Developing Highly Differentiated Antibody Therapeutics

argenx

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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

5

Multi-Asset Late-Stage Platform

- Phase 3 in MG and ITP
- Pre-commercial activities in MG

3

Highly Productive Development Engine

- Rapid development timeline
- New candidate each year

4

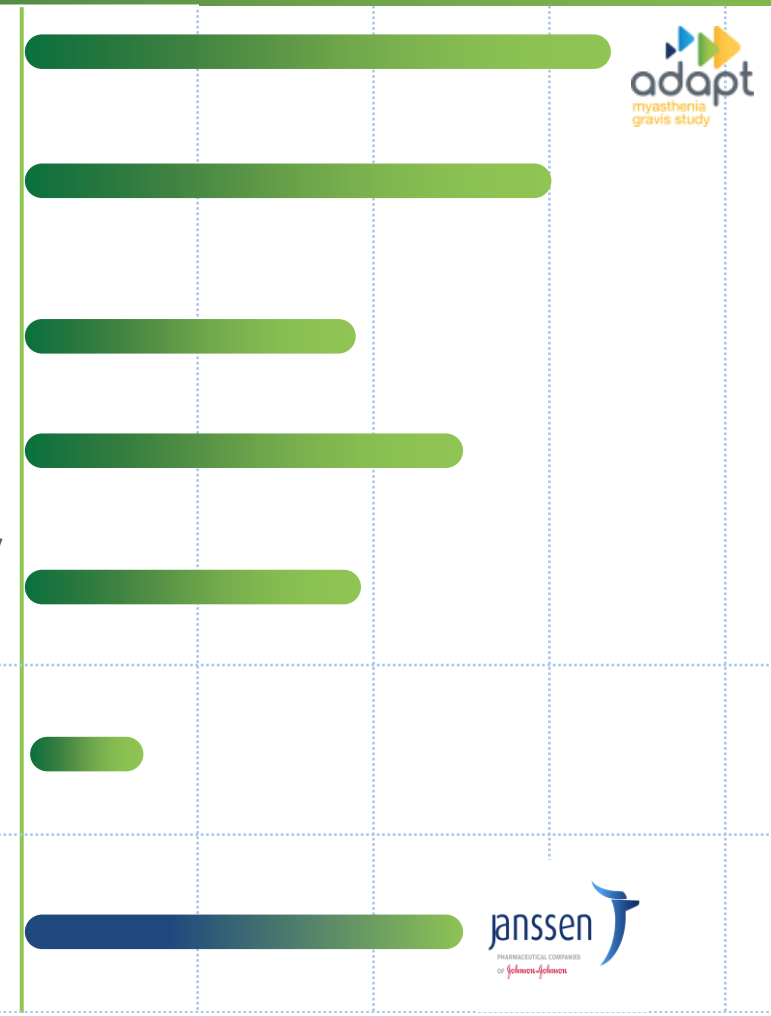
Maximum Value per Asset

- Pipeline-in-a-product strategy
- Strong biological rationale

Translate immunology breakthroughs into novel medicines which truly impact patients' lives

Deep Pipeline of Wholly-Owned Candidates for Orphan Indications

Product Candidate	Target	Indication	Next Milestone / Commentary
Wholly-Owned & Co-Development Product Candidates			
ARGX-113 Efgartigimod	FcRn	Myasthenia Gravis	3Q18: Phase 3 initiated
		Immune Thrombocytopenia (ITP)	2H19: Phase 3 initiation
		ITP Subcutaneous Formulation	1H19: Phase 2 initiation
		Pemphigus Vulgaris	1H19: Cohort 3 initiation
		Chronic Inflammatory Demyelinating Polyneuropathy	2H19: Phase 2 initiation
ARGX-117	Novel complement target	Severe Autoimmune Diseases	Antibody-mediated autoimmune diseases Complementary to ARGX-113
ARGX-110 Cusatuzumab	CD70	Acute Myeloid Leukemia	$500 million upfront Eligible for up to $1.3 billion in milestones; tiered royalties



Innovative Access Program Allows Strategic Partnering

Partner activity focused in therapeutic areas outside severe autoimmune and cancer

Product Candidate	Target	Indication	Preclinical	Phase 1	Phase 2	Phase 3	BLA	Next Milestone / Commentary
Partnered Product Candidates								
ARGX-112 LEO	IL-22R	Skin Inflammation						Eligible for up to ~€100mm in milestones; tiered royalties
ARGX-115 abbvie	GARP	Cancer Immunotherapy		AbbVie exercised option to develop and commercialize in August 2018				Received $60mm in upfront and preclinical milestone payments Eligible for up to $625mm milestones; tiered royalties
ARGX-116 STATEN BIOTECHNOLOGY	ApoC3	Dyslipidemia						Eligible for double-digit royalties and exclusive option to license the program; collaboration with Novo Nordisk

- **Innovative Access Program**: 7 live programs
- Antibody discovery alliance with Shire focused on multiple rare disease targets – 2 options exercised
- Additional programs include 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)

Efgartigimod:
A Pipeline-in-a-Product Opportunity

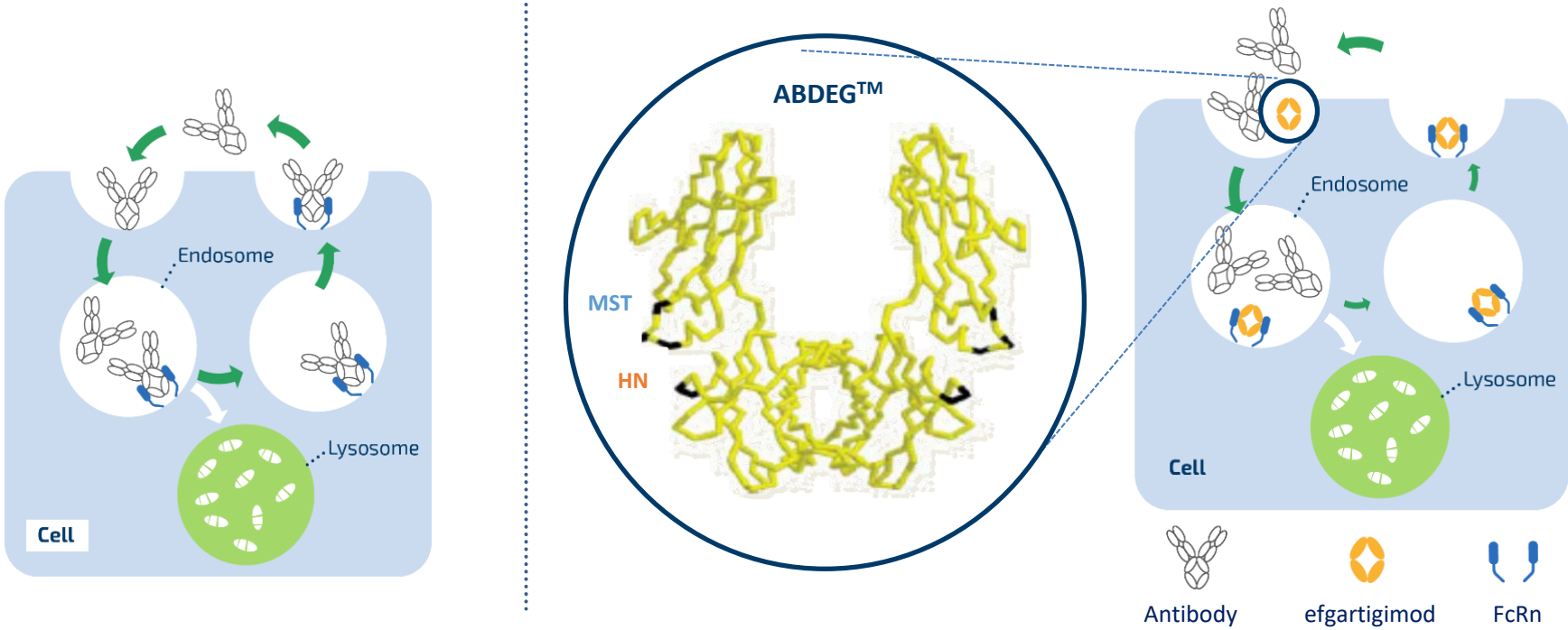
Efgartigimod: Human IgG1 Fc Fragment with ABDEG™ Mutations

Exploits Natural Fc/FcRn Interaction

IgG antibodies recycle through FcRn(1)…

efgartigimod potently blocks FcRn…

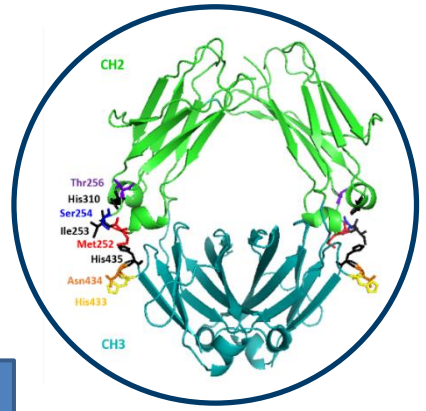
leading to IgG elimination(2)



(1) Roopenian et al. 2007, Nat Rev Immunol.
(2) Vaccaro et al. 2005, Nat Biotech.
(3) Ulrichts et al. 2018, J Clin Invest.
(4) argenx data

Efgartigimod Emerges as First-In-Class and Best-In-Class

Human IgG1 Fc fragment with ABDEG™ mutations



- **Natural ligand of FcRn**
- **Enhanced, pH dependent binding**

First-in-class features

Best-in-class clinical attributes

Reduced FcγR, C1q binding → **Clean safety & tolerability profile (~120 subjects)**
- No headache or GI AE profile

Endosomal recycling FcRn-efgart complex; no lysosomal degradation → **No decrease in albumin**

Can rebind FcRn → **Long half-life; unparalleled tissue penetration & distribution**

Long-lasting, potent PD effect; fast onset of clinical benefit

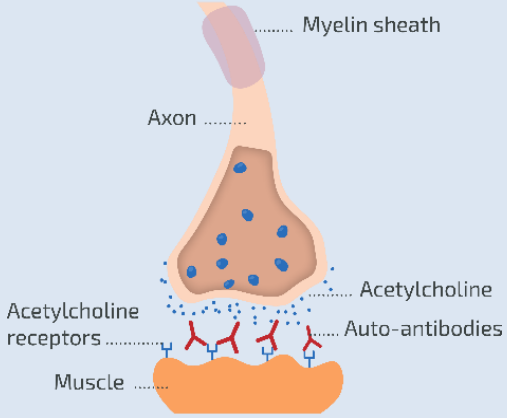
1/3 size of IgG; excellent physicochemical stability → **Lower dose enables convenient subQ administration, high concentration formulations and lower COGS**

Novel Treatment Modality in Severe Autoimmune Diseases

Efgartigimod Beachhead Indications

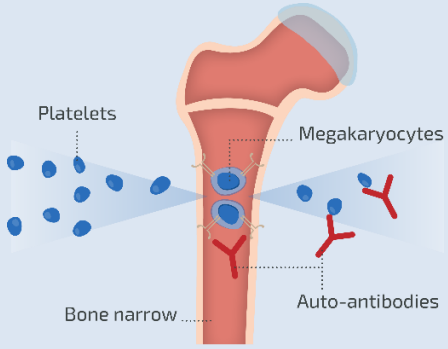
Myasthenia Gravis

- Block acetylcholine receptors
- Cross-link + internalize AChRs
- Complement recruitment



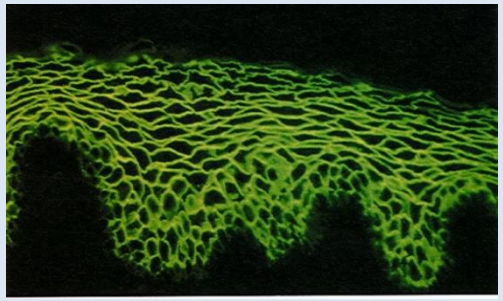
Immune Thrombocytopenia

- Enhance platelet clearance
- Platelet killing
- Inhibit platelet production
- Reduced platelet function



Pemphigus Vulgaris

- Sterically hinder epithelial adhesion affecting skin and mucosal integrity



Common Characteristics of Beachhead Indications

- Pathogenic auto-antibodies causal to disease biology
- Typical treatment options: corticosteroids, broad immunosuppressants, IVIG, plasmapheresis, Rituxan – with mixed response rates and serious side effects
- Orphan potential in U.S. (MG: 50-60K[1]; ITP: 50K[2]; PV: 30-40K[3])
- Potential pharmacoeconomic benefit to healthcare system given price of targeted therapies (e.g., Soliris for refractory MG ~$700K / year)[4]

(1) Philips et al. 2003, Ann N Y Acad Sci; Drachman et al. 1993, New Eng J Med
(2) Wall street research; Estimated 65K ITP patients in US with ~80% diagnosed with primary ITP
(3) IPPF (www.pemphigus.org)
(4) Source: Reprinted with permission by First Databank Inc.; WAC = Wholesale Aquisition Cost 8/21/17

Phase 2 Study of Efgartigimod in Generalized Myasthenia Gravis

- Consistent and favorable tolerability profile
- Fast, long-lasting 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 2 execution advances efgartigimod into Phase 3 (initiated)

Efgartigimod Safety And Tolerability Profile

2 hour infusion enabling outpatient administration

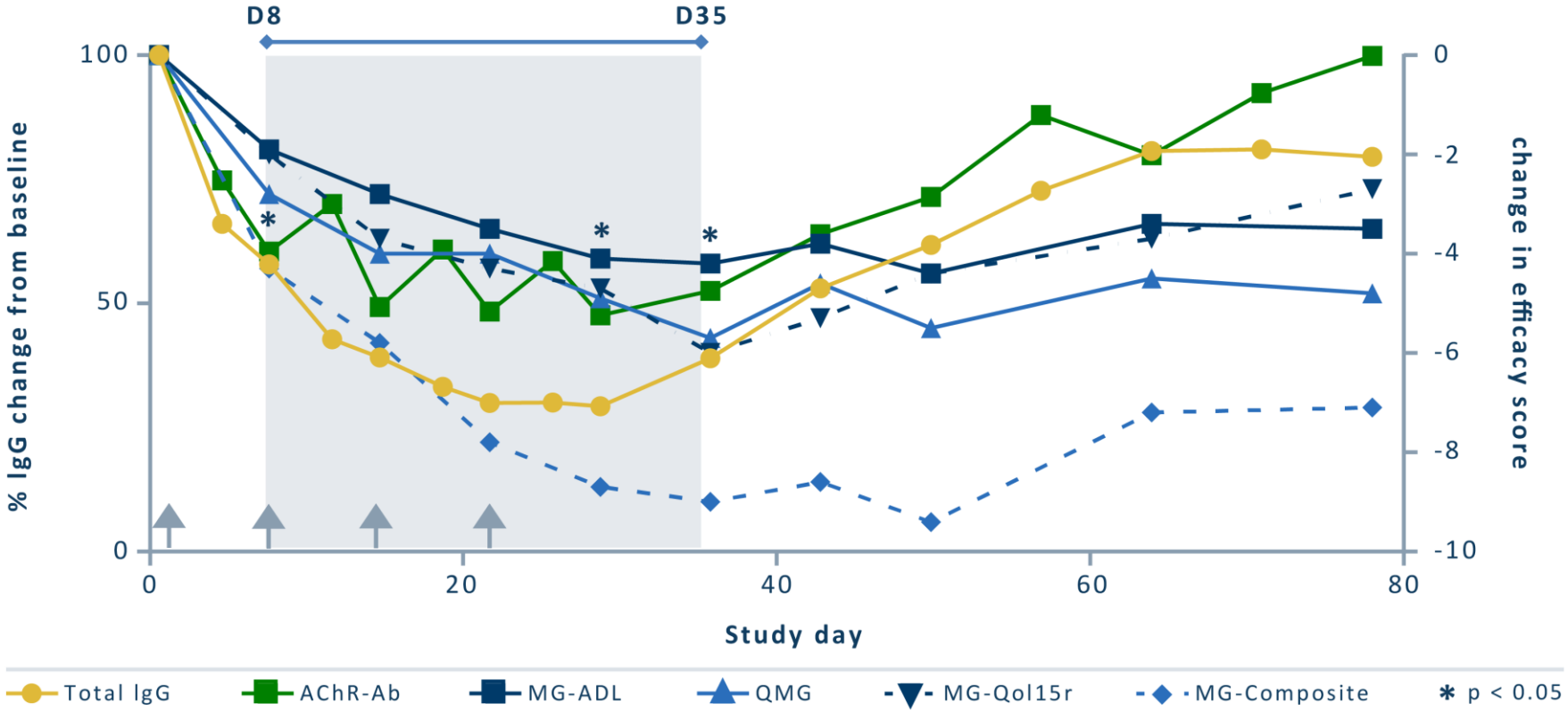
- Efgartigimod was well-tolerated in patients; confirmed findings from Phase 1 healthy volunteer trial
- TEAE profile was balanced between efgartigimod and placebo
- TEAEs were mostly mild (grade 1) in severity; no severe AEs were reported
- No deaths, serious AEs or TEAEs leading to discontinuation of treatment were reported during trial

Treatment Emergent Adverse Events (TEAEs) Reported in ≥ 2 patients	Placebo (N = 12)	Efgartigimod (N = 12)
TEAEs (Total)	**10 (83.3%)**	**10 (83.3%)**
• Headache	3 (25.0%)	4 (33.3%)
• Nausea	1 (8.3%)	1 (8.3%)
• Diarrhea	1 (8.3%)	1 (8.3%)
• Abdominal pain upper	1 (8.3%)	1 (8.3%)
• Arthralgia	2 (16.7%)	-
• B-lymphocyte decrease	-	2 (16.7%)
• Lymphocyte count decrease	-	2 (16.7%)
• Monocyte count decrease	-	2 (16.7%)
• Neutrophil count increase	-	2 (16.7%)
• Myalgia	-	2 (16.7%)
• Pruritus	2 (16.7%)	1 (8.3%)
• Rhinorrhea	1 (8.3%)	1 (8.3%)
• Tooth abscess	2 (16.7%)	-
• Toothache	2 (16.7%)	-
Efgartigimod deemed related TEAEs	**3 (25.0%)**	**8 (66.7%)**
• Headache	1 (8.3%)	3 (25.0%)
• Monocyte count decrease	0 (0.0%)	2 (16.7%)
• Rhinorrhea	1 (8.3%)	1 (8.3%)

argenx data

Total & Pathogenic IgG Reduction Correlates with Clinical Improvements

Assessment for all efficacy scales

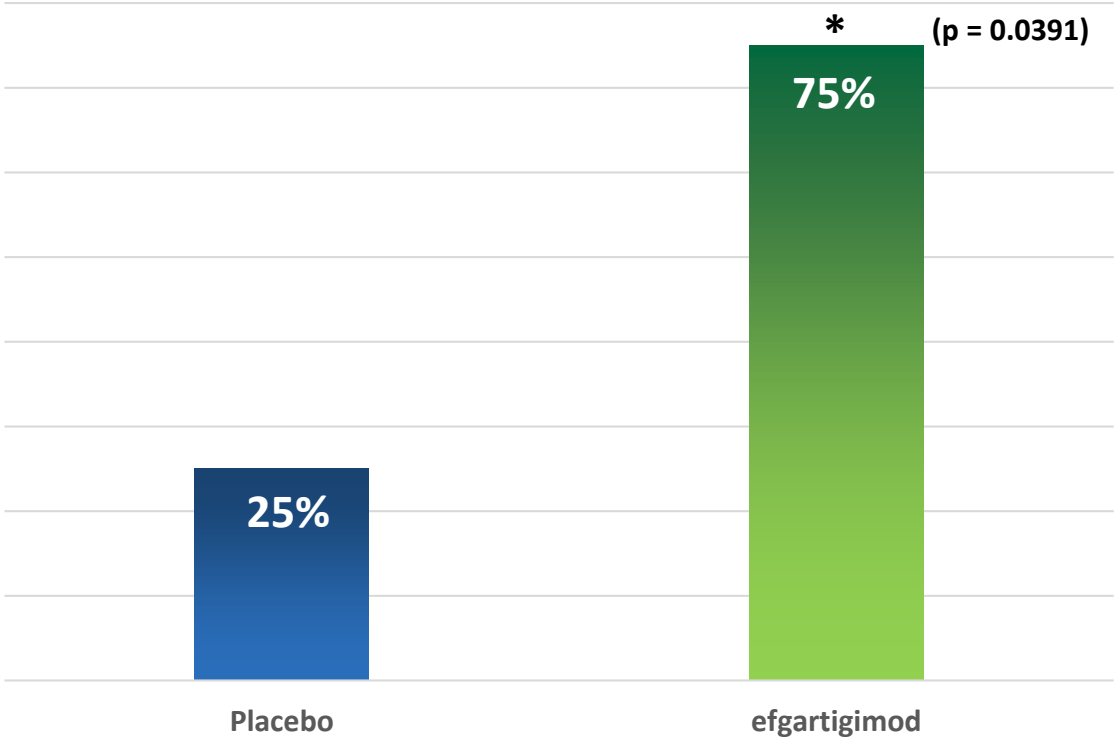


- Clinical improvement persists despite return of IgG levels
- Potential differentiation from PLEX where clinical benefit was reported to be lost 2-4 weeks after end of treatment [1]

(1) Kuks and Skallebaek, 1998, Transfus Sci

75% of Treated Patients Achieved Lasting Response

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



- 83% of patients treated with efgartigimod achieved a clinically meaningful response (MG-ADL ≥2)
- 75% of patients treated with efgartigimod had a clinically meaningful and statistically significant improvement in MG-ADL score for a period of at least 6 consecutive weeks versus 25% of patients on placebo

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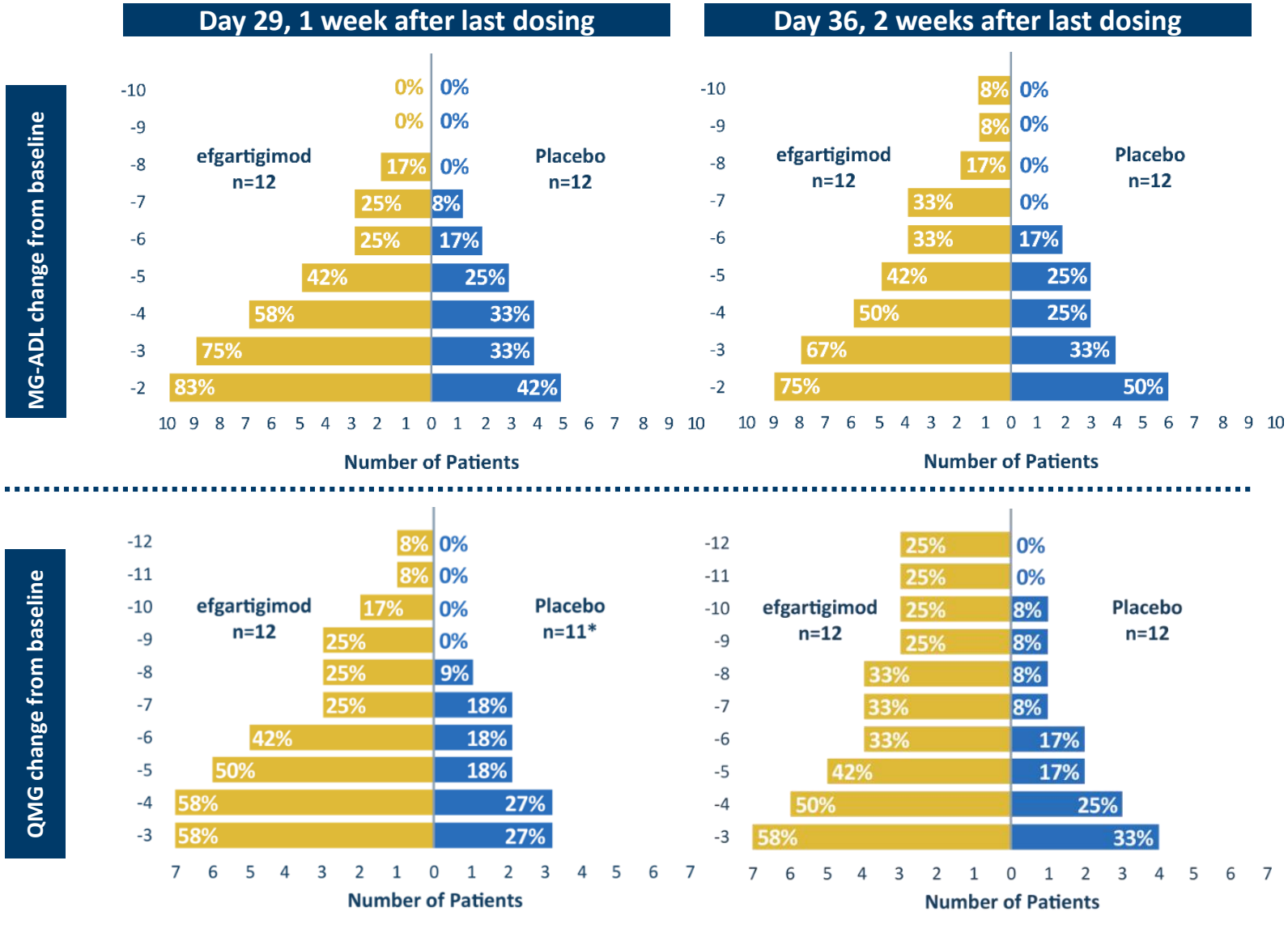
Robust Clinical Improvement Over Placebo Group



- Efgartigimod *vs.* placebo: increasing differentiation observed with increasing MG-ADL/QMG thresholds

* Missing data point of 1 patient

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

adapt
myasthenia gravis study

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

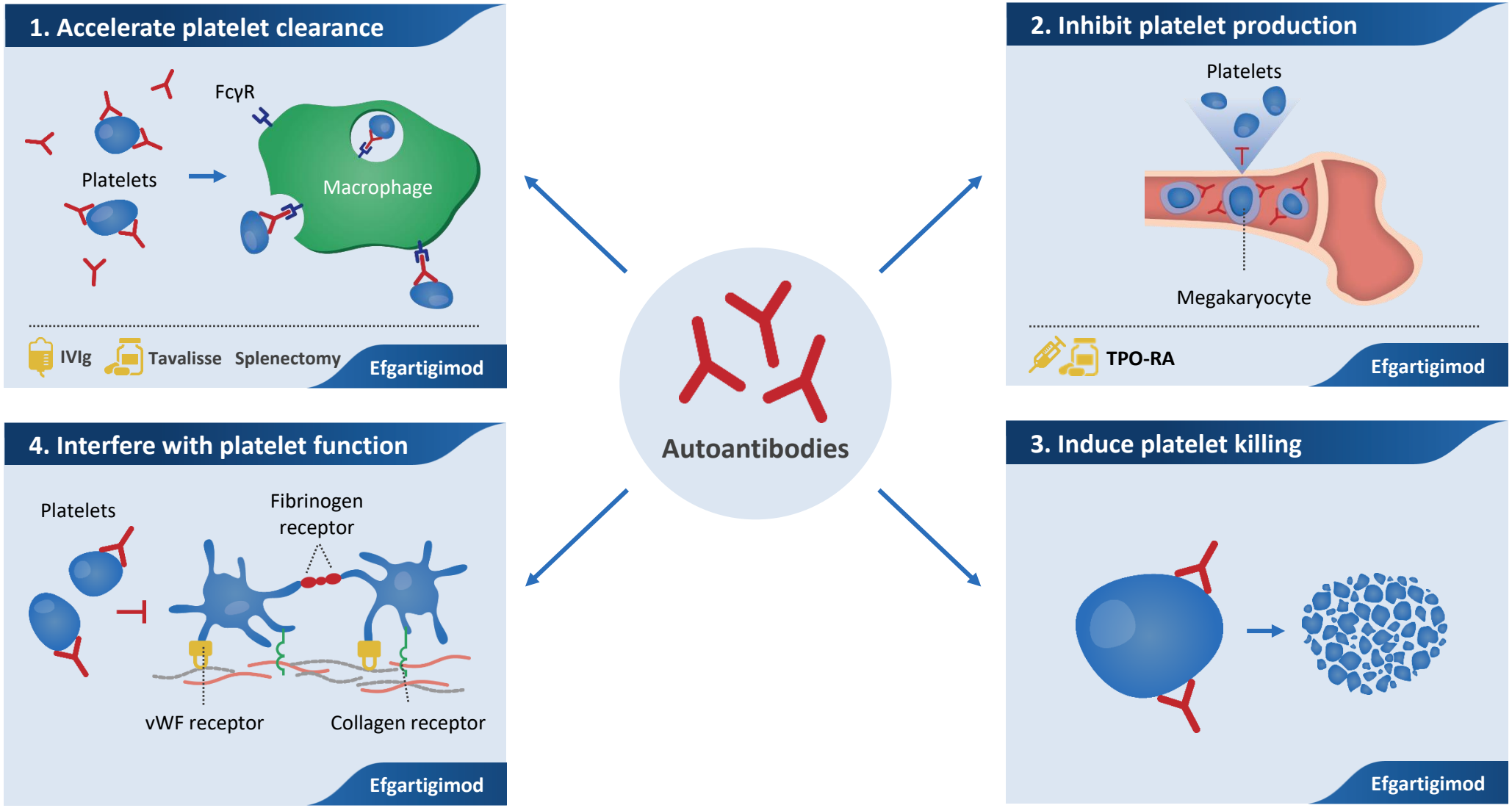
- Clinically meaningful increase in platelet counts over placebo
- 50% of patients came on study with platelets <$15x10^9$

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

Data enable Phase 3 in ITP (IV) and launch of Phase 2 in ITP (SC)

Efgartigimod Targets All Pathogenic AutoAb Actions Simultaneously

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



Favorable Tolerability Profile Consistent with Previous Studies

Treatment-emergent adverse events balanced between active and placebo arms

- **Tolerability profile consistent with Phase 2 myasthenia gravis (MG) and Phase 1 healthy volunteer (HV) trials**
- **TEAEs mostly mild in severity (grade 1)**
- **No deaths or TEAEs leading to discontinuation of treatment reported***

Bleeding TEAEs not included

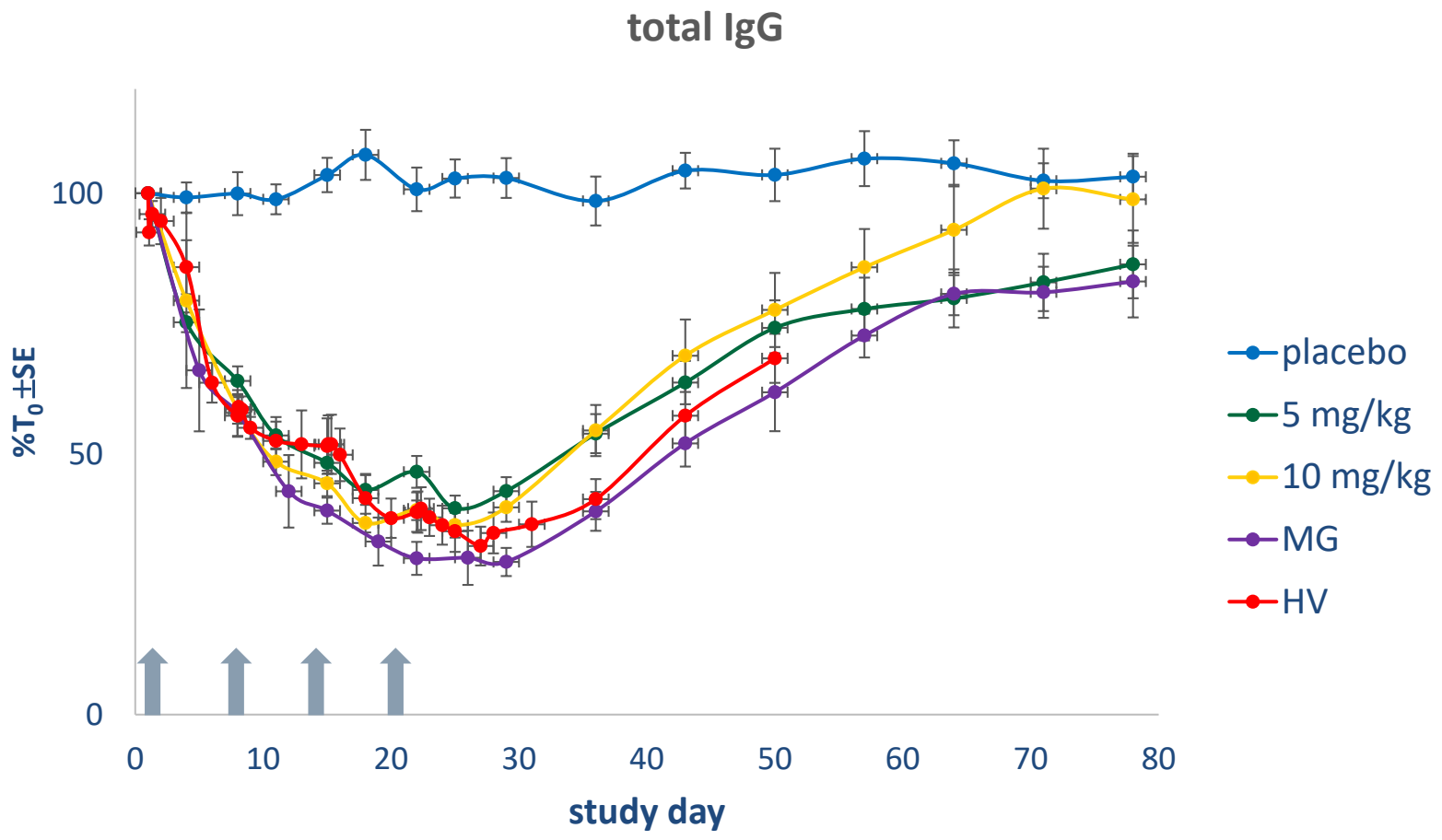
Treatment Emergent Adverse Events (TEAEs) Reported in ≥ 2 subjects	Placebo (N = 12)	Efgartigimod 5 mg/kg (N = 13)	Efgartigimod 10 mg/kg (N = 13)
Most common TEAEs N (%)			
• Headache	2 (16.7)	1 (7.7)	-
• Hypertension	1 (8.3)	-	2 (15.4)
• Vomiting	-	-	2 (15.4)
• Cystitis	-	1 (7.7)	1 (7.7)
• Rash	-	1 (7.7)	1 (7.7)
• Productive cough	1 (8.3)	1 (7.7)	-
TEAEs *deemed related* to study intervention N (%)			
• Headache	1 (8.3)	-	-
• Vomiting	-	-	1 (7.7)
• Pubic pain	1 (8.3)	-	-
• Vaginal discharge	1 (8.3)	-	-
• Amenorrhoea	1 (8.3)	-	-

* One thrombocytopenia downgraded per protocol after database lock

argenx data: Table 14.3.1.2a & 14.3.1.5a - Treatment-Emergent Adverse Events by System Organ Class, Preferred Term and Relationship to Study Drug - Main Study

Efgartigimod Leads to Lasting IgG Reduction Across Studies

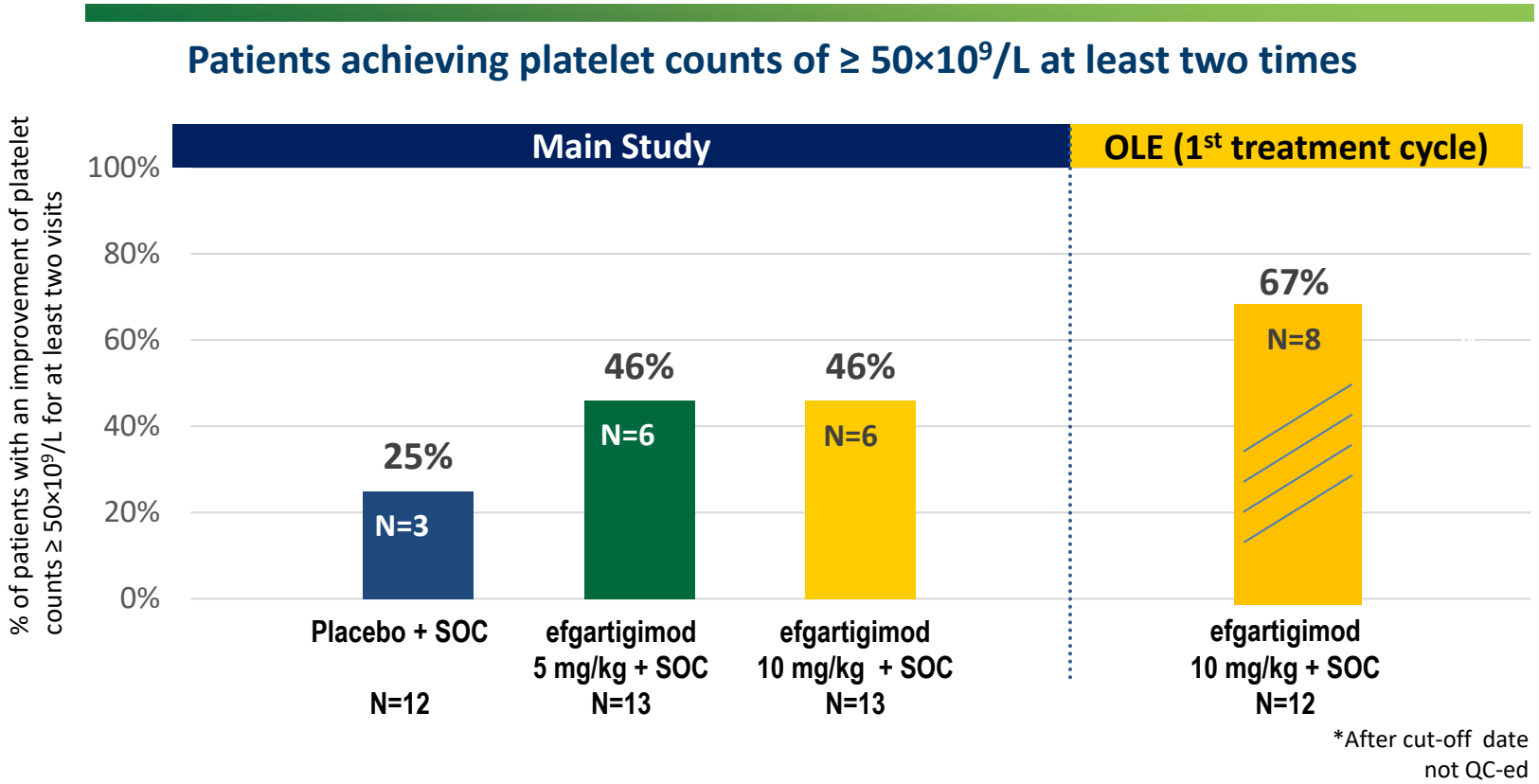
Total IgG levels in efgartigimod studies to date (Healthy Volunteers, MG, ITP)



- Pharmacodynamics (PD) closely align with Phase 1 trial in HV and Phase 2 trial in MG
- IgM, IgA and albumin levels not affected (data not shown)
- Half-life: approx. 5 days
- Pharmacokinetics (PK) very similar to Phase 1 trial in HV and Phase 2 trial in MG (data not shown)
- Low titer of anti-drug antibodies (ADA) seen in 16.7% placebo patients vs. 30.8% efgartigimod patients (10 mg/kg) with no apparent effect on PK/PD

Strong Improvement of Platelet Counts Across Doses

46-67% of patients exceeded platelet counts ≥ 50X10^9/L during at least two visits

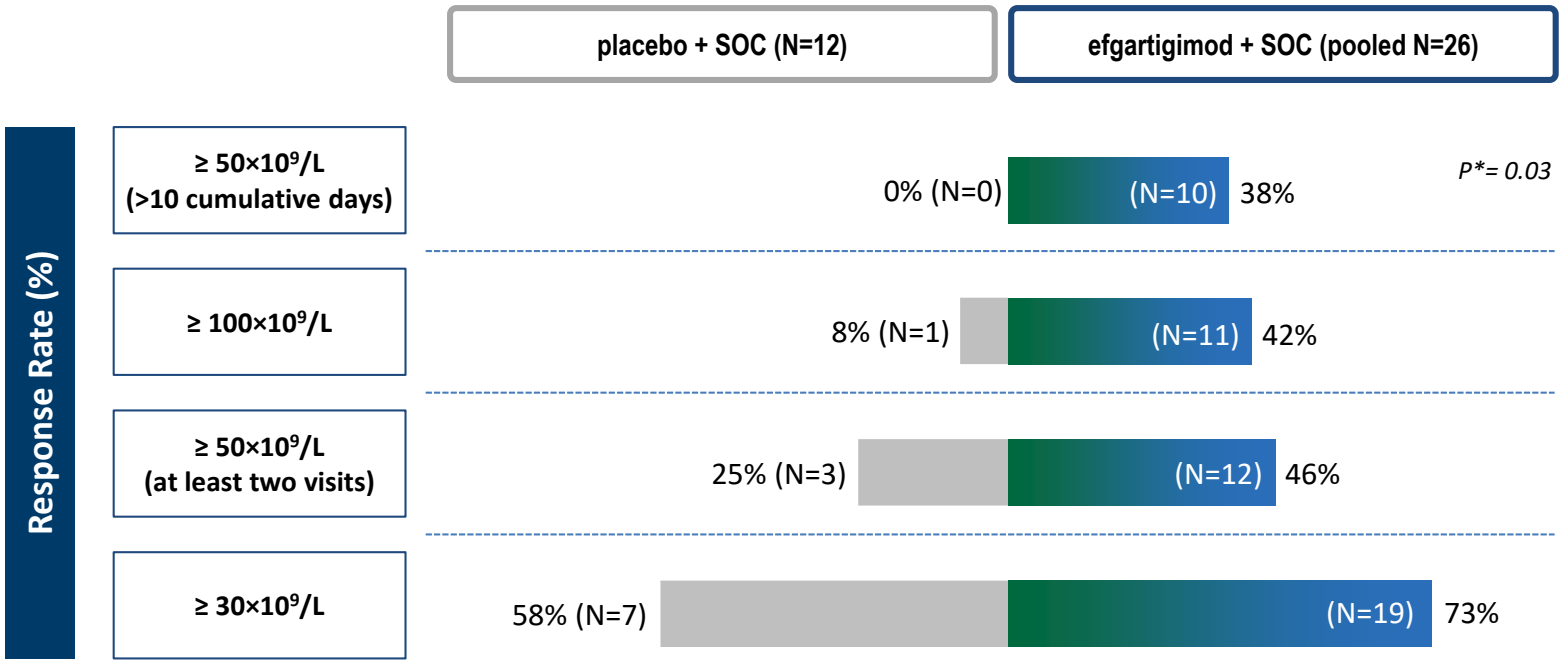


*After cut-off date not QC-ed

- OLE acts as true fourth cohort since patients' platelets had to fall below 30x10^9/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

Robust Improvement of Platelet Count

Post-hoc analysis of increasing thresholds of efficacy

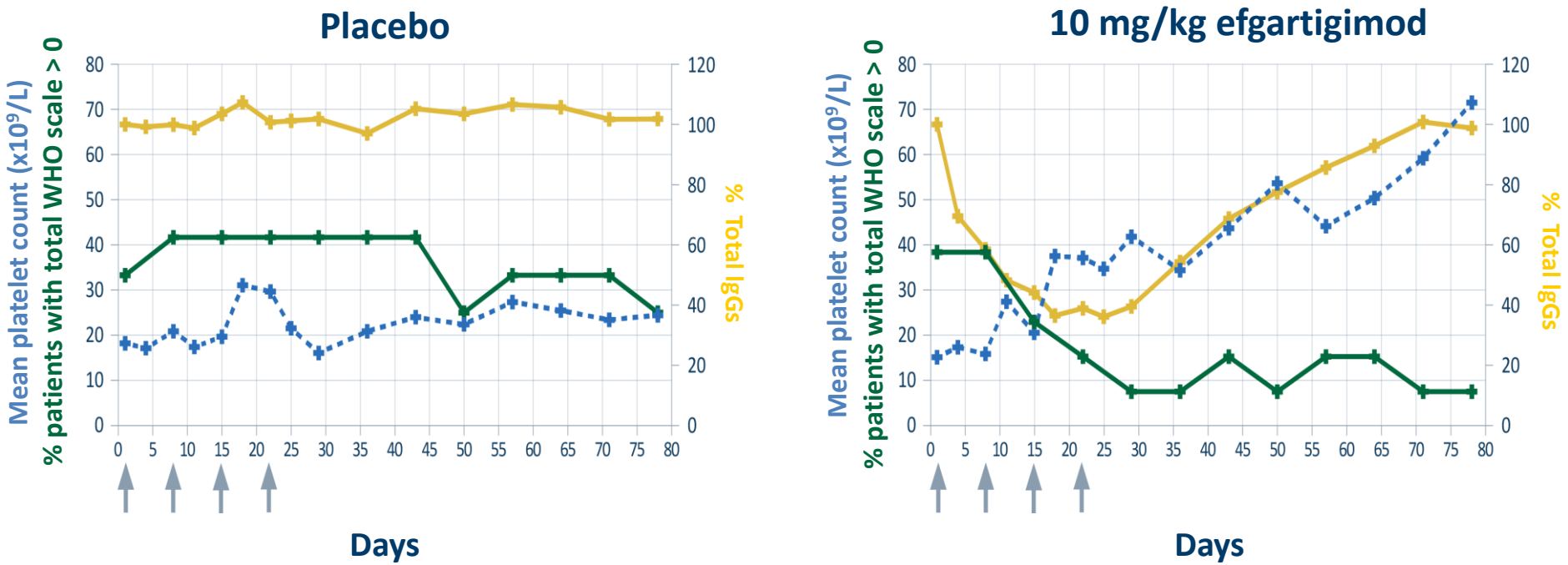


- Efgartigimod generated therapeutic effect at multiple relevant thresholds of efficacy
- Duration of platelets remaining ≥50x10^9/L ranged from 1 - 20 weeks with five patients above that platelet threshold for more than a month

Note: Increasing threshold analysis based exact logistic regression model with the baseline result as a factor

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

Mean platelet counts versus total WHO scale versus total IgGs



Pemphigus Vulgaris Phase 2 Interim Data

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

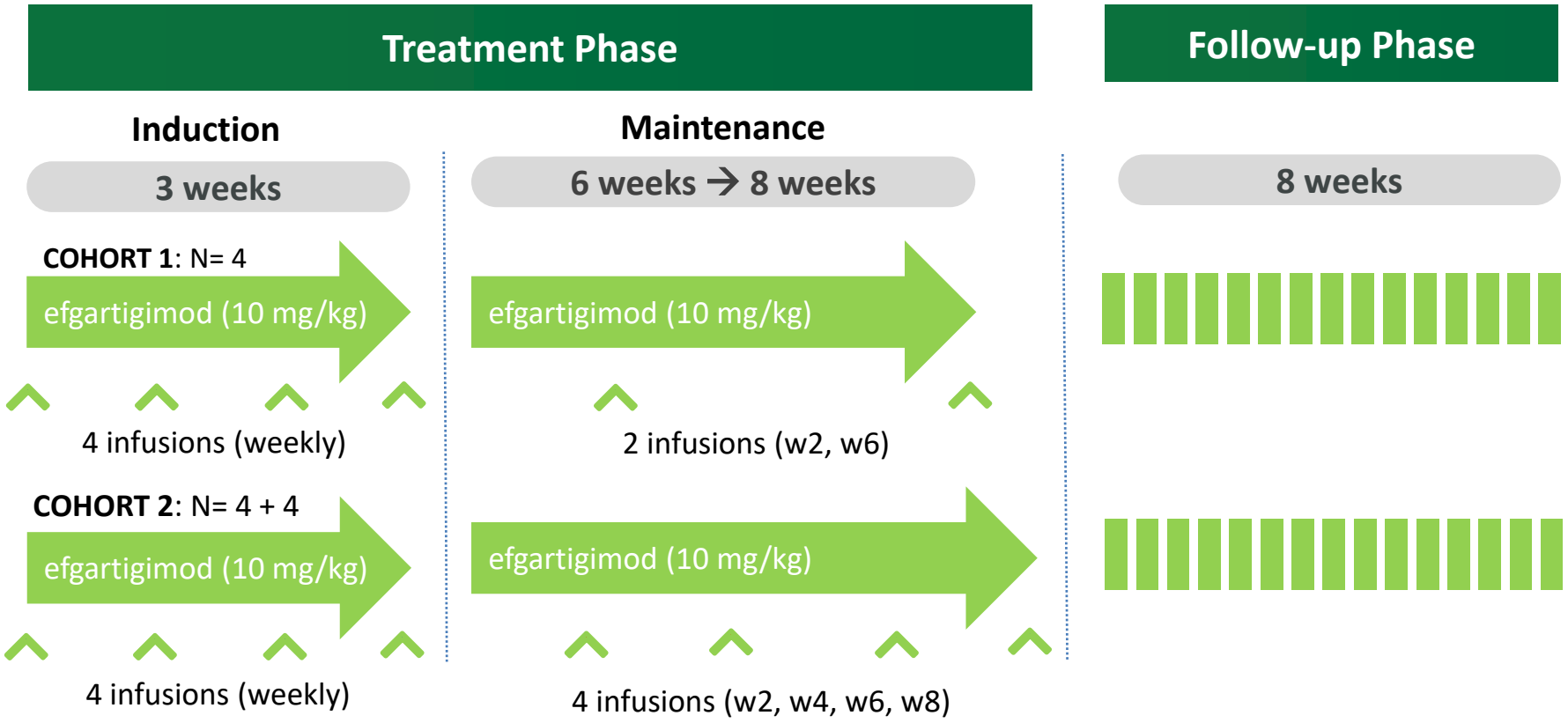
Patients with disease control:
- Mean max reduction in Pemphigus Disease Area Index (PDAI) score: 55%
- Mean max decrease in pathogenic IgGs: 57%

Favorable tolerability profile

No meaningful anti-drug antibody signals (ADA) reported

Pemphigus Vulgaris Phase 2 Adaptive Design

Third cohort to start in 1H 2019



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
Scleroderma
Lupus
Epidermolysis Bullosa Acquisita
Myasthenia Gravis
Multiple Sclerosis
Rheumatoid Arthritis
Anca Vasculitis
Pemphigus
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 ✓ → Neuromuscular Diseases

Immune Thrombocytopenia ✓ → Hematology Disorders

Pemphigus Vulgaris → Blistering Diseases

Therapeutic Area Beachheads with Expansion Possibilities into Adjacent Indications

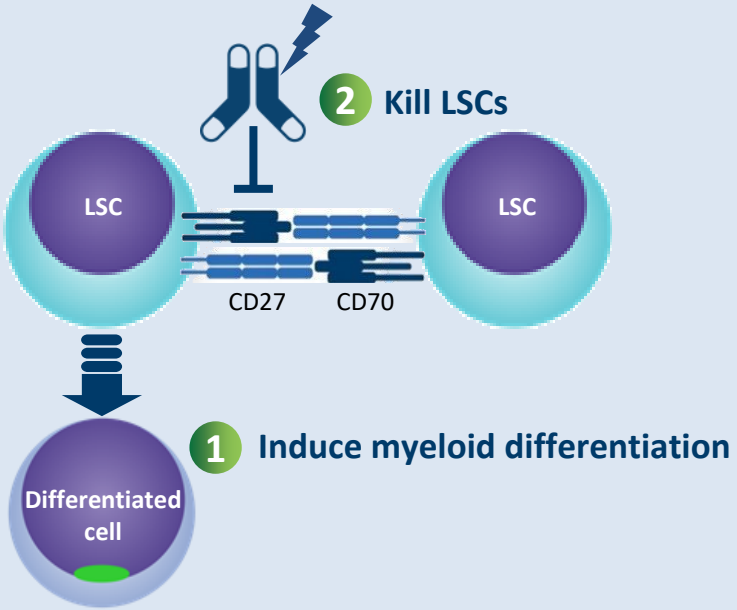
Chronic Inflammatory Demyelinating Polyneuropathy (CIDP)

Phase 2 CIDP study to start in 2H 2019

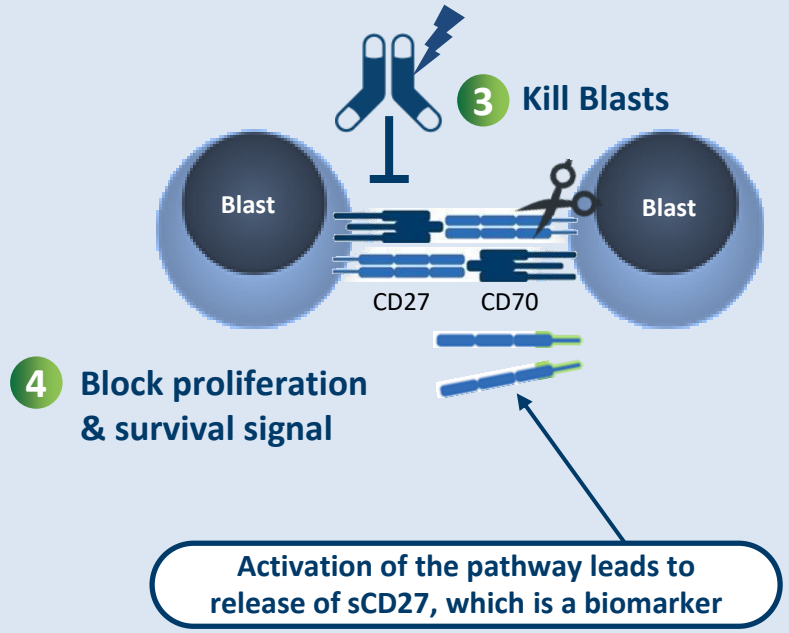
ARGX-110 (Cusatuzumab): Phase 1 / 2 Mono & Combo Therapy

Cusatuzumab Mode-of-Action Targets both Leukemic Stem Cells and Blasts

Cusatuzumab induces LSC differentiation



Cusatuzumab kills blasts

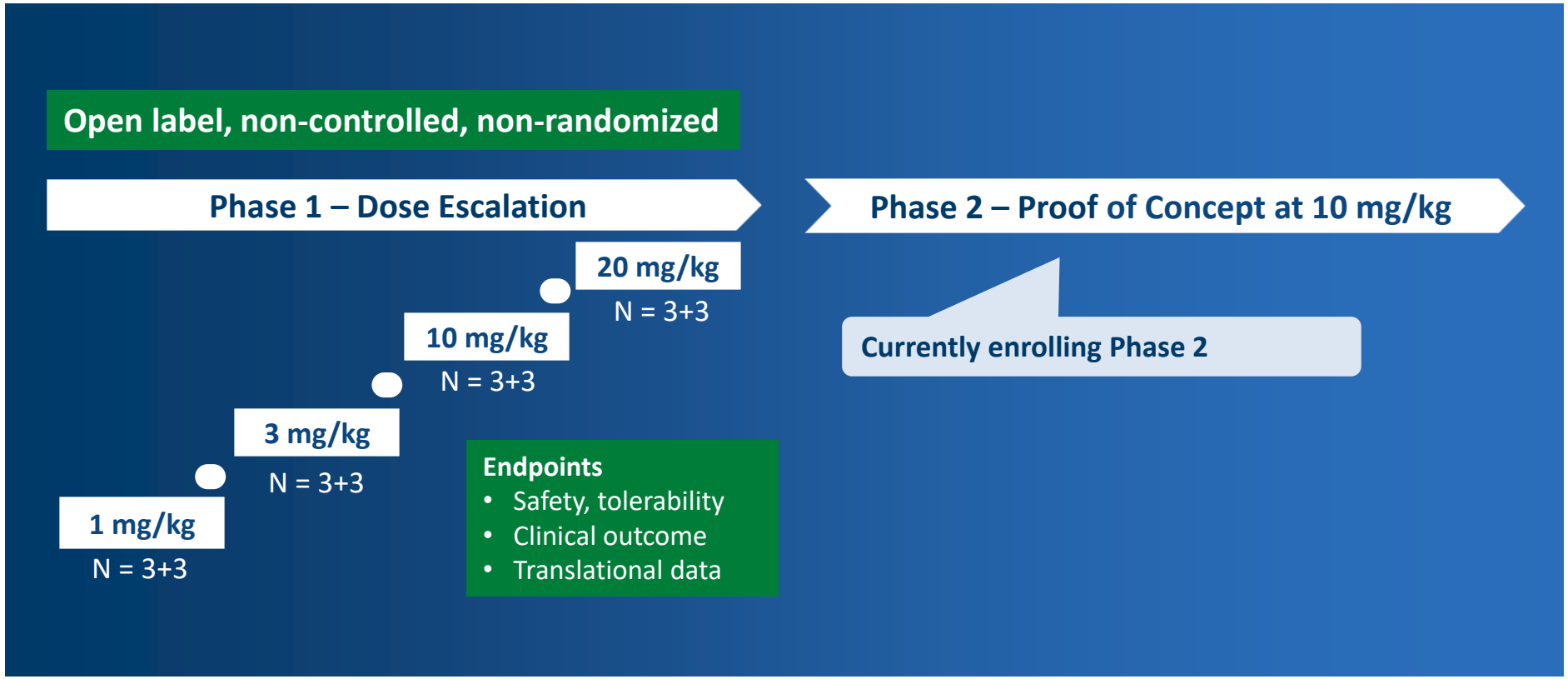


- Cusatuzumab is a potentially first-in-class anti-CD70 ADCC enhanced SIMPLE Antibody™ which selectively targets LSCs and blasts in AML and other heme indications

Silence et al. 2014, mAbs., Riether et al. 2017, J Exp Med.

Ongoing Phase 1/2 Combination Trial

Newly diagnosed AML patients unfit for intensive chemotherapy



- Efficacy seen across doses in Phase 1 dose escalation
- Up to 21 patients to enroll in initial Phase 2 study with potential to expand enrollment to 40
- 10 mg/kg selected for Phase 2 to saturate bone marrow and maintain clean tolerability profile

Overall Conclusions: Phase 1 Dose Escalation

Favorable tolerability profile

- No obvious toxicity on top of Vidaza toxicity
- No dose-limiting toxicity observed

Encouraging proof-of-biology data in 12 patients (4 dose cohorts; 3 pts each)

- 92% response rate (11/12) mainly CR/CRi
- 3 patients responded after cusatuzumab monotherapy
- Significant blast reduction in bone marrow after cusatuzumab monotherapy
- MRD negativity in 42% (5/12) treated patients

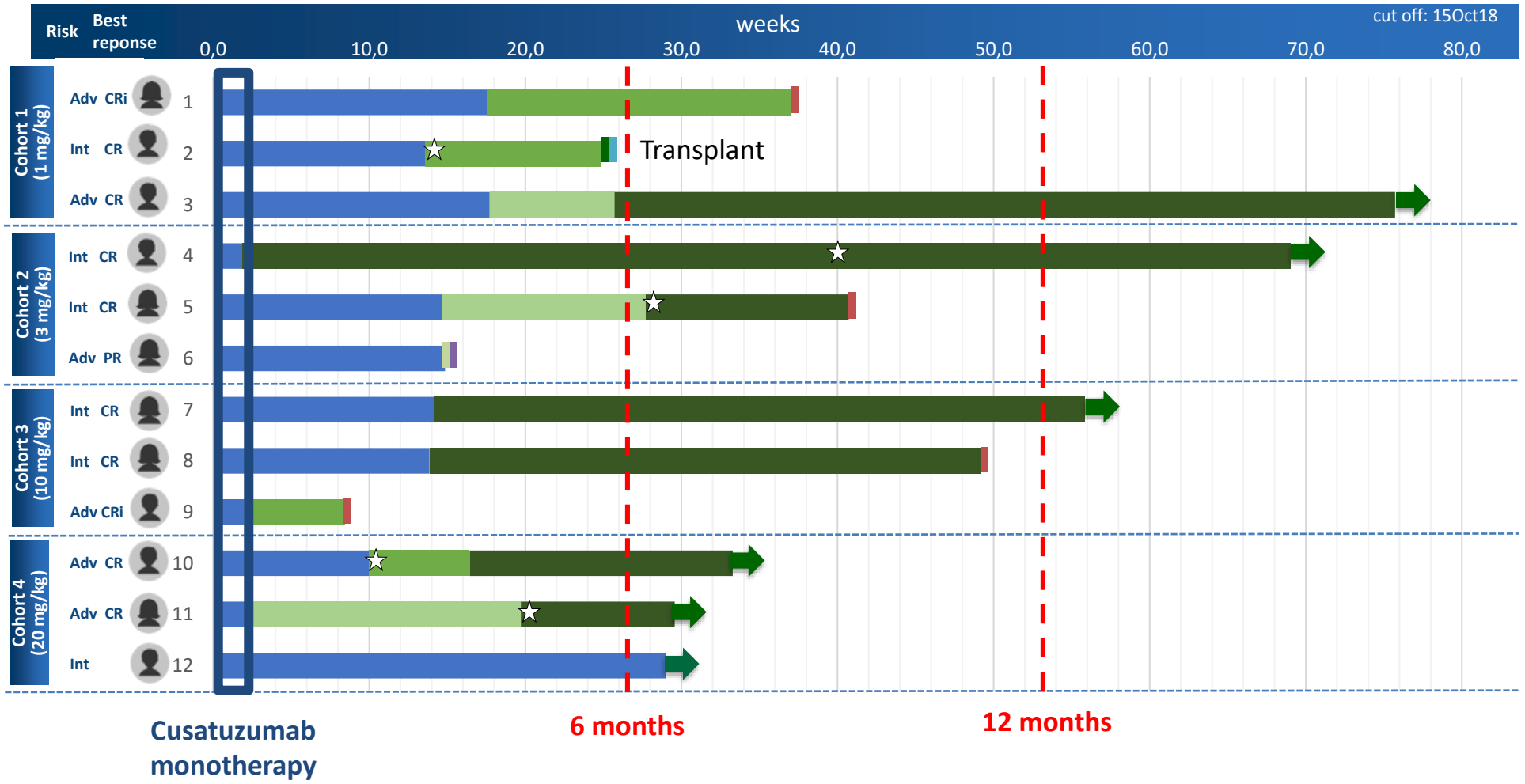
Supported by **translational dataset**

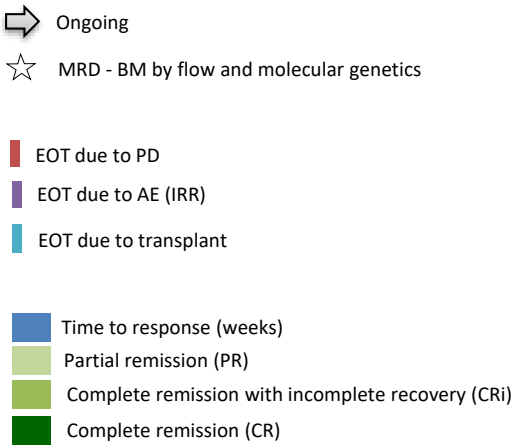
- Decreased sCD27 levels
- Reduced LSC colony formation
- Increased myeloid differentiation – asymmetric division

Recommended Phase 2 dose: 10 mg/kg

92% (11/12) Response Rate – CR/CRi/PR

Three patients on study for more than 12 months





Business Development & Financials

Cusatuzumab strategic alliance with Janssen Pharmaceuticals

argenx objectives	Janssen alliance
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

AbbVie Partnership for Novel Target GARP

Strategic Antibody Collaboration Details

- **GARP** is a protein specifically present on the surface of activated regulatory T-cells (Tregs)
- **AbbVie** exercised option in August 2018 to:
  - Obtain exclusive, worldwide license to develop and commercialize ARGX-115
  - Fund further GARP-related research by argenx beyond ARGX-115
- **argenx** can study ARGX-115 in combo with its pipeline programs

Financial Highlights

- **$60mm** received to date
- **$625mm in potential** development, regulatory and commercial **milestones**
- **Tiered royalties** on sales at percentages ranging from **mid-single digits to low teens**
- **Co-promotional** rights for ARGX-115-based products in the **European Economic Area and Switzerland**

Cuende et al., 2015, Science Trans Med.

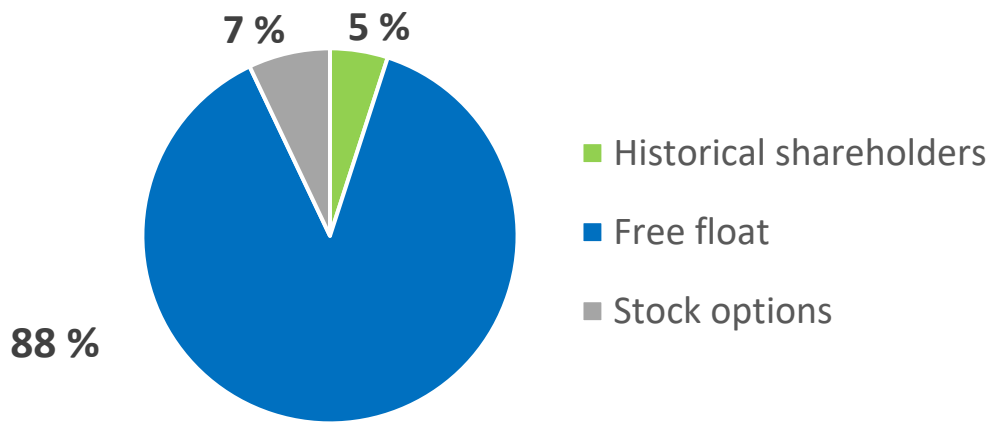
Financial Profile and Investor Composition

Shareholder base > 70% U.S. investors

Additional Key Statistics – Sept 30, 2018

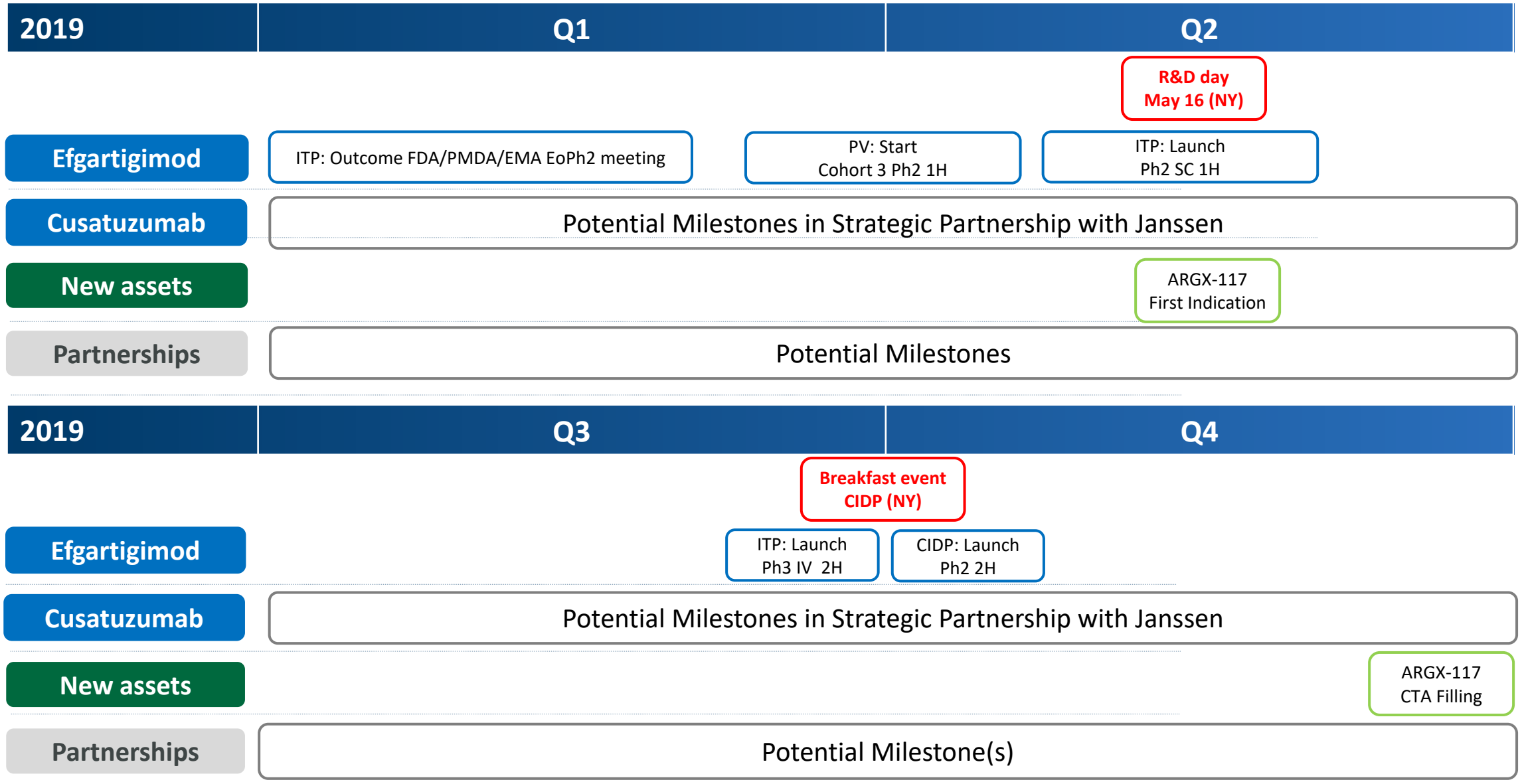
- **Cash position:** €582.3 mm (+ $500 mm Janssen deal, Dec 2019)
- **Capital raised since inception: €730 mm (ex. grants)**
  - 2017: raised $115 mm (€102 mm) in Nasdaq IPO
  - 2017: raised $266 mm (€226 mm) in public offering
  - 2018: raised $300 mm (€256 mm) in public offering
- **Non-dilutive funding since inception: €104mm (incl. grants)**
  - 2018: $10mm second preclinical milestone AbbVie
- **120 employees & consultants —89 R&D, 31 SG&A**

Blue-Chip Investor Base – Sept 30, 2018



- **U.S. shareholding** expanded **above 70%**
- **Outstanding shares: 35,934,457**

Key Upcoming Expected Milestones & Communications



Thank you!