argenx announces VYVGART™ approval in Japan for the treatment of generalized myasthenia gravis

- *VYVGART™ (efgartigimod alfa) is the first-and-only FcRn blocker approved in Japan*

- *Ministry of Health, Labour and Welfare (MHLW) decision marks second regulatory approval of VYVGART as part of global launch strategy, following approval in U.S. on December 17, 2021*

Regulated Information/Inside Information

Breda, the Netherlands – Jan. 20, 2022 – argenx SE (Euronext & Nasdaq: ARGX), a global immunology company committed to improving the lives of people suffering from severe autoimmune diseases, today announced that Japan's Ministry of Health, Labour and Welfare (MHLW) has approved VYVGART™ (efgartigimod alfa) intravenous infusion for the treatment of adult patients with generalized myasthenia gravis (gMG) who do not have sufficient response to steroids or non-steroidal immunosuppressive therapies (ISTs). VYVGART is the first-and-only neonatal Fc receptor (FcRn) blocker approved in Japan.

"People living with gMG around the world continue to experience severe disease burden despite treatment with commonly-used therapies. We are extremely proud to deliver the first-and-only approved FcRn blocker in Japan to a broad population of gMG patients, regardless of antibody status," said Tim Van Hauwermeiren, Chief Executive Officer of argenx. "Our commercial teams are ready and motivated to be serving as many people as possible who are living with this debilitating disease and we look forward to collaborating with the Japanese government to enable patient access. With today's approval of VYVGART in Japan, the recent U.S. FDA approval, and ongoing review of our application in Europe, we continue to advance rapidly toward achieving our goal of delivering this innovative, targeted treatment option globally."

Generalized myasthenia gravis is a rare and chronic neuromuscular disease characterized by debilitating and potentially life-threatening muscle weakness. VYVGART is a human IgG1 antibody fragment that binds to FcRn, resulting in the reduction of circulating immunoglobulin G (IgG) autoantibodies. The action of IgG autoantibodies at the neuromuscular junction is a key driver of gMG.[2]

The approval of VYVGART is based on results from the global Phase 3 ADAPT trial, which were published in the July 2021 issue of *The Lancet Neurology*. The ADAPT trial met its primary endpoint, demonstrating that significantly more anti-acetylcholine receptor (AChR) antibody positive gMG patients were responders on the myasthenia gravis activities of daily living (MG-ADL) scale following treatment with VYVGART compared with placebo (68% vs. 30%; $p<0.0001$). Responders were defined as having at least a two-point reduction on the MG-ADL scale sustained for four or more consecutive weeks during the first treatment cycle.

There were also significantly more responders on the Quantitative Myasthenia Gravis (QMG) scale following treatment with VYVGART compared with placebo (63% vs. 14%; p<0.0001). Responders were defined as having at least a three-point reduction on the QMG scale sustained for four or more consecutive weeks during the first treatment cycle.

VYVGART had a demonstrated safety profile in the ADAPT clinical trial. The most common adverse events in ADAPT were respiratory tract infection (33% vs. 29% placebo), headache (32% vs. 29% placebo), and urinary tract infection (10% vs. 5% placebo).

VYVGART was approved by the U.S. Food and Drug Administration (FDA) on December 17, 2021 for the treatment of gMG in adult patients who are AChR antibody positive. A Marketing Authorization Application for efgartigimod for the treatment of gMG is currently under review by the European Medicines Agency (EMA), with a decision anticipated in the second half of 2022. argenx is evaluating efgartigimod in six high-need autoimmune conditions, set to expand to ten conditions by the end of 2022.

See the full Prescribing Information for VYVGART in the U.S., which includes the below Important Safety Information. For more information related to VYVGART in Japan, visit argenx.jp.

IMPORTANT SAFETY INFORMATION (U.S. PRESCRIBING INFORMATION)

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 VYVGART™

VYVGART™ (efgartigimod alfa-fcab) 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).

About Generalized Myasthenia Gravis

Generalized myasthenia gravis (gMG) is a rare and chronic autoimmune disease where IgG autoantibodies disrupt communication between nerves and muscles, causing debilitating and potentially life-threatening muscle weakness. Approximately 85% of people with MG progress to

gMG within 24 months[1], where muscles throughout the body may be affected. Patients with confirmed AChR antibodies account for approximately 85% of the total gMG population.[1]

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.

References

[1] Behin et al. New Pathways and Therapeutics Targets in Autoimmune Myasthenia Gravis. J Neuromusc Dis 5. 2018. 265-277
[2] Howard JF Jr, Utsugisawa K, Benatar M, et al. Safety and efficacy of efficacy of eculizumab in anti-acetylcholine receptor antibody-positive refractory generalised myasthenia gravis (REGAIN): a phase 3, randomised, double-blind, placebo-controlled, multicenter study. Lancet Neurol. 2017; 16: 976-86

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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 its global launch strategy; its expectation

concerning treatment options, scale of potential patients and impact and effect on patients; estimates concerning the commercialization potential of VYVGART; anticipated outcome of collaboration with the Japanese government; expected approval of VYVGART by European Medicines Agency of MAA in the second half of 2022; and evaluation of efgartigimod in up to ten high-need conditions by end of 2022. 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.