

Together We Discover

Reaching Patients Through
Immunology Innovation

Corporate Presentation

OCTOBER 2021



Forward Looking Statements

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Building a Leading Immunology Company

Committed to our
Patients and their
Communities

Enviably
Immunology
Pipeline

Rooted in Science
through our IIP

Global launch preparations underway in U.S., Japan and EU

Efgartigimod proof-of-concept in 4/4 indications with ambition to be in 15 by 2025

Growing neuromuscular, hematology and skin commercial franchises

ARGX-117: second pipeline-in-a-product opportunity

ARGX-119: next asset from Immunology Innovation Program

Strong partnerships in place for additional value creation

Pro-forma cash position of \$2.7B as of 2Q21

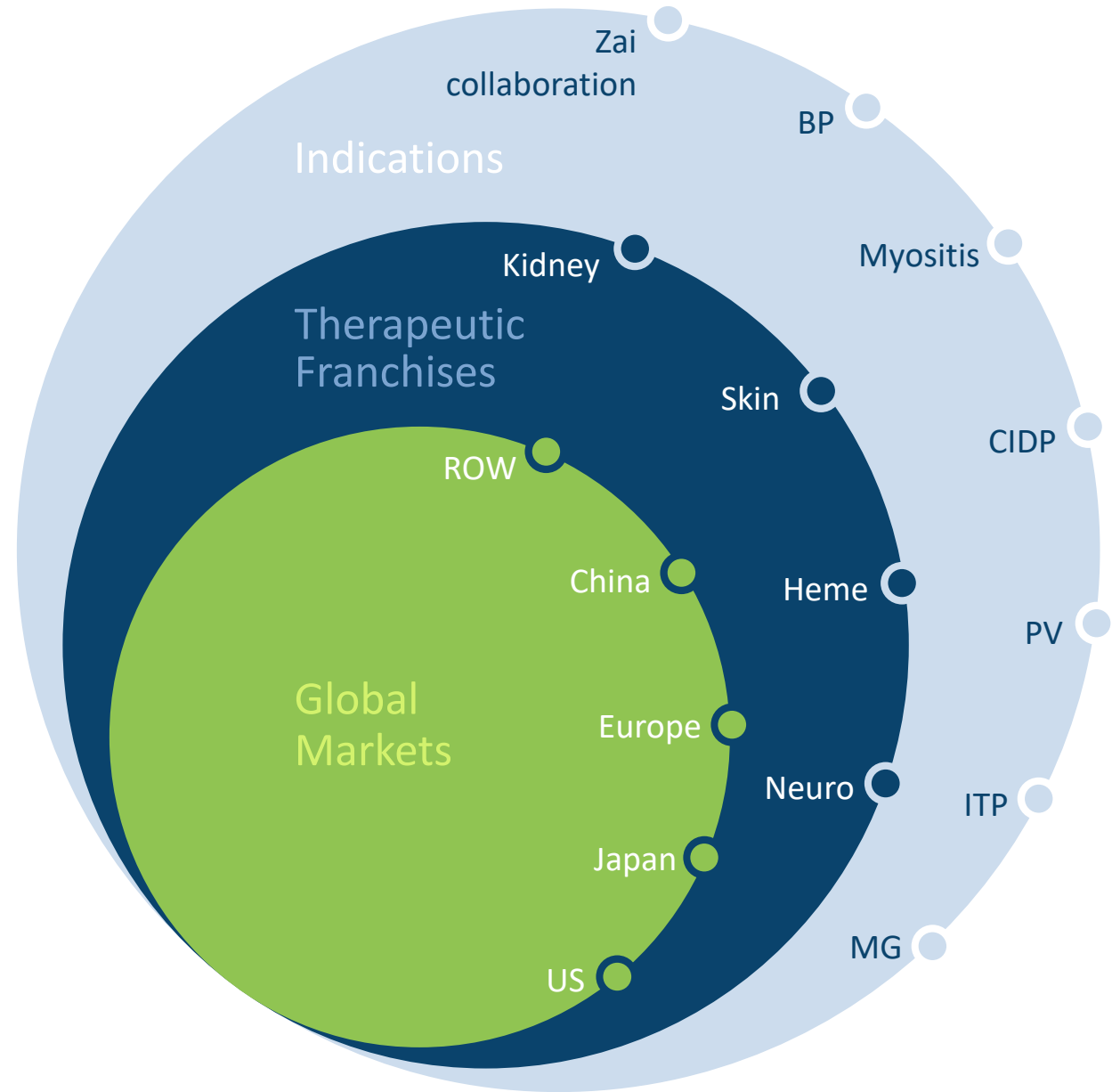
Deep Antibody Pipeline of Differentiated Candidates

Program	Indication			Preclinical	Phase 1	Phase 2	Phase 3	Registration	Update
Efgartigimod	IV	MG							
	SC	MG							Data 1H 2022
	IV	ITP							Data 1H 2022
	SC	ITP							
	SC	PV							
	SC	CIDP							
	SC	Myositis							
	SC	Bullous Pemphigoid (BP)							
ARGX-117	IV + SC	MMN							
	IV + SC	Kidney Diseases							
Cusatuzumab	+ AZA	Newly diag. AML (unfit)	CULMINATE						
	+ AZA + VEN	Newly diag. AML (unfit)	ELEVATE						
ARGX-118		Airway Inflammation							
ARGX-119		Neuromuscular Indications							
ARGX-120		Undisclosed							

Key: ■ NEURO ■ HEME ■ SKIN ■ KIDNEY

Uniquely Positioned For exponential expansion

- efgartigimod indications
- therapeutic franchises
- global markets

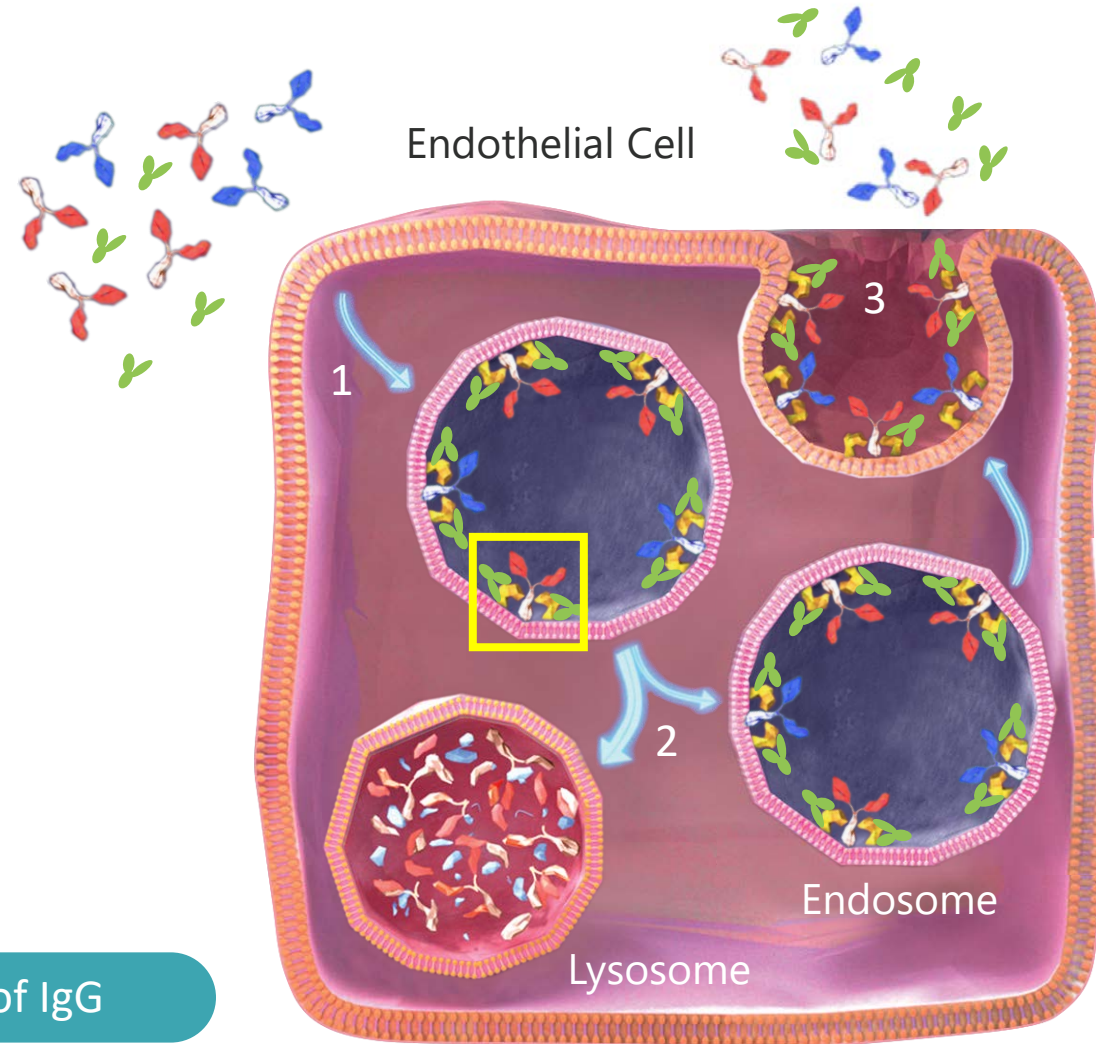


FcRn Plays a Key Role in IgG and Albumin Homeostasis

Studies have shown:

- 1** Circulating antibodies are taken up in the cell via pinocytosis. In the endosome, IgG antibodies bind to FcRn^{1,2}
- 2** Unbound IgGs enter the lysosomal degradation pathway, while FcRn-bound IgGs are rescued from degradation^{1,2}
- 3** The IgG antibodies bound by FcRn are then released back into circulation, thereby extending their half-life^{1,2}

Albumin is recycled by FcRn, independently of IgG



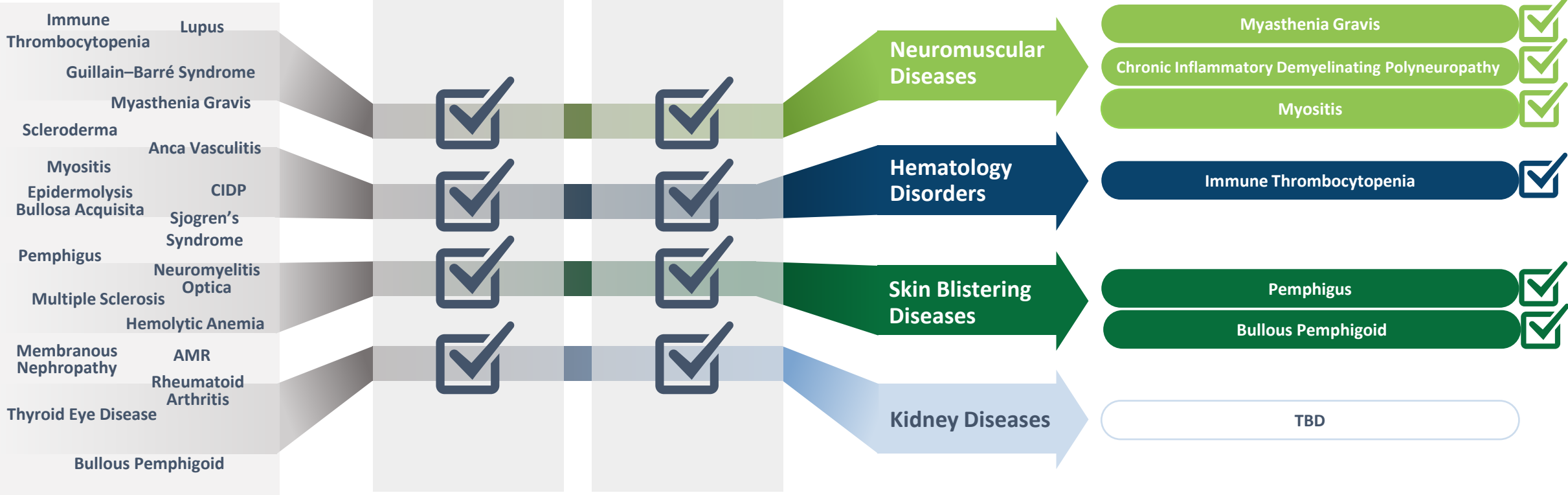
Efgartigimod: Broad Pipeline Opportunity

IgG-mediated Severe Autoimmune Diseases

Solid Biology Rationale

Feasible for Biotech

argenx Franchises & Indications
Proof-of-concept in 4/4 indications; 2/2 in neuromuscular franchise



Efgartigimod: Data Support Favorable Benefit to Risk Ratio

600+

subjects dosed

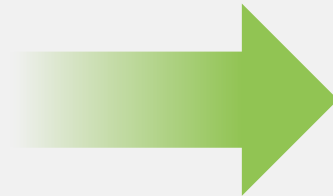
125+

patients on efgartigimod for
over 12 months

100

patients on efgartigimod
for over 18 months

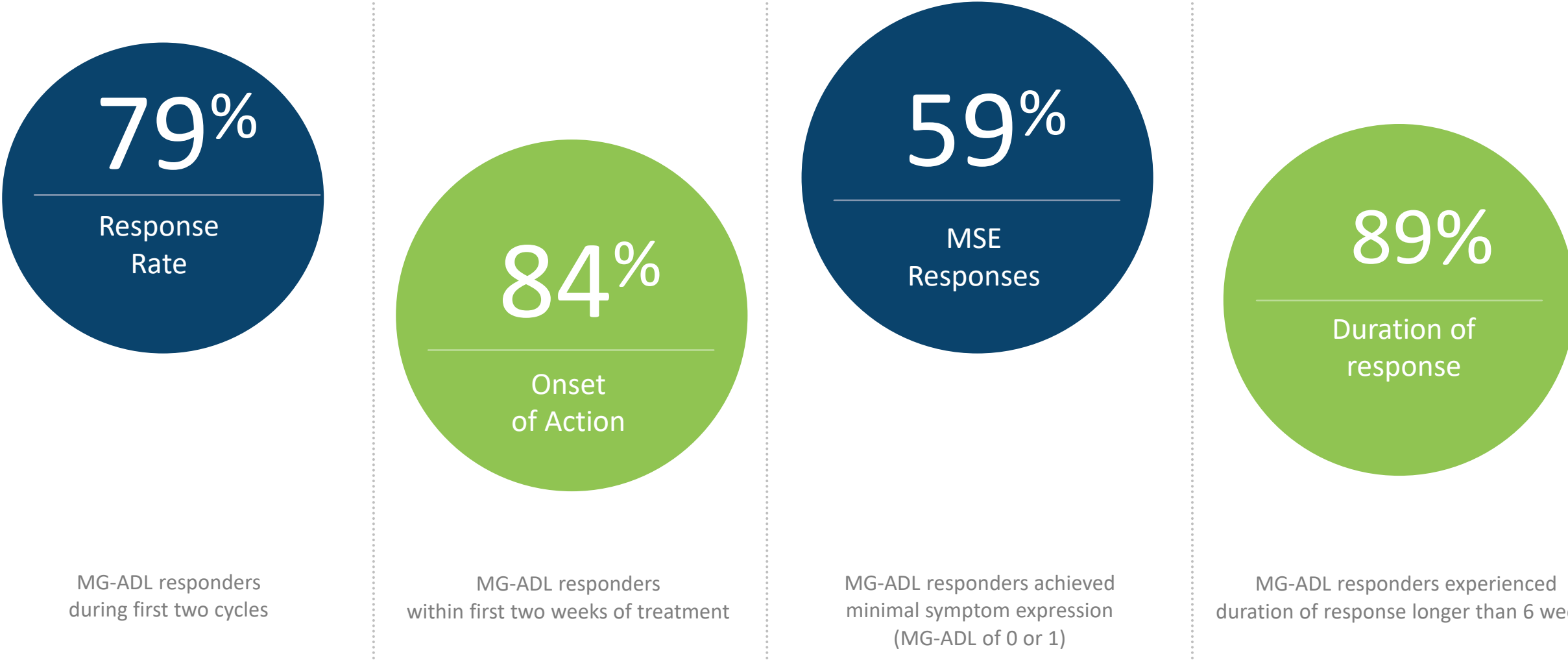
Clinical proof-of-concept in
four indications
(MG, ITP, PV, CIDP)



No evidence of dose-limiting
toxicities in healthy volunteers or
patients across trials

Opportunity to dose efgartigimod
to maximum PD effect

Promising Value Proposition to MG Patients



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*

Efgartigimod is an investigational agent that is not currently approved for use by any regulatory agency as efficacy and safety have not been established.

Preparing for a Successful Launch



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 filed with EMA and validated

China

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

Israel

Partnership agreement with Medison

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

Listening to and Learning from MG Community

MG United



A MYSTERY TO ME



MyRealWorld™ MG

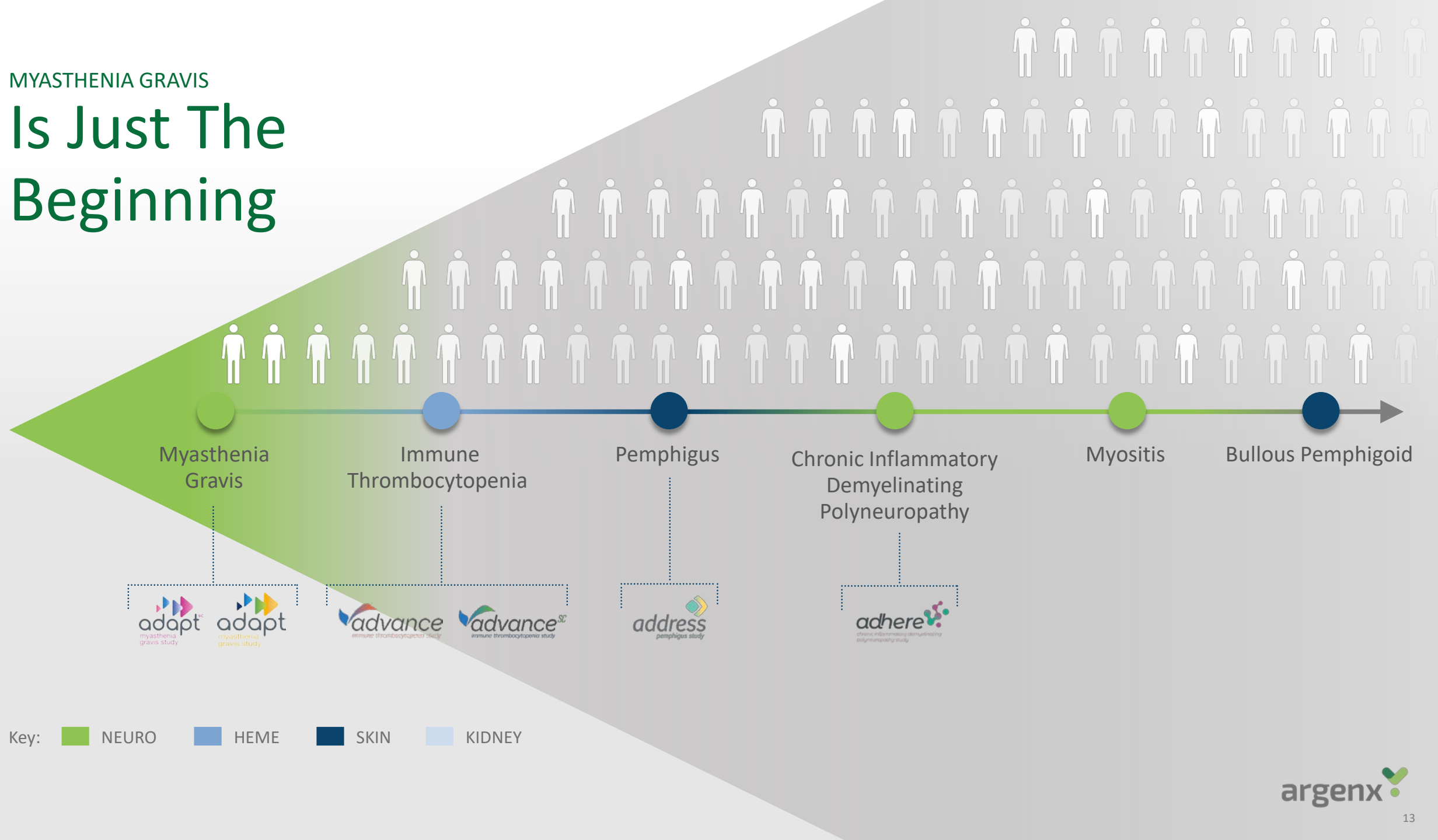


Cooking Together



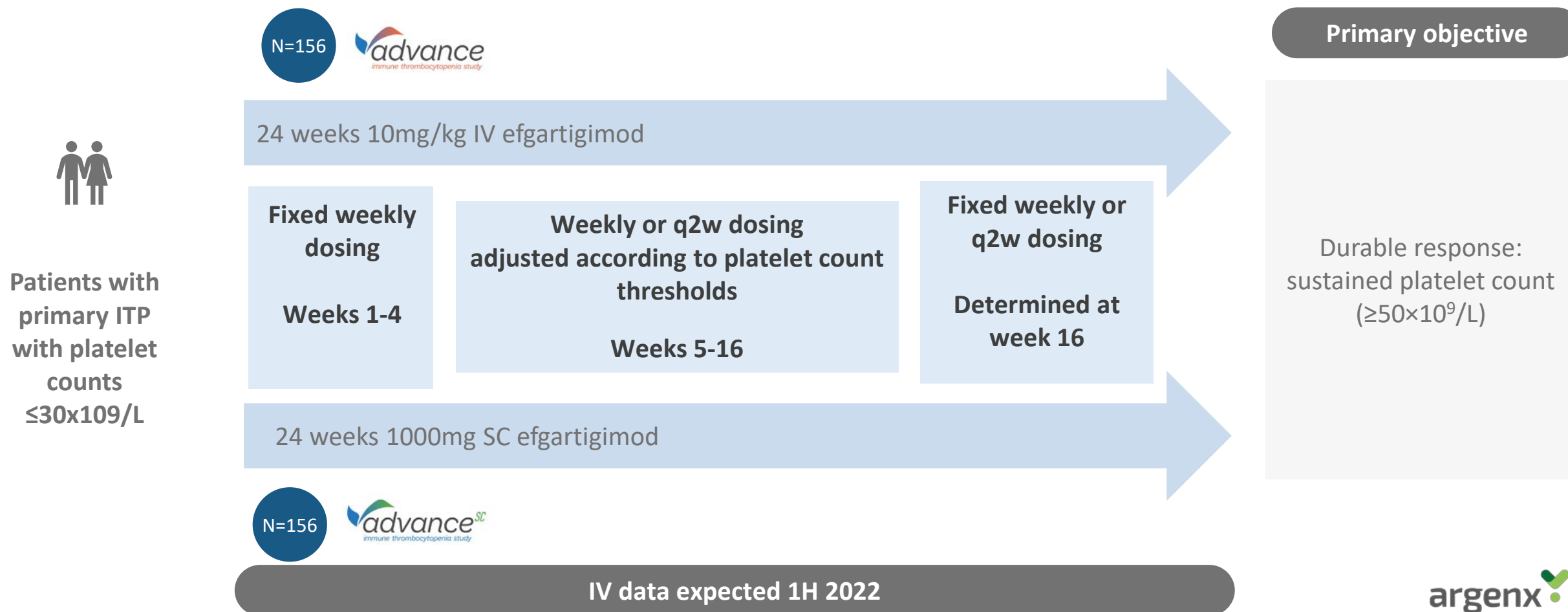
MYASTHENIA GRAVIS

Is Just The Beginning

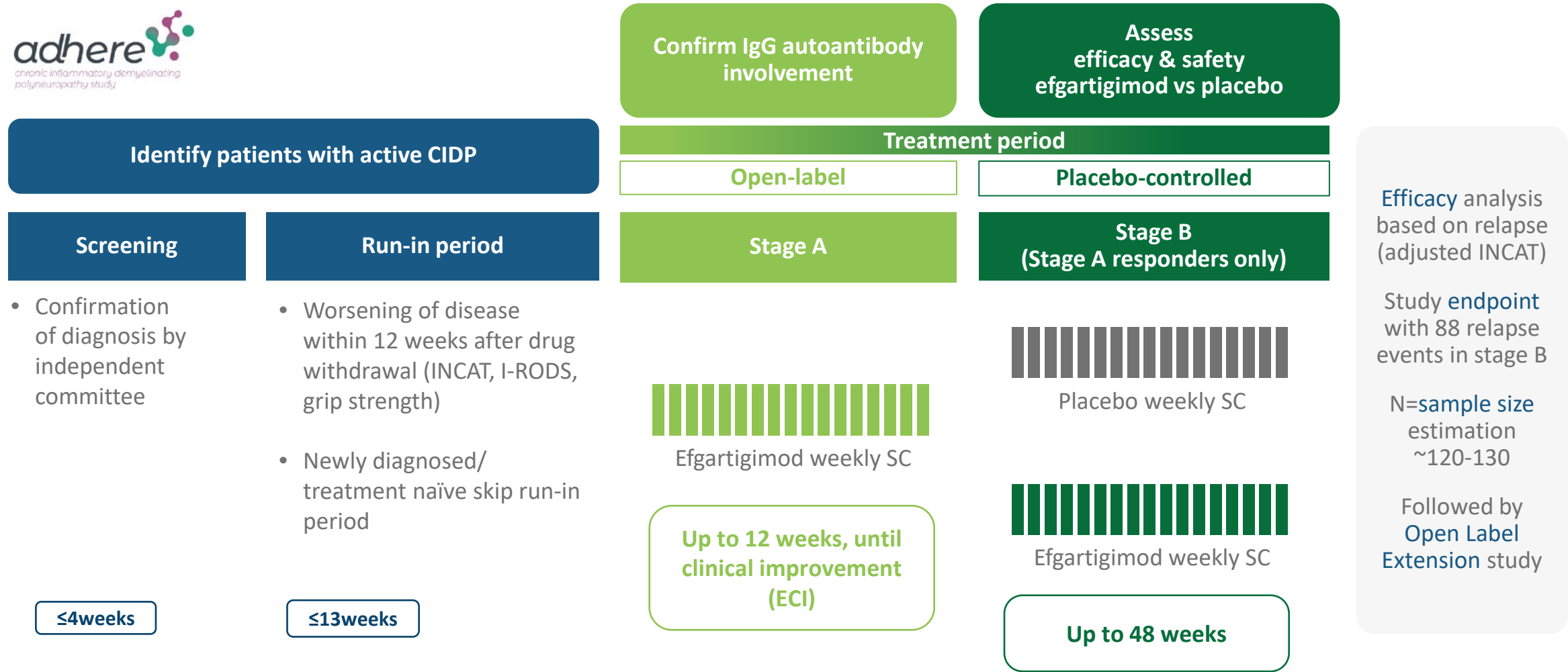


ITP Phase 3 ADVANCE: Two Trials Run in Parallel

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



Chronic Inflammatory Demyelinating Polyneuropathy: Phase 2/3 ADHERE Trial



Phase 3 Trial in Pemphigus: Focus on Fast Onset and Steroid-sparing



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)


Efgartigimod weekly SC


Placebo weekly SC

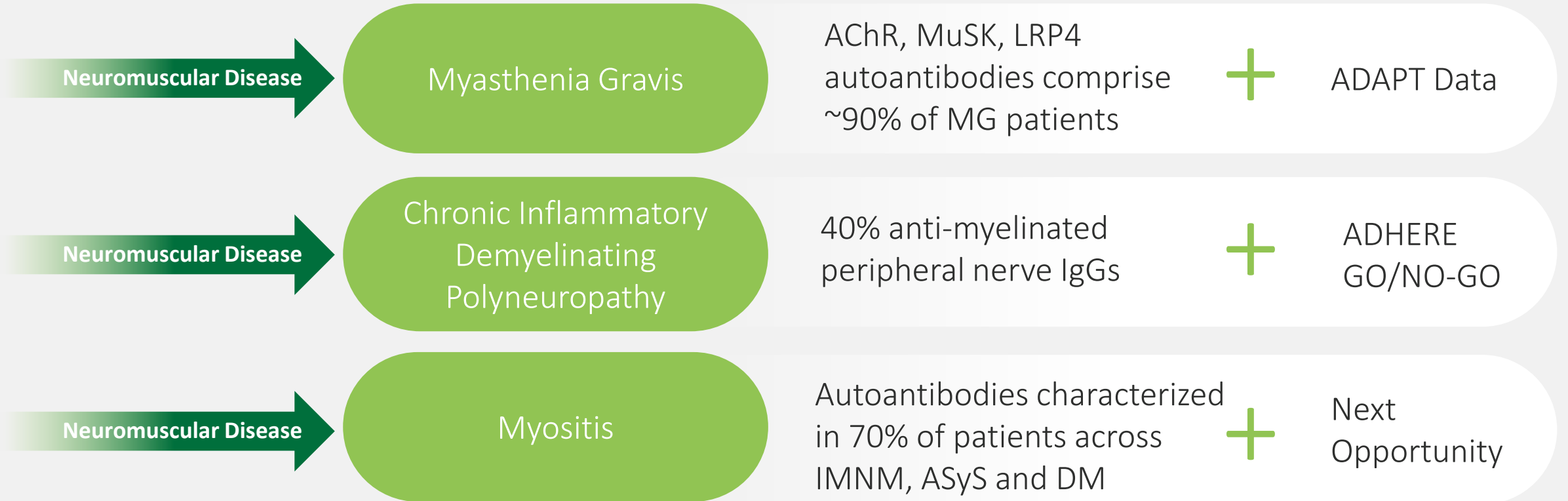
30 weeks

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

Myositis: IgG-Mediated Biology



IND filing by end of 2021 pending interactions with FDA

Bullous Pemphigoid: Expanding the Skin Franchise

Autoimmune
Blistering Diseases

Pemphigus

Pemphigoid

Autoantibody Driven

DSG1 and DSG3

BP180 and BP230

Convincing Rationale

IVIg, PLEX, Immunoadsorption demonstrate role of IgG

Unmet Patient Needs

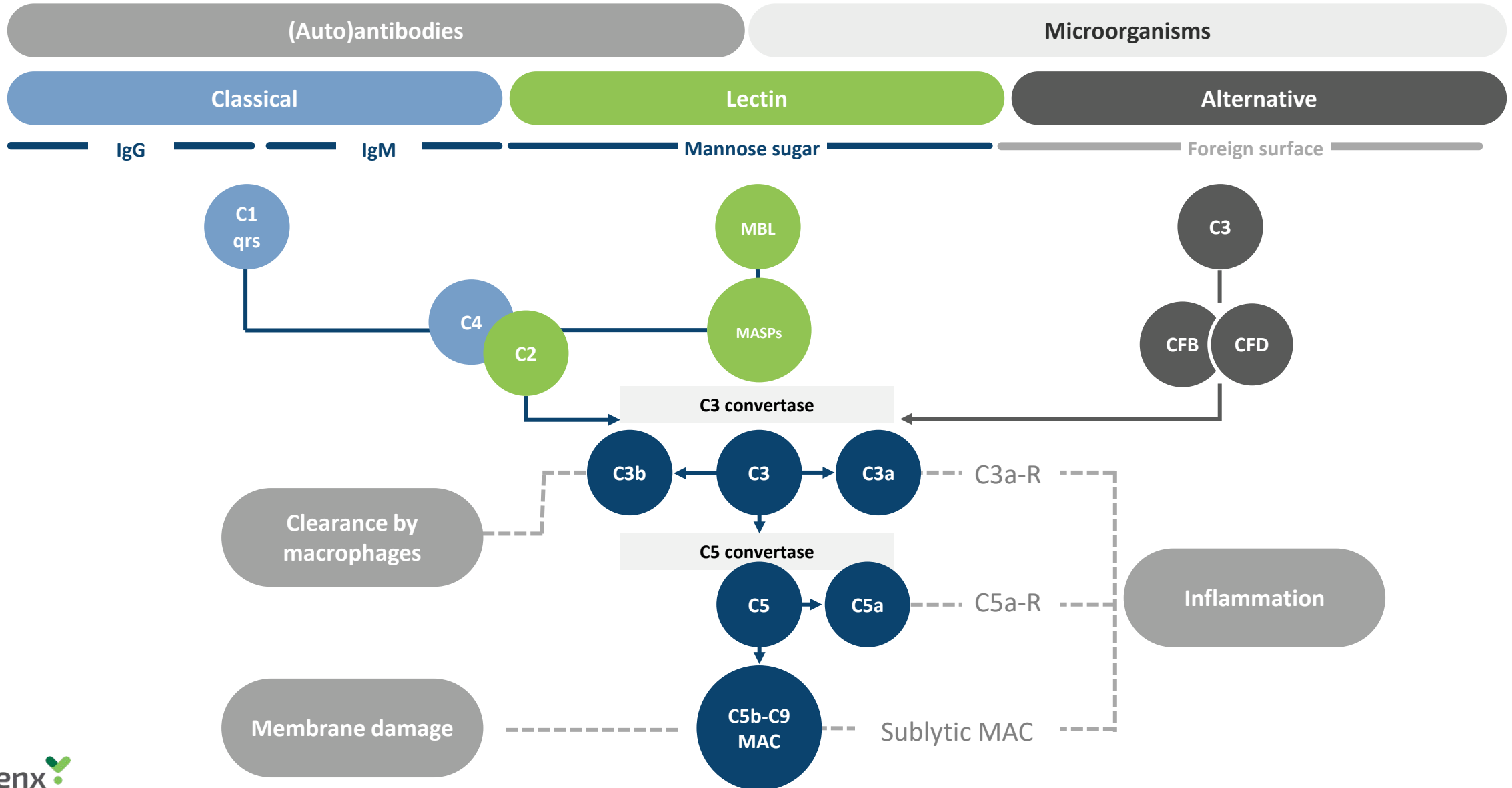
Fast-acting, tolerable therapies; ability to taper corticosteroids

Primary Endpoint

Complete or partial remission off corticosteroids

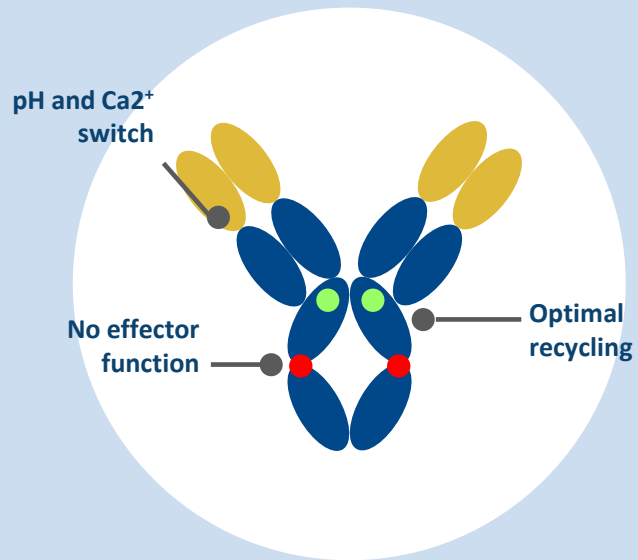
Registrational trial to start by end of year in parallel to ongoing pemphigus trial

ARGX-117 is First in Class Targeting C2

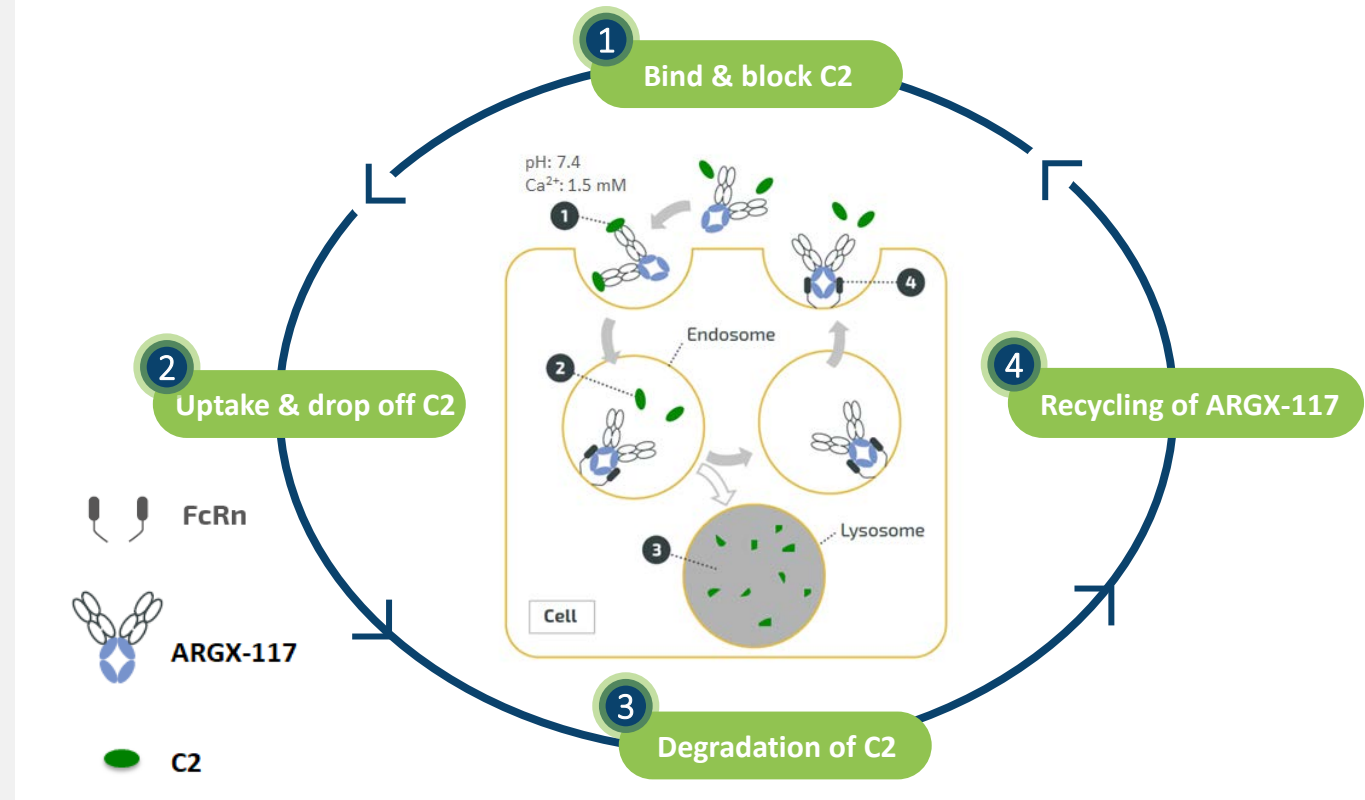


ARGX-117 Blocks and Actively Removes C2 from Circulation

Features of ARGX-117



Sweeping Antibody



Phase 1 Data Support Path Forward and Potential for Individualized Dosing Schedule in Patients

Favorable safety & tolerability profile supports advancing to Phase 2 patient trials

Consistent PK/PD profile across IV and SC dosing that may enable infrequent dosing

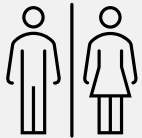
Phase 2 trial in multifocal motor neuropathy (MMN) to start by end of year

Phase 2 Multifocal Motor Neuropathy Trial Design

Patient Population

Trial to enroll approximately 45 MMN patients

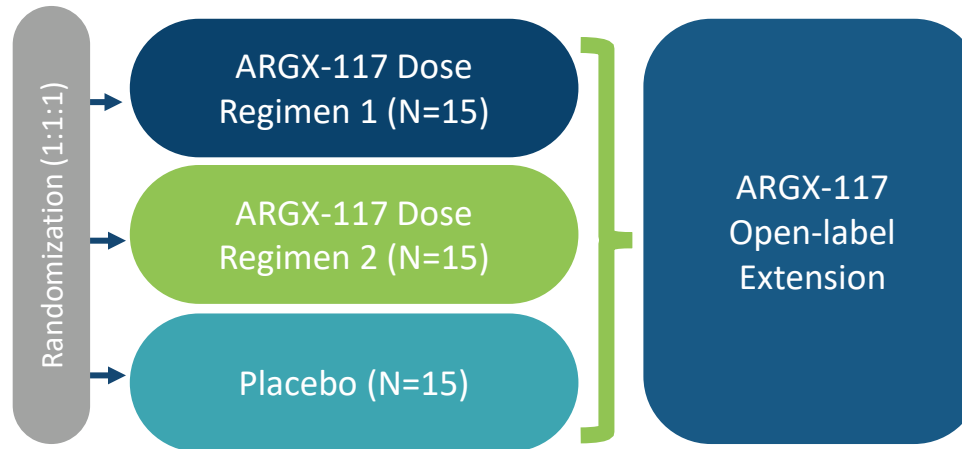
- Probable or definite MMN (per EFNS/PNS 2010)
- Independent adjudication committee
- Stable IVIg regimen
- IVIg treatment dependent



4-10 weeks



Total Duration: 16 weeks



Key Outcome Measures

Safety and tolerability

Time to IVIg re-treatment

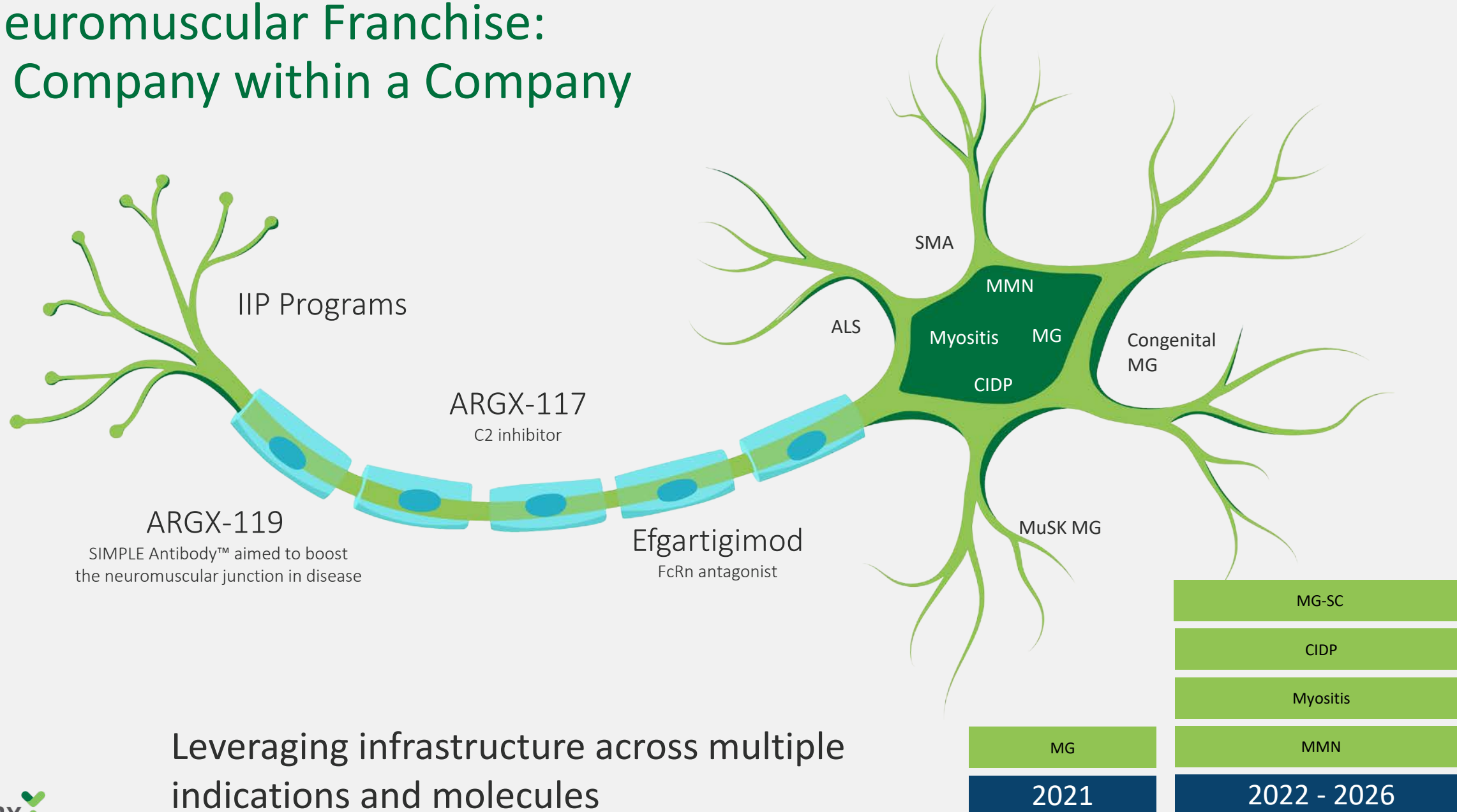
Measures of peripheral muscle strength: grip strength, mMRC sum score, 9-HPT, MMN-RODS

Patient-reported outcomes

PK, PD, biomarkers

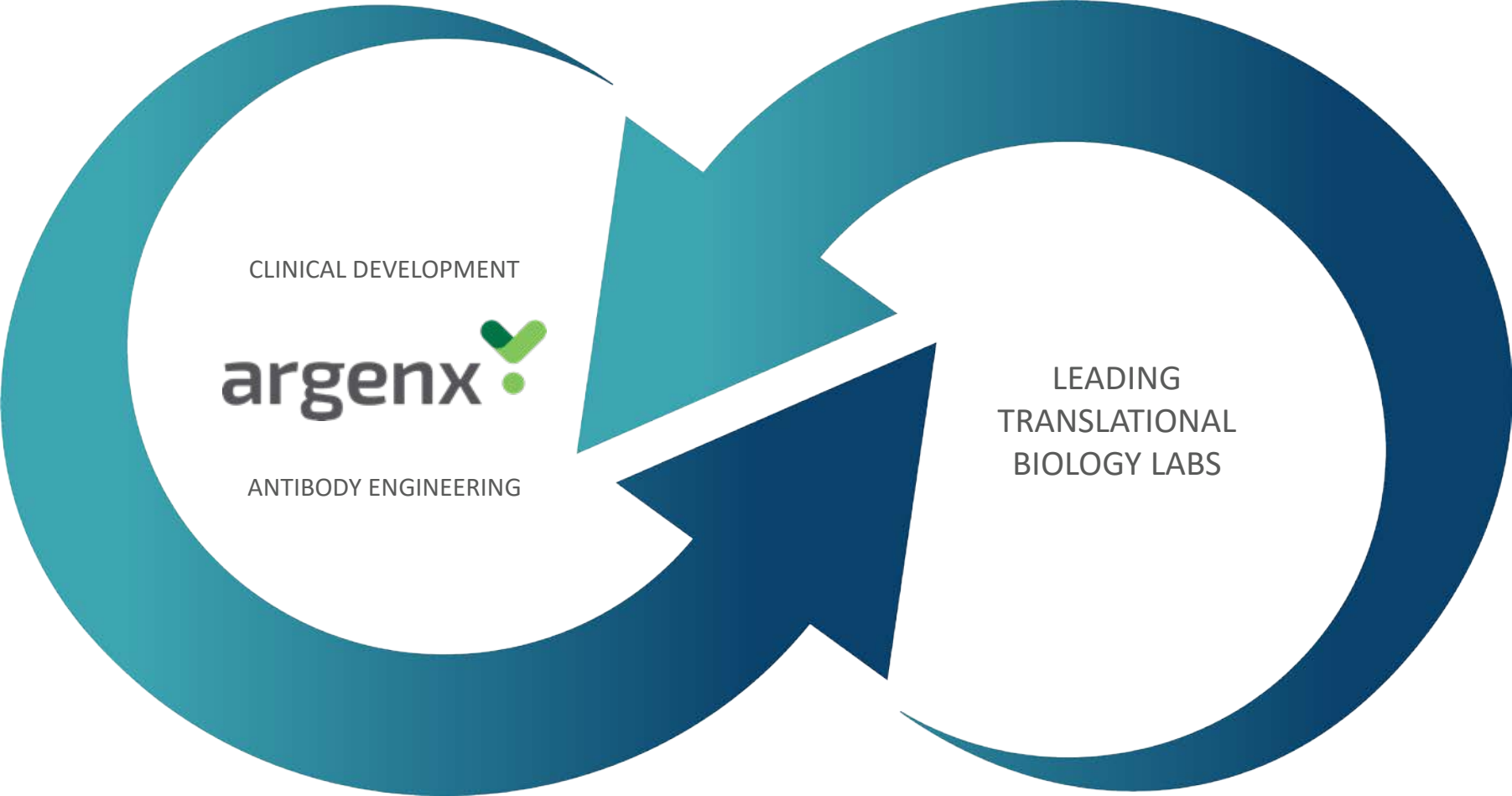
Trial on track to start by end of 2021

Neuromuscular Franchise: A Company within a Company



Leveraging infrastructure across multiple indications and molecules

Our Pipeline Starts with our Immunology Innovation Program



Internal Value Creation

First in Class | Unique Design | Multiple Indications

Efgartigimod

ARGX-117

ARGX-119

External Value Creation

ARGX-118

Staten
(ARGX-116)

AgomAb
(ARGX-114)

Dualyx

Cusatuzumab

Genor
(ARGX-109)

LEO
(ARGX-112)

AbbVie
(ARGX-115)

Building Tomorrow's Immunology Company



Rooted in groundbreaking immunology research, growing through collaboration



Together We Discover

Reaching Patients Through
Immunology Innovation

