argenx Provides Strategic Outlook Advancing Late-Stage Pipeline Towards ‘argenx 2021’ Vision

- Enrollment complete in Phase 3 ADAPT trial of efgartigimod in gMG; topline data expected mid-2020
- Positive Phase 2 data of efgartigimod in PV support advancement to registrational trial; proof-of-concept established in three initial efgartigimod indications and therapeutic areas
- Phase 3 ADAPT trial design to be discussed during 38th Annual J.P. Morgan Healthcare Conference
- Company presentation on Tuesday, January 14, 2020 at 2:00 p.m. PST

January 9, 2020

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 is providing a strategic outlook for 2020 outlining key priorities for its broad pipeline and path towards achieving its ‘argenx 2021’ integrated commercial vision.

“We begin 2020 in an exciting position, having met all our objectives for our clinical programs. This includes the completion of enrollment of our Phase 3 ADAPT trial of efgartigimod in gMG, the launch of key efgartigimod clinical trials in ITP and CIDP, and the initiation of cusatuzumab clinical trials in two AML settings with Janssen. In addition, we’re announcing today positive proof-of-concept data for efgartigimod in PV, our third ‘beachhead’ indication, further demonstrating our initial development strategy of targeting pathogenic autoantibodies and creating commercial opportunities in several therapeutic areas. Looking forward to the remainder of 2020, we plan up to five registrational efgartigimod trials and further expansion of the cusatuzumab global development plan with Janssen,” said Tim Van Hauwermeiren, Chief Executive Officer of argenx.

“Most importantly, we are continuing to execute on the ‘argenx 2021’ vision to become a global, integrated immunology company with our first launch of efgartigimod in gMG expected in 2021. At the core of this growth strategy is a commitment to expanding our early-stage pipeline with immunology breakthroughs and advancing our late-stage candidates while extending our reach to bring first-in-class medicines to patients,” continued Mr. Van Hauwermeiren.

argenx 2021 Vision
argenx continues to execute on its plan to become a fully integrated immunology company through its “argenx 2021” vision, including building two initial commercial franchises in neuromuscular indications and hematology/oncology and expanding its global presence encompassing Boston, Ghent and Tokyo. As part of this vision, argenx highlights:

- Leadership in FcRn and its therapeutic immunology potential:
  - On track to launch first FcRn antagonist with efgartigimod in generalized myasthenia gravis (gMG) in 2021
  - Up to five registrational trials expected to be ongoing in 2020 across four targeted indications (gMG, immune thrombocytopenia (ITP), chronic inflammatory demyelinating polyneuropathy (CIDP) and pemphigus vulgaris (PV))
  - Fifth efgartigimod indication expected to be announced in 2020
  - Further research underway exploring therapeutic potential of FcRn modulation
- Launch of MyRealWorld™ MG:
  - First-of-its-kind in MG, real-world evidence study launched; aiming to enroll approximately 2000 patients globally with support from regional patient advocacy organizations
  - Disease and treatment burden to be documented to support proposed value of efgartigimod
- Strong financial foundation:
  - Following November 2019 equity offering, argenx expects to report 2019 year-end cash and cash equivalents of approximately $1.5 billion

Pipeline Updates and 2020 Priorities
argenx today is reporting positive proof-of-concept data in PV, the third beachhead indication as part of the broad efgartigimod development strategy:

- 23 patients (10mg/kg or 25mg/kg efgartigimod) were evaluated for efficacy in an adaptive Phase 2 trial aiming to establish optimal treatment regimen
  - Clear correlation demonstrated between pathogenic IgG reduction and Pemphigus Disease Area Index score improvement
  - 78% (18/23) of patients achieved rapid disease control; median time to disease control for both monotherapy and combination therapy is 14-15 days
  - Fast clinical remission (CR) observed in 70% (5/7) of patients receiving optimized dosing regimen
    - CR achieved within 2-10 weeks
    - Optimized dosing regimen determined to be at least biweekly dosing of efgartigimod in combination with oral prednisone (0.25-0.5mg/kg)
  - Potential of corticosteroid sparing demonstrated
- Independent data review committee concluded safety and tolerability to be favorable
- Patients still in trial in extended dosing cohort; detailed results of Phase 2 data to be presented during medical meeting in 2020
- Data support advancing to registrational trial expected to start in second half of 2020

Within its neuromuscular franchise, argenx is evaluating:

- Efgartigimod in gMG:
  - Enrollment completed of 167 gMG patients in Phase 3 ADAPT trial with 10mg/kg IV efgartigimod; topline results from ADAPT now expected in mid-2020
  - Biologics License Agreement (BLA) for gMG expected to be filed in fourth quarter of 2020
  - Plan to engage with U.S. Food and Drug Administration (FDA) on potential bridging strategy for 1000mg subcutaneous (SC) ENHANZE®-efgartigimod in gMG
- Efgartigimod in CIDP:
  - Phase 2 ADHERE trial initiated in CIDP with SC ENHANZE®-efgartigimod

Within its hematology/oncology franchise, argenx is evaluating:

- Efgartigimod in ITP:
  - Phase 3 ADVANCE registrational trial initiated evaluating approximately 150 primary ITP patients dosed with 10mg/kg IV efgartigimod for both induction and maintenance of platelet response
  - ADVANCE SC trial expected to initiate in second half of 2020 evaluating 10mg/kg IV efgartigimod for induction of platelet response and fixed dose of SC efgartigimod for maintenance
  - Additional small confirmatory IV trial as part of registrational ITP program expected to initiate in first half of 2020
  - Orphan designation granted by European Medicines Agency (EMA) for ITP; designation granted by FDA in February 2019
- Cusatuzumab in collaboration with Janssen:
  - CULMINATE trial in combination with azacitidine to continue enrollment of newly diagnosed “unfit” AML patients
  - Phase 1b platform trial underway in various AML subpopulations and settings with initial trial evaluating combinations of cusatuzumab, venetoclax and azacitadine; additional trials expected to launch under platform trial in first half of 2020
  - Randomized Phase 2 trial in higher-risk myelodysplastic syndromes (MDS) expected to launch in first half of 2020
  - Data update from cusatuzumab development expected in 2020

argenx continues to expand its early-stage pipeline with first-in-class antibodies against immunologic targets:

- Phase 1 trial of ARGX-117 in healthy volunteers expected to begin in first quarter of 2020
  - Multiple doses and formulations (IV and SC with Halozyme ENHANZE® technology) to be evaluated as part of dose-finding work
  - Following analysis of Phase 1 data in fourth quarter of 2020, argenx expects to launch Phase 2 program in multifocal motor neuropathy (MMN) within its neuromuscular franchise and develop in additional indications
- Lead optimization work on ARGX-118 for airway inflammation to continue in 2020
- New product candidate ARGX-119 expected to be announced in 2020

JP Morgan Conference Presentation and Webcast

Tim Van Hauwermeiren, Chief Executive Officer, will present at the 38th Annual J.P. Morgan Healthcare Conference on Tuesday, January 14, 2020 at 2:00 p.m. PST in San Francisco, followed by a breakout question and answer session.

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

About efgartigimod
Efgartigimod is an IgG Fc fragment engineered to optimally antagonize the neonatal Fc Receptor (FcRn) for the treatment of IgG-mediated autoimmune diseases. FcRn plays a central role in rescuing IgG from degradation in the lysosome through a recycling pathway. Through inhibition of FcRn, efgartigimod leads to fast depletion of the disease-causing IgG autoantibodies. Efgartigimod binds in the same way as endogenous IgG, the natural ligand of FcRn, and has been engineered with ABDEG mutations to increase its affinity for FcRn while preserving the characteristic pH-dependent binding, contributing to its long serum half-life, pharmacodynamic effect and potentially enhanced tissue penetration. The development work on efgartigimod is conducted in close collaboration with Prof. E. Sally Ward (Department of Molecular and Cellular Medicine, Texas A&M University Health Science Center, College Station, TX; Center for Cancer Immunology, University of Southampton, Southampton, UK).

About argenx
argenx is a global immunology company developing antibody-based medicines for patients suffering from severe autoimmune diseases and cancer. By translating immunology breakthroughs into innovative drug candidates, argenx is building a world-class portfolio of first-in-class antibodies in both early and late clinical-stages of development. argenx is evaluating efgartigimod in multiple serious autoimmune indications and cusatuzumab in hematological malignancies in collaboration with Janssen, along with advancing earlier stage assets within its therapeutic franchises.
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