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Xencor Presents Interim Data from an Ongoing, Open-label, Phase 2 Study of XmAb®5871 in IgG4-Related Disease at EULAR 2017

- 14 of 15 patients (93%) achieved a response to therapy, 12 of them within 2 weeks of first dose -
- 6 patients attained disease remission (an IgG4-RD Responder Index of 0) -
- Every other week intravenous administration of XmAb5871 in patients with active IgG4-RD has been well tolerated -

MONROVIA, Calif., June 16, 2017 /PRNewswire/ -- Xencor, Inc. (NASDAQ: XNCR), a clinical-stage biopharmaceutical company developing engineered monoclonal antibodies for the treatment of autoimmune diseases, asthma and allergic diseases and cancer, today announced interim data from an ongoing, open-label, pilot Phase 2 study of XmAb®5871 in patients with active IgG4-Related Disease (IgG4-RD). Data show that 93% of patients achieved a response to therapy, 12 of them within two weeks of their first dose. The data were presented today by John H. Stone, M.D., MPH, director of rheumatology at Massachusetts General Hospital, at the Annual European Congress of Rheumatology (EULAR 2017).

"We continue to be very encouraged by the rapid initial response in IgG4-RD disease activity observed in this interim data set, in which now 14 of 15 patients evaluated for IgG4-RD Responder Index (RI) achieved a response, with initial responses typically occurring after the first dose and deepening over time," said Paul Foster, M.D., chief medical officer of Xencor. "We expect to report final study results for these 15 patients in 2017."

"The rapid clinical responses in many patients and the continued decline in the inflammatory response indicate that XmAb5871 is clearly a promising therapy for IgG4-RD," said Dr. John H. Stone, the principal investigator of the study. "We have learned a lot about how to study this disease in the context of this small trial."

As of a data cutoff of April 18, 2017, all 15 planned patients with active IgG4-RD have been enrolled and dosed with XmAb5871 (median number of infusions = 12, range 5-12). Patients had a median IgG4-RD RI of 12 (range 2-30) with a median of five organs involved (range 1-10) at the time of study entry. Organ site involvement occurring at a frequency of greater than or equal to 45% included lymph nodes, submandibular glands, parotid glands and lacrimal glands. In addition, 40% of patients suffered from constitutional symptoms at time of study entry.

Every other week intravenous administration of XmAb5871 has been well tolerated. Adverse events (AEs) were consistent with that previously reported, with all XmAb5871-related AEs graded as mild or moderate and no AE reported in more than 2 patients. No severe AEs deemed related to XmAb5871 were reported. One patient discontinued the study as the result of an AE of a moderate hypersensitivity reaction, as previously reported in November 2016.

Interim Efficacy Data:

As of April 18, 2017, 10 patients have completed the study, 3 of whom discontinued early as reported previously in November 2016, and 5 patients are ongoing. Fourteen of the 15 patients (93%) dosed with XmAb5871 have had a response to XmAb5871 therapy of greater than or equal to a two-point reduction in the IgG4-RD RI (protocol defined response), 12 of them within two weeks of the first dose. At two weeks following the last dose, five patients had an IgG4-RD RI of 0 and were on no corticosteroid therapy between months 2-6 (protocol definition of remission). In addition, a sixth patient achieved remission in the post-therapy follow-up period. All five of the patients that either entered the study on corticosteroids or that were administered corticosteroids at the beginning of the study have been able to taper and discontinue corticosteroids within 2 months of the start of the study.

The early discontinuations included the patient with early study termination due to an AE, one patient that had a response

to therapy (IgG4-RD RI reduction of six points), but lost response following the sixth infusion, and one patient that had no response to therapy as defined by a greater than or equal to two-point decrease in the IgG4-RD RI. This latter patient had an atypical presentation of larynx involvement as the only organ involved. The patient discontinued the study after six infusions. Neither of the two patients that discontinued early due to loss of response or lack of response responded to subsequent rituximab treatment.

Interim Biomarker Data:

Circulating $CD19^+$ plasmablast levels were elevated in the majority of patients at baseline and levels were reduced quickly following XmAb5871 treatment. XmAb5871 also reduced the numbers of circulating $SLAMF7^+$ $CD4^+$ cytotoxic T lymphocytes, an immune cell population potentially important in IgG4-RD, in those patients with measurable levels at baseline.

The poster presentation is available on the 'Investors' page of Xencor's website under 'Events and Presentations' at www.xencor.com.

About XmAb®5871-03

XmAb5871-03 is an open-label, single-arm study of up to 15 patients with histopathologically proven IgG4-RD with active disease as defined by disease activity in one or more organ systems and an IgG4-RD RI of greater than or equal to three. Participants will receive XmAb5871 by intravenous infusion every other week for up to a total of 12 infusions. Primary and secondary objectives are to evaluate the effect of every other week intravenous administration of XmAb5871 on the IgG4-RD RI in patients with active IgG4-RD and to evaluate the safety, tolerability, pharmacokinetics and immunogenicity of XmAb5871 in patients with active IgG4-RD over an up to six-month period. The primary endpoint of the completed study will be the proportion of patients on Day 169 with an improvement of disease activity score as defined by a decrease of IgG4-RD RI of greater than or equal to two points from the Day 1 pre-dose disease activity score.

About XmAb®5871

XmAb®5871 is a first-in-class monoclonal antibody that targets CD19 with its variable domain and that uses Xencor's XmAb immune inhibitor Fc domain to target FcγRIIb, a receptor that inhibits B-cell function. XmAb5871 is the first drug candidate that Xencor is aware of that targets FcγRIIb inhibition. Xencor has demonstrated in multiple animal models and in initial human clinical trials that XmAb5871 inhibits B-cell function without destroying these important immune cells, and demonstrated promising treatment effect in patients with rheumatoid arthritis, as well as ex vivo results showing inhibition of systemic lupus erythematosus (SLE) patient B-cell activation and humoral immunity.

Complete data results from a Phase 1b/2a study of XmAb5871 in patients with rheumatoid arthritis were presented at the American College of Rheumatology 2015 Annual Meeting as well as at the EULAR 2015 Annual Meeting. Ex vivo studies of SLE patient B cells were published in Journal of Immunology, 2011, 186(7):4223.

About IgG4-Related Disease

IgG4-Related Disease (IgG4-RD) is a rare fibro-inflammatory autoimmune disorder that is estimated to impact up to 40,000 patients in the United States. IgG4-RD affects multiple organ systems and is characterized by a distinct microscopic appearance of diseased organs, including the presence of IgG4-positive plasmablast cells. This objective diagnostic criterion is atypical for autoimmune diseases and offers advantages for accurately identifying patients. There are currently no approved therapies for this newly recognized disorder and corticosteroids are the current standard of care. John H. Stone, M.D, MPH, director, clinical rheumatology at Massachusetts General Hospital has developed and is validating the IgG4-RD Responder Index (RI), a proposed instrument to assess disease activity.

About Xencor's XmAb® Immune Inhibitor Technology

FcγRIIb (IIb), also called CD32b, is a receptor for Fc domains on B cells and other immune cells. When engaged, the IIb receptor blocks immune activation pathways and traffics bound soluble antigens out of circulation. Xencor has discovered a series of Fc domain variants with up to a 400-fold increase in binding affinity to FcγRIIb derived from just two amino acid changes. These XmAb® Immune Inhibitor Fc domains greatly heighten the properties of IIb receptor engagement and have potential as building blocks for drug candidates in autoimmune, allergic and inflammatory diseases.

About Xencor, Inc.

Xencor is a clinical-stage biopharmaceutical company developing engineered monoclonal antibodies for the treatment of autoimmune diseases, asthma and allergic diseases and cancer. Currently, 11 candidates engineered with Xencor's XmAb®

technology are in clinical development internally and with partners. Xencor's internal programs include: XmAb®5871 in Phase 2 development for the treatment of IgG4-Related Disease, and also for the treatment of Systemic Lupus Erythematosus; XmAb®7195 in Phase 1 development for the treatment of asthma and allergic diseases; XmAb®14045 in Phase 1 development for acute myeloid leukemia; XmAb®13676 in Phase 1 development for B-cell malignancies; XmAb®18087 in pre-clinical development for the treatment of neuroendocrine tumors; and XmAb®20717 in pre-clinical development for the treatment of multiple cancers. Xencor's XmAb antibody engineering technology enables small changes to the structure of monoclonal antibodies resulting in new mechanisms of therapeutic action. Xencor partners include Novartis, Amgen, MorphoSys, Merck, CSL/Janssen, Alexion and Boehringer Ingelheim. For more information, please visit www.xencor.com.

Forward Looking Statements:

Statements contained in this press release regarding matters that are not historical facts are forward-looking statements within the meaning of applicable securities laws, including the quotation from Xencor's officer and any expectations relating to its business, research and development programs, including ongoing clinical trials of XmAb5871, and the immune inhibitory Fc domain technology, partnering efforts or its capital requirements. Such statements involve known and unknown risks, uncertainties and other factors that may cause actual results, performance or achievements, including those of the complete clinical trial of XmAb5871, and the timing of events to be materially different from those implied by such statements, and therefore these statements should not be read as guarantees of future performance or results. Such risks include, without limitation, the risks associated with the process of discovering, developing, manufacturing and commercializing drugs that are safe and effective for use as human therapeutics and other risks described in Xencor's public securities filings. All forward-looking statements are based on Xencor's current information and belief as well as assumptions made by Xencor. Readers are cautioned not to place undue reliance on such statements and Xencor disclaims any intention or obligation to update or revise any forward-looking statements, whether as a result of new information, future events or otherwise.