argenx Announces U.S. Food and Drug Administration (FDA) Approval of VYVGART™ (efgartigimod alfa-fcab) in Generalized Myasthenia Gravis

- *VYVGART is the first-and-only FDA-approved neonatal Fc receptor blocker*
- *68% of anti-acetylcholine receptor (AChR) antibody positive gMG patients treated with VYVGART were responders (n=44/65) on the Myasthenia Gravis - Activities of Daily Living (MG-ADL) scale compared with 30% of patients treated with placebo (n=19/64) (p<0.0001) during the first treatment cycle in the Phase 3 ADAPT trial*
- *argenx has reached agreement in principle with various national and regional payers to structure value-based agreements*
- *argenx to host investor conference call today at 5:30pm ET*

BREDA, Netherlands--(BUSINESS WIRE)-- Regulatory News:

argenx SE (Euronext & Nasdaq: ARGX), a global immunology company committed to improving the lives of people suffering from severe autoimmune diseases and cancer, today announced that the U.S. Food and Drug Administration (FDA) has approved VYVGART™ (efgartigimod alfa-fcab) for the treatment of generalized myasthenia gravis (gMG) in adult patients who are anti-acetylcholine receptor (AChR) antibody positive. These patients represent approximately 85% of the total gMG population[1]. With this regulatory milestone, VYVGART is the first-and-only FDA-approved neonatal Fc receptor (FcRn) blocker.

“Today is the start of a new era for argenx and the gMG community as we honor our commitment to bring forward an innovative treatment option for people living with this debilitating disease. The approval of VYVGART represents many achievements: our first approved product; the first-and-only FDA-approved neonatal Fc receptor blocker; and the first approved therapy designed to reduce pathogenic IgGs, an underlying driver of gMG,” said Tim Van Hauwermeiren, Chief Executive Officer of argenx. “Importantly, we want to thank the patients, supportive caregivers, investigators and study teams who participated in the ADAPT trial, as well as our partners and dedicated employees for their hard work and collaboration – all of whom made this milestone possible.

“Our highly motivated commercial team is activated and ready to deliver VYVGART to patients. We believe the field of autoimmunity is on the precipice of an evolution, and we hope that this will be the first of many VYVGART launches, allowing us to help improve the lives of patients around the world," continued Mr. Van Hauwermeiren.

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) antibodies. The action of AChR autoantibodies at the neuromuscular junction is a key driver of gMG[2].

“The gMG community has long-awaited the FDA approval of VYVGART, especially for those patients who struggle with basic personal tasks such as speaking, chewing and swallowing food, brushing teeth and hair, and in some severe cases, breathing,” commented Samantha Masterson, President and Chief Executive Officer of the Myasthenia Gravis Foundation of America. “We thank argenx for its continued commitment to the gMG patient community, which led them to deliver this much-needed new treatment option with the potential to change the lives of many gMG patients.”

Proven clinical efficacy and safety profile

“People living with gMG have been in need of new treatment options that are targeted to the underlying pathogenesis of the disease and supported by clinical data,” said James F. Howard Jr., M.D., Professor of Neurology (Neuromuscular Disease), Medicine and Allied Health, Department of Neurology, The University of North Carolina at Chapel Hill School of Medicine and Principal Investigator for the ADAPT trial. “Today’s approval represents an important new advance for gMG patients and families affected by this debilitating disease. This therapy has the potential to reduce the disease burden of gMG and transform the way we treat this disease.”

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-AChR antibody positive gMG patients were responders on the 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 additionally 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).

Access and *My VYVGART Path*

argenx is committed to supporting affordable access to VYVGART. As part of this commitment, argenx is launching *My VYVGART Path*, a program designed to connect patients and clinicians to personalized support throughout the treatment journey. Program resources include disease and product education, access support and benefits verification, and financial assistance programs for eligible patients. Patients and healthcare providers can visit VYVGART.com for more information.

Adults living with gMG each experience the course of the disease differently, contributing to variability of disease severity and response to therapies. argenx prioritized early and active engagement with leading payers to address questions around broader budget predictability. The Company has reached agreements in principle with several national and regional commercial payers to structure a value-based agreement. The agreements are meant to provide predictability of cost for payers and appropriate access for patients.

“Generalized myasthenia gravis imposes a significant lifestyle and treatment burden on patients, families, and the overall healthcare system. This autoimmune disease affects each patient differently which can create variability in dosing and the resulting cost per patient,” said Steve Miller, M.D., Executive Vice President and Chief Clinical Officer at Cigna Corp. “The approval of VYVGART promises to address a treatment gap for patients suffering from this disease. argenx has put forth an innovative, value-based approach to contracting that will help payers with cost predictability as they face the challenge of ensuring real-world dosing remains affordable. This was a direct result of early engagement between argenx and Evernorth leading up to approval. Cigna looks forward to continued collaboration with argenx to enable appropriate access to the gMG patients who will benefit from VYVGART.”

Marketing Authorization Applications for efgartigimod for the treatment of gMG are currently under review with Japan’s Pharmaceuticals and Medical Devices Agency (PMDA) and the European Medicines Agency (EMA), with anticipated decisions from each agency in the first quarter and second half of 2022, respectively.

argenx has an exclusive partnership agreement with Zai Lab for the development and commercialization of efgartigimod in Greater China. Zai Lab is on track to file for approval in Greater China by mid-2022. Under the terms of the strategic agreement with Zai Lab, argenx will receive a $25 million milestone payment with this U.S. approval of VYVGART. In addition, argenx has an exclusive partnership with Medison for the commercialization of efgartigimod in gMG in Israel. Medison is on track to file for approval in Israel in the second quarter of 2022.

Conference Call and Webcast

A webcast of the live call may be accessed on the Investors section of the argenx website at argenx.com/investors. A replay of the webcast will be available on the argenx website for approximately one year following the call.

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 9752

Switzerland 41 43 210 11 32

See Important Safety Information and full Prescribing Information below for additional information.

IMPORTANT SAFETY 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 Phase 3 ADAPT Trial
The Phase 3 ADAPT trial was a 26-week randomized, double-blind, placebo-controlled, multi-center, global trial evaluating the safety and efficacy of VYVGART in adult patients with gMG. A total of 167 adult patients with gMG in North America, Europe and Japan enrolled in the trial. Patients were randomized in a 1:1 ratio to receive VYVGART or placebo, in addition to stable doses of their current gMG treatment. ADAPT was designed to enable an

individualized treatment approach with an initial treatment cycle followed by subsequent treatment cycles based on clinical evaluation. The primary endpoint was the comparison of percentage of MG-ADL responders in the first treatment cycle between VYVGART and placebo treatment groups in the anti-AChR antibody positive population.

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 IgG. 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-AChR antibody positive.

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 and cancer. 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 will commercialize the first-and-only FDA approved neonatal Fc receptor blocker, VYVGART (efgartigimod alfa-fcab) for the treatment of adult gMG patients who are anti-acetylcholine receptor (AChR) antibody positive. 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 at https://www.linkedin.com/company/argenx/ and Twitter at https://twitter.com/argenxglobal.

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

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 expectations for treatment options, estimating the commercialization potential of VYVGART; beliefs regarding the evolution in the field of autoimmunity; impact and effect on patients and scale of potential patients; impact on gMG community; expectations for agreements with commercial payers; statements regarding promises to address treatment gaps for patients and approach to contracting; anticipated decisions from PMDA and EMA in the first quarter and second half of 2022, respectively; expectations for Zai Lab to file for approval in Greater China in the first half of 2022, and Medison to file for approval in Israel in the second quarter of 2022; its aim to translate breakthroughs into a world-class portfolio; and its evaluation of efgartigimod and other experimental medicines. 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.

Contacts

Media:
Kelsey Kirk
kkirk@argenx.com

Joke Comijn (EU)
jcomijn@argenx.com

Investors:
Beth DelGiacco
bdelgiacco@argenx.com

Michelle Greenblatt
mgreenblatt@argenx.com

Source: argenx SE

Smart Multimedia Gallery

Photo

Tim Van Hauwermeiren, co-founder and CEO of argenx NV (Photo: Business Wire)

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