



Together We Discover

Corporate Presentation

August 2023



Forward Looking Statements

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Our mission is to transform severe autoimmunity

Redefining autoimmune
diseases as IgG-mediated

Raising expectations for what
'well-controlled' means for
patients

Redefining What 'Well-Controlled' Means for the Patient

We want to transform treatment for patients

Minimize treatment burden

Achieve broad and sustained responses

Regain control of their lives, including professionally and socially



Crystal
Living With CIDP

I was the type of woman that would run first thing in the morning before work, and then CIDP hit, and it was like hitting the wall at a hundred miles an hour.

VYVGART

for Generalized Myasthenia Gravis

NOW TWO FDA-APPROVED PRODUCTS

VYVGART[®]
(efgartigimod alfa-fcab)
Injection for Intravenous Use
400 mg/20 mL vial

VYVGART[®] **Hytrulo**
(efgartigimod alfa and
hyaluronidase-qvfc)
Subcutaneous Injection
180 mg/mL and 2000 U/mL vial

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

VYVGART Simplifies Treatment of gMG in the Community

Predictable Safety and Tolerability:
18 months and over 3000* real world patients

Simple Treatment Initiation:
broad access, no vaccination requirements

Choice in HOW to be Treated:
IV infusion or SC injection

Demonstrated Efficacy:
78% responders across first two treatment cycles in ADAPT

VYVGART[®]
(efgartigimod alfa-fcab)
Injection for Intravenous Use
400 mg/20 mL vial

VYVGART[®] Hytrulo
(efgartigimod alfa and hyaluronidase-gvfc)
Subcutaneous Injection
180 mg/mL and 2000 U/mL vial

Options for Site of Care:
infusion center, HCP office, home administration

Lowering the bar for VYVGART treatment initiation and expanding access to gMG patients

Optimizing Core Launch Strategies

VYVGART launched in US,
Japan, Germany, Italy

SUBMISSIONS OR APPROVALS IN
10+ COUNTRIES

Consistent prescriber
growth to increase breadth
of patients

>2,100 PRESCRIBERS
UNITED STATES



\$489M

IN NET VYVGART SALES YTD

**17,000 addressable
gMG patients**

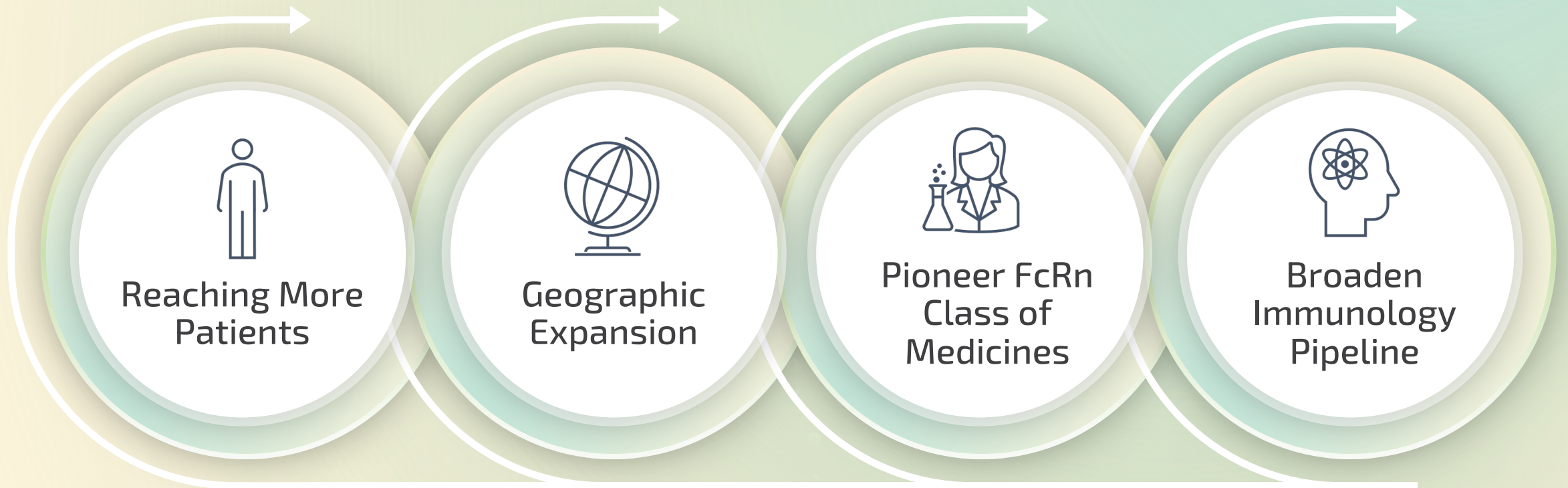
Consistent growth looking at
month over month new patient
starts

**Potential to drive
earlier line uptake**

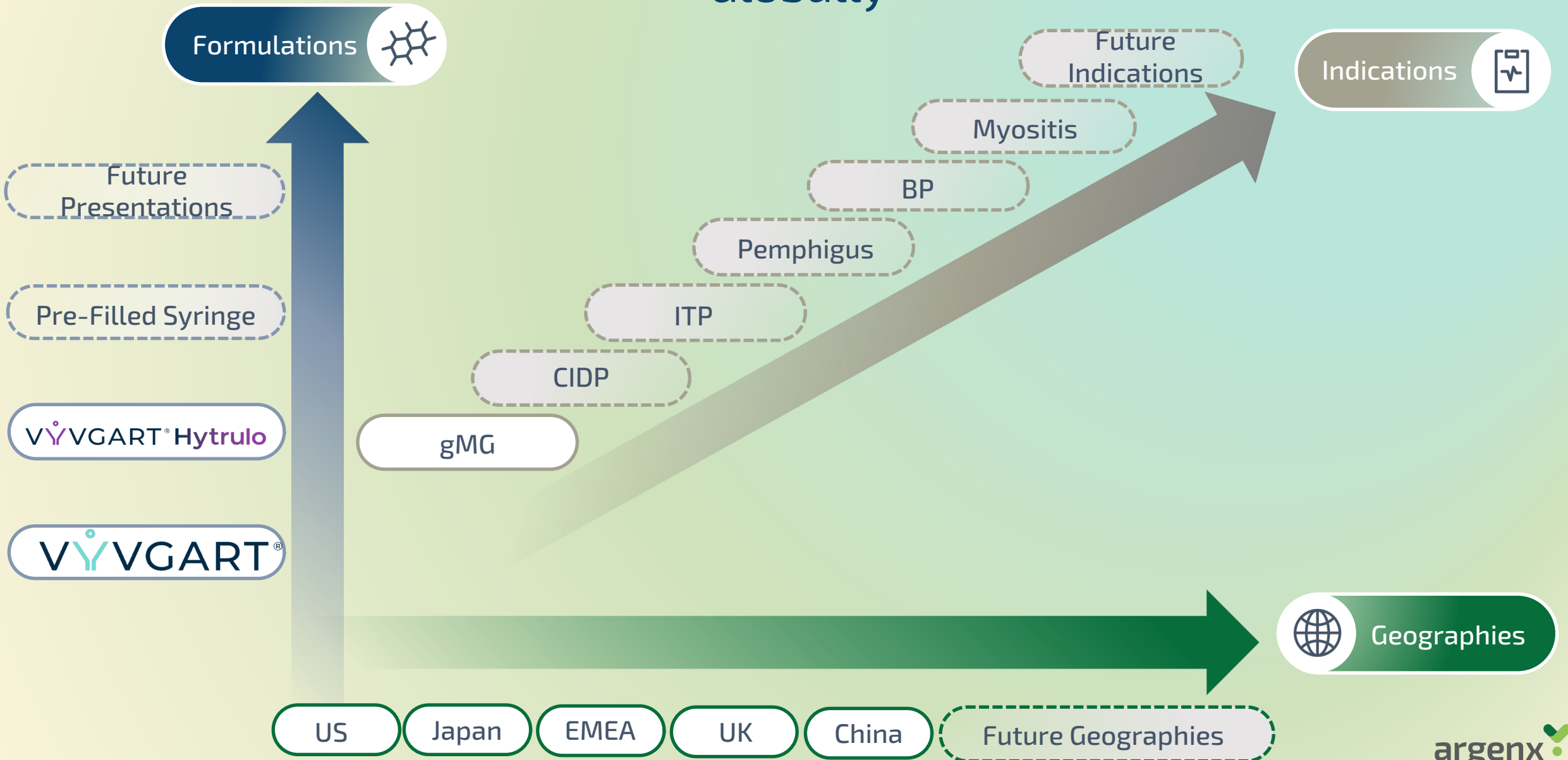
VYVGART Hytrulo launched in US

Driving Sustained Growth Across the Business

Consistent Execution + Serial Innovation



Multi-dimensional Expansion to Reach Autoimmune Patients Globally



Reaching gMG Patients Across the Globe

VYVGART®

Approvals Complete

U.S. DEC 2021
JAPAN JAN 2022
EUROPE SEPT 2022
UK MAR 2023
ISRAEL APRIL 2023
CHINA JUNE 2023

Approvals Pending

CANADA Q3 2023

VYVGART® Hytrulo

Approvals Complete

U.S. JUNE 2023

Approvals Pending

JAPAN BY Q1 2024
EUROPE Q4 2023
CHINA 2024

IgG Autoantibodies Serve as Unifying Biology Rationale for POC Indications

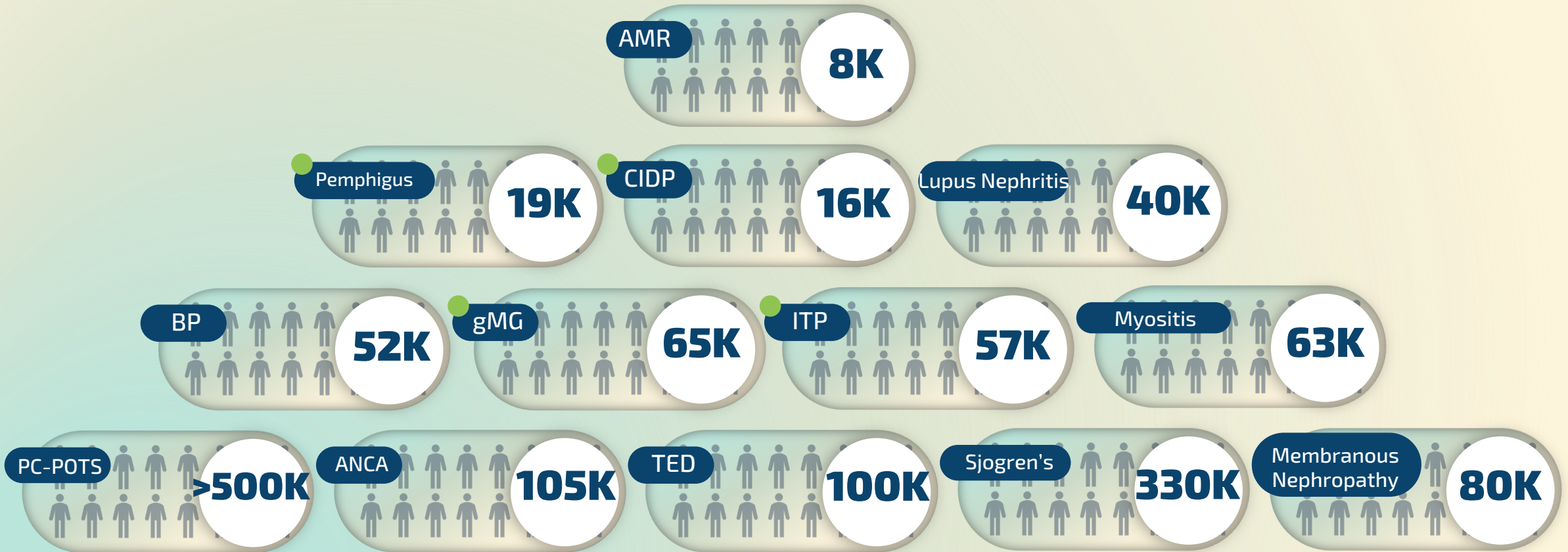


Pioneer FcRn
Class of
Medicines

Contactin1

DSG-1	Ro/SSA	Mi-2
La/SSB	THSd7A	MPO-ANCA
PLA2R	ANA	NF155
DSG-3	GPIIb/IIIa	HMGCR
BP230	DSA	NELL1
AChR	PR3-ANCA	GPCR
MuSK	BP180	LRP4
TSHR	JO-1	SRP
	Immune complexes	

gMG is just the beginning



● indications with successful proof-of-concept or Phase 3 data

ADHERE: Opportunity to Transform CIDP Patient Experience



Stage A

67% - 78%

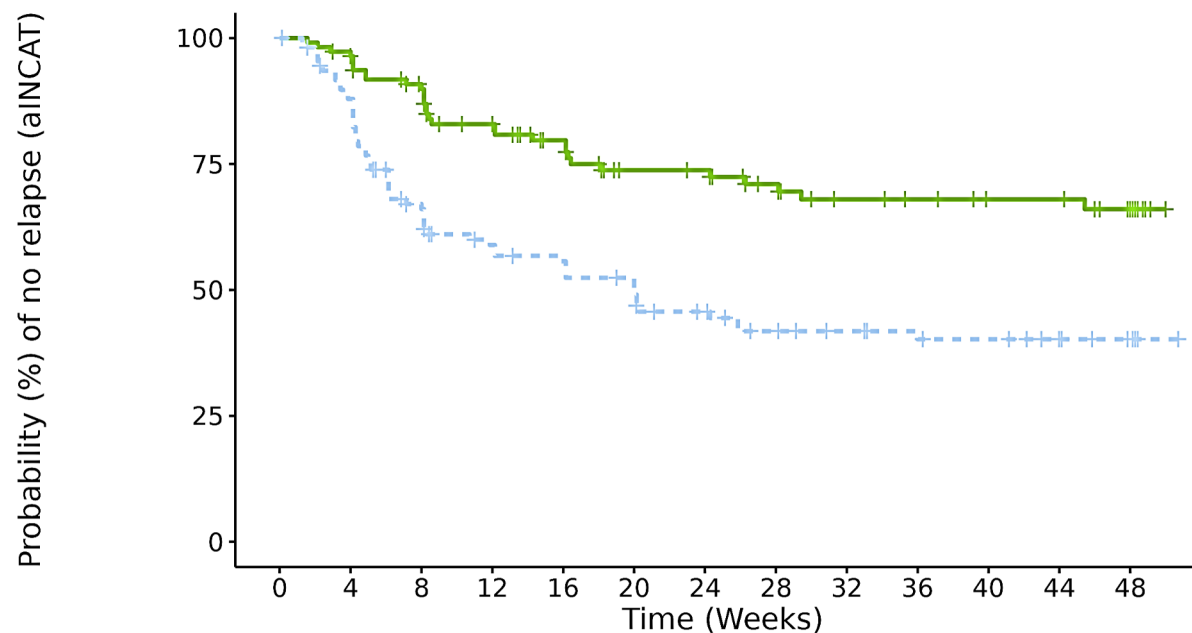
Response rates demonstrate IgG autoantibodies play significant role in underlying CIDP biology

Stage B

HR: 0.39

P = 0.000039

61% lower risk of relapse based on time to first adjusted INCAT deterioration with VYVGART Hytrulo compared to placebo



patients at risk

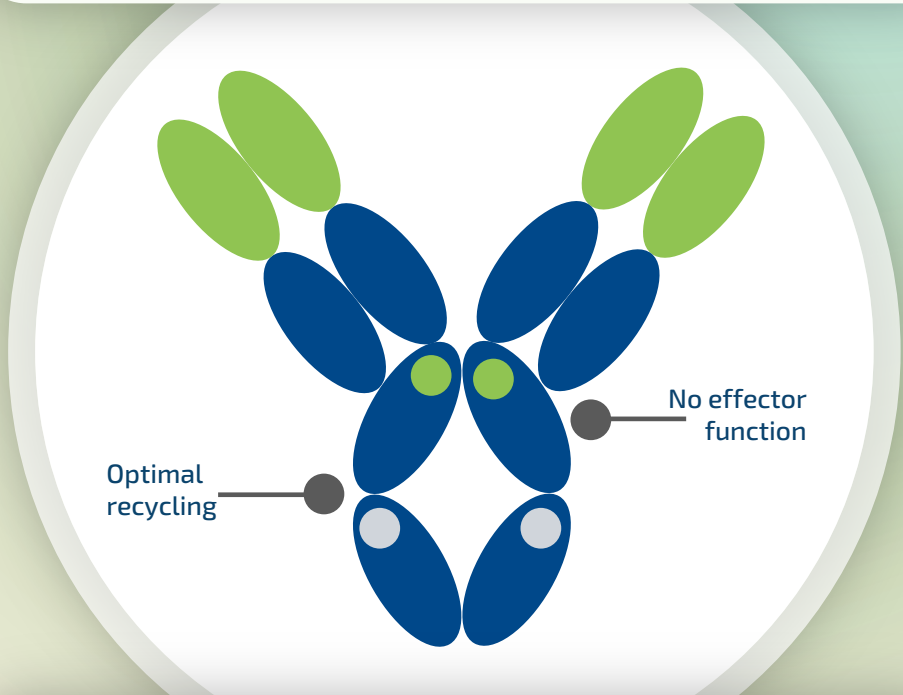
Vyvgart Hytrulo	111	107	93	80	68	56	55	48	42	40	36	36	28
Placebo	110	94	67	55	51	47	38	31	28	26	24	21	16

Empasiprubarat: Opportunity Across Multiple Autoimmune Diseases



Proof of Concept Established in MMN Our Path Forward

Anti-C2 Sweeping Antibody



Favorable safety and tolerability profile

Enrolling second cohort

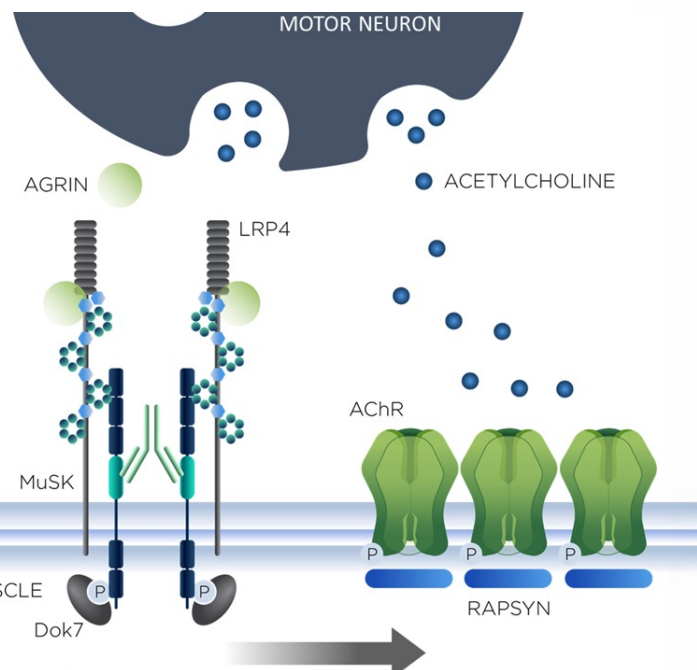
Topline ARDA results expected from both cohorts in 2024

On track to start proof-of-concept studies

Dermatomyositis	Delayed Graft Function in Kidney Transplant
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ARGX-119: MuSK Agonist with Broad Potential in Neuromuscular Disease



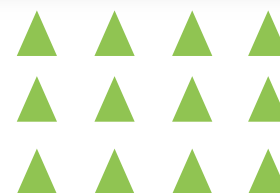
First-in-Human Phase 1 Healthy Volunteer Study

Single Ascending Dose



10 dose cohorts

Multiple Ascending Dose



3 dose cohorts / 4 weekly doses

clinical
Broaden
immunology
pipeline

First-in-
Patient

Congenital
Myasthenic
Syndrome (CMS)

First-in-patient trial in CMS to serve as proof of biology

Translational work ongoing in amyotrophic lateral sclerosis (ALS)

Positioned for Long-term Franchise Growth

Neurology

gMG, CIDP,
Myositis, TED, MMN,
CMS, Musk MG, ALS

Hematology
and
Rheumatology

ITP, Sjogren's, POTS,
Anca Vasculitis

Dermatology

Pemphigus, Bullous
Pemphigoid,
Dermatomyositis

Nephrology

Membranous
Nephropathy, Lupus
Nephritis, AMR, DGF

Innovation Ecosystem

discovery
Build out
innovation
ecosystem



Steady Cadence of Upcoming Milestones

Planned Commercial Milestones

VYVGART gMG Approval in China	✓
VYVGART gMG Approval in Canada	3Q 2023
VYVGART gMG Launches in EU	✓
VYVGART Hytrulo gMG Approval in US	✓
SC efgartigimod gMG Approval in EU	4Q 2023
SC efgartigimod gMG Approval in Japan	By 1Q 2024
VYVGART ITP Submission in Japan	✓

Planned Clinical Milestones

Efgartigimod

- ADHERE data in CIDP ✓
- ADDRESS data in Pemphigus **4Q 2023**
- ADVANCE (SC) data in ITP **4Q 2023**
- GO/NO-GO Bullous Pemphigoid (BP) **1Q 2024**
- POC data in Post-COVID POTS **1Q 2024**
- Initiate registrational trial in TED **4Q 2023**
- Initiate POC studies in ANCA and AMR **4Q 2023**

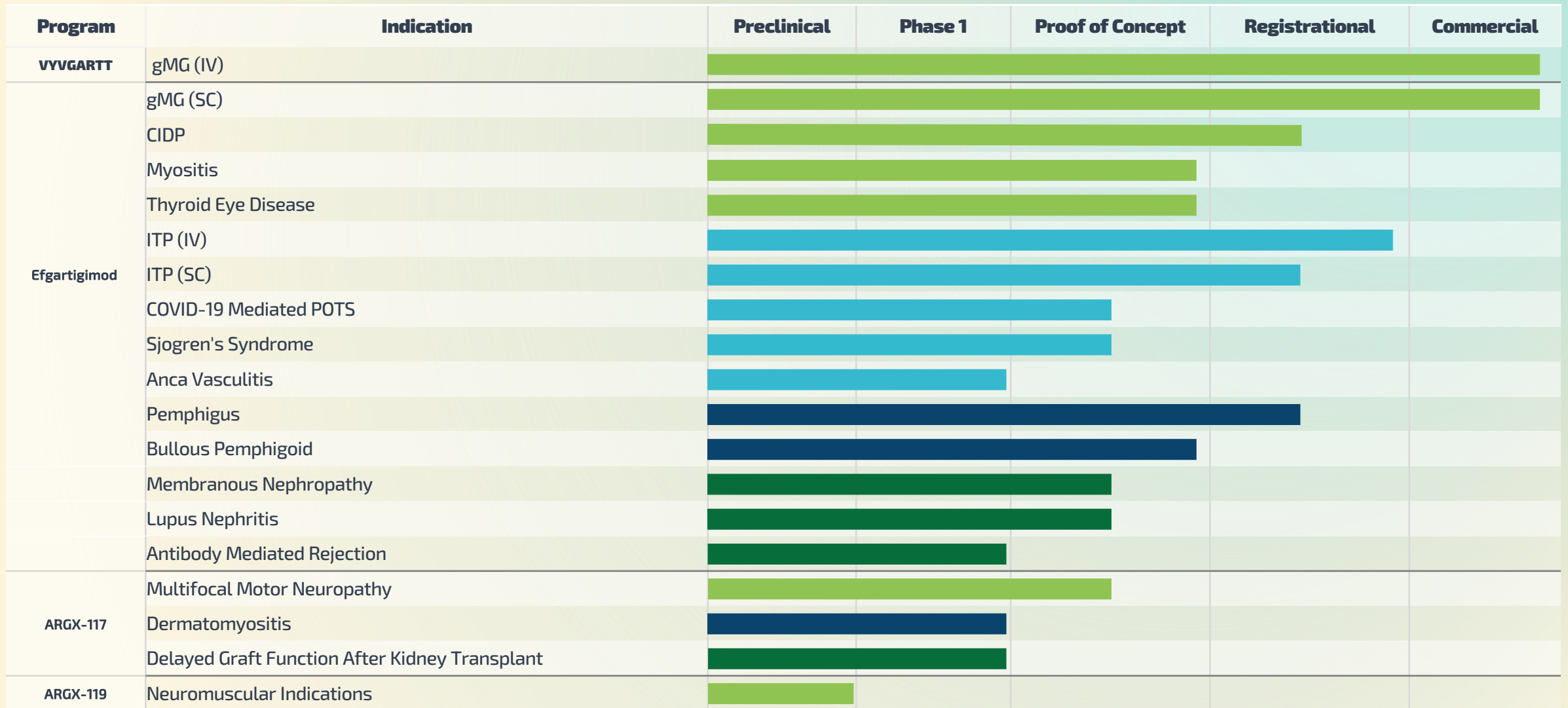
Additional pipeline

- ARGX-117: ARDA MMN interim results ✓
- ARGX-117: Initiate DGF POC study **By YE 2023**
- ARGX-119: Phase 1 study ✓

Our mission continues...



Breadth and Depth Within Autoimmune Pipeline



Key: ■ NEUROLOGY ■ HEMATOLOGY AND RHEUMATOLOGY ■ DERMATOLOGY ■ NEPHROLOGY

ITP ADVANCE-SC Trial



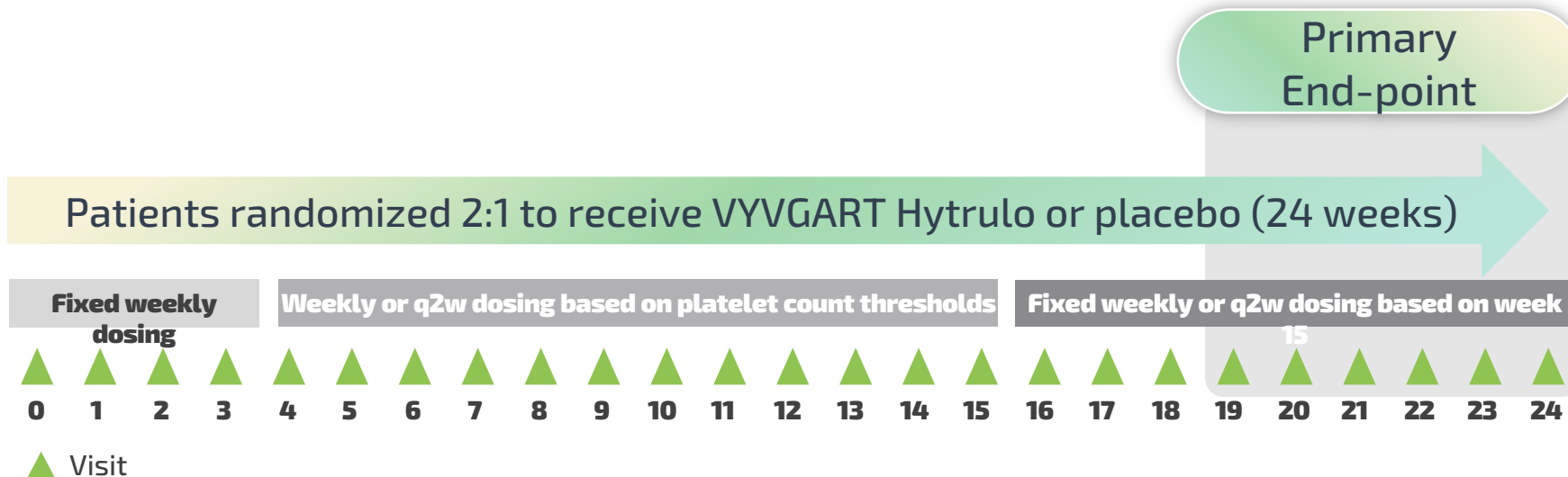
Adult Chronic or Persistent ITP patients

2 weeks screening

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

Stable concomitant ITP therapy allowed

Stratification: splenectomy, concomitant ITP therapy

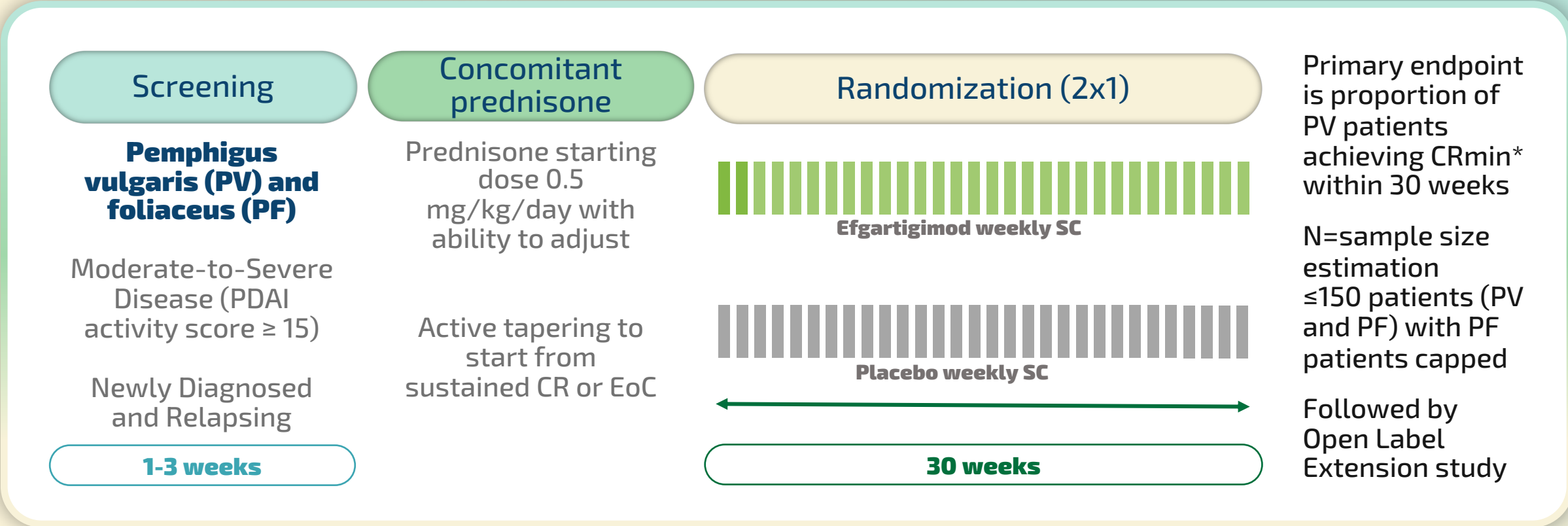


- Primary endpoint: Sustained platelet count ($\geq 50 \times 10^9/L$) in $\geq 4/6$ visits between weeks 19 and 24
- Stringent endpoint in line with regulatory feedback, addressing platelet count variability
- Secondary and exploratory endpoints centre around the extent of disease control to illustrate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

Topline data expected 4Q 2023

q2w = every other week

Pemphigus ADDRESS Trial: Focus on Fast Onset and Steroid-sparing



Topline data expected 4Q 2023

PC-POTS ALPHA Trial



42 patients

Total Duration: 24 weeks

Endpoints

Screening Criteria

Prior COVID-19 confirmed by documentation of historical PCR test

Eligibility is based on a **sustained HR increase ≥ 30 bpm** within 10 min of standing or head-up tilt testing; and/or HR reaching **> 120 bpm** within 10 min

≥ 3 vasomotor & ≥ 3 sympathetic symptoms lasting ≥ 12 w post COVID-19

COMPASS Score ≥ 35 at screening

28-day period

Randomization (2:1)



**Efgartigimod weekly IV
10mg/kg**



Placebo weekly IV



Topline data expected 1Q 2024

Co-primary endpoints

- MALMO POTS (MaPs) symptom score improvement from baseline to week 24
- Composite Autonomic Symptom Score (COMPASS) change from baseline to week 24

Secondary endpoints

- Changes from baseline to week 24 in Patient-Reported Outcomes Measurement Information (PROMIS) including fatigue and cognitive function
- Changes from baseline to week 24 in the Patient Global Impression of Severity and Change (PGI-S, PGI-C)

Followed by Open Label Extension study

Bullous Pemphigoid BALLAD Trial



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

Primary endpoint

- Proportion of participants who are in complete remission while receiving efgartigimod PH20 SC or placebo and have been off OCS for ≥ 8 weeks at week 36

Secondary endpoints

- Cumulative dose of OCS
- Proportion of participants who achieve IGA-BP score of 0 while off OCS for ≥ 8 weeks at week 36
- Proportion of participants with CDA and remain free of relapse
- Proportion of participants who are in CR while on minimal OCS (≤ 0.1 mg/kg/day) for ≥ 8 weeks at week 36
- Changes from baseline to week 36 in 24-hour average itch
- Quality of life

Followed by Open Label Extension study



Myositis ALKIVIA Trial



Screening

Stratified for

- IMNM
- Polymyositis (including ASyS)
- DM
- physician global assessment of disease activity (MDGA)

Eligibility criteria

- Muscle weakness
- Active myositis
- **MSA positive, or**
- **Historical biopsy** reviewed by Independent biopsy adjudication committee



Adults

Phase 2: 24 weeks

Separate Cohorts

Phase 2
90 Patients

Phase 3
150 Patients

Phase 3: 52 weeks

IMNM

1000mg efgartigimod SC

Placebo SC

ASyS

1000mg efgartigimod SC

Placebo SC

DM

1000mg efgartigimod SC

Placebo SC

GO/NO-GO Decision – Interim analysis of each subtype

Independent DSMB

Subtype selection

Sample size re-estimation

Proposed Endpoints

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

Empasiprubarb for Multifocal Motor Neuropathy

Advancing Phase 2 ARDA Study to Cohort 2



A phase 2, randomized, double-blind, placebo-controlled, parallel-group, multicenter study to evaluate the safety and tolerability, efficacy, pharmacokinetics, pharmacodynamics, and immunogenicity of two dose regimens of empasiprubarb (ARGX-117) in adults with multifocal motor neuropathy

