

Together We Discover

Reaching Patients Through
Immunology Innovation

Corporate Presentation

June 2022



Forward Looking Statements

This presentation has been prepared by argenx se (“argenx” or the “company”) for informational purposes only and not for any other purpose. Nothing contained in this presentation is, or should be construed as, a recommendation, promise or representation by the presenter or the company or any director, employee, agent, or adviser of the company. This presentation does not purport to be all-inclusive or to contain all of the information you may desire. This presentation also contains estimates and other statistical data made by independent parties and by us relating to market size and growth and other data about our industry. This data involves a number of assumptions and limitations, and you are cautioned not to give undue weight to such estimates.



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 expectations with respect to the timing and its expectations with respect to reporting data from registrational trials, and initiating trials in new indications and the filing of regulatory submissions; expected execution of commercial franchise launches, priorities and adoption; its plans for global commercialization launch of VYVGART; expectation of broad U.S. policy coverage of VYVGART; plans for Medison to file for approval in Israel in second quarter of 2022, and Zai Lab to file for approval in greater China by mid-2022; expected commercialization in Canada and expected partnership with GenPharm for commercialization in the Middle East. A further list and description of these risks, uncertainties and other risks can be found in argenx’s U.S. Securities and Exchange Commission (SEC) filings and reports, 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. Given these uncertainties, the reader is advised not to place any undue reliance on such forward-looking statements. These forward-looking statements speak only as of the date of publication of this document. argenx undertakes no obligation to publicly update or revise the information in this presentation, including any forward-looking statements, except as may be required by law.

Our Plan to Build a Leading Immunology Company

Driving revolution in autoimmunity with precision medicines

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

Committed to
our Patients and
their
Communities

Enviably
Immunology
Pipeline

Rooted in
Science through
our IIP

argenx 2025

We believe the future belongs to those who collaborate best

Catalyst-Filled Start to 2022 – Delivering on Our Objectives

- 1 US VYVGART® Sales
\$21.2M in net 1Q product sales
- 2 Global Launch
Japan launched in May
- 3 2 Phase 3 Datasets
Positive ADVANCE - IV data in ITP
Positive ADAPT-SC data in gMG - BLA submission by YE
- 4 Advancing Pipeline
10 efgartigimod indications in clinic by YE
2 ARGX-117 indications in clinic by YE
- 5 Strong Financial Position
\$806M raised; \$2.9B cash position

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 2Q22
	Pemphigus							Data 2H23
	Bullous Pemphigoid							Enrollment Ongoing
	ITP							Positive results May 2022
	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						
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 Pipeline								
Cusatuzumab	AML							
ARGX-118	Airway Inflammation							

Executing on VYVGART Launch Priorities

Meeting our stakeholders where they are



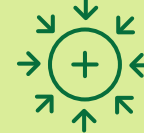
Empower Patients
to Demand Better



Best-in-Class
Patient
Support



Rapid HCP
Adoption of
VYVGART



Enable
Appropriate
Access

~\$21.2M

1Q 2022 Net Product Sales



10 mg/kg

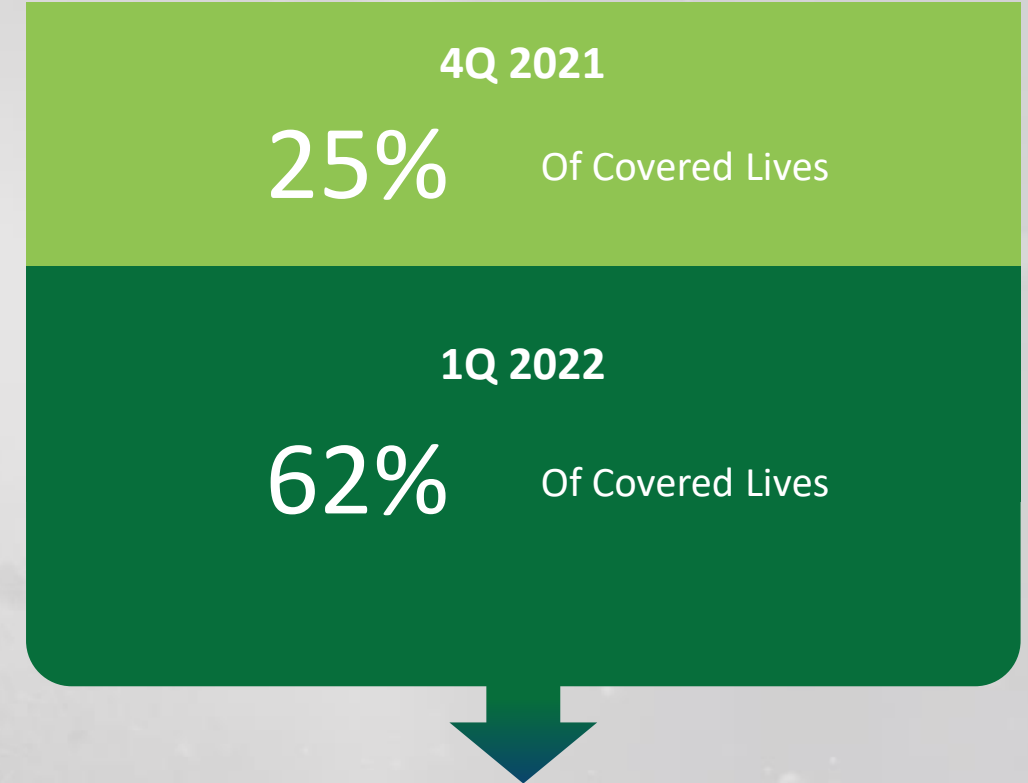
VYVGART[®]
(efgartigimod alfa-fcab)

Intravenous infusion

Key Pillars of VYVGART Value-Based Agreement



Most VYVGART-specific policies are favorable and aligned to label, with prior authorization median of (6) months



Seeking broad coverage by
end of 2Q 2022

Global gMG Launch Progressing



United States



VYVGART Approved December 17, 2021



Global

Japan

Launched May 2022

Europe

Anticipated decision
in 2H 2022

China (Zai Lab)

Anticipated filing
in mid-2022

Israel (Medison)

Filed for registration

Middle East (GenPharm)

Central/Eastern Europe (Medison)

Canada

Innovative Trial Accelerated Path Forward



Leveraged PK/PD from ADAPT

- Leveraged correlation between pharmacodynamic and clinical effect as observed in ADAPT

Met Primary Endpoint

- Met primary endpoint, demonstrating noninferior total IgG reduction at day 29 with subcutaneously administered efgartigimod compared to intravenous (IV) administration
- Secondary endpoints show clinical improvement consistent with IV administration

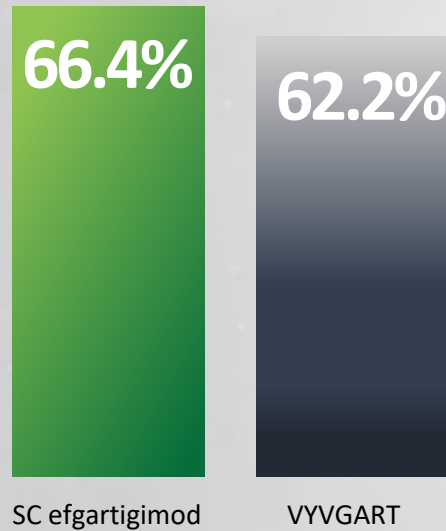
2023 Launch

- BLA to be submitted by end of 2022

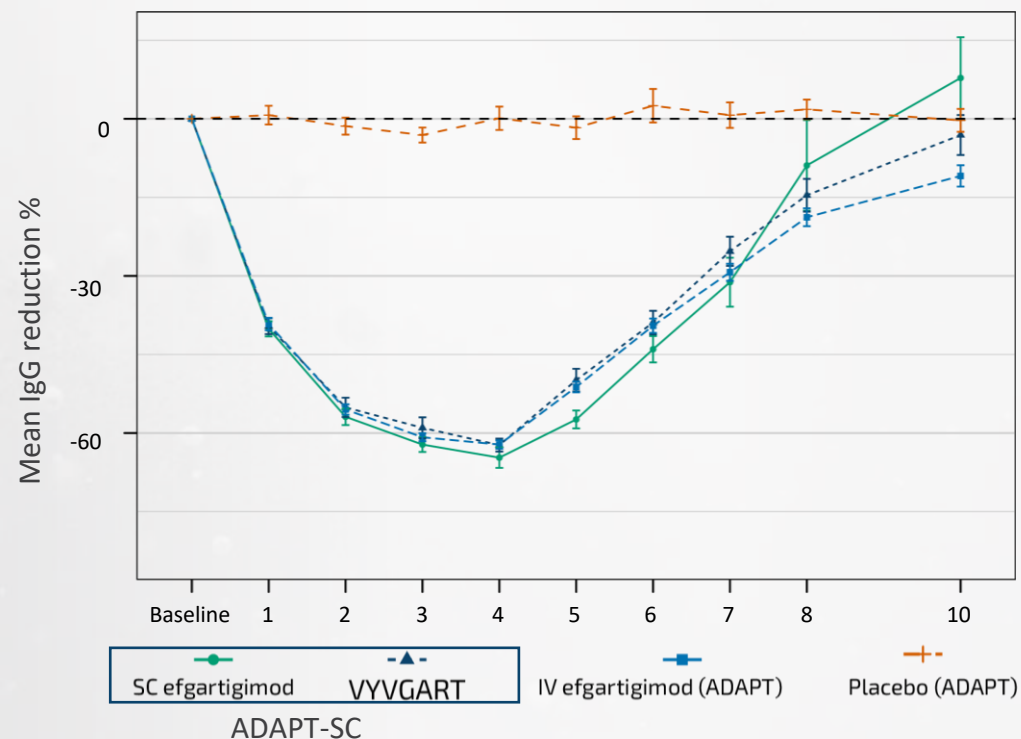
Primary Endpoint Met: SC Efgartigimod Noninferior to IV

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

Mean total IgG reduction from baseline at day 29

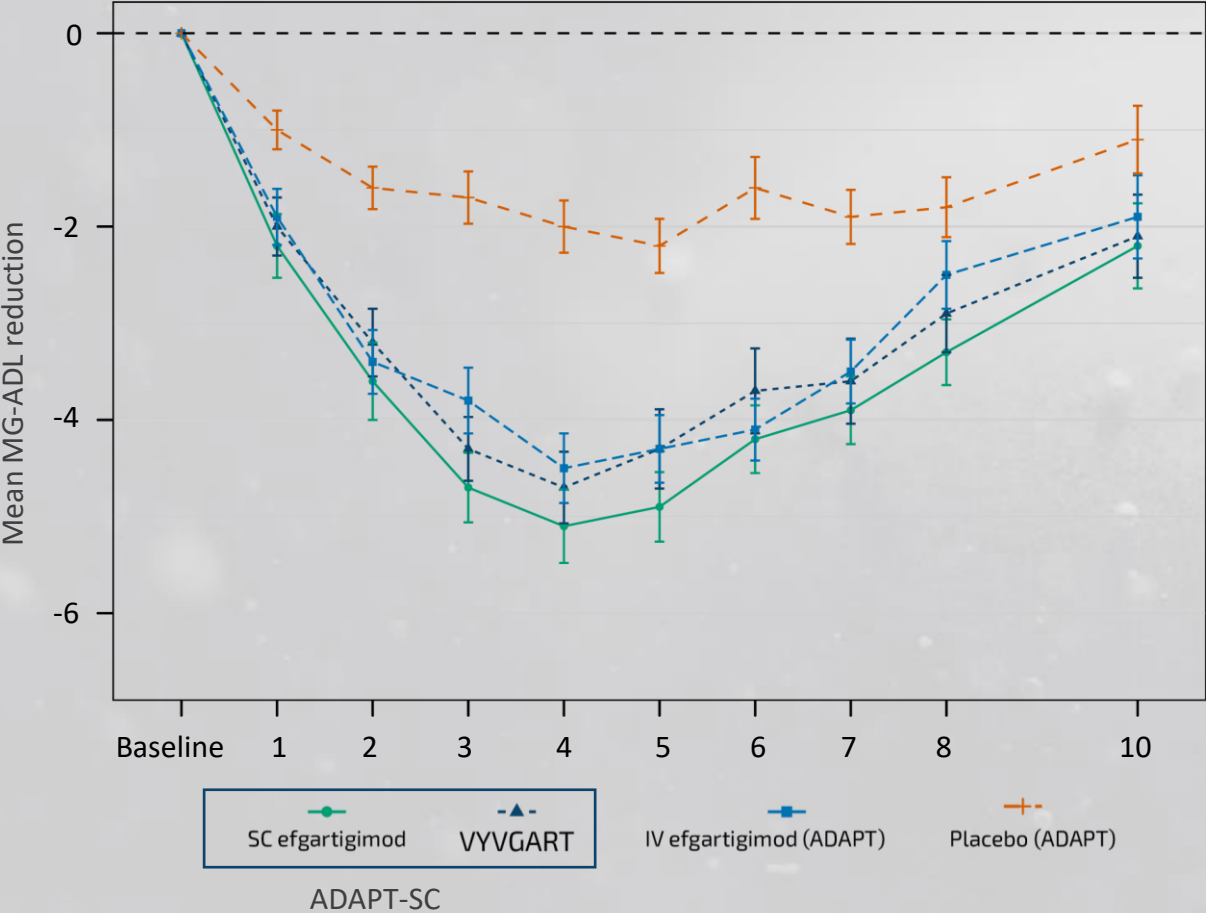


IgG reduction (%) in all ADAPT-SC and ADAPT participants

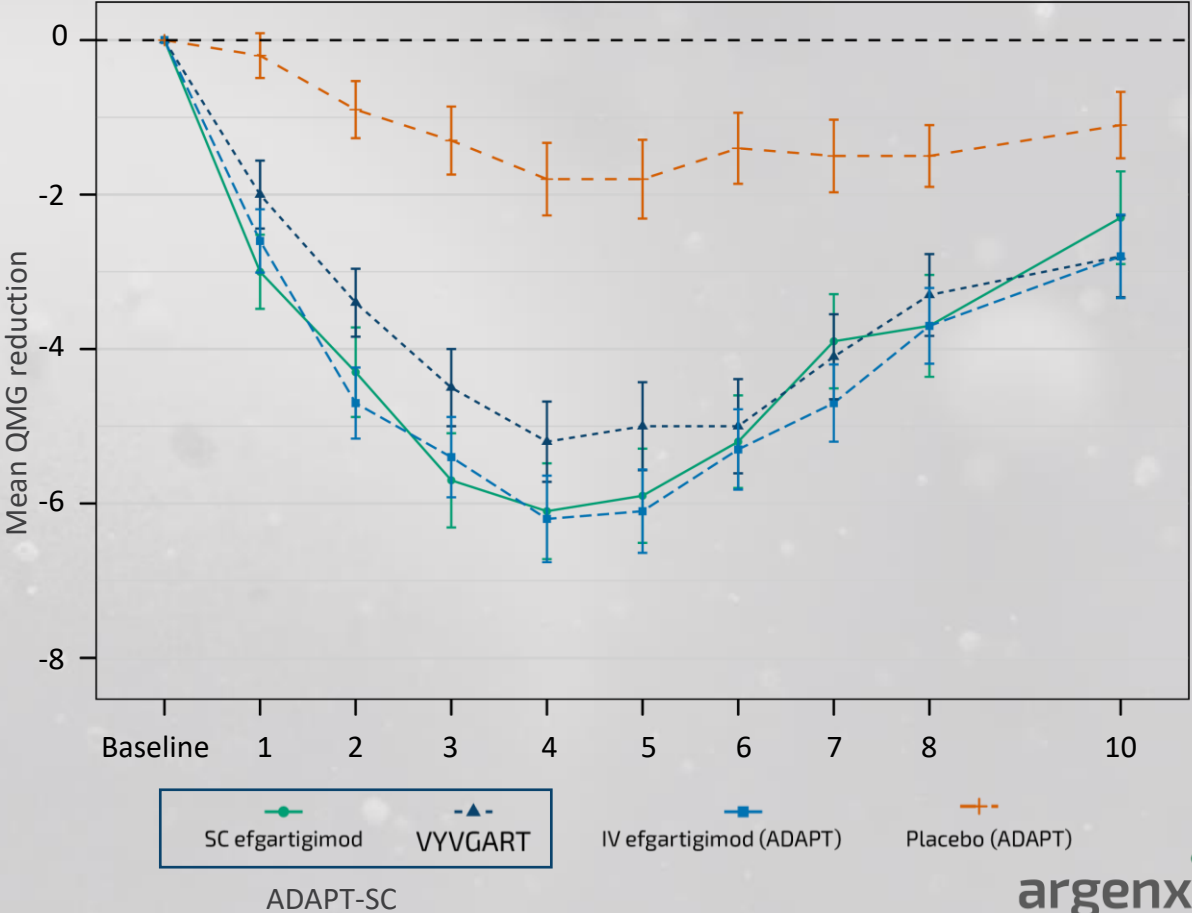


Fast, Deep and Durable Responses

Total change from baseline in MG-ADL score over time across ADAPT and ADAPT-SC



Total change from baseline in QMG score over time across ADAPT and ADAPT-SC



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

Results Support Path Forward



Primary endpoint met

- Statistically significant and clinically meaningful improvement in platelet counts over placebo

Meaningful patient benefit observed

- Fast and robust platelet count improvement over placebo
- Ability for every other week dosing confirmed

Favorable safety & tolerability observed

- Chronic administration of VYVGART was well-tolerated
- Safety profile consistent with previous clinical trials

Primary Immune Thrombocytopenia

LINDA
Living with primary ITP



Rare, serious autoimmune disease characterized by low platelet counts leading to risk of **bruising, bleeding and debilitating fatigue**

More than **70K** patients in the U.S.

75% of patients report impaired ability to perform daily tasks

50% of patients report high impact on emotional well-being including anxiety and depression

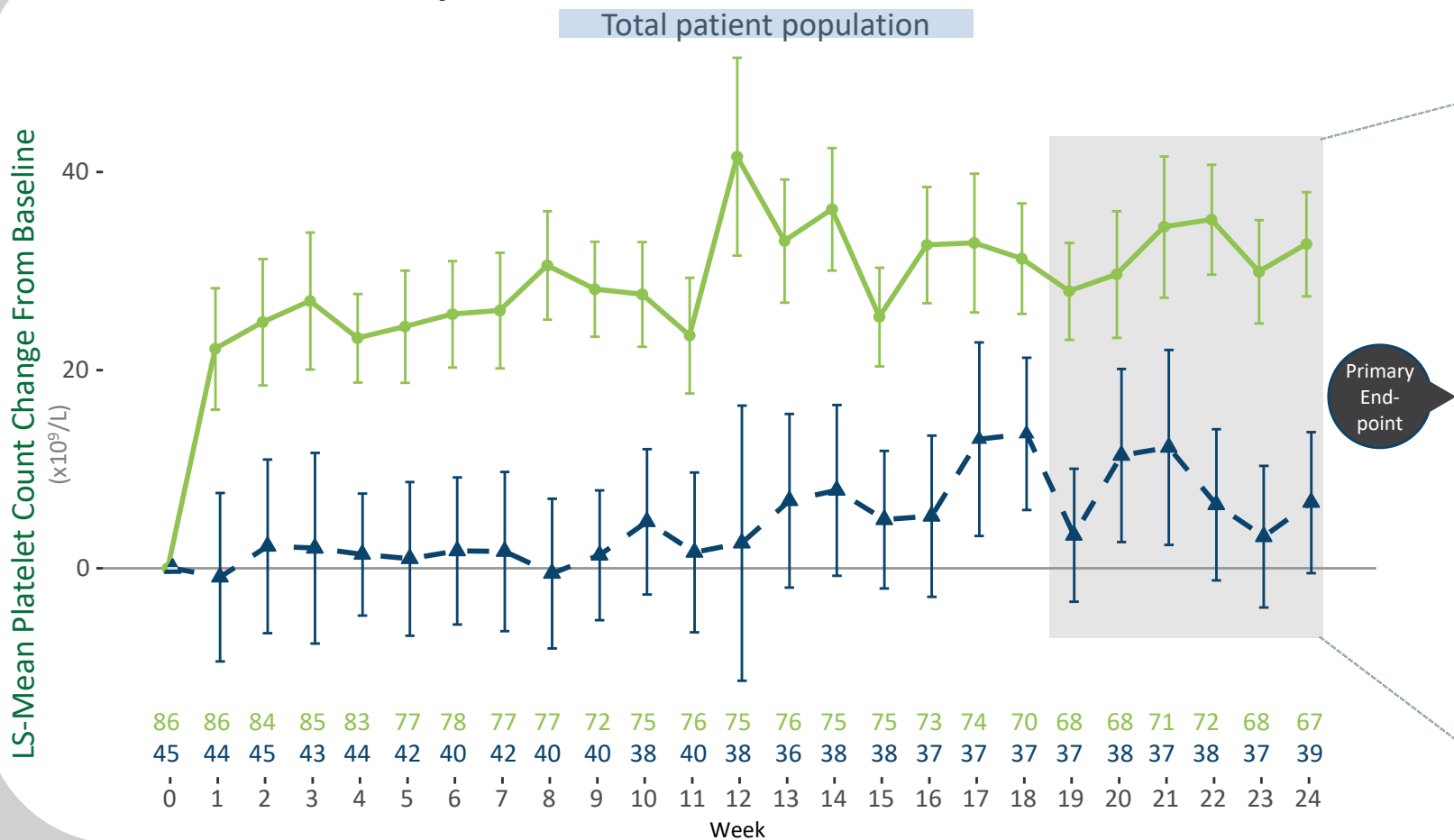
48% of patients reduced or considered reducing work hours

Current therapies often **fail to achieve sustained elevation of platelet counts**, necessitating retreatment or additional lines of therapy

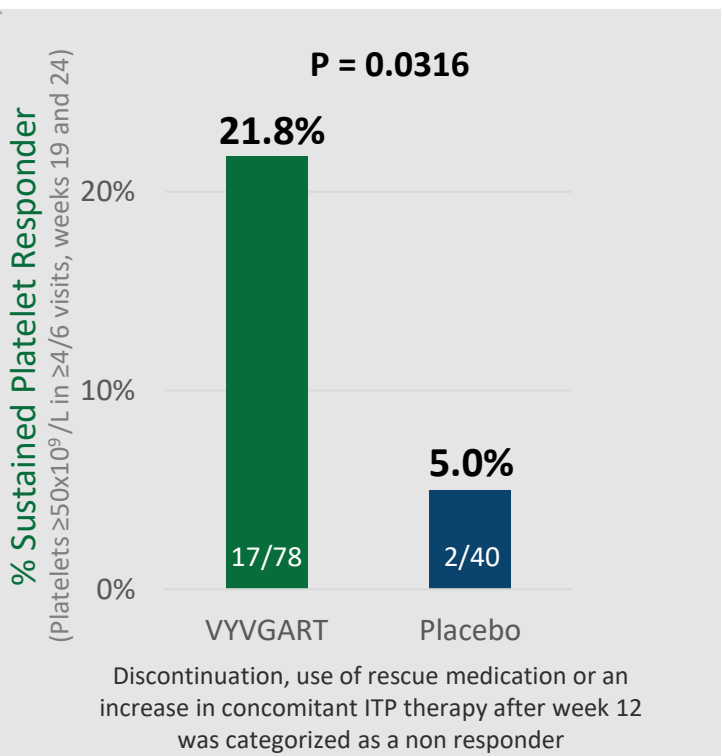
Treatment goal aims to achieve **sustained safe platelet levels**, with absence of bleeding and **without toxicity** of therapy

ADVANCE—IV Primary Endpoint Met

Clear Separation Between VYVGART and Placebo



Primary Endpoint: Statistically Significant Improvement over Placebo



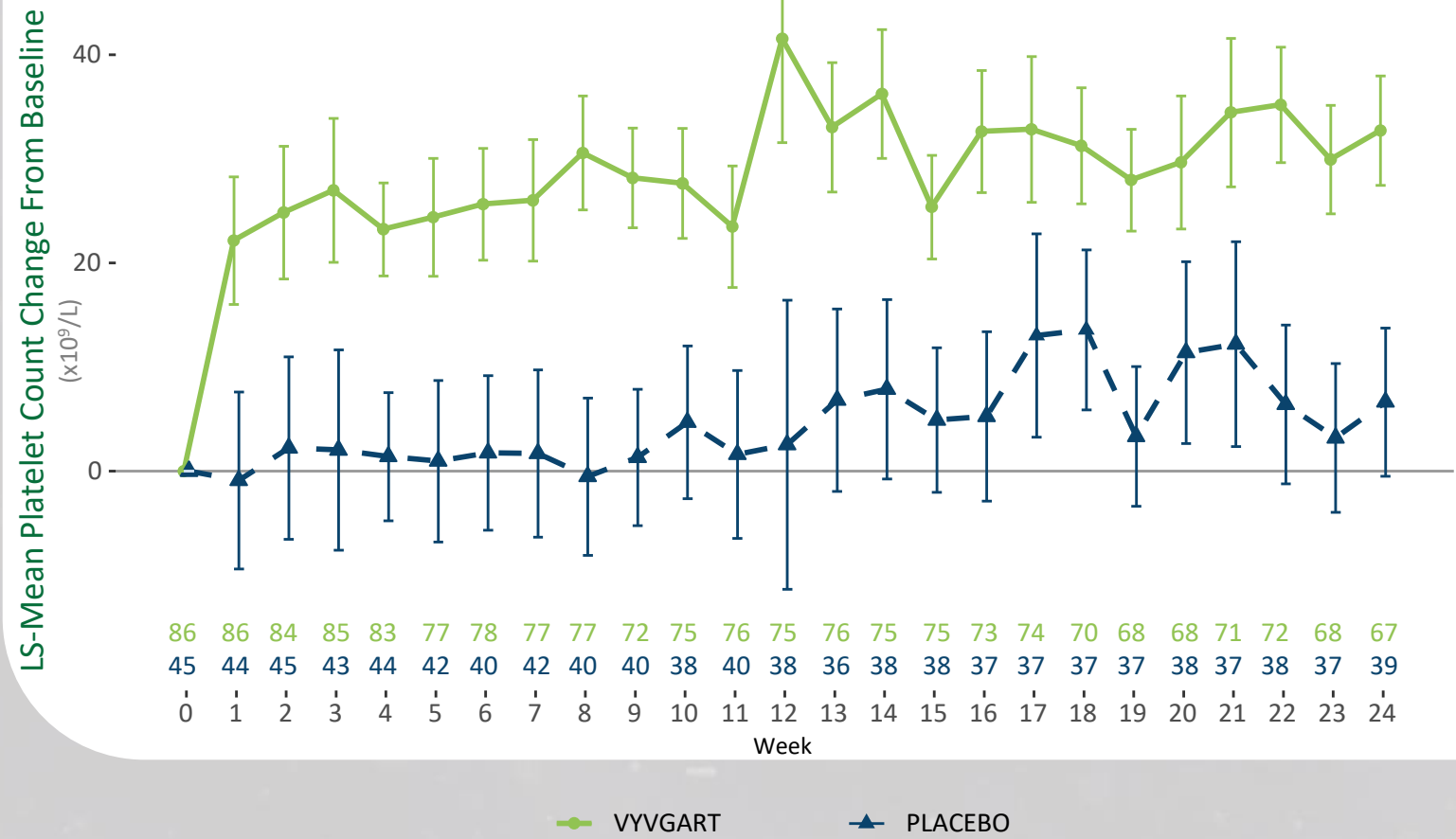
— VYVGART — PLACEBO

Primary Endpoint calculated with Cochran–Mantel–Haenszel test

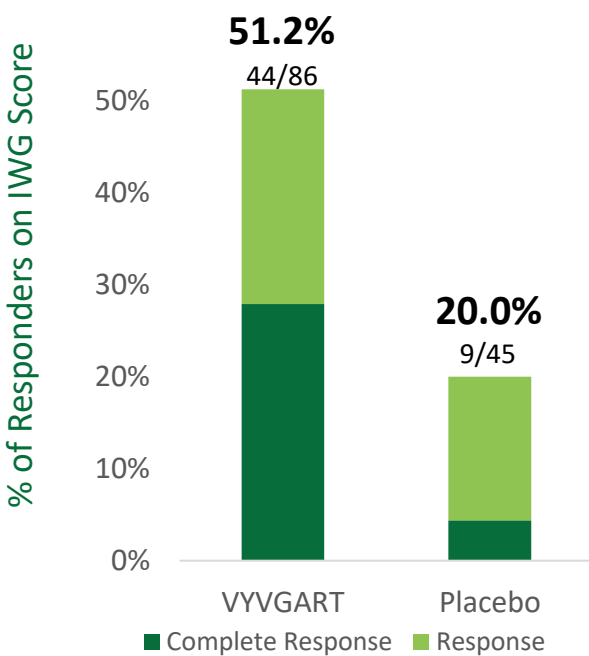
Per IWG criteria, 51% of VYVGART Patients Showed Real-World Relevant Platelet Count Improvement in Absence of Bleeding

Clear Separation Between VYVGART and Placebo

Total patient population



International Workgroup (IWG) Criteria reflecting real-world treatment goals



VYVGART Demonstrated Favorable Safety Profile

Safety and tolerability profile consistent with previous trials; First chronic dosing Phase 3 trial; Most AEs mild to moderate

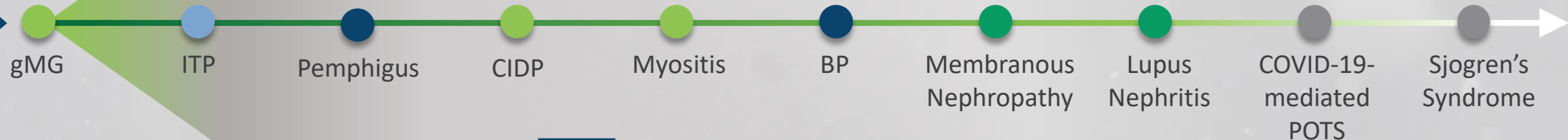
Number of patients	VYVGART (N=86) n (%)	Placebo (N=45) n (%)
Patients with an Adverse Event (AE) <i>n</i> (%)	80 (93.0)	43 (95.6)
Patients with an AE grade ≥ 3 <i>n</i> (%)	11 (12.8)	9 (20.0)
Most frequent AEs <i>n</i> (%)		
Blood urine present	31 (36.0)	17 (37.8)
Petechia	13 (15.1)	12 (26.7)
Contusion	17 (19.8)	6 (13.3)
Hematuria	14 (16.3)	6 (13.3)
Headache	14 (16.3)	7 (15.6)
Patients who discontinued treatment due to AEs <i>n</i> (%)	4 (4.7)	1 (2.2)
Patients with serious adverse events* (SAE) <i>n</i> (%)	7 (8.1)	7 (15.6)
Patients with AEs deemed related by the investigator <i>n</i> (%)	15 (17.4)	10 (22.2)
Special interest per protocol: Patients with AEs related to bleeding	61 (70.9)	39 (86.7)
Special interest per protocol: Patients with AEs related to infection	25 (29.1)	10 (22.2)

*No SAEs were considered by investigators to be related to therapy

ITP is the Second Indication with a Positive Phase 3 Read-out for VYVGART

LEVERAGE
FORMULATIONS
ACROSS MULTIPLE
INDICATIONS,
THERAPEUTIC
FRANCHISES, AND
MOLECULES

VYVGART®
(efgartigimod alfa-fcab)



4/4 (gMG, ITP, PV, CIDP) positive POC trials results



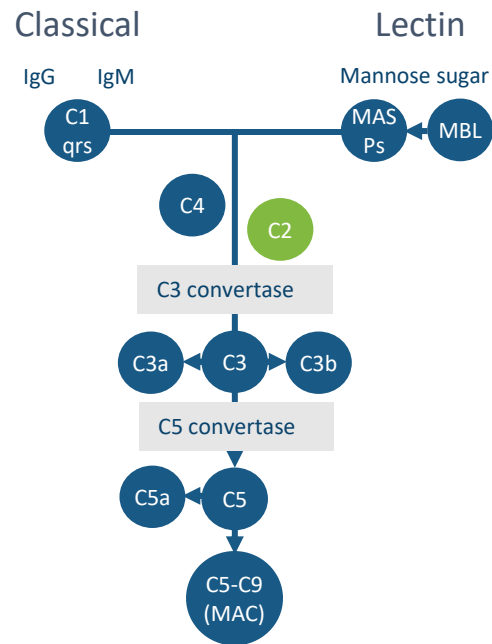
3/3 (gMG-IV, gMG-SC, ITP) positive Phase 3 trials results



First launches in US and Japan in gMG

Key: ● NEURO ● HEME ● SKIN ● KIDNEY

ARGX-117 Blocks C2



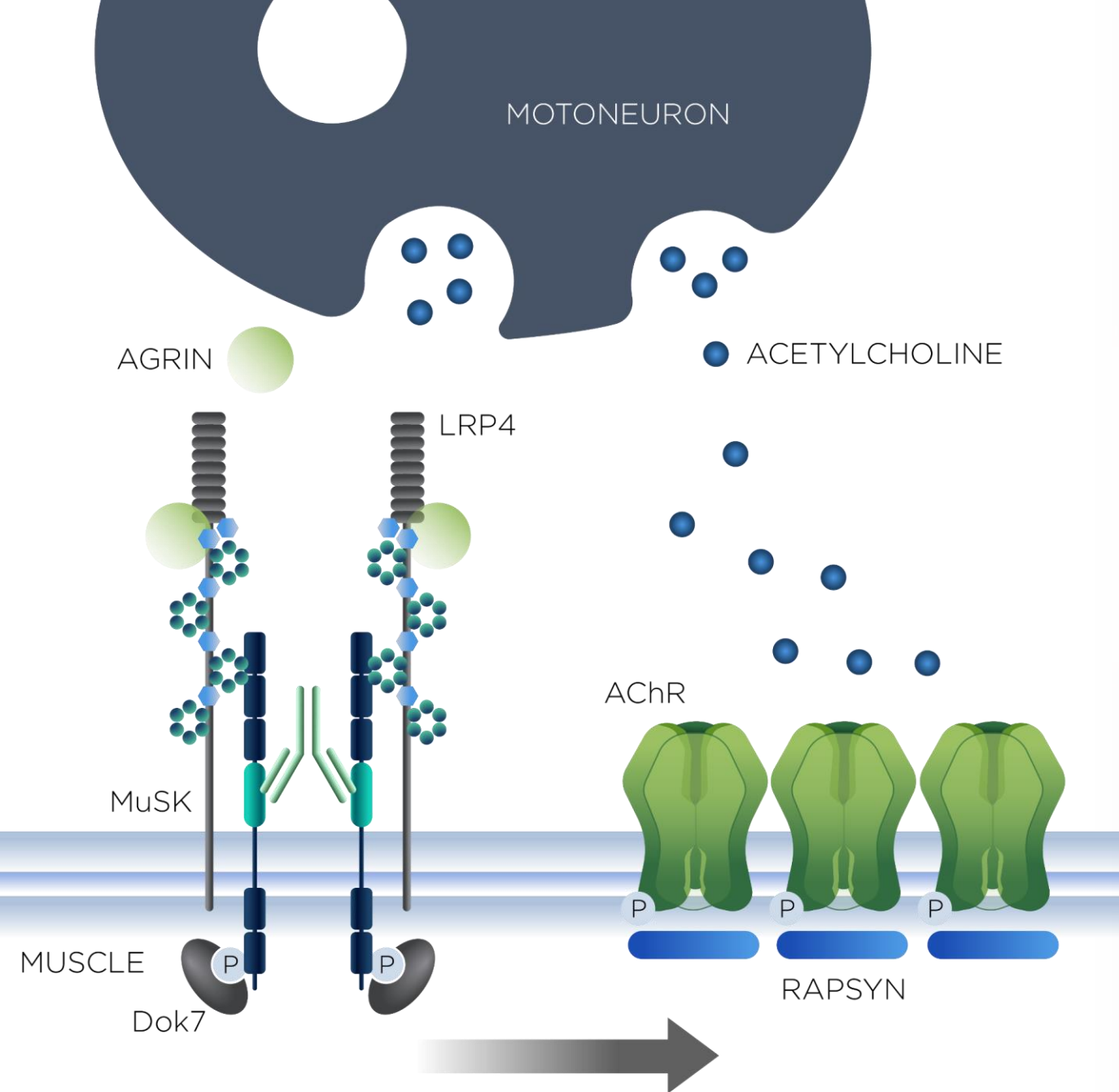
ARGX-117: Sweeping Antibody Designed to Target Unique Intervention in Complement

Targeting C2:

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

Multifocal Motor Neuropathy;
Phase 2 trial started

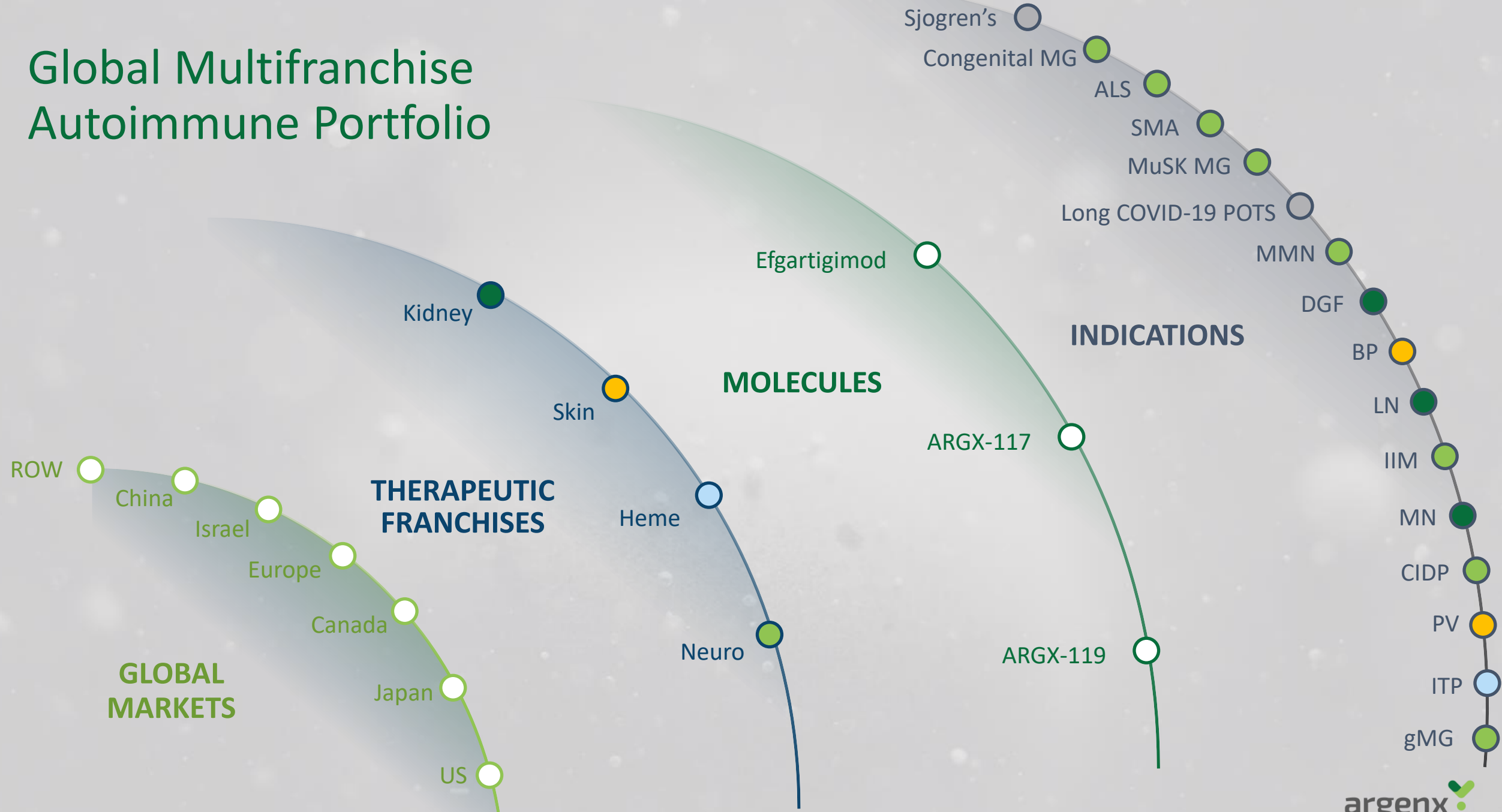
Delayed Graft Function in Kidney
Transplant



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>

Global Multifranchise Autoimmune Portfolio



gMG: generalized myasthenia gravis; ITP: immune thrombocytopenia; PV: pemphigus vulgaris; CIDP: chronic inflammatory demyelinating polyneuropathy; MN: membranous nephropathy; IIM: idiopathic inflammatory neuropathies; LN: lupus nephritis; BP: bullous pemphigoid; DGF: delayed graft function; MMN: multifocal motor neuropathy; POTS: postural orthostatic tachycardia syndrome; MusK: muscle specific kinase; SMA: spinal muscular atrophy; ALS: amyotrophic lateral sclerosis;

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

ADVANCE-IV Trial Design Based on Key Regulatory Input

131 adult Chronic or Persistent ITP patients

2 weeks screening

Mean platelet count $<30 \times 10^9/L$

Stable concomitant ITP therapy allowed

Stratification: splenectomy, concomitant ITP therapy

Patients randomized 2:1 to receive 10mg/kg VYVGART or placebo (24 weeks)

Primary End-point

Fixed weekly dosing

Weekly or q2w dosing based on platelet count thresholds

Fixed weekly or q2w dosing based on week 15



- Primary endpoint: Sustained platelet count ($\geq 50 \times 10^9/L$) in $\geq 4/6$ visits between weeks 19 and 24
- Stringent endpoint in line with regulatory feedback, addressing platelet count variability
- Secondary and exploratory endpoints center around the extent of disease control to illustrate real-world viability

Identical design for ADVANCE-SC with subcutaneous (SC) efgartigimod; topline 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

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

Topline data expected 2H 2023



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 2Q 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



Adults

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

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



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*