argenx Presents New Efgartigimod Data at EULAR 2025 Highlighting Positive Phase 2 Proof-of-Concept Results in Myositis and Sjogren's Disease

- *ALKIVIA data demonstrate significant improvement in muscle strength and physical function in myositis patients treated with efgartigimod*
- *RHO data show efgartigimod achieved sustained reduction in autoantibodies and improved functional outcomes in patients with Sjogren's disease; program granted U.S. FDA Fast Track designation*
- *argenx committed to new therapeutic areas in rheumatology with ongoing Phase 3 studies in myositis (ALKIVIA) and Sjogren's disease (UNITY)*

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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 the presentation of positive results from Phase 2 studies evaluating VYVGART® (IV: efgartigimod alfa-fcab and SC or Hytrulo: efgartigimod alfa and hyaluronidase-qvfc) in Sjogren's disease (SjD) and idiopathic inflammatory myopathies (IIM or myositis) at the European Congress of Rheumatology, EULAR 2025, from June 11 – 14 in Barcelona, Spain.

argenx also announced that the U.S. Food and Drug Administration (FDA) has granted efgartigimod Fast Track designation (FTD) for the treatment of primary Sjogren's disease.

"Our innovation model prioritizes strong biologic rationale and efficient clinical program design, which enables us to rapidly advance development in rheumatic diseases," said Luc Truyen, M.D., Ph.D., Chief Medical Officer, argenx. "The accumulating body of evidence about the role of IgG autoantibodies reinforces the therapeutic potential of efgartigimod as a new approach for treating several rheumatic diseases – aiming to go beyond symptom management by targeting the underlying disease. The data presented at EULAR highlight efgartigimod's potential as a precision therapy for patients living with myositis and Sjogren's disease, and we are hopeful this novel treatment will offer a new therapeutic option and lead to improved outcomes for patients."

ALKIVIA Data Show Efgartigimod Provides Functional Improvement in Patients with Myositis

Consistent and statistically significant treatment effect:
In the ongoing, seamless ALKIVIA Phase 2/3 study evaluating three myositis subtypes (IMNM, ASyS, DM), data from Phase 2 show that patients demonstrated significant improvement in muscle strength and physical function when treated with efgartigimod. The study's primary endpoint, mean Total Improvement Score (TIS) at 24 weeks, is a composite of six core measures of disease activity and muscle function. TIS improvement was observed in a majority of efgartigimod-treated patients across all six core measures, and the primary endpoint was met. Efgartigimod patients showed a significantly higher mean TIS of 50.45 compared to 35.65 in the placebo arm (2-sided P=0.0004). In addition, for patients treated with efgartigimod, 79% achieved a moderate improvement (TIS ≥40) and 34% achieved a major improvement (TIS ≥60), compared to 47% and 9.5% respectively of patients receiving placebo.

Favorable Time to TIS and Safety Profile: Among the study's secondary endpoints, patients receiving efgartigimod improved significantly faster than patients receiving placebo, leading to a median time to minimal improvement (TIS ≥20) of 30 days and time to moderate improvement (TIS ≥40) of 16 weeks. Comparatively, patients in the placebo arm reached minimal improvement (TIS ≥20) in 72 days, while there was no majority of placebo patients reaching moderate improvement (TIS ≥40) at any point in the 24-week study. Efgartigimod was well-tolerated and the proportion of patients experiencing at least one treatment-emergent adverse event (TEAE) was similar in the efgartigimod and placebo arms.

Evaluation of efgartigimod in myositis is ongoing in the Phase 3 portion of the ALKIVIA study.

"Myositis is a debilitating disease that can cause muscle weakness, affect multiple organs, and have a severe impact on patients' quality of life. Physicians struggle to treat it because current options are limited and have significant side effects," said Hector Chinoy, Ph.D., ALKIVIA study investigator and Professor of Rheumatology and Neuromuscular Disease at The University of Manchester. "Results from this study, the first of an FcRn inhibitor in myositis, demonstrate the potential of a transformative targeted treatment approach. Efgartigimod was well-tolerated and led to significant improvements compared to placebo, offering new hope for a treatment that targets autoantibodies as one of the potential key drivers of disease."

RHO Data Show Efgartigimod's Clinical Effect Across Endpoints in Sjogren's Disease

Improved systemic disease activity and reduction in symptoms: In the Phase 2 proof-of-concept RHO study, efgartigimod showed significant improvement in systemic disease activity and patient symptoms. 45.5% of patients receiving efgartigimod achieved improved outcomes on the CRESS composite primary endpoint at Week 24 – including systemic disease activity, salivary and tear gland function – compared to 11.1% among patients treated with placebo. Improvements among patients treated with efgartigimod were achieved in 4 out of 5 CRESS measures. In addition, disease activity among patients treated with efgartigimod showed a median change in clinESSDAI total score of -7.0 versus -4.0 in the placebo arm. A key secondary endpoint is the cSTAR composite of five disease measures, which showed patients treated with efgartigimod achieved a 54.5% response versus 33.3% in the placebo arm.

Potential for disease biology modulation: Biomarker response in RHO study also demonstrated rapid and sustained reduction of IgG with a ~60% reduction from Week 4 onwards. The efgartigimod group showed notable decreases in the disease-associated antibodies anti-Ro52 (-57% vs +13%) and Rheumatoid Factor (-26.6% vs -5.3%), as well as reduction in C1Q immune complexes (-4.5 vs -0.06 mc

eq/mL) compared to placebo.

Efgartigimod demonstrated a favorable safety profile among patients with Sjogren's disease. The observed safety and tolerability was consistent with other clinical trials, with no new safety signals observed. The Phase 3 UNITY trial is currently ongoing to assess efficacy and safety of efgartigimod in patients with moderate to severe Sjogren's disease.

"These data suggest that targeting FcRn and reducing IgGs has a meaningful impact on Sjogren's disease," said Isabelle Peene M.D., Ph.D., study investigator, Department of Rheumatology, Ghent University Hospital. "The clinical and biomarker findings add to our growing understanding of IgG autoantibodies in Sjogren's disease and could inform future treatment strategies for this complex, progressive and underserved condition."

More information on the data presented at the EULAR 2025 meeting can be found here.
Details for presentations are as follows:

Title	Presenter	Presentation
Efficacy and Safety of Efgartigimod PH20 SC in Adult Participants with Active Idiopathic Inflammatory Myopathy: Phase 2 Results from the ALKIVIA Study	Hector Chinoy	Oral Presentation #OP0002 Session: Abstract Plenary Wednesday, June 11 16:40-16:50 CEST
Treatment of Sjögren's disease by blocking FcRn: clinical and translational data from Rho, a phase 2 randomized, placebo controlled, double-blind, proof-of-concept study with efgartigimod	Isabelle Peene	Oral Presentation #OP0041 Session: Clinical Abstract Wednesday, June 11 16:30-16:40 CEST
Efficacy and safety of efgartigimod PH20 subcutaneous by prefilled syringe in adults with Sjögren's disease: A Phase 3, randomized, double-blind, placebo-controlled, multicenter trial with open-label extension (UNITY)	Simon Bowman	Poster #POS0844 Poster View III Thursday, June 12 12:00-13:30 CEST
Safety, tolerability, and efficacy of empasiprubart in adults with dermatomyositis (EMPACIFIC): A Phase 2, randomized, double-blind, placebo-controlled, multicenter study	Tetyana Storie	Poster #POS1049 Poster View VI Friday, June 13 12:00-13:30 CEST

ALKIVIA Study Design
The ALKIVIA study is a randomized, double-blind, placebo-controlled, multicenter, operationally seamless Phase 2/3 study of efgartigimod SC for the treatment of idiopathic inflammatory myopathies (IIM or myositis) across three subtypes, including immune-mediated necrotizing myopathy (IMNM), anti-synthetase syndrome (ASyS), and dermatomyositis (DM). The ALKIVIA study enrolled 240 patients in total and is being conducted in two phases, with an analysis of the Phase 2 portion of the clinical trial after the first 90 patients completed the study, followed by a Phase 3 portion if a signal is observed in the Phase 2 portion. The primary endpoint is the mean total improvement score (TIS) at the end of the treatment period (24 weeks in Phase 2 and 52 weeks in Phase 3) of all treated patients (IMNM, ASyS, DM) compared to placebo. Key secondary endpoints include response rates at the end of treatment, time to response, and duration of response in TIS, as well as change from baseline in individual TIS components. Other secondary endpoints include quality of life and other functional scores.

About Idiopathic Inflammatory Myopathies
Idiopathic inflammatory myopathies (myositis) are a rare group of autoimmune diseases that can be muscle specific or affect multiple organs including the skin, joints, lungs, gastrointestinal tract and heart. Myositis can be very severe and disabling and have a material impact on quality of life. Initially, myositis was classified as either DM or polymyositis, but as the underlying pathophysiology of myositis has become better understood, including through the identification of characteristic autoantibodies, new polymyositis subtypes have emerged. Two of these subtypes are IMNM and ASyS. Proximal muscle weakness is a unifying feature of each subtype. IMNM is characterized by skeletal muscle weakness due to muscle cell necrosis. ASyS is characterized by muscle inflammation, inflammatory arthritis, interstitial lung disease, thickening and cracking of the hands ("mechanic's hands") and Raynaud's phenomenon. DM is characterized by muscle inflammation and degeneration and skin abnormalities, including heliotrope rash, Gottron's papules, erythematous, calcinosis and edema.

RHO Study Design
The Phase 2 RHO study was a randomized, double-blinded, placebo-controlled multicenter proof of concept study to evaluate the safety and efficacy of efgartigimod in adults with Sjogren's Disease. In order to enter the study, patients needed to test positive for anti-Ro autoantibodies and maintain residual salivary flow. Thirty four patients were randomized 2:1 to receive either efgartigimod or placebo for up to 24 weeks. Multiple endpoints and biomarkers were evaluated in the signal-finding study, including the primary endpoint of CRESS (Composite of Relevant Endpoints for Sjogren's Syndrome). Within CRESS there are five components spanning: systemic disease activity as measured by the ESSDAI (EULAR Sjogren's Syndrome Activity Index), patient reported outcomes as measured by the ESSPRI (EULAR Sjogren's Syndrome Patient Reported Index), tear and salivary gland function and serology. To be a CRESS responder, patients needed to demonstrate a clinically meaningful benefit in at least 3 of the 5 composite items. Additional datapoints were gathered including the clinESSDAI, STAR (Sjogren's Tool for Assessing Response), biomarker data, and the change in lymphocytic infiltrate levels through parotid biopsies.

About Sjogren's Disease
Sjogren's Disease (SjD) is a chronic, slowly progressive inflammatory systemic autoimmune disease characterized by immune-mediated destruction of exocrine glands. SjD can be severely debilitating and have a negative impact on patient quality of life, with common symptoms reported as dry eyes and mouth, fatigue, and joint point. In addition, a substantial subset of patients suffer from extraglandular systemic disease. While the presence of anti-Ro and anti-La IgG autoantibodies are considered a hallmark of disease, the underlying cause of SjD is believed to be multi-factorial, triggered by environmental factors, leading to autoimmunity and chronic inflammation. SjD predominantly impacts women with a 9:1 female:male incidence ratio. Given the heterogeneous nature of the disease, the treatment journey can be challenging with long delays and high rates of misdiagnosis. There are no FDA- approved treatments targeting the disease itself, leaving current treatments to focus primarily on individual symptom management.

About Efgartigimod
Efgartigimod (efgartigimod alfa and hyaluronidase-qvfc) is a human IgG1 antibody fragment designed to reduce pathogenic immunoglobulin G (IgG) antibodies by binding to the neonatal Fc receptor (FcRn) and blocking the IgG recycling process. Efgartigimod is the first-approved FcRn blocker globally and is marketed as VYVGART® and VYVGART® Hytrulo in the United States and China for the treatment of generalized myasthenia gravis (gMG) and chronic inflammatory demyelinating polyneuropathy (CIDP), and as VYVDURA (Japan) or VYVGART SC for gMG in other regions globally. Efgartigimod is currently being evaluated in more than 15 severe autoimmune diseases where pathogenic IgGs are believed to be mediators of disease.

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 and is evaluating its broad potential in multiple serious autoimmune diseases while advancing several earlier stage experimental medicines within its therapeutic franchises. For more information, visit www.argenx.com and follow us on LinkedIn, Instagram, Facebook, and YouTube.

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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 "aim," "are," "believe," "can," "commit," and "will" and include statements argenx makes concerning its commitment to improving the lives of people suffering from severe autoimmune diseases and to new therapeutic areas in rheumatology; the discussion of its Phase 2 proof-of-concept results as well as the ongoing Phase 3 studies for efgartigimod in myositis and Sjogren's disease at EULAR 2025, including the planned agenda of such congress; its ability to rapidly advance development in rheumatic diseases; its goal to have efgartigimod not just manage symptoms but target the underlying disease and its potential as a precision therapy for myositis and Sjogren's patients; the potential for efgartigimod to be a transformative targeted treatment approach; and its hope that efgartigimod leads to improved outcomes and offer a new therapeutic options for such patients. 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 but not limited to, 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; the acceptance of its products and product candidates by its patients as safe, effective and cost-effective; the impact of governmental laws and regulations, including tariffs, export controls, sanctions and other regulations on its business; its reliance on 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.