argenx Announces 2021 Corporate Priorities and Highlights Recent Achievements Across Immunology Pipeline

- Submitted Biologics License Application (BLA) to U.S. Food and Drug Administration (FDA) for efgartigimod in generalized myasthenia gravis (gMG)
- Initiated 50-patient gMG bridging study of subcutaneous (SC) efgartigimod based on FDA feedback
- Enrolled first 30 patients, necessary for go/no-go decision, in ADHERE trial of SC efgartigimod for chronic inflammatory demyelinating polyneuropathy (CIDP)
- Announced interim data from Phase 2 CULMINATE trial of cusatuzumab in development with Janssen

Breda, the Netherlands – Jan. 8, 2021 – argenx (Euronext & Nasdaq: ARGX), a global immunology company committed to improving the lives of people suffering from severe autoimmune diseases and cancers, today announced its 2021 corporate priorities and highlighted recent achievements from its late-stage immunology pipeline driven by its FcRn antagonist, efgartigimod. Additionally, the Company announced interim data from the Phase 2 CULMINATE trial of cusatuzumab in development with Cilag GmbH International, an affiliate of Janssen, and provided financial guidance for 2021.

argenx previously announced an exclusive license agreement with Zai Lab Limited ("Zai Lab") for the development and commercialization of efgartigimod in Greater China and the acceleration of efgartigimod development through Phase 2 proof-of-concept trials in new autoimmune indications. Zai Lab will also contribute Chinese patients to argenx's global Phase 3 trials of efgartigimod. Under the terms of the agreement, argenx will receive $175 million in collaboration payments comprised of upfront Zai Lab equity, a guaranteed development cost-sharing payment, and a milestone payment upon U.S. efgartigimod approval. argenx will also be eligible for tiered royalties based on annual net sales of efgartigimod in Greater China.

"We are excited to enter a new chapter for argenx as we look toward commercialization and achieving our mission of reaching patients with debilitating rare diseases. We've submitted a BLA to the FDA for efgartigimod in gMG and expect to have global efgartigimod trials ongoing this year in six indications and two formulations. We hope to continue to demonstrate the broad opportunity of our FcRn antagonist within autoimmune diseases in 2021 and beyond," said Tim Van Hauwermeiren, Chief Executive Officer of argenx. "In parallel, establishing global commercial infrastructure within the U.S. and Japan continues to be a top priority. Now through our collaboration with Zai Lab in China and with the appointment of a general manager in Europe, we've solidified and accelerated our capabilities to bring efgartigimod and our future immunology candidates to patients worldwide."

2021 Corporate Priorities and Recent Progress

The Company will continue its transition to a fully integrated immunology company by executing on three corporate priorities in 2021, including: preparation for the potential FDA approval and U.S. commercial launch of efgartigimod for the treatment of patients with gMG; the progression of its clinical-stage autoimmune pipeline; and the continued growth of its broad and differentiated pipeline through its Immunology Innovation Program.

1. **Preparation for potential FDA approval and global commercial launch of efgartigimod for the treatment of patients with gMG**

    - Submitted BLA to FDA for efgartigimod for treatment of gMG and continued preparations for other global regulatory submissions
        - On track to submit application to Japan's Pharmaceuticals and Medical Devices Agency (PMDA) in first half of 2021
        - On track to submit to European Medicines Agency (EMA) in second half of 2021
        - Submission in China expected to occur shortly following potential approval in U.S.

    - Initiated bridging study for SC efgartigimod in gMG based on association between total IgG reduction and clinical benefit, and feedback from the FDA. The study is a registrational, non-inferiority trial comparing the pharmacodynamic effect of 1000mg SC efgartigimod with 10mg/kg IV efgartigimod and is expected to enroll approximately 50 patients.

    - Commercial preparation activities are underway and on track for potential 2021 launch, including continued engagement with key stakeholders, commercial inventory build, milestone-based hiring of field force around potential BLA acceptance and FDA approval, and development of a patient services program.

    - Appointed Anant Murthy, Ph.D., as General Manager of argenx Europe. In this role, Dr. Murthy will establish the commercial infrastructure for a European launch and lead market development activities in advance of a potential European Medicines Agency (EMA) approval of efgartigimod. Dr. Murthy brings ~20 years of international experience to argenx, most recently as Head of Market Access for EMEA and Canada and the General Manager of multiple European countries for Alnylam Pharmaceuticals.

2. **Progress clinical-stage autoimmune pipeline, including seven expected global trials of efgartigimod and Phase 1 trial of first-in-class C2 antibody ARGX-117**

    - ADVANCE (IV) and ADVANCE SC trials ongoing evaluating IV and SC efgartigimod in patients with primary immune thrombocytopenia (ITP); global program expected to support registration of both formulations

    - ADDRESS registrational trial ongoing evaluating SC efgartigimod in patients with pemphigus vulgaris (PV) and pemphigus foliaceous (PF)

- ADHERE trial ongoing evaluating SC efgartigimod in chronic inflammatory demyelinating polyneuropathy (CIDP);
  - Completed enrollment of first 30 patients
  - Decision whether to expand enrollment up to 130-140 patients expected in first quarter of 2021

- Clinical trials in fifth and sixth indications of efgartigimod to begin enrollment in 2021

- Ongoing Phase 1 healthy volunteer trial of IV and SC ARGX-117, a first-in-class C2 antibody, to evaluate safety and tolerability and establish dosing regimen
  - Data expected in mid-2021, after which argenx plans to launch Phase 2 proof-of-concept trials in severe autoimmune diseases, including multifocal motor neuropathy (MMN)

3. **Continued investment in broad and differentiated pipeline through Immunology Innovation Program**

- Preclinical work ongoing in early-stage pipeline, including continued progress on ARGX-118 and ARGX-119, and the optimization of ARGX-120

- Commitment to expand pipeline at cadence of one new candidate per year from Immunology Innovation Program

Interim Data from Cusatuzumab Phase 2 CULMINATE Trial

Development of cusatuzumab in acute myeloid leukemia (AML) remains ongoing as part of a global collaboration and license agreement with Cilag GmbH International, an affiliate of Janssen.

The Phase 2 CULMINATE trial (NCT04023526) is evaluating cusatuzumab in combination with azacitidine in newly-diagnosed, elderly AML patients who are ineligible for intensive chemotherapy. A total of 103 patients were randomized to receive either 10mg/kg (n=51) or 20mg/kg (n=52) cusatuzumab plus azacitidine as part of a dose identification. The 20mg/kg dose has been selected for ongoing and future trials.

A pre-planned interim analysis was conducted of the 52 patients (46.2% adverse ELN risk classification) receiving 20mg/kg cusatuzumab plus azacitidine treatment (intent-to-treat population (ITT)). The results from the ITT analysis showed a complete remission (CR) rate of 27% (14/52) and composite complete remission (CRc), including CRs with incomplete hematologic recovery, rate of 40% (21/52). The 30-day mortality rate of the ITT population was 9.6% (5/52). In a cohort where patients received at least two treatment cycles (20mg/kg cusatuzumab plus azacitidine), 42% (14/33) achieved CR and 64% (21/33) achieved CRc.

Cusatuzumab was observed to be well-tolerated and the safety profile was consistent with prior studies. Final results from the CULMINATE trial will be presented in a peer-reviewed forum.

The decision to initiate additional studies in the development of cusatuzumab, under the collaboration, will be determined following review of data from the ongoing Phase 1b ELEVATE trial (NCT04150887), which is evaluating cusatuzumab in combination with venetoclax and azacitidine in newly-diagnosed, elderly patients with AML who are ineligible for intensive chemotherapy.

Financial Guidance

As of December 31, 2020, argenx had approximately $2.0 billion in cash, cash equivalents and current financial assets. This preliminary cash balance does not include expenses or proceeds from recently announced business development transactions, including the purchase of a priority review voucher from Bayer HealthCare Pharmaceuticals, Inc. and the exclusive license agreement with Zai Lab for efgartigimod in Greater China.

Based on current plans to fund anticipated operating expenses and capital expenditures, argenx expects its cash burn to increase significantly in 2021, approximately doubling compared to 2020. The increased spend will support the Company's transition to an integrated immunology company in 2021, including the build-out of global commercial infrastructure and drug product inventory ahead of the expected launch of efgartigimod in gMG in the U.S, the advancement of its clinical-stage pipeline, including seven expected global trials of efgartigimod, and the continued investment in its Immunology Innovation Program.

J.P. Morgan Healthcare Conference Presentation and Webcast

argenx CEO Tim Van Hauwermeiren, will present these updates at the virtual 39th Annual J.P. Morgan Healthcare Conference on Monday, January 11, 2020 at 8:20 a.m. ET, followed by a question and answer session.

The live webcast of the presentation and question and answer session that follows may be accessed on the homepage of the argenx website at www.argenx.com. A replay of the webcast will be available for 90 days on the argenx website.

About Efgartigimod

Efgartigimod is an investigational antibody fragment designed to reduce disease-causing immunoglobulin G (IgG) antibodies and block the IgG recycling process. Efgartigimod binds to the neonatal Fc receptor (FcRn), which is widely expressed throughout the body and plays a central role in rescuing IgG antibodies from degradation. Blocking FcRn reduces IgG antibody levels representing a logical potential therapeutic approach for several autoimmune diseases known to be driven by disease-causing IgG antibodies, including: myasthenia gravis (MG), a chronic disease that causes muscle weakness; pemphigus vulgaris (PV), a chronic disease characterized by severe blistering of the skin; immune thrombocytopenia (ITP), a chronic bruising and bleeding disease; and chronic inflammatory demyelinating polyneuropathy (CIDP), a neurological disease leading to impaired motor function.

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 is evaluating efgartigimod in multiple serious autoimmune diseases, and cusatuzumab in hematological cancers in collaboration with Janssen. argenx is also advancing several earlier stage experimental medicines within its therapeutic franchises. argenx has offices in Belgium, the United States, and Japan. For more information, visit www.argenx.com and follow us on LinkedIn at https://www.linkedin.com/company/argenx/.

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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 its 2021 business and financial outlook and related plans, including with respect to financial guidance and cash burn; the therapeutic potential of its product candidates; the intended results of its strategy; the expected benefits of its collaborations, including with respect to the exclusive license agreement with Zai Lab; its and its collaboration partners' clinical development and regulatory plans, including the timing, design and outcome of ongoing and planned clinical trials and preclinical activities and the timing and outcome of regulatory filings and approvals; and the timing and progress of commercialization activities. 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 effects of the COVID-19 pandemic, the inherent uncertainties associated with 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 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.

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