argenx announces pipeline expansion and “argenx 2021” vision to build integrated immunology company at 2019 R&D Day

Company Name: ARGENX SE Market: Euronext ISIN: NL0010832176

Symbol: ARGX

RELEASE 22 MAY 2019 07:00 CET

- Expands pipeline with ARGX-117 and ARGX-118; two new pipeline candidates from its Innovative Access Program addressing first-in-class targets
- Highlights formation of two global therapeutic franchises in neuromuscular and hematology with targeted first commercial launch in 2021
- Appoints Wim Parys, M.D. as Chief Medical Officer, formerly of Janssen

May 22, 2019

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, will be hosting its second R&D Day today, Wednesday, May 22, 2019, in New York starting at 8:30am ET. During the event, argenx will present data on two new pipeline candidates that emerged from its Innovative Access Program (IAP) and will provide updates on the development plan of cusatuzumab (ARGX-110) in collaboration with Cilag GmbH International, an affiliate of the Janssen Pharmaceutical Companies of Johnson & Johnson. Today, argenx will also bring forward its “argenx 2021” vision, providing clarity on its strategy to become a fully integrated immunology company.

“At argenx we are making comprehensive investments across the organization and planning for nothing less than building the next great integrated immunology company, which we will carry out through our “argenx 2021” vision. We are putting all the elements in place for a sophisticated U.S. launch of efgartigimod for generalized myasthenia gravis patients, if approved. We are also working closely with Janssen to advance cusatuzumab into a broad development plan, starting with a first Phase 2 registration-directed trial in acute myeloid leukemia (AML) that is expected to start in the second half of 2019,” commented Tim Van Hauwermeiren, Chief Executive Officer of argenx.

"Today we will unveil two new and exciting candidates from our Innovative Access Program that will further enrich our immunology pipeline and provide us with ambitious therapeutic opportunities. As we strive to become a commercial organization, it is this R&D engine that is a key success differentiator of argenx, having allowed us to repeatedly identify first-in-class targets from our academic collaborators and translate these important targets into significant and sustainable value for our company and for our stakeholders."

argenx 2021 Vision

argenx today is announcing its plan to become a fully integrated, novel immunology company through its "argenx 2021" vision, which will include the building of two commercial franchises in neuromuscular and hematology. Within the two franchises are currently three expected pipeline candidates, including:

- Efgartigimod (ARGX-113) with the potential to address generalized myasthenia gravis (gMG) and chronic inflammatory demyelinating polyneuropathy within neuromuscular, and immune thrombocytopenia within hematology;
- Cusatuzumab with the potential to address acute myeloid leukemia (AML), high-risk myelodysplastic syndromes (MDS) and other hematological malignancies within the hematology franchise;
- Newly announced ARGX-117 with the potential to address multiple indications within both franchises.

By the end of 2021, argenx expects to launch efgartigimod in the U.S. in its first indication gMG, if approved. Through the building of commercial franchises, argenx plans to leverage capabilities and an organizational footprint for subsequent potential launches across its broad immunology pipeline.

Pipeline Expansion

argenx today announced the addition of two new therapeutic candidates, ARGX-117 and ARGX-118, to its proprietary antibody pipeline. Both emerged from argenx's IAP, in which it collaborates closely with academic experts, bringing the argenx cutting-edge antibody discovery technologies to the heart of novel target research.

ARGX-117 Targeting C2

ARGX-117 is a next-generation complement-targeting antibody against C2, an important component of both the classical and lectin pathways in the complement cascade. ARGX-117 has the following differentiated features:

- Unique design to precisely intervene at C2 in the complement cascade with the intention to balance optimal tolerability and activity
- Expected attractive pharmacokinetic and pharmacodynamic properties derived from proprietary antibody engineering that could potentially translate to convenient dosing schedule
- Pipeline-in-a-product opportunity with potential therapeutic applications in an array of complement-mediated diseases that fit within established franchises in neuromuscular and hematology, as well as in kidney indications

ARGX-117 was developed under a collaboration with the University Medical Center Utrecht/Broteio Pharma and was exclusively licensed by argenx in 2018. First-in-human clinical studies are expected to start in the first quarter of 2020.

argenx also announced that it has exercised its second exclusive license to Halozyme's ENHANZE® technology for ARGX-117 target C2. Under the terms of the collaboration, argenx will access Halozyme's well-established subcutaneous delivery technology for ARGX-117 to provide dosing optionality to patients. In exchange, argenx will pay Halozyme $10M for access to the second target in addition to potential future payments of up to $160 million, subject to achievement of specified milestones. Halozyme will also receive mid-single digit royalties on any future sales of commercialized products.

ARGX-118 Targeting Galectin-10

argenx is announcing today that it has exercised its option to exclusively acquire rights to ARGX-118, a highly differentiated antibody against Galectin-10, the protein of Charcot-Leyden crystals, which are implicated as a major contributor to severe asthma and to the persistence of mucus plugs. ARGX-118 has the following differentiated features:

- Acts on novel target intended to address mucus plugging, a large unmet need in airway inflammation
- Unique mechanism of action with observed crystal-dissolving properties
- Broad potential in severe airway inflammation diseases where mucus plugging plays a key role, including lung attack or asthma exacerbation, allergic bronchopulmonary aspergillosis, and chronic rhinosinusitis with nasal polyps

ARGX-118 was developed under a collaboration with VIB, a life sciences research institute based in Flanders, Belgium. The molecule is in the final stages of lead optimization work.

Appointment of Chief Medical Officer

argenx announced today the appointment of Wim Parys M.D. as Chief Medical Officer effective July 1, 2019. Dr. Parys will succeed outgoing Chief Medical Officer Nicolas Leupin, M.D. who will be departing from the Company. In his role, Dr. Parys will lead argenx's clinical development, clinical operations, regulatory affairs, pharmacovigilance and project management. Dr. Parys has served as an R&D consultant to argenx since February 2019.

Dr. Parys held several R&D leadership roles of increasing responsibility at Janssen, J&J and Tibotec, including Head of Development at Tibotec where he established and led Tibotec Inc. USA and Head of Development of Janssen's Infectious Diseases and Vaccines therapeutic area. Through his career, he led the development and regulatory submission of seven now-approved drugs. Most recently, Dr. Parys served as Head of R&D of the Global Public Health group of Janssen.

"With over 25 years of experience leading successful clinical programs and sophisticated regulatory submissions, we are confident Wim is the right leader to advance our rich pipeline at this pivotal time of growth for argenx. We believe his strategic vision will strengthen our clinical development activities and provide us key guidance as we advance to filing for our first drug approval," continued Mr. Van Hauwermeiren.

"We extend our deepest gratitude to Nicolas for his significant contributions and leadership to argenx over the last four years. He joined the Company as we were launching the first efgartigimod patient trials and his innovative thinking led to the successful late-stage results for this molecule as well as progress throughout our pipeline. We're confident his talents will lead to continued accomplishments."

Cusatuzumab Development Plan

argenx today announced that its partner Janssen intends to initiate in the second half of 2019 its first Phase 2 and registration-directed clinical trial of cusatuzumab under the companies' strategic collaboration. Cusatuzumab is an anti-CD70 monoclonal antibody for AML, high-risk MDS and other hematological malignancies. Details of the trial are as follows:

- Expected to enroll up to 150 patients with previously untreated AML who are not eligible for intensive chemotherapy
- Two-part trial: a dose selection part followed by a safety and efficacy part at the selected go-forward dose
- In the first part of the study, patients will be randomized to two dose levels of cusatuzumab (10mg/kg and 20mg/kg) in combination with azacytidine. The second part of the study is an expansion cohort at the selected dose to evaluate efficacy and safety of the combination

R&D Day Details

argenx will host its second R&D day today, Wednesday, May 22, 2019, in New York with presentations starting at 8:30am ET.

In addition to argenx management, guest speakers for the event will include:

- Prof. Erik Hack, M.D., Ph.D., Professor of Immunology, University Medical Center Utrecht
- Prof. Ludo van der Pol, M.D., Ph.D., Associate Professor, Utrecht University, Neurologist, University Medical Center Utrecht
- Prof. Bart Lambrecht, M.D., Ph.D., Director, VIB Center for Inflammation Research at Ghent University
- Dr. Rafael N. Villicana, M.D., Associate Professor, Medical Director, Kidney Transplantation, Loma Linda University Medical Center

Webcast information:

A live webcast of today's presentation will be available on the Company's website at www.argenx.com or via this link. A replay of the webcast will be available for 90 days following the presentation.

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. The company is 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. argenx's ability to execute on this focus is enabled by its suite of differentiated technologies. The SIMPLE Antibody™ Platform,

based on the powerful llama immune system, allows argenx to exploit novel and complex targets, and its three complementary Fc engineering technologies are designed to expand the therapeutic index of its product candidates.
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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 intended results of its strategy; argenx's advancement of, and anticipated clinical development, data readouts and regulatory milestones and plans, including the timing of planned clinical trials and expected data readouts, related to cusatuzumab, ARGX-117, including its first-in-human studies, and ARGX-118; the momentum of its product candidate pipeline; and its plans to become an integrated immunology company as outlined in the section titled "argenx 2021 Vision," including the building of two commercial franchises in neuromuscular and hematology. 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 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.