**argenx Announces Publication of Translational Data of Efgartigimod in Autoimmune Skin Blistering Diseases**

- *Translational data from Phase 2 study of efgartigimod in pemphigus further demonstrate argenx's scientific leadership in FcRn biology, providing new insights into pathophysiology of autoimmune skin blistering diseases and potential role of FcRn blockade*

- *Data to be included in symposium presentation at Society for Investigative Dermatology (SID) Annual Meeting being held today, May 19, 2022, 7:30am PT*

**May 19, 2022**

**Breda, the Netherlands** – argenx SE (Euronext & Nasdaq: ARGX), a global immunology company committed to improving the lives of people suffering from severe autoimmune diseases, today announced the publication of novel translational data from the open-label Phase 2 study of efgartigimod for the treatment of pemphigus that further support the potential role of FcRn blockade and potential of efgartigimod in autoimmune skin blistering disorders. The translational data, **"FcRn Antagonism Leads to a Decrease of Desmoglein-Specific B Cells: Secondary Analysis of a Phase 2 Study of Efgartigimod in Pemphigus Vulgaris and Pemphigus Foliaceus**" were published in the journal *Frontiers of Immunology*.

The novel translational data, along with previously published translational data in *Cells*, will also be presented in a symposium today at the Society for Investigative Dermatology (SID) Annual Meeting at 7:30am PT. The complete results from the Phase 2 study of efgartigimod for the treatment of pemphigus were published in the *British Journal of Dermatology*.

"The publication today highlights the value of our collaborative research model with leading academic centers of excellence, revealing an exciting set of new translational data on the pathophysiology of autoimmune skin blistering diseases and the therapeutic rationale of FcRn blockade in pemphigus and bullous pemphigoid," said Hans de Haard, Ph.D., Chief Scientific Officer of argenx. "We believe the secondary analysis of our Phase 2 study shows the potential role of FcRn blockade to extend beyond IgG antibody reduction to include an immunomodulatory effect on autoantigen-specific B-cells, which may account for the durable responses observed in the study. We understand the serious unmet need of people living with pemphigus and bullous pemphigoid and are committed to continuing this research on their behalf to bring forward an innovative new treatment option."

**Highlights from Published Translational Data:**

- **Pemphigus and BP are IgG-mediated autoimmune skin blistering diseases**
    - In pemphigus, IgG autoantibodies against Dsg-1 and Dsg-3 cause acantholysis or disruption of keratinocyte adhesion; new data suggest role of additional non-desmoglein IgG autoantibodies in pemphigus pathophysiology
    - In BP, IgG autoantibodies against hemidesmosomal proteins (BP180 and BP230) at the dermal-epidermal junction drive pathophysiology of disease

- **In a secondary analysis of a subset of patients in the Phase 2 study of efgartigimod in pemphigus:**
    - **Efgartigimod treatment resulted in sustained reduction of antigen-specific B-cells** in participants with pemphigus vulgaris (PV) and foliaceus, maintained following treatment cessation, and which correlated with sustained clinical improvement while total IgG returned to near baseline levels
    - **Median CD19+ B-cells remained within normal limits** at all timepoints measured, and no new safety signals were detected
    - **No observed effect on total leukocytes, neutrophils, monocytes, or lymphocytes** in patients treated with long-term efgartigimod therapy, however frequency of CD19+ B-cells in circulation was observed to decrease following efgartigimod treatment
- **Efgartigimod was shown to efficiently rescue the loss of keratinocyte adhesion** upon anti-Dsg-3 antibodies (anti-Dsg3) and PV IgG treatment, indicating that stabilization of keratinocyte adhesion may present a novel treatment paradigm for pemphigus

**About Efgartigimod**

Efgartigimod is an antibody fragment designed to reduce pathogenic immunoglobulin G (IgG) antibodies by binding to the neonatal Fc receptor and blocking the IgG recycling process. Efgartigimod is being investigated in several autoimmune diseases known to be mediated by disease-causing IgG antibodies, including neuromuscular disorders, blood disorders, and skin blistering diseases.

**About Pemphigus**

Pemphigus is a rare group of chronic blistering autoimmune diseases that affect the skin and mucous membranes, and are characterized by painful blisters, erosions and acantholysis, or disruption of keratinocyte adhesion. Blisters often break open, causing serious pain and increased risk of infection. Pemphigus vulgaris and pemphigus foliaceous are the most common forms of pemphigus.

**About Bullous Pemphigoid (BP)**

BP is a severe, rare, chronic and recurrent autoimmune disorder characterized by fluid-filled blisters, itching, and redness of the skin. It is the most common subepidermal autoimmune blistering disease. In severe cases with widespread blistering, risk of infection can be life-threatening. Fear of infection and the unpredictable nature of flare ups often has a significant impact on quality of life and psychological well-being.

**About argenx**

argenx is a global immunology company committed to improving the lives of people suffering from severe autoimmune diseases. Partnering with leading academic researchers through its Immunology Innovation Program (IIP), argenx aims to translate immunology breakthroughs into a world-class portfolio of novel antibody-based medicines. argenx developed and is commercializing the first-and-only approved neonatal Fc receptor (FcRn) blocker in the U.S. and Japan. The Company is evaluating efgartigimod in multiple serious autoimmune diseases and advancing several earlier stage experimental medicines within its therapeutic franchises. For more information, visit www.argenx.com and follow us on LinkedIn, Twitter, and Instagram.

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**Forward-looking Statements**

*The contents of this announcement include statements that are, or may be deemed to be, "forward-looking statements." These forward-looking statements can be identified by the use of forward-looking terminology, including the terms "believes," "hope," "estimates," "anticipates," "expects," "intends," "may," "will," or "should" and include statements argenx makes concerning the potential for efgartigimod in treatment of autoimmune skin blistering disorders, the potential extension of the role of FcRn blockade and argenx's ability to complete the research and development of an innovative treatment option for pemphigus and bullous pemphigoid. By their nature, forward-looking statements involve risks and uncertainties and readers are cautioned that any such forward-looking statements are not guarantees of future performance. argenx's actual results may differ materially from those predicted by the forward-looking statements as a result of various important factors. A further list and description of these risks, uncertainties and other risks can be found in argenx's U.S. Securities and Exchange Commission (SEC) filings and reports, including in argenx's most recent annual report on Form 20-F filed with the SEC as well as subsequent filings and reports filed by argenx with the SEC. Given these uncertainties, the reader is advised not to place any undue reliance on such forward-looking statements. These forward-looking statements speak only as of the date of publication of this document. argenx undertakes no obligation to publicly update or revise the information in this press release, including any forward-looking statements, except as may be required by law.*