argenx Initiates Second Cohort of Phase 2 ARDA Study of Empasiprubart in Multifocal Motor Neuropathy

- *Independent Data Monitoring Committee recommended study continuation based on the favorable safety profile observed in the first dose cohort*

- *Early efficacy signals support proof-of-concept of empasiprubart in multifocal motor neuropathy*

Regulated Information/Inside Information

Amsterdam, the Netherlands—June 20, 2023—argenx SE (Euronext & Nasdaq: ARGX), a global immunology company committed to improving the lives of people suffering from severe autoimmune diseases, today announced its plan to advance to a second dose cohort with the Phase 2 ARDA study of empasiprubart (ARGX-117) in multifocal motor neuropathy (MMN). The decision follows a planned interim analysis of the first dose cohort by an Independent Data Monitoring Committee (IDMC) meeting held on June 19, 2023.

"We are encouraged by the favorable safety profile and early efficacy signals from the ARDA study, as well as the IDMC recommendation to advance the study to the next cohort. Looking forward, we hope to build on the promising safety and efficacy we observed in the first cohort while populating our PK/PD model to understand the full potential of empasiprubart in MMN," commented Luc Truyen, M.D., Ph.D., Chief Medical Officer, argenx. "People living with MMN continue to face significant burden associated with this chronic autoimmune disease, including progressive muscle weakness, which can lead to an inability to walk or move one's arms or hands. Our mission is to transform treatment paradigms for autoimmune patients by changing expectations on what well-controlled means. Based on these data, we are hopeful that we can accomplish this in MMN and other autoimmune indications with our first-in-class C2 inhibitor."

The IDMC reviewed interim safety data from all patients (n=22) currently enrolled in the first cohort of the ARDA study, including nine patients who completed the full 16-week treatment period. The IDMC confirmed a favorable safety and tolerability profile of empasiprubart consistent with results from the Phase 1 study and recommended advancing to the second cohort. An early efficacy assessment of all 22 patients supports proof-of-concept of empasiprubart in MMN. The data showed distinct separation between treated patients and placebo based on a suite of clinical outcome measures, including time to IVIg retreatment.

In total, the ARDA study is expected to enroll 48 patients across two cohorts. The study's objective, in addition to assessing safety and efficacy of empasiprubart, is to populate a PK/PD model to inform the Phase 3 study dose selection. Based on the IDMC recommendation to continue enrollment, argenx will aim to build on the promising efficacy and safety observations from the first cohort by evaluating a next dose level of empasiprubart.

Phase 2 ARDA Study Design

The Phase 2 ARDA study is a randomized, double-blinded, placebo-controlled multicenter study to evaluate the safety and tolerability, efficacy, pharmacokinetics, pharmacodynamics, and immunogenicity of two dose regimens of empasiprubart in adults with multifocal motor neuropathy (MMN). The study consists of an IVIg dependency and monitoring period and two 16-week treatment cohorts of 24 MMN patients receiving empasiprubart or placebo in a 2x1 randomization. The dosing for Cohort 2 will be established after a planned interim analysis of the first nine patients to complete the 16-week treatment period from Cohort 1. The primary endpoint is safety and tolerability. Additional endpoints include time to IVIg retreatment, biomarker analyses of C2 levels, and changes in measurements on key clinical efficacy scores (modified medical research council (mMRC)-14 sum score, grip strength, MMN-RODS) as well as several patient-reported quality of life outcome measures.

About Empasiprubart

Empasiprubart (ARGX-117) is a first-in-class humanized sweeping antibody that binds specifically to C2 thereby blocking both the classical and lectin pathways of the complement cascade. By blocking upstream complement activity, empasiprubart has the potential to reduce tissue inflammation representing a broad pipeline opportunity across multiple severe autoimmune indications. In addition to multifocal motor neuropathy, argenx is planning to evaluate empasiprubart in delayed graft function following kidney transplant and dermatomyositis.

About Multifocal Motor Neuropathy

Multifocal motor neuropathy (MMN) is a rare, chronic autoimmune disease of the peripheral nervous system. The disease is characterized by slowly progressive, asymmetric muscle weakness mainly of the hands, forearms and lower legs. MMN is often associated with anti-GM1 IgM autoimmunity, leading to activation of the classical complement pathway, driving subsequent axon damage. High-dose IV immunoglobulin (IVIg) is the only approved treatment for MMN and patients typically experience disease progression despite therapy, indicating an unmet need for efficacious and better tolerated therapeutic options.

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., Japan, the EU and the UK. 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:
Erin Murphy
EMurphy@argenx.com

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

Lynn Elton (EU)
LElton@argenx.com

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 safety profile and efficacy signals from the ARDA study; the prospects of empasiprubart as a treatment for MMN and other indications; and the expected enrollment, objectives and results of the ARDA study. 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 publicly update or revise the information in this press release, including any forward-looking statements, except as may be required by law.