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Company Name: AnGes Inc.

Representative: Ei Yamada, President & CEO

Eiger BioPharmaceuticals and AnGes Announce Exclusive Partnership for Regulatory Approval and Commercialization of Zokinvy® (lonafarnib) in Japan

Eiger BioPharmaceuticals Inc. (“Eiger”, NASDAQ: EIGR) and AnGes Inc. (“AnGes”, TYO: 4563) today announced that the companies entered into an agreement for the regulatory approval, marketing, and distribution of Zokinvy® (lonafarnib) for the treatment of Hutchinson-Gilford progeria syndrome (HGPS or progeria) and processing-deficient progeroid laminopathies (PL) in Japan.

Under the terms of the agreement, AnGes is responsible for obtaining and maintaining regulatory approval for Zokinvy in Japan and will be the exclusive partner for distribution and commercialization. Eiger will receive upfront and milestone payments up to \$1.5 million, as well as earn revenue from the sale of Zokinvy to AnGes.

“We are thrilled to build a new partnership with Eiger for Zokinvy in Japan. We are committed to obtaining regulatory approval in an expeditious manner to deliver Zokinvy to the HGPS and PL patients in Japan,” said Ei Yamada, President and CEO, AnGes. “In parallel with the regulatory approval process, we will persistently advance our preparations to initiate the diagnostic test for HGPS as part of developing the genetic disease diagnosis testing of new-born babies at AnGes Clinical Research Laboratory (ACRL) established last year.”

“We are pleased to enter into this partnership with AnGes to seek regulatory approval and commercialization of Zokinvy in Japan to help patients suffering from progeria and progeroid laminopathies,” said David Cory, President and CEO, Eiger. “This collaboration is representative of Eiger’s strategic approach to fully leverage our innovative therapies to enhance shareholder value and support underserved patients around the world.”

About progeria and progeroid laminopathies

Progeria, also known as Hutchinson-Gilford progeria syndrome, and progeroid laminopathies are separate and distinct ultra-rare, fatal, genetic premature aging

diseases that accelerate mortality in young patients. It is estimated that there are 400 children worldwide with progeria and 200 children with progeroid laminopathies.

Progeria is caused by a point mutation in the LMNA gene, yielding the farnesylated aberrant protein, progerin. Progeroid laminopathies are genetic conditions of accelerated aging caused by a constellation of mutations in the LMNA and/or ZMPSTE24 genes yielding farnesylated proteins that are distinct from progerin. While non-progerin producing, these genetic mutations result in disease manifestations with phenotypes that have overlap with, but are distinct from, progeria.

Without Zokinvy therapy, children with progeria commonly die of the same heart disease that affects millions of normally aging adults (arteriosclerosis), by an average age of 14.5 years. Disease manifestations include severe failure to thrive, scleroderma-like skin, global lipodystrophy, alopecia, joint contractures, skeletal dysplasia, global accelerated atherosclerosis with cardiovascular decline, and debilitating strokes.

About Zokinvy® (lonafarnib)

Zokinvy was approved in the U.S. in November 2020 to reduce the risk of death with HGPS (progeria), and to treat processing-deficient progeroid laminopathies. It is indicated for adults and children over 12 months of age.

Zokinvy blocks the accumulation of defective, farnesylated proteins which form tight associations with the nuclear envelope, leading to cellular instability and premature aging in children and young adults with progeria and processing-deficient progeroid laminopathies.

Zokinvy is a first-in-class disease-modifying agent that has demonstrated a statistically significant survival benefit in children and young adults with progeria. In patients with progeria, Zokinvy reduced the incidence of mortality by 60% ($p=0.0064$) and increased average survival time by 2.5 years. The most commonly reported adverse reactions were gastrointestinal (vomiting, diarrhea, nausea), and most were mild or moderate (Grade 1 or 2) in severity. Many progeria patients have received continuous Zokinvy therapy for more than 10 years.

Eiger licensed exclusive worldwide rights to lonafarnib from Merck, known as MSD outside of the United States and Canada. Merck will not receive any milestone payments for the development of lonafarnib for the treatment of progeria and has waived royalty obligations from Eiger for a specified quantity of lonafarnib.



For more information including prescribing information for Zokinvy in the U.S. please go to www.zokinvy.com. Please click [here](#) for the full U.S. important safety information. Eiger has filed a marketing authorization application with the European Medicines Agency and expects a CHMP opinion in Q2 2022. Zokinvy is not approved for any indication in Japan.

About Eiger

Eiger is a commercial-stage biopharmaceutical company focused on the development of innovative therapies to treat and cure hepatitis delta virus (HDV) and other serious diseases. The Eiger HDV platform includes two first-in-class therapies in Phase 3 that target critical host processes involved in viral replication. Eiger is also developing peginterferon lambda as a therapeutic for COVID-19 and is planning to submit an emergency use authorization application to FDA based on positive results from the investigator sponsored Phase 3 *TOGETHER* study.

All five Eiger rare disease programs have been granted FDA Breakthrough Therapy designation: lonafarnib and peginterferon lambda for HDV, Zokinvy for progeria, and avexitide for both congenital hyperinsulinism and post-bariatric hypoglycemia.

For additional information about Eiger and its clinical programs, please visit www.eigerbio.com.

About AnGes Inc.

AnGes, Inc., a biopharmaceutical company focuses on the development of gene-based medicines. In September 2019, AnGes commenced the commercialization in Japan of Collategene[®] (Hepatocyte Growth Factor, HGF, plasmid gene therapy) for the treatment of Chronic arterial occlusive disease with lower limb ulcers. Collategene[®] is the world's first marketed drug using Plasmid DNA. AnGes is currently focusing on the development of DNA vaccines for COVID-19 and Hypertension, Tie2 tyrosine kinase receptor agonist for COVID-19 treatment, and NF- κ B decoy oligonucleotide and Chimera decoy oligonucleotide as next generation product for Chronic Discogenic Lumbar Back Pain. Furthermore, AnGes acquired EmendoBio to expand its capabilities in Genome Editing Technologies in December 2020. For more information, visit <https://www.anges.co.jp/en/>.

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