



argenx Highlights Breadth of Autoimmune Pipeline with New Multifocal Motor Neuropathy Data at 2024 Peripheral Nerve Society Annual Meeting

ARDA study data show potential for empasiprubart to drive functional improvement and reduced risk of relapse for multifocal motor neuropathy (MMN) patients

ADHERE+ data show durability of functional improvements with VYVGART® Hytrulo (efgartigimod alfa and hyaluronidase-qvfc), which is FDA approved for use in adults with chronic inflammatory demyelinating polyneuropathy (CIDP)

June 25, 2024 – 4:30pm EDT

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 that new data from across the company’s autoimmune pipeline were presented at the 2024 Peripheral Nerve Society (PNS) Annual Meeting in Montréal, Quebec.

“argenx is on a mission to transform the treatment of severe autoimmunity,” said Luc Truyen, M.D., Ph.D., Chief Medical Officer, argenx. “We have established MMN and CIDP as autoantibody-mediated diseases, and by developing novel medicines that precisely target disease biology, we are fulfilling our mission to create truly transformative outcomes for patients. We are excited to present data for our novel therapies that may offer benefits beyond symptom management – to safely help patients regain control of their lives without harsh side effects, residual impairments, or treatments dependent on high frequency infusions. argenx continues to build scientific and clinical evidence that supports the advancement of our innovative pipeline and shows the potential to expand therapeutic choices while reducing the risk of relapse and accumulating disability.”

Phase 2 ARDA Data Support Empasiprubart as Novel Targeted Treatment for MMN

For the first time at a medical congress, argenx presented Cohort 1 data from the Phase 2 ARDA study, which support proof of concept for empasiprubart as a potential new treatment option for MMN, a chronic, progressive autoimmune disease with only one approved treatment option. ARDA is the largest interventional study to-date in MMN.

- **Improved function and reduced risk of IVIg retreatment:** Compared with placebo, treatment with empasiprubart reduced the risk of IVIg retreatment by 91% (HR: 0.09 [95% CI: 0.02–0.44]), with improvement of grip and muscle strength, and improved patients’ ability to perform daily activities.
- **Favorable tolerability profile:** Empasiprubart was well-tolerated and most adverse events were mild or moderate.

ADHERE Oral Presentation Highlights Consistent Sustained Efficacy and Safety of VYVGART Hytrulo for Patients with CIDP

argenx also presented new data from the pivotal ADHERE and open-label extension ADHERE+ studies evaluating VYVGART Hytrulo in patients with CIDP. The ADHERE data supported the recent FDA approval of VYVGART Hytrulo as a safe and effective new treatment option for CIDP, demonstrating sustained functional benefit across all disease scores regardless of disease stage or treatment history. ADHERE met its primary endpoint ($p < 0.0001$) demonstrating a 61% reduction (HR: 0.39 95% CI: 0.25; 0.61) in the risk

of relapse versus placebo. Ninety-nine percent of trial participants elected to participate in the ADHERE+ open-label extension.

- **Sustained functional improvements:** Demonstrated improvements in functional ability (mean aINCAT scores) on VYVGART Hytrulo from Stage A baseline were maintained through Stage B of ADHERE and to Week 24 of ADHERE+. Mean aINCAT scores improved from Run-in Period baseline, indicating some patients gain functional benefit on VYVGART Hytrulo compared to baseline assessments while on prior treatment.
- **Secondary endpoints show consistent improvements across functional and strength measures:** VYVGART Hytrulo-treated patients demonstrated maintenance of functional benefit in Stage B, whereas placebo patients experienced clinically meaningful worsening across all disease scores, including aINCAT, I-RODS and grip strength
- **High retention and compliance in long-term extension study:** 98.9% of ADHERE+ participants demonstrated treatment compliance with VYVGART Hytrulo and 86% were still ongoing at Week 24 of the OLE.
- **Favorable safety profile:** Safety profile of VYVGART Hytrulo was similar between ADHERE and ADHERE+ with no increased rate of adverse events with increased exposure.

VYVGART Hytrulo was approved on June 21, 2024, for the treatment of adult patients with CIDP by the U.S Food and Drug Administration (FDA).

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 the presence of anti-GM1 IgM autoantibodies, leading to activation of the classical complement pathway, driving subsequent axon damage. High-dose 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 Chronic Inflammatory Demyelinating Polyneuropathy (CIDP)

Chronic inflammatory demyelinating polyneuropathy (CIDP) is a rare and serious autoimmune disease of the peripheral nervous system. Although confirmation of disease pathophysiology is still emerging, there is increasing evidence that IgG antibodies play a key role in the damage to the peripheral nerves. People with CIDP experience fatigue, muscle weakness and a loss of feeling in their arms and legs that can get worse over time or may come and go. These symptoms can significantly impair a person's ability to function in their daily lives. Without treatment, one-third of people living with CIDP will need a wheelchair.

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 before 2:1 randomization into a double-blind treatment phase for 16-weeks. Two sequential cohorts of 27 MMN patients receiving empasiprubart or placebo were enrolled investigating two different dose levels of empasiprubart. 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 ADHERE Trial Design

The ADHERE trial was a multicenter, randomized, double-blind, placebo-controlled trial evaluating VYVGART® Hytrulo (efgartigimod alfa and hyaluronidase-qvfc) for the treatment of chronic inflammatory demyelinating polyneuropathy (CIDP). ADHERE enrolled 322 adult patients with CIDP who were treatment naïve (not on active treatment within the past six months or newly diagnosed) or being treated with immunoglobulin therapy or corticosteroids. The trial consisted of an open-label Stage A followed by a randomized, placebo-controlled Stage B. In order to be eligible for the trial, the diagnosis of CIDP was confirmed by an independent panel of experts. Patients entered a run-in stage, where any ongoing CIDP treatment was stopped and in order to be eligible for Stage A had to demonstrate active disease, with clinically meaningful worsening on at least one CIDP clinical assessment tool, including INCAT, I-RODS, or mean grip strength. Treatment naïve patients were able to skip the run-in period with proof of recent worsening. To advance to Stage B, patients needed to demonstrate evidence of clinical improvement (ECI) with VYVGART Hytrulo. ECI was achieved through improvement of the INCAT score, or improvement on I-RODS or mean grip strength if those scales had demonstrated worsening during the run-in period. In Stage B, patients were randomized to either VYVGART Hytrulo or placebo for up to 48 weeks. The primary endpoint was measured once 88 total relapses or events were achieved in Stage B and was based on the hazard ratio for the time to first adjusted INCAT deterioration (i.e. relapse). After Stage B, all patients had the option to roll-over to an open-label extension study to receive VYVGART Hytrulo.

About Empasiprubart

Empasiprubart (ARGX-117) is a first-in-class humanized monoclonal antibody that binds C2 and blocks activation of both the classical and lectin pathways of the complement cascade, leaving the alternative pathway intact for its antimicrobial properties. By blocking complement activity upstream of C3 and C5, empasiprubart has the potential to reduce tissue inflammation and cellular damage, representing a broad pipeline opportunity across multiple severe autoimmune indications. In addition to multifocal motor neuropathy, argenx is evaluating empasiprubart in delayed graft function following kidney transplant and dermatomyositis.

About VYVGART Hytrulo (efgartigimod alfa and hyaluronidase-qvfc)

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. It is the first-and-only approved FcRn blocker administered by subcutaneous injection.

VYVGART Hytrulo is the proprietary name in the U.S. for subcutaneous efgartigimod alfa and recombinant human hyaluronidase PH20. It is marketed as VYVDURA® in Japan and VYVGART SC in Europe.

See FDA-approved Important Safety Information below and full [Prescribing Information](#) for VYVGART Hytrulo for additional information.

What is VYVGART® HYTRULO (efgartigimod alfa and hyaluronidase-qvfc)?

VYVGART HYTRULO is a prescription medicine used for the treatment of adult patients with chronic inflammatory demyelinating polyneuropathy (CIDP).

IMPORTANT SAFETY INFORMATION

Do not use VYVGART HYTRULO if you have a serious allergy to efgartigimod alfa, hyaluronidase, or any of the other ingredients in VYVGART HYTRULO. VYVGART HYTRULO can cause serious allergic reactions and a decrease in blood pressure leading to fainting.

VYVGART HYTRULO may cause serious side effects, including:

- **Infection.** VYVGART HYTRULO may increase the risk of infection. The most common infections for efgartigimod alfa-fcab-treated patients were urinary tract and respiratory tract infections. Signs or symptoms of an infection may include 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.
- **Allergic Reactions (hypersensitivity reactions).** VYVGART HYTRULO can cause allergic reactions such as rashes, swelling under the skin, and shortness of breath. Hives were also observed in patients treated with VYVGART HYTRULO. Serious allergic reactions, such as trouble breathing and decrease in blood pressure leading to fainting have been reported with efgartigimod alfa-fcab.
- **Infusion-Related Reactions.** VYVGART HYTRULO can cause infusion-related reactions. The most frequent symptoms and signs reported with efgartigimod alfa-fcab were high blood pressure, chills, shivering, and chest, abdominal, and back pain.

Tell your doctor if you have signs or symptoms of an infection, allergic reaction, or infusion-related reaction. These can happen while you are receiving your VYVGART HYTRULO treatment or afterward. Your doctor may need to pause or stop your treatment. Contact your doctor immediately if you have signs or symptoms of a serious allergic reaction.

Before taking VYVGART HYTRULO, tell your doctor if you:

- take any medicines, including prescription and non-prescription medicines, supplements, or herbal medicines,
- have received or are scheduled to receive a vaccine (immunization), or
- have any allergies or medical conditions, including if you are pregnant or planning to become pregnant, or are breastfeeding.

What are the common side effects of VYVGART HYTRULO?

The most common side effects in efgartigimod-alfa-fcab-treated patients were respiratory tract infection, headache, and urinary tract infection. Additional common side effects with VYVGART HYTRULO are injection site reactions, including rash, redness of the skin, itching sensation, bruising, pain, and hives.

These are not all the possible side effects of VYVGART HYTRULO. 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 HYTRULO 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 approved neonatal Fc receptor (FcRn) blocker in the U.S., Japan, Israel, the EU, the UK, Canada and China. 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](#).

Contacts

Media:

Ben Petok

bpetok@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 “aims,” “continues,” “may,” or “potential,” and include statements argenx makes regarding the potential for empasiprubarat to drive functional improvement and reduced risk of relapse for MMN patients; certain results or previews of its clinical studies and potential opportunities thereof; the potential benefits beyond symptom management of its therapies; and its 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 the results of argenx’s clinical trials; expectations regarding the inherent uncertainties associated with the development of novel drug therapies; preclinical and clinical trial and product development activities and regulatory approval requirements in products and product candidates; the acceptance of argenx’s products and product candidates by patients as safe, effective and cost-effective; the impact of governmental laws and regulations on our business; disruptions caused on our reliance of third-party suppliers, service providers and manufacturers; inflation and deflation and the corresponding fluctuations in interest rates; and regional instability and conflicts. 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.