PRESS RELEASE

Ipsen and Exicure enter into exclusive collaboration targeting rare neurodegenerative disorders

- Ipsen obtains exclusive options to Spherical Nucleic Acids (SNAs™) currently under discovery evaluation for Huntington's disease and Angelman syndrome
- Exicure will be responsible for discovery and certain pre-clinical development activities. In the event Ipsen exercises its option to the two programs, Ipsen will be responsible for further development and worldwide commercialization
- Exicure will receive a $20m upfront payment and is eligible to receive up to $1B in option exercise fees and milestone payments should Ipsen opt into both programs, as well as tiered royalties

Paris (France), and Chicago, IL, Cambridge, MA (USA) Monday 2 August 2021 – Ipsen (Euronext: IPN; ADR: IPSEY) and Exicure Inc. (NASDAQ: XCUR) have signed an exclusive collaboration agreement to research, develop, and commercialize novel Spherical Nucleic Acids (SNAs) as potential investigational treatments for Huntington's disease and Angelman syndrome.

Oligonucleotides are synthetic structures of nucleic acids that can be used to modulate gene expression via a range of processes, including gene activation, inhibition, and splice-modulation. These molecules have demonstrated potential in many different therapeutic areas.[1] Achieving efficient oligonucleotide delivery to target organs and tissues, including the brain, remains a major limitation to their use.[1,2] Exicure's SNAs provide distinct chemical and biochemical properties to oligonucleotides. In preclinical models, SNAs have been shown to enhance the cell penetration, biodistribution and organ persistence properties of oligonucleotides,[3,4] which may potentially enhance drug delivery to previously inaccessible target tissues, including deep brain regions.[5,6]

Philippe Lopes-Fernandes, Chief Business Officer at Ipsen, said "Neuroscience is deeply rooted within Ipsen as a key strategic driver for our business. We are pleased to partner with Exicure to progress development of investigational treatment options for Huntington's disease and Angelman syndrome, two areas of significant unmet need. This collaboration marks an important step in maximizing the potential of this novel technology, bringing together the expertise of Exicure and the robust heritage of Ipsen in neuroscience. With this new collaboration we will deepen our commitment to people living with neurological conditions around the world."

"We are thrilled to partner with Ipsen, a leading global company with significant expertise and commitment to developing treatments for patients with rare neurological diseases," said David Giljohann, Ph.D., Chief Executive Officer, Exicure, Inc. "In collaboration with Ipsen, we have the opportunity to apply our technology to Huntington's disease and Angelman syndrome, both indications requiring deep brain penetration and technological advances to reach previously hard-to-drug targets. We believe our platform technology with its deep penetration and persistence of medicinal effect will allow Exicure and Ipsen to overcome challenges from first-generation oligonucleotides and bring new medicines to patients in need."

Under the agreement, Ipsen will receive exclusive options to license SNA-based therapeutics arising from two collaboration programs for Huntington's disease and Angelman syndrome. Ipsen will pay Exicure a cash upfront payment of $20m upon closing and Exicure will be responsible for discovery and certain pre-clinical development activities. In the event Ipsen exercises its option, Ipsen will be responsible for further development and commercialization of the licensed products. Exicure will receive a $20m upfront payment and is eligible to receive up to $1B in option exercise fees and milestone payments should Ipsen opt into both programs, as well as tiered royalties.

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Huntington's disease
Huntington's disease (HD) is a progressive, fatal neurodegenerative disorder and the most common monogenic neurological disorder in the developed world, affecting about 40,000 individuals in the US.[7] HD is caused by an expanded CAG trinucleotide repetition in the huntingtin *(HTT)* gene in chromosome 4. HD is characterized by involuntary movements, psychiatric disorders, cognitive deterioration, and early mortality, with death often occurring within 10 to 20 years after motor symptoms appear. Mean age of onset of motor symptoms is around 40 years of age, with longer CAG repeats causing earlier disease onset.[8] There is currently no approved therapy to address the underlying molecular cause of HD to slow or stop disease progression.[9]

Angelman syndrome
Angelman syndrome (AS) is a severe neurodevelopmental disorder. The prevalence of Angelman syndrome is estimated to be 1 in 12,000-20,000 people in the general population.[10] The disorder is characterized by severe intellectual deficit, speech impairment, epilepsy, ataxic movements and behavioral abnormalities. AS results from loss of function of the maternally inherited copy of the ubiquitin-protein ligase E3A (UBE3A) gene on chromosome 15.[11] Disruption of UBE3A function in neurons prevents synapse formation and remodeling, leading to significant neurodevelopmental disability. There is currently no approved disease-modifying therapy for AS and standard-of-care treatment is supportive, such as medications for seizures and behavioral abnormalities.[12]

Ipsen
Ipsen is a global, mid-sized biopharmaceutical company focused on transformative medicines in Oncology, Rare Disease and Neuroscience; it also has a well-established Consumer Healthcare business. With Total Sales of over €2.5bn in FY 2020, Ipsen sells more than 20 medicines in over 115 countries, with a direct commercial presence in more than 30 countries. The Company's research and development efforts are focused on its innovative and differentiated technological platforms located in the heart of leading biotechnological and life-science hubs: Paris-Saclay, France; Oxford, U.K.; Cambridge, U.S.; Shanghai, China. Ipsen has c.5,700 colleagues worldwide and is listed in Paris (Euronext: IPN) and in the U.S. through a Sponsored Level I American Depositary Receipt program (ADR: IPSEY). For more information, visit ipsen.com.

Exicure, Inc.
Exicure, Inc. is a clinical-stage biotechnology company developing therapeutics for neurology, immuno-oncology, inflammatory diseases, and other genetic disorders based on its proprietary Spherical Nucleic Acid, or SNA technology. Exicure believes that its proprietary SNA architecture has distinct chemical and biological properties that may provide advantages over other nucleic acid therapeutics and may have therapeutic potential to target diseases not typically addressed with other nucleic acid therapeutics. Exicure is in preclinical development of XCUR-FXN a lipid-nanoparticle SNA–based therapeutic candidate, for the intrathecal treatment of Friedreich's ataxia (FA). Exicure's therapeutic candidate cavrotolimod (AST-008) is in a Phase 1b/2 clinical trial in patients with advanced solid tumors. Exicure is based in Chicago, IL and in Cambridge, MA.

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Ipsen's forward-looking statements

The forward-looking statements, objectives and targets contained herein are based on Ipsen's management strategy, current views and assumptions. Such statements involve known and unknown risks and uncertainties that may cause actual results, performance or events to differ materially from those anticipated herein. All of the above risks could affect Ipsen's future ability to achieve its financial targets, which were set assuming reasonable macroeconomic conditions based on the information available today. Use of the words 'believes', 'anticipates' and 'expects' and similar expressions are intended to identify forward-looking statements, including Ipsen's expectations regarding future events, including regulatory filings and determinations. Moreover, the targets described in this document were prepared without taking into account external growth assumptions and potential future acquisitions, which may alter these parameters. These objectives are based on data and assumptions regarded as reasonable by Ipsen. These targets depend on conditions or facts likely to happen in the future, and not exclusively on historical data. Actual results may depart significantly from these targets given the occurrence of certain risks and uncertainties, notably the fact that a promising product in early development phase or clinical trial may end up never being launched on the market or reaching its commercial targets, notably for regulatory or competition reasons. Ipsen must face or might face competition from generic products that might translate into a loss of market share. Furthermore, the Research and Development process involves several stages each of which involves the substantial risk that Ipsen may fail to achieve its objectives and be forced to abandon its efforts with regards to a product in which it has invested significant sums. Therefore, Ipsen cannot be certain that favorable results obtained during pre-clinical trials will be confirmed subsequently during clinical trials, or that the results of clinical trials will be sufficient to demonstrate the safe and effective nature of the product concerned. There can be no guarantees a product will receive the necessary regulatory approvals or that the product will prove to be commercially successful. If underlying assumptions prove inaccurate or risks or uncertainties materialize, actual results may differ materially from those set forth in the forward-looking statements. Other risks and uncertainties include but are not limited to, general industry conditions and competition; general economic factors, including interest rate and currency exchange rate fluctuations; the impact of pharmaceutical industry regulation and health care legislation; global trends toward health care cost containment; technological advances, new products and patents attained by competitors; challenges inherent in new product development, including obtaining regulatory approval; Ipsen's ability to accurately predict future market conditions; manufacturing difficulties or delays; financial instability of international economies and sovereign risk; dependence on the effectiveness of Ipsen's patents and other protections for innovative products; and the exposure to litigation, including patent litigation, and/or regulatory actions. Ipsen also depends on third parties to develop and market some of its products which could potentially generate substantial royalties; these partners could behave in such ways which could cause damage to Ipsen's activities and financial results. Ipsen cannot be certain that its partners will fulfil their obligations. It might be unable to obtain any benefit from those agreements. A default by any of Ipsen's partners could generate lower revenues than expected. Such situations could have a negative impact on Ipsen's business, financial position or performance. Ipsen expressly disclaims any obligation or undertaking to update or revise any forward-looking statements, targets or estimates contained in this press release to reflect any change in events, conditions, assumptions or circumstances on which any such statements are based, unless so required by applicable law. Ipsen's business is subject to the risk factors outlined in its registration documents filed with the French Autorité des Marchés Financiers. The risks and uncertainties set out are not exhaustive and the reader is advised to refer to Ipsen's 2020 Registration Document, available on ipsen.com.

Exicure's forward-looking statements

This press release contains forward-looking statements within the meaning of the Private Securities Litigation Reform Act of 1995. All statements in this press release other than statements of historical fact could be deemed forward looking including, but not limited to, statements regarding the company's exclusive collaboration with Ipsen; the ability of SNAs to potentially enhance drug delivery to previously inaccessible target tissues and other benefits of SNAs including as potential treatment options for Huntington's disease and Angelman syndrome; the ability of the company's technology to overcome challenges from first-generation oligonucleotides and bring new drugs to patients in need; the ability of the company to realize contingent milestone payments and royalties under the collaboration agreement with Ipsen; and the advancement, timing and success of the company's preclinical and clinical programs. The forward-looking statements in this press release speak only as of the date of this press release, and the company undertakes no obligation to update these forward-looking statements. Forward-looking statements are based on management's current beliefs and assumptions that are subject to risks and uncertainties and are not guarantees of future performance. Actual results could differ materially from those contained in any forward-looking statement as a result of various factors, including, without limitation: the risks that the ongoing COVID-19 pandemic may disrupt the company's business and/or the global healthcare system more severely than it has to date or more severely than anticipated; unexpected costs, charges or expenses that reduce the company's capital resources; the company's preclinical or clinical programs do not advance or result

in approved products on a timely or cost effective basis or at all; the results of early clinical trials are not always being predictive of future results; the cost, timing and results of clinical trials; that many drug candidates do not become approved drugs on a timely or cost effective basis or at all; the ability to enroll patients in clinical trials; possible safety and efficacy concerns; the ability of the company to collaborate successfully with strategic partners; regulatory developments; exposure to litigation, including patent litigation, and/or regulatory actions; and the ability of the company to protect its intellectual property rights. For a discussion of other risks and uncertainties, and other important factors, any of which could cause the company's actual results to differ from those contained in the forward-looking statements, see the section titled "Risk Factors" in the company's Annual Report on Form 10-K for the year ended December 31, 2020, as updated by the company's subsequent filings with the Securities and Exchange Commission. All information in this press release is as of the date of the release, and the company undertakes no duty to update this information, except as required by law.

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