



November 13, 2016

Xencor Presents Preliminary Data from an Ongoing, Open-label, Phase 2 Study of XmAb[®]5871 in IgG4-Related Disease (IgG4-RD) at the American College of Rheumatology 2016 Annual Meeting

- **9 of 11 patients (82%) achieved an initial response to therapy within 2 weeks of first dose --**
- **5 patients have 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 -**
- **Xencor management to host conference call today at 6:00 p.m. EST -**

MONROVIA, Calif., Nov. 13, 2016 /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 preliminary data from XmAb5871-03, an ongoing, open-label, pilot Phase 2 study of XmAb5871 in patients with active IgG4- RD. Data show that 82% of patients achieved an initial response to therapy within two weeks of their first dose. The data are being presented by John H. Stone, M.D., MPH, director of rheumatology at Massachusetts General Hospital, at the American College of Rheumatology (ACR) 2016 Annual Meeting in the Miscellaneous Rheumatic and Inflammatory Disease session on Sunday, November 13, 2016 (today) at 2:45 p.m. EST.



"We are very encouraged by the rapid initial response in IgG4-RD disease activity observed in this preliminary data set, in which nine of the 11 patients evaluated for IgG4-RD Responder Index (RI) post-treatment achieved a response within two weeks of their first dose, with responses typically deepening over time," said Paul Foster, M.D., chief medical officer of Xencor. "We expect to report complete study results in 2017."

"I am impressed by the steady 'dialing down' of the inflammatory response that XmAb5871 seems to exert in IgG4-RD and believe it is a promising potential therapy," said Dr. John H. Stone, the principal investigator of the study. "We are learning a lot about how to study this disease in the context of this small trial."

As of a data cutoff of October 31, 2016, 12 patients with active IgG4-RD have been enrolled and dosed with XmAb5871 (median number of infusions = 7, range 1-12). Patients had a median IgG4-RD RI of 10 (range 2-30) with a median of four organs involved (range 1-10) at the time of study entry. Organ site involvement occurring at a frequency of greater than or equal to 50% included lymph nodes, submandibular glands, parotid glands and lacrimal glands.

Preliminary Safety Data:

Every other week intravenous administration of XmAb5871 has been well tolerated. As of October 31, 2016, no serious adverse events (AEs) have been reported. Treatment related AEs have occurred in five patients (42%). Treatment-related AEs that occurred in more than one patient were abdominal pain/discomfort in three patients (25%), occurring as part of Grade 1 (mild) infusion-related gastrointestinal symptoms (nausea and/or vomiting and/or diarrhea) during the first infusion, and Grade 1 (mild) headache in two patients (16.7%). One patient discontinued the study as the result of an AE. The patient developed a Grade 2 (moderate) hypersensitivity reaction with rash and arthritis, commonly referred to as serum sickness, following the fifth infusion. The event resolved quickly without the need for medical management. This patient was subsequently found to have developed anti-drug antibodies.

Preliminary Efficacy Data:

Eleven of the 12 patients dosed with XmAb5871 have had at least one IgG4-RD RI performed following dosing as of the data cutoff date. Nine of 11 patients (82%) have had an initial response to XmAb5871 therapy of at least a three-point

reduction in the IgG4-RD RI within two weeks of the first dose. Five patients attained disease remission (an IgG4-RD RI of 0) during the study. Two patients entering the study on corticosteroids have been able to taper and discontinue steroid use during the study.

In addition to the patient with early study termination due to an AE, two other patients have discontinued treatment prior to receipt of all 12 planned infusions. One patient had a response to therapy (IgG4-RD RI reduction of six points), but lost response following the sixth infusion, at which point this patient discontinued treatment. One patient had no response to therapy as defined by a greater than or equal to two-point decrease in the IgG4-RD RI. This patient had an atypical presentation of larynx involvement as the only organ involved. The patient discontinued the study after six infusions. Neither of these two patients have responded to subsequent rituximab treatment.

The slide presentation will be available at 3:00 p.m. Eastern Time today on the 'Investors' page of Xencor's website under 'Events and Presentations' at www.xencor.com.

Conference Call Information

Xencor management will host a conference call and webcast today at 6:00 p.m. Eastern Time to discuss data presented today. The live call may be accessed by dialing (877) 359-9508 for domestic callers or (224) 357-2393 for international callers, and providing the conference ID number 16658995. A live webcast of the conference call will be available online from the investors section of the Xencor website at www.xencor.com. The webcast will be archived on the Company's website for 30 days.

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, 10 candidates engineered with Xencor's XmAb[®]

technology are in clinical development internally and with partners. Xencor's internal programs include: XmAb5871 in Phase 2 development for the treatment of IgG4-Related Disease, and also for the treatment of Systemic Lupus Erythematosus; XmAb7195 in Phase 1 development for the treatment of asthma and allergic diseases; XmAb14045 in Phase 1 development for acute myeloid leukemia; and XmAb13676 for B-cell malignancies and XmAb18087 for the treatment of neuroendocrine tumors, both in pre-clinical development. 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, Novo Nordisk 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.

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To view the original version on PR Newswire, visit:<http://www.prnewswire.com/news-releases/xencor-presents-preliminary-data-from-an-ongoing-open-label-phase-2-study-of-xmab5871-in-igg4-related-disease-igg4-rd-at-the-american-college-of-rheumatology-2016-annual-meeting-300361524.html>

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