Safe Harbor: Certain statements contained in this presentation, other than present and historical facts and conditions independently verifiable at the date hereof, may constitute forward-looking statements. Examples of such forward-looking statements include those regarding our investigational product candidates and preclinical studies and clinical trials, and the status, plans, timing of expected data readouts and related presentations and related results thereof, including the design of our trials and the availability of data from them, the timing and achievement of our product candidate development activities, future results of operations and financial positions, including potential milestones, business strategy, plans and our objectives for future operations. When used in this presentation, the words “anticipate,” “believe,” “can,” “could,” “estimate,” “expect,” “intend,” “is designed to,” “may,” “might,” “will,” “plan,” “potential,” “predict,” “objective,” “should,” or the negative of these and similar expressions identify forward-looking statements. Such statements, based as they are on the current analysis and expectations of management, inherently involve numerous risks and uncertainties, known and unknown, many of which are beyond the Company’s control. Such risks include, but are not limited to: the impact of general economic conditions, general conditions in the biopharmaceutical industries, changes in the global and regional regulatory environments in the jurisdictions in which the Company does or plans to do business, market volatility, fluctuations in costs and changes to the competitive environment. Consequently, actual future results may differ materially from the anticipated results expressed in the forward-looking statements. In the case of forward-looking statements regarding investigational product candidates and continuing further development efforts, specific risks which could cause actual results to differ materially from the Company’s current analysis and projections include: failure to demonstrate the safety, tolerability and efficacy of our product candidates; final and quality controlled verification of data and the related analyses; the expense and uncertainty of obtaining regulatory approval, including from the U.S. Food and Drug Administration and European Medicines Agency; the possibility of having to conduct additional clinical trials; our ability to obtain and maintain intellectual property protection for our product candidates; and our reliance on third parties such as our licensors and collaboration partners regarding our suite of technologies and product candidates. Further, even if regulatory approval is obtained, biopharmaceutical products are generally subject to stringent on-going governmental regulation, challenges in gaining market acceptance and competition. These statements are also subject to a number of material risks and uncertainties that are described in the Company’s filings with the U.S. Securities and Exchange Commission (“SEC”), 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. The reader should not place undue reliance on any forward-looking statements included in this presentation. These statements speak only as of the date made and the Company is under no obligation and disavows any obligation to update or revise such statements as a result of any event, circumstances or otherwise, unless required by applicable legislation.
Agenda

- Recent news
- Update clinical programs
- Ongoing collaborations
- Financial results
- Q&A
Rapidly Emerging Leadership in Immunology
Pioneering differentiated therapeutic antibodies in severe autoimmune diseases and cancer

1. **Novel Target Biology**
   - Integrated via advanced technology suite
   - First- and best-in-class potential

2. **Innovative Access Program**
   - Robust science
   - Collaborative
   - Efficient pipeline expansion

3. **Highly Productive Development Engine**
   - Rapid development timeline
   - New candidate each year

4. **Maximum Value per Asset**
   - Pipeline-in-a-product strategy
   - Strong biological rationale

5. **Multi-Asset Late-Stage Platform**
   - Phase 3 in MG and ITP
   - Pre-commercial activities in MG

Translate immunology breakthroughs into novel medicines which truly impact patients’ lives
<table>
<thead>
<tr>
<th>Product Candidate</th>
<th>Target</th>
<th>Indication</th>
<th>Preclinical</th>
<th>Phase 1</th>
<th>Phase 2</th>
<th>Phase 3</th>
<th>BLA</th>
<th>Next Milestone / Commentary</th>
</tr>
</thead>
<tbody>
<tr>
<td>Argex-113 Efgartigimod</td>
<td>FcRn</td>
<td>Myasthenia Gravis</td>
<td>adap</td>
<td>adap</td>
<td>adap</td>
<td>adap</td>
<td>adap</td>
<td>3Q18: Phase 3 initiated</td>
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<tr>
<td></td>
<td></td>
<td>Immune Thrombocytopenia</td>
<td>adap</td>
<td>adap</td>
<td>adap</td>
<td>adap</td>
<td>adap</td>
<td>2H19: Phase 3 initiation</td>
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<td></td>
<td></td>
<td>ITP Subcutaneous Formulation</td>
<td>adap</td>
<td>adap</td>
<td>adap</td>
<td>adap</td>
<td>adap</td>
<td>1H19: Phase 2 initiation</td>
</tr>
<tr>
<td></td>
<td></td>
<td>Pemphigus Vulgaris</td>
<td>adap</td>
<td>adap</td>
<td>adap</td>
<td>adap</td>
<td>adap</td>
<td>1H19: Cohort 3 initiation</td>
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<td></td>
<td></td>
<td>Chronic Inflammatory Demyelinating Polyneuropathy</td>
<td>adap</td>
<td>adap</td>
<td>adap</td>
<td>adap</td>
<td>adap</td>
<td>2H19: Phase 2 initiation</td>
</tr>
<tr>
<td>Argex-117 Novel complement target</td>
<td>Severe Autoimmune Diseases</td>
<td>adap</td>
<td>adap</td>
<td>adap</td>
<td>adap</td>
<td>adap</td>
<td>Antibody-mediated autoimmune diseases</td>
<td></td>
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<td></td>
<td></td>
<td></td>
<td></td>
<td></td>
<td></td>
<td></td>
<td>Complementary to Argex-113</td>
<td></td>
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<tr>
<td>Argex-110 Cusatuzumab</td>
<td>CD70</td>
<td>Acute Myeloid Leukemia</td>
<td>adap</td>
<td>adap</td>
<td>adap</td>
<td>adap</td>
<td>adap</td>
<td>$500 million upfront</td>
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<td></td>
<td></td>
<td></td>
<td></td>
<td></td>
<td></td>
<td>Eligible for up to $1.3 billion in milestones; tiered royalties</td>
<td></td>
</tr>
</tbody>
</table>
### Partnered Product Candidates

<table>
<thead>
<tr>
<th>Product Candidate</th>
<th>Target</th>
<th>Indication</th>
<th>Preclinical</th>
<th>Phase 1</th>
<th>Phase 2</th>
<th>Phase 3</th>
<th>BLA</th>
<th>Next Milestone / Commentary</th>
</tr>
</thead>
<tbody>
<tr>
<td>ARGX-112</td>
<td>IL-22R</td>
<td>Skin Inflammation</td>
<td></td>
<td></td>
<td></td>
<td></td>
<td></td>
<td>Eligible for up to ~€100mm in milestones; tiered royalties</td>
</tr>
<tr>
<td>ARGX-115</td>
<td>GARP</td>
<td>Cancer Immunotherapy</td>
<td></td>
<td></td>
<td></td>
<td></td>
<td></td>
<td>Received $60mm in upfront and preclinical milestone payments; Eligible for up to $625mm milestones; tiered royalties</td>
</tr>
<tr>
<td>ARGX-116</td>
<td>ApoC3</td>
<td>Dyslipidemia</td>
<td></td>
<td></td>
<td></td>
<td></td>
<td></td>
<td>Eligible for double-digit royalties and exclusive option to license the program; collaboration with Novo Nordisk</td>
</tr>
</tbody>
</table>

- **Innovative Access Program**: 7 live programs
- Antibody discovery alliance with **Shire** focused on multiple rare disease targets – 2 options exercised
- Additional programs include ARGX-114, HFG-mimetic SIMPLE Antibody® directed against the MET receptor (developed by Agomab); ARGX-111 targeting c-MET in solid tumors and blood cancers (P1 concluded, wholly-owned, available for partnering) and ARGX-109 (gerilimzumab) targeting IL-6 for rheumatoid arthritis (P1 concluded, partnered with Genor Biopharma)
**Myasthenia Gravis Phase 3 ADAPT Trial Design**

**Same Primary Endpoint as Successful Phase 2 Trial**

- Randomized, double-blind, placebo-controlled, multicenter trial enrolling 150 patients in North America, Europe and Japan
- 10 mg/kg intravenous (IV) dose of efgartigimod over 26-week period
- Enrolling AChR positive and AChR negative patients with disease driven primarily by MuSK and LRP4 autoantibodies
- Patients in the ADAPT trial will be able to roll over into an open-label extension trial for a period of one year
- First patient dosed in September 2018
- Based on PMDA feedback, this Phase 3 trial, if data is positive, to also serve as a basis for Japan registrational submission

**Primary endpoint**

**Myasthenia Gravis Activities of Daily Living (MG-ADL) Score**

**Secondary endpoints**

**Efficacy, Safety, Tolerability, Quality of Life and Impact on Normal Daily Activities Measures**
Mean platelet counts versus total WHO scale versus total IgGs

Immune Thrombocytopenia Ph2 Clinical Trial
Reduction of Total IgGs Correlates with Increased Platelet Counts and Reduced Bleeding Events

Placebo

10 mg/kg efgartigimod

Mean platelet count (x10⁹/L)

% patients with total WHO scale > 0

Days

% total IgGs

Mean platelet counts (x10⁹/L)

% patients with total WHO scale > 0

Days

% total IgGs

% patients with total WHO scale > 0
Strong Improvement of Platelet Counts Across Doses

46-67% of patients exceeded platelet counts \(\geq 50 \times 10^9/L\) during at least two visits.

- **OLE acts as true fourth cohort** since patients’ platelets had to fall below \(30 \times 10^9/L\) to be eligible for a treatment cycle; patients still in response from primary study were not eligible.

- **Responses seen across newly diagnosed (in 5mg/kg arm), persistent and chronic ITP patients**

*After cut-off date not QC-ed*
IDMC recommendation for cohort 3 to reach clinical remission (with/without minimal therapy):

- Weekly infusions 25 mg/kg (induction phase) until disease control (DC) with minimum of 5
- Biweekly dosing after DC
- Start maintenance based on DC
- Treatment duration limited to 34 weeks (induction + maintenance)
Efgartigimod: a Pipeline-in-a-Product Opportunity

Landscape of IgG-mediated severe autoimmune diseases (sampling)

- Immune Thrombocytopenia
  - Myasthenia Gravis
  - Multiple Sclerosis
- Scleroderma
- Lupus
- Epidermolysis Bullosa Acquisita
  - Pemphigus
  - Anca Vasculitis
  - Bullous Pemphigoid

Solid Biology Rationale
Disease proven to be predominantly mediated by pathogenic IgGs

Feasible for Biotech
Orphan potential, economically viable, efficient clinical & regulatory pathway

Proof-of Concept:

- Myasthenia Gravis
- Immune Thrombocytopenia
- Pemphigus Vulgaris

Therapeutic Area Beachheads with Expansion Possibilities into Adjacent Indications

- Neuromuscular Diseases
  - Chronic Inflammatory Demyelinating Polyneuropathy (CIDP)
- Hematology Disorders
- Blistering Diseases

Phase 2 CIDP study to start in 2H 2019
Efgartigimod: Human IgG1 Fc Fragment with ABDEG™ Mutations
Exploits Natural Fc/FcRn Interaction and retains pH dependent binding

IgG antibodies recycle through FcRn(1)...
efgartigimod potently blocks FcRn...
leading to IgG elimination(2)

(4) argenx data
Efgartigimod Emerges as First-In-Class and Best-In-Class

**First-in-class features**
- Human IgG1 Fc fragment
- With ABDEG™ mutations
- Reduced FcγR, C1q binding
- Endosomal recycling FcRn-efgart complex; no lysosomal degradation
- Can rebind FcRn
- 1/3 size of IgG; excellent physicochemical stability

**Best-in-class clinical attributes**
- Natural ligand of FcRn
- Enhanced, pH dependent binding
- Clean tolerability profile
  No headache or GI AE profile (~120 subjects)
- No decrease in albumin
  (Ulrichts et al., JCI, 2018)
- Long half-life
  Unparalleled tissue penetration & distribution
- Long-lasting, potent PD effect
  Fast onset of clinical benefit
- Lower dose enables convenient subQ administration, high concentration formulations and lower COGS
Efgartigimod Emerges as First-In-Class and Best-In-Class

- Human IgG1 Fc fragment
- With ABDEG™ mutations
- Clean tolerability profile: No headache or GI AE profile (~120 subjects)
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Best-in-class clinical attributes
- Natural ligand of FcRn
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Long half-life
- Unparalleled tissue penetration & distribution
- Long-lasting, potent PD effect
- Fast onset of clinical benefit
- Lower dose enables convenient subQ administration, high concentration formulations and lower COGS

Efgartigimod (25 mg/kg q7d):
No significant lowering of albumin

![Graph showing albumin levels over time](Image)

Ulrichts et al., JCI, 2018
Cusatuzumab Mode-of-Action Targets both Leukemic Stem Cells and Blasts

Cusatuzumab induces LSC differentiation

1 Induce myeloid differentiation
2 Kill LSCs

Cusatuzumab kills blasts

3 Kill Blasts
4 Block proliferation & survival signal

Activation of the pathway leads to release of sCD27, which is a biomarker

- Cusatuzumab is a potentially first-in-class anti-CD70 ADCC enhanced SIMPLE Antibody™ which selectively targets LSCs and blasts in AML and other heme indications
92% (11/12) Response Rate – CR/CRi/PR

Three patients on study for more than 12 months

Risk | Best response
--- | ---
0,0 | 10,0 | 20,0 | 30,0 | 40,0 | 50,0 | 60,0 | 70,0 | 80,0
--- | --- | --- | --- | --- | --- | --- | --- | ---
Cohort 1 (1 mg/kg)
Adv CR | Int CR | Adv CR | Int CR
1 | 2 | 3 | 4 | 5 | 6 | 7 | 8 | 9 | 10
Adv CRi | Adv CRi | Int CR
11 | 12

Time to response (weeks)
Partial remission (PR)
Complete remission with incomplete recovery (CRi)
Complete remission (CR)

EOT due to PD
EOT due to AE (IRR)
EOT due to transplant

Cusatuzumab monotherapy

Cut off: 15Oct18

Ongoing
MRD - BM by flow and molecular genetics

6 months
12 months
Accelerate & broaden development plan | Joint development plan focused on AML, MDS and other heme malignancies

Secure strong financials | Upfront $300m + $200m equity @ 20% premium, 1.3Bn in milestones, double digit royalties OUS

Retain commercial upside | 50% of US economics on a royalty basis, up to 50% commercial efforts

“We believe that cusatuzumab can become a foundational therapy for all lines of AML and high-risk MDS.” Brian Kenney, J&J spokesperson
Innovative Access Program
Success formula with proven track record

<table>
<thead>
<tr>
<th>argenx</th>
<th>Top Academic Labs</th>
</tr>
</thead>
<tbody>
<tr>
<td>Developing highly differentiated antibodies</td>
<td>Unravelling novel target biology</td>
</tr>
</tbody>
</table>

Pipeline of first-in-class assets

**Wholly owned**
- ARGX-113
- ARGX-110 (Co-dev Janssen)
- ARGX-117
- ARGX-118

**Partnered**
- ARGX-115 (AbbVie)
- ARGX-112 (Leo Pharma)
- ARGX-116 (Staten/Novo Nordisk)
- ARGX-114 (Agomab)

- University Southampton
- University Utrecht
- Bern University
- UCL-de Duve
- Ludwig Institute
- Penn University
- Columbia University
- University Torino
## argenx Y2018 Financials

<table>
<thead>
<tr>
<th></th>
<th>2018</th>
<th>2017</th>
<th>Variance</th>
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</thead>
<tbody>
<tr>
<td><strong>in thousands of €</strong></td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>Revenue</td>
<td>€ 21,482</td>
<td>€ 36,415</td>
<td>€ (14,933)</td>
</tr>
<tr>
<td>Other operating income</td>
<td>€ 7,749</td>
<td>€ 4,841</td>
<td>€ 2,908</td>
</tr>
<tr>
<td><strong>Total operating income</strong></td>
<td>€ 29,231</td>
<td>€ 41,256</td>
<td>€ (12,025)</td>
</tr>
<tr>
<td>Research and development expenses</td>
<td>€ (83,609)</td>
<td>€ (51,740)</td>
<td>€ (31,869)</td>
</tr>
<tr>
<td>Selling, general and administrative expenses</td>
<td>€ (27,471)</td>
<td>€ (12,448)</td>
<td>€ (15,023)</td>
</tr>
<tr>
<td><strong>Operating loss</strong></td>
<td>€ (81,849)</td>
<td>€ (22,932)</td>
<td>€ (58,917)</td>
</tr>
<tr>
<td>Financial income</td>
<td>€ 3,694</td>
<td>€ 1,250</td>
<td>€ 2,444</td>
</tr>
<tr>
<td>Financial expenses</td>
<td>—</td>
<td>—</td>
<td>—</td>
</tr>
<tr>
<td>Exchange losses</td>
<td>€ 12,308</td>
<td>€ (5,797)</td>
<td>€ 18,105</td>
</tr>
<tr>
<td><strong>Loss before taxes</strong></td>
<td>€ (65,847)</td>
<td>€ (27,479)</td>
<td>€ (38,368)</td>
</tr>
<tr>
<td>Income tax income expense</td>
<td>€ (794)</td>
<td>€ (597)</td>
<td>€ (197)</td>
</tr>
<tr>
<td><strong>Total comprehensive loss of the period</strong></td>
<td>€ (66,641)</td>
<td>€ (28,076)</td>
<td>€ (38,565)</td>
</tr>
<tr>
<td>Net increase in cash, cash equivalents and current financial assets compared to year-end 2017 and 2016</td>
<td>€ 204,795</td>
<td>€ 263,047</td>
<td></td>
</tr>
<tr>
<td>Cash, cash equivalents and current financial assets at the end of the period</td>
<td>€ 564,569</td>
<td>€ 359,775</td>
<td></td>
</tr>
</tbody>
</table>
**Financial Profile and Investor Composition**

Shareholder base > 70% U.S. investors

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**Additional Key Statistics – Dec 31, 2018**

- **Cash position:** €564.6 mm (+ $500 mm Janssen deal)
- **Capital raised since inception:** €717 mm (ex. grants)
  - 2017: raised $115 mm (€102 mm) in Nasdaq IPO
  - 2017: raised $266 mm (€226 mm) in public offering
  - 2018: raised $300 mm (€256 mm) in public offering
- **Non-dilutive funding since inception:** €107 mm (incl. grants)
  - 2018: $10mm second preclinical milestone AbbVie
- **132 employees & consultants** — 97 R&D, 35 SG&A

---

**Blue-Chip Investor Base**

- **Outstanding shares (Feb 22, 2019):** 37,907,551
- **Outstanding stock options (Feb 22, 2019):** 3,371,311
- **U.S. shareholding:** above 70%
### Key Upcoming Expected Milestones & Communications

<table>
<thead>
<tr>
<th>Year</th>
<th>Quarter</th>
<th>Efgartigimod</th>
<th>Cusatuzumab</th>
<th>New assets</th>
<th>Partnerships</th>
</tr>
</thead>
<tbody>
<tr>
<td>2019</td>
<td>Q1</td>
<td>ITP: Outcome FDA/PMDA/EMA EoPh2 meeting</td>
<td>Potential Milestones in Strategic Partnership with Janssen</td>
<td>Exclusive collaboration Halozyme for ENHANZE®</td>
<td>Potential Milestones</td>
</tr>
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<td></td>
<td></td>
<td>R&amp;D day May 22 (NY)</td>
</tr>
<tr>
<td></td>
<td>Q2</td>
<td>PV: Start Cohort 3 Ph2 1H</td>
<td>ITP: Launch Ph2 SC 1H</td>
<td>ARGX-117 First Indication</td>
<td>Potential Milestones</td>
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<td></td>
<td>Q3</td>
<td>ITP: Launch Ph3 IV 2H</td>
<td>ITP: Launch CIDP (NY)</td>
<td>Cusatuzumab</td>
<td>Potential Milestones in Strategic Partnership with Janssen</td>
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</tr>
<tr>
<td></td>
<td>Q4</td>
<td>CIDP: Launch Ph2 2H</td>
<td>ARGX-117 CTA Filling</td>
<td>New assets</td>
<td>Potential Milestone(s)</td>
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Thank you!