Eiger Initiates Rolling Submission of New Drug Application (NDA) with FDA for Lonafarnib for Treatment of Progeria and Progeroid Laminopathies

- **Rolling NDA Submission Planned for Completion in First Quarter 2020**
- **Marketing Authorization Application (MAA) to EMA Planned in First Quarter 2020**

PALO ALTO, Calif., December 16, 2019 — Eiger BioPharmaceuticals, Inc. (Nasdaq:EIGR), a late stage clinical biopharmaceutical company focused on the development and commercialization of targeted therapies for serious rare and ultra-rare diseases, today announced that it has initiated submission of a New Drug Application (NDA) for Lonafarnib for the treatment of Hutchinson-Gilford Progeria Syndrome (HGPS or Progeria) and Progeroid Laminopathies to the U.S. Food & Drug Administration (FDA) under the Rolling Review Process. Lonafarnib, an oral farnesyltransferase inhibitor (FTI), has demonstrated extended survival in children and young adults with Progeria, an ultra-rare and fatal disease that causes premature aging in children. Without treatment, children with Progeria die of heart disease at an average age of 14.5 years.

Lonafarnib has been granted Orphan Drug Designation for Progeria and Progeroid Laminopathies by the FDA and EMA and Breakthrough Therapy Designation and Rare Pediatric Disease Designation by the FDA. The FDA agreed that the Lonafarnib submission could be a rolling NDA, allowing completed portions of an NDA to be submitted and reviewed by the FDA on an ongoing basis. Eiger plans to complete the NDA rolling submission in first quarter of 2020.

“Our discussions with FDA have been constructive and collaborative at each step, and agency agreement to accept our NDA on a rolling basis marks an important milestone to begin the regulatory review process for Lonafarnib in Progeria and Progeroid Laminopathies,” said David Cory, Eiger President and CEO. “Eiger is committed to making a meaningful difference in the lives of patients with Progeria and Progeroid Laminopathies and we are thrilled to submit our first NDA for these children. We look forward to working closely with the FDA as we complete the rolling NDA process.”

About Progeria

Progeria, also known as Hutchinson-Gilford Progeria Syndrome (HGPS), is a rare and fatal genetic condition of accelerated aging in children. Progeria is caused by a point mutation in the *LMNA* gene, which encodes the lamin A protein, yielding the farnesylated aberrant protein, progerin. Lamin A protein is part of the structural scaffolding that holds the nucleus together. Children with Progeria die of the same heart disease that affects millions of normally aging adults (arteriosclerosis), but at 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. It is estimated that 400 children worldwide have Progeria.

About Progeroid Laminopathies

Progeroid Laminopathies are genetic conditions of accelerated aging caused by a constellation of mutations in the lamin A 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. Collectively, worldwide prevalence of Progeroid Laminopathies is likely greater than Progeria.

About Lonafarnib

Lonafarnib is a well-characterized, late-stage, orally active inhibitor of farnesyltransferase, an enzyme involved in modification of proteins through a process called prenylation. Progerin is a farnesylated aberrant protein that researchers believe cannot be cleaved, resulting in tight association with the nuclear envelope, which leads to changes in nuclear envelope morphology and subsequent cellular damage.

Lonafarnib blocks the farnesylation of progerin and has been dosed in over 80 children with Progeria at Boston Children's Hospital in Phase 1/2 and Phase 2 studies funded by The Progeria Research Foundation (www.progeriaresearch.org). In patients with HGPS, lonafarnib monotherapy was associated with a lower mortality rate after 2.2 years of follow-up compared with no treatment (3.7% vs 33.3%, respectively) with a hazard ratio of 0.12 or a reduction in risk of mortality of 88%.

Lonafarnib has been granted Orphan Drug Designation for Progeria by the FDA and EMA and Breakthrough Therapy Designation and Rare Pediatric Disease Designation by the FDA. Lonafarnib is not approved for any indication, and is licensed by Eiger from Merck Sharp & Dohme Corp.

About The Progeria Research Foundation

The Progeria Research Foundation was established in 1999 by the family of Sam Berns, a child with Progeria. Within four years of its founding, the PRF Genetics Consortium, led by Francis Collins, MD, PhD, discovered the Progeria gene. PRF has also been the driving force behind studies to evaluate lonafarnib as a potential treatment for Progeria and supports scientists who conduct Progeria research. Today, PRF is the only non-profit organization in the world solely dedicated to finding treatments and the cure for Progeria and its age-related conditions, including heart disease. For more information, please visit www.progeriaresearch.org.

About Eiger

Eiger is a late stage biopharmaceutical company focused on the development and commercialization of a pipeline of first-in-class, well-characterized drugs for serious rare and

ultra-rare diseases for patients with high unmet medical needs and for which no approved therapies exist.

The company's lead program is in Phase 3, developing lonafarnib, a first-in-class prenylation inhibitor for the treatment of Hepatitis Delta Virus (HDV) infection. The company is rapidly advancing peginterferon lambda, a first-in-class interferon, toward registration for the treatment of HDV. Eiger has initiated a rolling NDA submission for lonafarnib to treat Hutchinson-Gilford Progeria Syndrome (HGPS or Progeria) and Progeroid Laminopathies with plans to complete NDA submission followed by an MAA submission in the first quarter of 2020. For additional information about Eiger and its clinical programs, please visit www.eigerbio.com.

Note Regarding Forward-Looking Statements

This press release contains "forward-looking" statements that involve substantial risks and uncertainties. All statements other than statements of historical facts, including statements regarding our future financial condition, timing for and outcomes of clinical results, business strategy and plans and objectives for future operations, are forward-looking statements. These forward-looking statements include terminology such as "believe," "will," "may," "estimate," "continue," "anticipate," "contemplate," "intend," "target," "project," "should," "plan," "expect," "predict," "could," "potentially" or the negative of these terms. Forward-looking statements are our current statements regarding our intentions, beliefs, projections, outlook, analyses or current expectations concerning, among other things, our ongoing and planned clinical development, including planned completion of a rolling NDA submission in first quarter of 2020 followed by submission of an MAA in first quarter 2020 for Progeria and Progeroid Laminopathies; our progression and enrollment of our Phase 3 D-LIVR study in HDV; our planned advancement of lambda and lonafarnib boosted with ritonavir for HDV; our ability to transition into a commercial stage biopharmaceutical company; our ability to finance the continued advancement of our development pipeline products; and the potential for success of any of our product candidates. These statements concern product candidates that have not yet been approved for marketing by the U.S. Food and Drug Administration (FDA). No representation is made as to their safety or effectiveness for the purposes for which they are being investigated.

Various important factors could cause actual results or events to differ materially from the forward-looking statements that Eiger makes, including the risks described in the "Risk Factors" sections in the Quarterly Report on Form 10-Q for the quarter ended September 30, 2019 and Eiger's subsequent filings with the SEC. Eiger does not assume any obligation to update any forward-looking statements, except as required by law.

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