Nxera Pharma Secures Exclusive Rights to Fabry Disease Phase 3 Oral Therapy Candidate Lucerastat for Japan and Selected Asia Pacific Markets

- Nxera has exercised its option to license lucerastat for Japan, South Korea, Taiwan and selected other Asia Pacific markets (excluding China), paying Idorsia CHF 7 million upfront and contributing to the global Phase 3 program.
- Lucerastat is an investigational oral therapy for Fabry disease with more than 400 patient-years of clinical experience, adding a late-stage rare disease asset to Nxera's portfolio in Japan and Asia Pacific.
- Idorsia today initiated FAB-KLEAR, a pivotal Phase 3 study, as part of a global program expected to support a regulatory filing as early as 2029. Nxera will take part in the program and be responsible for managing clinical development in Japan.

Tokyo, Japan and Cambridge, UK, 28 October 2026 – Nxera Pharma Co., Ltd. ("Nxera" or the "Company"; TSE: 4565) today announces that it has exercised its option to license lucerastat from Idorsia Ltd ("Idorsia"; SIX: IDIA) and has entered into a co-development and commercialization agreement (the "Agreement"). Under the Agreement, Nxera will hold exclusive rights to develop and commercialize lucerastat in Japan, South Korea, Taiwan and selected other Asia Pacific markets, excluding China (the "Territory").

Idorsia granted Nxera the option in July 2023, as part of the Company's (then Sosei Heptares) acquisition of Idorsia's Asia Pacific (ex-China) operations. Nxera will pay Idorsia an upfront payment of CHF 7 million, in line with the pre-agreed option terms, and will also contribute to the global Phase 3 development program. Idorsia is eligible to receive royalties on net sales in the Territory.

Fabry disease is a rare, inherited condition in which a fatty substance called Gb3 builds up in cells, progressively damaging the kidneys, heart and nervous system. Lucerastat is designed to reduce the production of Gb3 and to work regardless of a patient's genetic mutation or prior treatment. In Idorsia's Phase 3 MODIFY study and its long-term extension, lucerastat showed sustained reductions in Gb3 and was associated with a slower rate of kidney function decline compared with the period before treatment. Some patients in the clinical trial have now received lucerastat for more than six years.

Idorsia's global Phase 3 program includes FAB-KLEAR, a kidney biopsy study in 16 adult male patients, and a 74-patient study comparing lucerastat with standard-of-care enzyme replacement therapies on kidney function. The program has been aligned with the US FDA on a streamlined registration strategy and is also in line with feedback received from the European Medicines Agency. Nxera will take part in the comparative

kidney function study and will be responsible for managing clinical development in Japan.

Christopher Cargill, President and CEO of Nxera Pharma, commented: "The clinical evidence generated to date, together with the defined Phase 3 development program, gives us confidence in the potential of lucerastat for people living with Fabry disease. We look forward to working closely with Idorsia to advance development and bring this potential new treatment option to patients across our territories. Lucerastat adds a late-stage rare disease asset to our growing portfolio in Japan and across Asia Pacific."

Amer Joseph, MD, Chief Medical Officer and Head of Global Clinical Development at Idorsia, commented: "The recently published Phase 3 MODIFY study and its long-term extension have provided strong evidence of lucerastat's disease-modifying potential. While the primary endpoint of neuropathic pain reduction was not met, a subjective outcome that is proving difficult to reach in Fabry disease, lucerastat demonstrated robust and sustained biomarker reductions and a promising signal for slowing renal function decline in patients. The next step is to generate the baseline-controlled data required by authorities for registration."

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About Lucerastat
Lucerastat is an investigational, oral substrate reduction therapy designed to treat Fabry disease independently of α-galactosidase A activity, GLA mutation status or prior enzyme replacement therapy. It acts by inhibiting glucosylceramide synthase, reducing the synthesis of glycosphingolipids, including globotriaosylceramide (Gb3). In the 118-patient Phase 3 MODIFY study, lucerastat significantly reduced plasma and urinary Gb3 compared with placebo. In an interim analysis of the open-label extension, treatment was associated with a slower rate of kidney function decline compared with the two years before enrollment (eGFR slope: -1.64 on lucerastat vs. -3.50 historical), and lucerastat was well tolerated with long-term treatment. Lucerastat has received orphan drug designation in the US and the EU.

About Fabry Disease
Fabry disease is a rare, X-linked lysosomal storage disorder caused by mutations in the GLA gene, resulting in absent or markedly reduced activity of the enzyme α-galactosidase A. This leads to a build-up of Gb3 throughout the body, causing multisystem disease that mainly affects the kidneys, heart and nervous system, and reduces life expectancy and quality of life. Fabry disease is frequently underdiagnosed or misdiagnosed. According to DelveInsight, approximately 16,000 patients were diagnosed with Fabry disease across the seven major markets (US, EU4, UK and Japan) in 2020, a number expected to rise to roughly 21,000 by 2034, and the Fabry disease market is projected to reach around US$4 billion by 2034.

About Idorsia
The purpose of Idorsia is to discover, develop and commercialize innovative medicines to help more patients. To achieve this, we will develop Idorsia into a leading biopharmaceutical company, with a strong scientific core. Headquartered near Basel, Switzerland – a European biotech hub – Idorsia has a highly experienced team of dedicated professionals, covering all disciplines from bench to bedside; QUVIVIQ™ (daridorexant), a different kind of insomnia treatment with the potential to revolutionize this mounting public health concern; strong partners to maximize the value of our portfolio; a promising in-house development pipeline; and a specialized drug discovery engine focused on small-molecule drugs that can change the treatment paradigm for many patients. Idorsia is listed on the SIX Swiss Exchange (ticker symbol: IDIA).

About Nxera Pharma
Nxera Pharma is a technology-powered biopharma company in pursuit of new specialty medicines to

improve the lives of patients with unmet needs in Japan and globally. The Company has built an agile, new-generation commercial business in Japan to develop and commercialize innovative medicines, including several launched products, to address this high-value, large and growing market and those in the broader APAC region. In addition, the Company is advancing an extensive pipeline internally and in partnership with leading pharma and biotech companies, powered by its unique NxWave™ GPCR structure-based drug discovery platform. Nxera Pharma operates at key locations in Tokyo and Osaka (Japan), London and Cambridge (UK), Basel (Switzerland) and Seoul (South Korea) and is listed on the Tokyo Stock Exchange (ticker: 4565).

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Forward-looking statements

This press release contains forward-looking statements, including statements about the discovery, development, and commercialization of products. Various risks may cause Nxera Pharma Group's actual results to differ materially from those expressed or implied by the forward looking statements, including: adverse results in clinical development programs; failure to obtain patent protection for inventions; commercial limitations imposed by patents owned or controlled by third parties; dependence upon strategic alliance partners to develop and commercialize products and services; difficulties or delays in obtaining regulatory approvals to market products and services resulting from development efforts; the requirement for substantial funding to conduct research and development and to expand commercialization activities; and product initiatives by competitors. As a result of these factors, prospective investors are cautioned not to rely on any forward-looking statements. We disclaim any intention or obligation to update or revise any forward-looking statements, whether as a result of new information, future events or otherwise.