argenx Reports Topline Results from ADDRESS Study of Efgartigimod SC in Pemphigus

- *ADDRESS study did not meet primary or secondary endpoints*
  - *Pemphigus deprioritized as efgartigimod indication*
    - *Update on BALLAD study GO/NO GO decision*
- *Conference call scheduled for today, December 20, 2023, at 8:30am ET (2:30pm CET)*

Regulated Information – Inside Information

December 20, 2023, 7:00am CET

AMSTERDAM, 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 topline results from the ADDRESS study evaluating efgartigimod subcutaneous (SC) (efgartigimod alfa and hyaluronidase-qvfc) in adults with pemphigus vulgaris (PV) and pemphigus foliaceus (PF). The ADDRESS results show the proportion of PV patients achieving the primary endpoint of complete remission on a minimal dose of steroids (CRmin) was not significantly different between efgartigimod SC and placebo.

argenx will not pursue additional development in pemphigus and plans to prioritize clinical development of efgartigimod in its ongoing severe autoimmune indications.

“We are disappointed by today’s results, particularly for pemphigus patients who have seen little innovation in this treatment space,” said Luc Truyen, M.D., Ph.D., Chief Medical Officer at argenx. “At argenx, we are in the business of transformation, providing new medicines that go beyond incremental benefit and raise the bar for what patients can expect from a treatment. While we will not move forward into pemphigus, our job today is the same as yesterday – continue to be execution-focused and data-driven, apply learnings across our ongoing development programs, and pursue optimal development of efgartigimod, empasiprubart and our earlier stage programs. 2023 was a remarkable year of growth for argenx across the business and we are poised to build on our success in 2024.

“We are grateful to the pemphigus community and all involved in the ADDRESS study, including patients, healthcare professionals, and our argenx teams,” continued Dr. Truyen.

ADDRESS Study Results

The Phase 3 ADDRESS study enrolled 222 adult patients with newly diagnosed or relapsing moderate-to-severe PV (n=190) or PF (n=32). Patients were randomized to efgartigimod SC or placebo with both treatment groups receiving concomitant steroids at a starting dose of 0.5mg/kg, which is a lower dose than recommended by current treatment guidelines and was tapered according to protocol upon achievement of complete remission.

- **Consistent pharmacodynamic (PD) effect of efgartigimod SC**: Treatment with efgartigimod SC led to total immunoglobulin G (IgG) and desmoglein autoantibody (DSG-1 and DSG-3) reductions up to 75%. The observed PD effect was consistent with previous clinical trials of efgartigimod.

- **Unexpected PD effect of corticosteroids**: There was a higher than expected response to background treatment with corticosteroids, which showed a reduction of DSG-1 and DSG-3 levels of up to 70% in the placebo arm and correlated to sustained clinical benefit. The level of autoantibody reduction driven by corticosteroids in both treatment arms was sufficient for patients to achieve CRmin. The significant PD effect of corticosteroids was specific to DSG-1 and DSG-3 autoantibodies, while the observed effect on total IgG reduction was in line with the literature (up to 10%).
- **Study did not meet primary endpoint**: Treatment with efgartigimod SC led to CRmin in 35.5% (44/124) of patients compared to 30.3% (20/66) with placebo (p=0.5956). Secondary endpoints were also not met, including CRmin in the overall pemphigus population (PV and PF), cumulative dose of corticosteroids and time to disease control or complete remission
- **Consistent and favorable safety profile**: Efgartigimod SC was well-tolerated in ADDRESS. The observed safety and tolerability profile was consistent with other clinical trials and the confirmed safety profile of VYVGART and VYVGART Hytrulo.

Update on BALLAD Study

argenx is reviewing the BALLAD study in light of the ADDRESS results and the comparable biology between pemphigus and bullous pemphigoid, and has decided not to make a GO/NO GO decision at this time but rather wait for learnings from all currently enrolled patients and consider a new trial design for the path forward.

Conference Call Details

argenx will host a conference call today at 2:30 pm CET (8:30am ET) to discuss the ADDRESS results. A webcast of the live call and replay may be accessed on the Investors section of the argenx website.

Dial-in Numbers:

Please dial in 15 minutes prior to the live call.

Belgium	32 800 50 201
France	33 800 943355
Netherlands	31 20 795 1090
United Kingdom	44 800 358 0970
United States	1 888 415 4250
Japan	81 3 4578 9081
Switzerland	41 43 210 11 32

About the ADDRESS Study

The ADDRESS study was a randomized, double-blind, placebo-controlled, multicenter, global trial evaluating the efficacy and safety of efgartigimod SC (efgartigimod alfa and hyaluronidase-qvfc) in adult patients with pemphigus. Enrolled patients had newly diagnosed or relapsing moderate-to-severe pemphigus vulgaris (PV) or pemphigus foliaceus (PF) with PDAI scores of ≥15. Patients were randomized in a 2:1 ratio to receive efgartigimod SC or placebo for a total of 30 weeks as part of the primary trial. All patients were on concomitant corticosteroids at a starting dose of 0.5mg/kg/day, which could be tapered according to protocol upon achievement of complete remission (PDAI = 0). The primary endpoint was measured by the proportion of PV patients who achieved sustained complete remission on a minimal dose

of corticosteroids (CRmin) within 30 weeks. Key secondary endpoints included proportion of overall population (PV and PF) who achieved CRmin, cumulative corticosteroid dose, and time to disease control and complete remission. At the end of the 30-week study, eligible patients entered a double-blind 8-week follow-up as part of the ADDRESS open-label extension study during which CRmin off treatment (efgartigimod SC or placebo) was assessed.

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 the BALLAD Study

The BALLAD study is a randomized, double-blind, placebo-controlled, multicenter trial evaluating the efficacy and safety of VYVGART Hytrulo (efgartigimod alfa and hyaluronidase-qvfc) in adult patients with bullous pemphigoid (BP). Enrolled patients have moderate to severe BP and are a more fragile and older population than pemphigus patients. Patients were randomized in a 1:1 ratio to receive VYVGART Hytrulo or placebo for a total of 36 weeks as part of the primary trial. All patients are on concomitant corticosteroids at a starting dose of 0.5 mg/kg/day, which could be tapered according to protocol upon achievement of complete remission (BPDAI= 0). The primary endpoint is measured at 36 weeks by the proportion of BP patients who achieved clinical remission while receiving efgartigimod SC or placebo but off corticosteroids therapy for at least 8 weeks. Key secondary endpoints include cumulative dose of corticosteroids from baseline, proportion of patients who achieve an Investigator Global Assessment of BP (IGA-BP) score of 0 or 1, changes from baseline in the BP Disease Area Index (BPDAI) activity score, proportion of patients who are in CR on minimal corticosteroids for at least 8 weeks at week 36 and time to control of disease or complete remission.

About Bullous Pemphigus

Bullous pemphigoid is a rare chronic blistering autoimmune disease and is the most common form of pemphigoid diseases. It is characterized by autoantibodies against structural proteins of the dermal-epidermal junction and, clinically, by tense blisters and erosions of skin or mucous membranes close to the skin surface. The disease has a strong impact on a person's quality of life and is associated with a high mortality.

About VYVGART Hytrulo (efgartigimod SC)

VYVGART Hytrulo is a subcutaneous combination of efgartigimod alfa, a human IgG1 antibody fragment marketed for intravenous use as VYVGART®, and recombinant human hyaluronidase PH20 (rHuPH20), Halozyme's ENHANZE® drug delivery technology to facilitate subcutaneous injection delivery of biologics. In binding to the neonatal Fc receptor (FcRn), VYVGART Hytrulo results in the reduction of circulating IgG. VYVGART Hytrulo is the proprietary name in the U.S. for subcutaneous efgartigimod alfa and recombinant human hyaluronidase PH20. It is marketed in Europe as VYVGART and may be marketed under different proprietary names following approval in other regions.

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 approved neonatal Fc receptor (FcRn) blocker in the U.S., Japan, Israel, the EU, the UK, China and Canada. 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.

For further information, please contact:

Media:
Ben Petok
bpetok@argenx.com

Investors:
Alexandra Roy (US)
aroy@argenx.com

Lynn Elton (EU)
lelton@argenx.com

This press release contains inside information within the meaning of Article 7(1) of the EU Market Abuse Regulation (Regulation 596/2014).

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 "plans," "aims," "believes," "continues," "hope," "estimates," "anticipates," "expects," "intends," "may," "will," "should," or "commitment" and include statements argenx makes concerning argenx's topline results from the ADDRESS study of efgartigimod SC in adults with PV and PF, our plan to pursue optimal development of efgartigimod, empasiprubart and our earlier stage programs, and our goal of translating immunology breakthroughs into a world-class portfolio of novel antibody-based medicines. 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, including but not limited to argenx's additional analyses of the dataset from the ADDRESS and BALLAD studies, expectations regarding the inherent uncertainties associated with development of novel drug therapies, preclinical and clinical trial and product development activities and regulatory approval requirements, the acceptance of our products and product candidates by our patients as safe, effective and cost-effective, and the impact of governmental laws and regulations on our business. 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.