



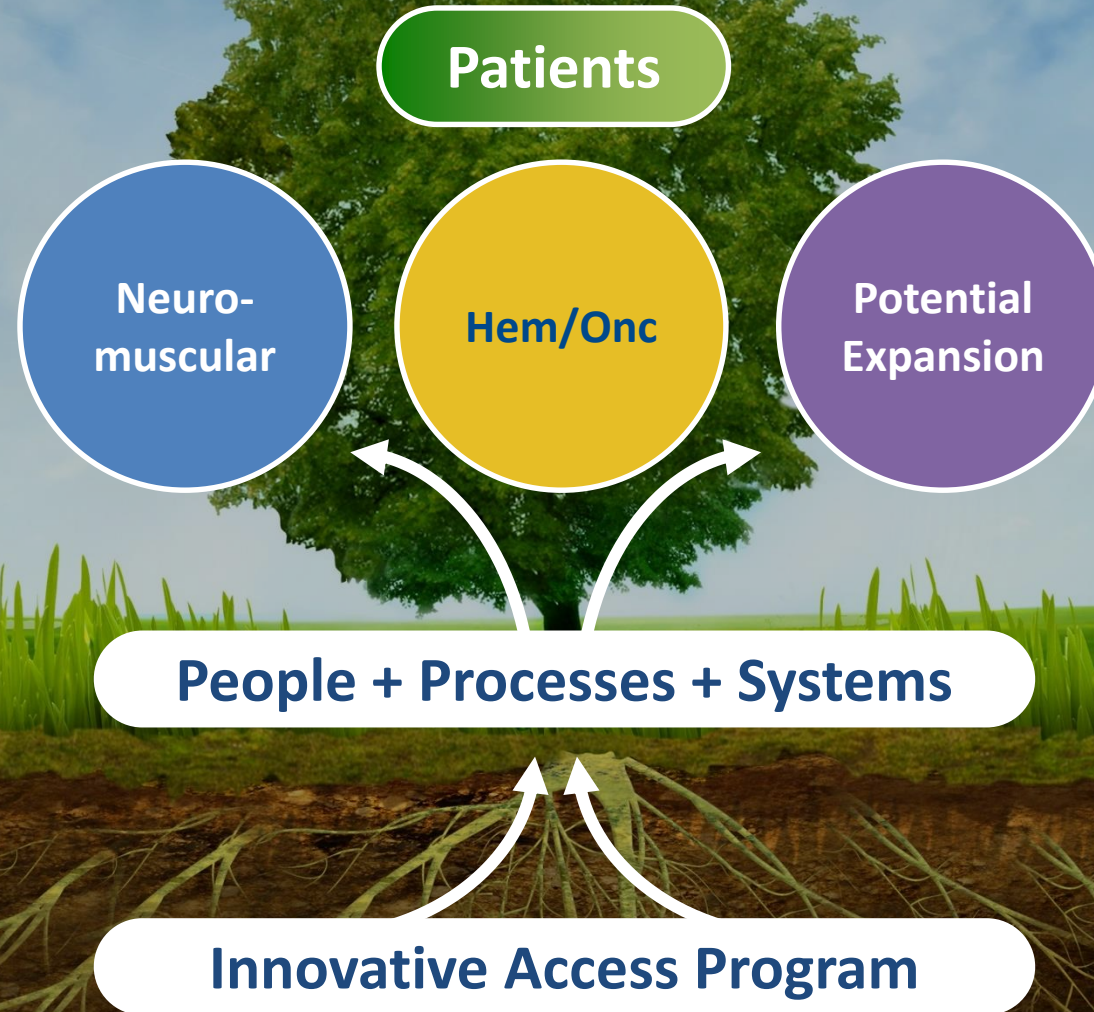
June 2019

Forward-Looking Statements

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product candidates and continuing further development efforts, specific risks which could cause actual results to differ materially from the Company’s current analysis and expectations 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.

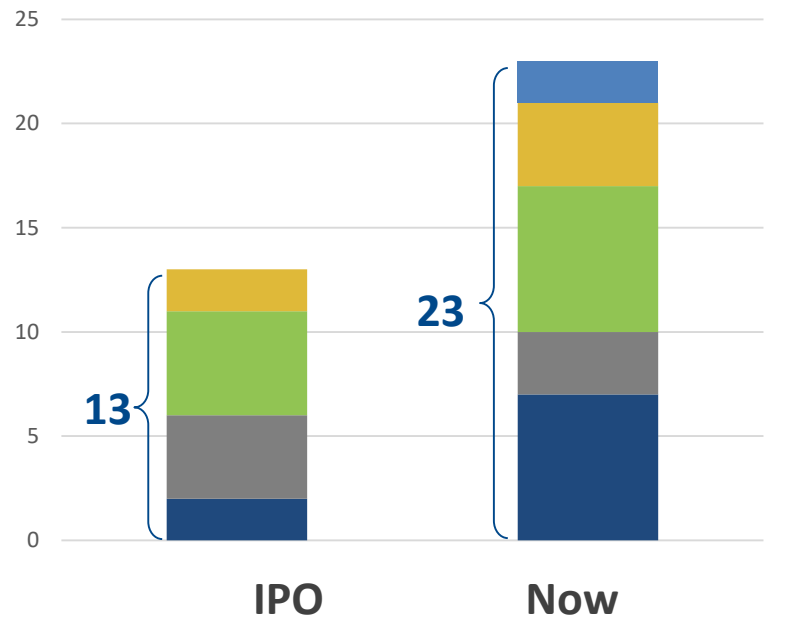


Impressive Value Creation Since IPO

Accelerating & expanding development programs

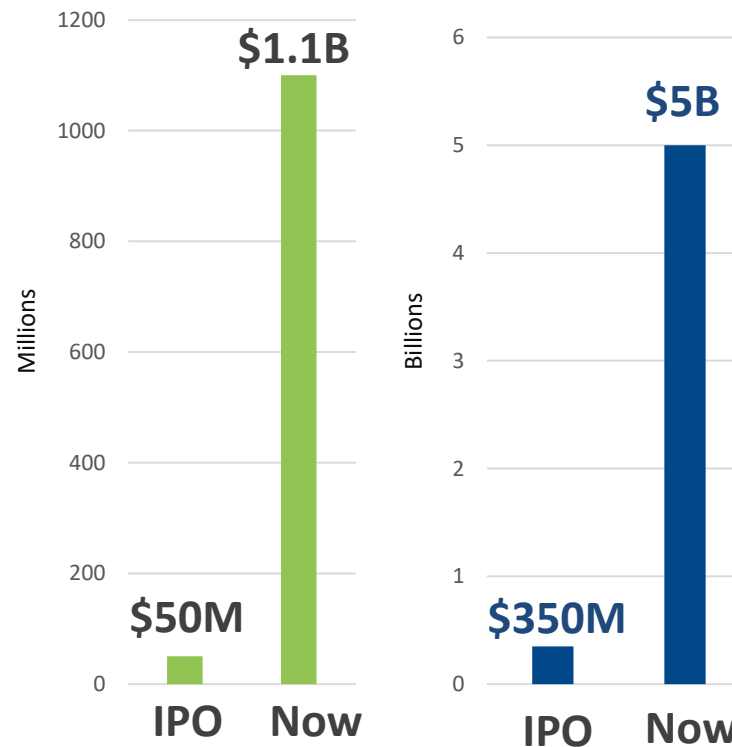
Key Data

MG & ITP | Interim PV | AML



■ IAP ■ Preclinical ■ Phase 1 ■ Phase 2 ■ Phase 3

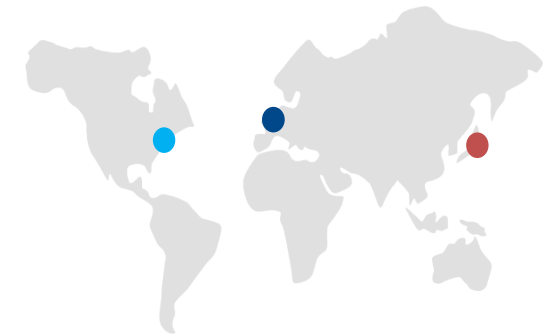
Well-capitalized to advance to the next level








Cash

Market Cap

Global expansion



- Ghent
- Boston (2018)
- Tokyo (2019)

-  **Late-stage immunology company** → **Two Phase 3 trials by end of 2019**
-  **Wholly-owned pipeline-in-a-product assets** → **Potential across multiple high-value indications**
-  **Proof-of-concept in two indications** → **Success in beachhead indications de-risks concept**
-  **Validating oncology collaborations** → **Maintained 50% of cusatuzumab commercial rights**
-  **Proven engine to grow pipeline** → **Innovative Access Program in action**

Multiple Value-Creating Milestones Through 2020

2019

ENHANZE[®] HV Data

Phase 3 ITP IV/SC Start

Phase 2 CIDP Start

Phase 2 AML Start

ARGX-117 CTA Filing

2020



Phase 2 PV Data

Phase 3 ADAPT MG Data

5th Indication

Development Update

ARGX-119

-  Efgartigimod
-  Cusatuzumab
-  New Assets

\$1.1B in Cash; Funded Through 2021

Accessing Novel Targets Through Collaboration

argenx

Top Academic Institutions & Biotechs



Antibody Expertise

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

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 Janssen)

ARGX-117
ARGX-118

PARTNERED

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

5-10 ongoing programs at any given time

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

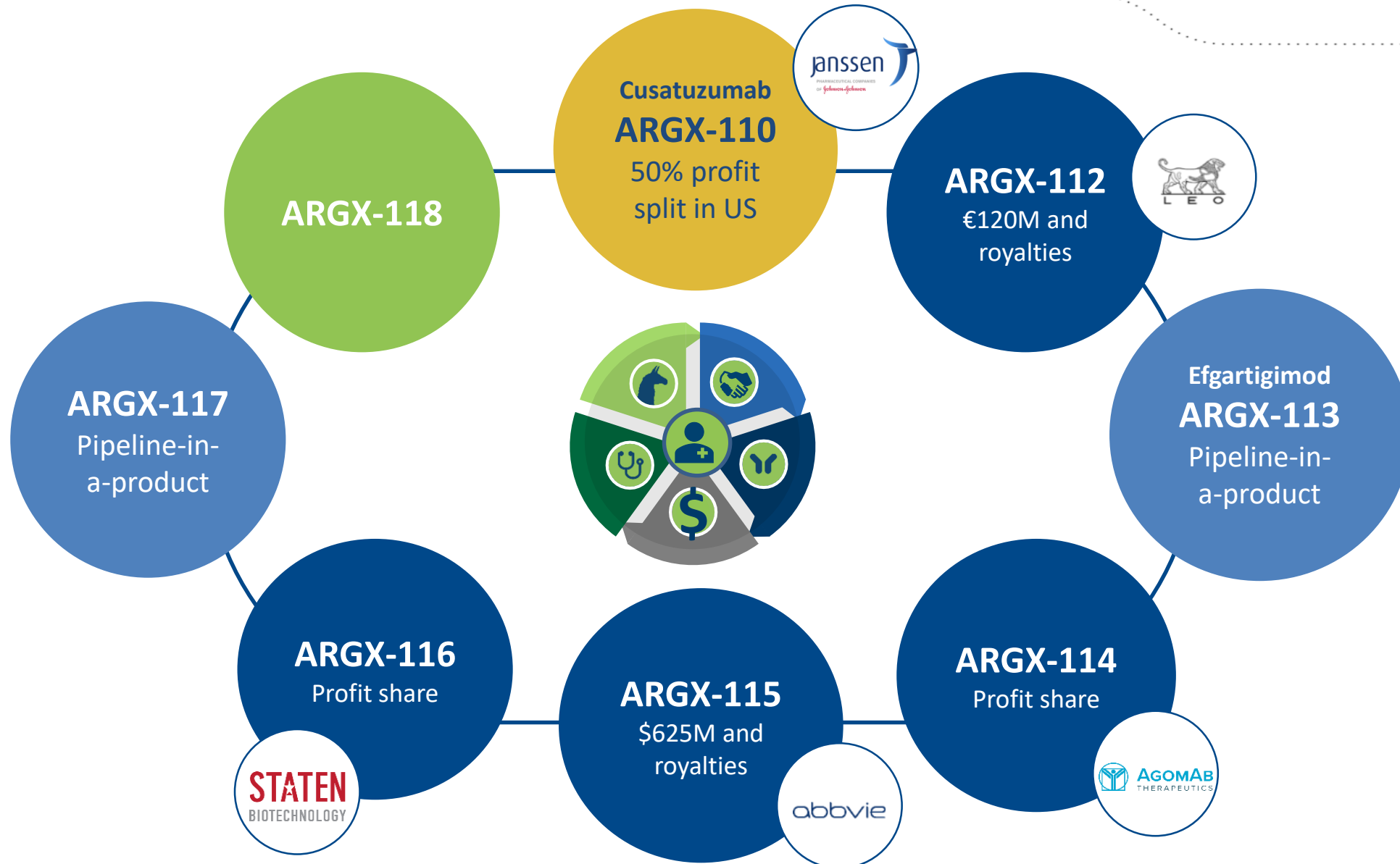
Leveraging unique insights in complement disease biology


→ Pipeline-in-product opportunity

ARGX-117



Serial Value Creation from Novel Targets





Late-stage Development Product Candidates: Efgartigimod and Cusatuzumab

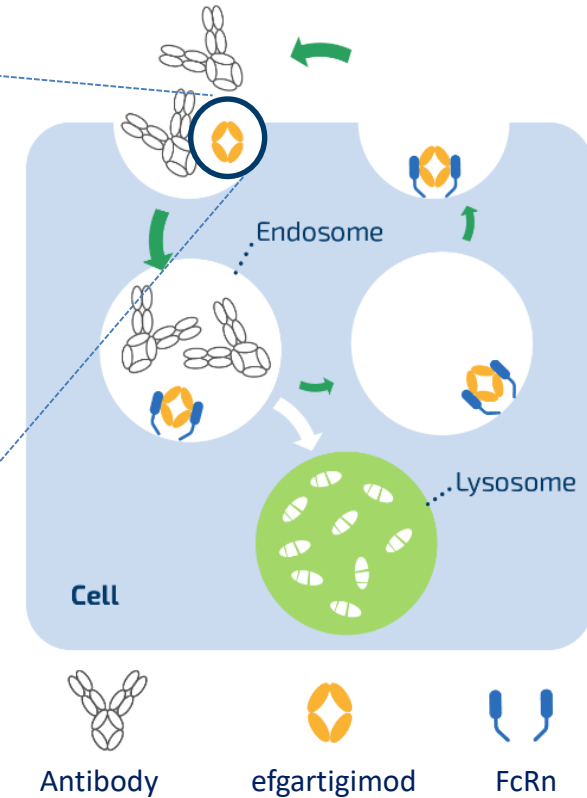
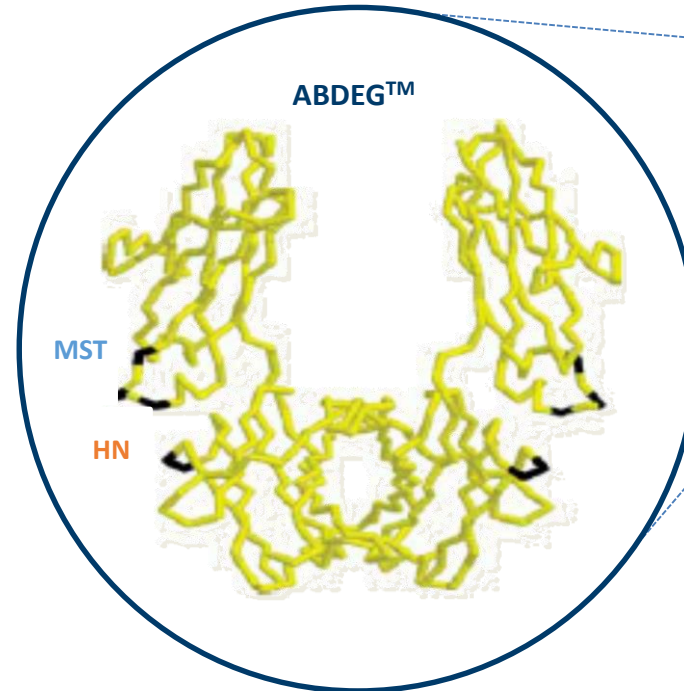
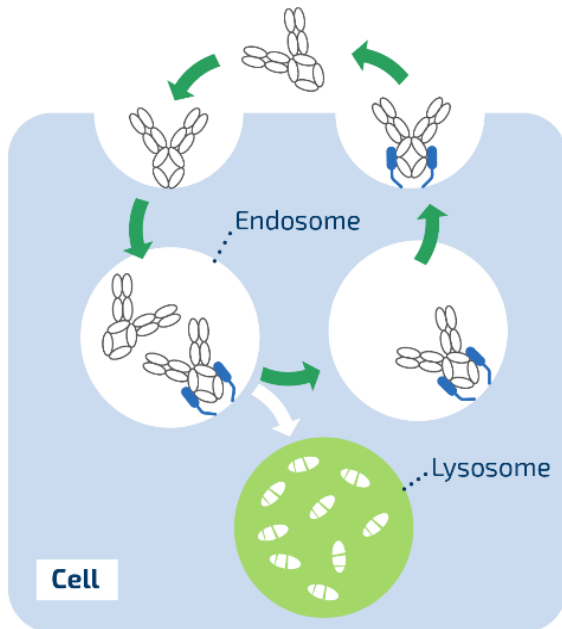
Efgartigimod: Human IgG1 Fc Fragment with ABDEG™ Mutations

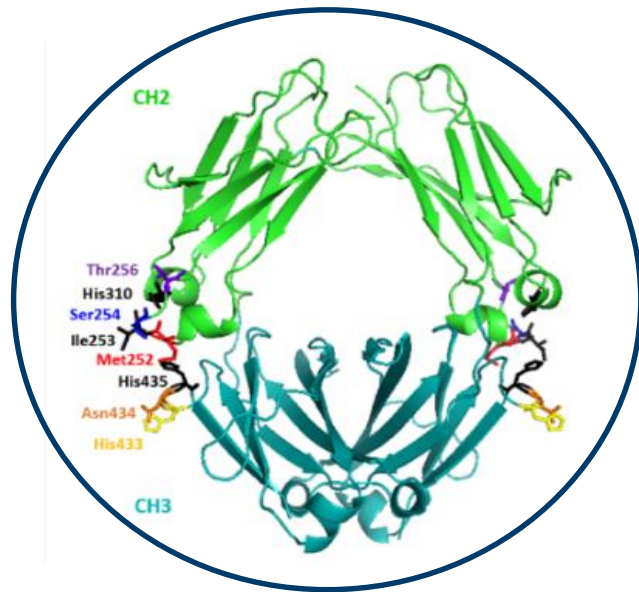
Exploits Natural Fc/FcRn Interaction and retains pH dependent binding

IgG antibodies recycle through FcRn⁽¹⁾...

efgartigimod potently blocks FcRn...

leading to IgG elimination⁽²⁾





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 product (10mg/kg): 60min infusion, no premedication, no infusion reactions
- SC maintenance product (165mg/ml): 2ml push
- SC Enhanze® product through Halozyme

Efgartigimod: a Pipeline-in-a-Product Opportunity

Landscape of IgG-mediated severe autoimmune diseases (sampling)

Immune
Thrombocytopenia

Scleroderma

Lupus

Epidermolysis
Bullosa Acquisita

Myasthenia Gravis

Rheumatoid Arthritis

Pemphigus

Multiple Sclerosis

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

Hematology Disorders

Blistering Diseases

Chronic Inflammatory
Demyelinating Polyneuropathy
(CIDP)

Phase 2 CIDP study to start in 2H 2019



Consistent and favorable tolerability profile



Rapid and sustained benefit; clinically meaningful and statistically significant



Strong correlation between IgG level reduction and disease improvement; supporting focus on IgG-mediated diseases



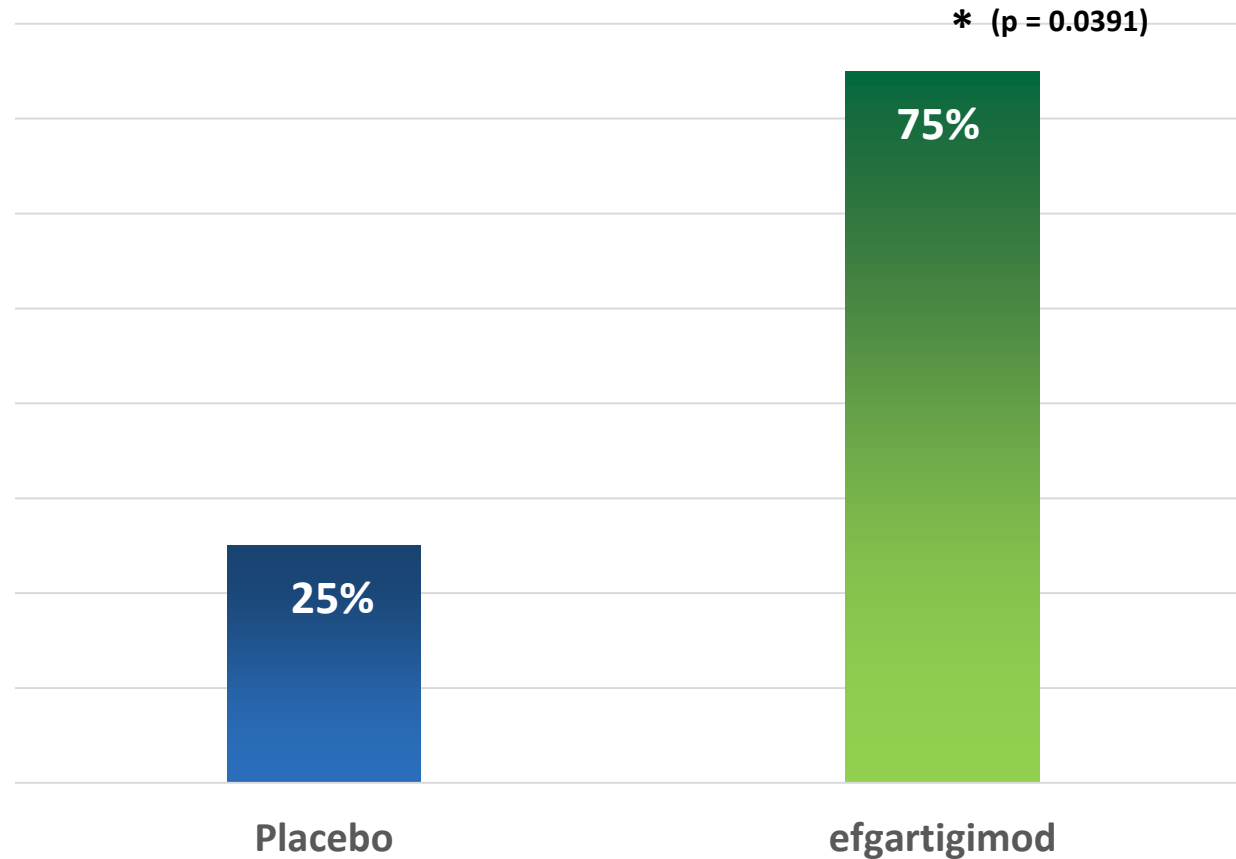
Significant reduction of AChR autoantibodies



Phase 3 ADAPT study ongoing with data expected in 2020

75% of Treated Patients Achieved Lasting Response

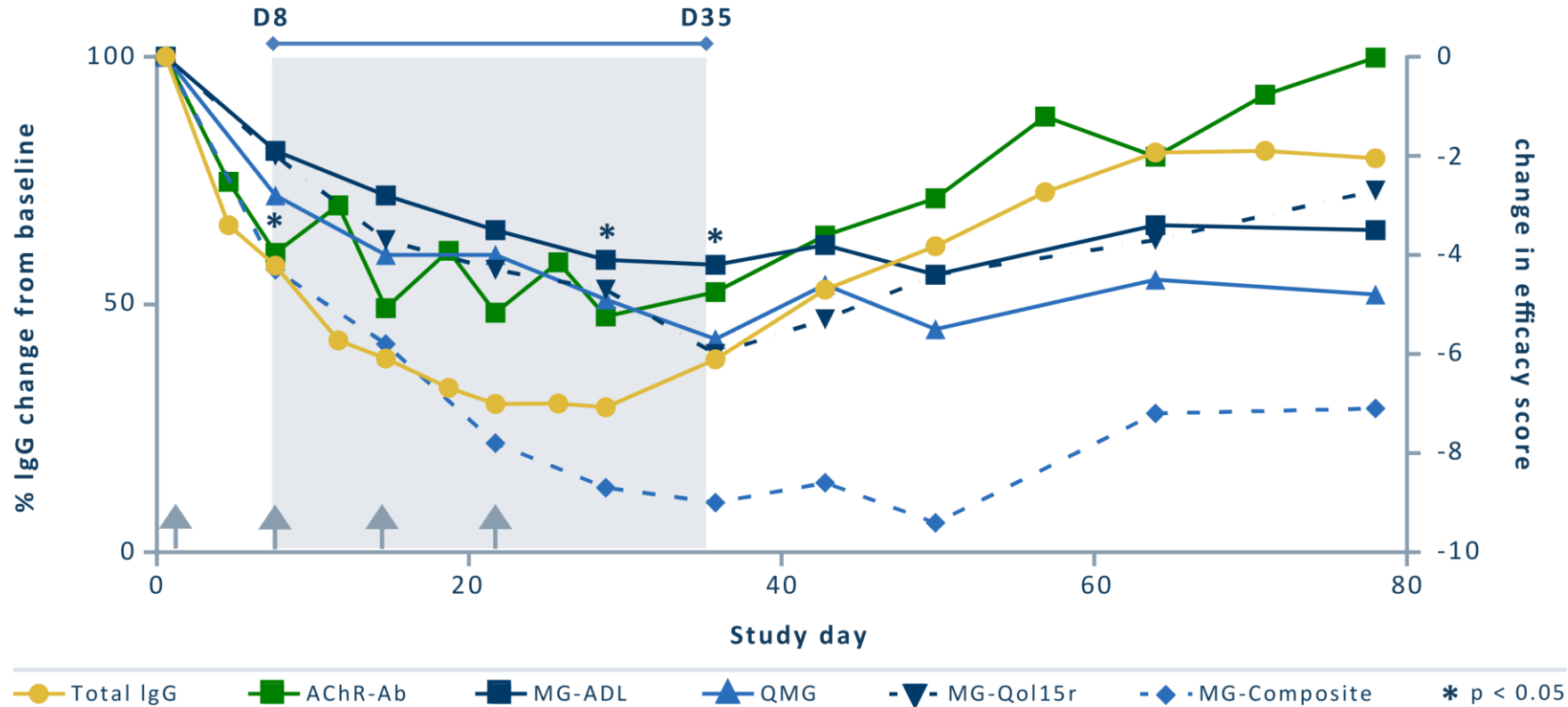
Patients with MG-ADL ≥ 2 for a period of at least 6 weeks



- 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 & Pathogenic IgG Reduction Correlates with Clinical Improvements

Assessment for all efficacy scales

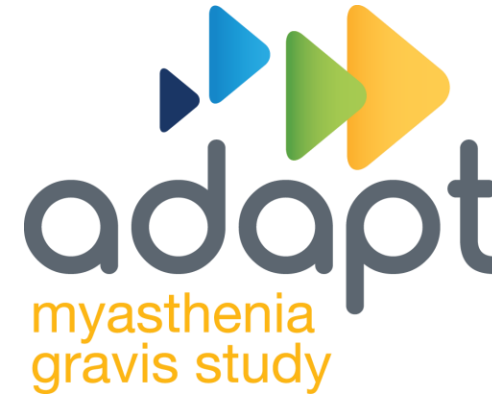


- Clinical improvement persists despite return of IgG levels
- Potential differentiation from PLEX where clinical benefit was reported to be lost 2-4 weeks after end of treatment ⁽¹⁾

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**

ITP Phase 2 Results Establish Hematologic Beachhead

Novel approach beyond boosting platelet production or broad immuno-suppression



Favorable and consistent safety and tolerability profile

- No trends seen for infections or headaches across all studies
- No decreases in IgM, IgE, IgA or albumin



Robust efficacy signal in relapsed/refractory population after short drug exposure

- Clinically meaningful increase in platelet counts over placebo
- 50% of patients came on study with platelets $<15 \times 10^9$



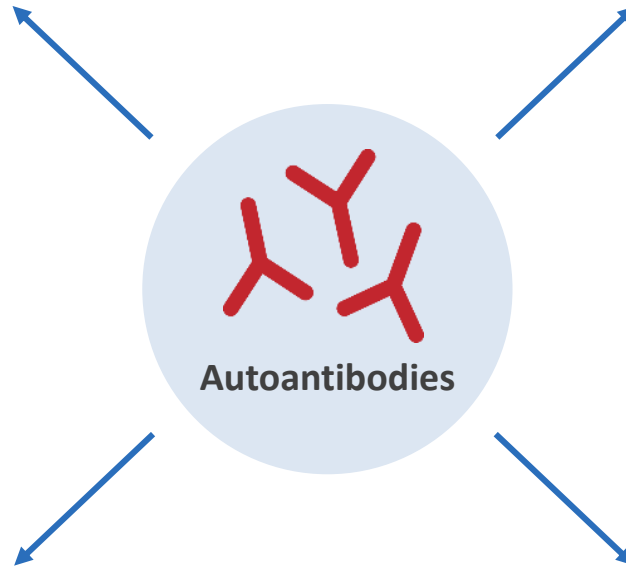
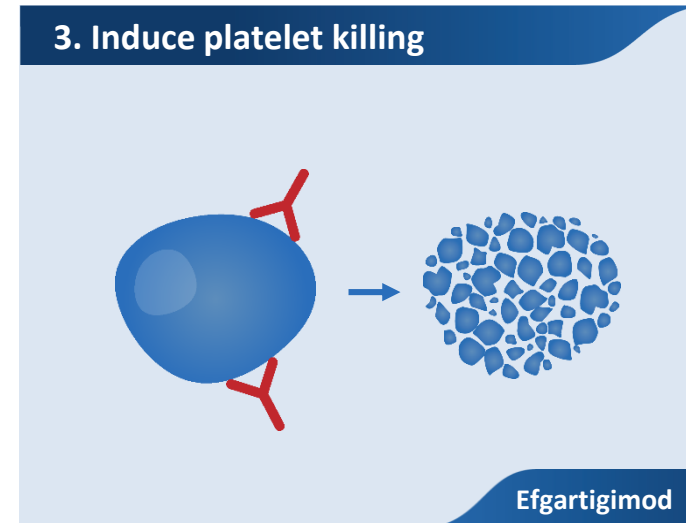
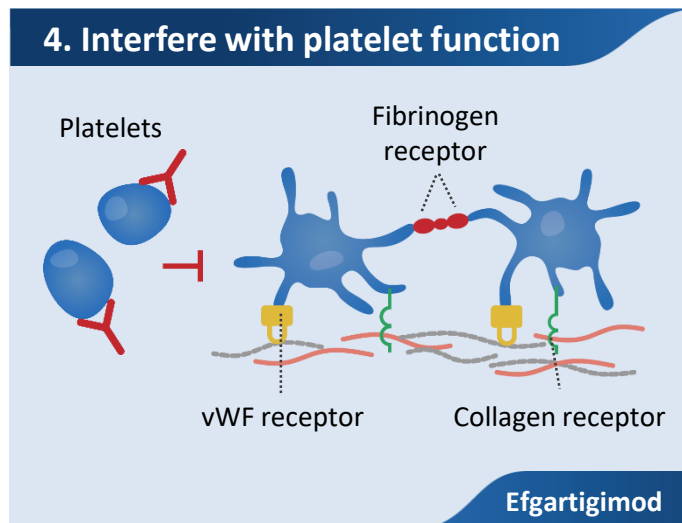
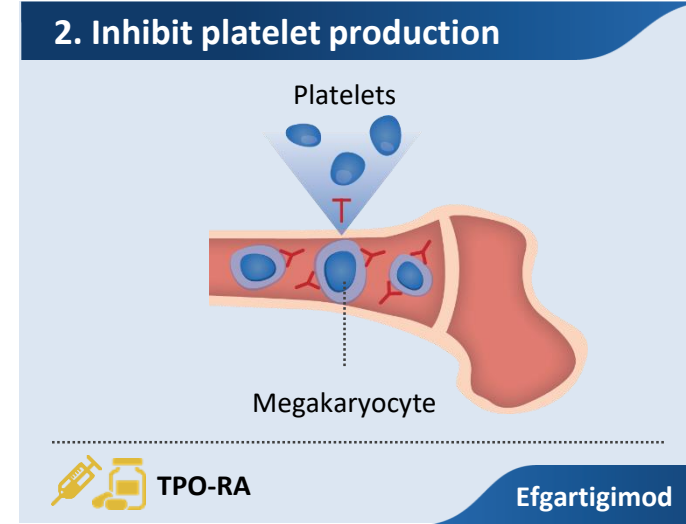
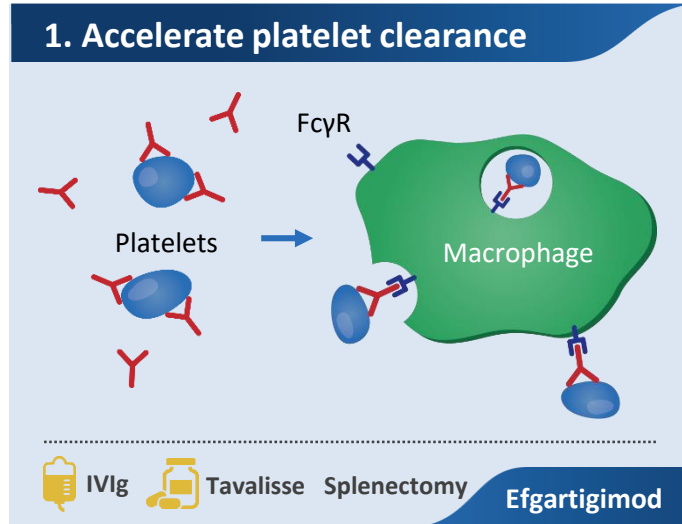
Strong correlation between IgG reduction, platelet count improvement and reduction of bleeding events



Phase 3 ITP Program to start 2H19 with update on path forward in 3Q19

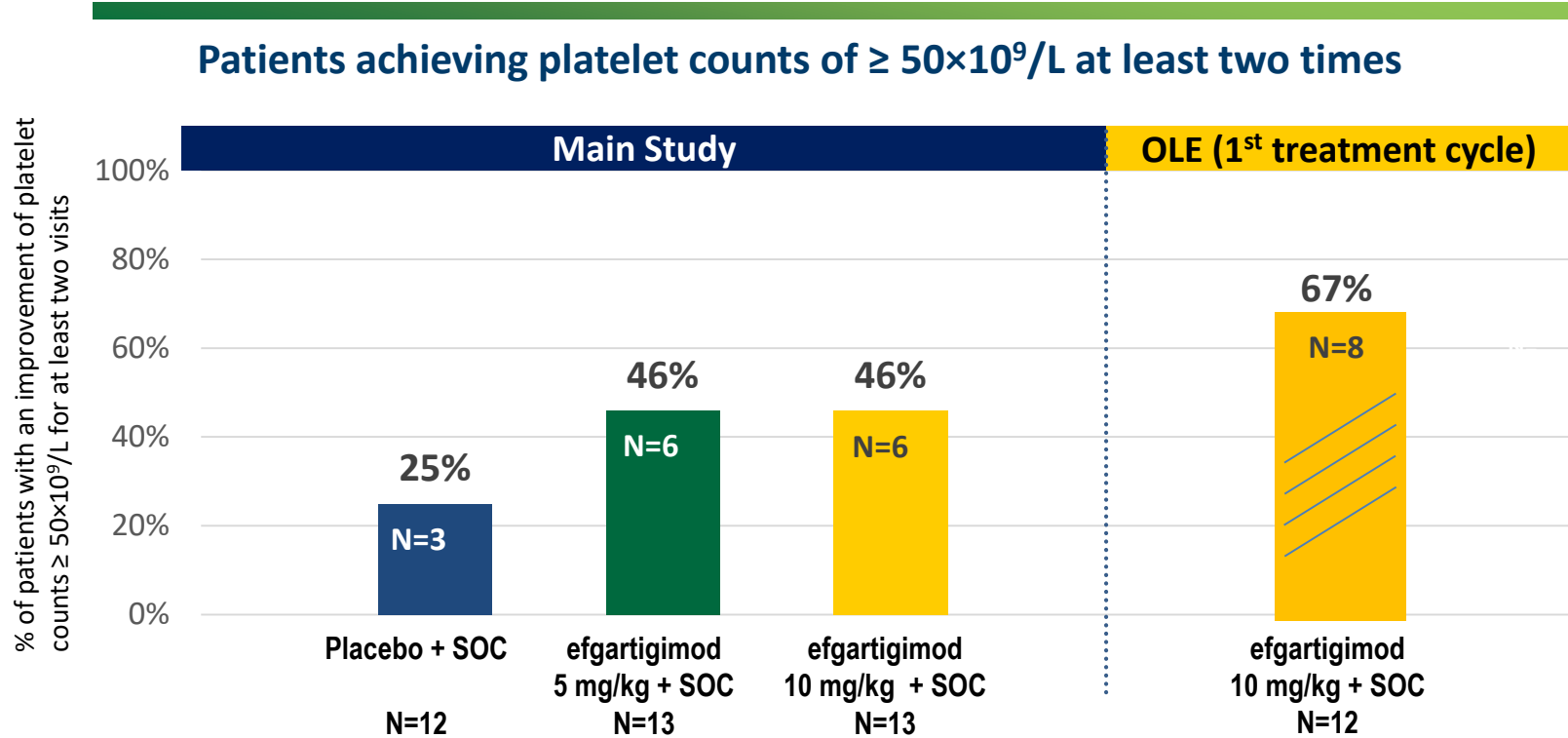
Efgartigimod Targets All Pathogenic AutoAb Actions Simultaneously

Potential to eliminate therapeutic cycling based on trial-and-error



Strong Improvement of Platelet Counts Across Doses

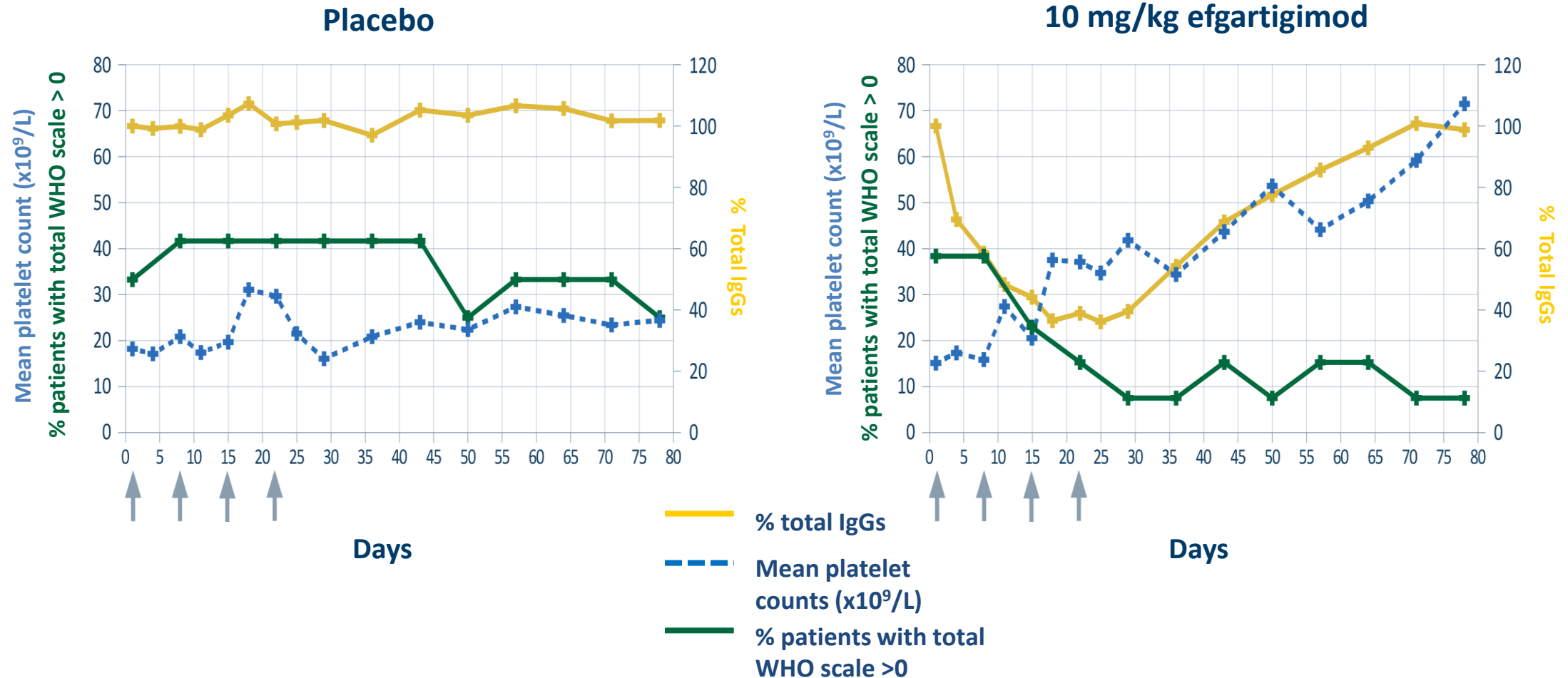
46-67% of patients exceeded platelet counts $\geq 50 \times 10^9/L$ during at least two visits



- 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

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

Mean platelet counts versus total WHO scale versus total IgGs





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

Pemphigus Vulgaris Phase 2 Adaptive Design

Phase 2 data expected in 2020

Treatment Phase

Induction

3 weeks

COHORT 1: N= 4

efgartigimod (10 mg/kg)

4 infusions (weekly)

Maintenance

6 weeks → 8 weeks

efgartigimod (10 mg/kg)

2 infusions (w2, w6)

COHORT 2: N= 4 + 4

efgartigimod (10 mg/kg)

4 infusions (weekly)

efgartigimod (10 mg/kg)

4 infusions (w2, w4, w6, w8)

Follow-up Phase

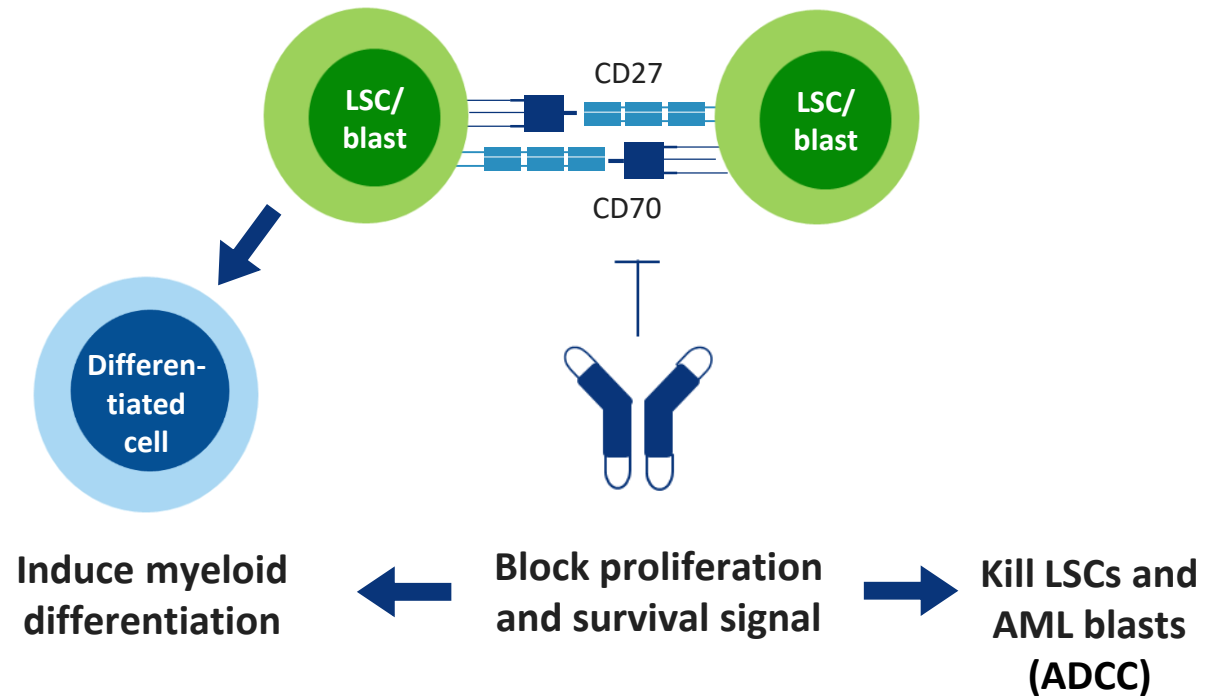
8 weeks

IDMC recommendation for Cohort 3 to reach clinical remission (with/without minimal therapy):

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

Cusatuzumab: Potential Foundational, Novel Therapy for Acute Myeloid Leukemia

Multiple MOA of Cusatuzumab



- Novel target and mechanism of action¹ (inhibition of CD70 pathway)
- Intrinsic activity shown as a single-agent in AML
- Potential for combination therapy²
- 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 patients³
- IAP, Bern University – Prof. Ochsenbein

Cusatuzumab Strategic Alliance with Janssen Pharmaceuticals



argenx objectives

Accelerate & broaden development plan

Secure strong financials

Retain commercial upside

Janssen alliance

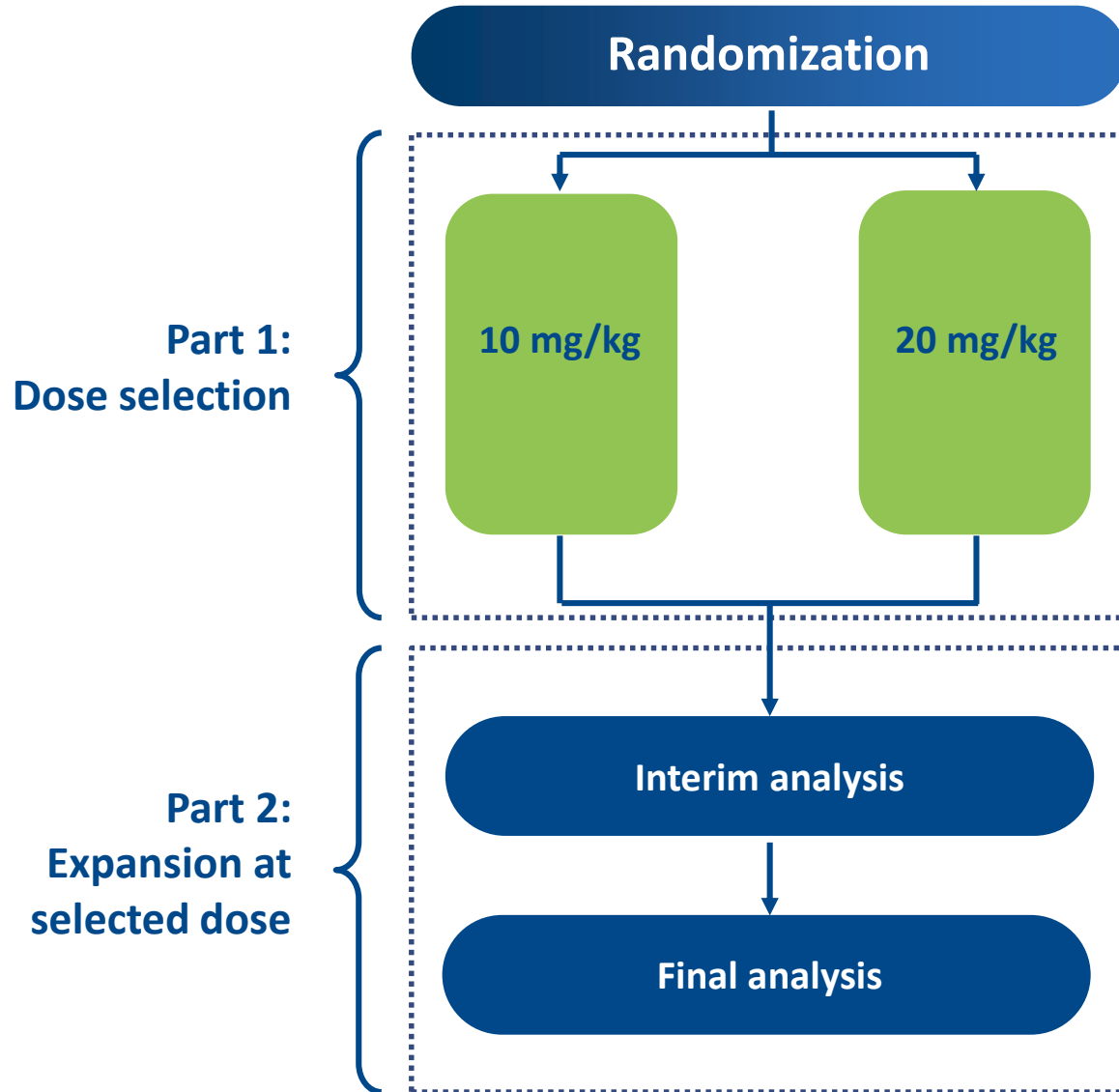
Joint development plan focused on AML, MDS and other heme malignancies

Upfront \$300M + \$200M equity @ 20% premium, \$1.3B in milestones, double digit royalties OUS

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

Anticipated Phase 2 study start in US in 2H19

Phase 2 Study in Newly Diagnosed, Unfit AML Patients



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)

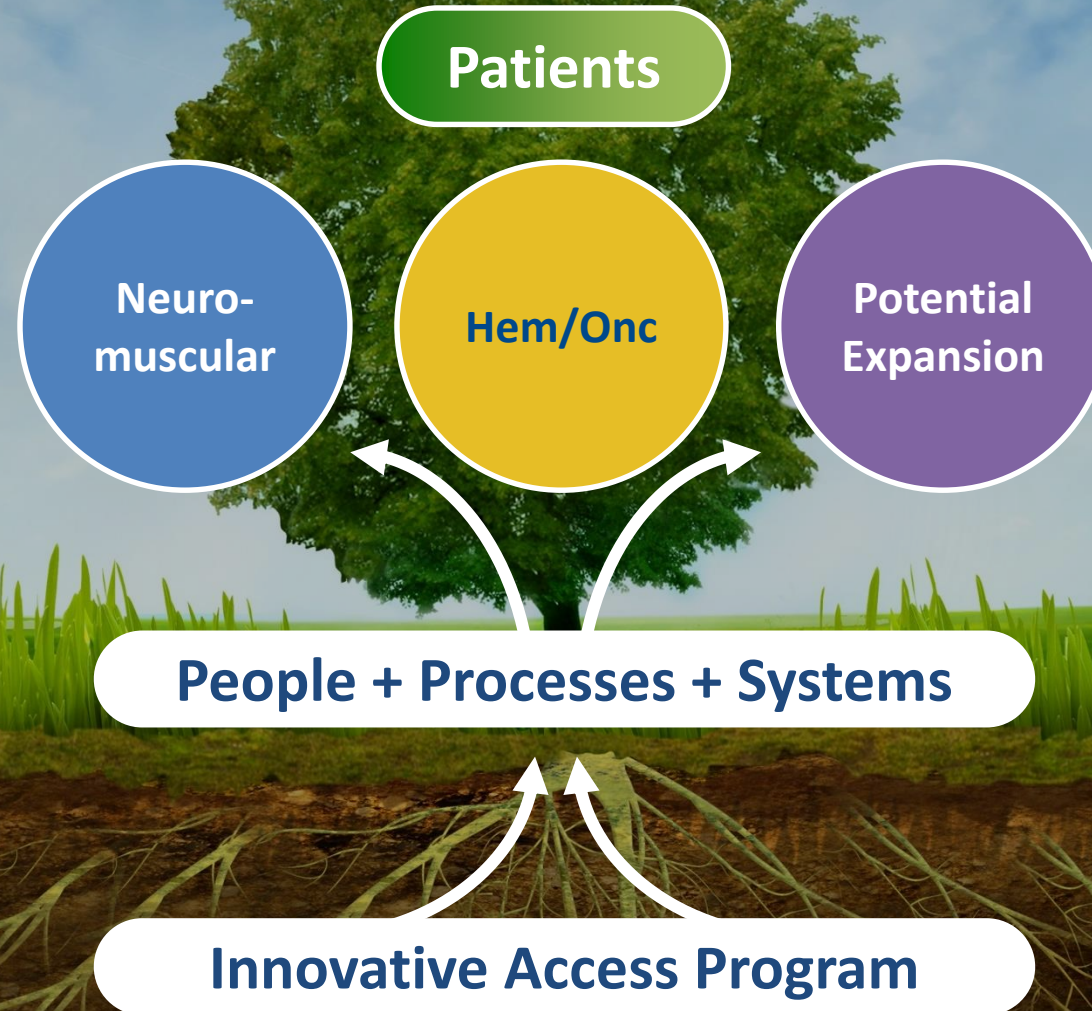
Secondary Objectives:

- 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 in US: second half 2019


Commercial





Building Immunology Franchises

Neuro Inflammation Poly Neuropathies Myositis

 Expansion Opportunities

MG CIDP MMN

Neuro-muscular

BMT AIHA ANCA Leukemia Lymphomas

 Expansion Opportunities

MALIGNANT HEME BENIGN HEME

AML  MDS ITP

janssen 
50/50

Hem/Onc

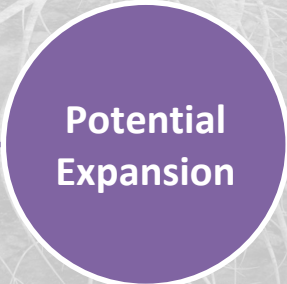
Complement or IgG-mediated Diseases in Skin and Kidney

- Pemphigus
- Dermatomyositis
- Bullous Pemphigoid



- Lupus Nephritis
- Nephropathies

- AMR
- IRI



Building the Experienced, Diverse Organization

Business Analytics



Distribution



Finance



Patient Advocacy



Human Resources



Legal / Compliance



Marketing



Market Access



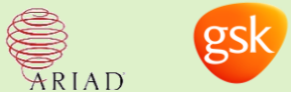
Pharmacovigilance



Strategic Insight



Sales Leadership



Regulatory Affairs



Medical Affairs



Japan GM



EU Commercial Dev Leader





June 2019