argenx reports positive topline results from Phase 2 proof-of-concept trial of ARGX-113 (efgartigimod) in generalized myasthenia gravis

- Favorable tolerability profile consistent with Phase 1 data
- ARGX-113 treatment resulted in a strong clinical improvement over placebo during the entire duration of the study
- 75% of ARGX-113 treated patients had a clinically meaningful and statistically significant improvement through at least 6 consecutive study weeks versus 25% of patients on placebo
- Management to host conference call today at 8:00 am EST

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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 positive topline results from its Phase 2 proof-of-concept clinical trial of ARGX-113 (efgartigimod) in myasthenia gravis (MG) patients with confirmed generalized muscle weakness. The data showed a clinically meaningful and statistically significant benefit of ARGX-113 over placebo. In addition, ARGX-113 was found to have a favorable tolerability profile consistent with that observed in the Phase 1 study.

The results will be discussed during a conference call and webcast presentation today at 2:00 pm CET/ 8:00 am EST. The conference call can be accessed by dialing 1-631-510-7498 in the U.S. or selecting from the numbers below if international. The confirmation code is **5472459**. The webcast may be accessed on the homepage of the argenx website at www.argenx.com or by clicking here.

"There remains a clear unmet need for a safe and fast-acting treatment for patients with generalized MG, who continue to face serious, potentially life-threatening symptoms associated with their disease," said Professor James F. Howard Jr., MD, Distinguished Professor of Neuromuscular Disease, professor of neurology, medicine and allied health, and chief of the Neuromuscular Disorders Section in the University of North Carolina School of Medicine, Chapel Hill, NC USA. "These data demonstrate a rapid and sustained benefit in disease score after treatment with ARGX-113, supporting further development of the drug as a potential new option to fill the current treatment gap for MG patients."

Phase 2 Trial Design
The Phase 2 double-blind, placebo-controlled, proof-of-concept trial enrolled 24 MG patients with generalized muscle weakness, and a total Myasthenia Gravis Activity-of-Daily-Living (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 corticosteroids and/or immunomodulatory agents. The primary endpoints of the study were safety and tolerability. Secondary endpoints included efficacy as measured by the change from baseline of the MG-ADL, Quantitative Myasthenia Gravis (QMG), and Myasthenia Gravis Composite (MGC) disease severity scores; impact on quality of life as measured by the Myasthenia Gravis Quality of Life (MGQoL) score; and an assessment of pharmacokinetics (PK) and pharmacodynamic (PD) markers. All 24 patients were evaluable.

Top-line Results
Primary endpoint analysis revealed ARGX-113 to be well tolerated in all patients, with most adverse events (AEs) characterized as mild and deemed unrelated to the study drug. No serious or severe AEs were reported. The observed tolerability profile is consistent with the Phase 1 healthy volunteer study.

The secondary endpoint measures relating to efficacy showed ARGX-113 treatment resulted in rapid onset of action and strong clinical improvement over placebo during the entire duration of the study. Specifically, we observed that:

- 75% of patients treated with ARGX-113 had a clinically meaningful and statistically significant improvement in MG-ADL scores (at least a 2-point reduction from baseline) for a period of at least 6 consecutive weeks versus 25% of patients on placebo (p = 0.0391).
- Clinical benefit in the ARGX-113 treatment group maximized as of 1 week after the administration of the last dose, achieving statistical significance over the placebo group (p = 0.0356) on the MG-ADL score. Increasing differentiation was observed between the ARGX-113 treatment group versus placebo with increasing MG-ADL thresholds.
- Patients in the treatment arm showed rapid disease improvement, with clear separation from placebo 1 week after the first infusion.
- All patients in the treatment arm showed a rapid and deep reduction of their total IgG levels and disease improvement was found to correlate with reduction in pathogenic IgG levels.
- ARGX-113 demonstrated strong clinical improvement over placebo as measured by all four predefined clinical efficacy scales - MG-ADL, QMG, MGC and MG-QoL15.

"These results strengthen our conviction that reducing pathogenic autoantibodies may offer an innovative approach to treat myasthenia gravis and could give rise to potential therapeutic benefits in other neuromuscular conditions that are similarly mediated. Further, through our deeper understanding of the drug's mechanism, we see promise of its potential across other disease categories as well, including autoimmune blood disorders or skin blistering diseases which we are evaluating in our two ongoing Phase 2 studies in immune thrombocytopenia and pemphigus vulgaris," commented Nicolas Leupin, Chief Medical Officer of argenx. "We look forward to refining our plan forward and optimizing the broad potential of ARGX-113."

argenx plans to present the full data from the trial at the American Academy of Neurology annual meeting (Los Angeles, April 21-27, 2018).

argenx is conducting two additional ongoing Phase 2 clinical trials of ARGX-113 in immune thrombocytopenia (ITP) and pemphigus vulgaris (PV). Topline data for the ITP trial and interim data from the PV trial are both expected in the second half of 2018.

About ARGX-113
ARGX-113 (efgartigimod) 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. It consists of the Fc-portion of an antibody that has been modified by the argenx proprietary ABDEG(TM) technology. ARGX-113 blocks antibody recycling through FcRn binding and induces rapid 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).

About argenx
argenx is a clinical-stage biotechnology company developing a deep pipeline of differentiated antibody-based therapies for the treatment of severe autoimmune 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(TM) 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.
www.argenx.com

Dial-in numbers:
Please dial in 5-10 minutes prior to 2:00 pm CET/ 8:00 am EST using the number and conference ID below.

Confirmation Code: 5472459

- Standard International Dial-In Number: +44 (0) 1452 555566
- Local Call Dial-In Numbers:

United States, New York	16315107498
United Kingdom	08444933800
Belgium	081700061
Austria	019286568
Finland	0923195187
France	0176742428
Germany	06922224918
Italy	0236008146
Luxembourg	20880695
Netherland	0207176886
Spain	914143669
Sweden	0850336434
Switzerland	0565800007

A question and answer session will follow the presentation of the results. Go to www.argenx.com to access the live audio webcast. The archived webcast will also be available (30 days) for replay shortly after the close of the call from the "Downloads" section of the argenx website.

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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," "estimates," "anticipates," "expects," "intends," "may," "will," or "should," and include statements argenx makes concerning the encouraging clinical data of ARGX-113; argenx's advancement of, and anticipated clinical development and regulatory milestones and plans related to ARGX-113; and the intended results of its strategy. 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 argenx's expectations regarding its the inherent uncertainties associated with competitive developments, preclinical and clinical trial and product development activities and regulatory approval requirements; argenx's reliance on collaborations with third parties; estimating the commercial potential of argenx's product candidates; argenx's ability to obtain and maintain protection of intellectual property for its technologies and drugs; argenx's limited operating history; and argenx's ability to obtain additional funding for operations and to complete the development and commercialization of its product candidates. 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 the final prospectus related to argenx's initial U.S.

public offering filed with the SEC pursuant to Rule 424(b) of the Securities Act of 1933, as amended, 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.