Together We Discover

Reaching Patients Through
Immunology Innovation

Corporate Presentation

MAY 2021

Achieving ‘argenx 2021’ Vision

argenx 2021: Reaching patients

- Commercial franchises
- Global expansion

Late-stage pipeline

- Positive Phase 3 ADAPT data
- FcRn leadership; 4/4 POC MG CIDP ITP PV
- Cusatuzumab strategic alliance

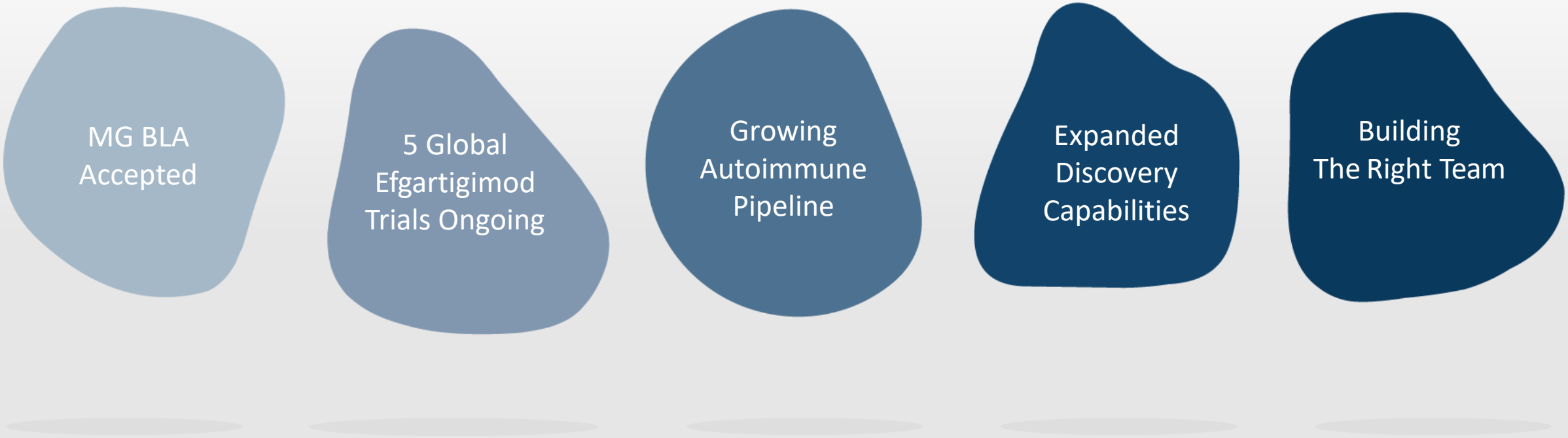
Immunology breakthroughs

- Immunology Innovation Program

Strong balance sheet

- Pro-forma cash position of ~$3.1B

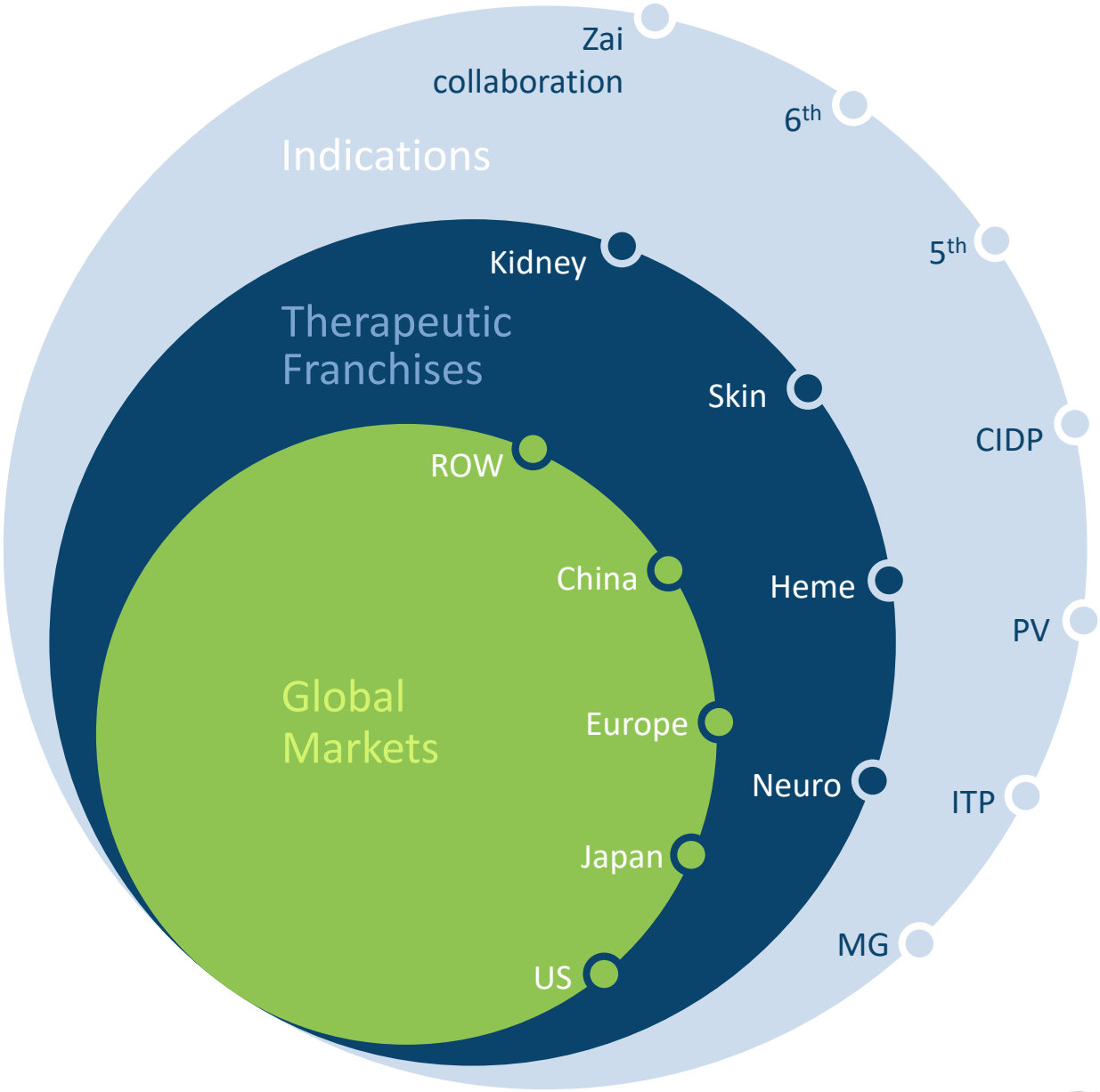
Demonstrated Execution Across Business



Uniquely Positioned

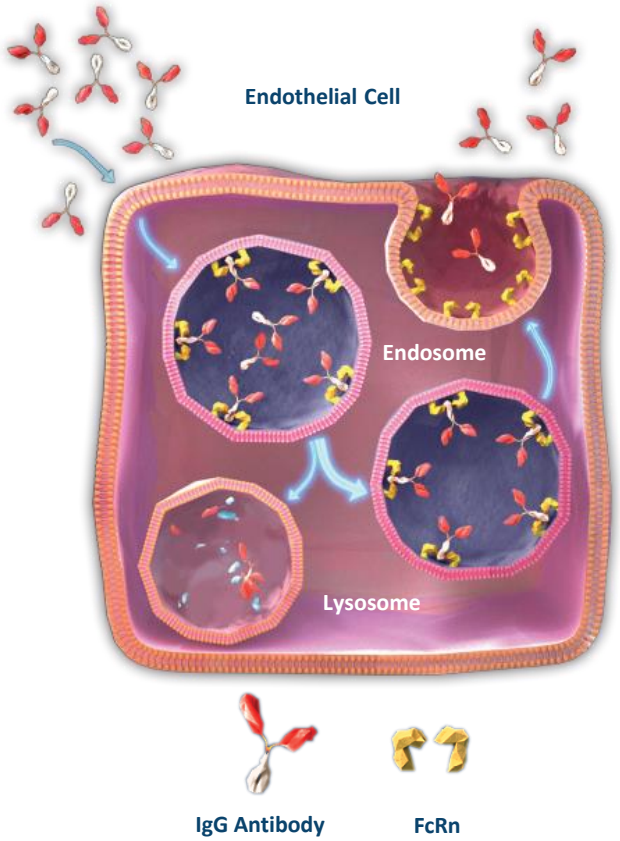
For exponential expansion

- efgartigimod indications
- therapeutic franchises
- global markets

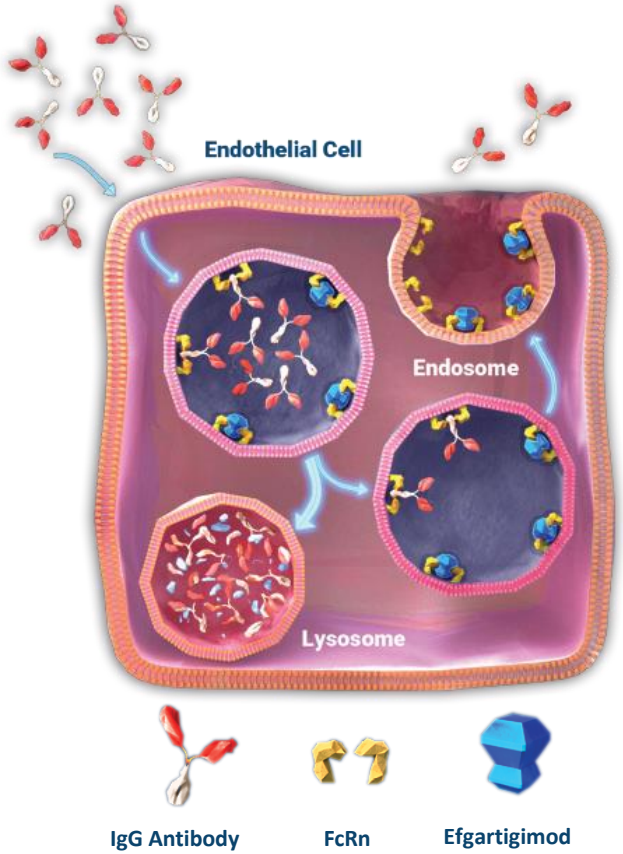


FcRn Biology is Foundational to the Immune System

FcRn recycles IgG antibodies extending their abundancy



Efgartigimod Blocks FcRn leading to IgG elimination



- Human IgG1 **Fc fragment** uniquely modulates FcRn, preserving characteristic pH dependent binding of endogenous IgG
- No impact on IgM, IgA or human serum albumin
- Does not affect IgG production, an important component to a vaccine response

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

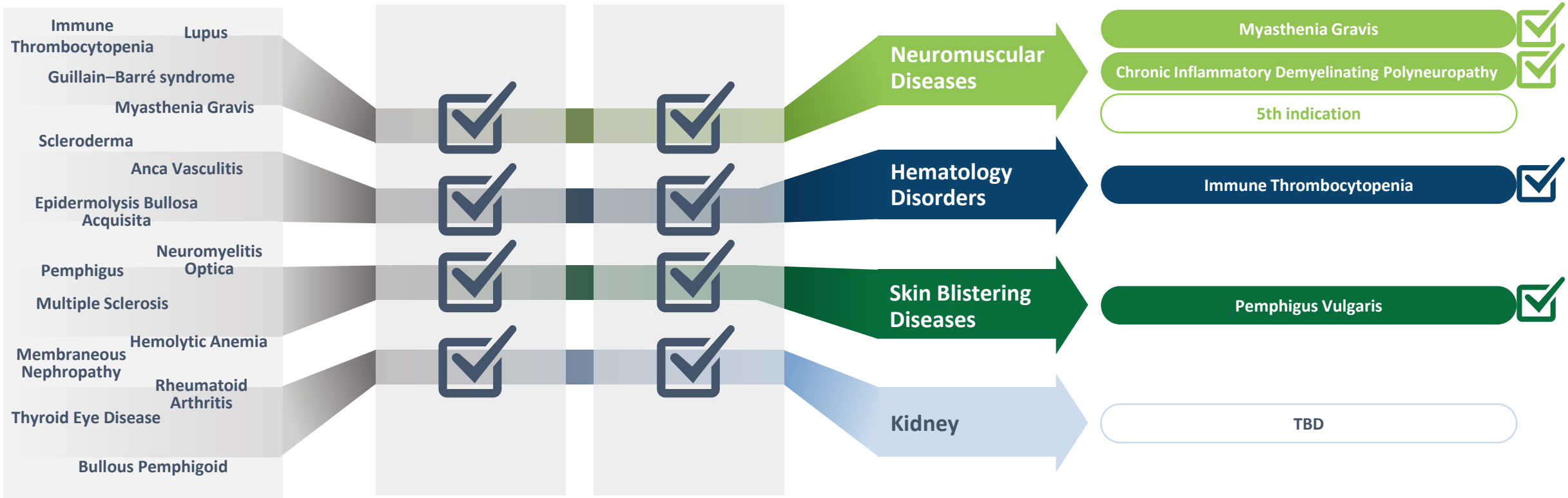
Efgartigimod: Broad Pipeline Opportunity

Landscape of IgG-mediated Severe Autoimmune Diseases
(sampling)

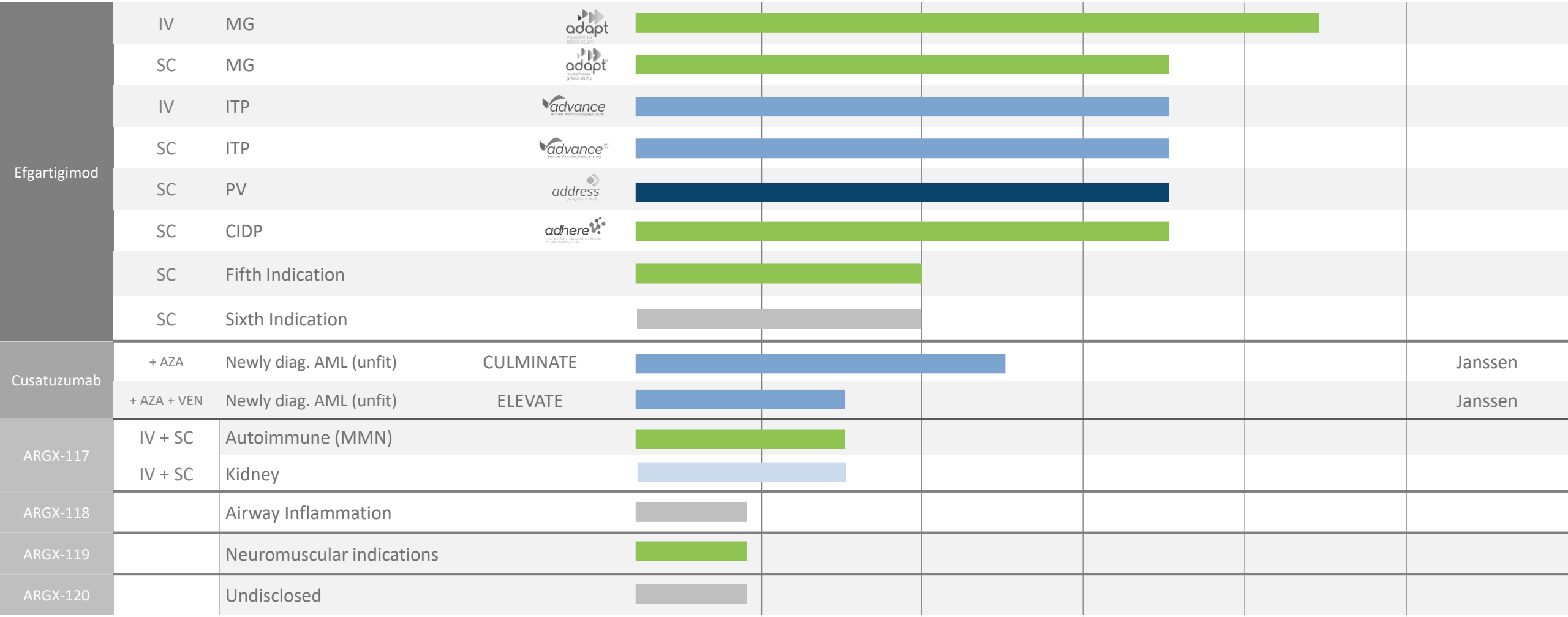
Solid Biology Rationale:
Predominantly mediated by pathogenic IgGs

Feasible for Biotech:
Orphan indication, efficient clinical & regulatory pathway

argenx Franchises & Indications
Efgartigimod to date achieved proof-of-concept in 4/4 indications; 2/2 in neuromuscular franchise



Deep Antibody Pipeline of Differentiated Candidates



Efgartigimod:
First-in-Class FcRn Antagonist

- Proof-of-concept in four indications (MG, ITP, PV, CIDP)
- IV and SC injection in development
- 350+ subjects or patients dosed
- Safety profile comparable to placebo in ADAPT trial

Patients on drug for more than 2 years

ERI
Living with MG

CHRIS
Living with MG

TERESA
Living with MG

MYASTHENIA GRAVIS

Patient Experience

GLENN
Living with MG

"A person with MG on a good day operates at 70%. On bad days, you can get 10% out of your battery."

1/2
of Patients
have been diagnosed with depression or anxiety in addition to gMG

Symptoms can vary from patient to patient, day to day, **or even throughout the same day**...this unpredictability contributes to emotional burden of disease

2.6
Years
mean time from symptom expression to diagnosis

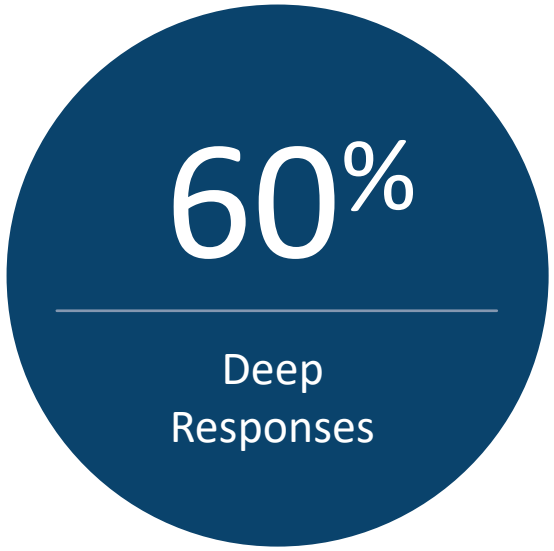
- MG affects more than patients' muscles
- Surveyed neurologists ranked severe MG only behind ALS as most severe disease they treat

Information from argenx market research

Promising Value Proposition to MG Patients

MG-ADL responders during first two cycles

MG-ADL responders within first two weeks of treatment



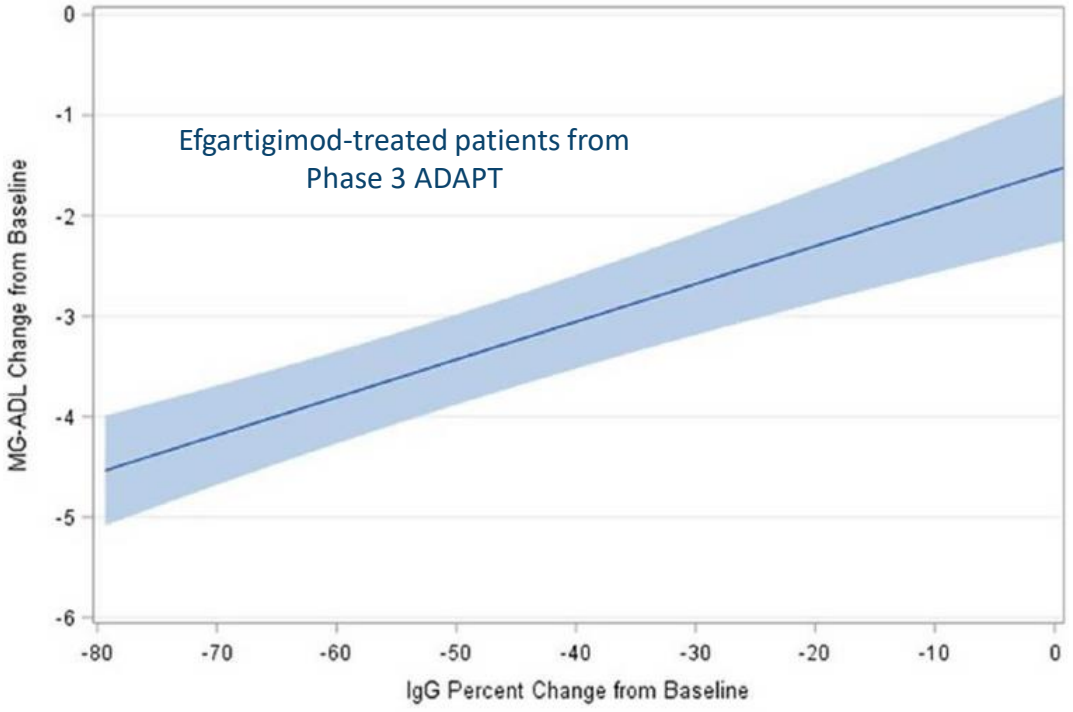
MG-ADL responders achieved minimal symptom expression (MG-ADL of 0 or 1)

Patients likely to benefit from individualized dosing

Primary endpoint: MG-ADL responder ≥2-point improvement for at least four consecutive weeks during the first cycle*
First secondary endpoint: QMG responder ≥3-point improvement for at least four consecutive weeks during the first cycle*

SC Bridging Strategy Leverages Correlation Between Pharmacodynamic and Clinical Effect

- Established association of total IgG and MG-ADL following efgartigimod treatment



- Bridging study (n=50) underway to support registration of SC efgartigimod

  - Study designed to demonstrate non-inferiority of PD effect of 1000 mg SC efgartigimod to 10mg/kg IV efgartigimod
  - Phase 1 HV data showed 1000 mg SC efgartigimod has similar PD effect as 10mg/kg IV efgartigimod
  - Additional patients from ADAPT+ to transition to SC efgartigimod

Chronic Inflammatory Demyelinating Polyneuropathy: Phase 2/3 ADHERE Trial

adhere
chronic inflammatory demyelinating polyneuropathy study

Identify patients with active CIDP

Confirm IgG autoantibody involvement

Assess efficacy & safety efgartigimod vs placebo

Treatment period

Open-label | **Placebo-controlled**

Screening	Run-in period	Stage A	Stage B (Stage A responders only)
• Confirmation of diagnosis by independent committee	• Worsening of disease within 12 weeks after drug withdrawal (INCAT, I-RODS, grip strength) • Newly diagnosed/ treatment naïve skip run-in period	Efgartigimod weekly SC	Placebo weekly SC
		Up to 12 weeks, until clinical improvement (ECI)	Efgartigimod weekly SC
			Up to 48 weeks
≤4weeks	**≤13weeks**		

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

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ITP Phase 3 ADVANCE: Two Trials Run in Parallel

Phase 3, multicenter, randomized, double-blind, placebo-controlled trial

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

N=156 advance immune thrombocytopenia study

24 weeks 10mg/kg IV efgartigimod

Fixed weekly dosing	Weekly or q2w dosing adjusted according to platelet count thresholds	Fixed weekly or q2w dosing
Weeks 1-4	Weeks 5-16	Determined at week 16

24 weeks 1000mg SC efgartigimod

N=156 advance SC immune thrombocytopenia study

Primary objective

Durable response: sustained platelet count ($\geq 50\times10^9$/L)

Efgartigimod Phase 3 Trial in Pemphigus - Focus on Potential to Drive Fast-Onset and Steroid Sparing

address
pemphigus study

Screening

Pemphigus vulgaris (PV) and foliaceus (PF)

Moderate-to-Severe Disease (PDAI activity score ≥ 15)

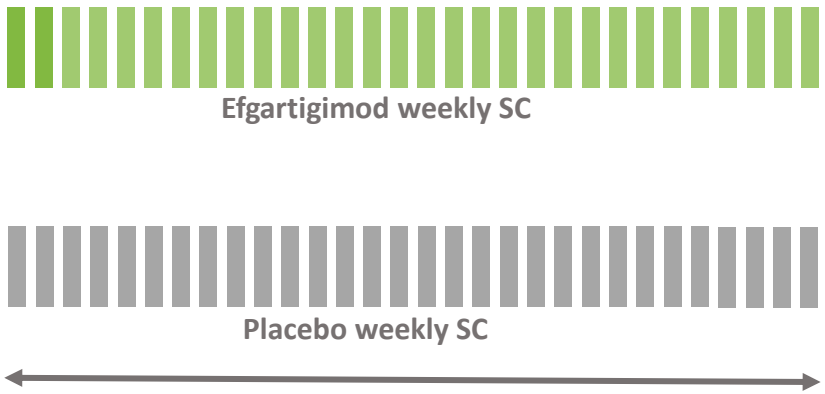
Newly Diagnosed and Relapsing

1-3 weeks

Concomitant prednisone

- Prednisone starting dose 0.5 mg/kg/day with ability to adjust
- Active tapering to start from sustained CR or EoC

Randomization (2x1)



30 weeks

Primary endpoint is proportion of PV patients achieving CRmin* within 30 weeks

N=sample size estimation ≤150 patients (PV and PF) with PF patients capped

Followed by Open Label Extension study

CR=complete clinical remission; CRmin=complete remission on minimal therapy; EoC=end of consolidation; SC=subcutaneous.

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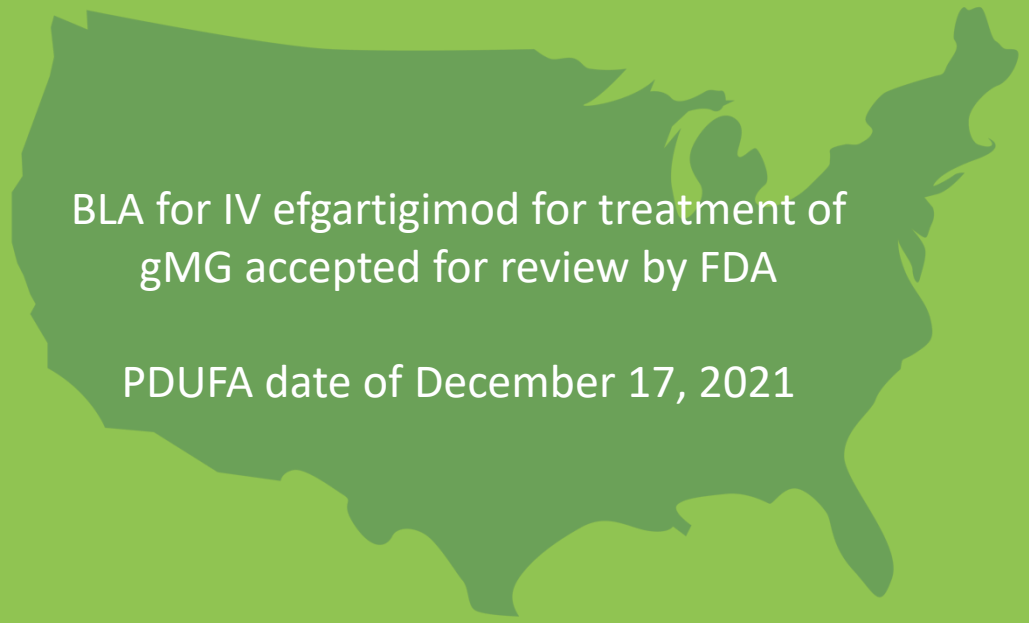
Preparing for a Successful Launch

Efgartigimod Regulatory Timelines

United States



BLA for IV efgartigimod for treatment of gMG accepted for review by FDA

PDUFA date of December 17, 2021

Global

Japan

J-MAA expected to be filed with PMDA in first half of 2021

EU

MAA expected to be filed with EMA in second half of 2021

China

Zai Lab Limited to discuss potential accelerated regulatory pathway for approval in China with NMPA

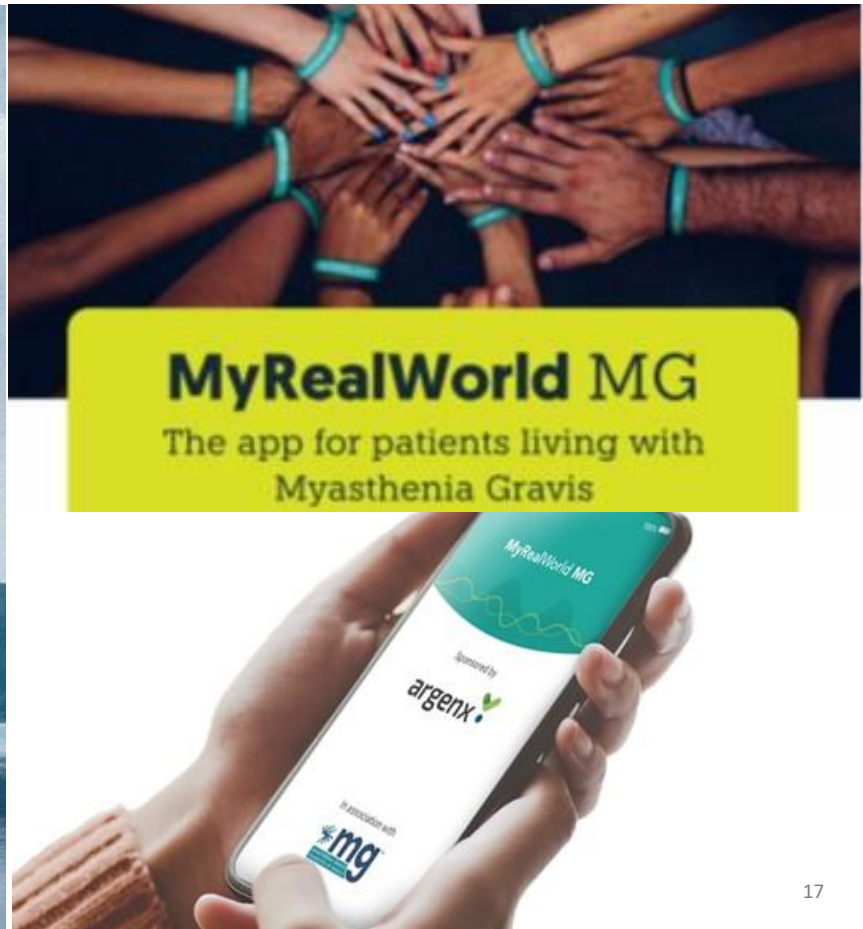
Launched Pre-Approval Access Program in the United States and Europe

Listening to and Learning from MG Community

MG United

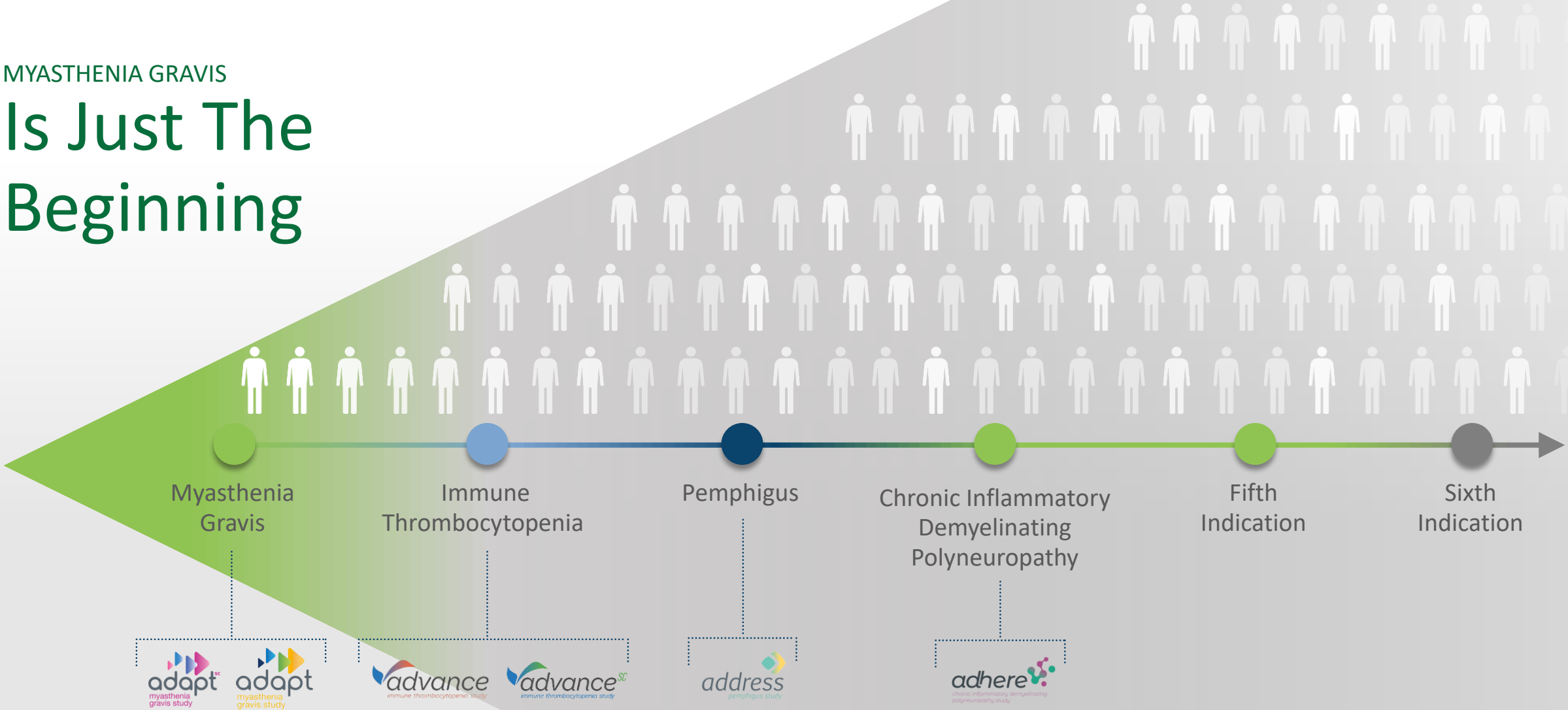
A Mystery To Me

MyRealWorld™ MG



MYASTHENIA GRAVIS

Is Just The Beginning

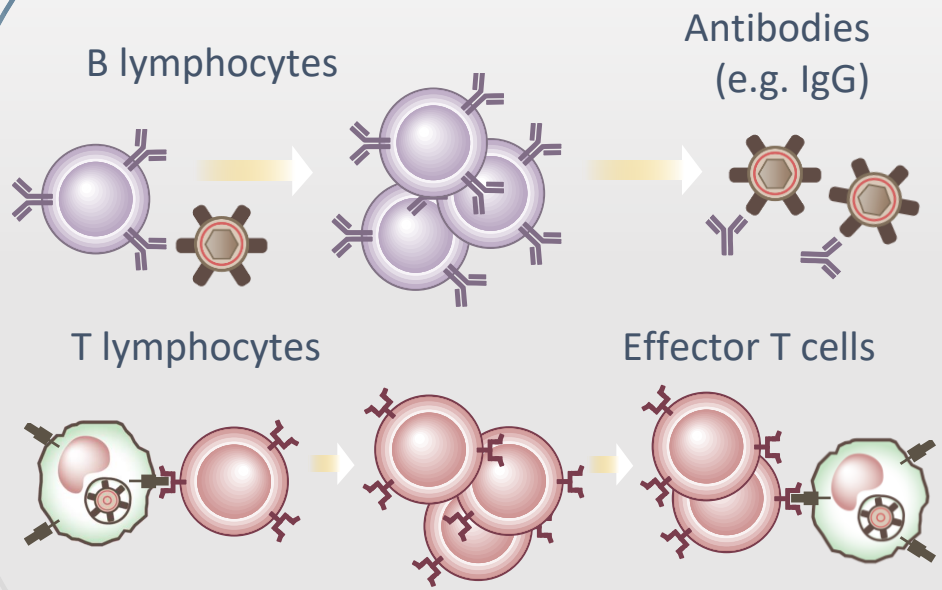


Key: NEURO HEME SKIN KIDNEY

Expanding Reach Within Immune System With ARGX-117

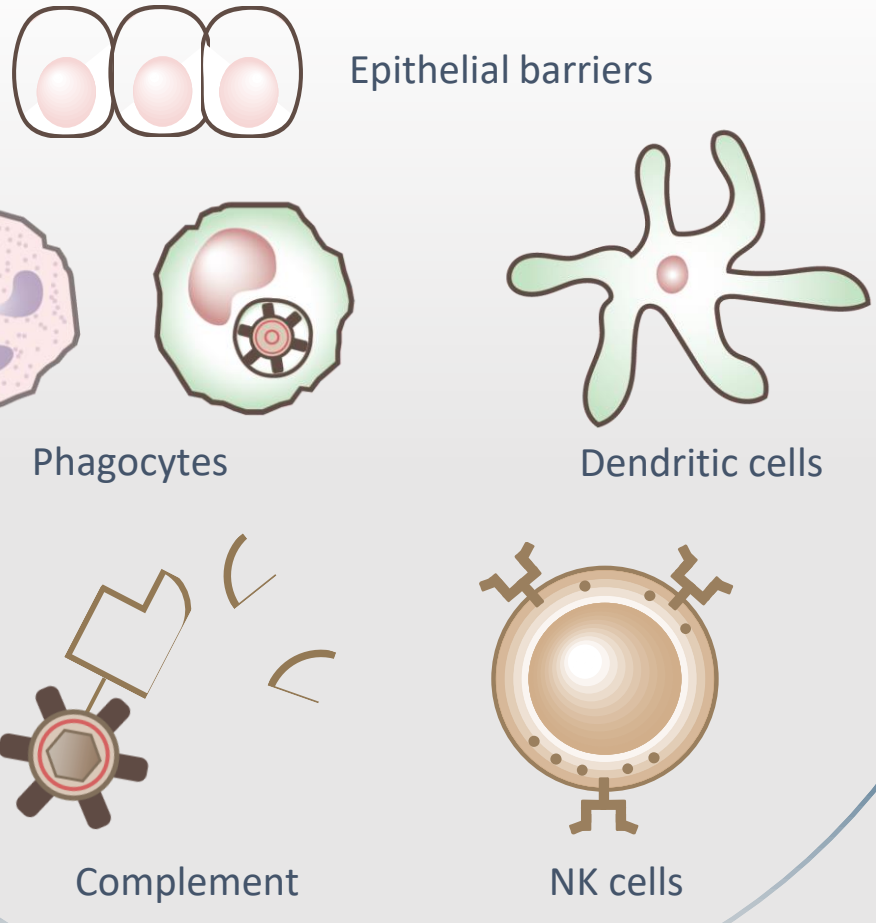
Efgartigimod

Adaptive Immune System



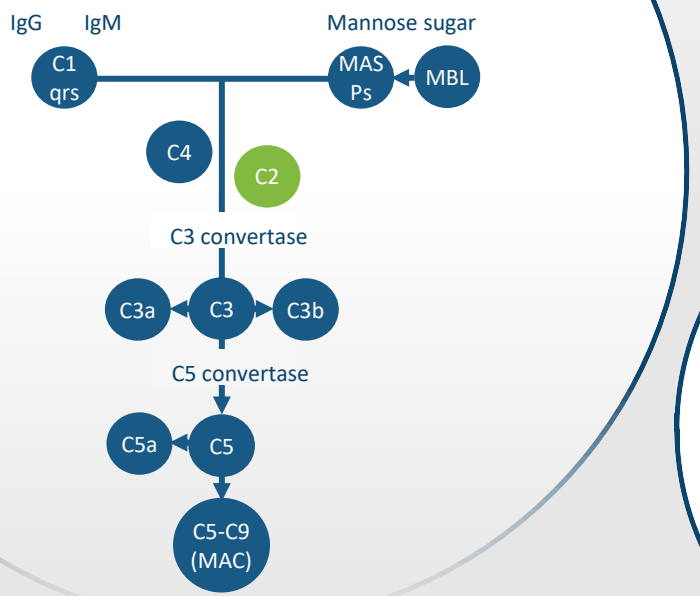
ARGX-117

Innate Immune System



ARGX-117: Broad Opportunity By Targeting C2

Unique Intervention



Phase 1 Healthy Volunteer Data Expected Mid-2021

SC and IV Formulations

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Halozyme

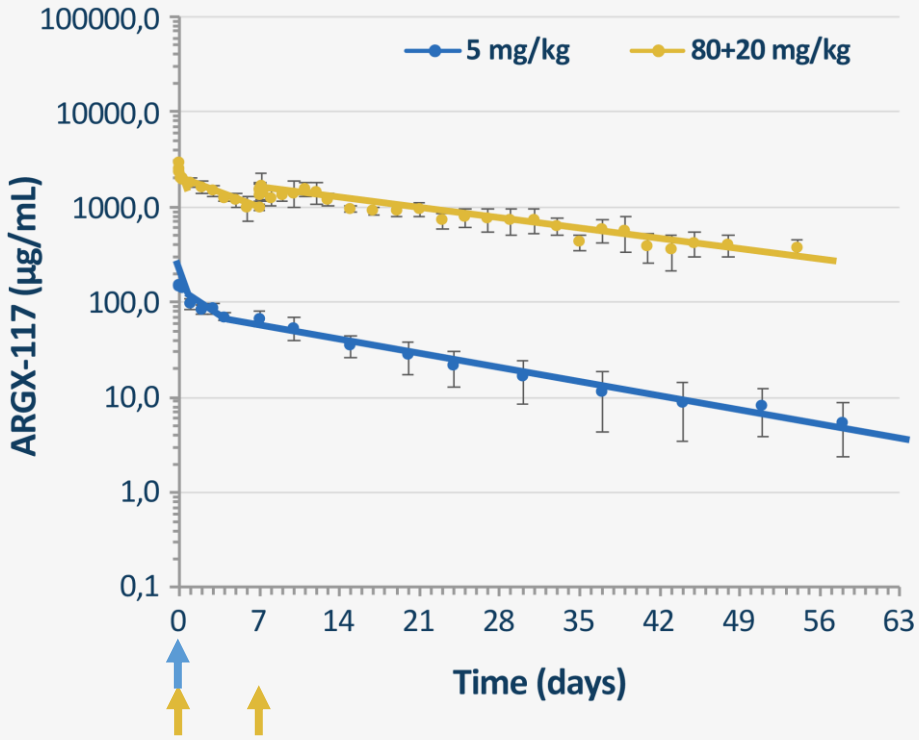
Option exercised for C2

Phase 2 Indications

- Multifocal Motor Neuropathy
- Kidney Indications

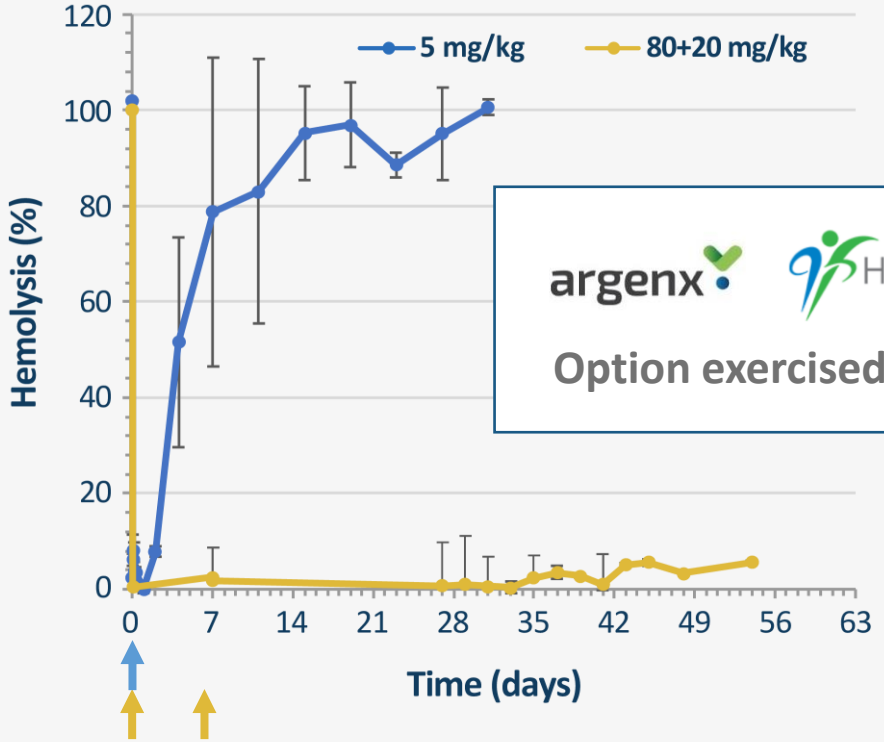
ARGX-117: Potential Dosing Optionality

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

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Feasibility: Orphan Potential & Economically Viable Indication

Unmet need for new therapies that slow down progression of disease and reduce reliance on IVIg

MMN

Multifocal Motor Neuropathy

"ALS patient that didn't die"

Slowly progressive

Asymmetric distal limb weakness mainly affecting upper limbs

Patients become dependent

Prevalence

~13,000 patients in the US

Often underdiagnosed

Predominantly men under 50

Diagnosis / Metrics

Anti-GM1 IgM antibody presence

Nerve conduction block

Defined clinical endpoints (i.e. 9-HPT, grip strength, Guy's neurological disability score)

Treatment

First line therapy is frequent, high dose of IVIg over 2-5 days

Patients unhappy with short duration of effect, disease progression despite strict adherence, side effects of IVIg

Payors aligned in need for new therapies

Cusatuzumab Strategy

Newly diagnosed elderly AML patients who are unfit for intensive chemotherapy

- Phase 2 CULMINATE Trial
  Cusatuzumab + Azacitidine

  Go-forward dose selected

20 mg/kg

CR Rates	CR	CRc
	n=14	n=21
ITT (n=52)	27%	40%
Patients who received ≥ 2 cycles (n=33)	42%	64%

30-day mortality: 5/52 (9.6%)

CRc: CR, CRi, CRh

46.2% Adverse Risk Classification (ELN)

- Phase 1b ELEVATE Trial in Triple Combination

cusatuzumab
+
azacitidine
+
venetoclax

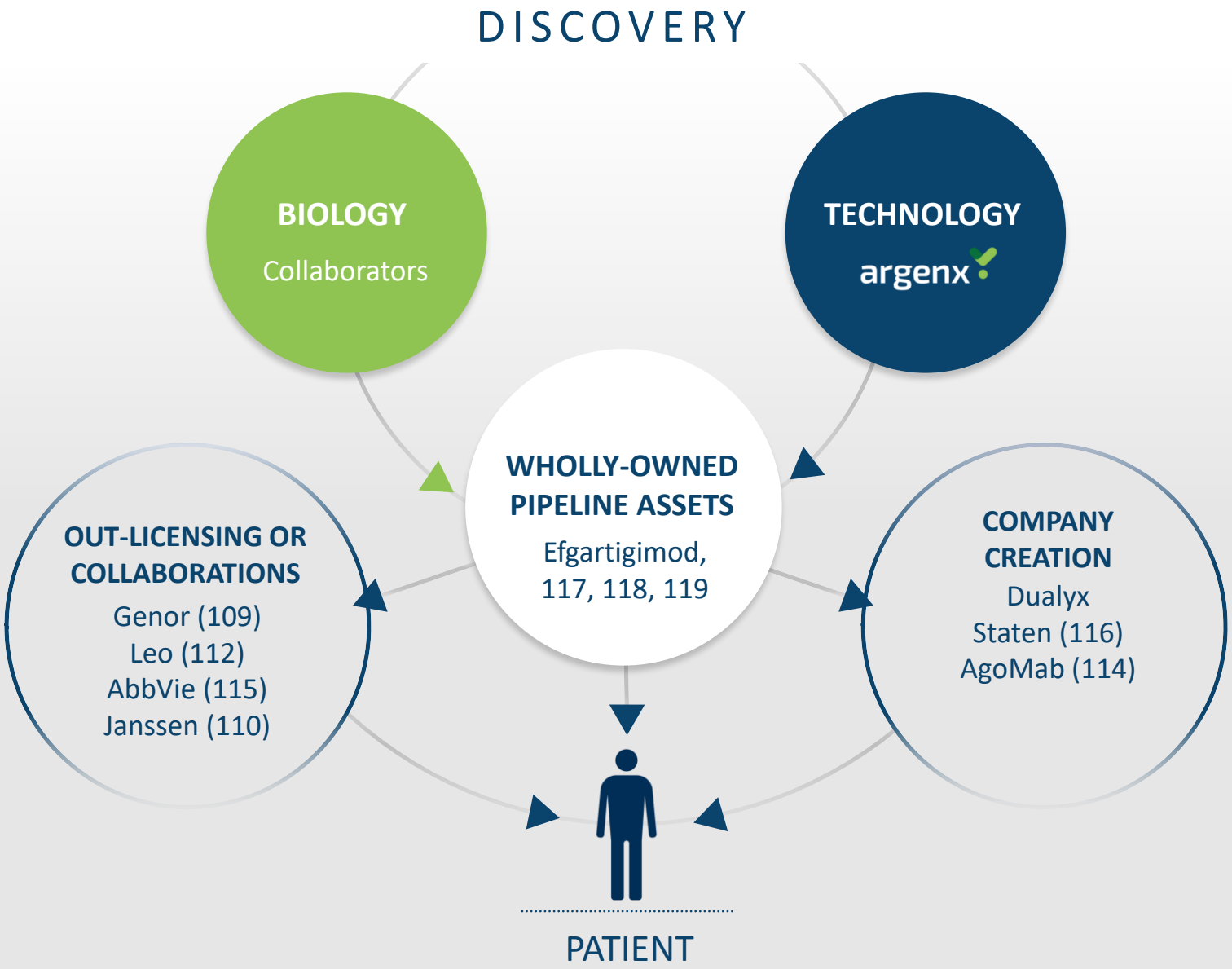
Decision to initiate additional studies will be determined following review of data from ELEVATE

CRi: Complete Remission with incomplete count recovery
CRh: Complete Remission with partial recovery of peripheral blood counts

Immunology Innovation Program (IIP)

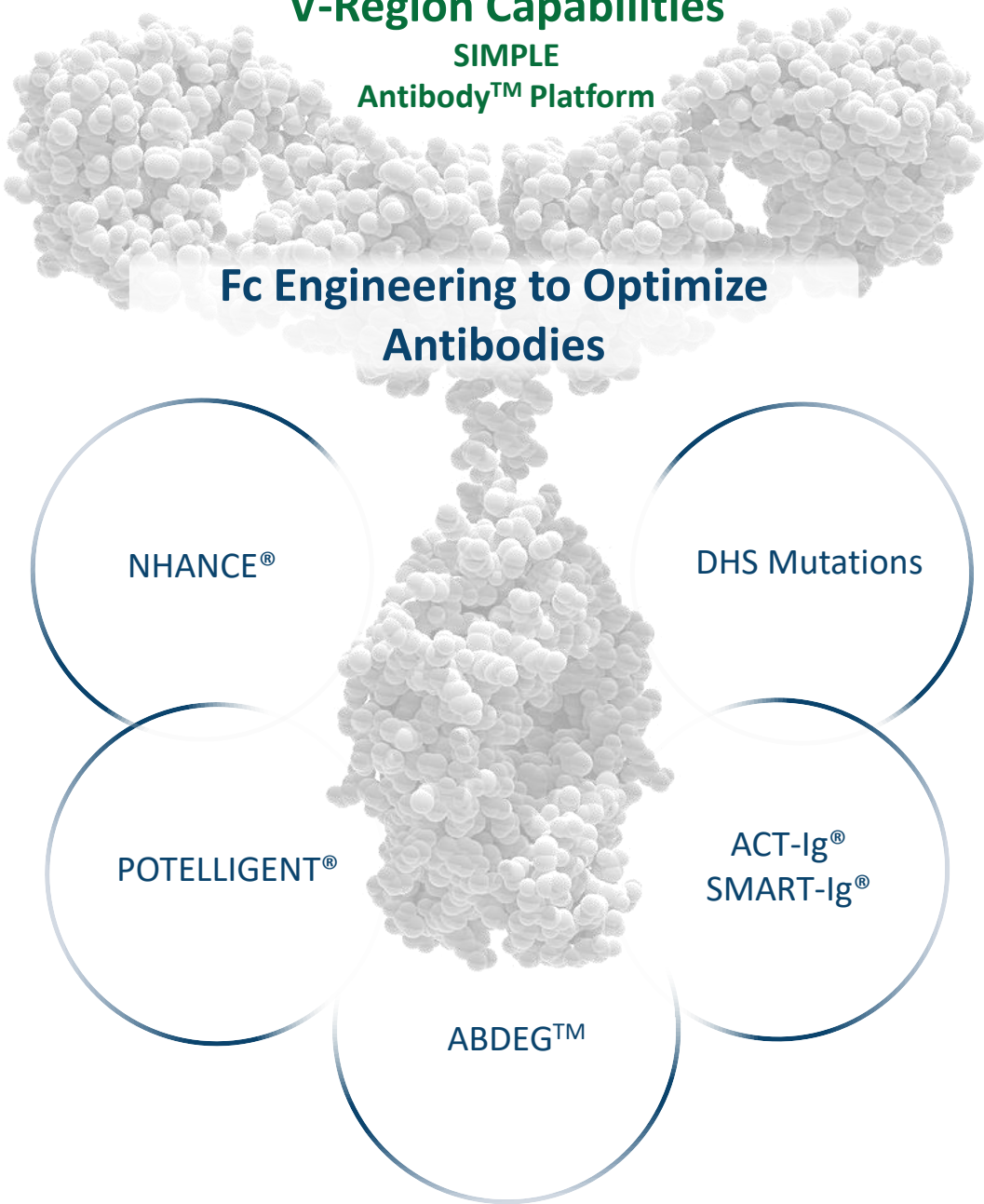
Optimizing the collision of great minds

Core Strategy To Grow Our Pipeline



Leading Antibody Discovery and Engineering Toolkit

SC Dosing Optionality
Halozyme — ENHANZE® Technology



Building Tomorrow's Immunology Company

Reach gMG patients with efgartigimod

Advance clinical development in multiple autoimmune indications

Strategic Priorities

Global expansion

Leverage IIP

Rooted in groundbreaking immunology research, growing through collaboration

Together We Discover

Reaching Patients Through
Immunology Innovation