Kyowa Hakko Kirin and Kyowa Kirin International Announce Burosumab Data Presentations at ASBMR 2017 Annual Meeting

Tokyo, Japan and London, UK — **August 24, 2017** — Kyowa Hakko Kirin Co., Ltd. (Kyowa Hakko Kirin) and Kyowa Kirin International PLC (Kyowa Kirin International), a wholly owned subsidiary of Kyowa Hakko Kirin, today announced upcoming presentations of data highlighting burosumab for the treatment of X-linked hypophosphatemia (XLH) and tumor-induced osteomalacia (TIO) at the American Society for Bone and Mineral Research (ASBMR) 2017 Annual Meeting taking place September 8-11 in Denver, Colorado.

Kyowa Hakko Kirin, Kyowa Kirin International and Ultragenyx Pharmaceutical Inc. have been collaborating in development and commercialization of burosumab globally based on the collaboration and licence agreement between Kyowa Hakko Kirin and Ultragenyx.

Two oral presentations will highlight a late breaking abstract on 24 week data from the adult Phase 3 study (n=134) and 64 week data from the pediatric Phase 2 study (n=52) in XLH patients

Oral Presentation #LB-1159: A Phase 3 Randomized, 24 Week, Double-Blind, Placebo-Controlled Study Evaluating the Efficacy of Burosumab, an Anti-FGF23 Antibody, in Adults with X-Linked Hypophosphatemia (XLH)

- Monday, September 11, 11:15 AM 11:25 AM MDT
- Mile High Ballroom, Colorado Convention Center

Oral Presentation #1154: Burosumab (KRN23), a Fully Human Anti-FGF23 Monoclonal Antibody for X-linked Hypophosphatemia (XLH): Final 64-Week Results of a Randomized, Open-label, Phase 2 Study of 52 Children

- Monday, September 11, 09:45 AM 10:00 AM MDT
- Mile High Ballroom, Colorado Convention Center

Three poster presentations will highlight additional burosumab data including functional patient reported outcomes from the pediatric Phase 2 study, Phase 2 study in Tumor Induced Osteomalacia (TIO), and the Phase 2 study in pediatric patients under 5 years old

Poster #FR0331 (shown twice): Effects of Burosumab (KRN23), a Fully Human Anti-FGF23 Monoclonal Antibody, on Functional Outcomes in Children with X-linked Hypophosphatemia (XLH): Final Results from a Randomized, 64-week, Open-label Phase 2 Study

- Friday, September 8, 05:00 PM 07:00 PM MDT and Saturday, September 9, 12:30 PM 02:30 PM MDT
- ASBMR Discovery Hall Exhibit Hall A & B1, Colorado Convention Center

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Poster #SU0325: Effects of Burosumab (KRN23), a Human Monoclonal Antibody to FGF23, in Patients with Tumor-Induced Osteomalacia (TIO) or Epidermal Nevus Syndrome (ENS)

- Sunday, September 10, 12:30 PM 02:30 PM MDT
- ASBMR Discovery Hall Exhibit Hall A, Colorado Convention Center

Poster #MO0695: The Effects of Burosumab (KRN23), a Fully Human Anti-FGF23 Monoclonal Antibody, on Phosphate Metabolism and Rickets in 1 to 4-Year-Old Children with X-linked Hypophosphatemia (XLH)

- Monday, September 11, 12:00 PM 02:00 PM MDT
- ASBMR Discovery Hall Exhibit Hall A & B1, Colorado Convention Center

About Burosumab (KRN23)

Burosumab is an investigational recombinant fully human monoclonal IgG1 antibody, discovered by Kyowa Hakko Kirin, against the phosphaturic hormone fibroblast growth factor 23 (FGF23). FGF23 is a hormone that reduces serum levels of phosphorus and active vitamin D by regulating phosphate excretion and active vitamin D production by the kidney. Burosumab is being developed by Ultragenyx and Kyowa Hakko Kirin to treat XLH and tumor-induced osteomalacia (TIO), diseases characterized by excess levels of FGF23. Phosphate wasting in XLH and TIO is caused by excessive levels and activity of FGF23. Burosumab is designed to bind to and thereby inhibit the biological activity of FGF23. By blocking excess FGF23 in patients with XLH and TIO, burosumab is intended to increase phosphate reabsorption from the kidney and increase the production of vitamin D, which enhances intestinal absorption of phosphate and calcium.

A clinical program studying burosumab in adults and pediatric patients with XLH is ongoing. Burosumab is also being developed for TIO, a disease characterized by typically benign tumors that produce excess levels of FGF23, which can lead to severe osteomalacia, fractures, bone and muscle pain, and muscle weakness.

About Kyowa Kirin

Kyowa Hakko Kirin Co., Ltd. is a research-based life sciences company, with special strengths in biotechnologies. In the core therapeutic areas of oncology, nephrology and immunology/allergy, Kyowa Hakko Kirin leverages leading-edge biotechnologies centered on antibody technologies, to continually discover innovative new drugs and to develop and market those drugs world-wide. In this way, the company is working to realize its vision of becoming a Japan-based global specialty pharmaceutical company that contributes to the health and wellbeing of people around the world.

Kyowa Kirin International PLC is a wholly owned subsidiary of Kyowa Hakko Kirin and is a rapidly growing specialty pharmaceutical company engaged in the development and

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commercialisation of prescription medicines for the treatment of unmet therapeutic needs in Europe and the United States. Kyowa Kirin International is headquartered in Scotland.

You can learn more about the business at: <u>www.kyowa-kirin.com</u>.