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argenx 2021:
Becoming a Global Integrated Immunology Biotech

- Patients
  - Neuro-muscular
  - Hem/Onc
  - Potential Expansion

People + Processes + Systems

Innovative Access Program
Impressive Value Creation Since IPO

Accelerating & expanding development programs

- MG & ITP
- Interim PV
- AML

Key Data

- IPO: $50M
- Now: $350M

$1.1B

- $50M
- $1.1B
- $5B

Well-capitalized to advance to the next level

Global expansion

- Ghent
- Boston (2018)
- Tokyo (2019)
argenx Today:
Building Leadership in Immunology

- **Late-stage immunology company**
  - Two Phase 3 trials in progress by end of 2019

- **Wholly-owned pipeline-in-a-product assets**
  - Proof-of-concept in two beachhead indications

- **Validating oncology collaborations**
  - Maintained 50% of cusatuzumab commercial rights

- **Innovative Access Program**
  - One new asset per year to grow pipeline

- **Well-funded with cash into 2021**
  - $1.05B in cash to execute on ambitious plan

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**Well-funded with cash into 2021**

- **50% of cusatuzumab commercial rights**
- **One new asset per year to grow pipeline**
- **$1.05B in cash to execute on ambitious plan**
<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 milestones</th>
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<td>ARGX-113</td>
<td>FcRn</td>
<td>Myasthenia Gravis (MG)</td>
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<td>Immune Thrombocytopenia (ITP)</td>
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<td>Chronic Inflammatory Demyelinating Polyneuropathy (CIDP)</td>
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<td>Severe Autoimmunity IV/ENHANZE® SC</td>
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<td>Lead selected</td>
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Deep Proprietary Pipeline of Highly Differentiated Product Candidates
Targeting high-value rapid-growth markets
Multiple Value-Creating Milestones Through 2020

2H19
- ENHANZE® HV Data
- Phase 3 ADVANCE ITP Start
- Phase 2 CIDP Start
- Phase 2 AML Start
- ARGX-117 CTA Filing

2020
- Phase 2 PV Data (1H)
- Phase 3 ADAPT MG Data (2H)
- 5th Indication
- Development Update
- ARGX-119

Well-capitalized to execute on ambitious development plan into 2021
Innovative Access Program
Unique discovery engine to identify novel target biology

Accessing Novel Targets Through Collaboration

argenx

Antibody Expertise
SIMPLE Antibody™, NHance®, ABDEG™, POTELLIGENT®

Top Academic Institutions & Biotechs
Disease Biology Expertise
Texas A&M, Bern, Utrecht, Louvain, Penn, Columbia, Torino, de Duve, VIB

Co-creating first-in-class assets

WHOLLY-OWNED
ARGX-113
ARGX-110
(Co-developed with Janssen)

PARTNERED
ARGX-115
ARGX-112
ARGX-116
ARGX-114

5-10 ongoing programs at any given time
Serial Value Creation from Novel Targets

ARGX-117
Pipeline-in-a-product

ARGX-118
Immunology breakthrough in airway inflammation

ARGX-110
Cusatuzumab
50% profit split in US

ARGX-112
€120M and royalties

ARGX-113
Efgartigimod
Pipeline-in-a-product

ARGX-114
Profit share

ARGX-115
$625M and royalties

ARGX-116
Profit share

ARGX-118
Immunology breakthrough in airway inflammation

ARGX-111
$625M and royalties

ARGX-112
€120M and royalties
Late-stage Development Product Candidates: Efgartigimod and Cusatuzumab
Efgartigimod: Human IgG1 Fc Fragment with Proprietary 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)**
Efgartigimod: Best-In-Class Potential With Broad Applicability

**Efficacy** – Set the bar high in Phase 2 studies
75% of gMG patients achieved durable responses
~50% response rate in heavily pre-treated ITP patients

**Safety** – No class effect
>150 patients treated
No safety signal detected (no trend in headaches or GI symptoms; no drop in albumin)

**Convenience** – Optionality for patients
IV (10mg/kg): 60min infusion, no premedication, no infusion reactions
SC maintenance product (165mg/ml): 2ml push
SC ENHANZE® product through strategic collaboration with Halozyme
Efgartigimod: Pipeline-in-a-Product Opportunity
Clinical proof-of-concept achieved for neuromuscular and hematology indications

Landscape of IgG-mediated severe autoimmune diseases (sampling)

- Immune Thrombocytopenia
  - Myasthenia Gravis
  - Multiple Sclerosis
- Scleroderma
- Lupus
  - Pemphigus
  - Epidermolysis Bullosa Acquisita
- Rheumatoid Arthritis
  - Anca Vasculitis
- Myasthenia Gravis
- Multiple Sclerosis
- Pemphigus Vulgaris
- 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 2H19
Efgartigimod in Myasthenia Gravis – Phase 3 ADAPT Trial Ongoing
Enrollment on track – data expected 2H20

- Randomized, double-blind, placebo-controlled, multicenter trial enrolling 150 patients in North America, Europe and Japan
- Enrolling AChR positive and AChR negative patients with disease driven primarily by MuSK and LRP4 autoantibodies
- 10 mg/kg IV dose over 26-week period
- Patients eligible to roll over into 1-year open-label extension trial

Data from completed Phase 2 trial published in Neurology demonstrating that:
- Treatment with efgartigimod resulted in clinically meaningful and sustained improvement in disease scores, consistent across four MG scales
- Efgartigimod has a clean tolerability profile in line with HV study with no withdrawals or apparent differences between patients or placebo groups
Efgartigimod in Myasthenia Gravis
Role of pathogenic autoantibodies very well-characterized

- Block acetylcholine from binding to AChR
- Cross-link and internalize AChRs reducing number of binding sites
- Recruit complement which can destroy muscle surface
Efgartigimod in Myasthenia Gravis – Strong Phase 2 Efficacy Results

75% of treated patients achieved lasting response

Patients with MG-ADL ≥ 2 for a period of at least 6 weeks

- 83% of efgartigimod patients achieved clinically meaningful response (MG-ADL ≥2)
- 75% of efgartigimod patients had clinically meaningful and statistically significant improvement in MG-ADL score for at least 6 consecutive weeks versus 25% of patients on placebo
Total and Pathogenic IgG Reduction Correlates with Clinical Improvements

Assessment for all efficacy scales in Phase 2

- Clinical improvement persists despite return of IgG levels
- Potential differentiation from PLEX where clinical benefit was reported to be lost 2-4 weeks after end of treatment
Efgartigimod in Immune Thrombocytopenia – Phase 3 ADVANCE Trial to Start

First of two registration Phase 3 trials to start in 2H19

Randomized, double-blind, placebo-controlled, multicenter trial enrolling up to 158 adult patients with primary ITP

Enrolling patients with platelet levels $<30 \times 10^9/L$ and stable dose and dosing frequency of SoC prior randomization

10 mg/kg IV dose over a 24-week treatment period

Patients eligible to roll over into 1-year open-label extension trial

Data from completed Phase 2 trial presented at the annual ASH conference demonstrating that:

- Treatment with efgartigimod resulted in clinically meaningful improvements in platelet counts and efgartigimod treatment showed a clear correlation between IgG reduction, platelet count improvement and bleeding event reduction
- Efgartigimod has a clean tolerability profile in line with HV study and treatment-emergent adverse events were balanced between active and placebo arms
Efgartigimod in Immune Thrombocytopenia
Targets all pathogenic autoantibody actions simultaneously and may limit therapeutic cycling

1. Accelerate platelet clearance
   - Platelets
   - FcγR
   - Macrophage
   - IVIg
   - Tavilisse
   - Splenectomy
   - Efgartigimod

2. Inhibit platelet production
   - Platelets
   - Megakaryocyte
   - TPO-RA
   - Efgartigimod

3. Induce platelet killing
   - Platelets
   - FcγR
   - TPO-RA
   - Efgartigimod

4. Interfere with platelet function
   - Platelets
   - Fibrinogen receptor
   - vWF receptor
   - Collagen receptor
   - Efgartigimod
Efgartigimod in Immune Thrombocytopenia – Strong Phase 2 Efficacy Results

Strong improvement of platelet counts across doses

46-67% of patients achieving platelet counts of $\geq 50 \times 10^9$/L at least two times

**Main Study**

- Placebo + SOC: 25% (N=3, N=12)
- Efgartigimod 5 mg/kg + SOC: 46% (N=6, N=13)
- Efgartigimod 10 mg/kg + SOC: 46% (N=6, N=13)

**OLE (1st treatment cycle)**

- Efgartigimod 10 mg/kg + SOC: 67% (N=8, N=12)

- 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.
Reduction of Total IgGs Correlates with Increased Platelet Counts and Reduced Bleeding Events

Mean platelet counts versus total WHO scale versus total IgGs

<table>
<thead>
<tr>
<th>Days</th>
<th>Placebo</th>
<th>10 mg/kg efgartigimod</th>
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</thead>
<tbody>
<tr>
<td>Mean platelet count (x10^9/L)</td>
<td>% patients with total WHO scale &gt; 0</td>
<td>Mean platelet count (x10^9/L)</td>
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<tr>
<td>0</td>
<td>80</td>
<td>10</td>
</tr>
<tr>
<td>5</td>
<td>70</td>
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<td>10</td>
<td>45</td>
</tr>
<tr>
<td>40</td>
<td>0</td>
<td>50</td>
</tr>
</tbody>
</table>

% total IgGs
Mean platelet counts (x10^9/L)
% patients with total WHO scale >0
Efgartigimod in Pemphigus Vulgaris – Phase 2 Ongoing
Cohort 3 enrolling – topline data expected 1H20

Phase 2, cohort 3 enrolling patients:

- Administration of extended dosing of efgartigimod
- To evaluate potential of efgartigimod to induce clinical remission

Results from Cohort 1

Rapid disease control in 4 out of 6 PV patients:
- 3 within 1 week
- 1 within 4 weeks

Patients with disease control:
- Mean max reduction in Pemphigus Disease Area Index (PDAI) score: 55%
- Mean max decrease in pathogenic IgGs: 57%

Favorable tolerability profile

No meaningful anti-drug antibody signals (ADA) reported
Multiple MOA of Cusatuzumab

- Novel target and mechanism of action\(^1\) (inhibition of CD70 pathway)
- Intrinsic activity shown as a single-agent in AML
- Potential for combination therapy\(^2\)
- Phase 1/2 study: 92% ORR with 10/12 patients with CR/Cri after cusatuzumab treatment in combination with azacitidine (AZA) in newly diagnosed AML patients\(^3\)
- IAP, Bern University – Prof. Ochsenbein

Differen-
tiated
cell

Induce myeloid differentiation

Block proliferation and survival signal

Kill LSCs and AML blasts (ADCC)
## Cusatuzumab in AML – Phase 2 to Start

Phase 2 and registration-directed trial in acute myeloid leukemia to start in 2H19

### Randomization

- **Part 1:** Dose selection
  - 10 mg/kg
  - 20 mg/kg

- **Part 2:** Expansion at selected dose
  - Interim analysis
  - Final analysis

### Combination Therapy:

- Cusatuzumab + Azacitidine

### Patient Population:

- Newly diagnosed AML patients unfit for intensive chemotherapy (n= up to 150)

### Primary Objective:

- To determine the efficacy (CR rate)

### Secondary Objectives:

- ORR = (CR + CRh + CRi)
- Time to response and duration of response
- Event-free survival
- Overall survival
- Safety
- PK/immunogenicity
- MRD

### Anticipated

- Phase 2 study start: second half 2019
## Cusatuzumab Strategic Alliance with Janssen Pharmaceuticals

<table>
<thead>
<tr>
<th>argenx objectives</th>
<th>Janssen alliance</th>
</tr>
</thead>
<tbody>
<tr>
<td>Accelerate &amp; broaden development plan</td>
<td>Joint development plan focused on AML, MDS and other heme malignancies</td>
</tr>
<tr>
<td>Secure strong financials</td>
<td>Upfront $300M + $200M equity @ 20% premium, $1.3B in milestones, double digit royalties OUS</td>
</tr>
<tr>
<td>Retain commercial upside</td>
<td>50% of US economics on a royalty basis, up to 50% commercial efforts</td>
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</table>
New Assets from Innovative Access Program: ARGX-117 and ARGX-118
Innovative Access Program
Pipeline recently expanded with addition of two preclinical assets

**Early Target Validation**

Power of SIMPLE Antibody™ technology
→ Charcot-Leyden Crystal dissolving antibodies

Unravelling novel airway inflammation biology
→ Galectin-10 first novel airway inflammation target in decades

ARGX-118

**Jumpstart Product Development**

Power of NHance® technology and engineering know-how
→ Turn unique mouse V-regions into highly differentiated product candidate

Leveraging unique insights in complement disease biology
→ Pipeline-in-product opportunity

ARGX-117
Targeting C2 Preserves Key Complement Functionality

1. Target upstream C5 to shut down all functions
2. C3 deficiency → infections
   - C3 levels extremely high
3. Intact alternative pathway → reduced infection risk
4. Lectin pathway implicated in disease
   - Role still being revealed
5. C1 deficiency → lupus
   - Lectin not blocked
6. C4 deficiency → lupus
   - C4 levels variable
7. Crossroad classical & lectin pathway
   - Manageable C2 levels
   - Mild deficiency

Classical
- IgG
- IgM

Lectin
- Mannose sugar

Alternative
- Foreign surface
ARGX-117: V and Fc Regions Act in Concert to Sweep C2

1. ARGX-117 binds C2 in circulation

2. ARGX-117/C2 complex taken up by pinocytosis entering endosome; ARGX-117 releases C2 at lower pH in endosome

3. C2 is degraded in lysosome

4. ARGX-117 remains bound going back into circulation to bind to new C2 because of NHance® mutations
ARGX-117: Dosing Optionality

**Pharmacokinetics**

- Half-life ARGX-117: 2-3 weeks
- Cynomolgus monkey data

**Pharmacodynamics**

- Blocking for 2 months after repeat dosing
- C2 levels cynomolgus monkey = 4x human

Option exercised for C2
Commercial
Building Immunology Franchises

**Neuro Inflammation**
- MG
- CIDP
- MMN

**Poly Neuropathies**

**Myositis**

Expansion Opportunities

**BMT**  **AIHA**  **ANCA**  **Leukemia Lymphomas**

**MALIGNANT HEME**
- AML
- MDS

**BENIGN HEME**
- ITP

Hem/Onc

50/50
Franchises Sit in High-Value Rapid-Growth Global Markets

- **Neuromuscular**
  - >$5B (CAGR ~10%+)
  - CIDP: 2.4B
  - MG: 2.2B
  - MMN: 0.4B

- **Hem/Onc**
  - ~$7B (CAGR ~10%)
  - AML: 2.2B
  - MDS: 1.7B
  - ITP: 2.8B

- ~370,000 patients
- Double digit CAGR