

**FOR IMMEDIATE RELEASE**

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## **Collaboration with Ultragenyx to Develop and Commercialize KRN23 for X-linked Hypophosphatemia**

Tokyo, Japan, September 4, 2013 — Kyowa Hakko Kirin Co., Ltd. (President and CEO: Nobuo Hanai; “Kyowa Hakko Kirin”), today announced that it has entered into a collaboration and license agreement with Ultragenyx Pharmaceutical Inc. (“Ultragenyx”), to develop and commercialize KRN23, a recombinant fully human monoclonal IgG1 antibody, intended to treat X-linked hypophosphatemia (XLH). Kyowa Hakko Kirin is currently completing a Phase 1/2 study in adults with XLH in the US and Canada. Initiation of a pediatric XLH program is planned for 2014.

XLH is a rare metabolic bone disorder caused by excessive loss of phosphate in the urine leading to severe hypophosphatemia and poor bone growth and mineralization. XLH patients have low serum phosphate levels due to high levels of FGF23, a hormone that represses the reabsorption of phosphate from the urine. KRN23 is intended to bind to and render FGF23 inactive, leading to an increase in kidney tubular absorption of phosphate and increased serum phosphate levels. KRN23 is potentially the first specific treatment that addresses the underlying problem of XLH.

Ultragenyx is a privately held, clinical-stage biotechnology company committed to bringing to market life-transforming therapeutics for patients with rare and ultra-rare metabolic genetic diseases. Under the terms of the agreement, Kyowa Hakko Kirin and Ultragenyx will collaborate on the development of KRN23 for the US, Canada and European Union (EU), with Ultragenyx leading development efforts in the XLH indication and the parties sharing development costs. In the US and Canada, Kyowa Hakko Kirin and Ultragenyx will share commercial responsibilities and profits. Kyowa Hakko Kirin will commercialize KRN23 in the EU. Ultragenyx will develop and commercialize the product in Mexico, Central and South America.

KRN23 was found and is developed by Kyowa Hakko Kirin as the only product targeting FGF23 in the world and is strategically important for Kyowa Hakko Kirin to expand its business in global markets. Kyowa Hakko Kirin believes that the collaboration accelerates development of KRN23 and maximizes its values in global markets, by utilizing Ultragenyx's expertise in the development of novel therapeutics for rare genetic diseases.

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#### **About Ultragenyx Pharmaceutical Inc.**

Ultragenyx is a privately held, clinical-stage biotechnology company committed to bringing to market life-transforming therapeutics for patients with rare and ultra-rare metabolic genetic diseases. Founded in 2010, the company is rapidly building a diverse portfolio of product candidates with the potential to address diseases for which the unmet medical need is high, the biology for treatment is clear, and for which there are no effective treatments.

The company is led by a management team experienced in the development and commercialization of rare disease therapeutics. Ultragenyx's strategy is predicated upon time and cost-efficient drug development, with the goal of delivering safe and effective therapies to patients with the utmost urgency.

For more information on Ultragenyx, please visit the company's website at [www.ultragenyx.com](http://www.ultragenyx.com).

For further information please access: [http://www.kyowa-kirin.com/news\\_releases/index.html](http://www.kyowa-kirin.com/news_releases/index.html)