Together We Discover

Reaching Patients Through
Immunology Innovation

First Quarter 2021 Financial Results

MAY 2021

Forward Looking Statements

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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 its statement that the submissions in China and the EU are on track and that it is well-positioned for a global launch of its first-in-class FcRn antagonist, including that BLA for IV efgartigimod for treatment of gMG accepted for review by the U.S. Food and Drug Administration (FDA) in March 2021 with target action date of December 17, 2021 under Prescription Drug User Fee Act (PDUFA), , J-MAA submitted to Japan's PMDA and accepted for review with anticipated Japan commercial launch in 2022, MAA expected to be filed with European Medicines Agency (EMA) in second half of 2021 and Zai Lab Limited to discuss potential accelerated regulatory pathway for approval in China with National Medical Products Administration (NMPA); statements regarding its commercial readiness; its statement that data expected mid-year from Phase 1 trial of C2 inhibitor, ARGX-117; its hope to reach patients this year; its statements regarding the therapeutic potential of Efgartigimod in patients with gMG as well as several other severe autoimmune diseases mediated by IgG autoantibodies; its plans to start enrollment in two additional efgartigimod indications this year, its expectation to have Phase 1 data mid-year for its C2 inhibitor, ARGX-117, our business and financial outlook and related plans; the therapeutic and commercial potential of our product candidates; the intended results of our strategy; the expected benefits of our collaborations, including with respect to our collaboration with Zai Lab; our and our collaboration partners' clinical development and regulatory plans, including the timing, design and outcome of ongoing and planned clinical trials and preclinical activities and the timing and outcome of regulatory filings and approvals; the timing, progress and benefits of marketing and commercialization activities; and the expected size of the markets for our product candidates. 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 COVID-19 pandemic on our business, 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 costsand 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 govern- mental 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.

Achieving ‘argenx 2021’ Vision

argenx 2021: Reaching patients

- Commercial franchises
- Global expansion 

Late-stage pipeline

- FcRn leadership, 4/4 POC MG CIDP ITP PV
- ARGX-117 pipeline-in-a-product opportunity MMN
- Cusatuzumab strategic alliance

Immunology breakthroughs

- Immunology Innovation Program

Strong balance sheet

- Pro-forma cash position of $3B

Promising Value Proposition to MG Patients

78%

Response Rate

MG-ADL responders during first two cycles

84%

Fast Onset of Action

MG-ADL responders within first two weeks of treatment

60%

Deep Responses

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

OVER 50%

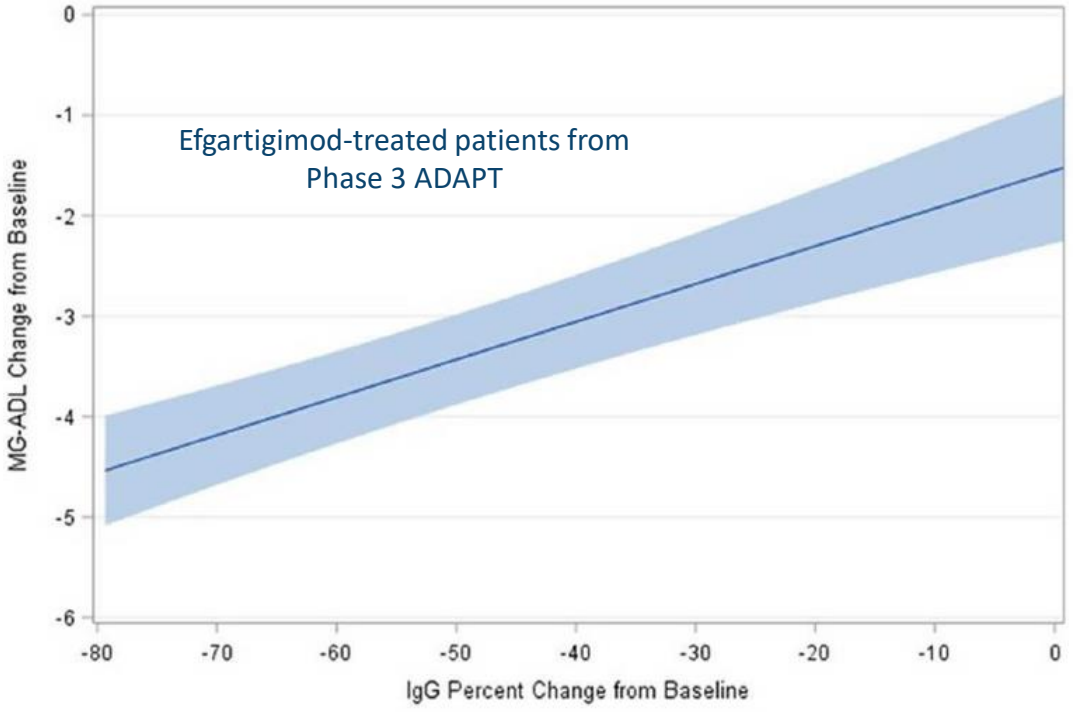
Supportive of Individualized Dosing

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
  - Primary endpoint assessment at day 29

Efgartigimod:
First-in-Class FcRn Antagonist

- Proof-of-concept in four indications (MG, ITP, PV, CIDP)
- IV and SC injection in development
- 400+ 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

Chronic Inflammatory Demyelinating Polyneuropathy: Phase 2/3 ADHERE Trial

adhere
chronic inflammatory demyelinating polyneuropathy study

Identify patients with active CIDP

Screening

- Confirmation of diagnosis by independent committee

≤4weeks

Run-in period

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

≤13weeks

Confirm IgG autoantibody involvement

Assess efficacy & safety efgartigimod vs placebo

Treatment period

Open-label | **Placebo-controlled**

Stage A

Efgartigimod weekly SC

Up to 12 weeks, until clinical improvement (ECI)

Stage B (Stage A responders only)

Placebo 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

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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)

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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)

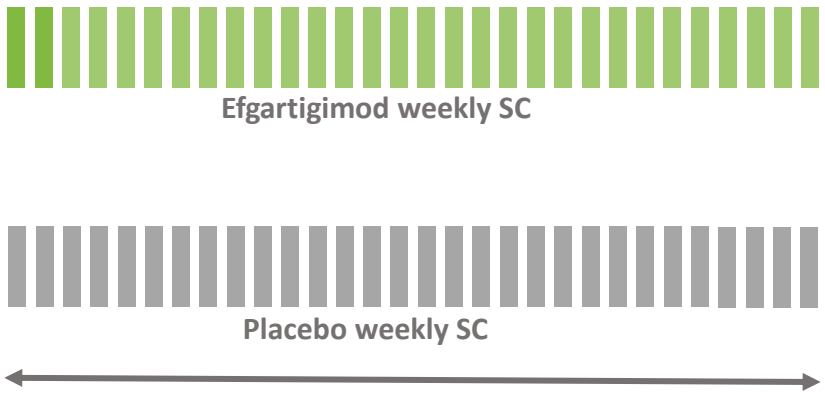
Newly Diagnosed and Relapsing

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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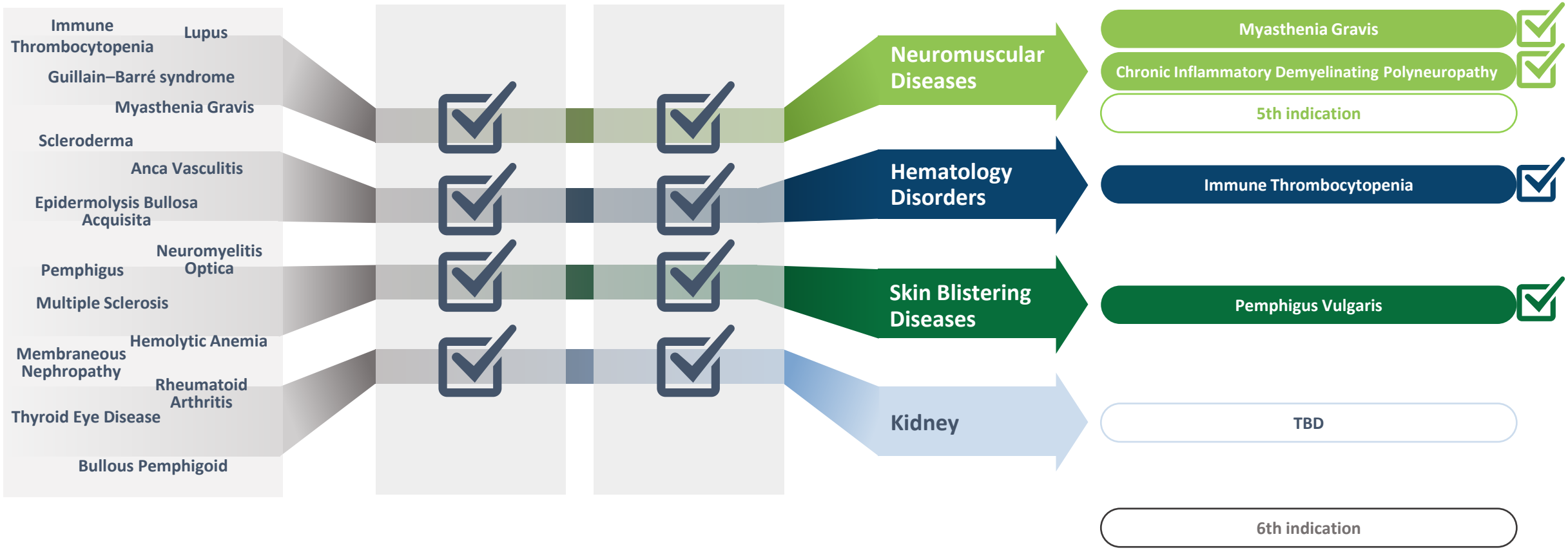
Efgartigimod: Broad Pipeline Opportunity

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



Reaching Patients Worldwide

argenx + Zai Lab

Expand

patients we reach + number of indications

Accelerate

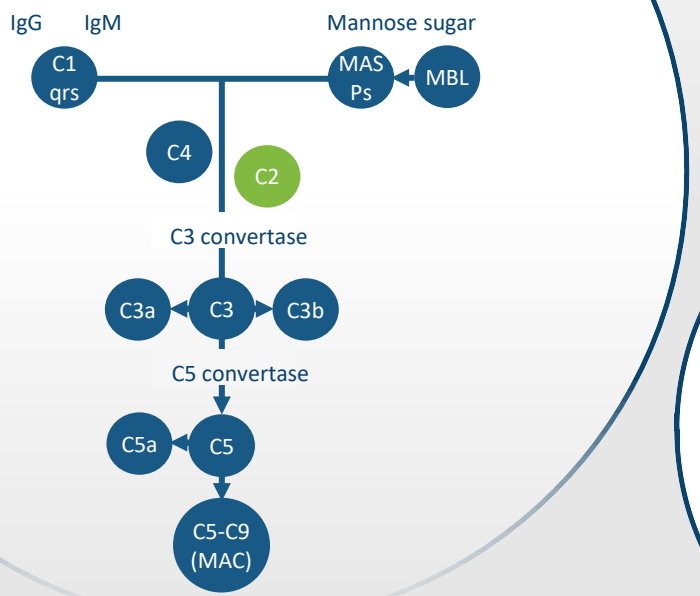
enrollment into global registrational trials

Mutual Passion

bringing innovative therapies to patients

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

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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

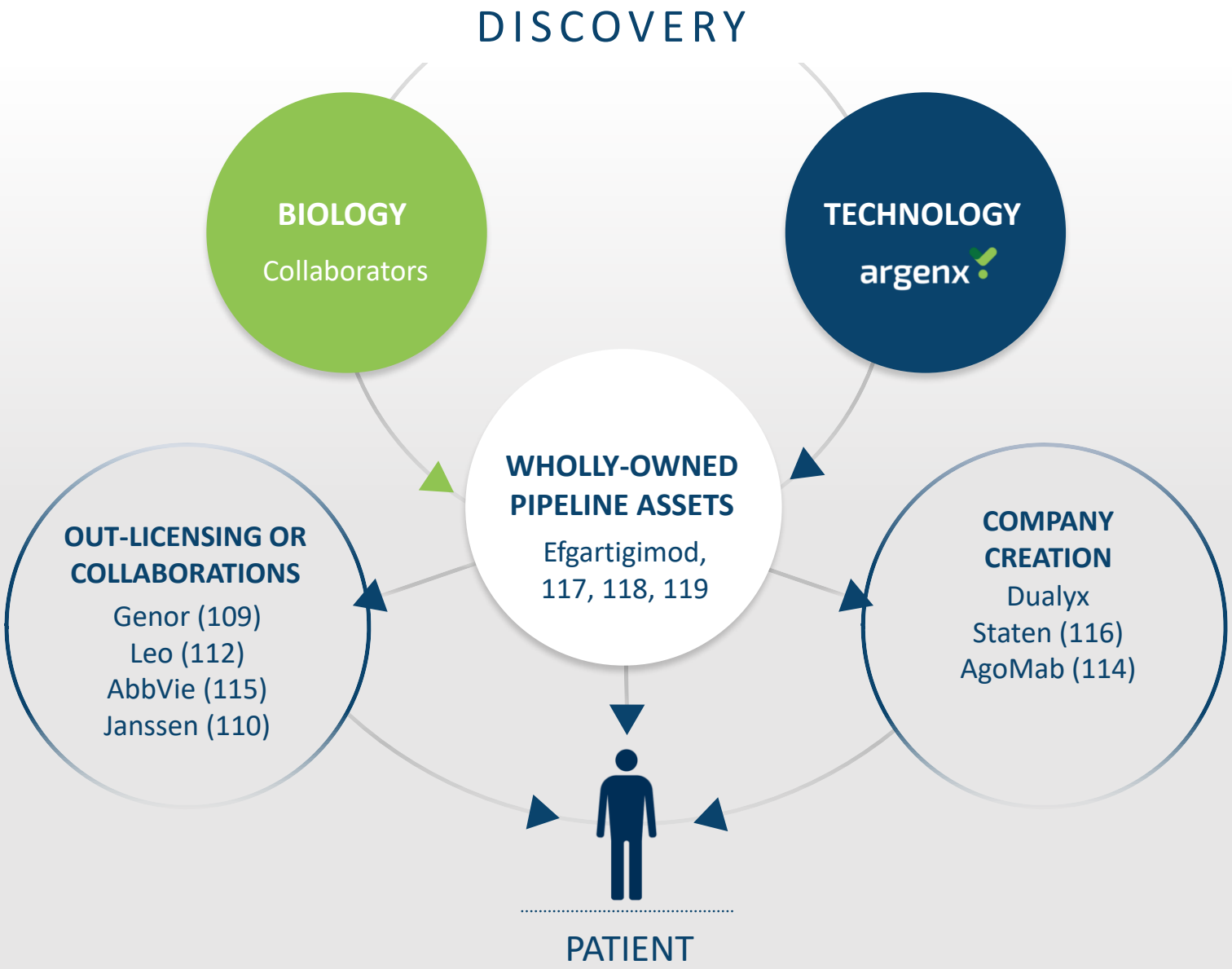
Decision to initiate additional studies will be determined following review of data from ELEVATE

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

Immunology Innovation Program (IIP)

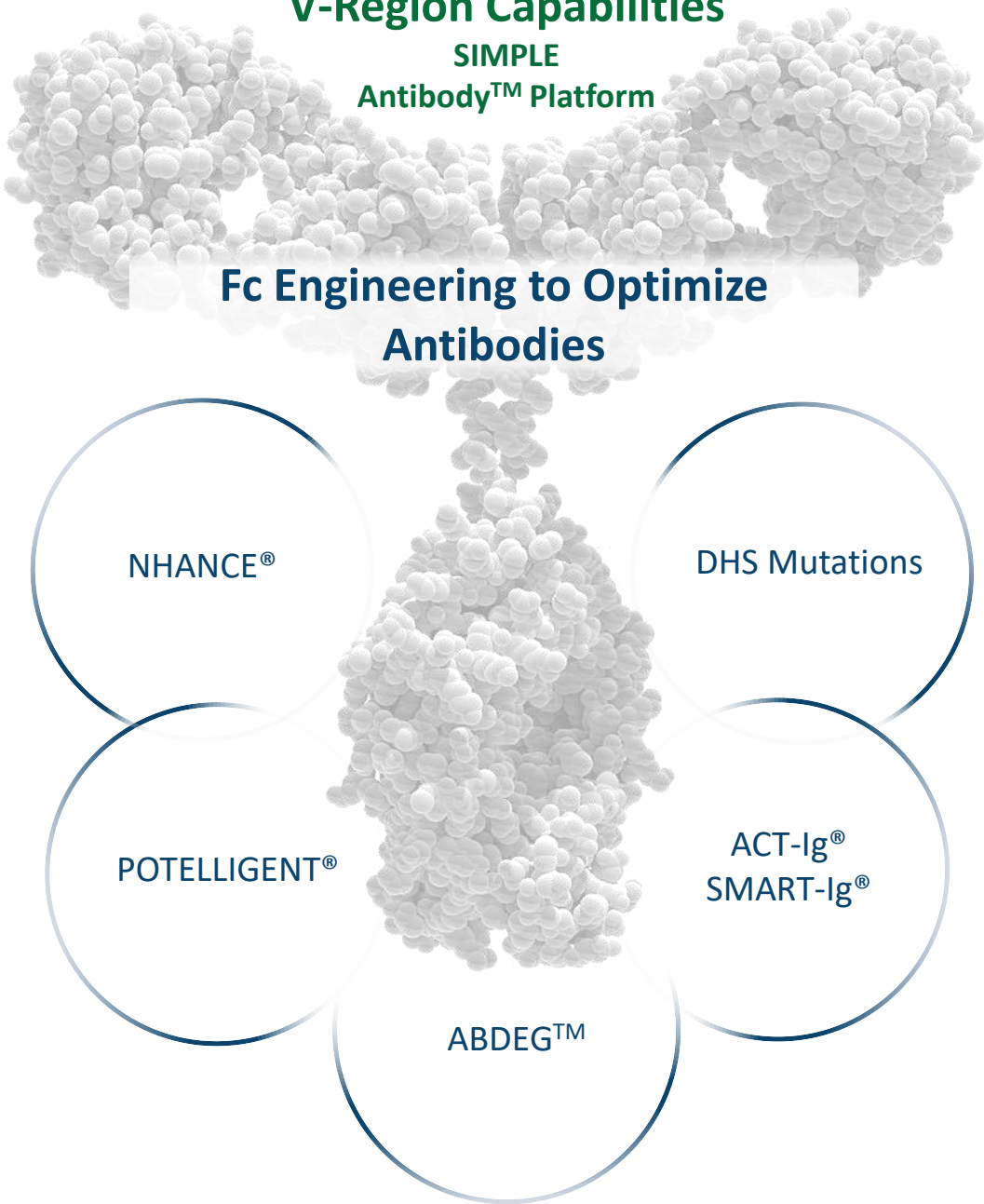
Optimizing the collision of great minds

Core Strategy To Grow Our Pipeline

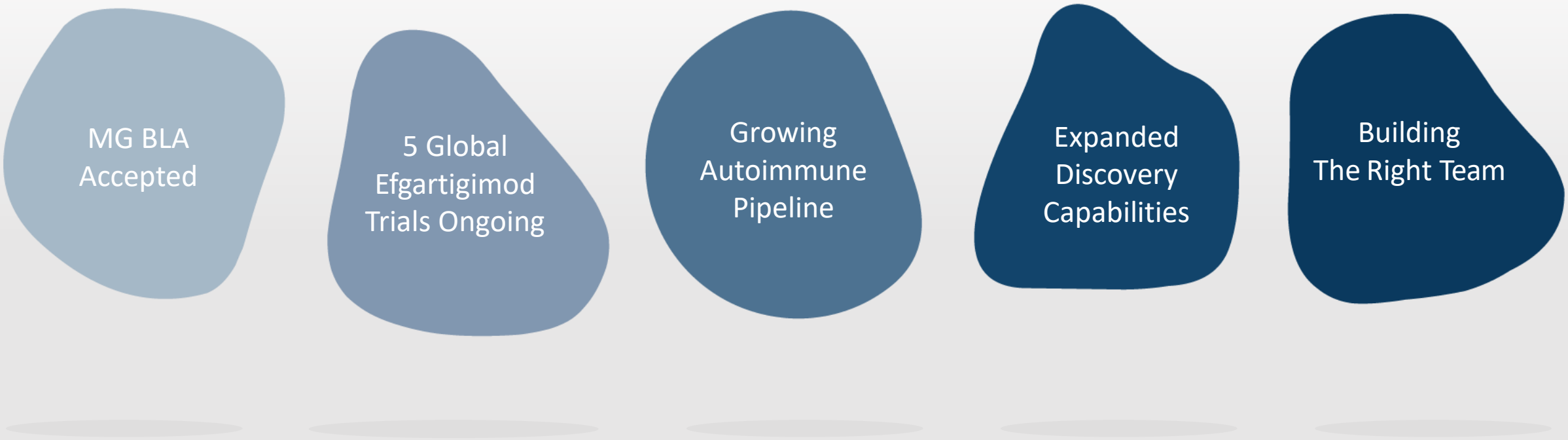


Leading Antibody Discovery and Engineering Toolkit

SC Dosing Optionality
Halozyme — ENHANZE® Technology



Demonstrated Execution Across Business



First Quarter 2021 Financial Results

(in thousands of $ except for shares and EPS)	Three Months Ended March 31, 2021	2020	Variance
Revenue	$ 158,155	$ 21,139	$ 137,017
Other operating income	9,260	4,672	4,588
Total operating income	**167,415**	**25,811**	**141,604**
Research and development expenses	(122,328)	(104,661)	(17,666)
Selling, general and administrative expenses	(56,253)	(27,609)	(28,644)
Total operating expenses	**(178,580)**	**(132,270)**	**(46,310)**
Change in fair value on non-current financial assets	11,152	0	11,152
Operating loss	$ **(13)**	$ **(106,459)**	$ **106,446**
Financial income/(expenses)	(420)	(3,591)	3,171
Exchange gain/(losses)	(28,817)	22,985	(51,802)
Loss before taxes	$ **(29,249)**	$ **(87,064)**	$ **57,815**
Income taxes	(11,184)	(1,200)	(9,984)
Loss for the period and total comprehensive loss	$ **(40,433)**	$ **(88,264)**	$ **47,831**
Weighted average number of shares outstanding	49,946,515	42,786,194	
Basic and diluted profit/(loss) per share (in $)	(0.81)	(2.06)	
Net increase/(decrease) in cash, cash equivalents and current financial assets compared to year-end 2020 and 2019	910,903	(70,318)	
Cash, cash equivalents and current financial assets at the end of the period	2,907,355	1,430,343	

Preparing for a Successful Launch

Listening to and Learning from MG Community

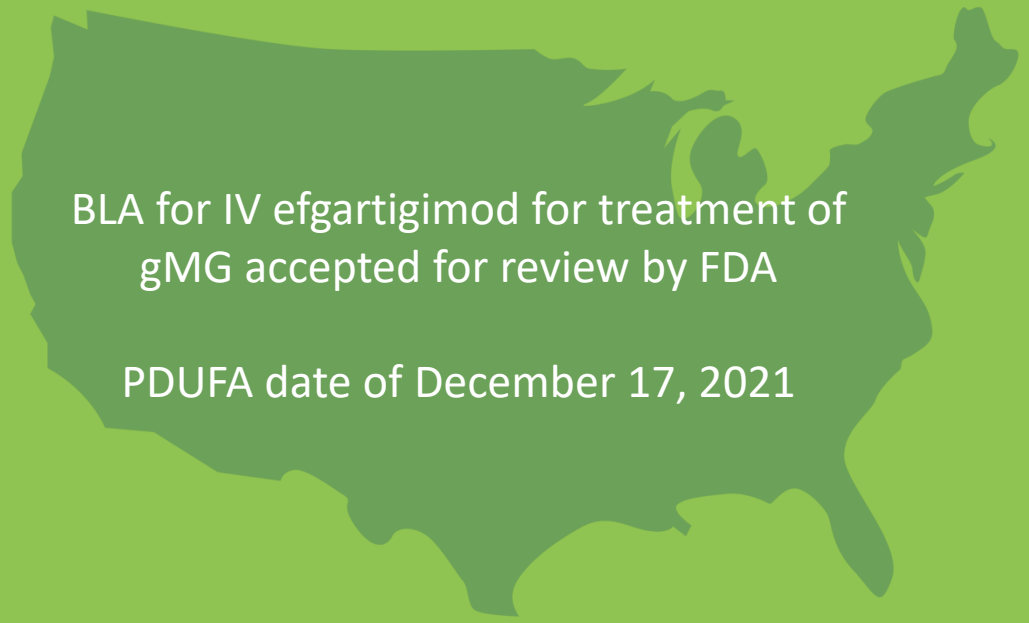
MG United

A Mystery To Me

MyRealWorld™ MG

Efgartigimod Regulatory Update

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 for IV efgartigimod for treatment of gMG accepted for review by PMDA

EU

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

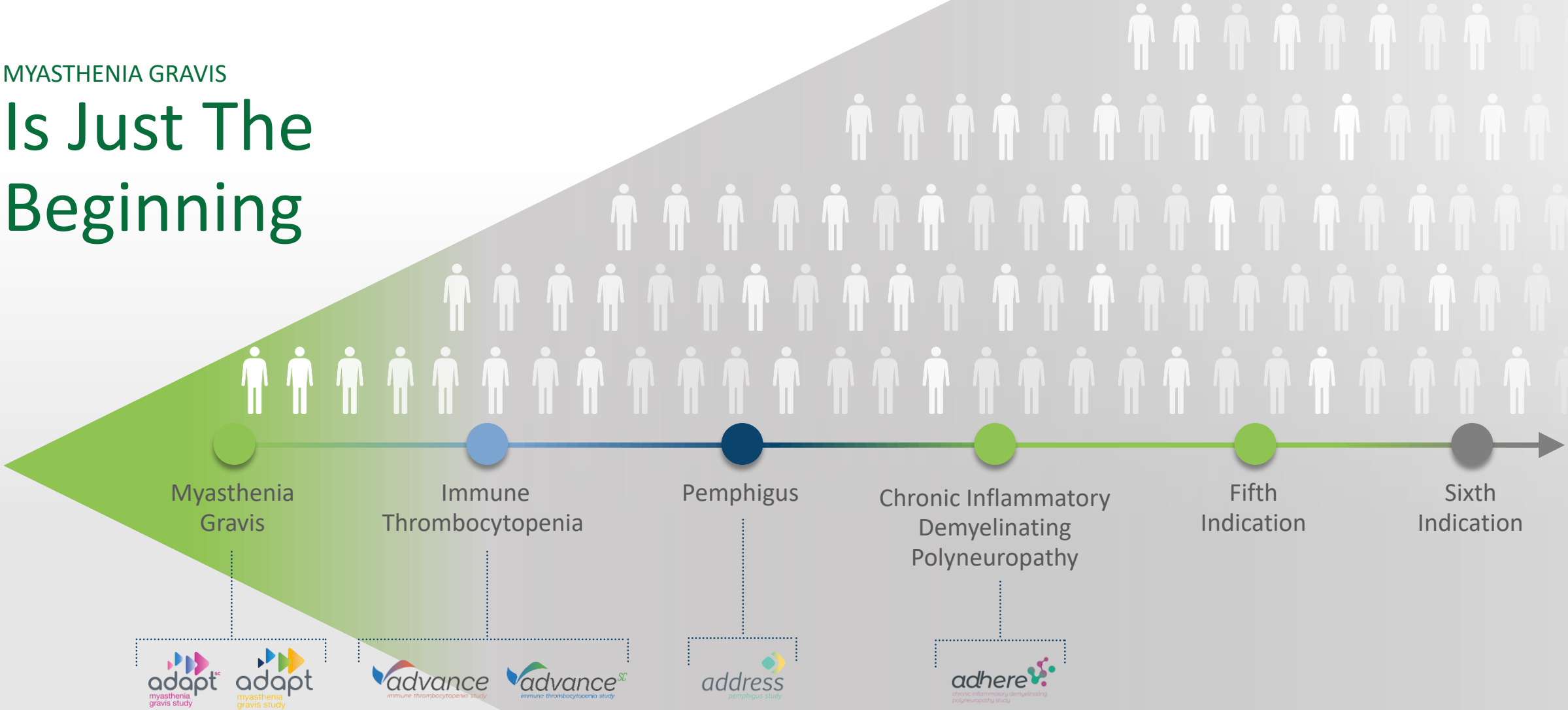
China

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

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

MYASTHENIA GRAVIS

Is Just The Beginning



Key: NEURO HEME SKIN KIDNEY

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Building Tomorrow's Immunology Company

Reach gMG patients with efgartigimod

Global expansion

Strategic Priorities

Advance clinical development in multiple autoimmune indications

Leverage IIP

Rooted in groundbreaking immunology research, growing through collaboration

Together We Discover

Reaching Patients Through
Immunology Innovation

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