argenx to present complete data from the Phase 2 proof-of-concept trial of efgartigimod (ARGX-113) in

- Eight-week follow-up data show separation of clinical efficacy scores between treatment group and placebo group through the duration of study

- Total and pathogenic IgG reduction correlates with disease score improvements

Company to host workshop and webcast today at 1:00 p.m. PT

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Breda, the Netherlands / Ghent, Belgium - argenx (Euronext & Nasdaq: ARGX), a clinical-stage biotechnology company developing a deep pipeline of differentiated antibody-based therapies for the treatment of severe autoimmune diseases and cancer, today announced that it will present complete data from the Phase 2 proof-of-concept trial of efgartigimod (ARGX-113) in generalized myasthenia gravis (MG) patients at the 2018 American Academy of Neurology (AAN) Annual Meeting in Los Angeles, CA. These data will be presented during the Clinical Trial Plenary Session by James F. Howard Jr., M.D., principal investigator on the trial and Distinguished Professor of Neuromuscular Disease, Professor of Neurology, Medicine & Allied Health, and Chief, Neuromuscular Disorders Section, The University of North Carolina School of Medicine.

"We are very encouraged by the full set of data on efgartigimod that will be presented today at AAN, particularly the correlation of total and pathogenic IgG reduction and clinical response. The data show an early separation between treatment and placebo groups on efficacy scores that persisted for the total duration of the study. We believe this may be as a differentiator from current therapies for managing IgG levels, including plasmapheresis, where benefit reversed more rapidly," commented Nicolas Leupin, CMO of argenx. "We continue to learn more about the novel mechanism of action of our drug candidate and look forward to reporting data from two additional indications this year, immune thrombocytopenia and pemphigus vulgaris, which like MG, are diseases mediated by pathogenic IgGs."

The tolerability of efgartigimod remained consistent with findings from the Phase 1 trial in healthy volunteers. The study drug candidate was well-tolerated in all patients with no serious or severe adverse events reported, and most adverse events were characterized as mild and deemed unrelated to the drug candidate.

Key Highlights from Full Phase 2 Dataset

- Full efficacy data through the eight-week follow-up phase show that administration of efgartigimod resulted in clinical improvement over the placebo through the entire duration of study (11 weeks). Clinical benefit in the efgartigimod treatment group maximized as of one week after the administration of the last dose, achieving statistical significance over the placebo group ($p = 0.0356$) on the Myasthenia Gravis Activity-of-Daily-Living (MG-ADL) score.
    - 75% of patients treated with efgartigimod had a clinically meaningful and statistically significant improvement in MG-ADL scores (at least a two-point reduction from baseline) for a period of at least six consecutive weeks, versus 25% of patients on the placebo ($p = 0.0391$).
    - Increasing differentiation was observed between the efgartigimod treatment group versus the placebo group, with increasing MG-ADL thresholds. Updated results will include the differentiation between the treatment and placebo groups for both the MG-ADL and Quantitative Myasthenia Gravis (QMG) thresholds at the 29-day point and the 36-day point.

    - Patients in the treatment arm showed disease improvement, with separation from the patients in the placebo group one week after the first infusion that persisted after the last dose.
    - Efgartigimod treatment resulted in clinical improvement over the placebo, as measured by all four predefined clinical efficacy scales - MG-ADL, QMG, Myasthenia Gravis Composite (MGC) and Myasthenia Gravis Quality of Life (MG-QoL).
- All patients in the treatment arm showed a reduction of total IgG levels. Clinically meaningful disease improvement was found to correlate with reduction in pathogenic IgG levels.
    - Total IgG reduction in patients was consistent with the Phase 1 healthy volunteer trial.
    - Reduction of IgG levels was consistent across IgG subtypes, including AChR autoantibodies (IgG1 and IgG3).
    - Updated results show mean maximum IgG reduction of up to 70.7% among treated patients.

argenx is conducting two additional ongoing Phase 2 clinical trials of efgartigimod in immune thrombocytopenia (ITP) and pemphigus vulgaris (PV). Topline data from the ITP trial and interim data from the PV trial are both expected in the second half of 2018.

An investor workshop is being held today at 1:00 p.m. PT in Los Angeles to discuss the complete efgartigimod clinical data presented by Dr. Howard. A live webcast of the presentation will be available on the Company's website www.argenx.com or by clicking here. A replay of the webcast will be available for 90 days following the presentation.

Phase 2 Trial Design
The Phase 2 trial evaluated 24 MG patients with generalized muscle weakness, and a total MG-ADL score >=5, with more than 50% of the score consisting of non-ocular items. Patients were randomized to receive four weekly doses of either standard of care plus 10 mg/kg of ARGX-113, or standard of care plus placebo. Standard of care therapies included acetylcholinesterase inhibitors, corticosteroids and/or immunomodulatory agents. The primary endpoints of the trial were safety and tolerability. Secondary endpoints included efficacy as measured by the change from baseline of the MG-ADL, QMG, and MGC disease severity scores; impact on quality of life as measured by the MGQoL score; and an assessment of pharmacokinetics (PK) and pharmacodynamic (PD) markers and immunogenicity.

About efgartigimod
Efgartigimod (ARGX-113) is an investigational therapy for IgG-mediated autoimmune diseases and was designed to exploit the natural interaction between IgG antibodies and the recycling receptor FcRn. ARGX-113 is the Fc-portion of an antibody that has been modified by the argenx proprietary ABDEG(TM) technology to increase its affinity for FcRn beyond that of normal IgG antibodies. As a result, ARGX-113 blocks antibody recycling through FcRn binding and leads to fast depletion of the autoimmune disease-causing IgG autoantibodies. The development work on ARGX-113 is done in close collaboration with Prof. E. Sally Ward (University of Texas Southwestern Medical and Texas A&M University Health Science Center, a part of Texas A&M University (TAMHSC)).

About argenx
argenx is a clinical-stage biotechnology company developing a deep pipeline of differentiated antibody-based therapies for the treatment of severe auto-immune diseases and cancer. We are focused on developing product candidates with the potential to be either first-in-class against novel targets or best-in-class against known, but complex, targets in order to treat diseases with a significant unmet medical need. Our ability to execute on this focus is enabled by our suite of differentiated technologies. Our SIMPLE Antibody™ Platform, based on the powerful llama immune system, allows us to exploit novel and complex targets, and our three antibody engineering technologies are designed to enable us to expand the therapeutic index of our product candidates.
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