Together We Discover

Reaching Patients Through
Immunology Innovation

Corporate Presentation

OCTOBER 2021

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Building a Leading Immunology Company

- 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

We believe the future belongs to those who collaborate best

Deep Antibody Pipeline of Differentiated Candidates

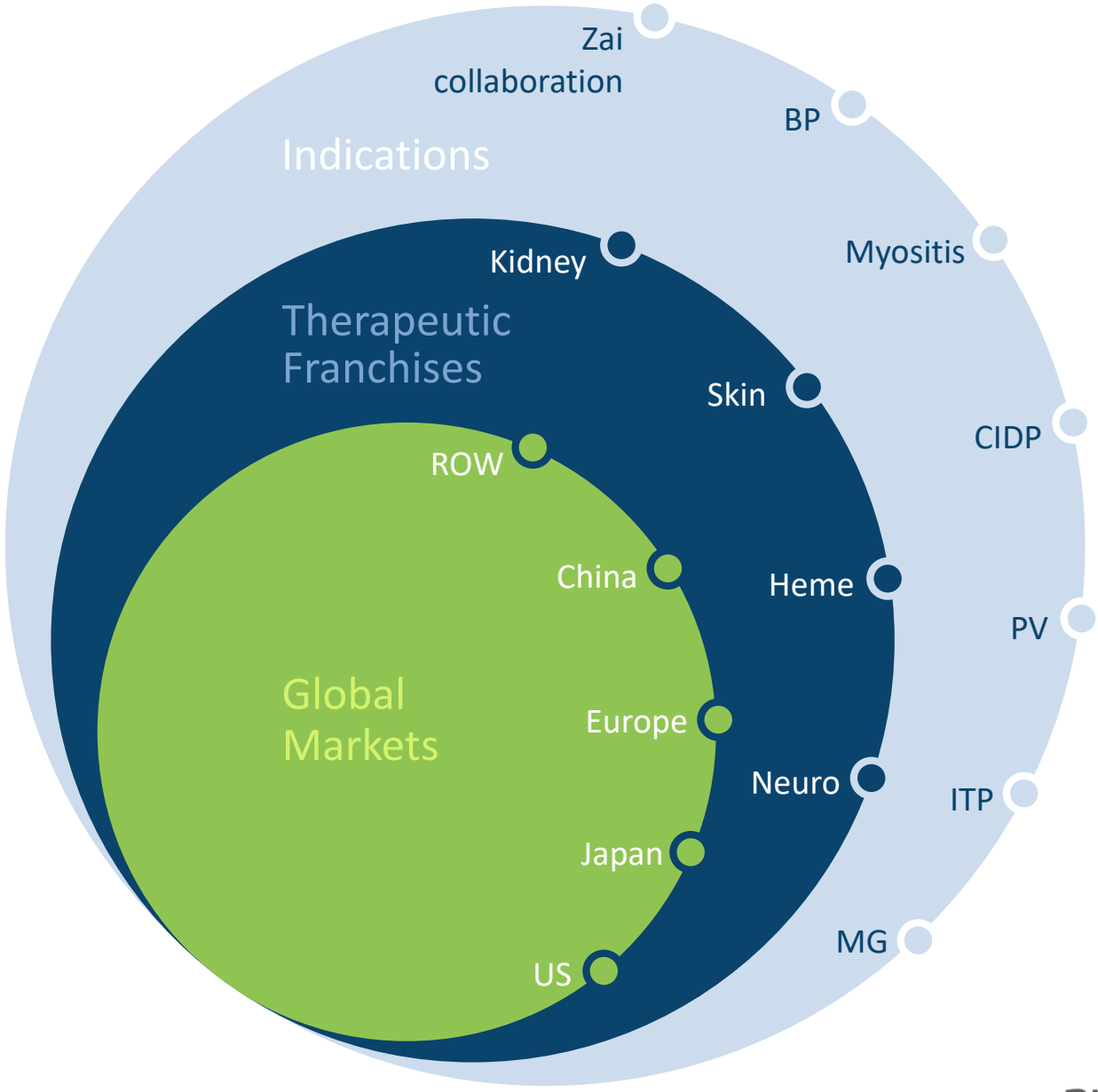
Program		Indication		Preclinical	Phase 1	Phase 2	Phase 3	Registration	Update
Efgartigimod	IV	MG	adapt						
	SC	MG	adapt						Data 1H 2022
	IV	ITP	advance						Data 1H 2022
	SC	ITP	advance						
	SC	PV	address						
	SC	CIDP	adhere						
	SC	Myositis							
	SC	Bullous Pemphigoid (BP)							
ARGX-117	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

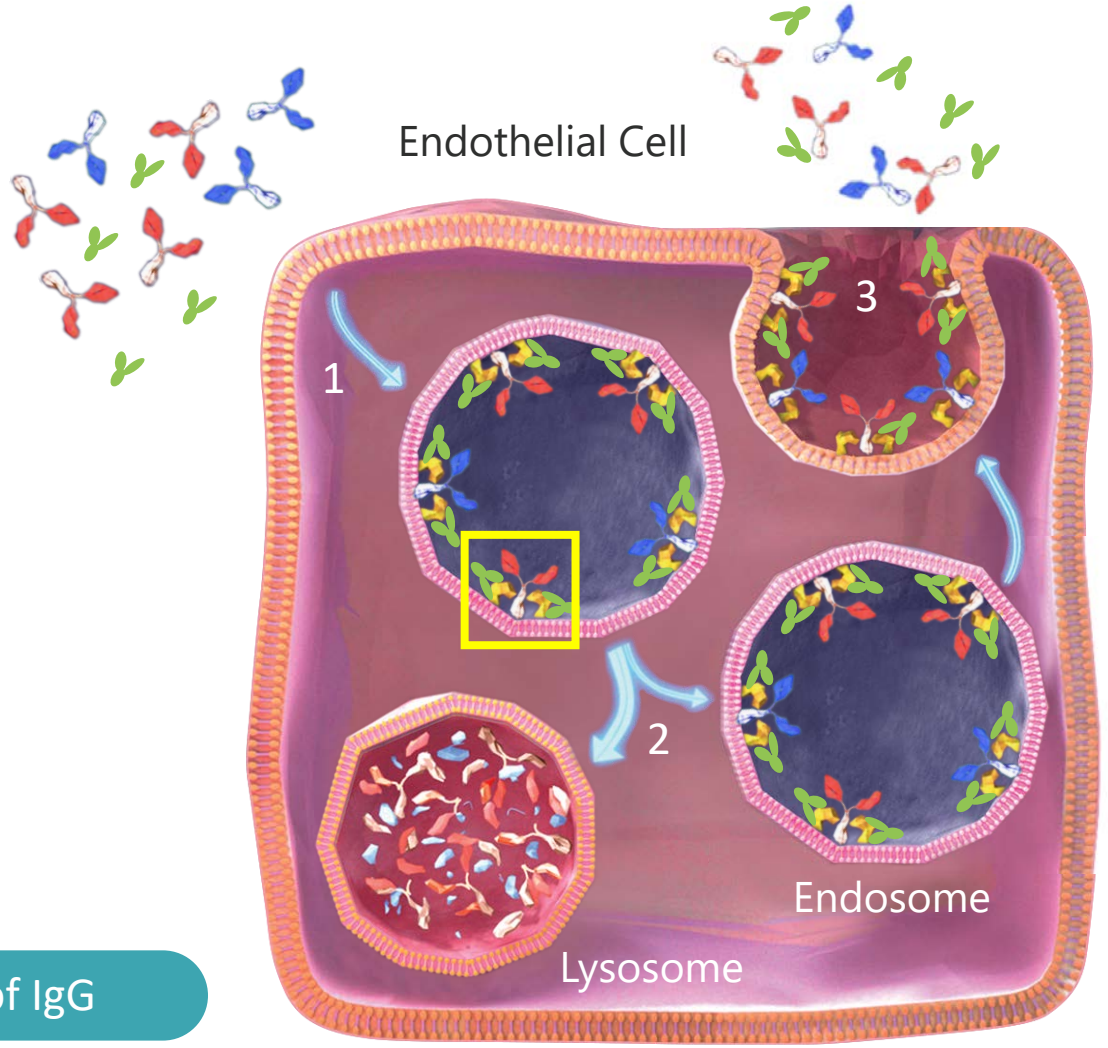
Studies have shown:

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



FcRn, neonatal fragment crystallizable receptor; IgG, immunoglobulin G.
(1) Sesarman et al., Cell Mol Life Sci. 2010; (2) Habib et al., Supp Neuro Review. 2020.

Efgartigimod: Broad Pipeline Opportunity

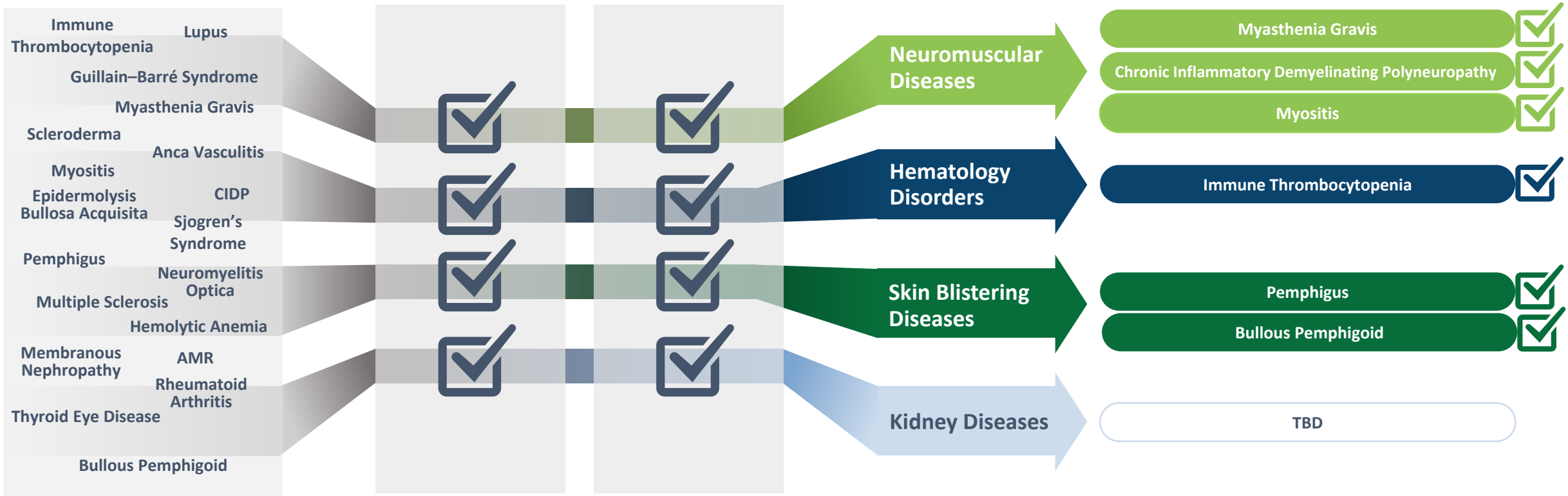
IgG-mediated Severe Autoimmune Diseases

Solid Biology Rationale

Feasible for Biotech

argenx Franchises & Indications

Proof-of-concept in 4/4 indications; 2/2 in neuromuscular franchise



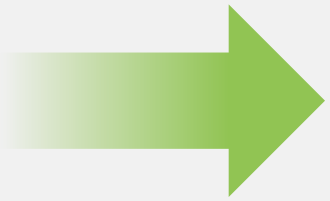
Efgartigimod: Data Support Favorable Benefit to Risk Ratio

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

79%

Response Rate

MG-ADL responders during first two cycles

84%

Onset of Action

MG-ADL responders within first two weeks of treatment

60%

MSE Responses

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

89%

Duration of response

MG-ADL responders experienced duration of response longer than 6 weeks

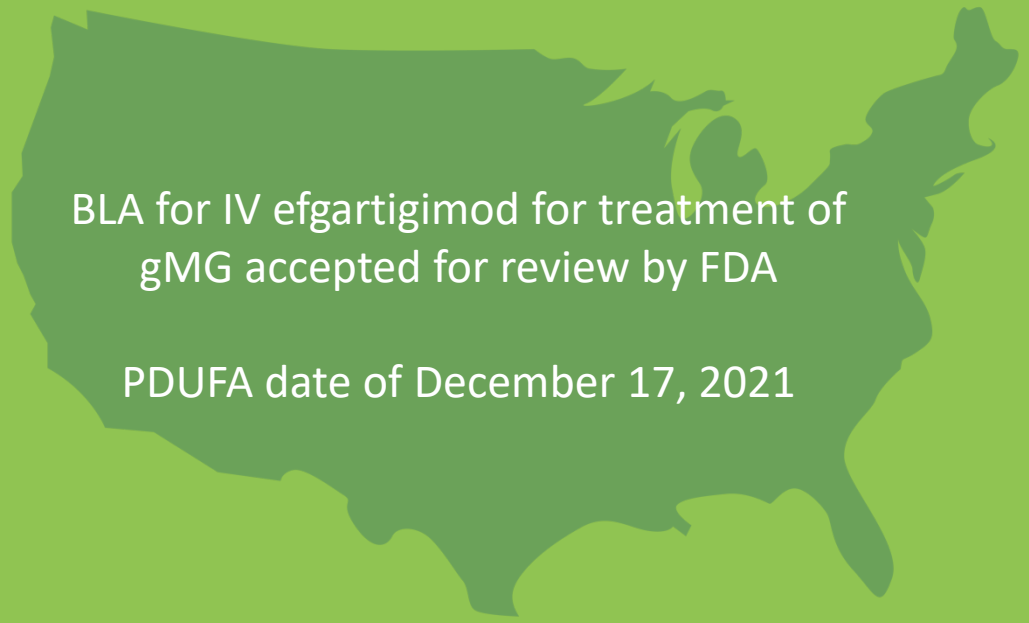
Primary endpoint: MG-ADL responder ≥2-point improvement for at least four consecutive weeks during the first cycle*
First secondary endpoint: QMG responder ≥3-point improvement for at least four consecutive weeks during the first cycle*

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

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

Listening to and Learning from MG Community

MG United

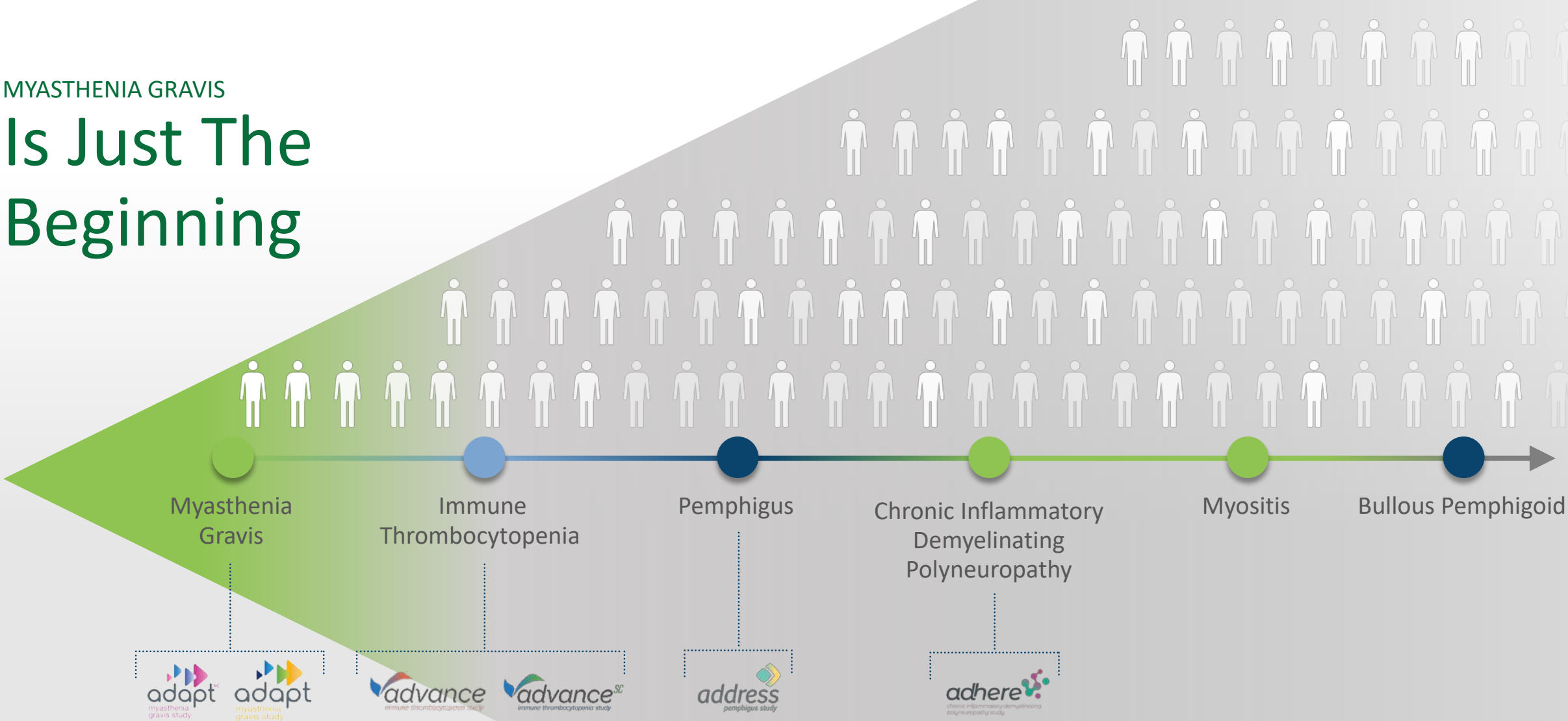
A Mystery To Me

MyRealWorld™ MG

Cooking Together

MYASTHENIA GRAVIS

Is Just The Beginning



Key: NEURO HEME SKIN KIDNEY

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

IV data expected 1H 2022

Primary objective

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

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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

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

address
pemphigus study

Screening

Pemphigus vulgaris (PV) and foliaceus (PF)

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

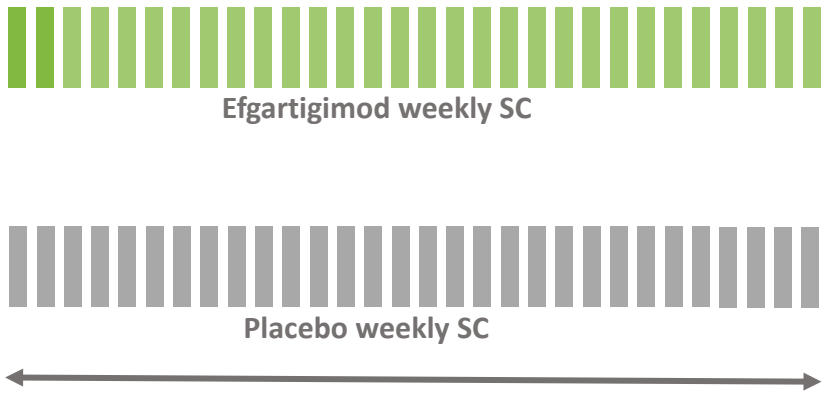
Newly Diagnosed and Relapsing

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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Myositis: IgG-Mediated Biology

Neuromuscular Disease	Myasthenia Gravis	AChR, MuSK, LRP4 autoantibodies comprise ~90% of MG patients	+	ADAPT Data
Neuromuscular Disease	Chronic Inflammatory Demyelinating Polyneuropathy	40% anti-myelinated peripheral nerve IgGs	+	ADHERE GO/NO-GO
Neuromuscular Disease	Myositis	Autoantibodies characterized in 70% of patients across IMNM, ASyS and DM	+	Next Opportunity

IND filing by end of 2021 pending interactions with FDA

IMNM: Immune-Mediated Necrotizing Myopathy ASyS: Anti-Synthetase Syndrome DM: Dermatomyositis

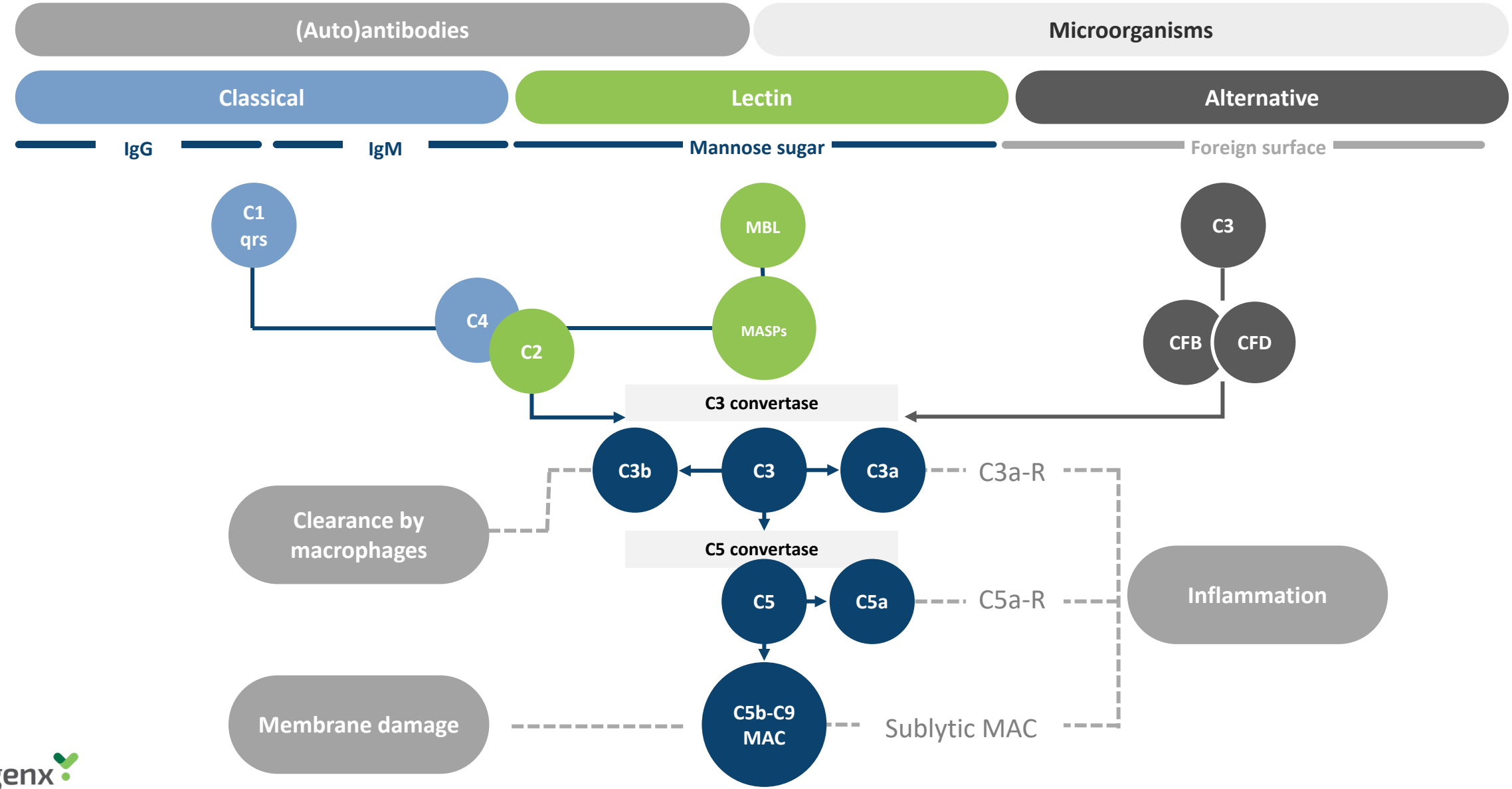
Gilhus et al., Nature Rev/Disease Primers. 2019; Gilhus et al., NEJM, 2016; Querol et al., Nat Rev Neurol. 2017; Schmidt, Journal Neuromusc Diseases. 2018; Aquilar-Vazquez et al., Frontiers in Immunology. 2021; McHugh, Managing Myositis. 2019

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	

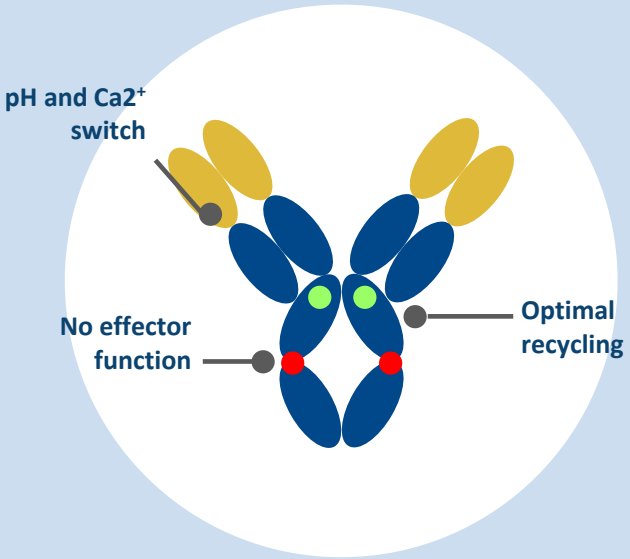
Registrational trial to start by end of year in parallel to ongoing pemphigus trial

ARGX-117 is First in Class Targeting C2

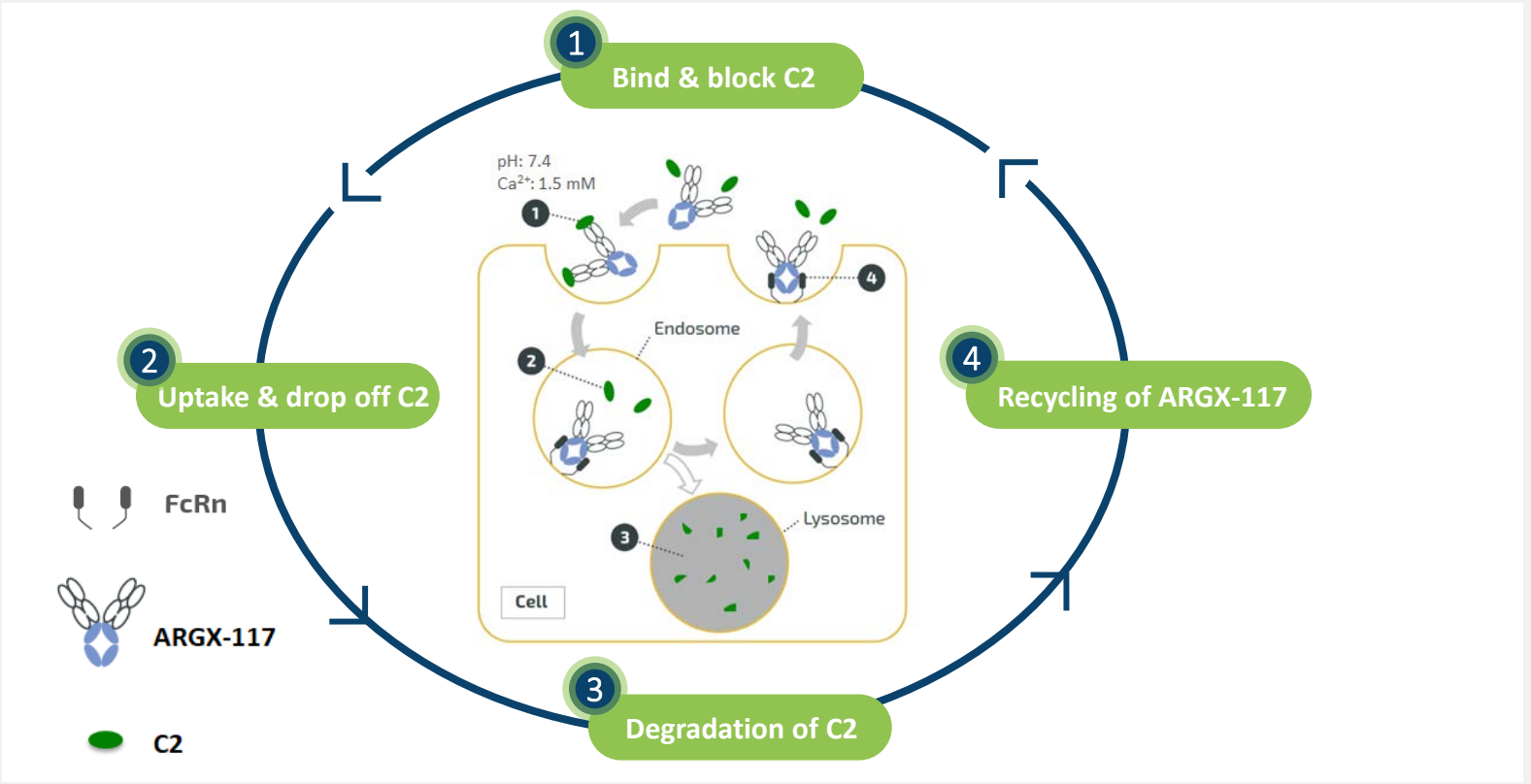


ARGX-117 Blocks and Actively Removes C2 from Circulation

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

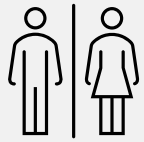
Phase 2 trial in multifocal motor neuropathy (MMN) to start by end of year

Phase 2 Multifocal Motor Neuropathy Trial Design

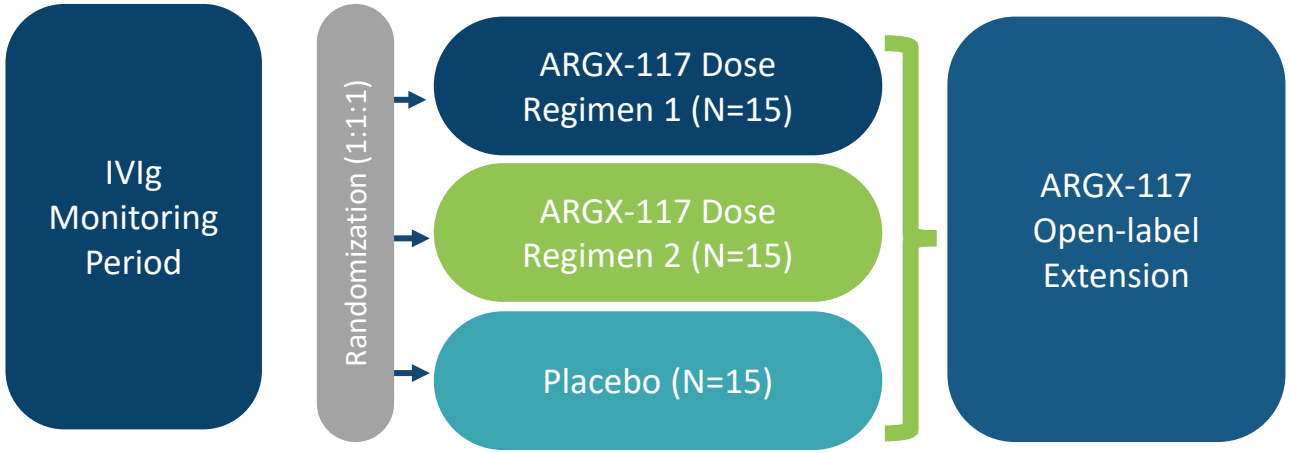
Patient Population

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

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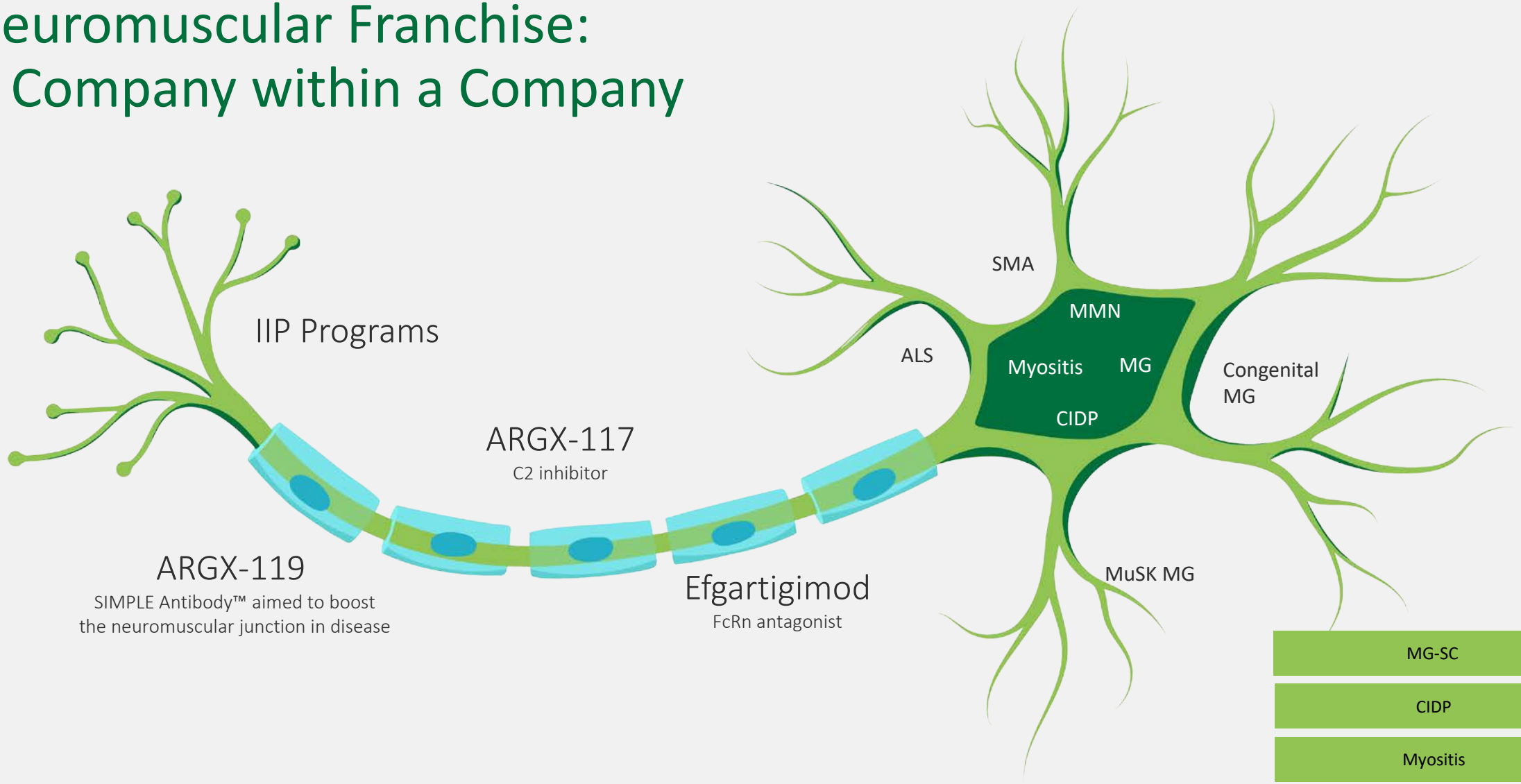
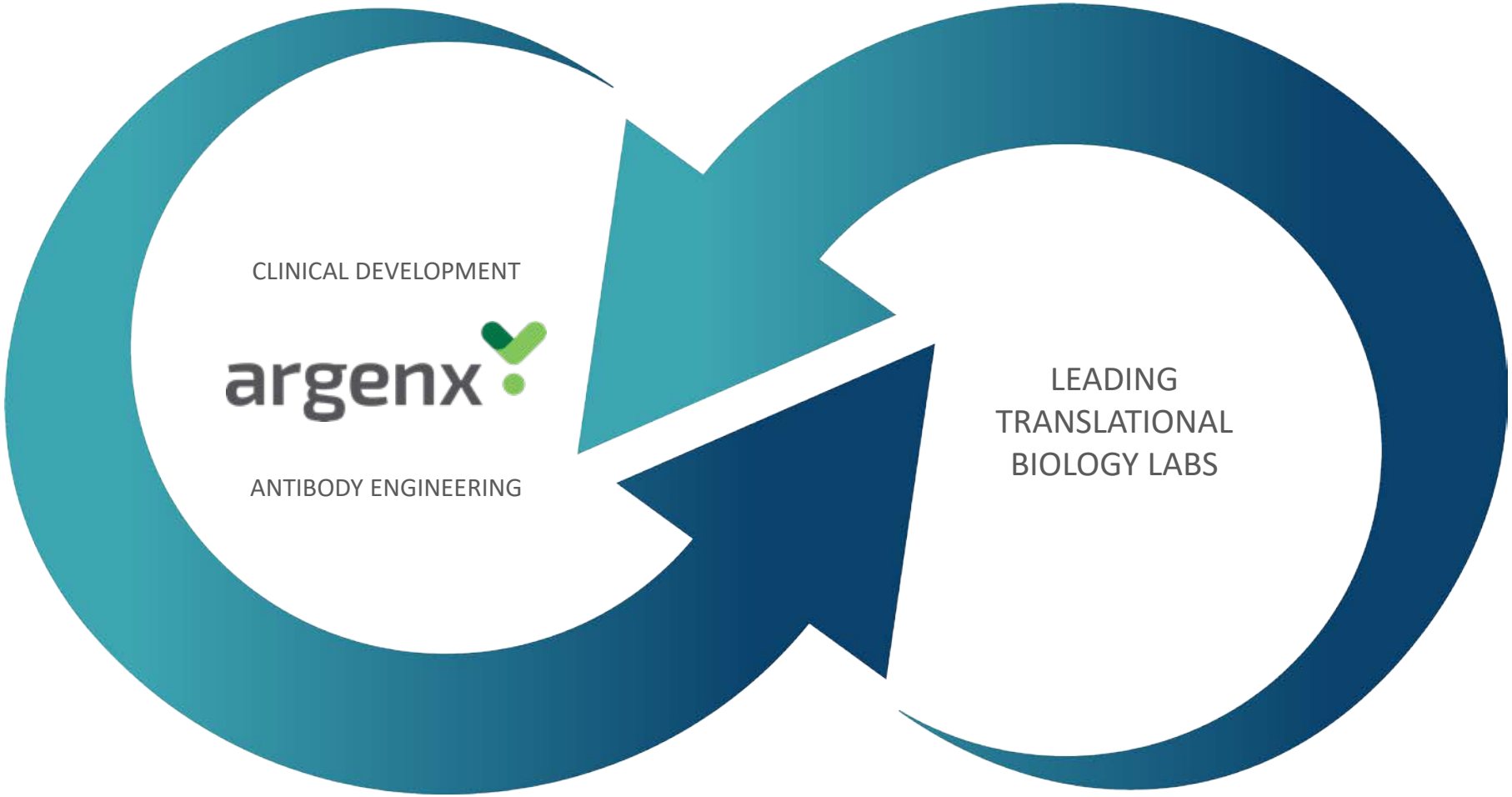
Neuromuscular Franchise: A Company within a Company



Leveraging infrastructure across multiple indications and molecules

MG	MG-SC
	CIDP
	Myositis
	MMN
2021	2022 - 2026

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

Strategic Priorities

- Reach gMG patients with efgartigimod
- Global expansion
- 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