BofA Securities Health Care Conference 2020

Corporate Presentation

May 2020

Forward-Looking Statements

This presentation has been prepared by argenx se ("argenx" or the "company") for informational purposes only and not for any other purpose. Nothing contained in this presentation is, or should be construed as, a recommendation, promise or representation by the presenter or the company or any director, employee, agent, or adviser of the company. This presentation does not purport to be all-inclusive or to contain all of the information you may desire. This presentation also contains estimates and other statistical data made by independent parties and by us relating to market size and growth and other data about our industry. This data involves a number of assumptions and limitations, and you are cautioned not to give undue weight to such estimates.

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, our ability to obtain regulatory approval of our product candidates, the expected size of the markets for our prouct candidates, 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 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.

Navigating The Impact Of COVID-19

Our People

Ensuring alignment across our team

Supporting each other in new environment

Our Patients

Adapting for our patients

Implementing opportunities for home infusions and telehealth visits

Our Business

Focused on continuity

ADAPT readout on track for mid-2020 and BLA filing by end of year

Late-stage Biotech Building Towards Commercial Success

argenx 2021: Reaching patients	Therapeutic franchises
	Global expansion
Late-stage pipeline	FcRn leadership MG CIDP ITP PV
	Cusatuzumab strategic alliance
Immunology breakthroughs	Two new pipeline assets from IIP
Strong balance sheet	1.3B€ in cash at end of 2019

Growing Franchises With Multiple Late-Stage Programs

Neuro-muscular

- MG — adapt (myasthenia gravis study)
- CIDP — adhere (chronic inflammatory demyelinating polyneuropathy study)

ARGX-117

Hem/Onc

- ITP — advance (immune thrombocytopenia study); advance SC (immune thrombocytopenia study); advance 2 (immune thrombocytopenia study)
- AML
- MDS

Janssen

Skin / Kidney

- PV

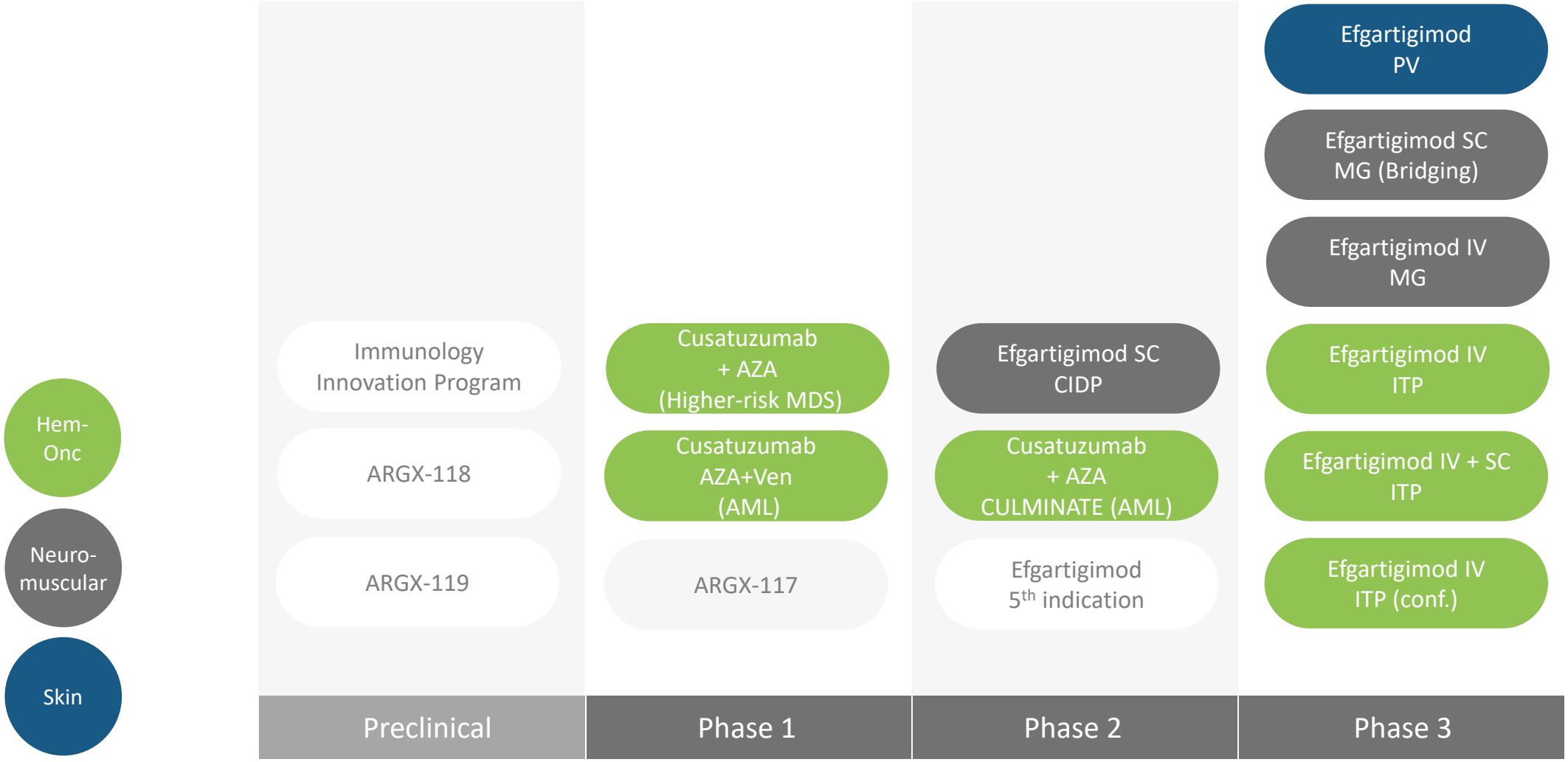
Advancing to Phase 3 on positive Phase 2 data

- Kidney

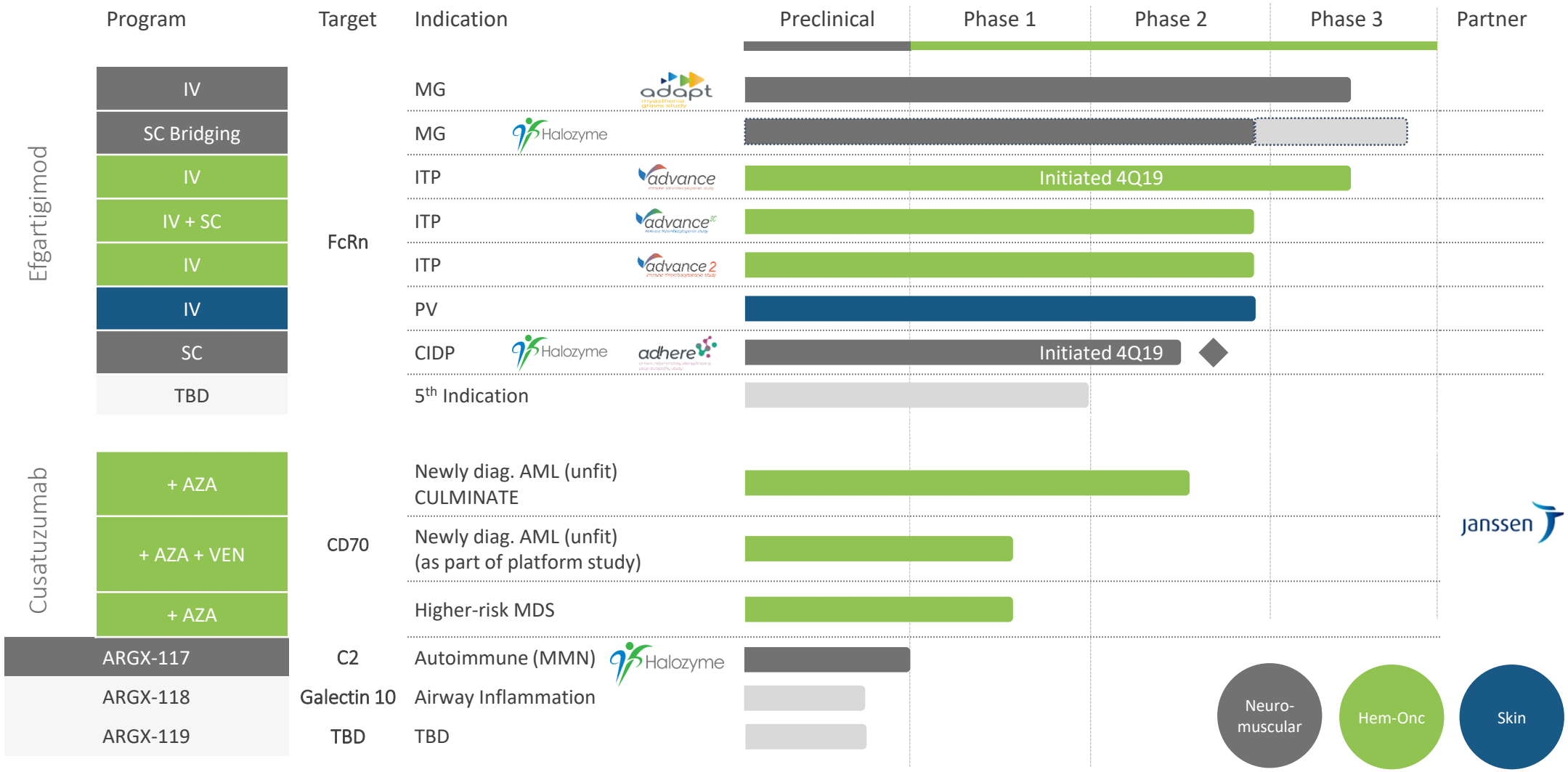
Severe autoimmune conditions

GOAL OF 5 LAUNCHES IN 5 YEARS

2020 View Of Pipeline: Poised To Have Five Phase 3 Trials Underway



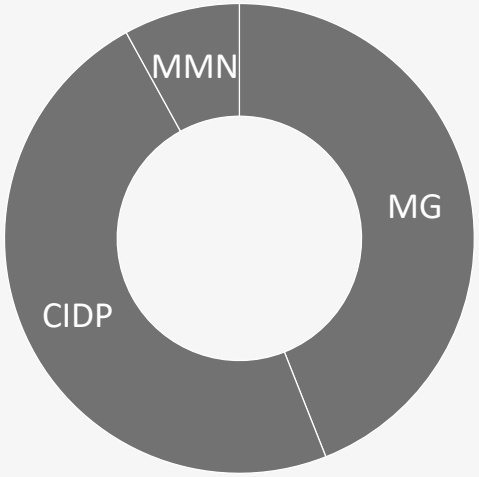
Deep Antibody Pipeline Of Differentiated Candidates



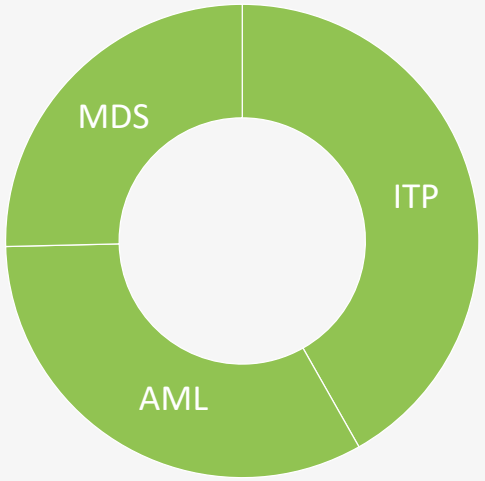
MG: Myasthenia Gravis **ITP:** Immune Thrombocytopenia **PV:** Pemphigus Vulgaris **CIDP:** Chronic Inflammatory Demyelinating Polyneuropathy **AML**: Acute Myeloid Leukemia **MDS:** Myelodysplastic Syndromes

Franchises Sit In High-Value Rapid-Growth Markets

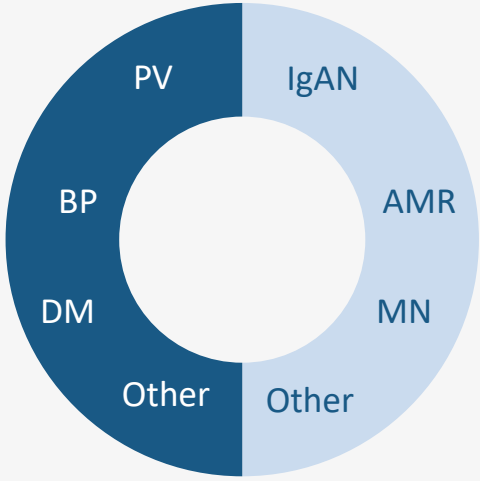
Neuromuscular
>$5B (CAGR ~10%+)



Hem/Onc
~$7B (CAGR ~10%)



Skin / Kidney



~370,000 patients

> 600,000 patients

argenx estimates based on proprietary market research

IgAN: IgA nephropathy
BP: Bullous pemphigoid
AMR: Antibody-mediated rejection
DM: Dermatomyositis
MN: Membranous nephropathy

The Right Team In Place To Launch Efgartigimod

argenx

COO leading commercial organization	Commercial leaders hired across all key functions	Field-based medical research liaisons in place	Stepwise salesforce ramp-up

Significant product launch experience

SOLIRIS® (eculizumab) Injection for Intravenous Use
MOZOBIL (plerixafor injection)
Kanuma™ sebelipase alfa
DACOGEN decitabine for injection
Vyvanse
Xembify® (immune globulin subcutaneous human-klhw) 20%
Tegsedi® (inotersen) injection 284 mg/1.5 mL
ALUNBRIG™ BRIGATINIB 30mg TABLETS
ORENCIA® (abatacept) Injection for Intravenous Use Injection for Subcutaneous Use
KEVEYIS® dichlorphenamide 50 mg tablets
CELEBREX®
actos® pioglitazon HCl
Sensipar® (cinacalcet) Tablets
Strensiq® (asfotase alfa) for injection
VPRIV® velaglucerase alfa for injection
Skyrizi™ risankizumab-rzaa 75mg/0.83mL Injection
ALPROLIX™ [Coagulation Factor IX (Recombinant), Fc Fusion Protein]
Enbrel® etanercept
ICLUSIG® (ponatinib) tablets 45 mg, 15 mg
Aranesp® (darbepoetin alfa)
SPIRIVA® (tiotropium)
Gattex® (teduglutide) for injection
TYSABRI®
gamunex-C® immune globulin injection (human), 10% caprylate/chromatography purified
ULTOMIRIS™ (ravulizumab-cwvz) injection for intravenous use
Amevive® (alefacept)
HUMIRA® adalimumab
Halaven® (eribulin mesylate) Injection 0.5 mg/mL
Vectibix®
Uloric® (febuxostat) 40 mg | 80 mg tablets
Zantac®
NUPLAZID™ (pimavanserin) tablets
Cablivi® caplacizumab
GOCOVRI® (amantadine) extended release capsules
Tecfidera® (dimethyl fumarate)

Preparing for global launch

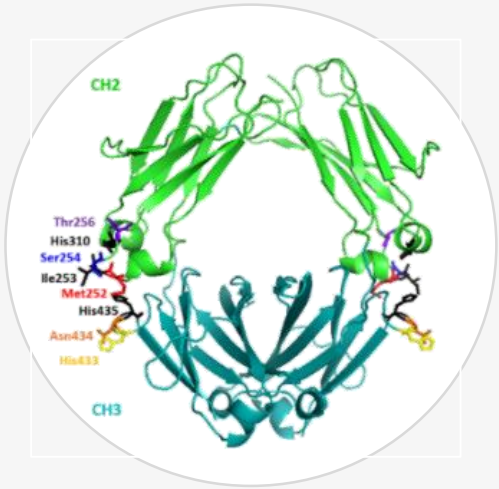
USA

JAPAN

EU

Building Innovation At Every Step

Preclinical / Discovery



Molecule Design:
Immunology Innovation Program

Clinical

adapt
myasthenia gravis study

Clinical Development:
Thoughtful ADAPT Design

Commercial

MyReal World™ MG

Commercial Approach:
Real-World Evidence Study

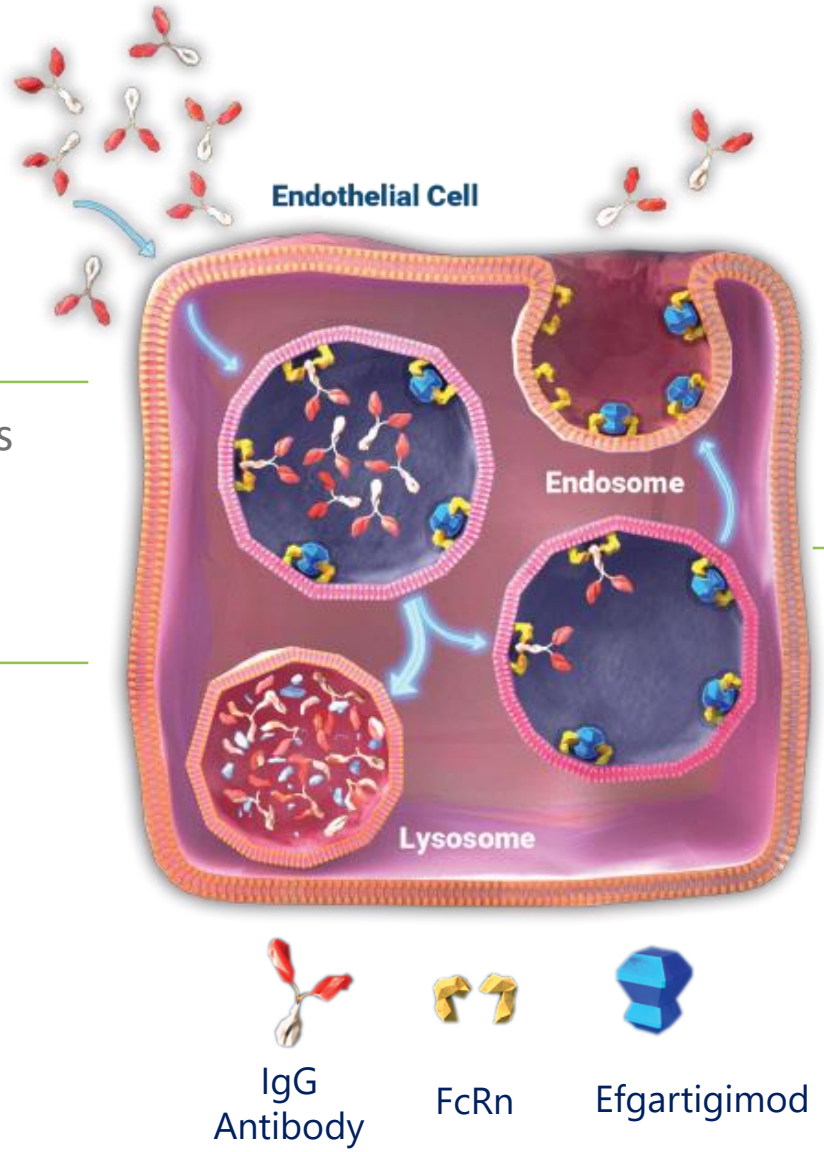
Efgartigimod: Unique Molecule Design Leads To Differentiated Profile In Phase 2

Efficacy

3/3 beachhead indications

Safety

No class effect



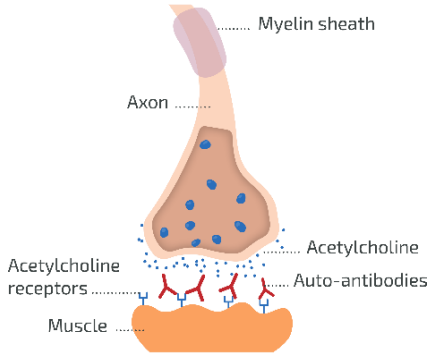
Convenience

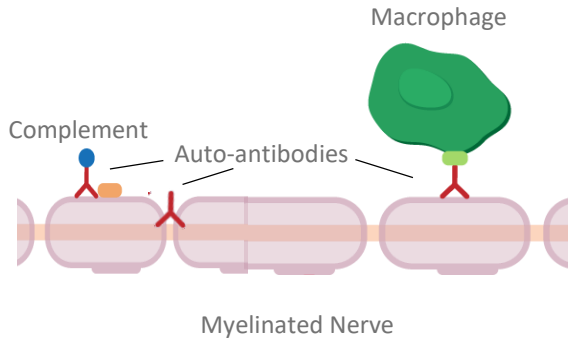
Potential optionality for patients

Beachhead Strategy Based On Unifying Biologic Rationale

Neuromuscular

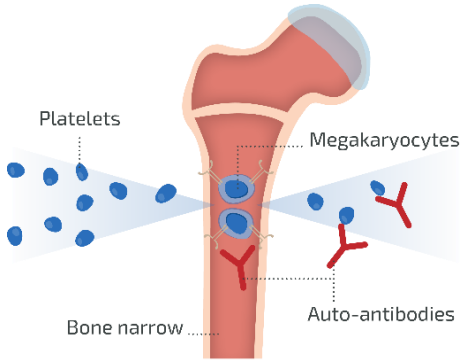
Myasthenia Gravis → Chronic Inflammatory Demyelinating Polyneuropathy





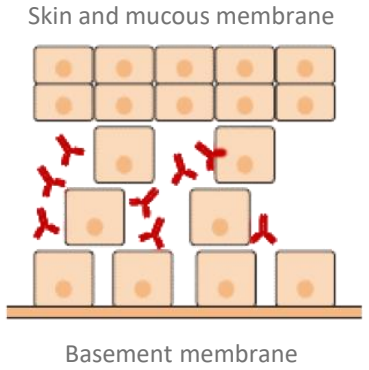
Hem/Onc

Immune Thrombocytopenia



Skin

Pemphigus Vulgaris



Potential Therapeutic Utility in Diseases Mediated by Pathogenic IgGs

- Block acetylcholine receptors
- Cross-link + internalize acetylcholine receptors
- Recruit complement

- Block nerve conduction
- Recruit macrophages
- Activate complement

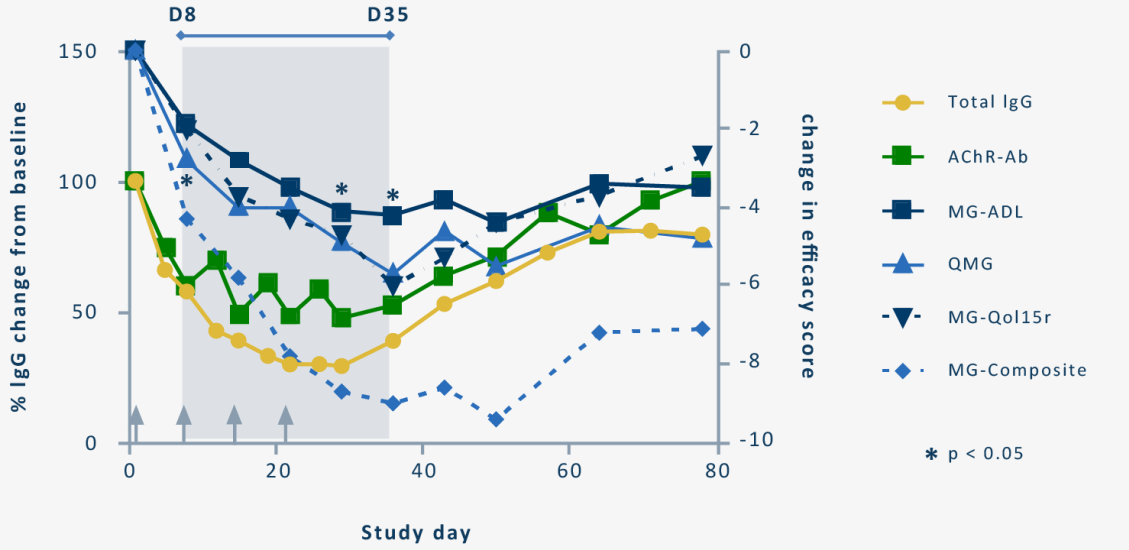
- Enhance platelet clearance
- Kill platelets
- Inhibit platelet production
- Reduce platelet function

- Acantholysis
- Steric hindrance
- Deplete desmoglein

MG and ITP Phase 2 Data Support Registrational Plan

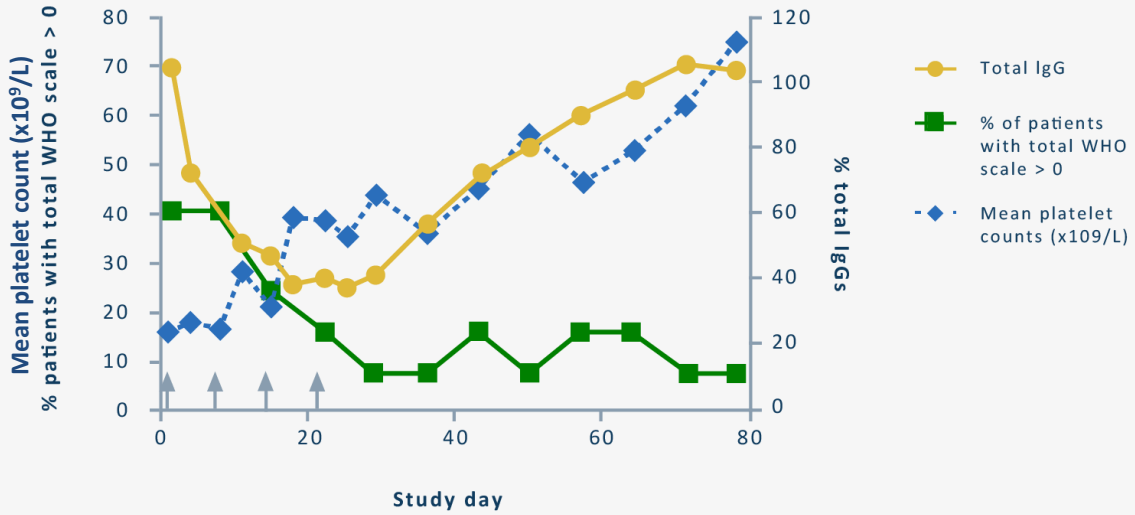
MG | 10 mg/kg efgartigimod

Neurology®



ITP | 10 mg/kg efgartigimod

American Journal of Hematology



- Reduction of total and pathogenic IgGs led to clinically meaningful improvements in disease scores (MG-ADL, QMG, QoL and Composite for MG; platelet count and bleeding events for ITP)
- Favorable tolerability profile with adverse events balanced between active and placebo arms

PV Phase 2 Data Support Registrational Plan

Baseline Characteristics

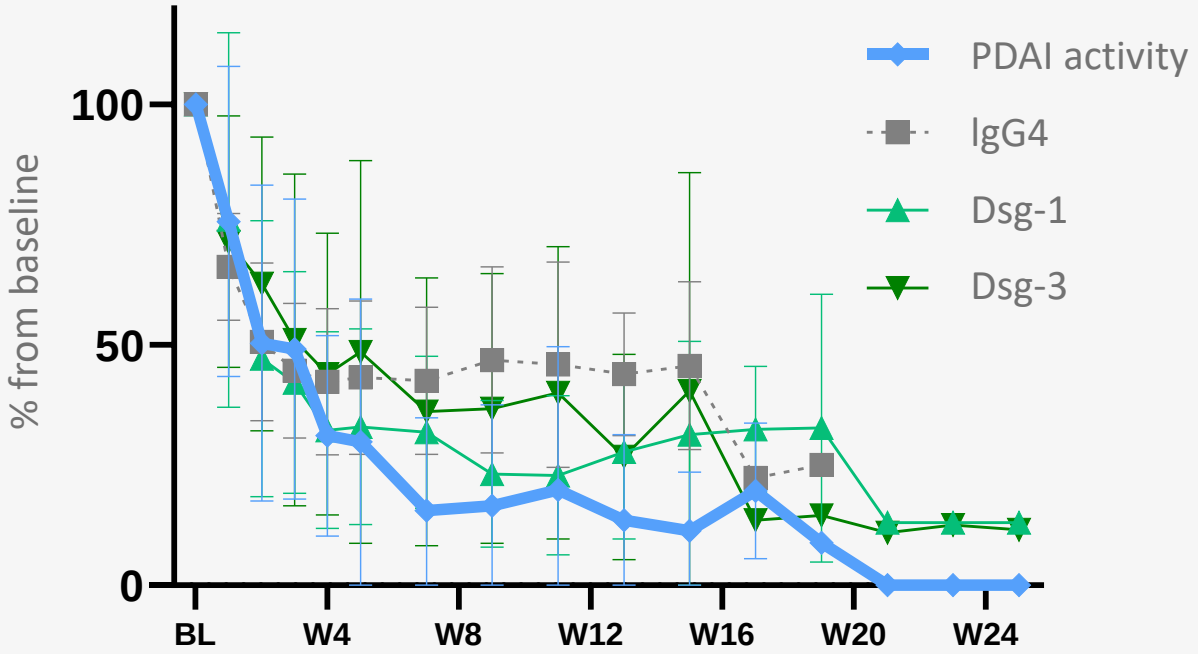
Pemphigus Vulgaris subtype
Mucosal-dominant (N = 8)
Mucocutaneous (N = 10)
Cutaneous (N = 1)
Pemphigus Foliaceus (N = 4)

Severity
Mild: PDAI < 15 (N = 9)
Moderate: PDAI 15-44 (N = 14)

Disease history
Newly diagnosed (N = 9)
Relapsing (N = 14)

23*

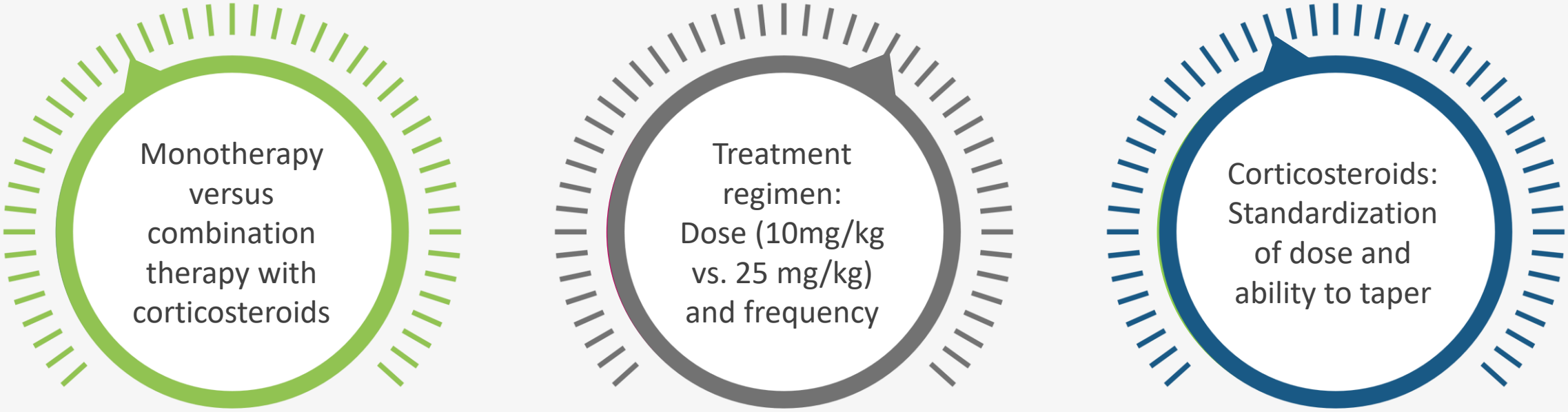
IgG Reduction Correlated to PDAI Score Improvement in Responders



Weekly or biweekly maintenance dosing at variable frequency

* Eligible for efficacy analysis
Data cut off 7 Nov 2019; 8 patients on study at time of interim analysis
Data show efgartigimod treatment phases with at least biweekly dosing ; excludes IgG4 for one patient (outlier)

Adaptive Phase 2 Trial Of Efgartigimod In PV



Synergy With Low-Dose Steroids In PV Phase 2

Fast onset of action	**78% disease control** (18/23 patients) – majority **after 1-2 infusions** Median time to DC: 14 to 15 days (mono/combo therapy)
Deep responses	**70% complete clinical remission** (5/7 patients) on optimized dosing* Time to CR: **2-10 weeks**
	Mean maximum PDAI improvement in responders >60% to >85% (mono/combo therapy)
	Strong steroid sparing potential demonstrated
Favorable tolerability	Determined by independent monitoring committee
Potential synergy	Efgartigimod clears a-Dsg antibodies/Steroids stimulate Dsg synthesis

* At least biweekly efgartigimod + corticosteroids @ 0.25-0.5mg/kg

ADAPT Trial: Built For MG Patients Based On Strengths Of Efgartigimod

We listened to stakeholders…

…and built on observed attributes of efgartigimod

Request to be tailored, convenient, cost-effective

Phase 2 MG data:

Fast onset of action

- Responded within first four weeks

Clinical response in 83% of patients

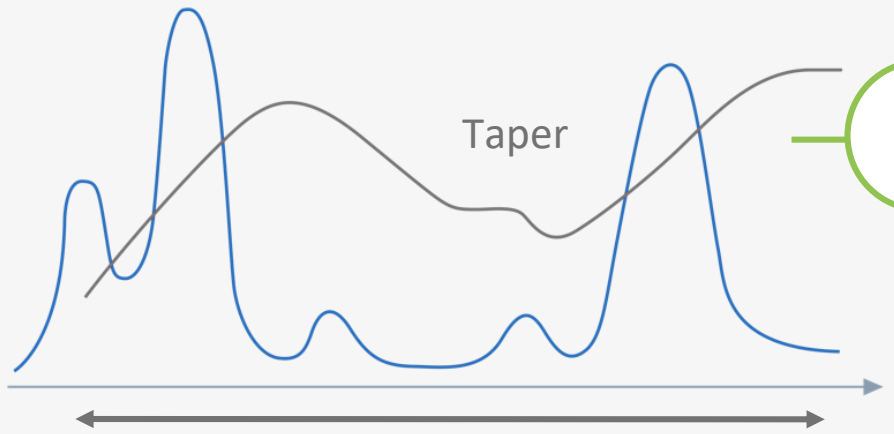
Durable response in 75% of patients

- Sustained for at least 6 weeks

Promising tolerability

Potential To Offer Tailored Treatment Approach In MG

Current Standard of Care

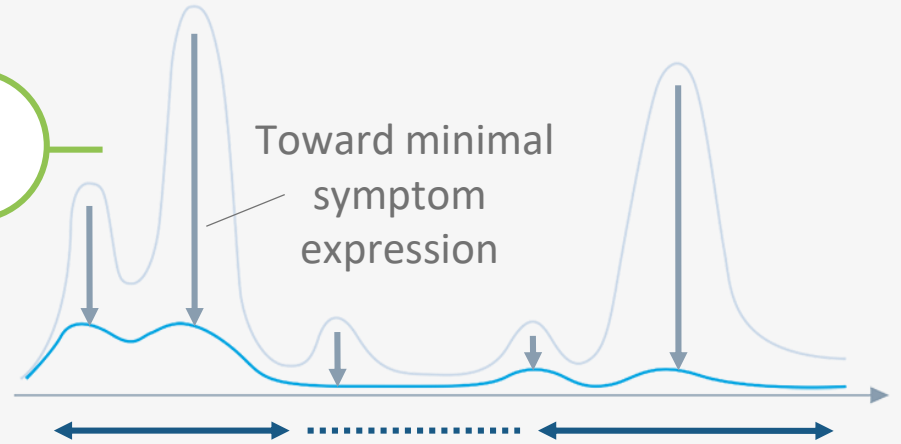


Chronic dosing

- Fast-acting steroids and slow-acting immunosuppressants
- Balancing symptom suppression and side effects

Typical MG Symptoms

Goal of Efgartigimod Dosing



Patient-tailored dosing

- Tailored regimen matches variability of MG
- Time between cycles is individualized
- Period of sustained therapeutic benefit between cycles can offer flexibility

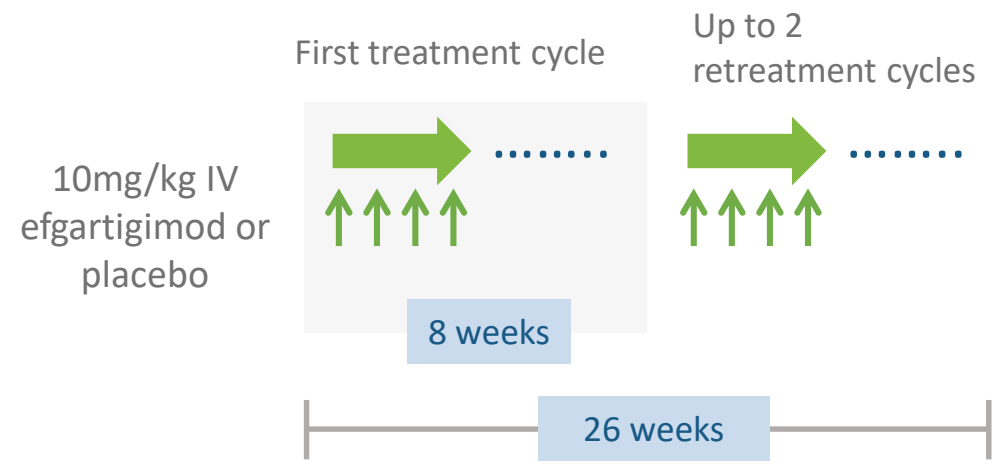
Innovative ADAPT Trial Designed To Meet Clinical Practice

Patient population consistent with Phase 2

gMG patients (MG-ADL≥5)

Stratified for AChR+ or AChR- and background therapy (n=167 total)

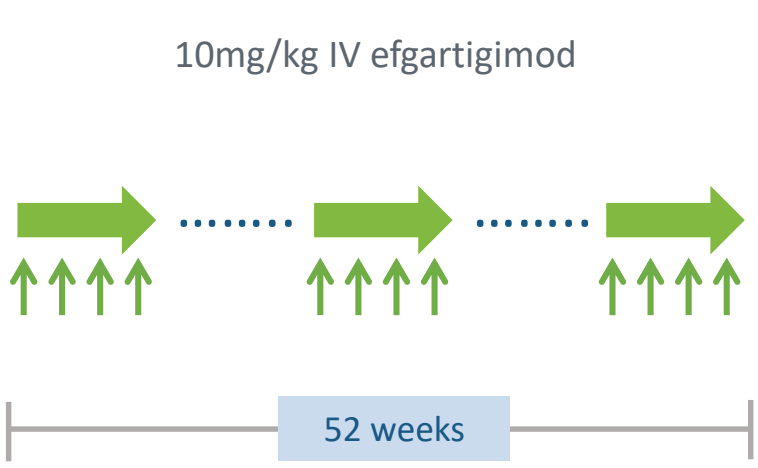
Primary endpoint readout at week 8
Duration of benefit measured over 26 weeks



Individualized treatment cycles

Time between cycles determined by duration of sustained treatment benefit

Open-label Extension
Retreat as needed to simulate clinical practice

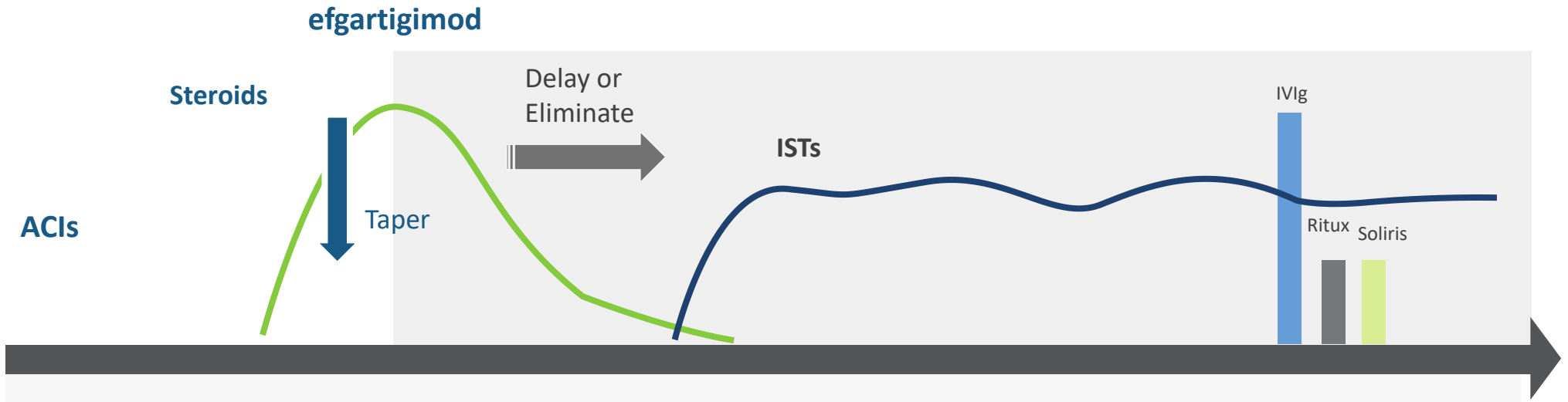


Primary endpoint (AChR+): % responders after first treatment cycle

Responder: ≥2 ADL points for at least 4 consecutive weeks **any time** within initial treatment cycle

Potential To Disrupt Current MG Treatment Paradigm

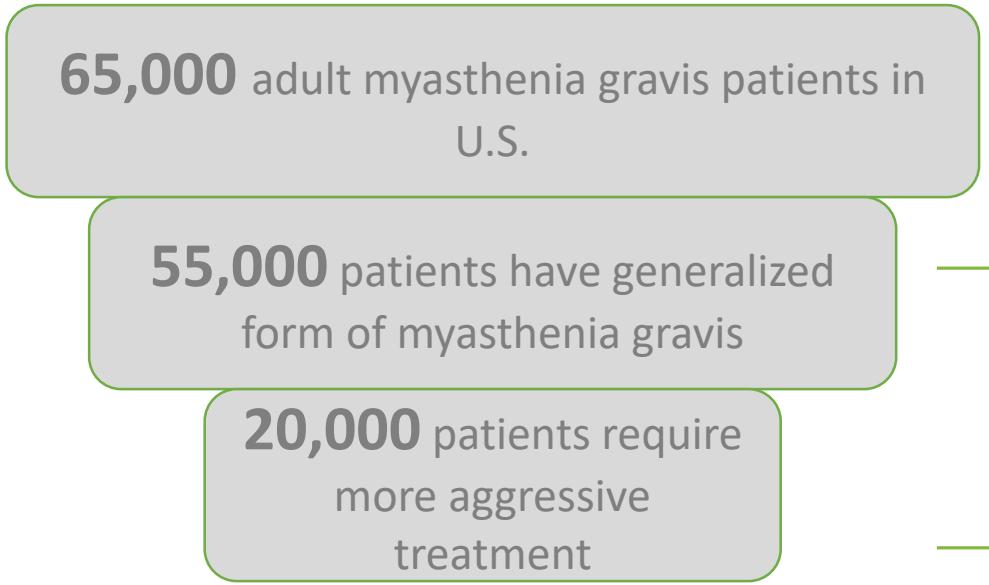
Vision: Efgartigimod positioned to be used early and more broadly within existing paradigm



Current MG Treatment Paradigm

ACIs (mestinon) at diagnosis	Steroids most common add-on	ISTs used for steroid sparing	Later agents used for severe/refractory/crisis

Addressable MG Market: Patients Who Need Therapy Beyond Steroids



85%
of MG patients have generalized form of disease

36%
of patients require treatment beyond steroids and ACIs

Estimated Efgartigimod Addressable Market — 20,000

MyRealWorld™ MG: Understanding Potential Value Proposition Of Efgartigimod

First of its kind in MG

Global prospective – longitudinal - observational

Voice of ≥ 2000 patients - digitally

Patient perspective on diagnosis, treatment, symptom, economic and humanistic burden

ITP Phase 3 ADVANCE: Evaluating IV + SC Maintenance Dosing

Patients with primary ITP with platelet counts ≤30x109/L

Trial		Primary objective
N=156 advance	* 24 weeks 10mg/kg IV (potential to adjust frequency upon response)	Durable response: sustained platelet count (≥50×10^9/L)
n≥50 advance 2	* 24 weeks 10mg/kg IV (potential to adjust frequency upon response)	Cumulative duration of platelet count response (≥50×109/L)
n≥117 advance SC	Up to 12 weekly doses 10mg/kg IV * → ≥ 20 weeks SC	Maintenance of response of SC compared to placebo after initial response with IV

* randomization

Dosing cadence to depend on response

CIDP Phase 2 ADHERE: Potential For Development Acceleration

adhere
chronic inflammatory demyelinating polyneuropathy study

Identify patients with active CIDP

Confirm IgG autoantibody involvement

Document efficacy & safety efgartigimod vs placebo

Treatment period

Open-label | Placebo-controlled

Screening (≤4weeks)

- Confirmation of diagnosis by independent committee

Run-in period (≤13weeks)

- Worsening of disease within 12 weeks after drug withdrawal (INCAT, I-RODS, grip strength)
- Newly diagnosed/ treatment naïve skip run-in period

Stage A

Efgartigimod weekly SC

Up to 12 weeks, until clinical improvement (ECI)

Stage B (Stage A responders only)

Efgartigimod weekly SC

Efgartigimod weekly SC

Up to 48 weeks

Go/No Go N=30

Efficacy analysis based on relapse (adjusted INCAT)

Study endpoint with 88 relapse events in stage B

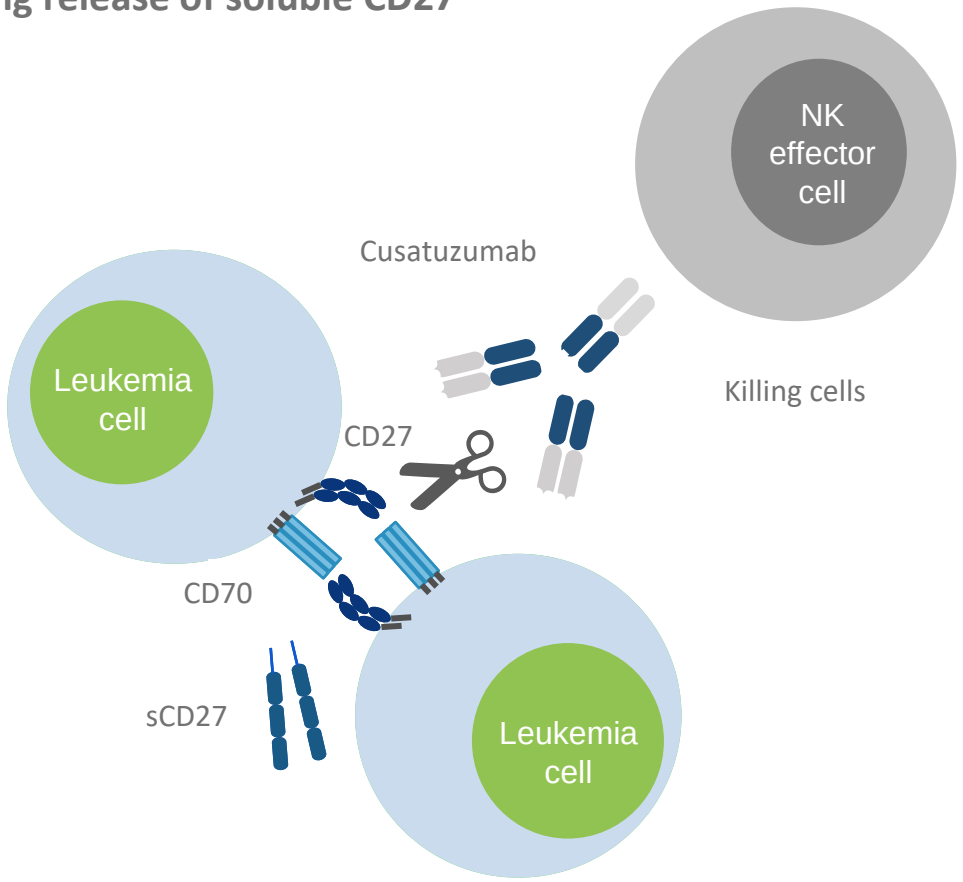
N=sample size estimation ~120-130

Followed by Open Label Extension study

Cusatuzumab Strategic Alliance With Janssen

Blocking CD70-CD27 signalling

Blocking release of soluble CD27



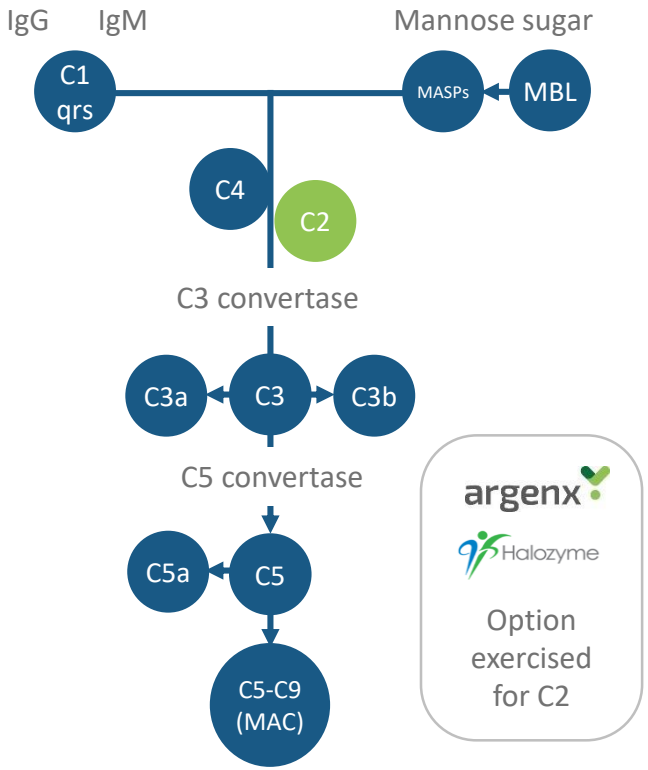
- Joint development plan focused on AML, MDS and other heme malignancies
- Upfront $300M + $200M equity @ 20% premium, up to $1.3B in milestones, double digit royalties OUS
- 50% of US economics on a royalty basis, up to 50% commercial efforts

- First trials underway (CULMINATE and ELEVATE)
- Additional trials to start, including in AML settings and subpopulations platform study, and in MDS (BEACON)

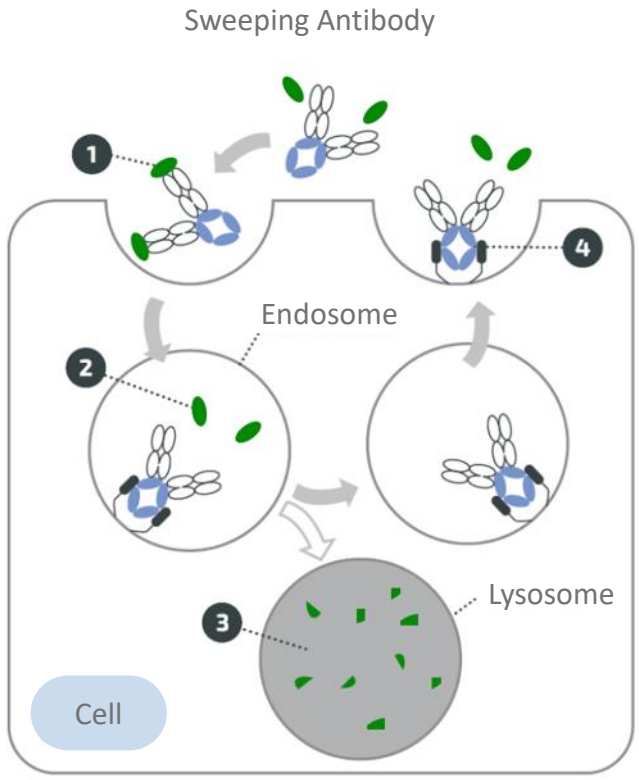
- Achieved first milestone payment under collaboration for enrollment progress in CULMINATE

ARGX-117: Sweeping Antibody Targeting C2

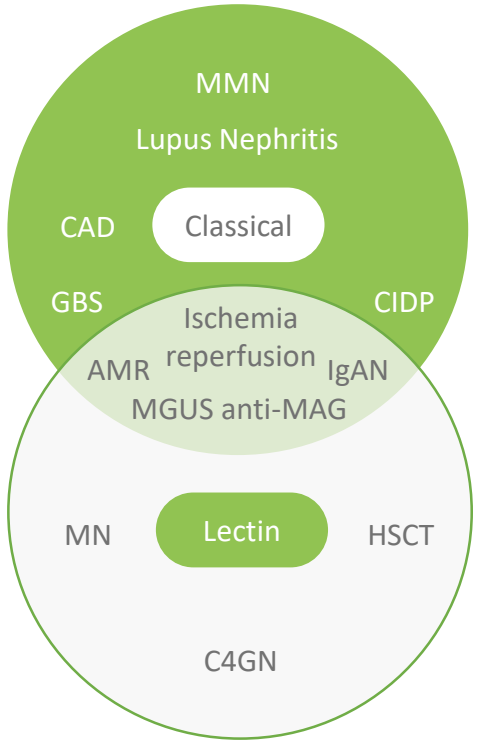
Unique Intervention in Complement Cascade



Showcase of Antibody Engineering Capabilities



Pipeline-in-a-Product Potential



Immunology Innovation Program

Accessing First-in-Class Targets by Collaborating with Leading Research Biologists

argenx

Antibody Expertise

Academic Institutions & Biotechs

Disease Biology Expertise

Co-creating immunology solutions: building beyond each individual contribution

8 assets from Immunology Innovation Program have delivered value to argenx

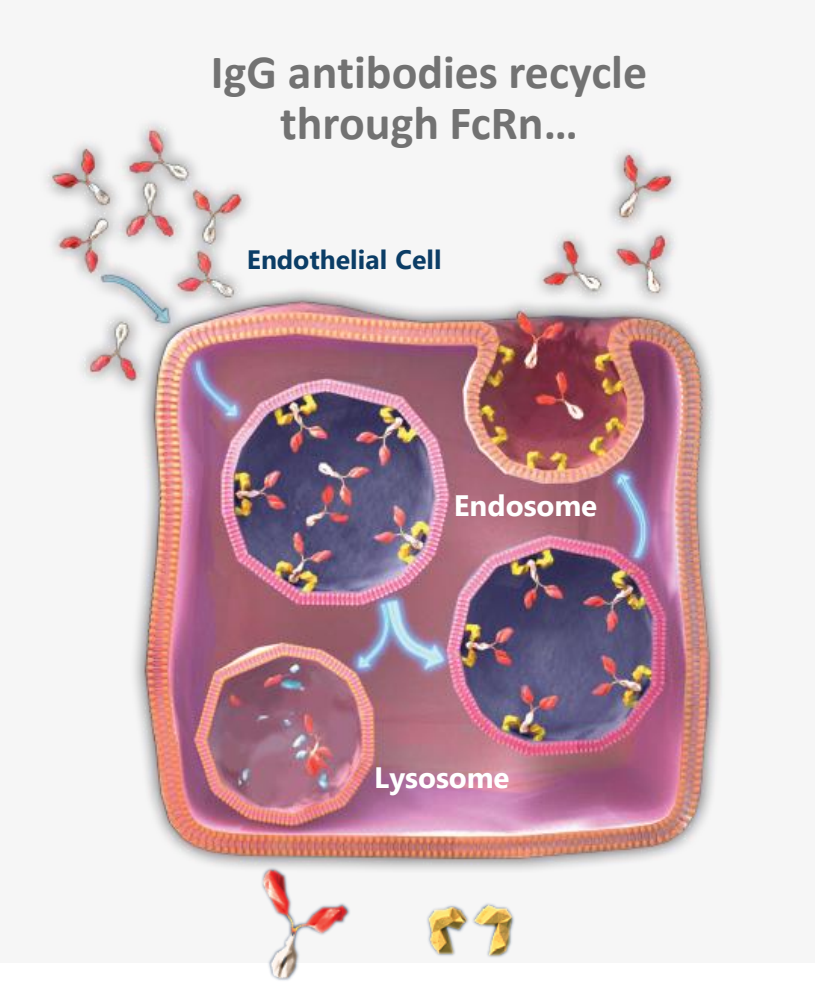
Our Key Priorities In 2020

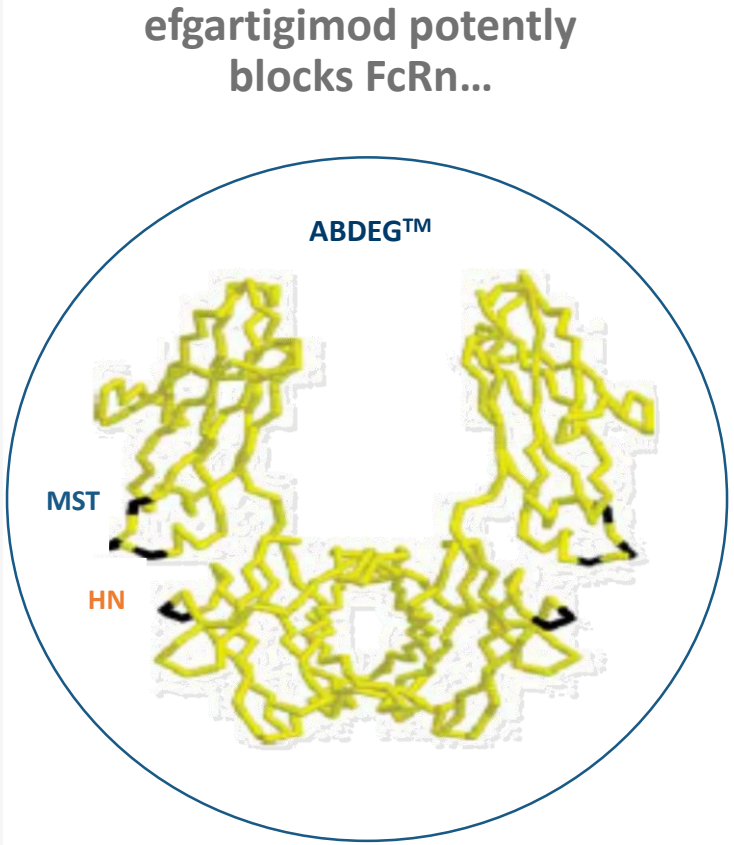
1. Prepare for launch
2. Execute pipeline:
   5 registrational and 7
   Phase 1-2 trials
3. Expand through Immunology
   Innovation Program

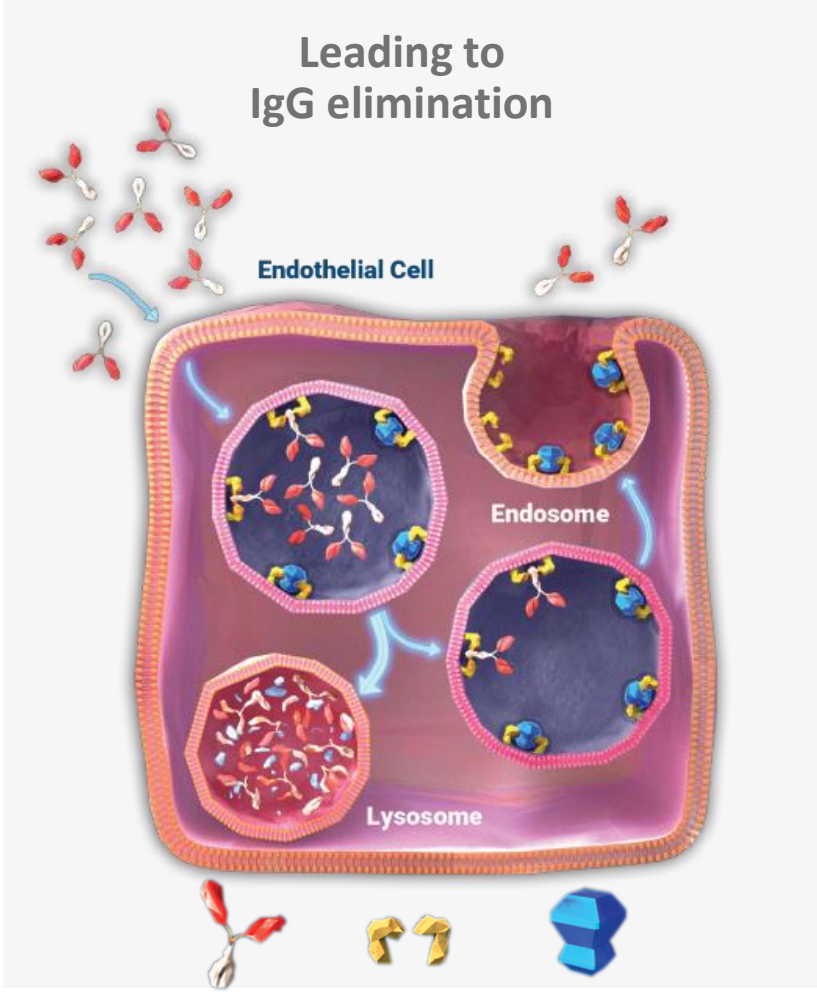
Thank You

visit argenx.com

Efgartigimod: Human IgG1 Fc Fragment With Proprietary ABDEG™ Mutations







Roopenian et al. 2007, Nat Rev Immunol. Vaccaro et al. 2005, Nat Biotech. Ulrichts et al. 2018, J Clin Invest.

Efgartigimod Portfolio Comprised Of Multiple Formulations

Standalone Products
(Built to be Interchangeable)

10 mg/kg

IV Efgartigimod

adapt myasthenia gravis study

advance immune thrombocytopenia study

advance 2 immune thrombocytopenia study

60-minute infusion

Halozyme

1000mg fixed

ENHANZE® Efgartigimod SC

adhere chronic inflammatory demyelinating polyneuropathy study

MG Strategy

Subcutaneous injection

+

10 mg/kg 360 mg fixed (2ml)

IV Efgartigimod + SC Efgartigimod

Induction Maintenance

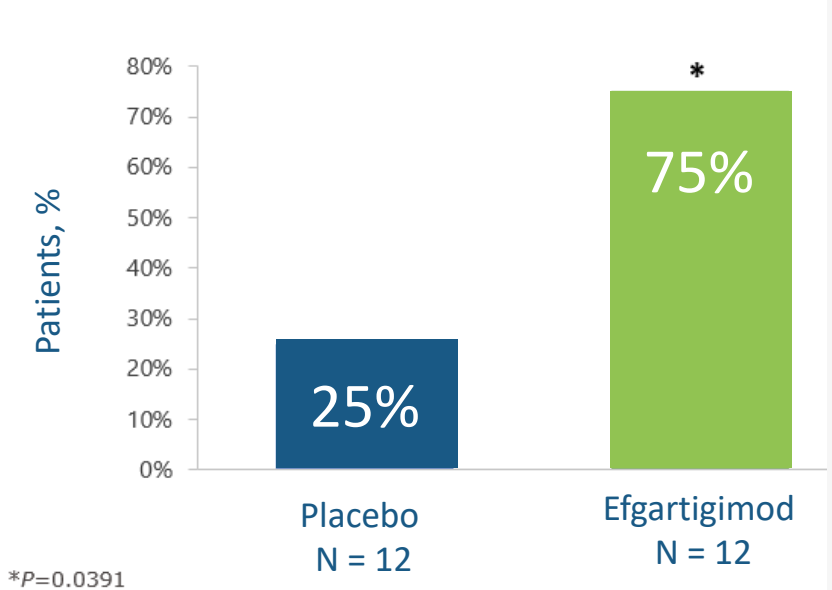
advance SC immune thrombocytopenia study

IV infusion induction
SC injection maintenance

Three Formulations Available for Use in Future Studies

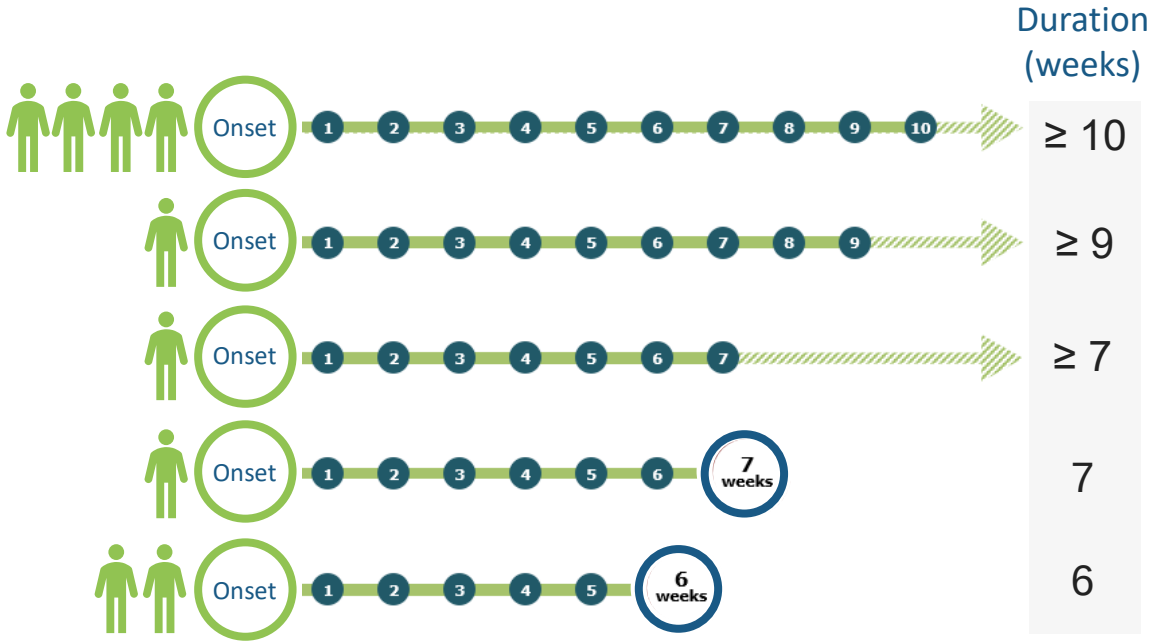
gMG Phase 2: Sustained Improvement In 75% Of Treated Patients

75% of efgartigimod group showed clinically meaningful improvement in MG-ADL for at least 6 consecutive weeks compared to 25% of placebo group



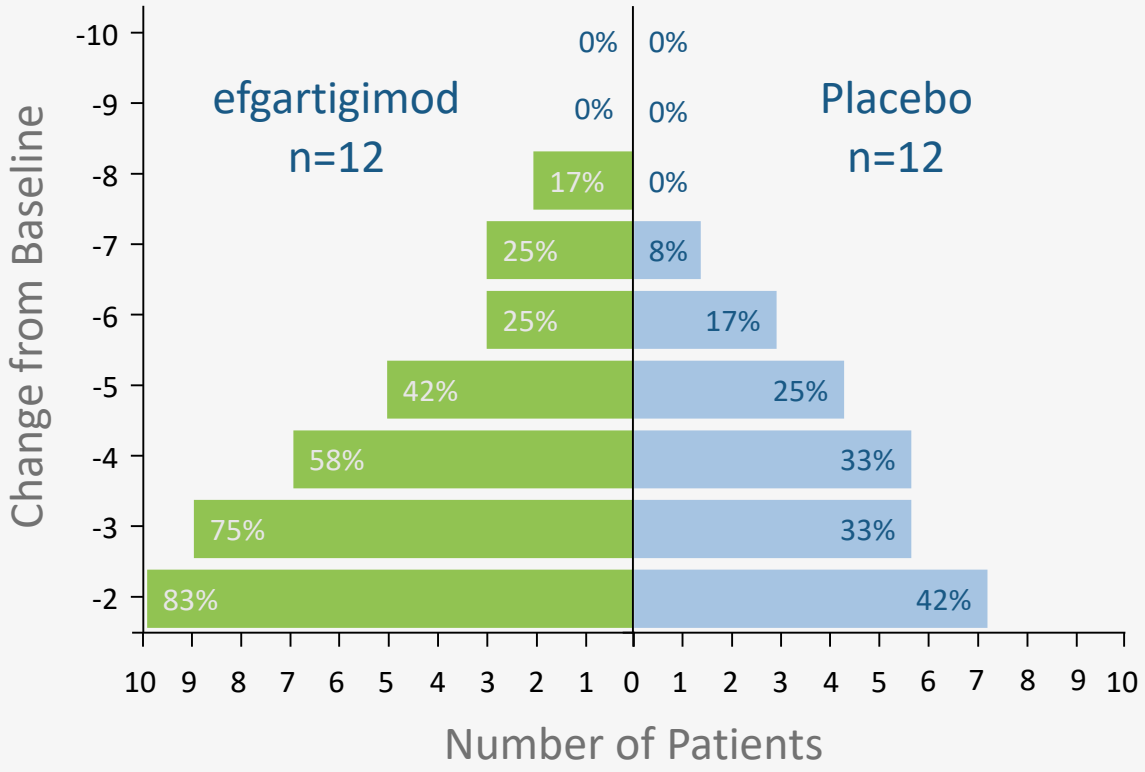
*P=0.0391
MG-ADL: myasthenia gravis activities of daily living.

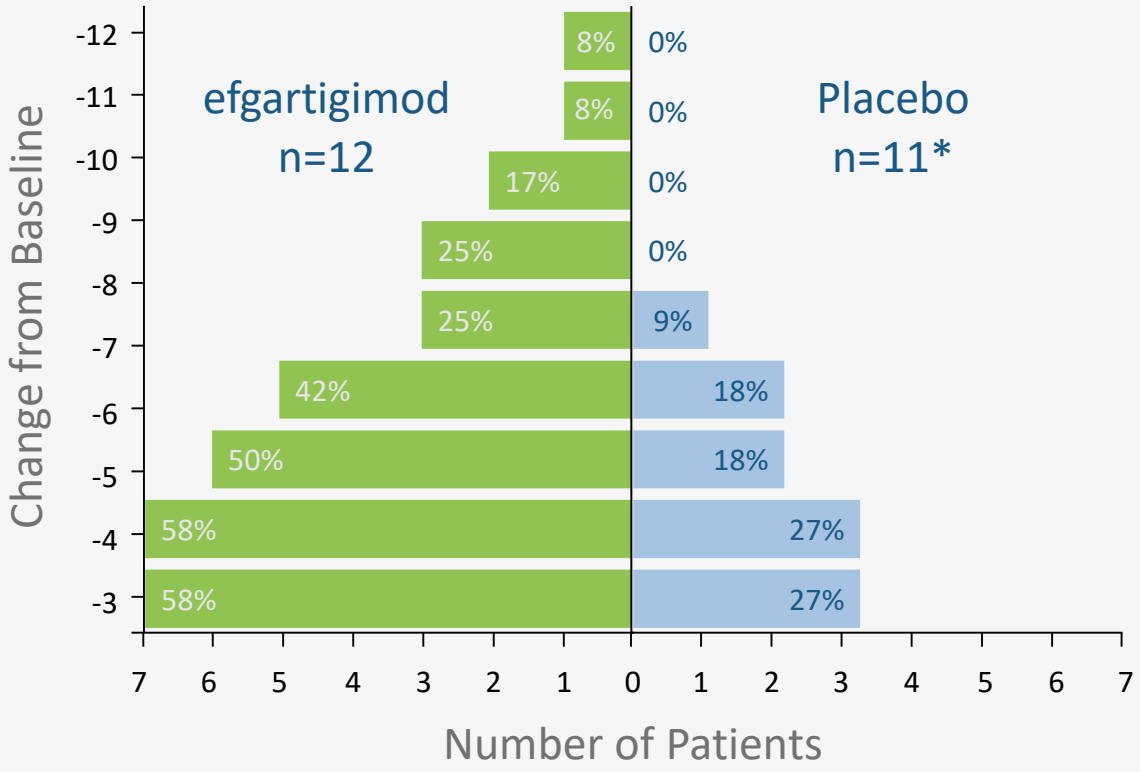
6/12 (50%) efgartigimod group had ongoing improvement at end of study



*Clinically meaningful improvement ongoing at end of study

gMG Phase 2: Proportion Of Patients With Increasing Threshold Of Benefit

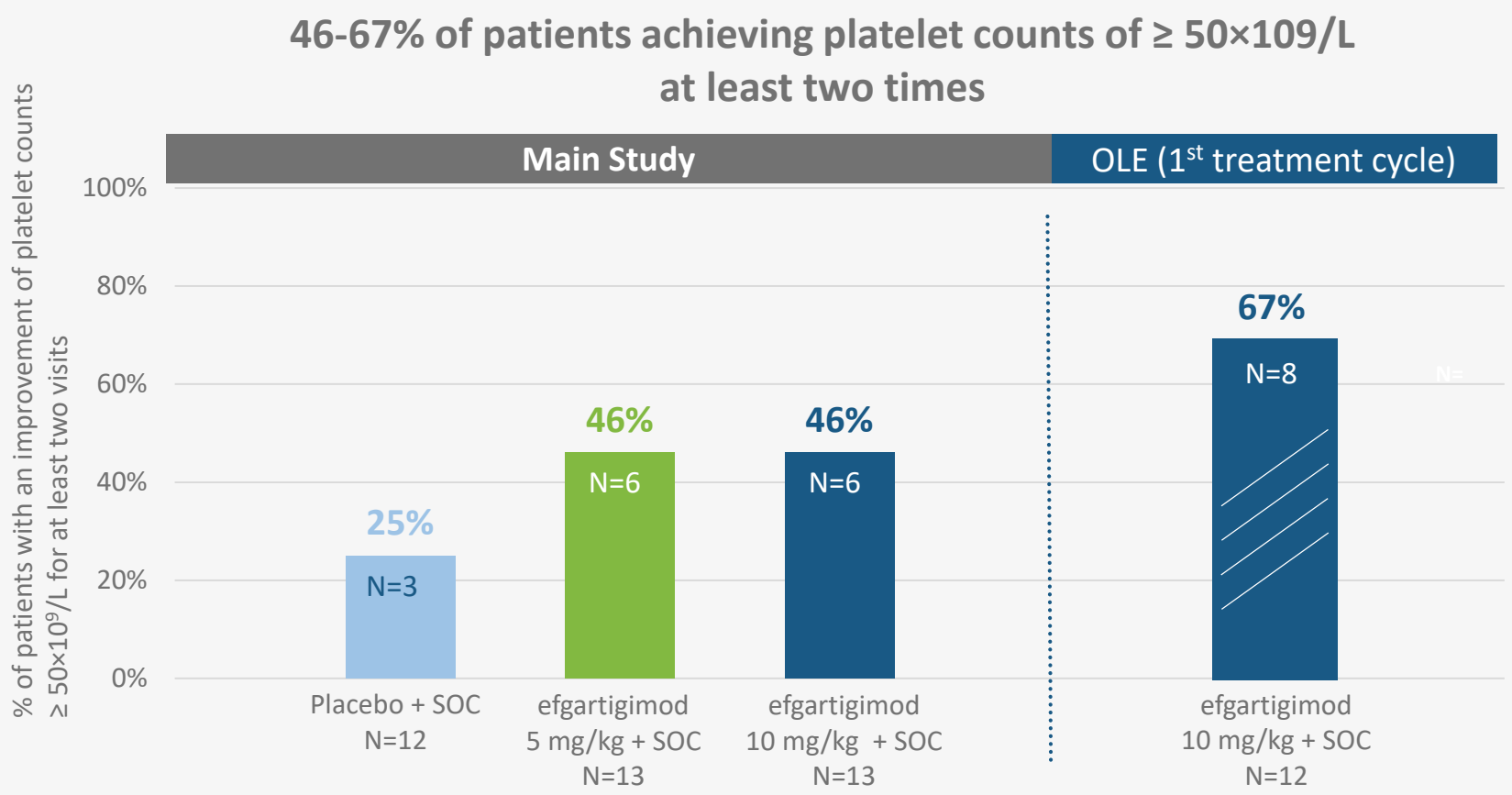




Day 29, 1 week after last infusion

Howard Jr JF. Randomized phase 2 study of FcRn antagonist efgartigimod in generalized myasthenia gravis. *Neurology* 2019;92:e1-e13

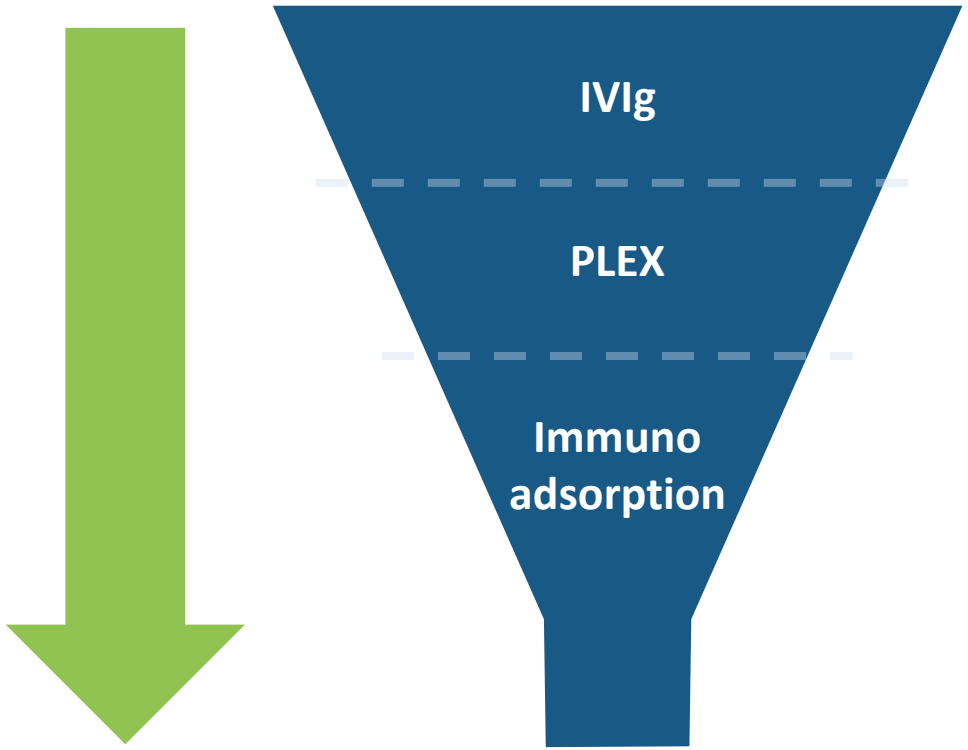
ITP Phase 2: Improvement Of Platelet Counts Across Doses



- OLE acts as fourth cohort since patients' platelets had to fall below 30x109/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

CIDP: Therapeutic Efficacy Shown With Increasing Selectivity For IgG Reductions

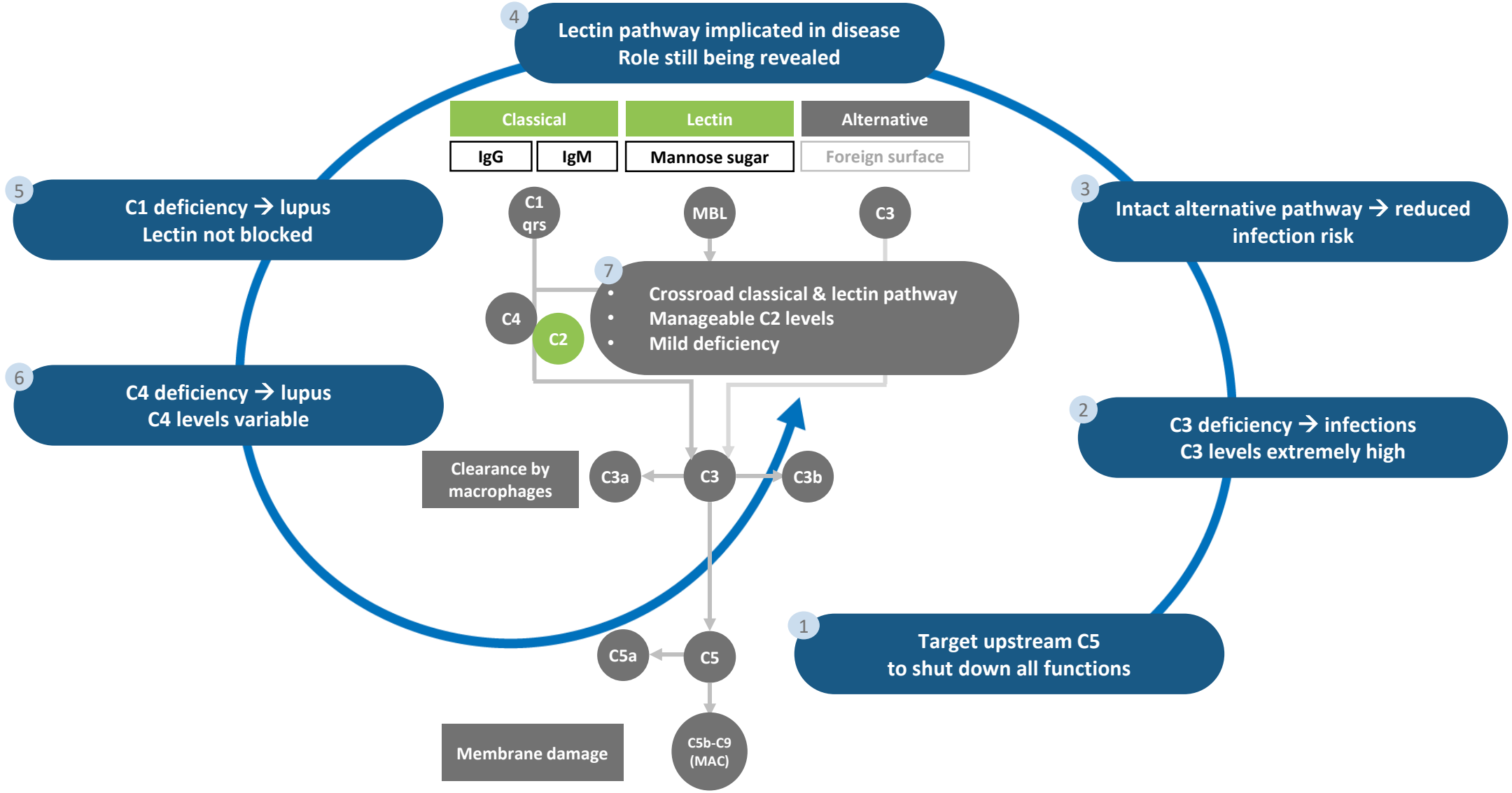
Selectivity for IgG	MoA	Response rate
IVIg	Several/undefined MoA, including increased catabolism of IgGs	50-70%
PLEX	Removal of humoral serum factors	33-80%
Immuno adsorption	Immunoadsorption (IA) with tryptophan matrix: IgG, IgM, immune complex depletion	Comparable to PLEX
	IA with protein A: IgG depletion	80-100%



Clinical evidence for the role of pathogenic autoantibodies in CIDP

Oaklander et al (2017), Cochrane Database Syst Rev Lieker et al (2017), J Clin Apher, 32(6):486-493 Zinman et al (2005) Transfus Apher Sci. 2005 Nov;33(3):317-24.

C2 At Intersection Of Classical And Lectin Pathways

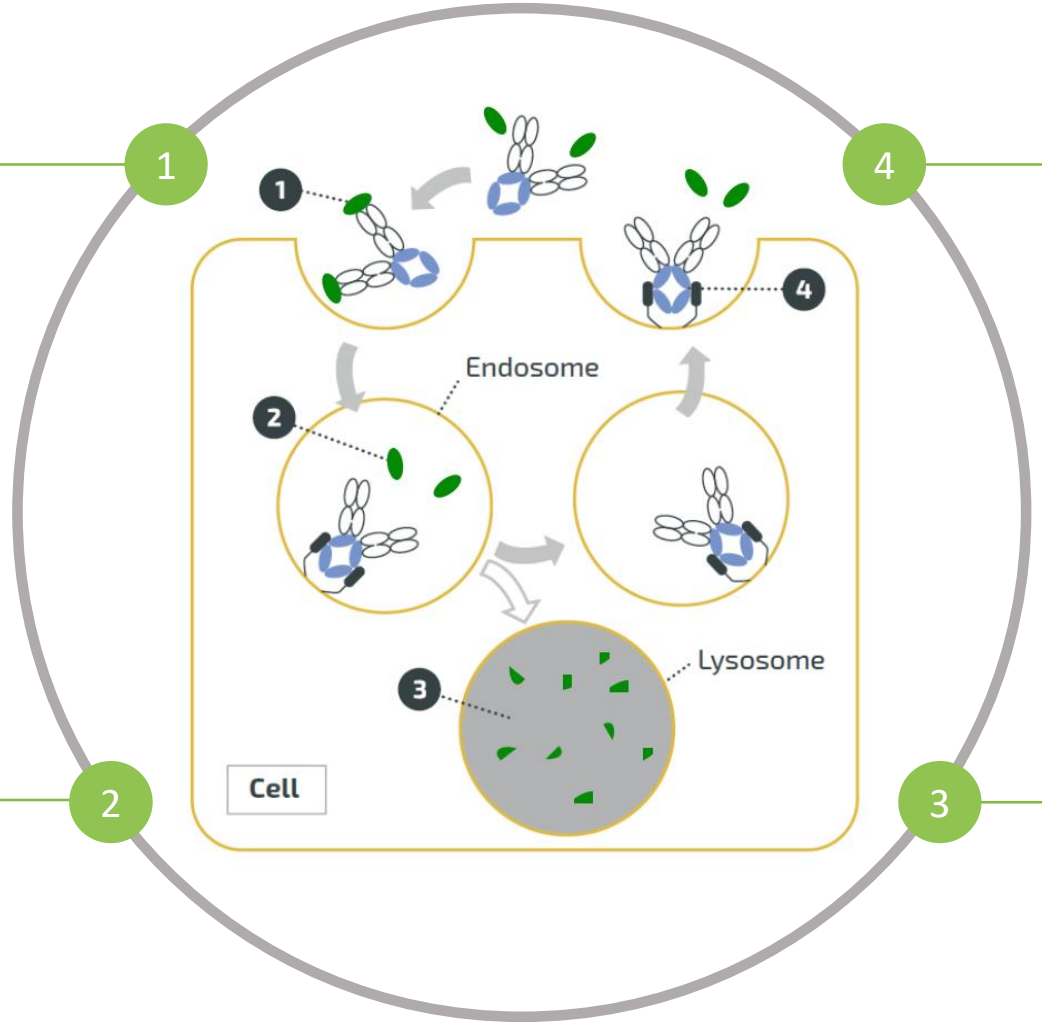


ARGX-117: Sweeping Antibody To Eliminate Serum C2

ARGX-117 binds C2 in circulation

ARGX-117/C2 complex taken up by pinocytosis entering endosome;

ARGX-117 releases C2 at lower pH in endosome



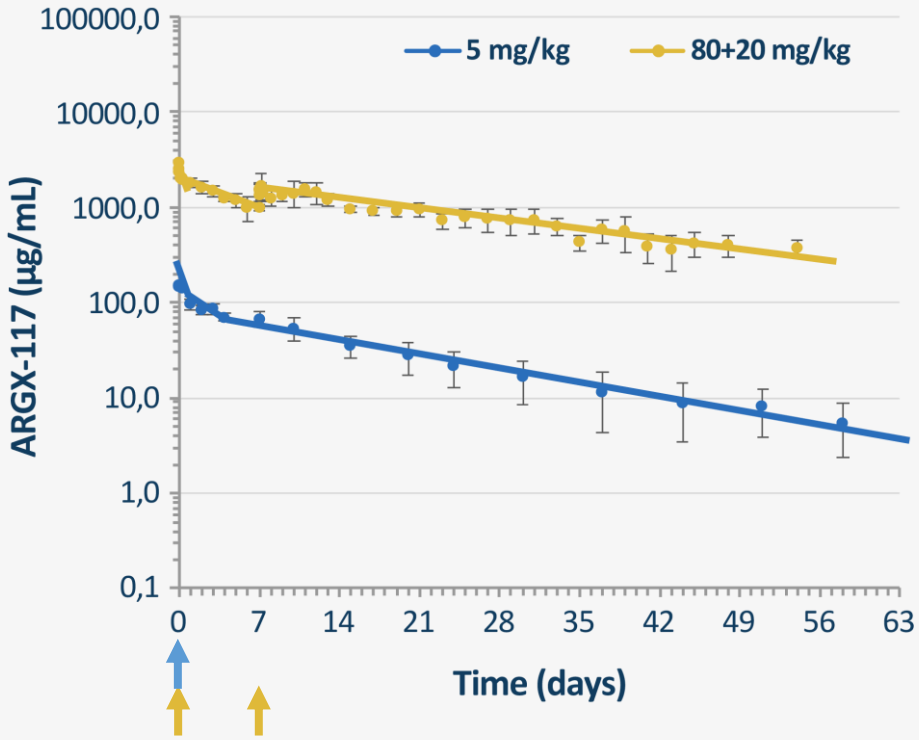
ARGX-117 remains bound going back into circulation to bind to new C2 because of NHance® mutations

C2 is degraded in lysosome

FcRn ARGX-117 C2

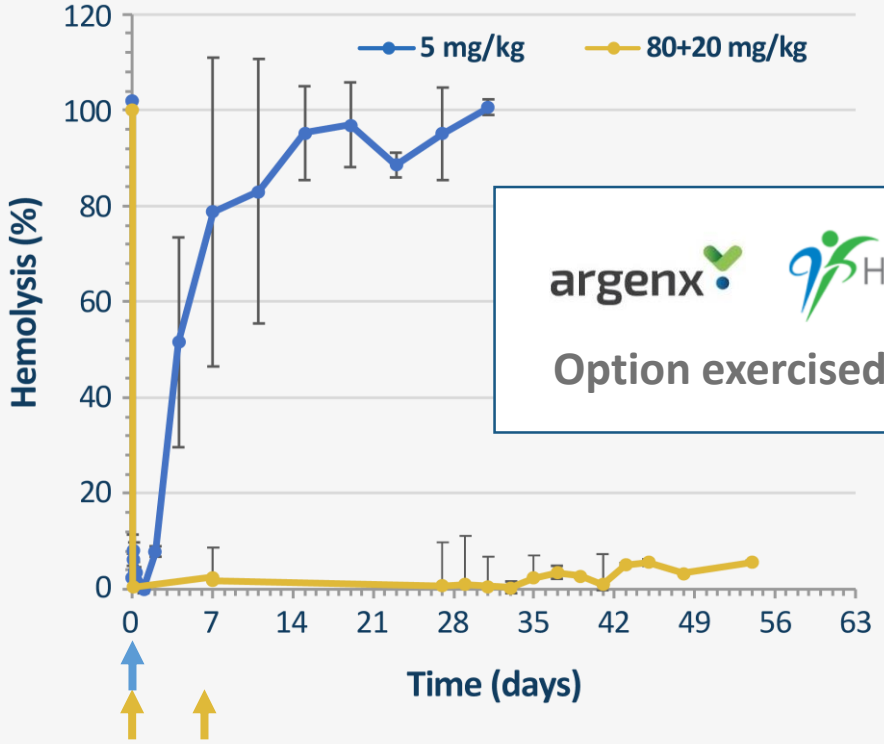
ARGX-117: Potential Dosing Optionality

Pharmacokinetics



Half-life ARGX-117: 2-3 weeks

Pharmacodynamics



argenx Halozyme

Option exercised for C2

Blocking for 2 months after repeat dosing

Cynomolgus monkey data

C2 levels cynomolgus monkey = 4x human