

Inventiva selected to present the positive results from its Phase IIa clinical study with odiparcil in MPS VI at the 16th Annual WORLDSymposium™

- ▶ The Phase IIa clinical study results in MPS VI adult patients showed the capacity of odiparcil to improve key clinical parameters such as corneal clouding and cardiac and respiratory functions
- ▶ The study also met its safety primary objective, confirming the good safety profile of odiparcil

Daix (France), February 3, 2020 – Inventiva (Euronext: IVA), a clinical-stage biopharmaceutical company developing oral small molecule therapies for the treatment of diseases in the areas of fibrosis, lysosomal storage disorders and oncology, today announced that Dr. Nathalie Guffon (MD, Hôpital Femme-Mère-Enfant, Lyon, France), investigator of the iMProveS (improve MPS treatment) Phase IIa clinical study, will present a poster, entitled “Treatment of mucopolysaccharidosis type VI patients with odiparcil alone or in addition to enzyme replacement therapy: A Phase IIa study”, at the 16th Annual WORLDSymposium™ taking place from February 10 to 13, 2020 at the Hyatt Regency Orlando, Orlando, Florida.

Dr. Nathalie Guffon will present the positive results of the iMProveS Phase IIa clinical study published on December 18, 2019, which showed that odiparcil improved key clinical parameters in several patients such as corneal clouding and cardiac and respiratory functions. In addition, the clinical study met its safety primary objective further supporting the good overall safety profile of odiparcil already observed in previous Phase I and Phase II clinical studies. Based on the iMProveS clinical study results, Inventiva has decided to continue the clinical development of odiparcil for the treatment of MPS VI.

Pierre Broqua, Chief Scientific Officer and cofounder of Inventiva, said: “We are very excited by the results of the iMProveS study showing that odiparcil is able to treat hard-to-reach tissues and improve cardiac and respiratory functions. Given that enzyme replacement therapy as current standard of care shows limited efficacy in these organs, we believe that odiparcil could become a reference treatment for MPS VI. We look forward to present our promising results at the upcoming congress and to pursue the clinical development of odiparcil.”

The presentation details are as follows:

Presentation title:	“Treatment of mucopolysaccharidosis type VI patients with odiparcil alone or in addition to enzyme replacement therapy: A Phase IIa study” (poster number: LB-17)
Presenter:	Dr. Nathalie Guffon
Date:	February 12, 2020
Time:	4.30pm - 6.30pm (Eastern Time)
Location:	Hyatt Regency Orlando, 9801 International Drive, Orlando, Florida, United States

About the WORLDSymposium™

The WORLDSymposium™ is a leading annual research conference dedicated to lysosomal diseases. Since 2002, the W.O.R.L.D. (We're Organizing Research on Lysosomal Diseases) meeting has grown to an international research conference that attracts over 1600 participants from more than 50 countries around the globe every year.

About the iMProveS Phase IIa clinical trial

The iMProveS (*improve MPS treatment*) study was a 26-week Phase IIa clinical trial taking place in four European sites and evaluating odiparcil for the treatment of patients with mucopolysaccharidosis ("MPS") type VI. The primary endpoint of the trial was safety, as assessed by clinical and biological standard tests. Secondary endpoints included changes from baseline in leukocyte, skin and urinary glycosaminoglycan ("GAG") content, improvements of activity and mobility, evaluation of cardiovascular, lung and respiratory function and vision and hearing impairments.

For this trial, 20 patients, aged 16 years or older, had been enrolled. Patients receiving enzyme replacement therapy (ERT) and one of two odiparcil doses (500mg or 250mg twice a day) or placebo had been randomized in the double-blind placebo-controlled cohort. Patients not being treated with ERT and only receiving the high dose of odiparcil (500mg twice a day) had been enrolled in the open label cohort.

About odiparcil and mucopolysaccharidoses

Odiparcil is an orally-available small molecule that acts on the underlying cause of the symptoms of mucopolysaccharidosis ("MPS"), a group of rare, progressive genetic disorders. MPS is characterized by the accumulation of glycosaminoglycans ("GAGs"), polysaccharides which are important for the modulation of cell to cell signalling and the maintenance of tissue structure and function, in the lysosomes of cells. Due to genetic mutations, lysosomes in patients with MPS contain deficient versions of the enzymes necessary to break down GAGs. As a result, GAGs accumulate within the lysosomes, causing the latter to swell and interfere with the ordinary functioning of cells, leading to the symptoms associated with MPS. MPS is categorized by subtypes, depending on the enzyme that is deficient and the corresponding GAGs that accumulate. By modifying how GAGs are synthesized, odiparcil facilitates the production of soluble GAGs that can be excreted in the urine, rather than accumulating in cells. Specifically, odiparcil acts on chondroitin sulfate ("CS") and dermatan sulfate ("DS"), either or both of which accumulate in patients with MPS I, II, IVa, VI and VII.

A Phase I/II clinical study in children with MPS VI is currently under preparation following the positive results of a Phase IIa clinical study in adult MPS VI patients published at the end of 2019.

Odiparcil has been granted Orphan Drug Designation (ODD) by the U.S. Food and Drug Administration (FDA) and the European Medicines Agency (EMA) and has also obtained Rare Pediatric Disease Designation (RPDD) in the U.S. for the treatment of MPS VI.

About Inventiva

Inventiva is a clinical-stage biopharmaceutical company focused on the development of oral small molecule therapies for the treatment of diseases with significant unmet medical needs in the areas of fibrosis, lysosomal storage disorders and oncology.

Leveraging its expertise and experience in the domain of compounds targeting nuclear receptors, transcription factors and epigenetic modulation, Inventiva is currently advancing two clinical candidates – lanifibranor and

odiparcil – in non-alcoholic steatohepatitis (“NASH”) and mucopolysaccharidosis (“MPS”), respectively, as well as a deep pipeline of earlier stage programs.

Lanifibranor, its lead product candidate, is being developed for the treatment of patients with NASH, a common and progressive chronic liver disease. Inventiva is currently evaluating lanifibranor in a Phase IIb clinical trial for the treatment of this disease for which there are currently no approved therapies.

Inventiva is also developing odiparcil, a second clinical stage asset, for the treatment of patients with MPS, a group of rare genetic disorders. A Phase I/II clinical study in children with MPS VI is currently under preparation following the positive results of a Phase IIa clinical study in adult MPS VI patients published at the end of 2019.

In parallel, Inventiva is in the process of selecting an oncology development candidate for its Hippo signalling pathway program. Furthermore, the Company has established a strategic partnership with AbbVie in the area of autoimmune diseases. AbbVie has started the clinical development phase of ABBV-157, a drug candidate for the treatment of moderate to severe psoriasis resulting from its collaboration with Inventiva. This collaboration entitles Inventiva to receive milestone payments upon the achievement of pre-clinical, clinical, regulatory and commercial milestones, in addition to royalties on any approved products resulting from this partnership.

The Company has a scientific team of approximately 70 people with deep expertise in the fields of biology, medicinal and computational chemistry, pharmacokinetics and pharmacology as well as in clinical development. It also owns an extensive library of approximately 240,000 pharmacologically relevant molecules, around 60% of which are proprietary, as well as a wholly-owned research and development facility.

Inventiva is a public company listed on compartment C of the regulated market of Euronext Paris (Euronext: IVA – ISIN: FR0013233012). www.inventivapharma.com

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statements, forecasts and estimates. Furthermore, forward-looking statements, forecasts and estimates only speak as of the date of this press release. Readers are cautioned not to place undue reliance on any of these forward-looking statements.

Please refer to the "Document de référence" filed with the Autorité des Marchés Financiers on April 12, 2019 under n° R.19-006 for additional information in relation to such factors, risks and uncertainties.

Inventiva has no intention and is under no obligation to update or review the forward-looking statements referred to above. Consequently, Inventiva accepts no liability for any consequences arising from the use of any of the above statements.