argenx R&D Spotlight

Exploring FcRn Biology and the Opportunity for Efgartigimod in Autoimmune Myositis

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A Playbook in Motion

Beth DelGiacco /// Vice President, Corporate Affairs

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Agenda

A Playbook in Motion	*Beth DelGiacco*
The Opportunity in Autoimmune Myositis	*Karen Massey*
Autoantibodies in Myositis: From Bystanders to Therapeutic Targets	*Leentje De Ceuninck*
Innovative Approach to Clinical Trial Design	*Dr. Luc Truyen*
Voices from the Frontline: Myositis Patient Journey	*Physician Panel* *Moderated by Joshua Bryson*
Commercial Opportunity	*Sandrine Piret-Gerard*
Delivering the Future	*Karen Massey*
Q&A Session	

Beth DelGiacco
VP, Corporate Affairs

Karen Massey
Chief Executive Officer

Leentje De Ceuninck, Ph.D.
Principal Scientist

Dr. Luc Truyen, M.D., Ph.D.
Chief Medical Officer

Joshua Bryson, Ph.D.
Head of U.S. Medical Affairs
& Evidence Generation

Sandrine Piret-Gerard
Chief Commercialization Officer

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Key Opinion Leaders Here Today

Avery LaChance, MD, MPH, FAAD
Harvard Medical School

Arjun Seth, MD
Northwestern Medical Group

Rohit Aggarwal, MD, MS
University of Pittsburgh

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Key Take Aways For Today

01 Autoimmune myositis is a white space opportunity aligned with our playbook: strong biology and high unmet need

02 ALKIVIA enabled a data-rich approach to our path forward: IMNM/DM opportunity

03 We are refining our US regulatory filing strategy: strength of each subtype on its own

04 IMNM/DM are strategic entry points for argenx and FcRn into rheumatology

05 Well-positioned to achieve our two goals: near-term label expansion and long-term leadership in autoimmune myositis

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The Opportunity in Autoimmune Myositis

Karen Massey /// Chief Executive Officer

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The Human Cost of Autoimmune Myositis Goes Far Beyond Muscle Weakness

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Autoimmune Myositis is a Quintessential argenx Opportunity

Clear Biology

IMNM
anti-SRP
anti-HMGCR

DM
anti-Mi-2
anti-MDA5
anti-TIF1γ,
NXP2, SAE

High Unmet Need

A Blockbuster Opportunity*

IMNM

20K

Zero approved or late-development therapies

Breakthrough Therapy Designation for IMNM

DM

40K

Heterogeneous disease; Limited treatments

*U.S. Prevalence

ALKIVIA is Designed to Unlock the Potential of Efgartigimod in Myositis

ALKIVIA: Adaptive Study in an Evolving Space

alkivia
Idiopathic Inflammatory Myopathy Study

- Built on biology insights
- Innovative trial design
- Fastest path to patients in one or more subtypes

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Two Primary Goals in Autoimmune Myositis

Label Expansion

Broaden access to reach more patients with VYVGART today

VYVGART Hytrulo®
(efgartigimod alfa and hyaluronidase-qvfc)

Long Term Leadership

Shape and expand autoimmune myositis market

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Autoantibodies in Myositis: From Bystanders to Therapeutic Targets

Leentje De Ceuninck /// Principal Scientist

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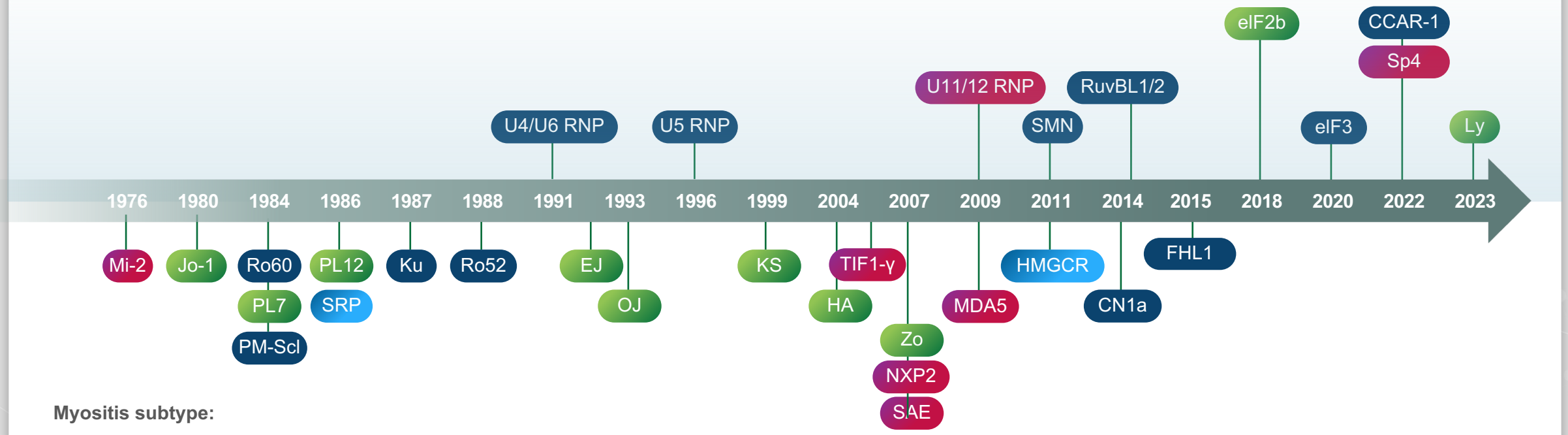
50 Years Since Identification of First Myositis-Specific Autoantibody

Hallmarks in disease diagnosis and classification



Myositis subtype:

DM | IMNM | ASyS | Other[a]

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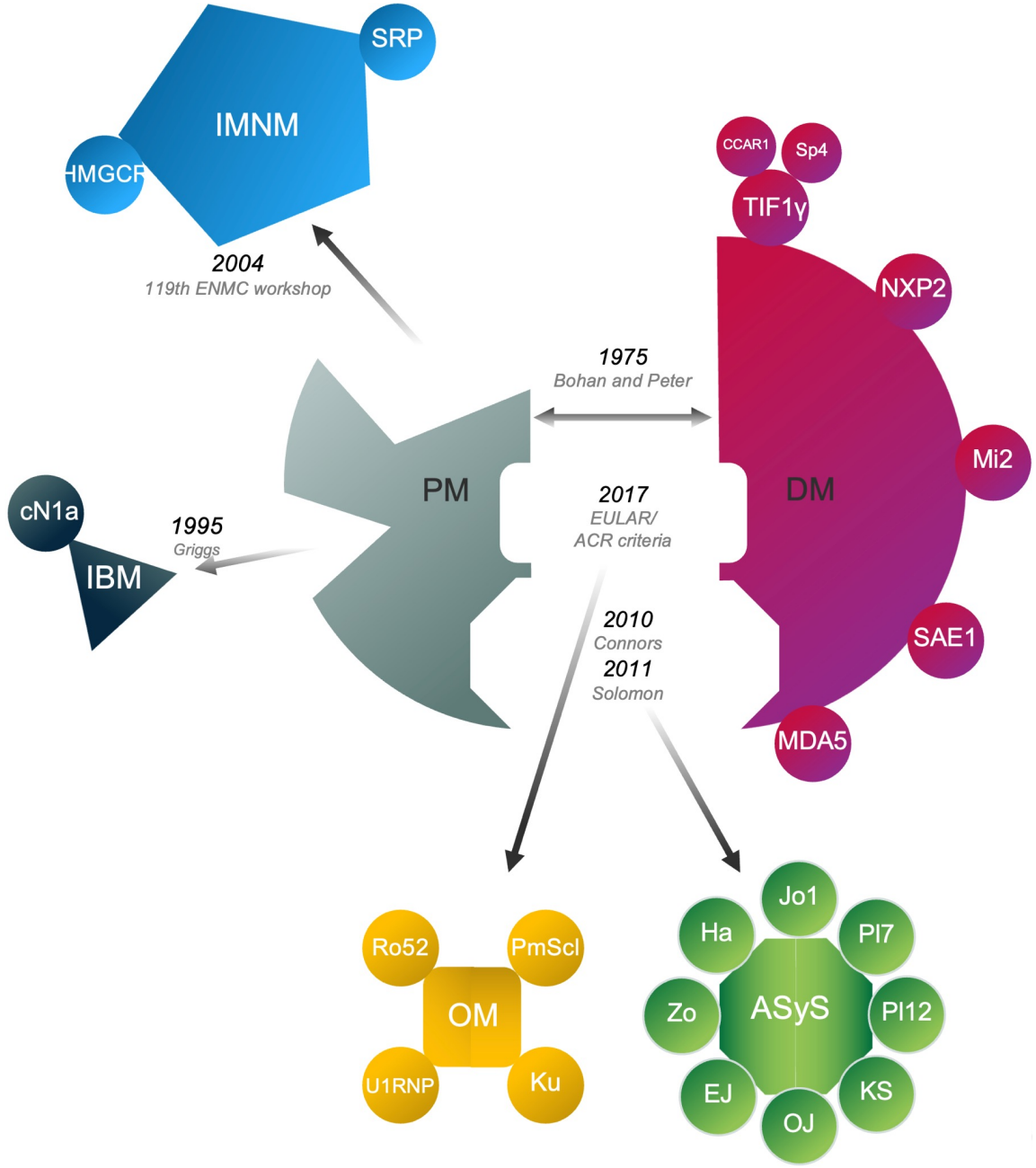
Autoimmune Myositis Increasingly Understood as a Group of Antibody and Phenotype-defined Syndromes

- POLYMYOSITIS (PM)
- DERMATOMYOSITIS (DM)
- INCLUSION BODY MYOSITIS (IBM)
- IMMUNE MEDIATED NECROTISING MYOPATHY (IMNM)
- OVERLAP MYOSITIS (OM)
- ANTISYNTHETASE SYNDROME (ASyS)



Animation is courtesy of Dr. Khoo and Dr. Chinoy

Khoo T, Lilleker JB, Thong BY, Leclair V, Lamb JA, Chinoy H.
Epidemiology of the idiopathic inflammatory myopathies.
Nat Rev Rheumatol. 2023 Nov;19(11):695-712. doi: 10.1038/s41584-023-01033-0.
Epub 2023 Oct 6.
PMID: 37803078.

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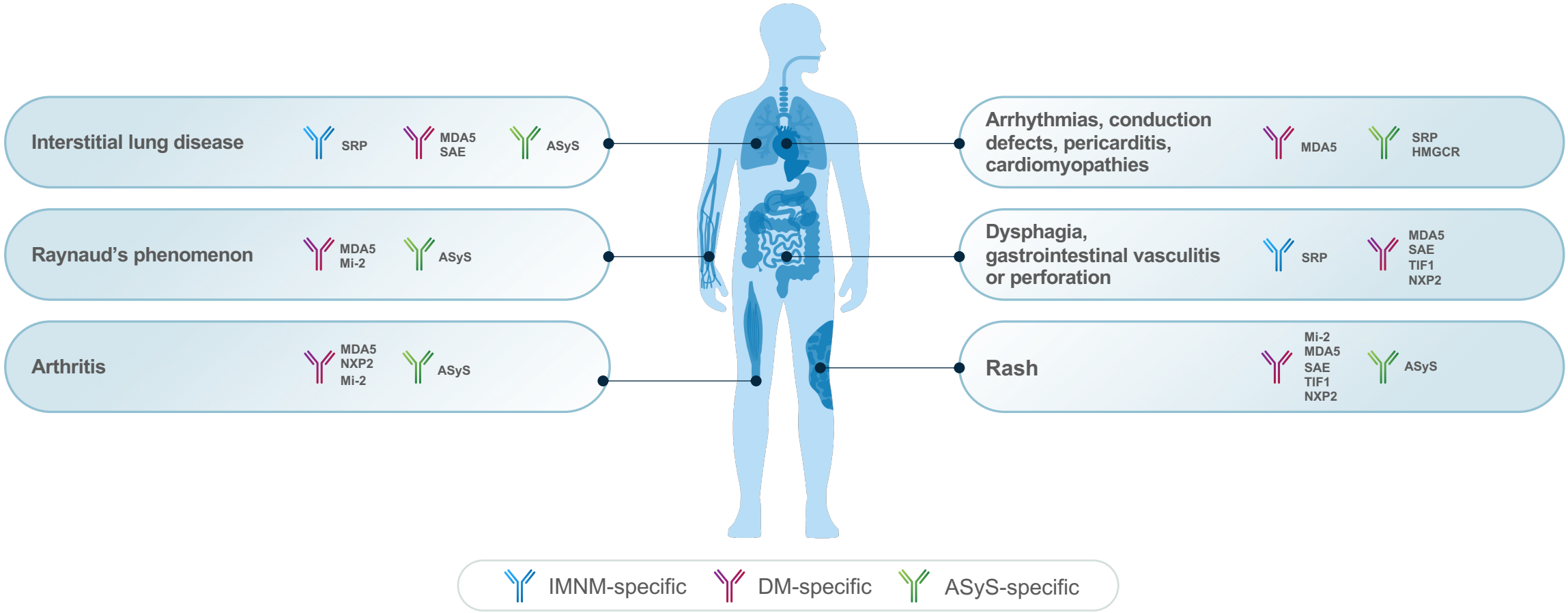
Myositis-Specific Antibodies Correlate with Distinct Clinical Phenotypes

Most Common Hallmark: Proximal Muscle Weakness



AMA, antimitochondrial antibodies; ARS, aminoacyl-transfer RNA synthetases; ASyS, antisynthetase syndrome; cN1A, cytosolic 5′-nucleotidase 1A; DM, dermatomyositis; HMGCR, 3-hydroxy-3-methylglutaryl coenzyme A reductase; IMNM, immune-mediated necrotizing myopathy; MDA5, melanoma differentiation-associated protein 5; MSA, myositis-specific antibody; NXP2, nuclear matrix protein 2; SAE, small ubiquitin-like modifier activating enzyme; SRP, signal recognition particle; TIF1, transcriptional intermediary factor 1.

*Some patients with myositis show no detectable MSAs and are considered seronegative.[2]

Figure adapted from Allameen NA, et al. 2025.[1] Used with permission from Springer Nature Limited. Copyright ©2024, Springer Nature Limited.

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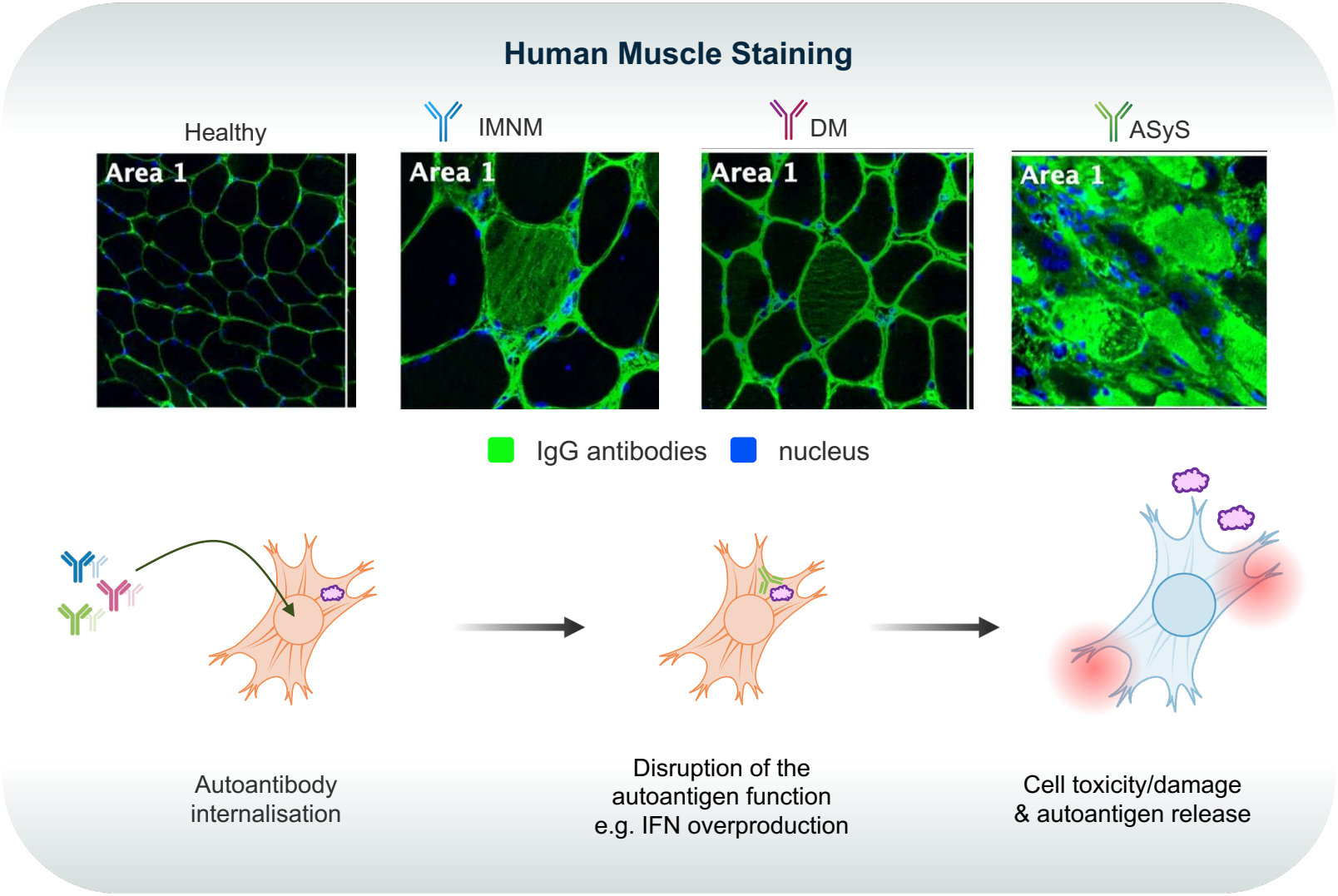
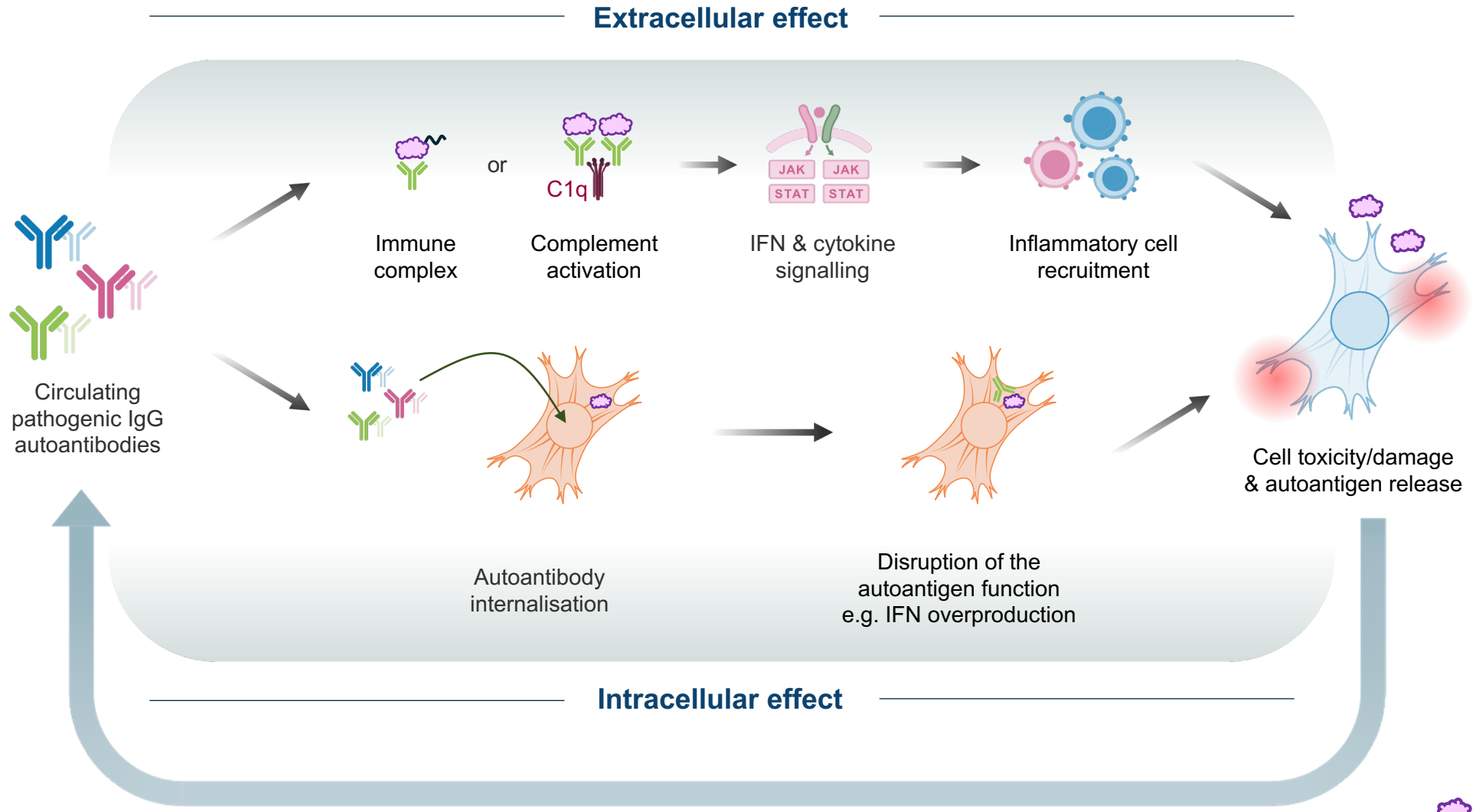
Intracellular Autoantibodies Disrupt Autoantigen Function

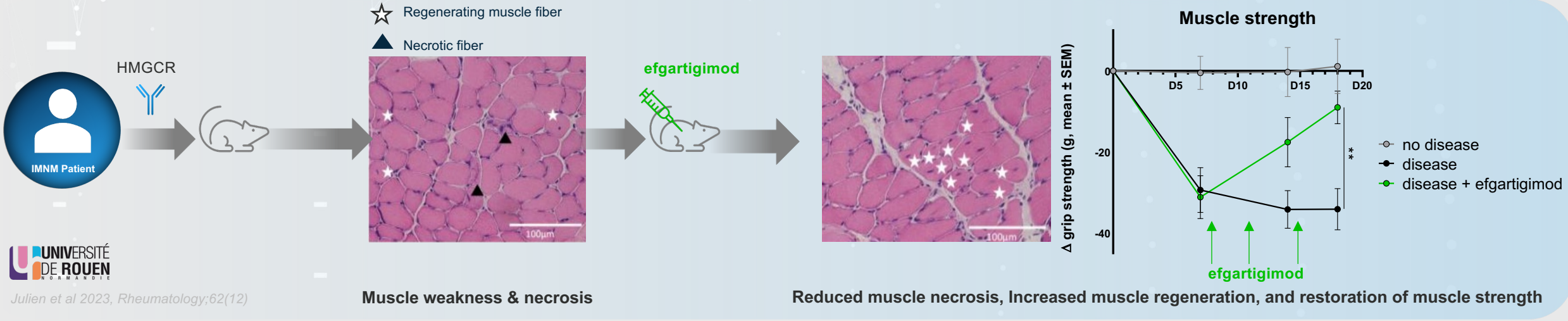


1. Musai J, et al. Curr Rheuamtol Rep. 2024;26(12):421–30; Pinal-Fernandez I, et al. Ann Rheum Dis. 2024;83(11):1549–60.

Autoantibodies in Autoimmune Myositis are Directly Pathogenic



Efgartigimod Restores Mouse Muscle Function and Reduces Lung Fibrosis

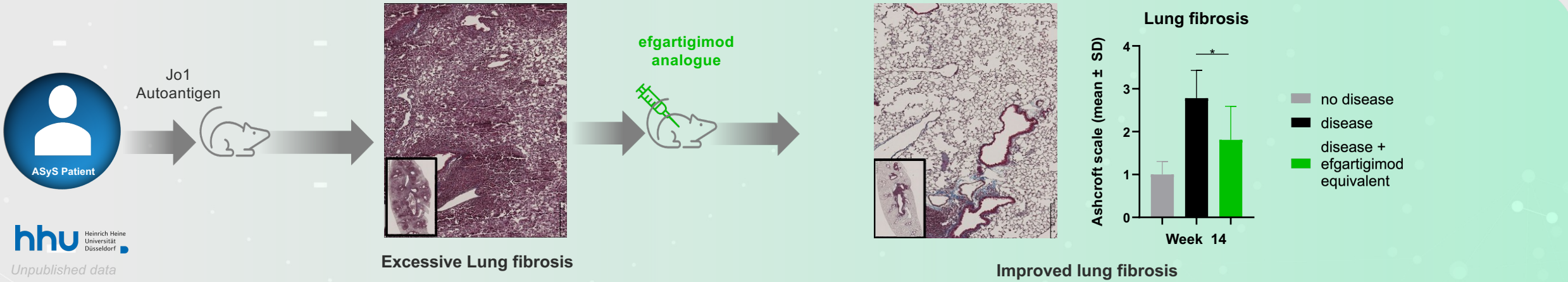


UNIVERSITÉ DE ROUEN NORMANDIE

Julien et al 2023, Rheumatology;62(12)

Muscle weakness & necrosis

Reduced muscle necrosis, Increased muscle regeneration, and restoration of muscle strength



hhu Heinrich Heine Universität Düsseldorf

Unpublished data

Excessive Lung fibrosis

Improved lung fibrosis

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Clinical Data and Biology Tell One Coherent Story

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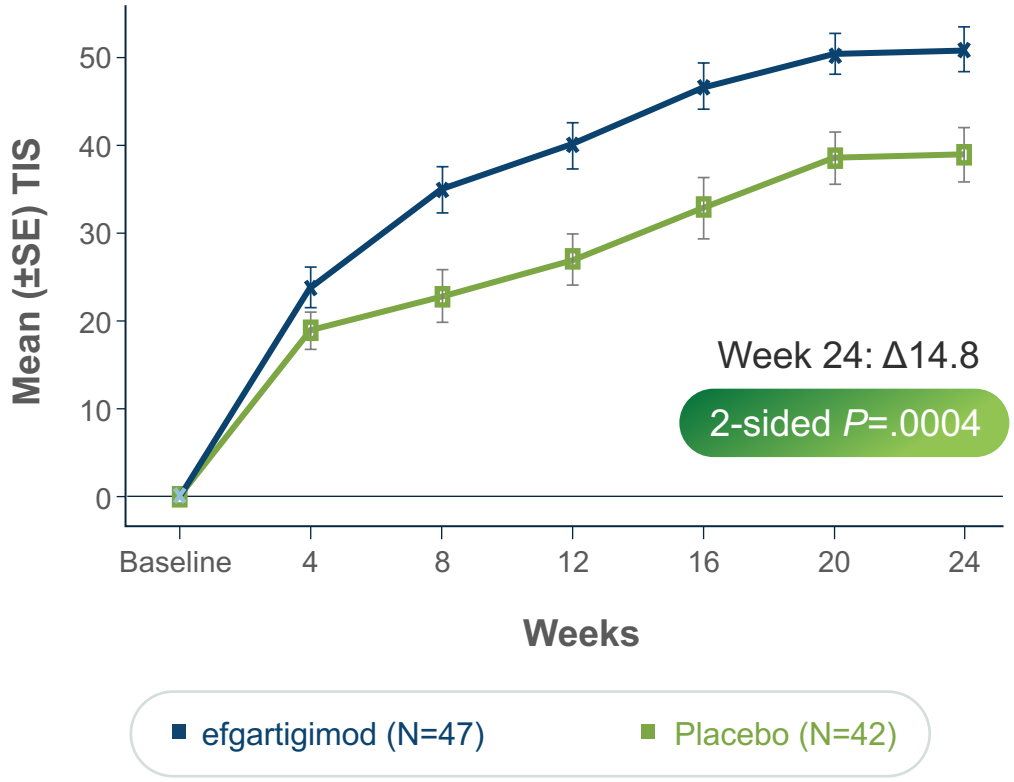
Idiopathic Inflammatory Myopathy Study

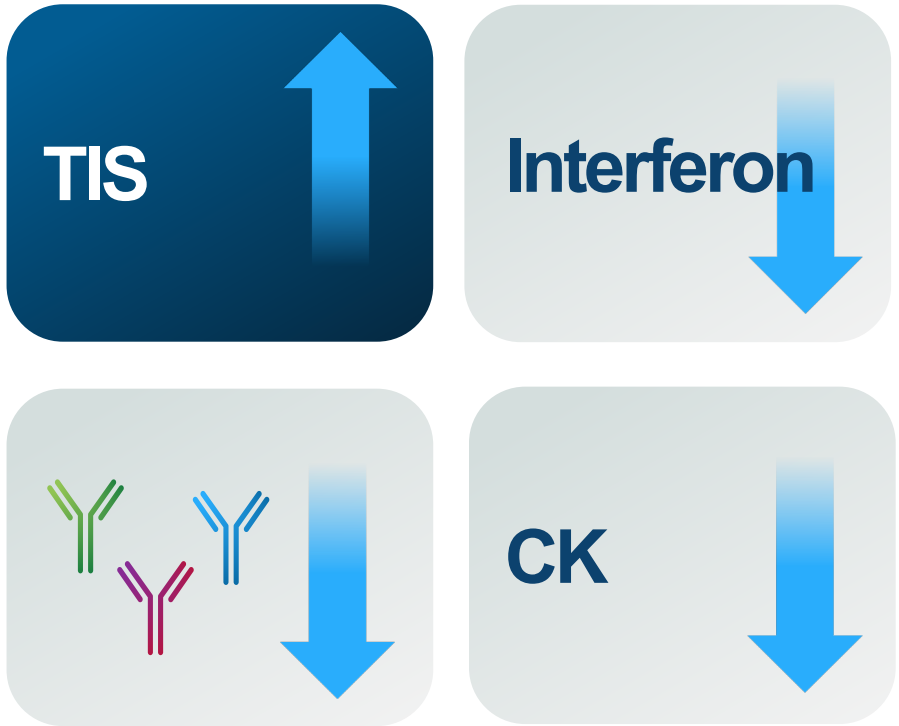
ALKIVIA Phase 2 Data

Efgartigimod Led to Significant Improvement in Mean TIS at Week 24



P-value is based on treatment policy (ANCOVA model)

Reflection of Biology in Biomarkers



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Innovative Approach to Clinical Trial Design

Luc Truyen /// Chief Medical Officer

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ALKIVIA Is Designed to Unlock the Potential of Efgartigimod in Autoimmune Myositis

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Idiopathic Inflammatory Myopathy Study

Adaptive Study

Built On Biology Insights

Increasing knowledge on IgG autoantibodies as drivers of disease

Common persistent impairment of muscle function

Delivering Data On What Patients Care About

TIS endpoint spanning 6 core set measures, including MMT8 (muscle strength)

Steroid tapering

Innovative Trial Design

Operationally seamless Phase 2/3 study

Evaluating 3 subtypes of AIM (IMNM, DM, PM)

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ALKIVIA Phase 2/3 Adaptive Basket Study

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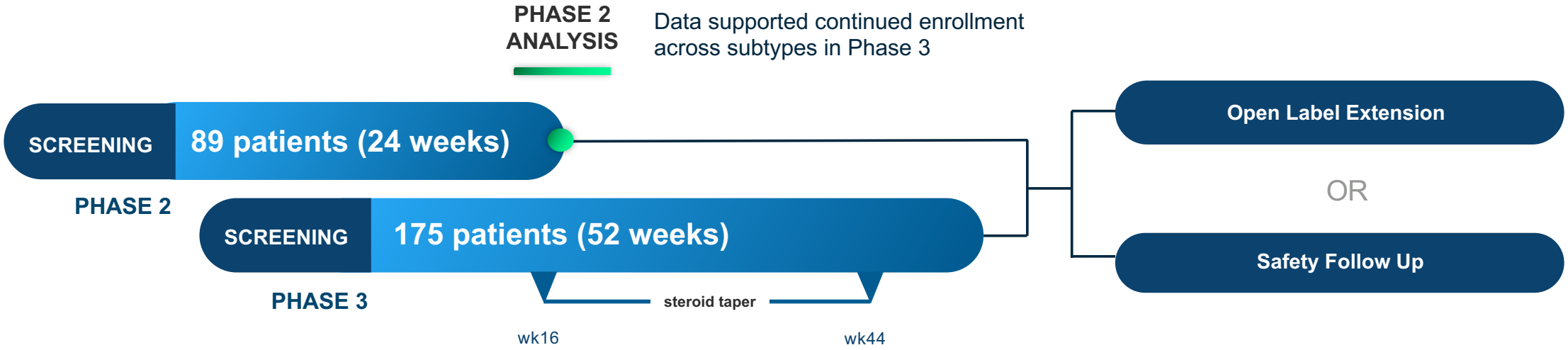
Idiopathic Inflammatory Myopathy Study



Primary endpoint: TIS (total improvement score)

TIS Core Set Measures
Capture Breadth of Response

- **Muscle strength**
- **Physical function**
- **Patient & physician global assessment**
- **Muscle enzymes**
- **Extra-muscular activity**

Trial Features

NO ENROLLMENT TARGET PER SUBTYPE
Enrollment reflects unmet need, prevalence

ACTIVE MUSCLE WEAKNESS
Despite standard of care

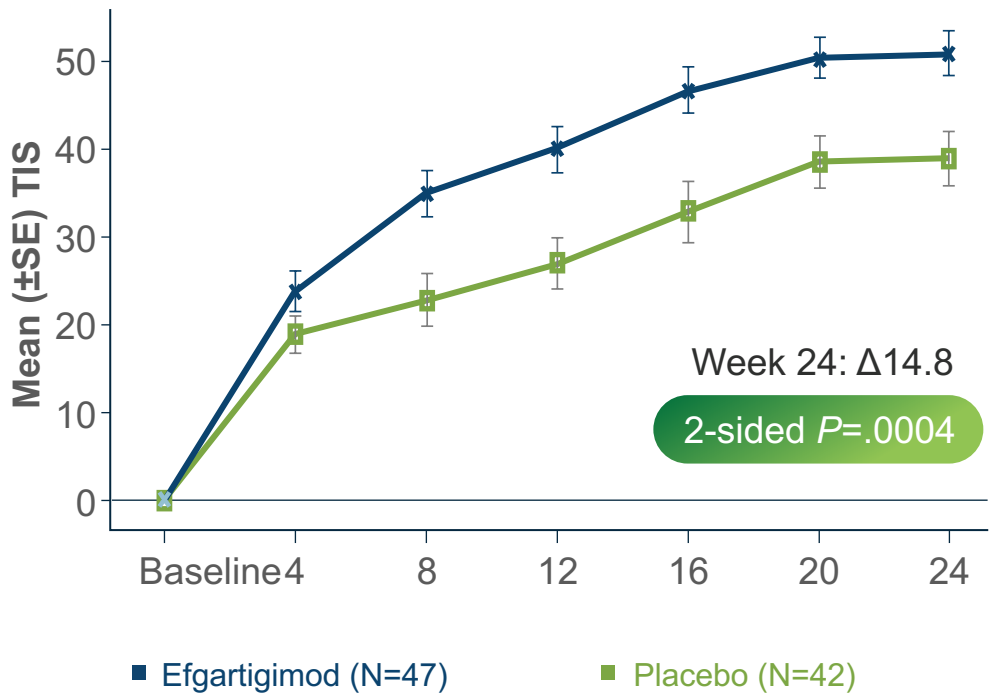
TAPERING PROTOCOL*
Supports real world demand for steroid reduction

PRIMARY ANALYSIS*†
Updated to evaluate strength of evidence of each subtype

*Phase 3 only † US

IMNM Showed Significant Improvement and DM Showed Clear Signal in Phase 2

Phase 2 Met Primary Endpoint
Mean TIS across Total Population



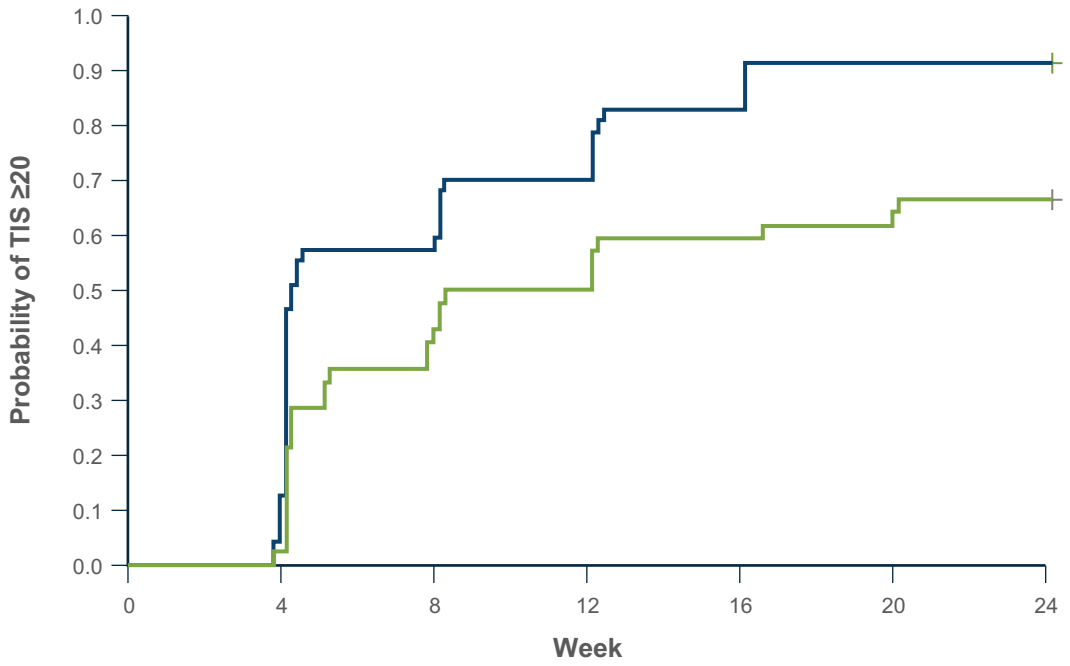
IMNM Key Contributor to Response in Phase 2

Baseline Characteristics	Phase 2
Patients (total)	89
IMNM-subtype	61%
DM-subtype	29%
PM-subtype	10%
MMT8*	120
Steroid dose (median)	10 mg
Age (mean)	56
Time since diagnosis	4-5 years
MSA+	85%

* MMT8 is a scale of 0-150 where 150 equals normal muscle function

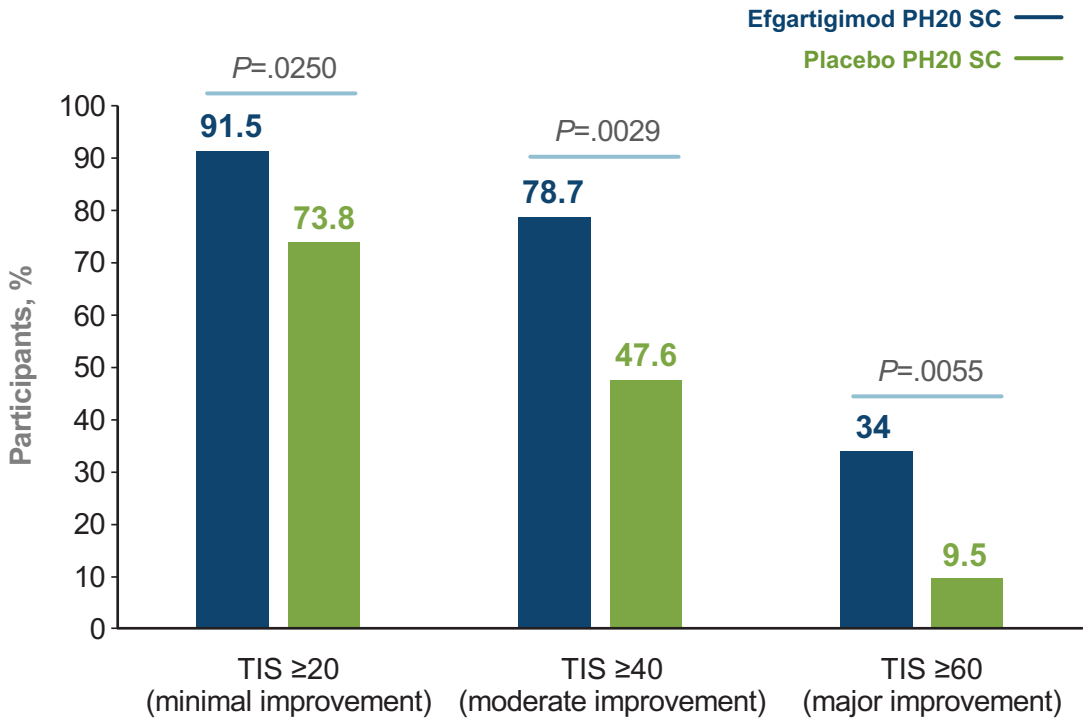
Phase 2 Results Show Efgartigimod Signature Speed and Depth of Response

Speed



Median time to TIS ≥20 was significantly shorter in efgartigimod versus placebo **(30 days vs 72 days; *P*=.0020)**

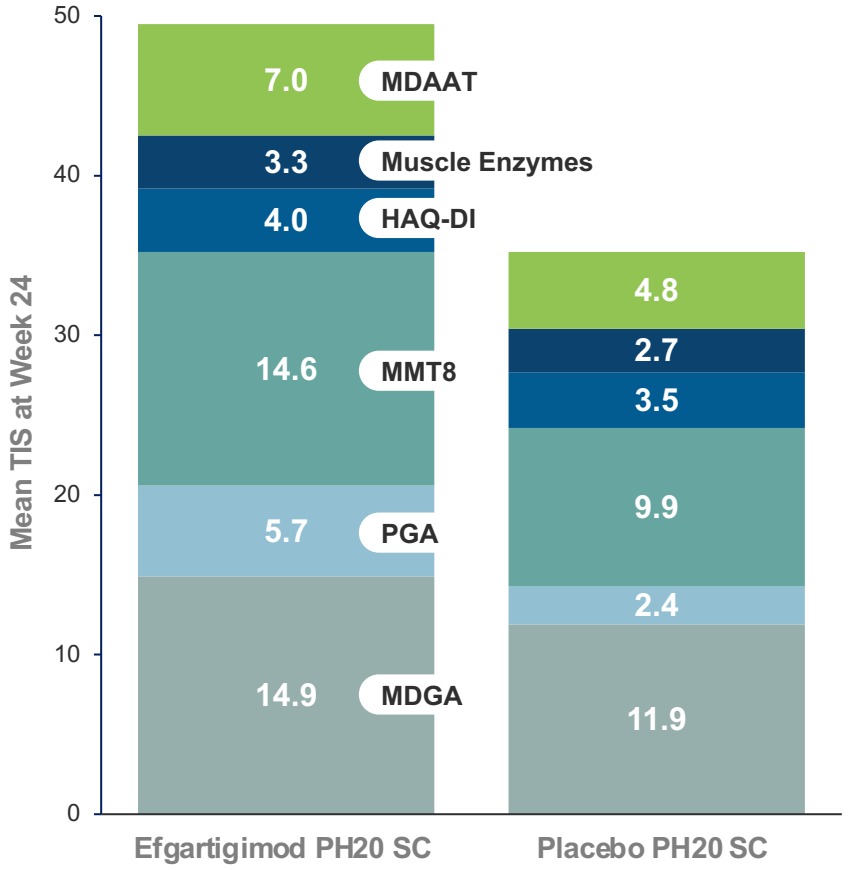
Depth of Response



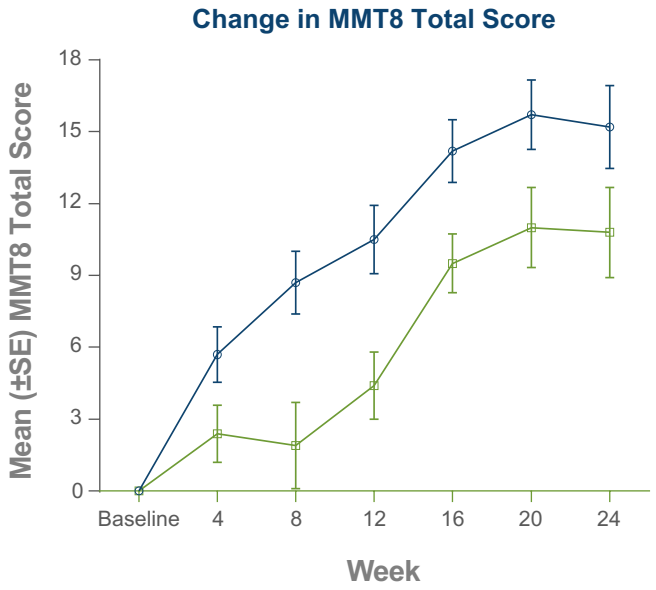
A higher proportion of patients treated with efgartigimod **had a moderate (TIS ≥40) and major (TIS ≥60) clinical improvement** at Week 24 compared with placebo

Efgartigimod Treatment Improved all 6 Core Set Measures of the TIS

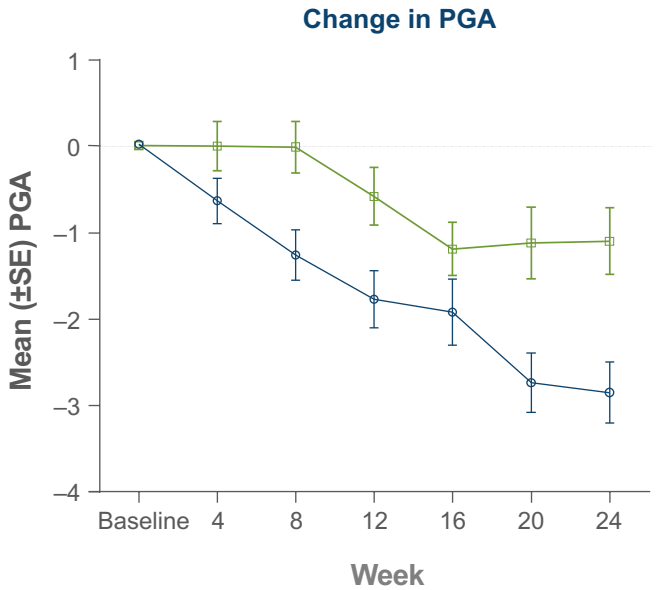
Percentage Contribution of Individual CSM to TIS at Week 24



Significant improvement in muscle strength



Significant reduction in patient-reported disease activity



Efgartigimod PH20 SC Placebo PH20 SC

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ALKIVIA Phase 2 Study Demonstrated Safety Consistent with Known Profile

	Efgartigimod PH20 SC (N=47; PYFU=22)			Placebo PH20 SC (N=42; PYFU=18)		
	n (%)	m	ER	n (%)	m	ER
≥1 AE	41 (87.2)	320	14.7	37 (88.1)	168	9.2
≥1 SAE	8 (17.0)	11	0.5	9 (21.4)	10	0.5
≥1 grade ≥3 AE	7 (14.9)	10	0.5	12 (28.6)	13	0.7
≥1 AE leading to study drug discontinuation	3 (6.4)	3	0.1	4 (9.5)	4	0.2
≥1 AESI (infection)	20 (42.6)	24	1.1	20 (47.6)	31	1.7
≥1 injection site reaction	21 (44.7)	155	7.1	9 (21.4)	41	2.2
≥1 fatal AE	2 (4.3)*	2	<0.1	0	0	0
Most common AEs (occurring in >10% of participants)						
COVID-19	4 (8.5)	4	0.2	5 (11.9)	5	0.3
Diarrhea	6 (12.8)	9	0.4	2 (4.8)	2	0.1
Injection site bruising	5 (10.6)	7	0.3	4 (9.5)	9	0.5
Injection site erythema	11 (23.4)	36	1.6	2 (4.8)	8	0.4
Injection site pain	3 (6.4)	25	1.1	5 (11.9)	14	0.8
Injection site rash	8 (17.0)	41	1.9	0	0	0
Injection site reaction	5 (10.6)	8	0.4	1 (2.4)	5	0.3
Urinary tract infection	1 (2.1)	1	<0.1	5 (11.9)	6	0.3

Participants treated with efgartigimod PH20 SC demonstrated a mean maximum IgG reduction of 72% from baseline

n, number of patients
m, number of events
ER, event rate
*Both deaths (road traffic accident and septic shock) were considered unrelated to the study drug

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Our Strategy Prioritizes Bringing Innovation to Patients as Quickly as Possible

Key Insights Gained

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Idiopathic Inflammatory Myopathy Study

- IMNM drove the response in Phase 2
- DM showed clear signal in Phase 2
- Breakthrough Therapy Designation in IMNM
- Baseline characteristics with Phase 3 distribution similar to Phase 2

Updated Filing Strategy*

Data for each subtype stands on its own

IMNM

DM

If data support, file with urgency in one or more subtypes

Why This Matters

Innovation has no meaning unless it reaches patients

IMNM: No approved treatments
DM: Opportunity to bring differentiated benefit

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*US only

Panel Discussion

Moderated by Joshua Bryson

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Avery LaChance, MD, MPH, FAAD
Harvard Medical School

Arjun Seth, MD
Northwestern Medical Group

Rohit Aggarwal, MD, MS
University of Pittsburgh

Panelists

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Joshua Bryson, PhD
Head of U.S. Medical Affairs & Evidence Generation

Moderator

Avery LaChance, MD, MPH, FAAD
Harvard Medical School

Arjun Seth, MD
Northwestern Medical Group

Rohit Aggarwal, MD, MS
University of Pittsburgh

Panelists

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Commercial Opportunity

Sandrine Piret-Gerard /// Chief Commercialization Officer

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High Unmet Need for Innovation in Autoimmune Myositis

Persistent Disability

Up to **80%**

experience persistent disability

Daily Impairment

~45%

reported work impairment

Treatment Limitations

~50%

require therapies beyond current regimens

High Steroid Use

>85%

are on steroids

Clear need for therapies that can deliver:

- Rapid onset
- Sustained efficacy (esp. muscle weakness)
- Established safety
- Steroid sparing
- Convenient administration

Sources:

- George M, Romich E, Riley TR, et al. Characteristics, treatments and outcomes of patients with dermatomyositis using real-world data.
- Christopher-Stine L, et al. BMC Rheumatol. 2025;9(1):23;
- Marie I. Curr Rheumatol Rep. 2012;14(3):275–285

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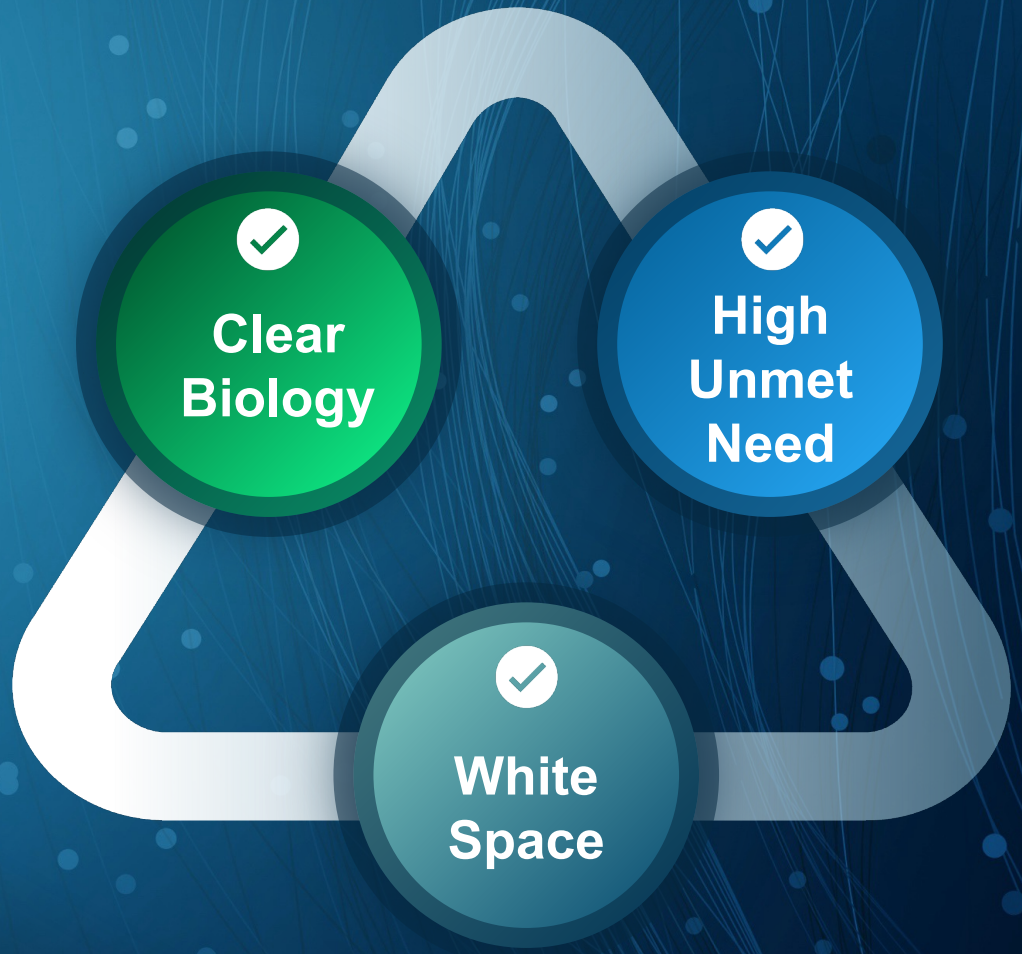
Autoimmune Myositis Fits the argenx Playbook

Sandy, Myositis Patient

Our Playbook



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Unique Opportunity to be the First and Only Therapy in IMNM

CLEAR BIOLOGY

- Strongest body of evidence of autoantibodies as disease driver

HIGH UNMET NEED

“Stop the Damage Now”

- Rapid, sustained efficacy on muscle

WHITE SPACE

- No approved treatments and no others in sight

9/10

Rheumatologists are likely to prescribe VYVGART for IMNM if approved*

* argenx market research as of 1Q 2026

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Opportunity for Differentiated Entry in Dermatomyositis

CLEAR BIOLOGY

- Broader spectrum of autoantibody involvement
- Heterogeneous disease manifestations

HIGH UNMET NEED

“Help me manage for the long haul”

- Efficacy on muscle and skin
- Safety/tolerability profile fit for long-term use

WHITE SPACE

- IVIg only approved treatment
- Different MOAs for different needs

8/10

Rheumatologists are likely to prescribe VYVGART for DM if approved*

* argenx market research as of 1Q 2026

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IMNM Alone has the Potential to be a Multi-Blockbuster Opportunity

20k
IMNM
Patients diagnosed

+

40k
DM
Patients diagnosed

+

Biologics market expansion

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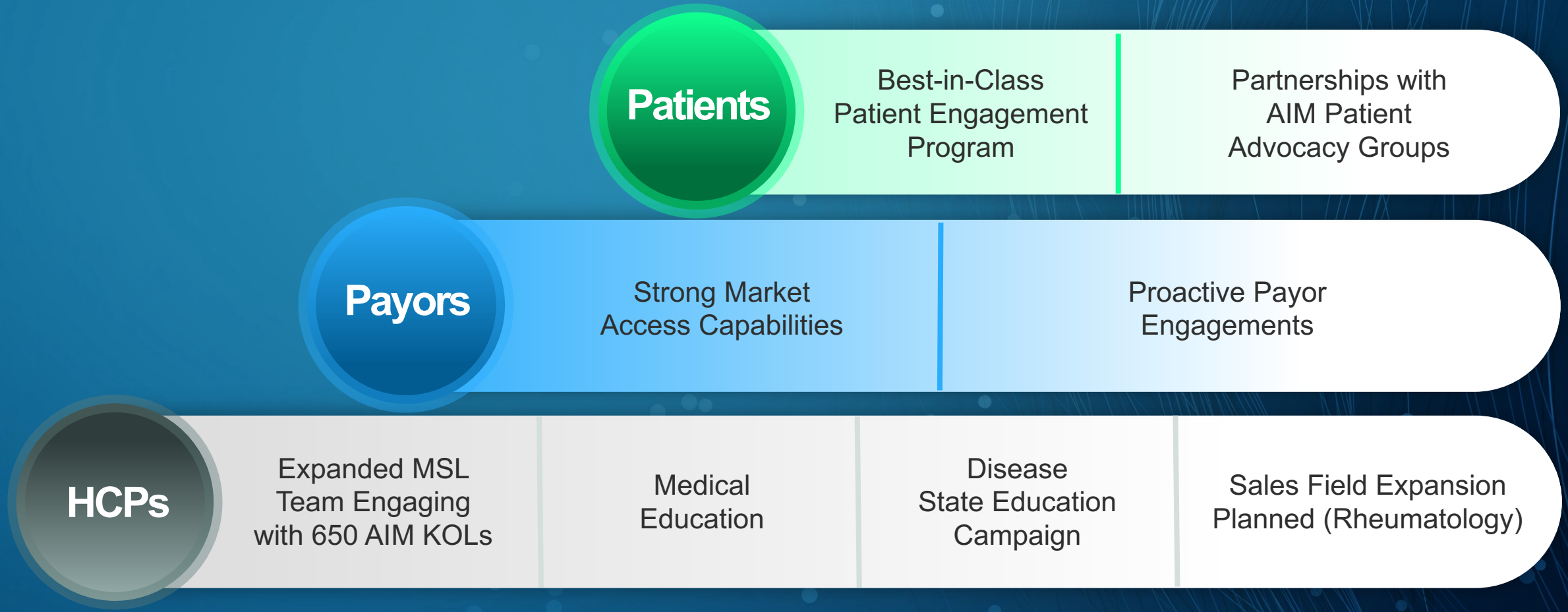
Augmenting our Commercialization Engine to Prepare for a Successful Entry into Rheumatology



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VISION 2030 is Our Engine for Long-term Growth

50k
patients on treatment

Scale VYVGART in MG and CIDP

10
labeled indications

Expand across indications and assets

5
late-stage molecules

Build durable and diversified portfolio in FcRn and beyond

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Q&A

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