

Together We Discover

Reaching Patients Through
Immunology Innovation

Corporate Presentation

APRIL 2022



Forward Looking Statements

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Our Plan to Build a Leading Immunology Company

Committed to
our Patients and
their
Communities

Enviably
Immunology
Pipeline

Rooted in
Science through
our IIP

Focused on global, expanding autoimmune market

Reaching patients globally

Vibrant neuromuscular, hematology, skin and kidney franchises

Efgartigimod in 15 indications (commercial and development)

ARGX-117 in multiple late-stage trials

Proof-of-concept demonstrated with ARGX-119

New asset each year from IIP

argenx 2025

We believe the future belongs to those who collaborate best

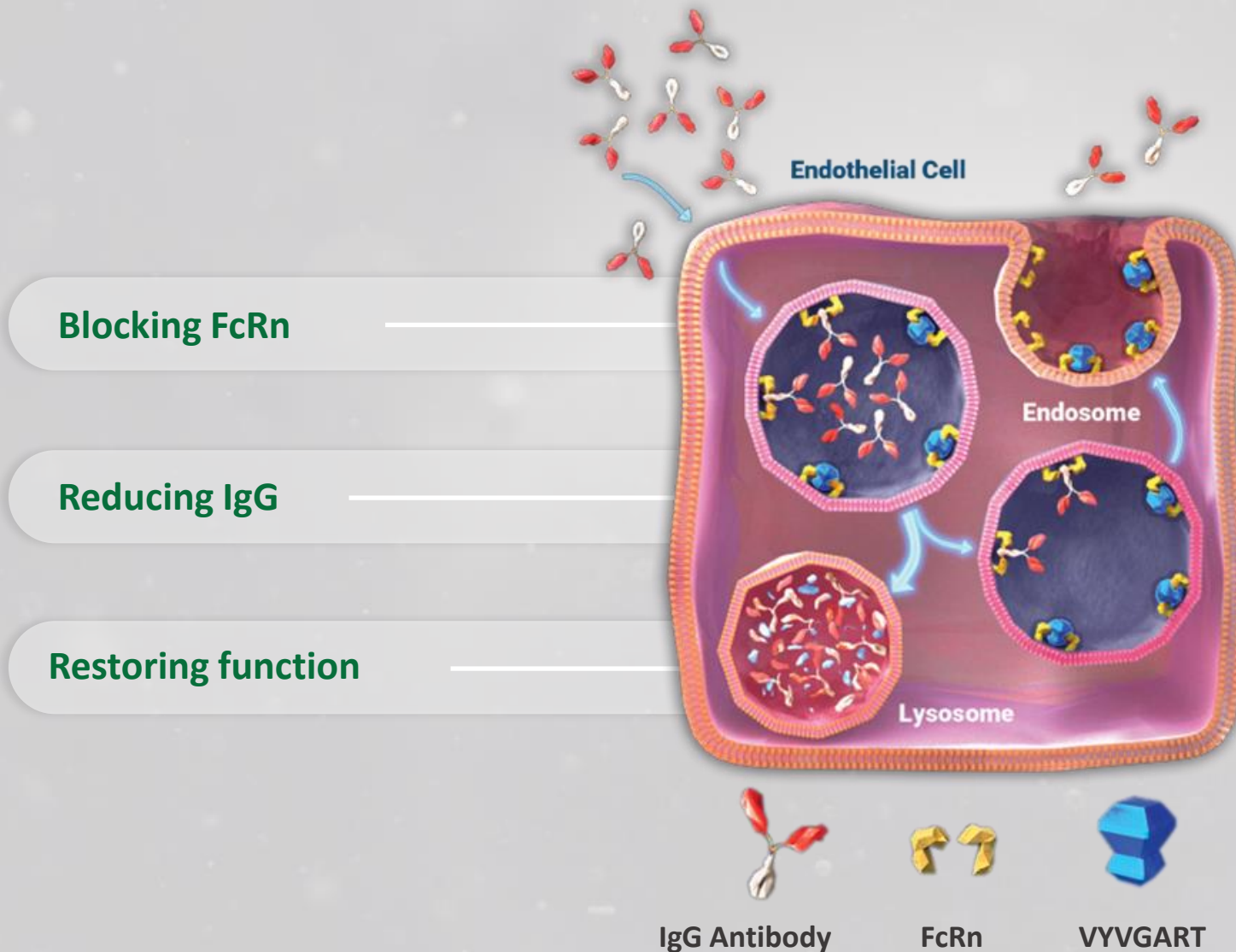
VYVGART IS NOW FDA-APPROVED


(efgartigimod alfa-fcab)

VYVGART is indicated for the treatment of generalized myasthenia gravis (gMG) in adult patients who are anti-acetylcholine receptor (AChR) antibody positive

Please see Important Safety Information and the full Prescribing Information for VYVGART at <https://vyvgart.com>

VYVGART: First FDA-Approved FcRn Antagonist



Mechanism of Action: VYVGART is a human IgG1 antibody fragment that binds to the neonatal Fc receptor (FcRn), resulting in the reduction of circulating IgG.

GENERALIZED MYASTHENIA GRAVIS Patient Experience

gMG is characterized by debilitating muscle weakness and fatigue. Despite taking an average of 2.3 current treatments, **61% of patients have poor well-being** according to WHO-5 Index

1/2
of Patients

have been diagnosed with depression or anxiety in addition to gMG

2.6
Years

mean time from symptom expression to diagnosis

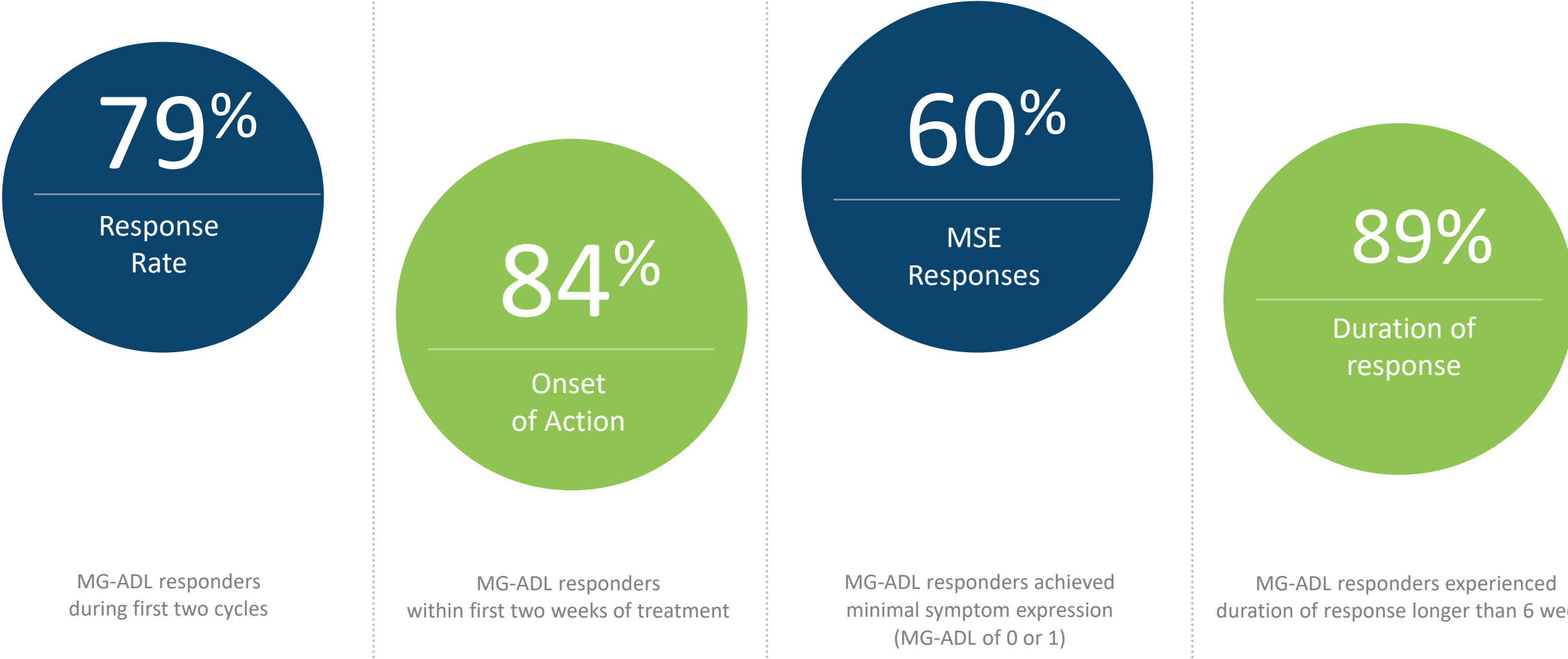
Symptoms can vary from patient to patient, day to day, **or even throughout the same day**...this unpredictability contributes to emotional burden of disease

51%
of Patients

stopped working completely from disease impact

Surveyed neurologists ranked severe gMG only behind ALS as most severe disease they treat

Phase 3 ADAPT Trial Results of Efgartigimod in gMG



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*

Challenging the gMG Treatment Paradigm

Meeting our stakeholders where they are



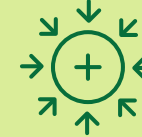
Empower Patients
to Demand Better



Best-in-Class
Patient
Support



Rapid HCP
Adoption of
VYVGART



Enable
Appropriate
Access

Global gMG Launch Underway



United States



VYVGART Approved December 17, 2021



Global

Japan
Approved
January 20, 2022

Europe
Anticipated decision
in 2H 2022

China (Zai Lab)
Anticipated filing
in mid-2022

Israel (Medison)
Anticipated filing
in 2Q 2022

Middle East (GenPharm)

Canada

Listening to and Learning from MG Community

MG United



A MYSTERY TO ME



MyRealWorld™ MG



Cooking Together



Differentiated Treatment Options for gMG Patients

ADAPT-NXT
Continuous Dosing

SC efgartigimod
single injection



VYVGART®
(efgartigimod alfa-fcab)

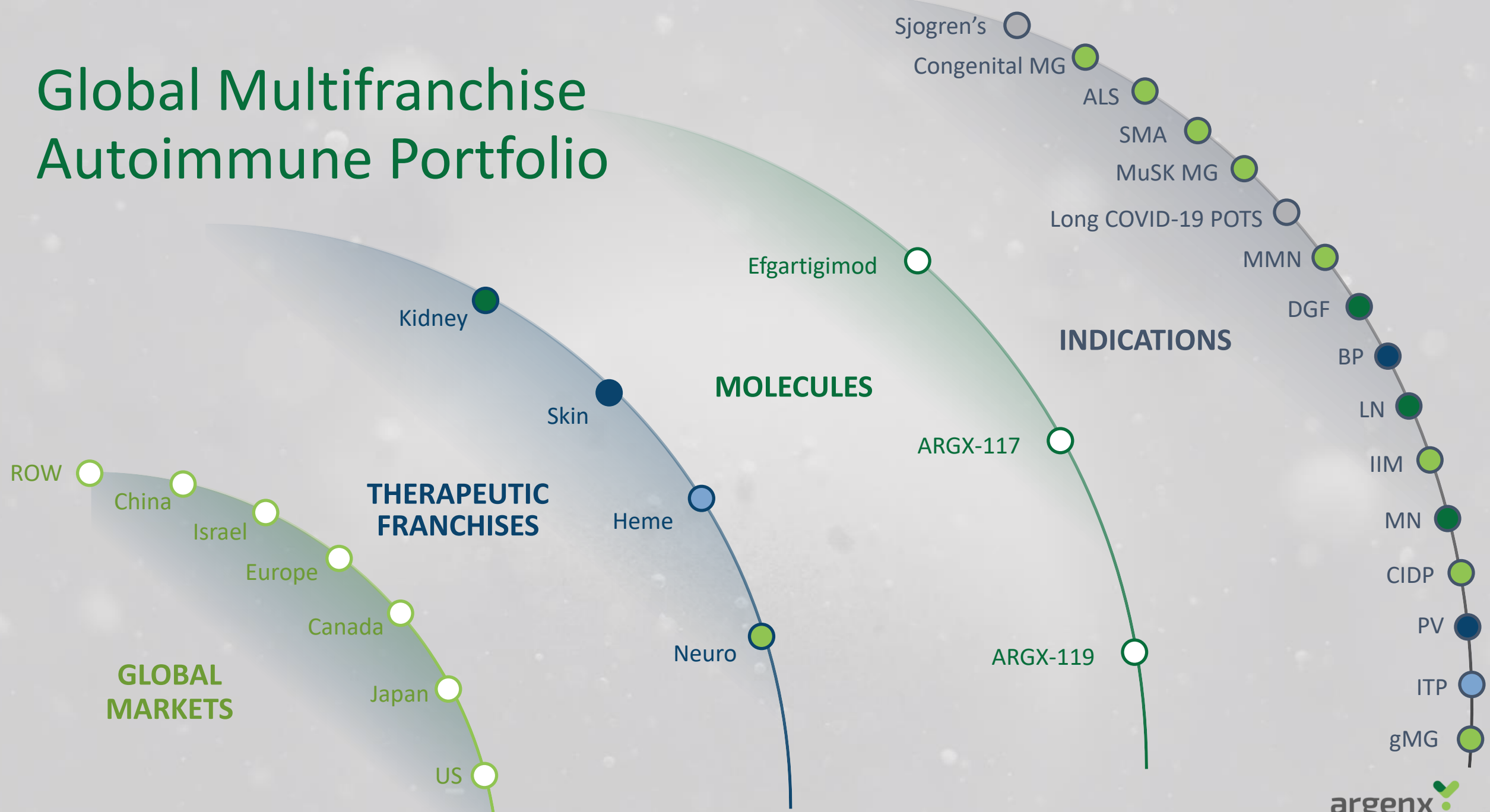
**Future Delivery
Vehicles and
Formulations**
(e.g., pre-filled syringe,
Elektrofi, autoinjector)

Eri, living with gMG

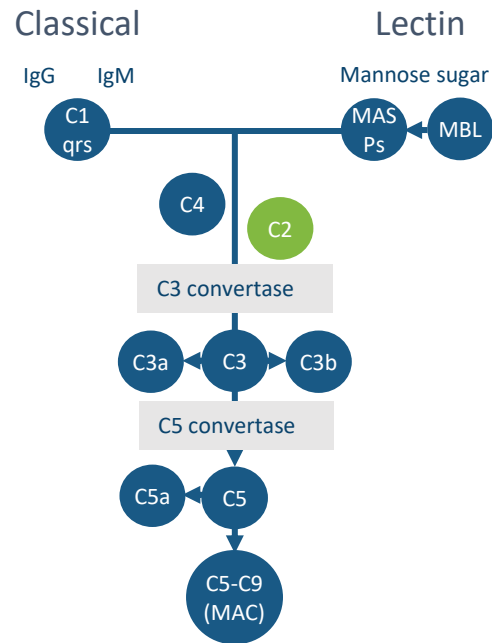
Autoimmune Pipeline								
Program	Indication		Preclinical	Phase 1	Proof of Concept	Registrational	Commercial	Update
VYVGART	gMG							US Launch Ongoing
Efgartigimod	gMG							Submit BLA by end of 2022
	CIDP							Data 1Q23
	Myositis							Trial to initiate 1Q22
	Pemphigus							Timeline under review
	Bullous Pemphigoid							Enrollment Ongoing
	ITP							Data 2Q22
	ITP							Data 1Q23
	Membranous Nephropathy							POC trial to start in 2022
	Lupus Nephritis							POC trial to start in 2022
	Sjogren's Syndrome							POC trial to start in 2H22
	COVID-19 Mediated Postural Orthostatic Tachycardia Syndrome							POC trial to start in mid-2022
ARGX-117	Multifocal Motor Neuropathy							Enrollment Ongoing
	Delayed Graft Function After Kidney Transplant							POC trial to start in 2022
ARGX-119	Neuromuscular Indications							Phase 1 to start in 2022
ARGX-120	Undisclosed							
Non-Autoimmune Programs								
Cusatuzumab	AML							
ARGX-118	Airway Inflammation							

Key:  NEUROMUSCULAR  HEMATOLOGY  DERMATOLOGY  NEPHROLOGY

Global Multifranchise Autoimmune Portfolio



ARGX-117 Blocks C2



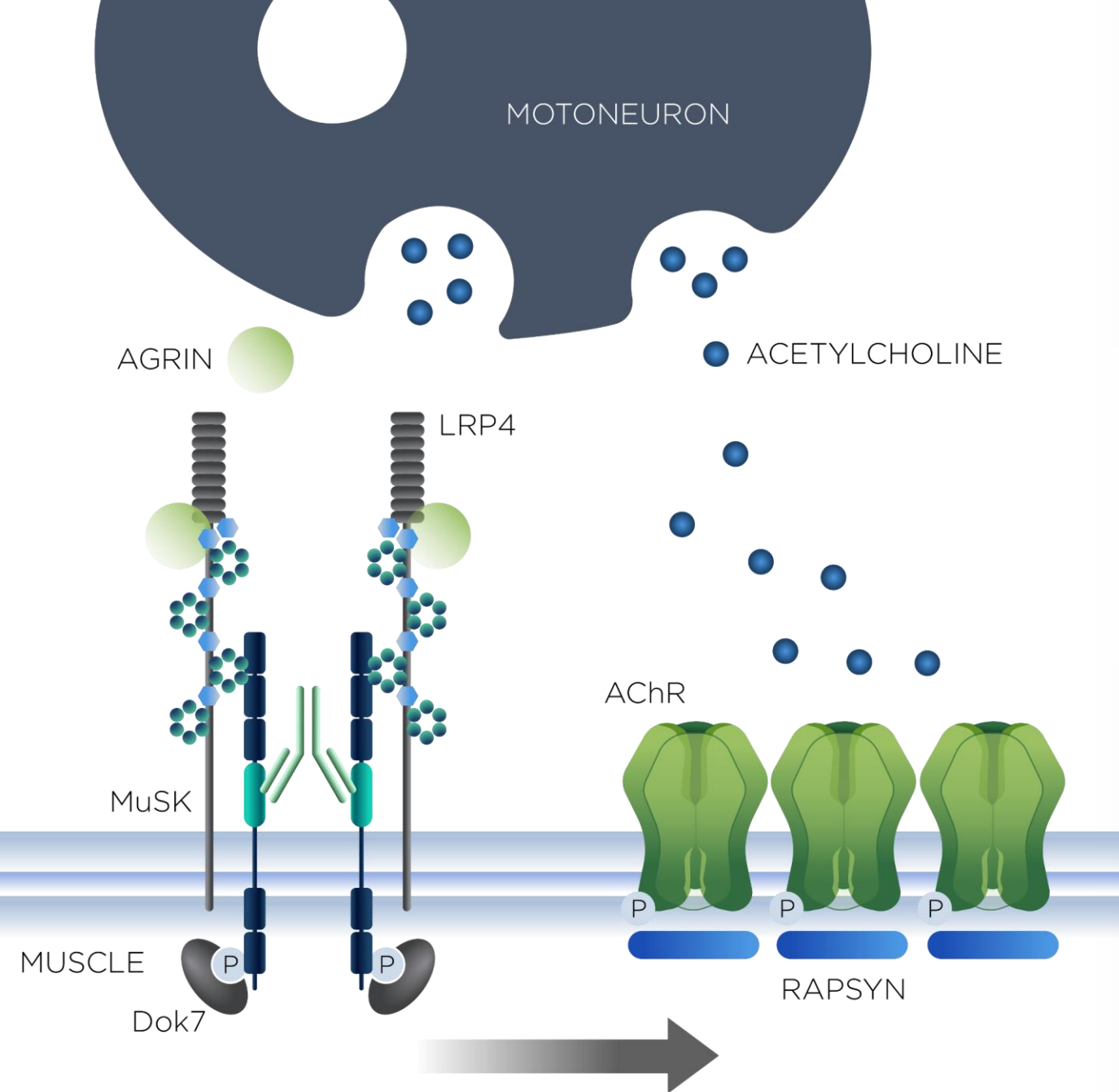
ARGX-117: Sweeping Antibody Designed to Target Unique Intervention in Complement

Targeting C2:

- Leaves alternative pathway intact against infection
- Blocks all complement effector functions

Multifocal Motor Neuropathy;
Phase 2 trial started

Delayed Graft Function in Kidney
Transplant



ARGX-119: MuSK Agonist with Broad Potential in Neuromuscular Disease

Oury, J., Zhang, W., Leloup, N. et al. Mechanism of disease and therapeutic rescue of Dok7 congenital myasthenia. *Nature* 595, 404–408 (2021).
<https://doi.org/10.1038/s41586-021-03672-3>

Pipeline Growth Driven By Immunology Innovation Program



Internal Value Creation Strategy

FIRST IN CLASS | UNIQUE DESIGN | MULTIPLE INDICATIONS

Efgartigimod

ARGX-117

ARGX-119

External Value Creation Strategy

ARGX-118

Staten
(ARGX-116)

AgoMAb
(ARGX-114)

Dualyx

Cusatuzumab

Genor
(ARGX-109)

LEO
(ARGX-112)

AbbVie
(ARGX-115)

One Team



One Plan

Determined to Achieve the
Unthinkable

Like the Argonauts,
we're on a bold mission:

**Engineer potentially
life-changing immunology
therapies for patients.**

Appendix

ADAPT-SC Bridging Study Design

DESIGN

110 adult gMG patients

MGFA Class II, III, IV

Anti-AChR antibody positive
(n=91) and negative (n=19)

MG-ADL score $\geq 5^a$

On ≥ 1 stable gMG treatment^b

Efgartigimod treatment naive

2 weeks screening

**Patients randomized 1:1 to receive
1000 mg SC efgartigimod or VYVGART**



3 weeks on treatment 7 weeks follow-up

10 weeks

**All patients receive one
treatment cycle**

(4 weekly single injections or 1-hour infusions)

**Primary
Endpoint**

Mean total IgG reduction from baseline
at day 29 compared to VYVGART

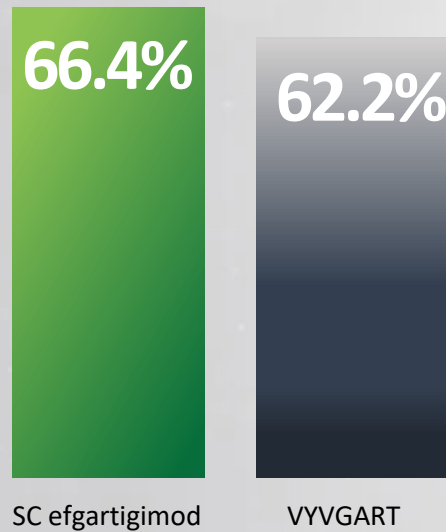
**Open-label
extension**

105 total patients
rolled over

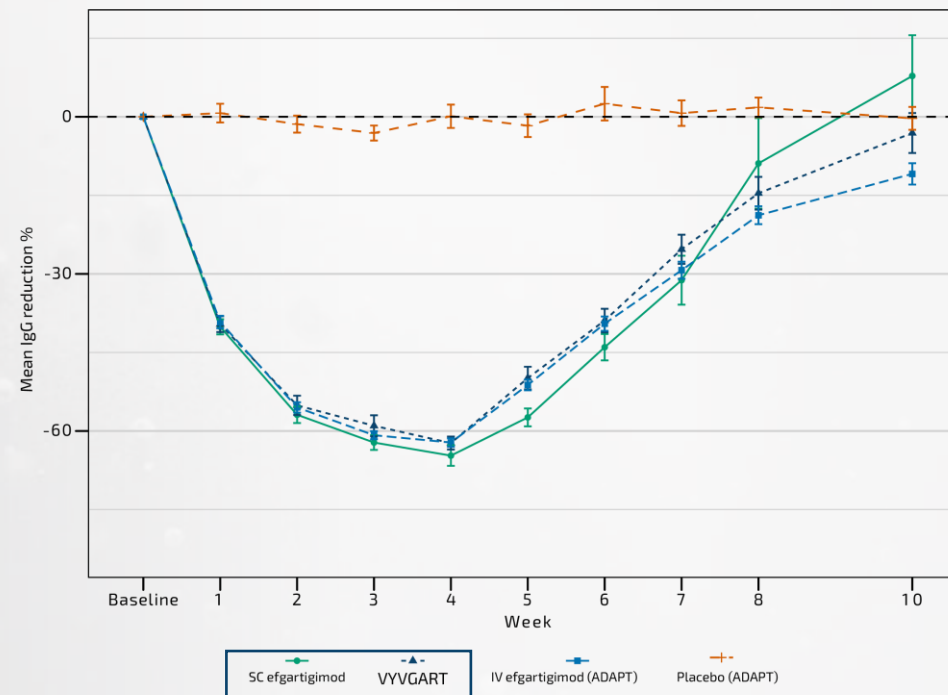
Primary Endpoint Met: SC Efgartigimod Noninferior to IV

Primary endpoint of noninferiority met [$p < 0.0001$]

Mean total IgG reduction from baseline at day 29



IgG reduction (%) in all ADAPT-SC and ADAPT participants



ADAPT-SC

Immune Thrombocytopenia ADVANCE Trials: Two Run in Parallel

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



Patients with
primary ITP
with platelet
counts
 $\leq 30 \times 10^9/L$

N=156



24 weeks 10mg/kg IV efgartigimod

Fixed weekly
dosing

Weeks 1-4

Weekly or q2w dosing adjusted
according to platelet count
thresholds

Weeks 5-16

Fixed weekly
or q2w dosing

Determined
at week 16

N=156



24 weeks 1000mg SC efgartigimod

Primary objective

Durable response:
sustained platelet
count ($\geq 50 \times 10^9/L$)
in 4/6 visits
between weeks 19
and 24

Topline IV data expected 2Q 2022; topline SC data expected 1Q 2023

Chronic Inflammatory Demyelinating Polyneuropathy ADHERE Trial



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

Stage B
(Stage A responders only)

Efgartigimod weekly SC

Placebo weekly SC

Efgartigimod weekly SC

Up to 12 weeks, until clinical improvement (ECI)

Up to 48 weeks

Efficacy analysis based on relapse (adjusted INCAT)

Study endpoint with 88 relapse events in stage B

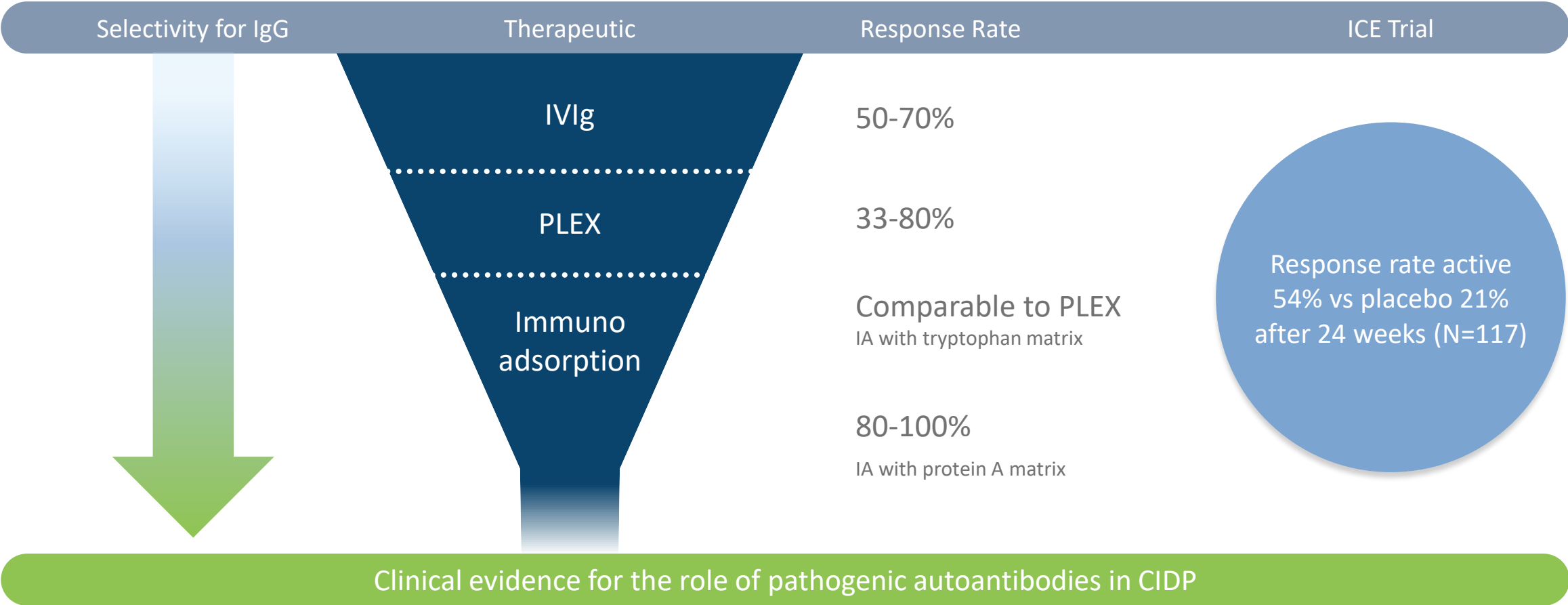
N=sample size estimation
~120-130

Followed by Open Label Extension study

Topline data expected 1Q 2023



CIDP: Therapeutic Activity Shown With Increasing Selectivity For IgG Reductions



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.

Pemphigus ADDRESS Trial: 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

Timing of topline data currently under review



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

Myositis: Adaptive Phase 2/3 Trial Design

180 patients

Total Duration: 52 weeks

Proposed Endpoints

Independent adjudication committee

Stratified for

- IMNM
- ASyS
- DM
- Severity of muscle weakness (MMT)

Eligibility criteria

- Muscle weakness
- No malignancies



Adults

IMNM

1000mg efgartigimod SC (+ SOC)

Placebo SC (+ SOC)

ASyS

1000mg efgartigimod SC (+ SOC)

Placebo SC (+ SOC)

DM

1000mg efgartigimod SC (+ SOC)

Placebo SC (+ SOC)

Interim Analysis of First 30 Patients of Each Subtype

Independent DSMB

Subtype selection

Sample size re-estimation

Primary

Mean Total Improvement Score (TIS)
Change (ACR/EULAR endorsed)

Key Secondary

- Response based on TIS, time to reach TIS, TIS duration
- Quality of Life
- Individual Core Set Measures TIS
 - MMT-8 score
 - Physician Global Assessment
 - Patient Global Assessment
 - HAQ-Disability Index score
 - Extramuscular Global Assessment
- Muscle enzyme serum level

Trial to initiate in 1Q 2022

Bullous Pemphigoid Registrational Trial Underway

160 patients

Total Duration: 36 weeks

Endpoints

Bullous Pemphigoid

Moderate-to-severe
disease (Investigator
Global Assessment
Score (IGA))

Newly diagnosed and
relapsing



Randomization (1:1)



Efgartigimod weekly SC



Placebo weekly SC

Concomitant Oral Corticosteroids (OCS)

OCS starting dose 0.5 mg/kg/day with ability to adjust
Active tapering to start from sustained control of disease activity (CDA)

Interim Analysis of First 40 Patients

Proposed primary endpoint

- Proportion of participants in complete remission while on minimal OCS (≤ 0.1 mg/kg/day) for ≥ 8 weeks at week 26

Secondary endpoints

- Cumulative dose of OCS
- Proportion of participants who achieve IGA BP score of 0 or 1 while off OCS for ≥ 8 weeks at week 36
- Proportion of participants who are in CR while off OCS for ≥ 8 weeks at week 36
- Changes from baseline to week 36 in 24-hour average itch
- Proportion of participants with CDA and remain free of relapse
- Quality of life

Followed by Open Label Extension study

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

Multifocal Motor Neuropathy Phase 2 Trial Underway

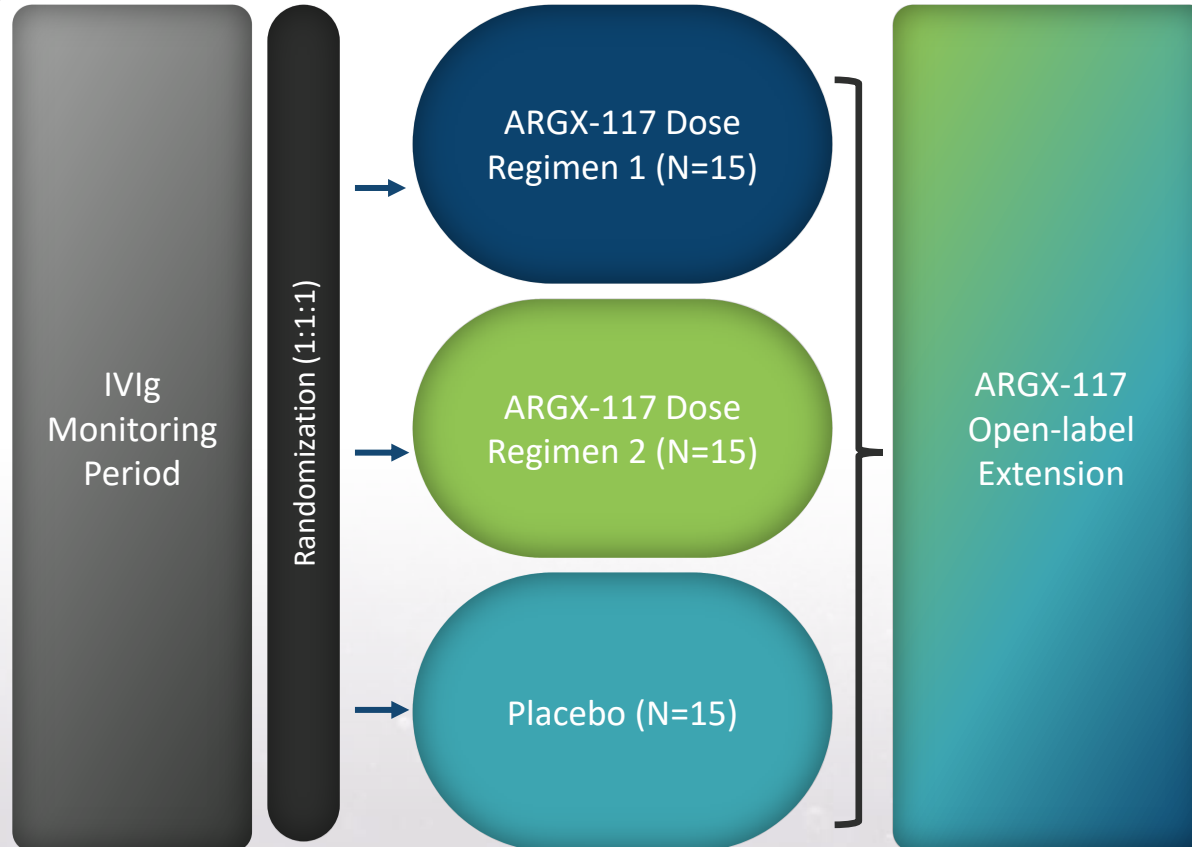
Patient Population

Trial to enroll approximately 45 MMN patients

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



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

Membranous Nephropathy (MN): Most Common Cause of Nephrotic Syndrome

No Current FDA-Approved Therapies

PREVALENCE

~70,000 patients in the U.S.

Mean age of diagnosis is 50-60 years

More predominant in men

HALLMARK

Thickening of the glomerular capillary walls due to immune complex deposition

20-30% progress to end-stage renal disease

DIAGNOSIS

Nephrotic syndrome, characterized by proteinuria and decreased serum albumin, presents in 80% of patients

70% of MN patients have IgG4 autoantibodies against PLA2R

PLA2R-negative patients can have anti- THSd7A or anti- NELL1 antibodies

UNMET NEED

No approved therapies; all used off-label

Currently used therapies include cyclophosphamide, rituximab and calcineurin inhibitors



Lupus Nephritis (LN):

Severe Organ Manifestation of Systemic Lupus Erythematosus

Need for new therapies with better tolerability profile

PREVALENCE

~45,000 patients in the U.S.

60% of adult patients with SLE develop renal symptoms

HALLMARK

Form of glomerulonephritis

5-20% of LN patients progress to end-stage renal disease

DIAGNOSIS

LN can be presenting manifestation for diagnosis of SLE

Presence of anti-dsDNA antibodies and related anti-nuclear antibodies; some closely associated with different histological classes

UNMET NEED

Oral corticosteroids and broad immunosuppressants are current standard of care

Therapies not uniformly effective

Substantial cause of morbidity and death among patients with SLE



Sjogren's Syndrome:

Systemic Autoimmune Disorder of Exocrine Glands

No Current FDA-Approved Therapies

PREVALENCE

~300,000 patients in the U.S.

9:1 female to male

Men less likely to have systemic manifestations

Age of typical onset: 40-50 years

HALLMARK

Inflammation of exocrine glands results in the development of sicca symptoms, such as dryness of the eyes and mouth

Some patients present with extra-glandular manifestations, including neuropathy, cutaneous, and pulmonary manifestations

DIAGNOSIS

Delayed diagnosis, often due to multifactorial nature of disease

Can take multiple years for correct diagnosis, typically by rheumatologist

Antibody presence (% of patients):
>80% ANA ; 70% anti-Ro/SSA; 50 % anti-La/SSB

UNMET NEED

IVIg treatment used to treat severe cases

Mild to moderate patients typically treated with secretagogues or sialagogues (e.g., eye drops, Biotene products)

Oral corticosteroids can be used for flare-ups



COVID-19 Mediated Postural Orthostatic Tachycardia Syndrome: Emerging Data Linking IgG Autoimmunity to Long COVID

No Current FDA-Approved Therapies

PREVALENCE

~500,000 – 1,000,000 POTS patients in the U.S. before COVID

~80% of patients are women between 15-50 years of age

HALLMARK

Excessive heart rate increase in an upright or standing position

Can prevent a person from being upright for more than a couple of minutes

Shortness of breath, headache, fatigue, poor concentration, heart palpitations, weakness, anxiety

DIAGNOSIS

Despite well-defined diagnostic criteria, diagnosis can be challenging due to non-specific and multi-organ nature of symptoms

IgG autoantibodies against anti-adrenergic [β 1-AR, β 2-AR]

UNMET NEED

No approved therapies

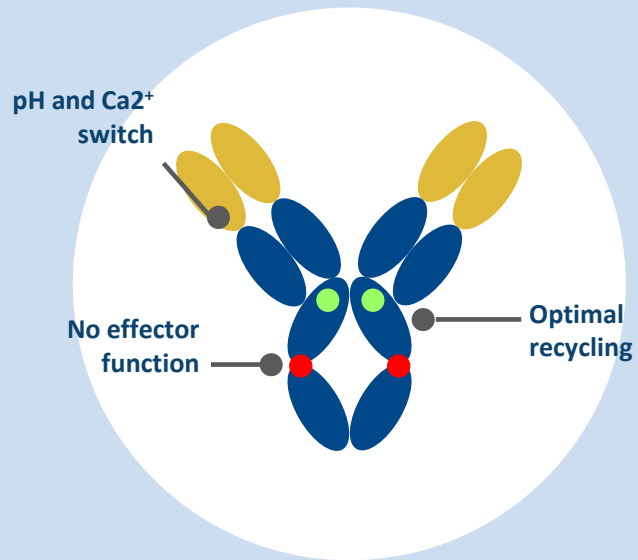
Symptomatic treatments focus on blood volume, kidney sodium levels, heart rate reduction and vessel constriction

POTS is associated with significant employment and economic loss

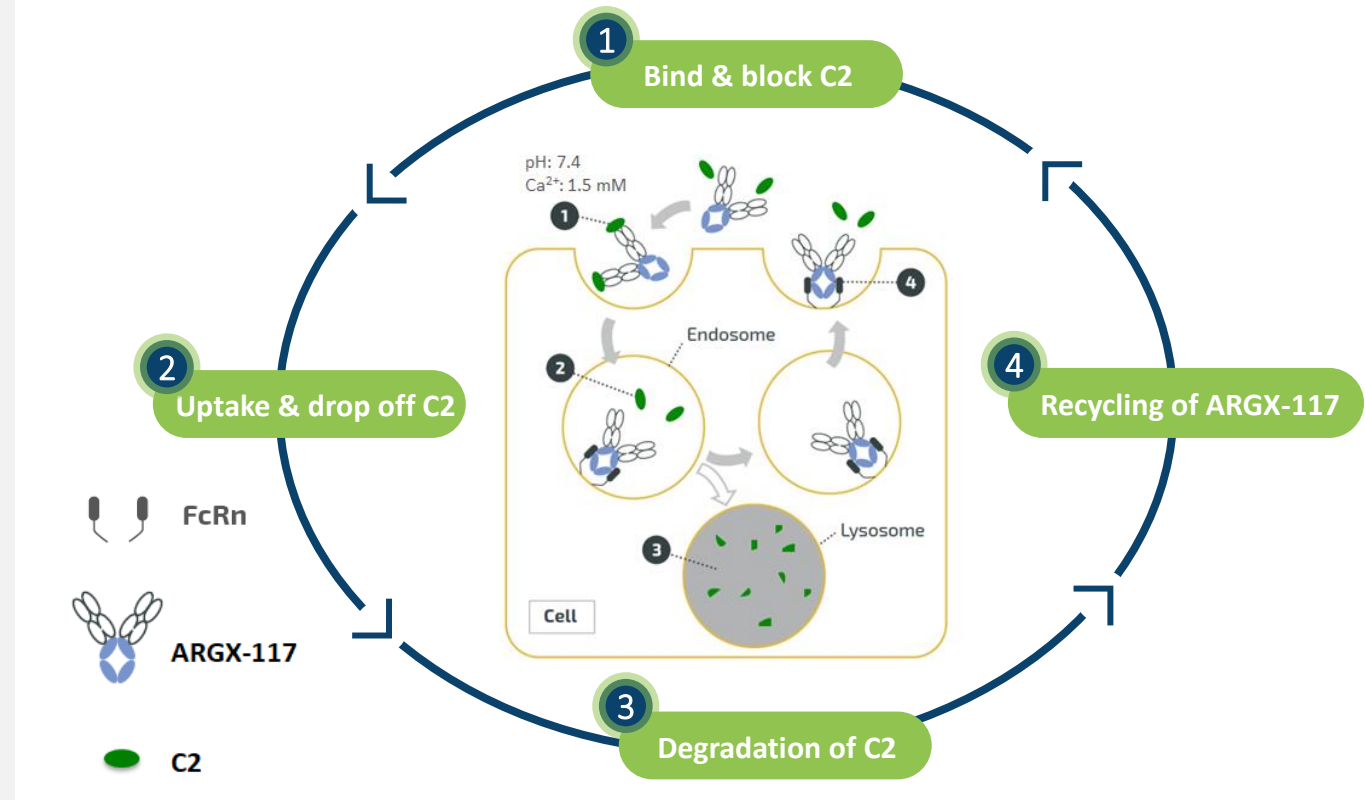


ARGX-117 Blocks and Actively Removes C2 from Circulation

Features of ARGX-117



Sweeping Antibody



Multifocal Motor Neuropathy:

Serious, Debilitating Neuromuscular Disease

Need for new therapies that slow progression of disease and reduce reliance on IVIg

PREVALENCE

~13,000 patients in the U.S., UK, Germany and France – increasing

Often underdiagnosed

Predominantly men

Median age 40 years

HALLMARK

Slowly progressive muscle weakness due to motor neuron degeneration

Mainly affects hands and forearms
- patients become dependent

DIAGNOSIS

Often misdiagnosed as ALS

Takes ~1.5 years for correct diagnosis

Nerve conduction blocks

Anti-GM1 IgM antibody presence

UNMET NEED

Steroids ineffective

First line and only approved therapy is frequent, high doses of IVIg over 2-5 days

>\$0.5B IVIg sales and growing



Kevin Budding, Lill Eva Johansen, Inge Van de Walle, Kim Dijkxhoorn, Elisabeth de Zeeuw, Lauri M. Bloemenkamp, Jeroen W. Bos, Marc D. Jansen, Chantall A.D. Curial, Karen Silence, Hans de Haard, Christophe Blanchetot, Liesbeth Van de Ven, Jeanette H.W. Leusen, R. Jeroen Pasterkamp, Leonard H. van den Berg, C. Erik Hack, Peter Boross, W. Ludo van der Pol Neurol Neuroimmunol Neuroinflamm Jan 2022, 9 (1) e1107; DOI: 10.1212/NXI.0000000000001107; argenx market research

Delayed Graft Function After Kidney Transplant

Complication Leading to Negative Graft Outcome

Need for new therapies that prevent graft loss and shortens hospitalization

PREVALENCE

Occurs in up to 40% of kidney transplant recipients

Over 2 million people worldwide currently receive dialysis treatment or are recipients of a kidney transplant

HALLMARK

Ischemia reperfusion injury (IRI) is a prime contributor to development of DGF

Negatively affects short and long-term graft outcome

MBL/C4d co-staining is observed in kidney biopsies

DIAGNOSIS

Delayed graft function (DGF), commonly defined as the need for dialysis within the first week post-transplantation

Increased serum creatinine levels

UNMET NEED

No current FDA-approved therapies to prevent DGF

DGF is associated with higher prolonged hospitalization

Growing market due to expanded criteria donor kidneys



*Pascual J, et al. Am J Kidney Dis. 2008; Biglarnia et al, Nat Rev Nephrol, 2018; Yarlagadda SG et al, Nephrol Dial Transplant . 2009
Castellano, Am J Pathol 2010, 176:1648–1659*