track record of drug development in the US and Europe. Together, IntraBio's team translates innovative scientific research in the fields of lysosomal biology, autophagy, and neurology into novel drugs for a broad spectrum of genetic and neurodegenerative diseases so to significantly improve the lives of patients and their families.

IntraBio Inc. is a US corporation with its principal offices in Oxford, United Kingdom.