argenx Announces Positive Phase 3 Data from ADVANCE Trial of VYVGART® (efgartigimod alfa-fcab) in Adults with Primary Immune Thrombocytopenia

Study met primary endpoint, demonstrating a higher proportion of sustained platelet response with VYVGART treatment compared to placebo (p=0.0316); responders observed across patient types regardless of prior therapy or disease severity

Statistically significant separation from placebo in key platelet-derived secondary endpoints

Safety and tolerability profile of VYVGART is consistent with previous clinical trials; ADVANCE is first registrational trial with chronic dosing out to 24 weeks

Topline data expected in first quarter 2023 from ADVANCE-SC, the second pivotal trial required for registration in primary immune thrombocytopenia (ITP)

argenx to host investor call today at 8:30am ET / 2:30pm CET

Regulated Information/Inside Information

Breda, the Netherlands—May 5, 2022—argenx SE (Euronext & Nasdaq: ARGX), a global immunology company committed to improving the lives of people suffering from severe autoimmune diseases, today announced positive data from the Phase 3 ADVANCE trial of VYVGART® (efgartigimod alfa-fcab) in adults with primary ITP. ADVANCE met its primary endpoint demonstrating that a higher proportion of chronic ITP patients receiving VYVGART achieved a sustained platelet count response compared to placebo. ADVANCE is the first Phase 3 clinical trial of a neonatal Fc receptor (FcRn) blocker in ITP.

"Immune thrombocytopenia is a rare, debilitating autoimmune disease that can be very difficult to treat, especially in patients who have an insufficient response to previous ITP therapies. There is no clear standard of care and many patients continue to experience significant symptoms and decreased quality of life," said Catherine Broome, M.D., Associate Professor of Medicine at Georgetown University Lombardi Comprehensive Cancer Center, and Principal Investigator in the ADVANCE study. "These data are very promising as they show that platelet counts can rapidly improve to clinically meaningful levels following VYVGART treatment in a proportion of a heavily pretreated patient population. We are excited that targeting pathogenic IgG autoantibodies could represent a new, potential approach to help alleviate the disease burden in this patient community."

The ADVANCE trial enrolled 131 adult patients with chronic and persistent ITP. Patients were heavily pretreated and 67% of patients had received three or more prior ITP therapies, including 59% who had prior thrombopoietin receptor agonist (TPO-RAs) experience, 34% with prior rituximab experience and 37% with a history of splenectomy. Patients were insufficiently controlled at baseline with mean platelet counts of $17x10^9$/L across all patients. Of patients who completed the full ADVANCE study, 94% (63/67) of VYVGART-treated patients and 97% (38/39) of placebo patients continued to the ADVANCE+ open-label extension study.

Highlights of Phase 3 ADVANCE Data

Primary endpoint met

ADVANCE met its primary endpoint demonstrating a significantly higher proportion of patients with chronic ITP receiving VYVGART (17/78; 21.8%) compared to placebo (2/40; 5%) achieved a sustained platelet response (p=0.0316), defined as having platelet counts greater than or equal to $50x10^9$/L on at least four of the last six scheduled visits between weeks 19 and 24 of treatment.

Primary endpoint responders were observed across patient types regardless of age, disease severity, time since diagnosis, prior ITP treatment or background medication.

Key platelet-derived secondary endpoints demonstrated statistical significance

Key platelet-derived secondary endpoints showed VYVGART-treated patients had a statistically significant benefit compared to placebo on (1) cumulative number of weeks where platelet counts were at least $50x10^9$/L in the chronic ITP population (p=0.0009) and (2) sustained platelet response in the overall population, including both chronic and persistent ITP patients (p=0.0108). Numerically fewer WHO-classified bleeding events occurred in treated patients throughout the trial but the difference from placebo was not statistically significant. A higher proportion of treated patients in the overall population achieved a durable, sustained platelet response compared to placebo, defined as a sustained platelet response on at least six of the last eight scheduled visits between weeks 17 and 24 of treatment (p=0.0265), but was not considered statistically significant based on hierarchical testing.

Additional secondary endpoints provided clinically meaningful data on platelet count responses throughout 24-week trial

Additional secondary endpoint data from the ADVANCE trial are consistent with primary and secondary platelet-derived endpoints and provide additional context on metrics that often drive treatment decisions.

- **International Working Group (IWG) responder status**: 51.2% of VYVGART-treated patients were classified as IWG responders and 27.9% as complete responders compared to 20% of placebo patients as IWG responders and 4.4% as complete responders. IWG responders are defined as having a platelet count of at least $30x10^9$/L, a two-fold increase in platelet count from baseline, and the absence of bleeding for two separate, consecutive weekly visits. Complete responders are patients with platelet counts of $100x10^9$/L and the absence of bleeding for two separate, consecutive weekly visits.
- **Mean platelet count change from baseline**: VYVGART-treated patients demonstrated a rapid onset of platelet count improvement with statistically significant separation from placebo observed at week one and maintained through 20 out of 24 weeks of the trial.
- **Switch to biweekly dosing**: Ten VYVGART-treated patients switched to a biweekly (every two weeks) dosing schedule after achieving platelet counts of $100x10^9$/L for three out of four consecutive visits, compared to one placebo patient. Nine of the ten treated patients achieved a sustained platelet response.

Consistent safety and tolerability profile

ADVANCE is the second registrational trial of VYVGART and the first to evaluate chronic weekly dosing. VYVGART was well-tolerated in this 24-week study and the observed safety and tolerability profile was consistent with previous clinical trials.

“In listening to and learning from people in the ITP community, we understand the impact of living with this disease can extend beyond physical signs, taking a serious toll on a person’s quality of life. These compelling preliminary data emphasize the potential for VYVGART to drive responses in ITP regardless of

prior lines of therapy, history of splenectomy or time from diagnosis. We look forward to learning more about the potential approach of targeting pathogenic IgGs in ITP through our ADVANCE-SC trial, which is on track to read out in the first quarter of next year," said Luc Truyen, MD, Ph.D., Chief Medical Officer at argenx. "The totality of data generated thus far continue to support the key attributes of VYVGART, including its onset of action and safety profile. These data further reinforce our confidence in FcRn blockade as a precision tool with the potential to reach a broad spectrum of IgG-mediated severe autoimmune diseases."

The Phase 3 ADVANCE trial is the first of two registrational trials being conducted as part of the ongoing ITP development program. ADVANCE-SC is evaluating subcutaneous efgartigimod for the treatment of primary ITP. Topline data from the ADVANCE-SC study are expected in the first quarter of 2023.

Phase 3 ADVANCE Trial Design

The Phase 3 ADVANCE trial was a randomized, double-blind, placebo-controlled, multicenter, global trial evaluating the efficacy and safety of VYVGART in adult patients with chronic or persistent primary ITP. A total of 131 adult patients with primary ITP in North America, Europe and Japan enrolled in the trial and received VYVGART or placebo for a total of 24 weeks as part of the primary trial. Enrolled patients had a confirmed ITP diagnosis and a mean entry platelet count of less than $30x10^9$/L. Patients were on a stable dose of at least one ITP treatment prior to randomization and had received at least one prior therapy. Concomitant medications permitted included corticosteroids, nonsteroidal immunosuppressive drugs, fostamatinib or TPO-RAs. If patients were on 'watch and wait' at baseline, they had to have received at least 2 prior treatments for ITP.

Patients were randomized in a 2:1 ratio to receive VYVGART or placebo for a total of 24 weeks as part of the primary trial. Randomized patients received weekly infusions from weeks 1-4 and were eligible to adjust frequency to bi-weekly depending on platelet count. Administration frequency was fixed from study visits 16-24. The primary endpoint was measured by the proportion of patients with chronic ITP with a sustained platelet count response defined as achieving platelet counts of greater than or equal to $50x10^9$/L for at least four of the last six scheduled visits between weeks 19 and 24. Patients who received rescue therapy at week 12 or later, or for whom dose and/or frequency of concurrent ITP therapies increased at week 12 or later, were considered non-responders. Key secondary endpoints included extent of disease control over 24-week treatment period, proportion of overall population with sustained platelet count response, incidence and severity of WHO-classified bleeding events and an extended primary endpoint analysis between weeks 17 and 24.

About Immune Thrombocytopenia (ITP)

Immune thrombocytopenia (ITP) is an autoimmune disorder where immunoglobulin G (IgG) autoantibodies destroy platelets and reduce platelet production, which can lead to an increased risk of excessive bleeding and bruising. In severe cases, frequent bleeding events can cause anemia or even brain hemorrhage in rare cases. ITP is also associated with debilitating fatigue and significant impacts on mental health, including anxiety, fear and depression. Many ITP patients are inadequately controlled on current therapies so there remains a significant unmet need for additional treatment options.

About VYVGART® (efgartigimod alfa-fcab)

VYVGART is a human IgG1 antibody fragment that binds to the neonatal Fc receptor (FcRn), resulting in the reduction of circulating immunoglobulin G (IgG) autoantibodies. It is the first and only approved FcRn blocker. VYVGART is approved in the United States for the treatment of adults with generalized

myasthenia gravis (gMG) who are anti-acetylcholine receptor (AChR) antibody positive and in Japan for the treatment of adults with gMG who do not have sufficient response to steroids or non-steroidal immunosuppressive therapies (ISTs). VYVGART is being studied in adults with primary immune thrombocytopenia (ITP) and other IgG autoantibody-mediated diseases.

Important Safety Information for VYVGART® (efgartigimod alfa-fcab) intravenous (IV) formulation (U.S. prescribing information) for the treatment of generalized myasthenia gravis (gMG) in adult patients who are anti-acetylcholine receptor (AChR) antibody positive.

What is VYVGART® (efgartigimod alfa-fcab)?
VYVGART is a prescription medicine used to treat a condition called generalized myasthenia gravis, which causes muscles to tire and weaken easily throughout the body, in adults who are positive for antibodies directed toward a protein called acetylcholine receptor (anti-AChR antibody positive).
What is the most important information I should know about VYVGART?
VYVGART may cause serious side effects, including:

- **Infection.** VYVGART may increase the risk of infection. In a clinical study, the most common infections were urinary tract and respiratory tract infections. More patients on VYVGART vs placebo had below normal levels for white blood cell counts, lymphocyte counts, and neutrophil counts. The majority of infections and blood side effects were mild to moderate in severity. Your health care provider should check you for infections before starting treatment, during treatment, and after treatment with VYVGART. Tell your health care provider if you have any history of infections. Tell your health care provider right away if you have signs or symptoms of an infection during treatment with VYVGART such as fever, chills, frequent and/or painful urination, cough, pain and blockage of nasal passages/sinus, wheezing, shortness of breath, fatigue, sore throat, excess phlegm, nasal discharge, back pain, and/or chest pain.
- **Undesirable immune reactions (hypersensitivity reactions).** VYVGART can cause the immune system to have undesirable reactions such as rashes, swelling under the skin, and shortness of breath. In clinical studies, the reactions were mild or moderate and occurred within 1 hour to 3 weeks of administration, and the reactions did not lead to VYVGART discontinuation. Your health care provider should monitor you during and after treatment and discontinue VYVGART if needed. Tell your health care provider immediately about any undesirable reactions.

Before taking VYVGART, tell your health care provider about all of your medical conditions, including if you:

- Have a history of infection or you think you have an infection.
- Have received or are scheduled to receive a vaccine (immunization). Discuss with your health care provider whether you need to receive age-appropriate immunizations before initiation of a new treatment cycle with VYVGART. The use of vaccines during VYVGART treatment has not been studied, and the safety with live or live-attenuated vaccines is unknown. Administration of live or live-attenuated vaccines is not recommended during treatment with VYVGART.
- Are pregnant or plan to become pregnant and are breastfeeding or plan to breastfeed.

Tell your health care provider about all the medicines you take, including prescription and over-the-counter medicines, vitamins, and herbal supplements.

What are the common side effects of VYVGART?
The most common side effects of VYVGART are respiratory tract infection, headache, and urinary tract infection.

These are not all the possible side effects of VYVGART. Call your doctor for medical advice about side effects. You may report side effects to the US Food and Drug Administration at 1-800-FDA-1088.

Please see the full Prescribing Information for VYVGART and talk to your doctor.

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 expected long-term safety, tolerability and efficacy of VYVGART® (efgartigimod alfa-fcab) in adult patients with Primary Immune thrombocytopenia. 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.