2018 Full Year Results

argenx
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February 28, 2019

Forward-Looking Statements

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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

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	Preclinical	Phase 1	Phase 2	Phase 3	BLA	Next Milestone / Commentary
Wholly-Owned & Co-Development Product Candidates								
ARGX-113 Efgartigimod	FcRn	Myasthenia Gravis				adapt		3Q18: Phase 3 initiated
		Immune Thrombocytopenia						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			janssen			$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						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

AbbVie exercised option to develop and commercialize in August 2018

- **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

adapt
myasthenia gravis study

Primary endpoint

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

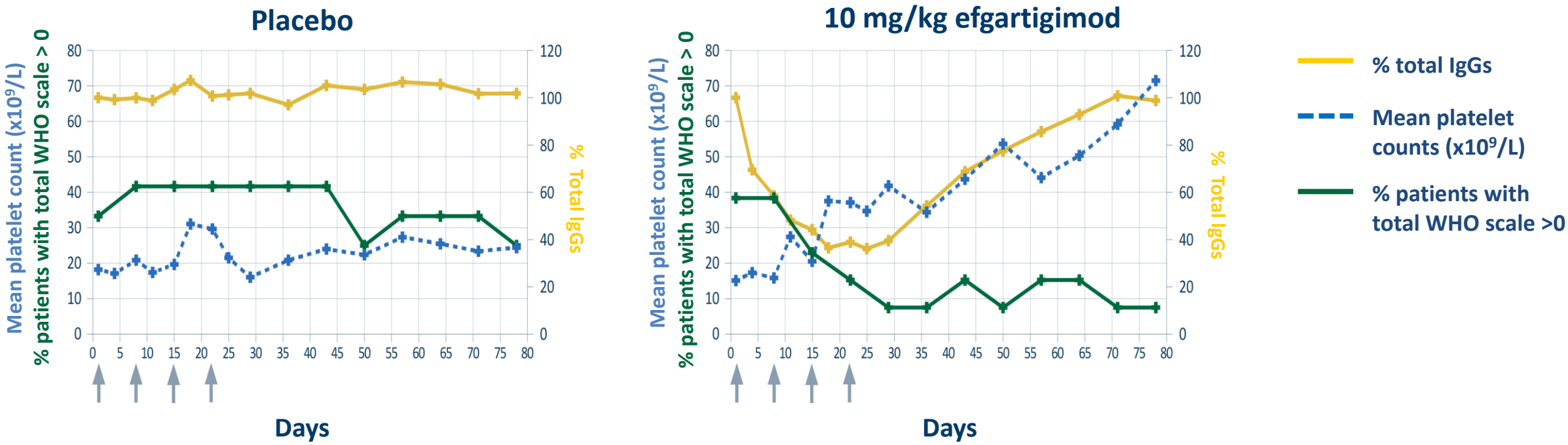
Secondary endpoints

Efficacy, Safety, Tolerability, Quality of Life and Impact on Normal Daily Activities Measures

Immune Thrombocytopenia Ph2 Clinical Trial

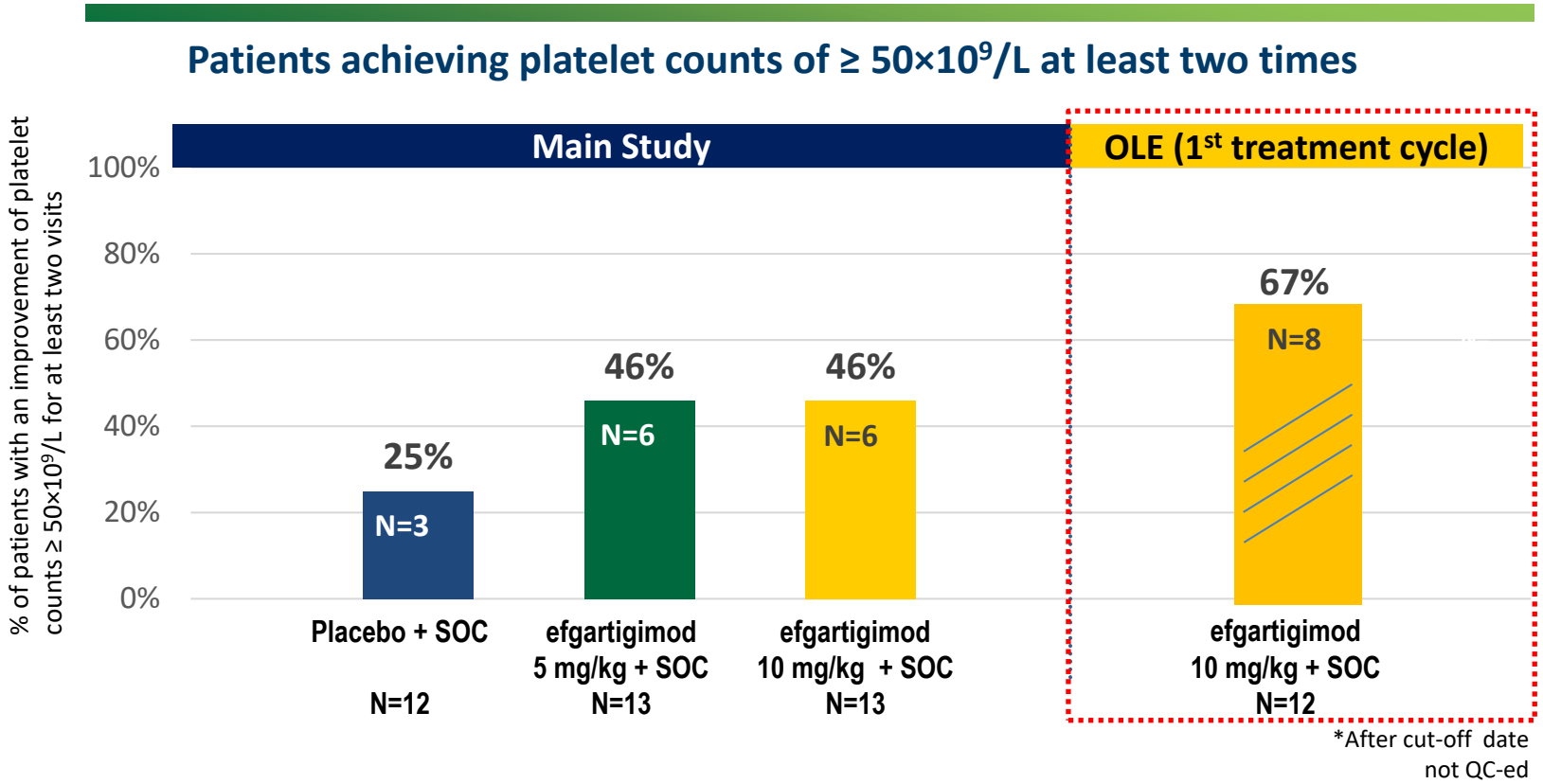
Reduction of Total IgGs Correlates with Increased Platelet Counts and Reduced Bleeding Events

Mean platelet counts versus total WHO scale versus total IgGs



Strong Improvement of Platelet Counts Across Doses

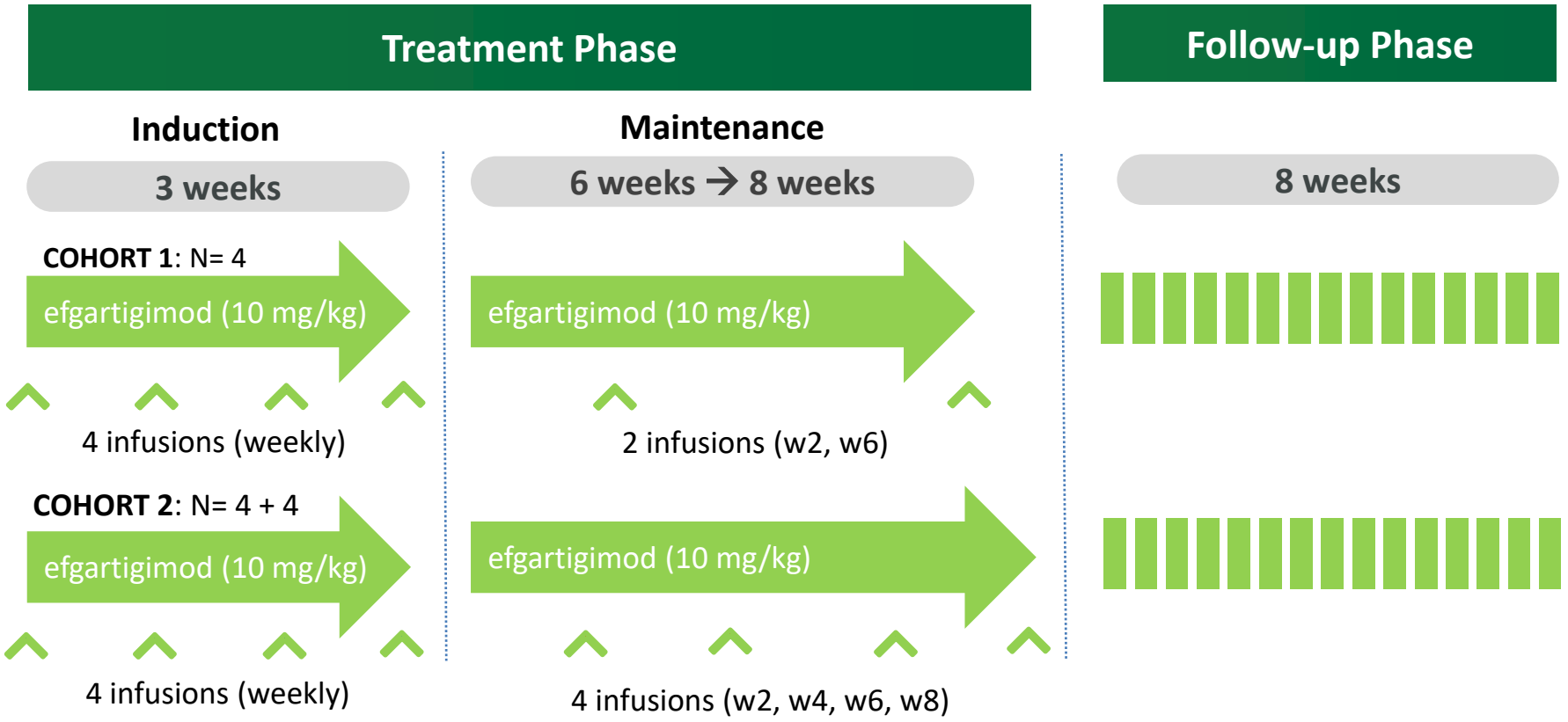
46-67% of patients exceeded platelet counts ≥ 50X10^9/L during at least two visits



- 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

Pemphigus Vulgaris Phase 2 Adaptive Design

Cohort 3 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

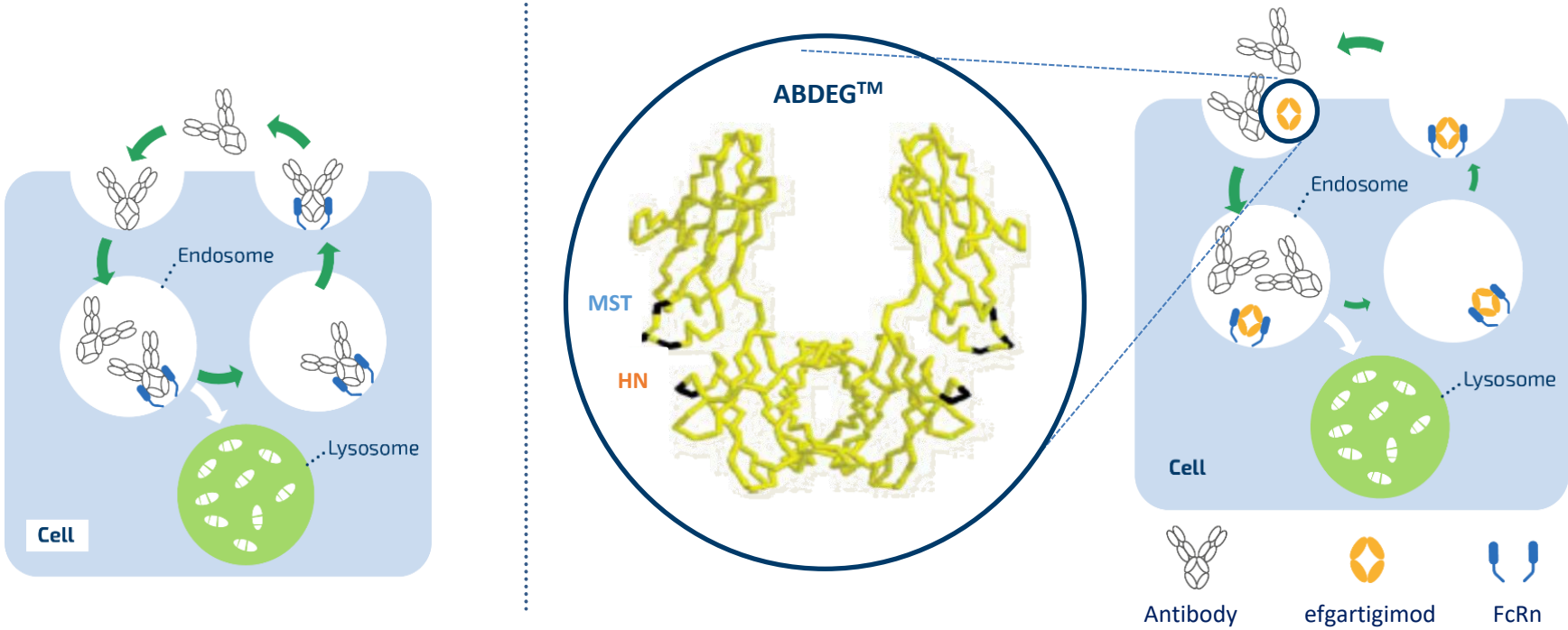
Efgartigimod: Human IgG1 Fc Fragment with ABDEG™ Mutations

Exploits Natural Fc/FcRn Interaction and retains pH dependent binding

IgG antibodies recycle through FcRn(1)…

efgartigimod potently blocks FcRn…

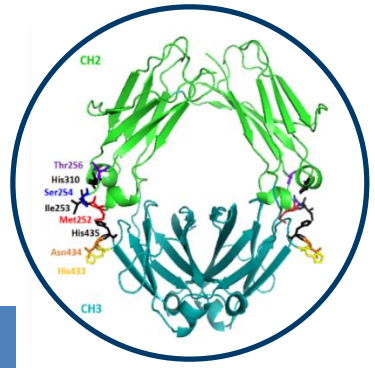
leading to IgG elimination(2)



(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

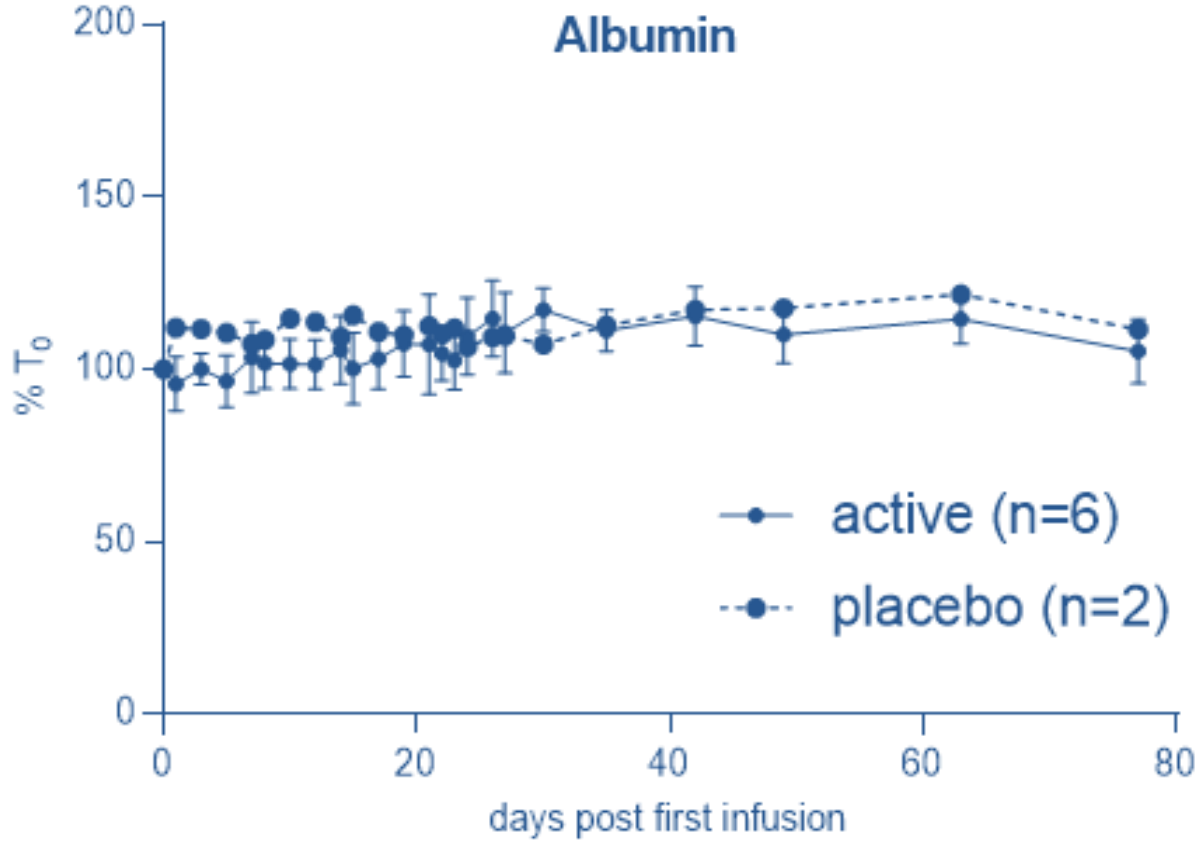
- **Reduced FcγR, C1q binding**
- **Endosomal recycling FcRn-efgart complex; no lysosomal degradation**
- **Can rebind FcRn**
- **1/3 size of IgG; excellent physicochemical stability**

Best-in-class clinical attributes

- **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

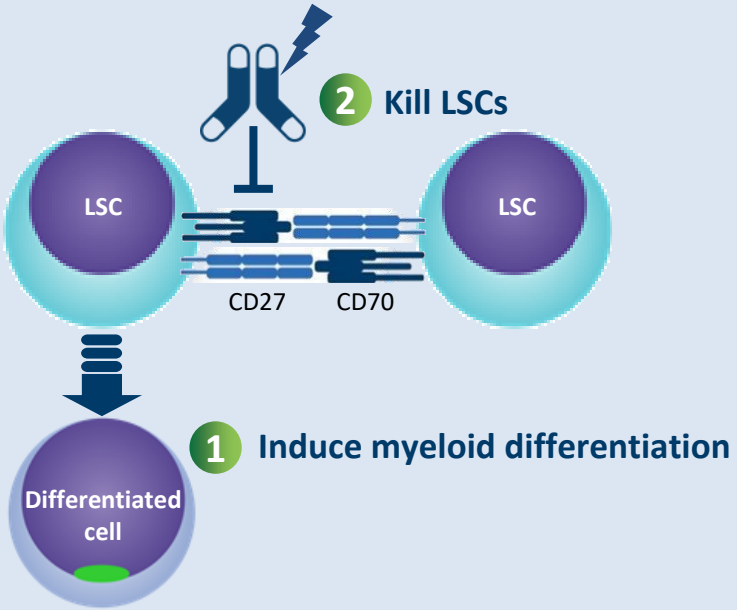
Efgartigimod (25 mg/kg q7d):
No significant lowering of albumin



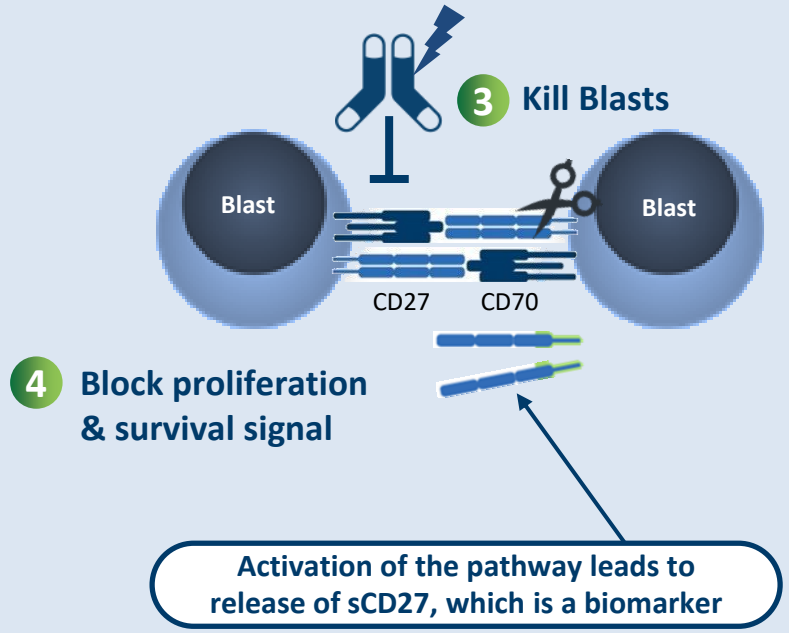
Ulrichts et al., JCI, 2018

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

Cusatuzumab induces LSC differentiation



Cusatuzumab kills blasts

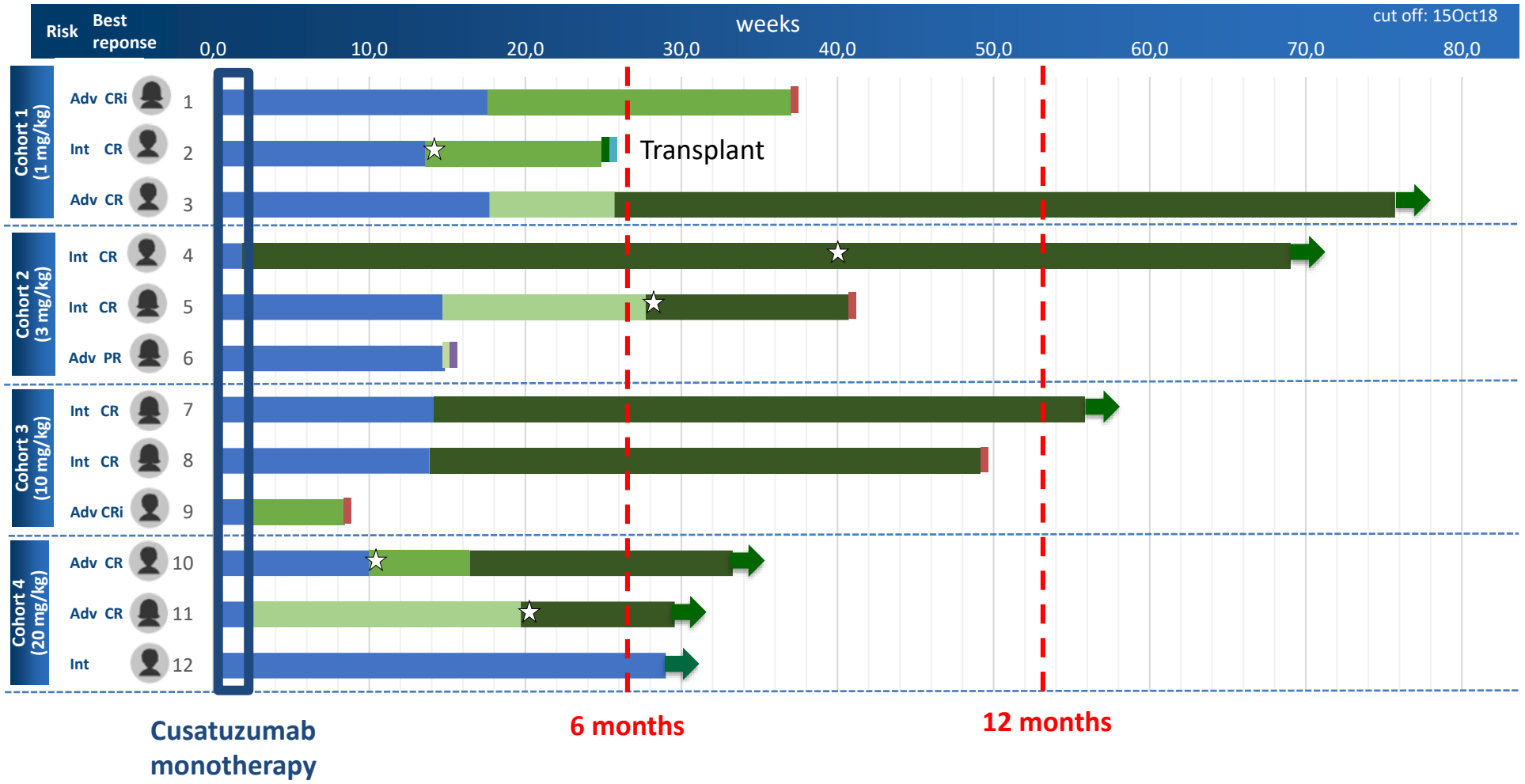


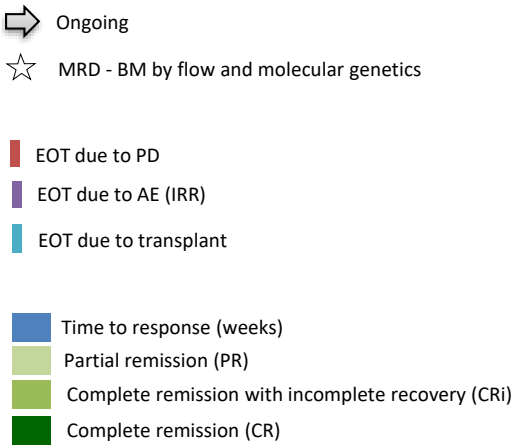
- Cusatuzumab is a potentially first-in-class anti-CD70 ADCC enhanced SIMPLE Antibody™ which selectively targets LSCs and blasts in AML and other heme indications

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

92% (11/12) Response Rate – CR/CRi/PR

Three patients on study for more than 12 months





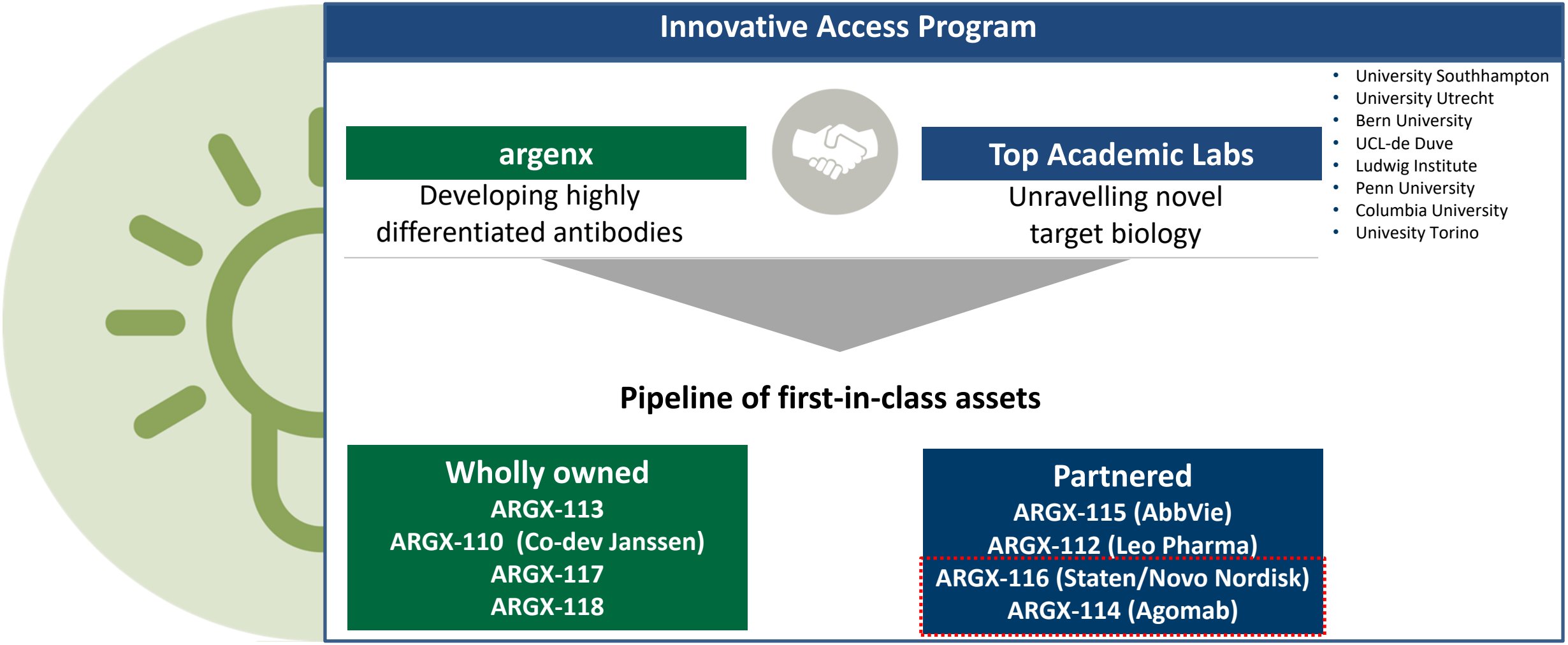
Cusatuzumab strategic alliance with Janssen Pharmaceuticals

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

Innovative Access Program

Success formula with proven track record



argenx Y2018 Financials

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

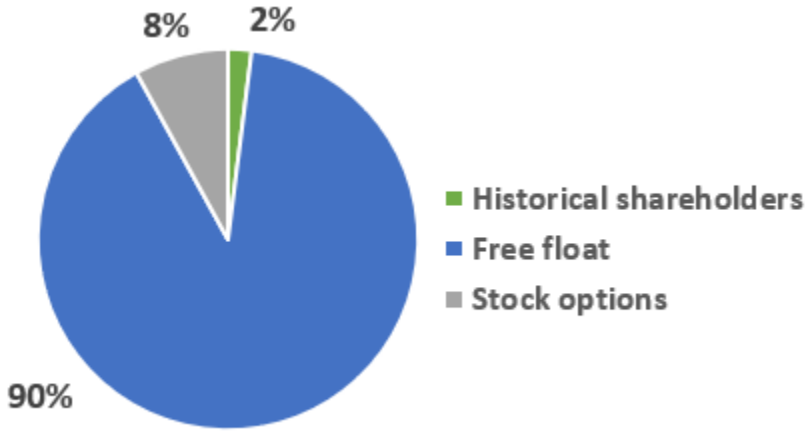
Financial Profile and Investor Composition

Shareholder base > 70% U.S. investors

Additional Key Statistics – Dec 31, 2018

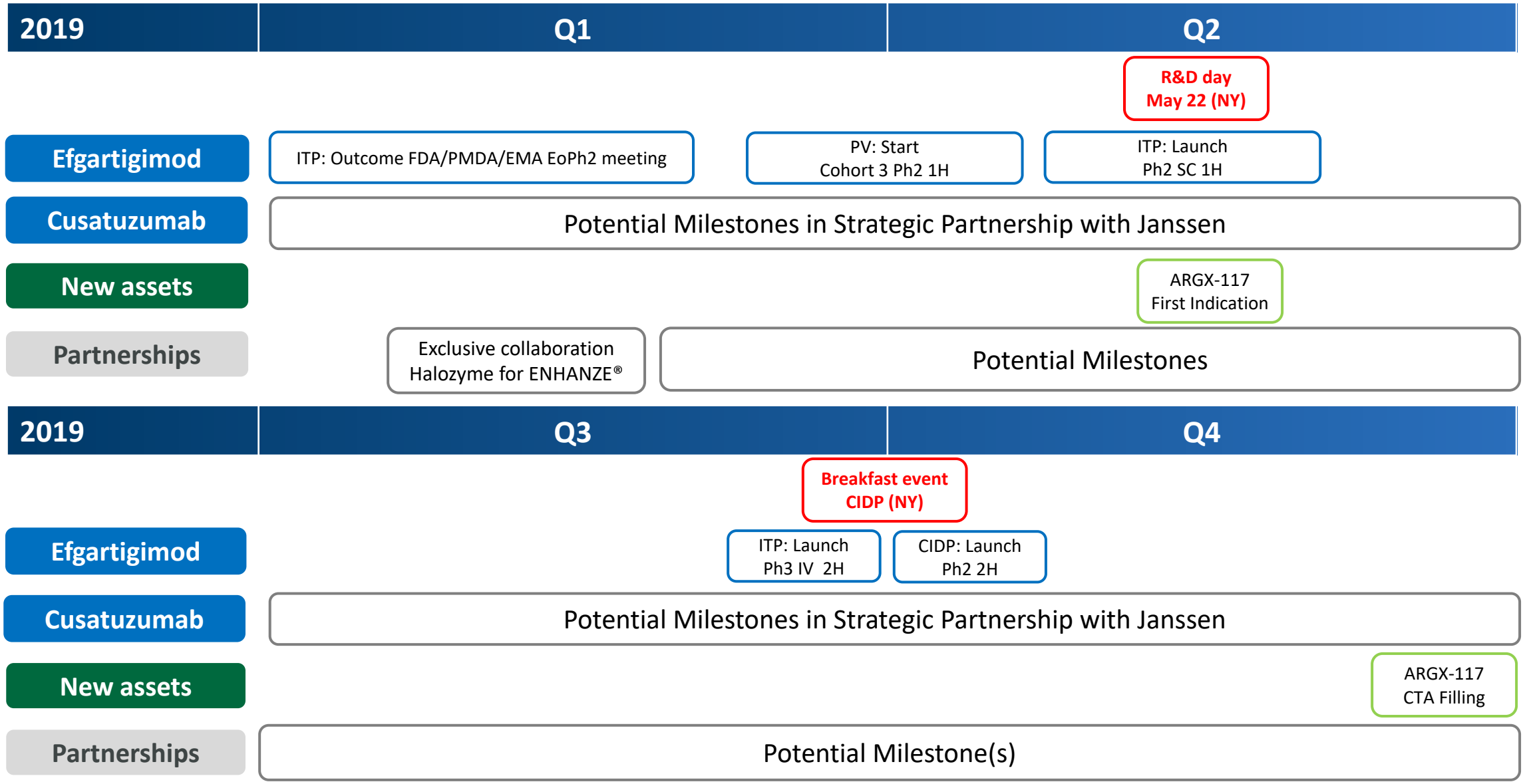
- **Cash position: €564.6 mm** (+ $500 mm Janssen deal)
- **Capital raised since inception: €717 mm (ex. grants)**
  - 2017: raised $115 mm (€102 mm) in Nasdaq IPO
  - 2017: raised $266 mm (€226 mm) in public offering
  - 2018: raised $300 mm (€256 mm) in public offering
- **Non-dilutive funding since inception: €107mm (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



Thank you!