May 8, 2017

# Horizon Pharma plc Announces Agreement to Acquire River Vision Development Corp. and Teprotumumab (RV001), a Biologic in Late-Stage Development for Rare Eye Disease

- *Pivotal Trial of Teprotumumab in Thyroid Eye Disease (TED) Expected to Begin in Second Half of 2017* -

- *Teprotumumab Phase 2 Trial Results Recently Published in The New England Journal of Medicine* -

DUBLIN, Ireland, May 08, 2017 (GLOBE NEWSWIRE) -- Horizon Pharma plc (NASDAQ:HZNP), a biopharmaceutical company focused on improving patients' lives by identifying, developing, acquiring and commercializing differentiated and accessible medicines that address unmet medical needs, today announced that it has agreed to acquire River Vision Development Corp. ("River Vision") and its development-stage medicine teprotumumab (RV001), a fully human monoclonal antibody (mAb) currently in development for Thyroid Eye Disease (TED), a rare, autoimmune inflammatory disorder.

Under the terms of the agreement, Horizon Pharma will acquire all outstanding equity of River Vision for an upfront cash payment of $145 million, plus potential future milestone and earn-out payments contingent on the satisfaction of certain regulatory milestones and sales thresholds. The acquisition is expected to close today.

"This acquisition is an important step in our strategy of pursuing and acquiring development-stage medicines targeting rare diseases," said Timothy P. Walbert, chairman, president and chief executive officer, Horizon Pharma plc. "With no approved medicines to treat Thyroid Eye Disease, there is a significant unmet treatment need among the approximately 10,000 patients in the United States with moderate to severe disease and we look forward to beginning the pivotal study with teprotumumab in the second half of this year."

Horizon anticipates a potential peak annual sales opportunity for teprotumumab, if approved, in excess of $250 million in the United States.

***About Teprotumumab (RV001)***
Teprotumumab is a human mAb inhibitor of insulin-like growth factor type 1 receptor (IGF-1R), which has received Orphan Drug, Fast Track and Breakthrough Therapy designations from the U.S. Food and Drug Administration (FDA).

On May 4, 2017, *The New England Journal of Medicine* published Phase 2 study results evaluating the efficacy and safety of teprotumumab in patients with recent onset, moderate-to-severe TED. In the randomized double-blind, placebo controlled study, 88 patients were assigned to receive teprotumumab or placebo in eight intravenous infusions, 10mg/kg for their first infusion followed by 20mg/kg for the remaining seven infusions, every three weeks during the six-month treatment course. The primary endpoint was response in the study eye defined as a reduction in clinical activity score of ≥ 2 points and reduction of proptosis, which is protrusion of the eyeball from the socket of ≥ 2 mm at week 24.

In the intent-to-treat population, 29/42 (69%) patients receiving teprotumumab and 9/45 (20%) patients receiving placebo were responders at week 24 ($p<0.001$). Therapeutic effects were rapid with responder rates of 46% for patients treated with teprotumumab and 5% for patients treated with placebo at week six ($p < 0.001$). Treatment with teprotumumab was well tolerated with the majority of adverse events being mild. The only treatment-related adverse event was hyperglycemia in diabetic patients, which was controlled by adjusting diabetes medication.

***About Thyroid Eye Disease***
Thyroid eye disease (TED) is an eye condition in which the eye muscles and fatty tissue behind the eye become inflamed. This can cause the eyes to be pushed forward ('staring' or 'bulging' eyes) and the eyes and eyelids to become swollen and red. In some cases there is swelling and stiffness of the muscles that move the eyes so that the eyes are no longer in line with each other and/or the lids are unable to close.

***About Horizon Pharma plc***
Horizon Pharma plc is a biopharmaceutical company focused on improving patients' lives by identifying, developing, acquiring and commercializing differentiated and accessible medicines that address unmet medical needs. The Company markets 11 medicines through its orphan, rheumatology and primary care business units. For more information, please visit www.horizonpharma.com. Follow @HZNPplc on Twitter or view careers on our LinkedIn page.

***About River Vision Development Corp.***
River Vision Development Corp. was founded by Narrow River Management, LP (Narrow River) for the sole purpose of developing teprotumumab, which was developed by Genmab and subsequently licensed from Roche, for TED and other potential indications.

***Forward-Looking Statements***
This press release contains forward-looking statements, including, but not limited to, statements related to the anticipated acquisition of River Vision Development Corp. and the benefits thereof, Horizon Pharma's strategy, plans, objectives, expectations (financial or otherwise) and intentions, including with respect to teprotumumab, the timing of a planned Phase 3 clinical trial of teprotumumab and other statements that are not historical facts. These forward-looking statements are based on Horizon Pharma's current expectations and inherently involve significant risks and uncertainties. Actual results and the timing of events could differ materially from those anticipated in such forward looking statements as a result of these risks and uncertainties, which include, without limitation, risks related to Horizon's ability to complete the transaction on the proposed terms; risks associated with clinical development, such as the risk that clinical trials are not initiated or completed on time and the fact that prior clinical results may not predict the outcome of future trials; risks associated with acquisitions, such as the risk that the businesses will not be integrated successfully, that such integration may be more difficult, time-consuming or costly than expected or that the expected benefits of the transaction will not occur; risks related to future opportunities and plans for River Vision and teprotumumab, including uncertainty related to the future development of teprotumumab and whether Horizon Pharma can ultimately obtain regulatory approval to market teprotumumab; disruption from the proposed transaction, making it more difficult to conduct business as usual; and the calculations of, and factors that may impact the calculations of, the acquisition price in connection with the proposed acquisition and the allocation of such acquisition price to the net assets acquired in accordance with applicable accounting rules and methodologies, as well as other risks related to Horizon's business detailed from time-to-time under the caption "Risk Factors" and elsewhere in Horizon Pharma's SEC filings and reports, including in its Annual Report on Form 10-K for the year ended December 31, 2016 and subsequent quarterly reports on Form 10-Q. Horizon Pharma undertakes no duty or obligation to update any forward-looking statements contained in this press release as a result of new information, future events or changes in its expectations.

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