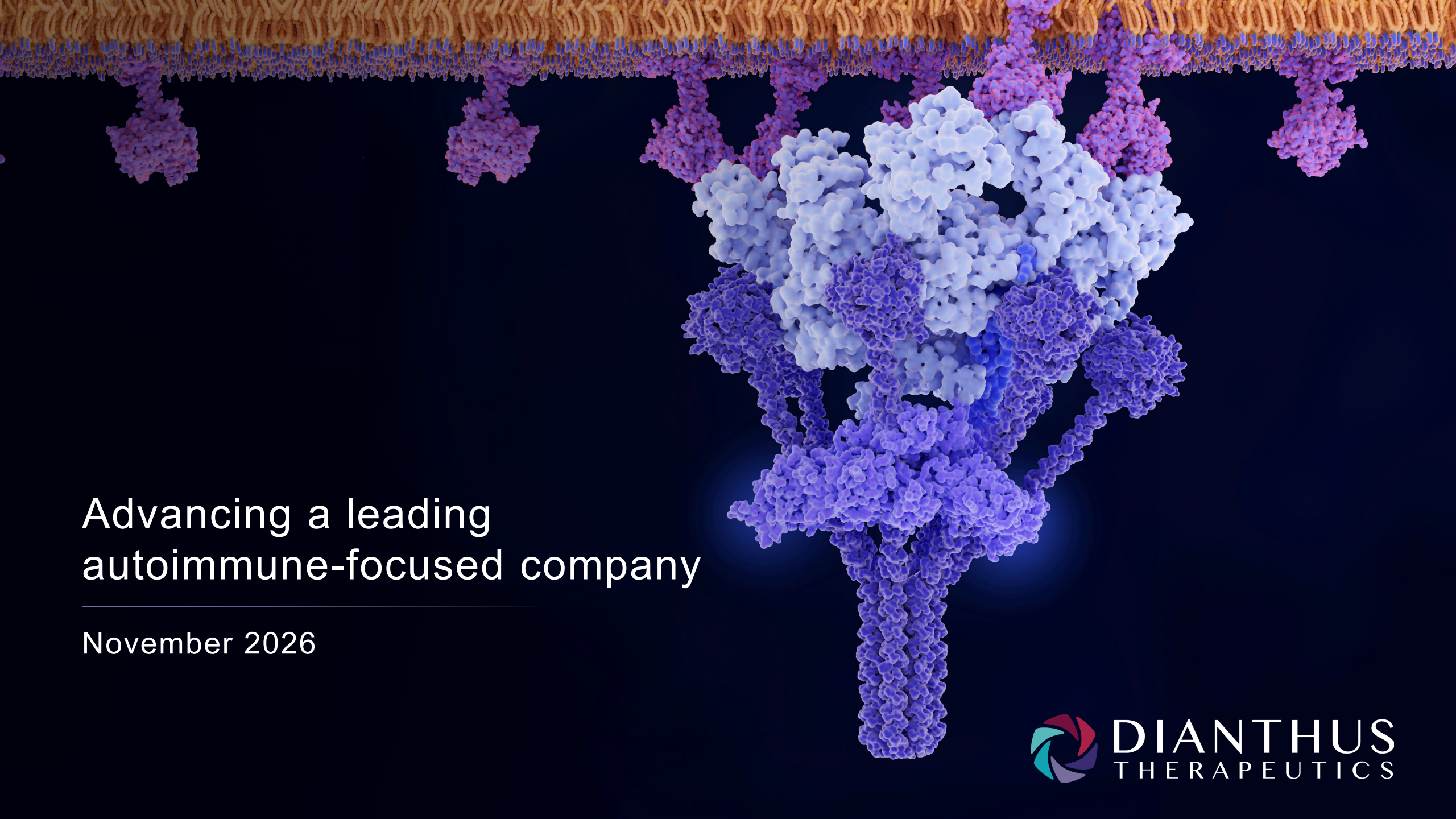


# Advancing a leading autoimmune-focused company

November 2026

DIANTHUS THERAPEUTICS

# Forward-looking statements

Certain statements in this presentation, other than purely historical information, may constitute “forward-looking statements” within the meaning of the federal securities laws, including for purposes of the safe harbor provisions under the United States Private Securities Litigation Reform Act of 1995, express or implied statements regarding future plans and prospects, including statements regarding the expectations or plans for discovery, preclinical studies, clinical trials and research and development programs, in particular with respect to claseprubart, DNTH212, and DNTH312, and any developments or results in connection therewith, including the target product profile and administration of claseprubart, DNTH212, and DNTH312; the anticipated timing of the initiation and results from those studies and trials; expectations regarding the clinical trial designs or indications; expectations regarding the time period over which the Company’s capital resources are expected to be sufficient to fund its anticipated operations; and expectations regarding market size, patient population size, and potential market opportunities, in particular with respect to claseprubart, DNTH212 and DNTH312. Claseprubart, DNTH212, and DNTH312 are investigational agents that are not approved as therapies in any indication in any jurisdiction worldwide. The words “opportunity,” “potential,” “milestones,” “runway,” “will,” “anticipate,” “achieve,” “near-term,” “catalysts,” “pursue,” “pipeline,” “believe,” “continue,” “could,” “estimate,” “expect,” “intend,” “may,” “might,” “plan,” “possible,” “predict,” “project,” “should,” “strive,” “would,” “aim,” “target,” “commit,” and similar expressions (including the negatives of these terms or variations of them) generally identify forward-looking statements, but the absence of these words does not mean that statement is not forward looking.

Actual results could differ materially from those included in the forward-looking statements due to various factors, risks and uncertainties, including, but not limited to, that preclinical testing of claseprubart, DNTH212, and DNTH312 and data from clinical trials may not be predictive of the results or success of ongoing or later clinical trials, that the preliminary interim analysis based on a limited number of patients from the Part A open-label portion of the claseprubart CAPTIVATE study in patients with CIDP may not be predictive of the results or success of the remaining patients treated in Part A or patients treated in Part B of the CAPTIVATE study, that the development of claseprubart, DNTH212, or DNTH312 may take longer and/or cost more than planned, that the Company or its partner may be unable to successfully complete the clinical development of the Company’s compounds, that the Company or its partner may be delayed in initiating, enrolling or completing its planned clinical trials, and that the Company's compounds may not receive regulatory approval or become commercially successful products. These and other risks and uncertainties are identified under the heading "Risk Factors" included in the Company’s Annual Report on Form 10-K for the period ended December 31, 2025, and other filings that the Company has made and may make with the SEC in the future. Nothing in this presentation should be regarded as a representation by any person that the forward-looking statements set forth herein will be achieved or that any of the contemplated results of such forward-looking statements will be achieved.

The forward-looking statements in this presentation speak only as of the date they are made and are qualified in their entirety by reference to the cautionary statements herein. Dianthus undertakes no obligation to publicly update or revise any forward-looking statement, whether as a result of new information, future events or otherwise, except as required by law.

# Advancing a leading autoimmune-focused company with neuromuscular and rheumatology franchises

## Advancing a portfolio of autoimmune therapeutics with best-in-disease, pipeline-in-a-product potential and targeting patient-friendly, infrequent S.C. self-administration

### Claseprubart: Neuromuscular Franchise
*(aC1s mAb)*

- **Highly potent, ~8-week half-life, Classical Pathway (CP) inhibitor targeting active C1s**
- **Validated pipeline-in-a-product** potential with **positive Ph. 2 gMG results, early** Interim Responder Analysis **GO decision** in **Ph. 3 CIDP,** and clinical **PoC for CP inhibition in MMN**
- **Clinical and *in vitro* head-to-head data** support potential for a more **effective and convenient** biologic with no Boxed Warning/REMS
- Targeting **convenience** of a **single, self-administered S.C. 300mg/2mL autoinjector dosed every 2 or 4 weeks**

***Claseprubart milestones***:
**gMG:** Ph. 3 gMG top-line data (2H'28)
**CIDP:** Ph. 3 Part A top-line data (YE'27), Part B top-line data (YE'28)
**MMN:** Ph. 2 MMN top-line data (Dec. '26); Initiation of Ph. 3 MMN trial (2027)

### DNTH212: Rheumatology Franchise
*(BDCA2 and BAFF/APRIL bifunctional fusion protein)*

- **Bifunctional BDCA2 and BAFF/APRIL inhibitor** targeting two **validated** pathways
- Potential for **enhanced efficacy** from complementary mechanisms **targeting innate and adaptive immune systems**
- Demonstrated **superior *in vitro* pDC depletion** vs. litifilimab and **superior serum Ig inhibition** vs. povetacicept in NHPs
- **Pipeline-in-a-product opportunity across multiple diseases** with potential for **Q4W or less frequent S.C. self-administration**
- **Initial priority indications selected: SjD, SLE and DM; dosed first SLE patient** in **Ph. 1 Part B** in October '26

***DNTH212 milestones:***
Ph. 1 Part A SAD healthy volunteer preliminary data (Q1'27)
Ph. 1 Part B SLE patient preliminary data (2H'27)
Ph. 1 DNTH-led MAD initiation (Q2'27) and preliminary MAD data (2H'27)

## DNTH312 (next-generation claseprubart and TACI bifunctional fusion protein) expands our leadership across severe autoimmune diseases

Designed to deliver **potent aC1s and BAFF/APRIL inhibition** by targeting complementary, validated pathways, with **best-in-disease, pipeline-in-a-product potential** across multiple severe autoimmune diseases. DNTH312 expected to be **Ph. 1 ready by YE'27**

## Strong financial position with cash of ~$1.2B[1] and runway expected into 2030 to fund multiple near and long-term catalysts

SjD: Sjögren's Disease; SLE: Systemic Lupus Erythematosus; DM: Dermatomyositis; SAD: single ascending dose; MAD: multiple ascending dose
1. Cash, cash equivalents and investments as of September 30, 2026

# Two clinical-stage candidates with best-in-disease, pipeline-in-a-product potential

| | Claseprubart | | | DNTH212 |
|---|---|---|---|---|
| **Indications** | **gMG** | **CIDP** | **MMN** | **SjD, SLE and DM** |
| | ***>100,000 U.S. patients*** | ***>40,000 U.S. patients*** | ***>10,000 U.S. patients*** | **~625,000 U.S. patients** |
| **Market Insight** | Multi-billion $, growing market with opportunity for a best-in-disease, convenient therapy to expand first-line biologics use | Active C1s inhibition has shown robust efficacy in patients who were refractory, stable, and naïve to IVIg | Empasiprubart, a C2 inhibitor, demonstrated impressive efficacy in MMN, validating Classical Pathway inhibition | Targets both innate and adaptive immune systems, with superior *in vitro* pDC depletion vs. litifilimab and superior serum Ig inhibition vs. povetacicept in NHPs |
| **Our Opportunity** | Ph. 2 data support potential for best-in-disease profile demonstrating rapid, robust, continuous symptom control with convenient, Q2W or Q4W S.C. dosing and a potentially differentiated safety profile | Observed 75% response rate from first 40 participants to complete CAPTIVATE Part A, supporting best-in-disease potential | Demonstrated superiority vs. empasiprubart in head-to-head *in vitro* Classical Pathway potency experiment, with potential to be a best-in-disease therapeutic with limited competition | Validation of both BDCA2 and BAFF/APRIL targeted therapies support bifunctional approach, with potential for best-in-disease efficacy |
| **Next Milestones** | **Ph. 3 Top-line Data in 2H'28** | **Ph. 3 Part A Top-line Data by YE'27; Part B Top-Line Data by YE'28** | **Ph. 2 Top-line Data in Dec. '26; Ph. 3 to Initiate in 2027** | **Ph. 1 Part A HV Data in Q1'27; Ph. 1 Part B SLE Data in 2H'27; MAD to Initiate in Q2'27, Data in 2H'27** |

gMG: >100,000 gMG U.S. patients from Komodo claims data accessed 2013-2025; approx. 85% of gMG patients have AChR autoantibody-driven disease https://www.ncbi.nlm.nih.gov/pmc/articles/PMC7033452/#
CIDP & MMN: Komodo claims data 2013-2025, adjusted to account for 70% capture of real-world patient counts for biologic treated patients; CIDP adjusted to account for 27% misdiagnosed
Estimated SjD, SLE and DM U.S. patients per Dianthus meta-analysis and estimates

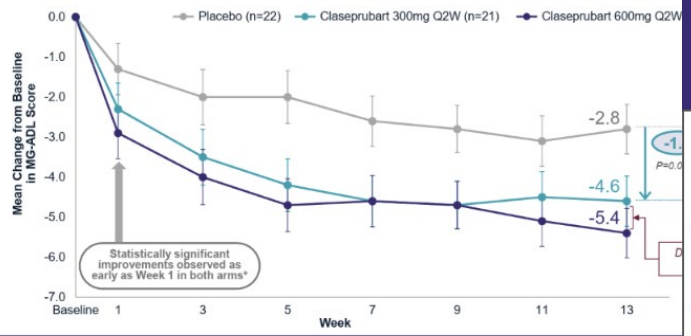
# Claseprubart:
# Building a Best-in-Disease Neuromuscular Franchise

# Building on the promise of claseprubart to be a potential pipeline-in-a-product and best-in-disease therapy in growing and underserved markets

## Ph. 2 MaGic Showed Rapid & Robust Improvements in gMG

Q3'25

**Claseprubart arms demonstrated rapid, sustained, and clinically meaningful improvements in MG-ADL score**




MG-ADL improvements for participants treated with cl
were rapid, sustained, clinically meaningful and statistically signifi

The change from baseline in MG-ADL was analyzed using a mixed effect model for repeated measures (MMRM) with treatment group, visit, treatment by visit interaction, stratification fac
standard error of the mean.
*One-sided p-values are presented for comparisons of claseprubart vs placebo, with any p-value below 0.1 considered nominally statistically significant.

## CAPTIVATE Part A Early GO Decision in CIDP

Q1'26

**Announced early GO decision reached with less than 40 planned participants completing Part A in M**

| | |
|---|---|
| CAPTIVATE Interim Analysis Objective | Targeting response rate of 50% or greater (≥20 patien Part A) based on precedent set with aC1s inhibition |
| GO Decision | GO decision reached early after 20 confirmed respon than 40 planned participants completing Part A |
| Safety / Tolerability Update | Independent DSMB reviewed the data to date and cor no related serious infections, no clinical symptoms o no related SAEs or discontinuations |

GO decision supports continued development of claseprubart at 30
targeting a potentially best-in-disease biologic

CAPTIVATE Interim Analysis data cut off as of March 4, 2026
Drug-Induced Lupus (DIL) is an autoimmune syndrome triggered by specific medications, such as statins, TNF-alpha inhibitors, and ACE inhibitors/beta-blockers. A key distinguishing
typically resolve once the offending medication is withdrawn.

## Ph. 2 MoMeNtum Top-line Data Anticipated Dec. '26

Dec. '26

**MoMeNtum is a small PoC Ph. 2 in MMN primarily focused on safety, top-line data anticipated in December '26**

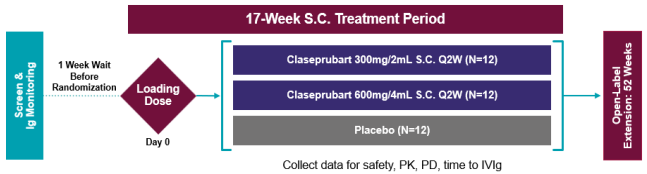
Efficacy endpoints, such as Grip Strength, are secondary exploratory endpoints

**Highlights**

- **Design:** 36 participants randomized to receive either claseprubart or placebo for 17 weeks
- **Inclusion:** ≥18 years old with MMN who are immunoglobulin responsive and dependent
- **Dosing:** I.V. Loading Dose followed by 300mg/2mL or 600mg/4mL S.C. Q2W starting Day 7
- **No ANA screening exclusion criteria or routine ANA testing** during the RCT or OLE

**Endpoints**

- **Primary:** Safety
- **Secondary:** Efficacy (time to IVIg retreatment, time to relapse, grip strength and other muscle strength and motor function measurements)




Collect data for safety, PK, PD, time to IVIg retreatment, time to relapse, grip strength and other muscle strength and motor function measurements

MOMENTUM

**Phase 2 MoMeNtum trial of claseprubart in MMN exceeded its original enrollment target of 36 patients; top-line results with 46 patients on track for December '26**

MoMeNtum Trial: https://clinicaltrials.gov/study/NCT06537999

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# Claseprubart has opportunity to compete as a potential 1L biologic in three large and growing US neuromuscular markets

## >100K patients

**AChR+ gMG patients**

Potential for US growth as only <20% of AChR+ patients treated with biologics

MAGIC — **Positive Ph. 2 data reported Sept. ’25**

## >140K patients

**>40K CIDP patients**

Potential for aC1s inhibition to transform treatment landscape

CAPTIVATE — **Early GO decision announced Mar. ’26**

## >150K patients

**>10K MMN patients**

PoC supporting Classical Pathway inhibition and limited competition

MOMENTUM — **Ph. 2 top-line data expected in Dec. ’26**

**Claseprubart has potential to capture meaningful market share across three synergistic multi-billion-dollar markets**

Figures represent U.S. estimated patients only. gMG: >100,000 gMG U.S. patients from Komodo claims data accessed 2013-2025; approx. 85% of gMG patients have AChR autoantibody-driven disease https://www.ncbi.nlm.nih.gov/pmc/articles/PMC7033452/#

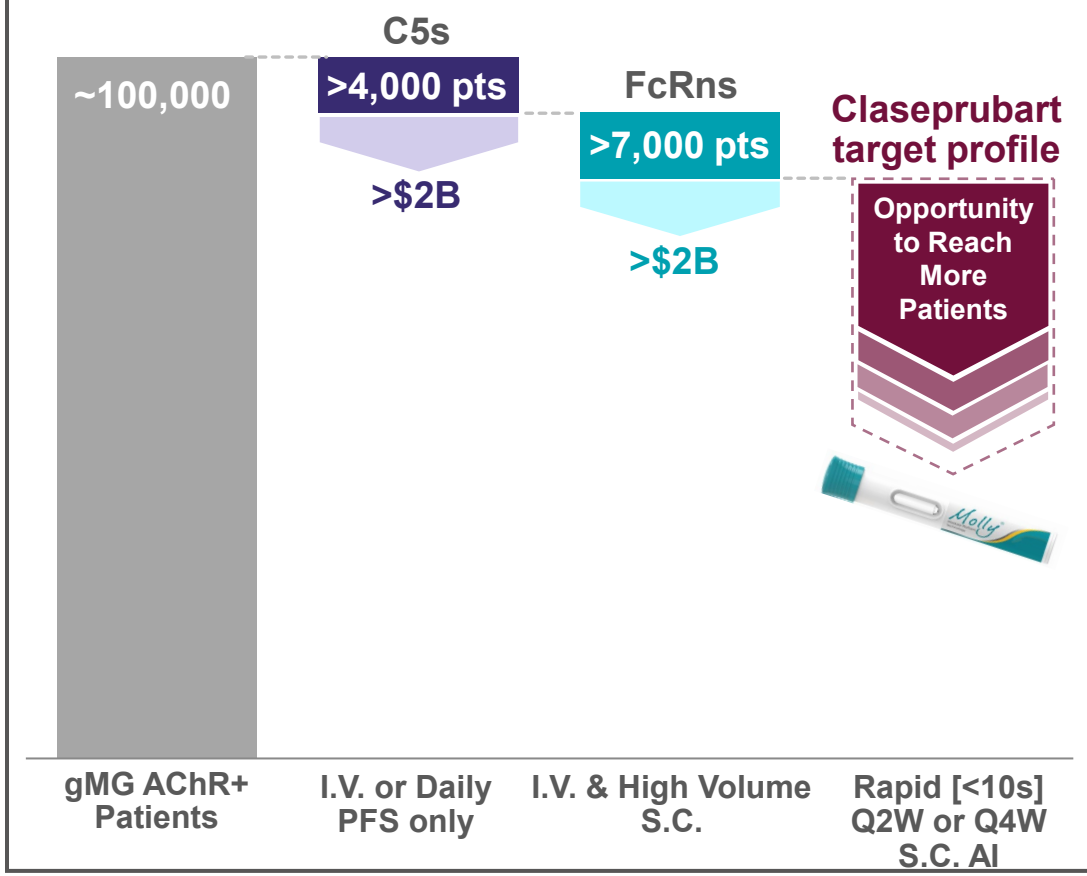
CIDP & MMN: Komodo claims data 2013-2025, adjusted to account for 70% capture of real-world patient counts for biologic treated patients; CIDP adjusted to account for 27% misdiagnosed

# Claseprubart: Opportunity to be a Best-in-Disease, First-Line Biologic for Generalized Myasthenia Gravis

# The US gMG market has significant potential to expand as <20% of AChR+ patients[1] are treated with biologics

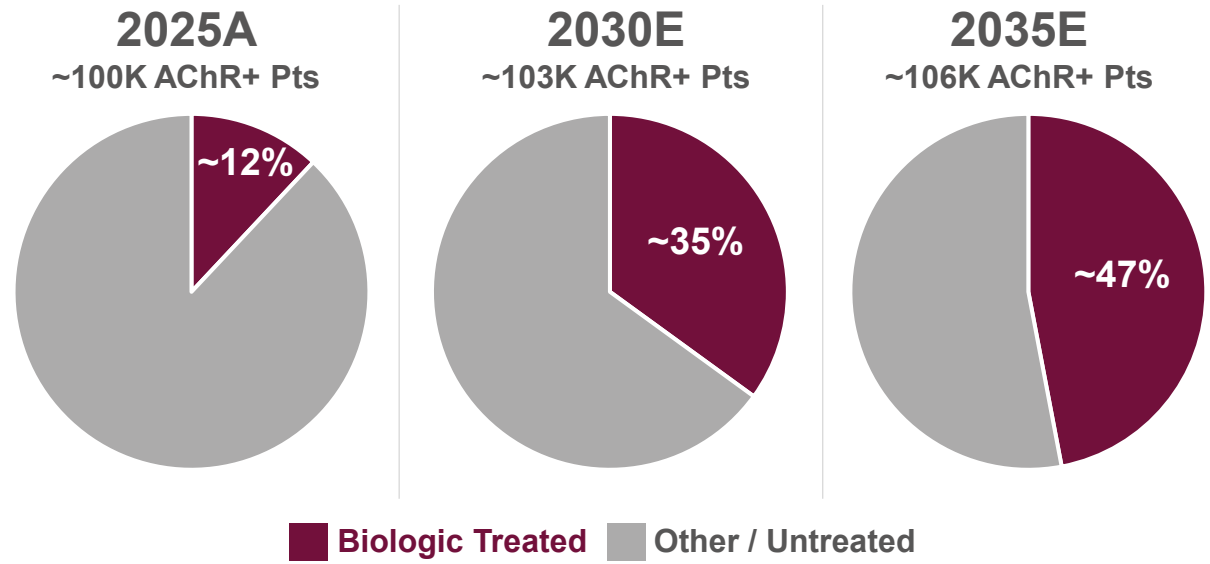
Significant opportunity for a highly differentiated, more patient-friendly biologic to expand use of biologics in gMG

## Current US gMG Market




## >$8B gMG Opportunity for Complement in 2035[2]

### Estimated US biologic penetration[3,4]




### Estimated US biologic sales[3,4]

| 2025A | 2030E | 2035E |
|---|---|---|
| ~$4B | ~$14B | ~$20B |
| ~12K pts | ~36K pts | ~50K pts |

Note: Positioning of claseprubart's target profile is illustrative. 1. Komodo claims data accessed 2013-2025, adjusted to account for 70% capture of real-world patient counts; AChR+ 85% of gMG. 2. Based on Dianthus market research and estimates. 3. 2025 US financial reports on gMG drugs. Soliris/Ultomiris adjusted for relative size of MG based on claims. Vyvgart adjusted for estimated CIDP sales. 4. Based on EvaluatePharma (Jan '26) and Dianthus market research and estimates. Assumes average biologic net price per patient of ~$400,000 per year. AstraZeneca 2024 Investor Day estimated >36,000 AChR+ treated patients in future state. AstraZeneca Q2'25 earnings transcript expected 50% of gMG patients to be on branded treatments by 2028.

# gMG biologic market growth driven by the introduction of new treatment options, despite limitations

## Continued Unmet Need Shown by Patient Switching and New Therapy Uptake

**50%** of the gMG market is anticipated to consist of branded biologics, driven by differentiated efficacy, safety and more convenient options[1]

**40%** of patients discontinue Vyvgart within one year, highlighting continued unmet need and the opportunity for differentiated long-term treatment approaches[2]

**20%** of Vyvgart franchise growth is being driven by uptake of Hytrulo as patients transition from IV to a complex (up to 90 seconds, multi-step injection) S.C. option[3]

**10%** projected market share for Uplizna in its first full year of launch, underscoring physician willingness to adopt a new therapy, despite a gradual onset of efficacy[4]

1. EvaluatePharma (Jan '26) and Dianthus market research and estimates. AstraZeneca Q2'25 earnings transcript expected 50% of gMG patients to be on branded treatments by 2028. 2. Komodo closed claims data (2022-2025); persistence assessed during periods of continuous patient claims visibility. 3. Komodo closed claims data (2024-2026) and argenx financial disclosures. 4. Komodo closed claims data (Q1 2026) and Amgen Q1/Q2 2026 earnings reports.

# Survey of US Neurologists confirms significant opportunity for differentiated biologic in gMG market

## Claseprubart aims to address the significant unmet needs in the gMG market

### Total Neurologists 81

| | |
|---|---|
| Neuromuscular specialists | 67% |
| Generalists | 33% |
| Academic | 40% |
| Community based | 60% |

### Sample Demographics

**~17** years in active clinical practice (post-residency), on average

**~93%** of professional time spent providing direct patient care, on average

**~40** gMG patients seen in the past 12 months, on average

**~81%**

of Neurologists believe patients would benefit from treatment options with **greater durability of symptom relief**

**~78%**

of Neurologists believe patients would benefit **from a more convenient treatment option**

**~67%**

of Neurologists would start more patients on a complement inhibitor if they **do not have a boxed warning or REMS requirement**

**~72%**

of Neurologists **prefer low-volume autoinjector** over high-volume prefilled syringe **due to ease of use & faster injection (i.e. <10 seconds)**

Dianthus 2025 Neurologist MG quantitative survey (N=81, N = 54 Neuromuscular specialists, N = 27 General Neurologists), fielded Q2

# Achieving a TPP that addresses market needs has the potential to be best-in-disease and grow the gMG market



**RAPID ONSET WITH DURABLE AND CONSISTENT EFFICACY**

Improve symptoms within one week and maintain efficacy between doses



**NO BOXED WARNING OR REMS**

Well-tolerated with comparable safety to FDA-approved C1s inhibitor Enjaymo



**RAPID SELF-ADMINISTERED AUTOINJECTOR CONVENIENCE**

Comparable convenience, simplicity, and speed to Dupixent 2mL autoinjector

# MaGic Ph. 2 Results Support a Potentially Best-in-Disease Treatment for gMG

# MaGic Ph. 2 top-line results support a potentially best-in-disease treatment for gMG

## Efficacy Endpoints

**Strong results support claseprubart potential as a best-in-disease complement inhibitor**

- Rapid, sustained, statistically significant symptom improvements as measured by MG-ADL, QMG, MSE, MGC, MG-QoL-15r

## Safety Endpoints

**Generally well tolerated, with a potentially differentiated safety profile**

- No encapsulated bacterial infections
- No symptoms indicative of autoimmune activation or DIL
- Supports no Boxed Warning or REMS

## Optimal Dose

**Comparable efficacy & safety across both 300mg/2mL and 600mg/4mL doses**

- Target dose of 300mg/2mL Q2W will be in Ph. 3 study
- Supports convenient, infrequent, self-administration with same autoinjector as Dupixent

**MaGic Ph. 2 results support a profile with the potential to displace C5 complement inhibitors and compete effectively with FcRns as first-line biologic treatment in growing MG market**

Autoinjector for claseprubart administration is anticipated to be SHL Medical's Molly technology, patented or patent pending in the US, China, India, Japan, Korea, Taiwan and at the European Patent Office.
C5 efficacy data are derived from different clinical trials conducted at different times, with differences in trial design and patient populations.
Drug-Induced Lupus (DIL) is an autoimmune syndrome triggered by specific medications, such as statins, TNF-alpha inhibitors, and ACE inhibitors/beta-blockers. A key distinguishing feature of DIL is its reversibility, as symptoms typically resolve once the offending medication is withdrawn.

# MaGic is a global Ph. 2 trial in AChR+ gMG patients

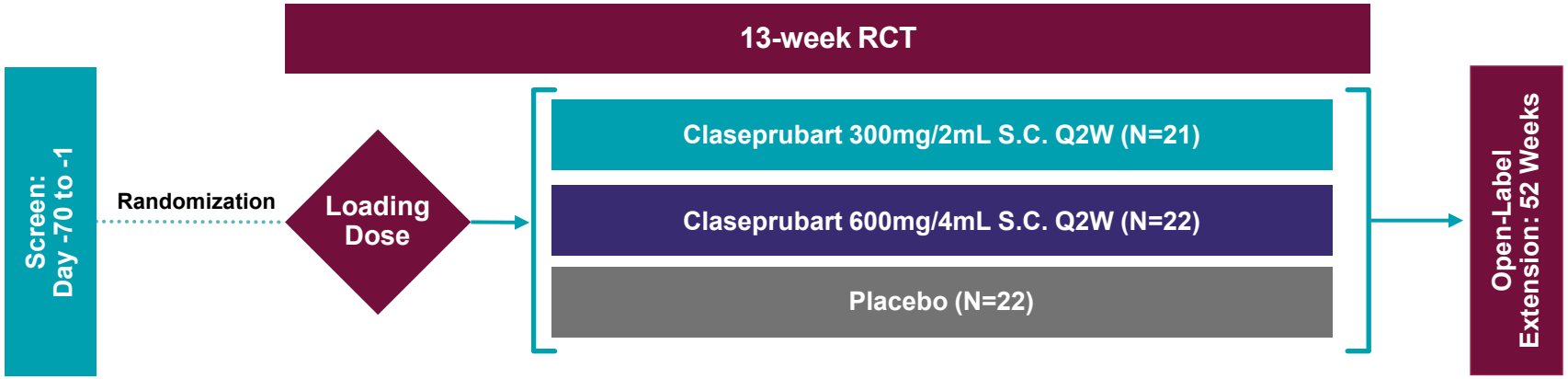
A global, multicenter, randomized, double-blind, placebo-controlled study to evaluate the safety, efficacy, and PK / PD of claseprubart administered S.C. following initial loading dose

### Highlights

- **Design:** ~60 male and female subjects randomized to receive either claseprubart or placebo for 13 weeks
- **Inclusion:** ≥18 years old with AChR antibody + gMG
- **Dosing:** 15 or 20mg/kg I.V. Loading Dose followed by 300mg/2mL or 600mg/4mL S.C. Q2W starting Day 7

### Endpoints

- **Primary:** Safety
- **Secondary / Exploratory:** Efficacy (MG-ADL, QMG, MSE, MGC, MG-QoL-15r)



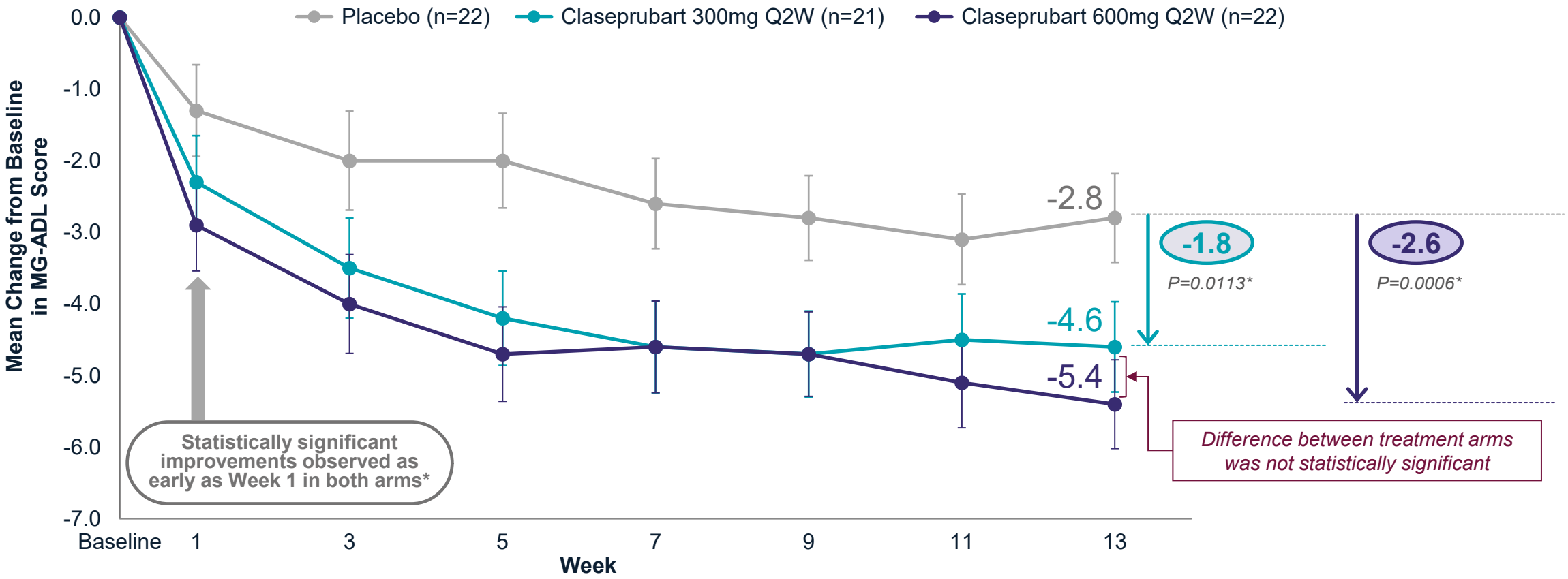

MAGIC

**Trial enrollment exceeded target, with 65 participants enrolled**



MaGic Trial: https://clinicaltrials.gov/study/NCT06282159

# Claseprubart arms demonstrated rapid, sustained, and clinically meaningful improvements in MG-ADL score

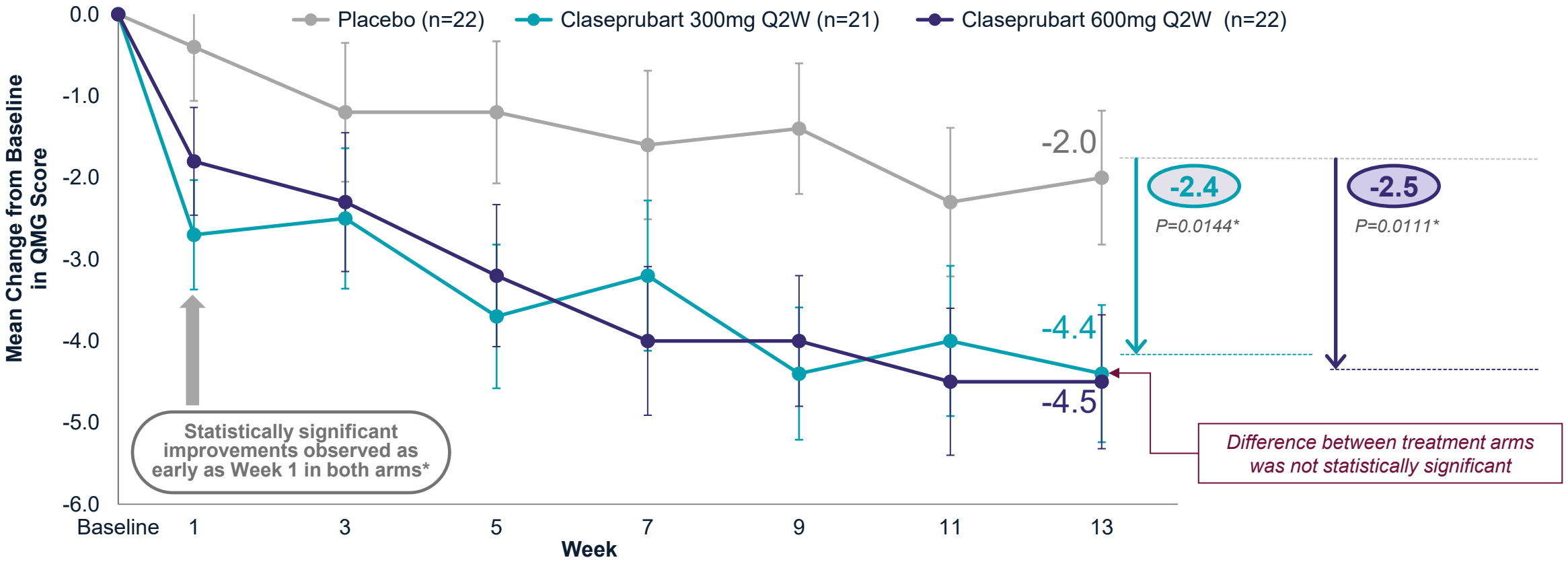



**MG-ADL improvements for participants treated with claseprubart were rapid, sustained, clinically meaningful and statistically significant as early as Week 1**

The change from baseline in MG-ADL was analyzed using a mixed effect model for repeated measures (MMRM) with treatment group, visit, treatment by visit interaction, stratification factors, and baseline measure included. Bars represent standard error of the mean.

*One-sided p-values are presented for comparisons of claseprubart vs placebo, with any p-value below 0.1 considered nominally statistically significant.

# Claseprubart arms demonstrated rapid, sustained, and clinically meaningful improvements in QMG score

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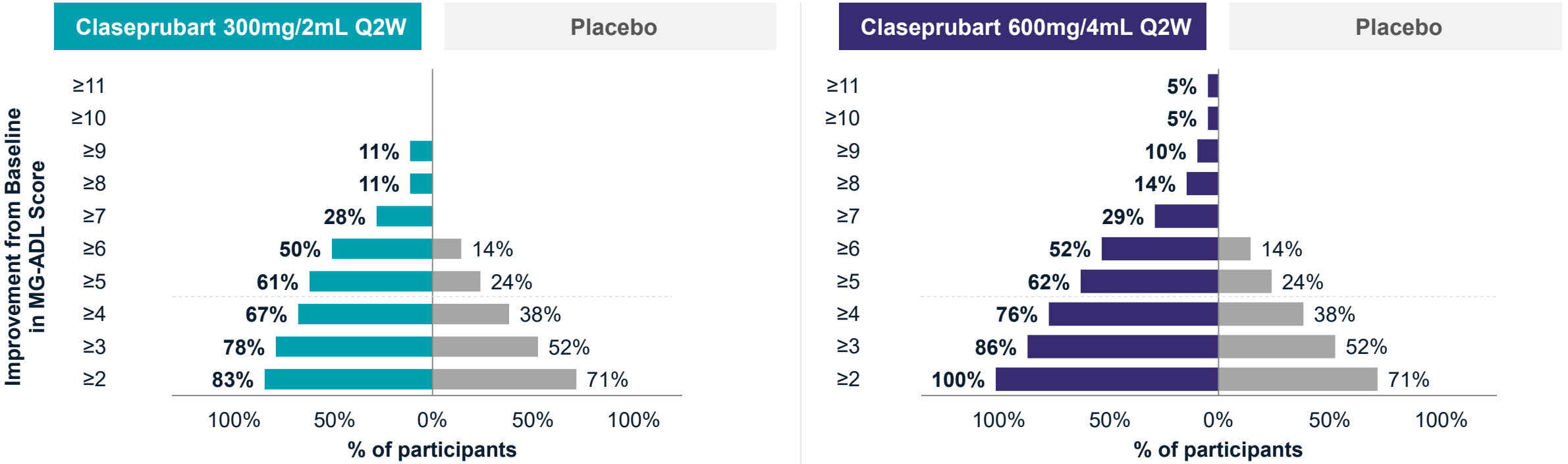
*One-sided p-values are presented for comparisons of claseprubart vs placebo, with any p-value below 0.1 considered nominally statistically significant.

# >60% of participants on claseprubart 300mg/2mL achieved ≥5 point improvement in MG-ADL

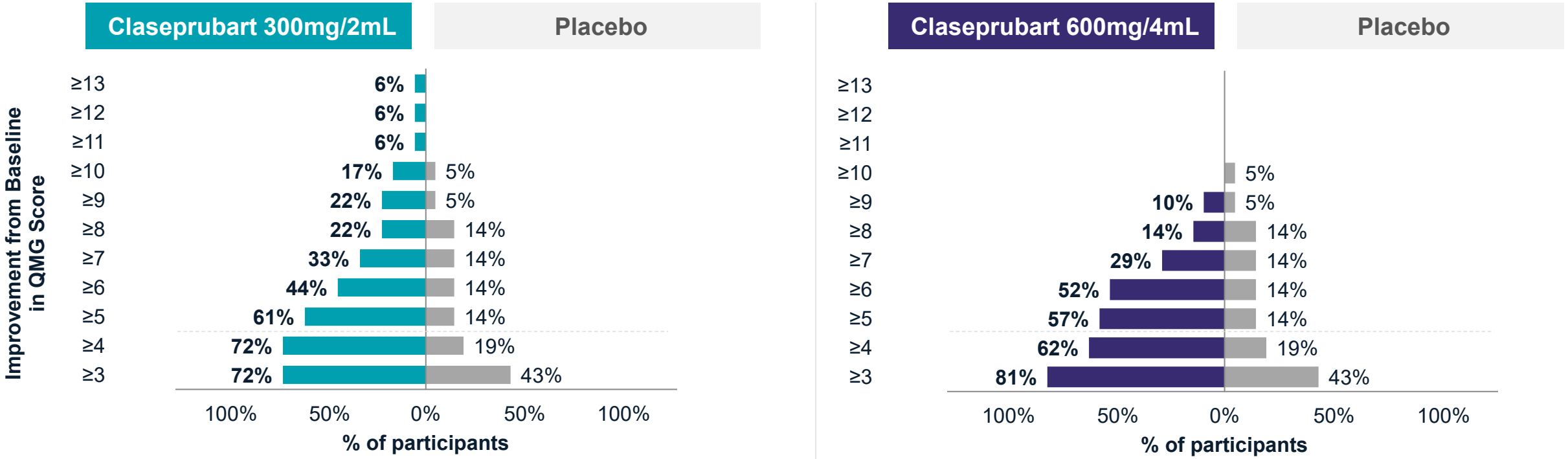
## Improvement in MG-ADL Total Score




**Participants across both treatment arms achieved robust improvements in MG-ADL at Week 13**

# >60% of participants on claseprubart 300mg/2mL achieved ≥5 point improvement in QMG

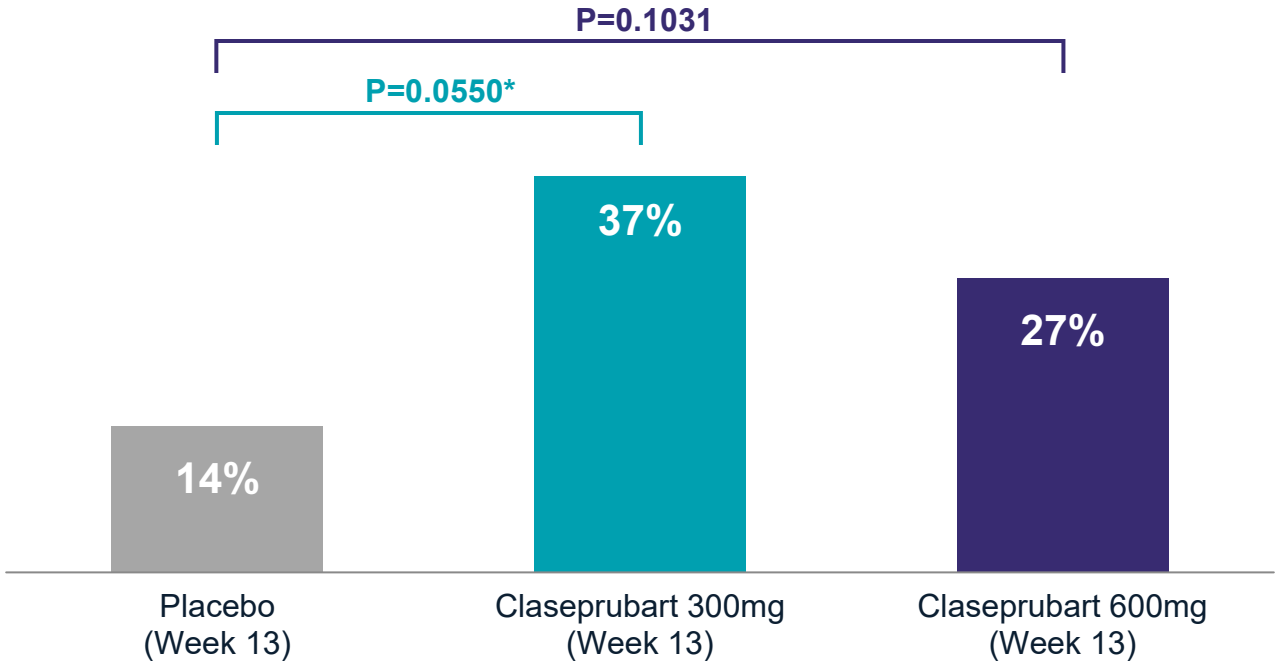
## Improvement in QMG Total Score




**Participants across both treatment arms achieved robust improvements in QMG at Week 13**

# 37% of 300mg/2mL claseprubart-treated participants achieved Minimal Symptom Expression on MG-ADL at Week 13

**Minimal Symptom Expression (MSE)**
% of Participants Achieving MG-ADL Score of 0 or 1 at Week 13




**MSE supports potential best-in-class profile**

The proportion of participants who achieve MSE was analyzed using a logistic regression with terms for treatment group, stratification factors, and baseline MG-ADL included.
*One-sided p-values are presented for comparisons of claseprubart vs placebo, with any p-value below 0.1 considered nominally statistically significant.

# Across key efficacy measures, claseprubart demonstrated robust and clinically meaningful responses

| | Placebo | Claseprubart 300mg/2mL Q2W | | Claseprubart 600mg/4mL Q2W | |
|---|---|---|---|---|---|
| | | Absolute | Placebo-adjusted | Absolute | Placebo-adjusted |
| MG-ADL mean change from baseline at Week 13 | -2.8 | -4.6 | -1.8 *(P=0.0113)** | -5.4 | -2.6 *(P=0.0006)** |
| QMG mean change from baseline at Week 13 | -2.0 | -4.4 | -2.4 *(P=0.0144)** | -4.5 | -2.5 *(P=0.0111)** |
| MSE at Week 13 | 14% | 37% | 23% *(P=0.0550)** | 27% | 13% *(P=0.1031)* |
| MGC mean change from baseline at Week 13 | -3.1 | -8.7 | -5.6 *(P=0.0008)** | -8.6 | -5.5 *(P=0.0008)** |
| MG-QoL-15r mean change from baseline at Week 13 | -3.9 | -6.1 | -2.2 *(P=0.0414)** | -5.4 | -1.5 *(P=0.1122)* |

**Claseprubart 300mg/2mL Q2W treatment arm achieved statistical significance vs. placebo across all five key efficacy measures**

*One-sided p-values are presented for comparisons of claseprubart vs placebo, with any p-value below 0.1 considered nominally statistically significant.

# Claseprubart was generally well tolerated, with a favorable, potentially differentiated safety profile in Ph. 2

| | Placebo (N=22) | Claseprubart 300mg/2mL Q2W (N=21) | Claseprubart 600mg/4mL Q2W (N=22) |
|---|---|---|---|
| **Clinical adverse events (AEs)[1]** | 11 (50.0%) | 13 (61.9%) | 15 (68.2%) |
| **Related serious AEs** | 1 (4.5%) | 0 (0%) | 0 (0%) |
| **RCT discontinuation due to related AE** | 0 (0%) | 0 (0%) | 0 (0%) |
| **Infections** | 10 (45.5%) | 5 (23.8%) | 6 (27.3%) |
| **Related serious infections** | 1 (4.5%) | 0 (0%) | 0 (0%) |
| **Injection site reactions[2]** | 0 (0%) | 2 (9.5%) | 2 (9.1%) |
| **Newly positive for anti-nuclear antibodies (ANA)[3]** | 0 (0%) | 1 (5.9%) | 8 (36.4%) |
| **Rashes** | 0 (0%) | 0 (0%) | 0 (0%) |
| **Arthralgia** | 1 (4.5%) | 1 (4.8%) | 0 (0%) |

**Comparable clinical safety profile to placebo with remarkably benign administration, no infection signal and no symptoms indicative of autoimmune activation or DIL[4]**

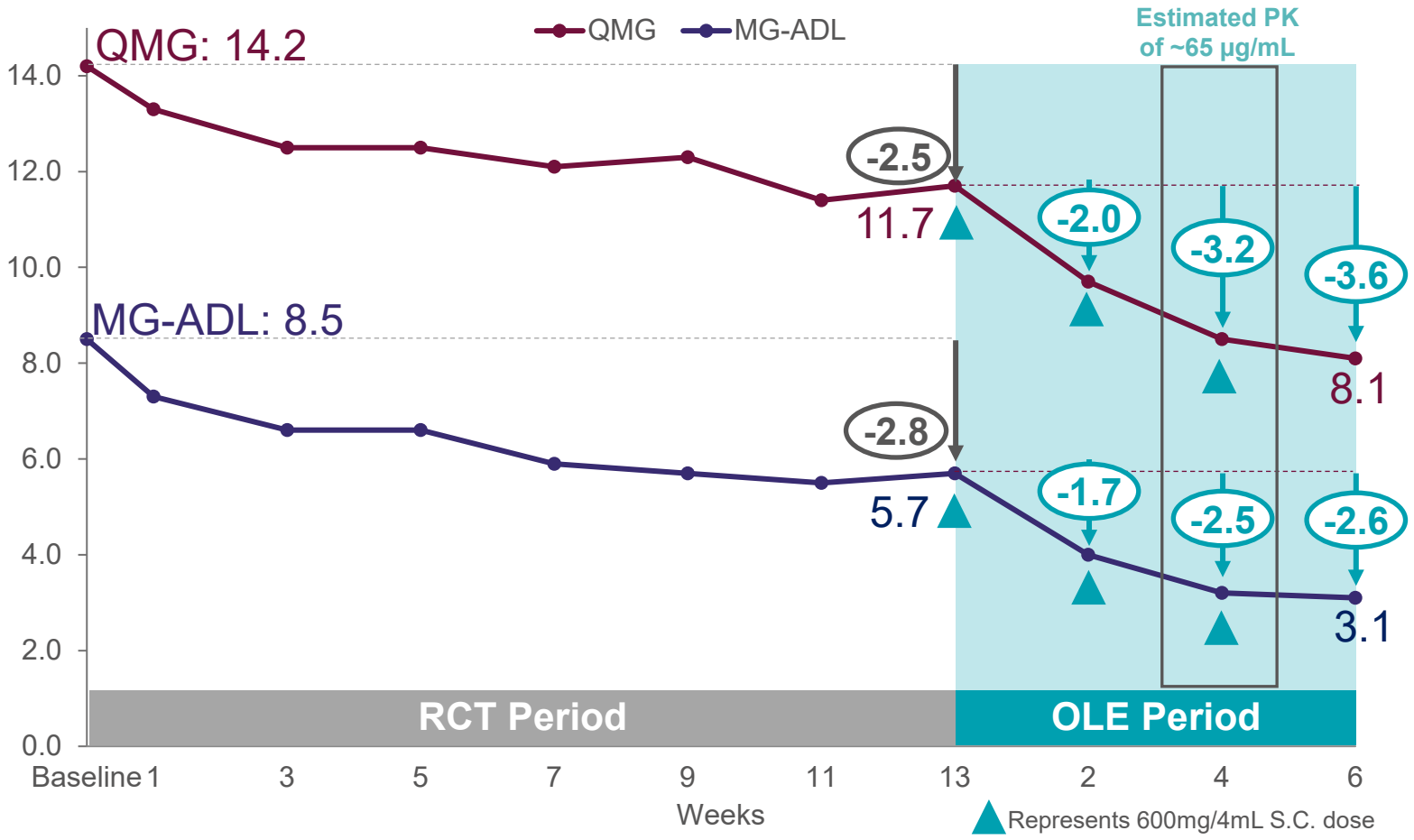
1. Excludes events in the investigations System Organ Class (MedDRA).
2. All injection site reactions were mild to moderate.
3. Represents participants who were ANA negative at baseline and tested positive at ≥1:320 at any point during RCT (percentages calculated from n=17 for 300mg arm and n=22 for 600mg arm). An ANA titer of ≥1:320 was an exclusion criterion for the clinical trial protocol. At end of RCT (Week 13), 2 of the 8 patients in 600mg arm tested negative for ANA, 2 of the 8 patients in 600mg arm remained positive but at <1:320.
4. Drug-Induced Lupus (DIL) is an autoimmune syndrome triggered by specific medications, such as statins, TNF-alpha inhibitors, and ACE inhibitors/beta-blockers. A key distinguishing feature of DIL is its reversibility, as symptoms typically resolve once the offending medication is withdrawn.

# Rationale for Q4W 300mg/2mL Dosing & Potential for Enhanced, Best-in-Disease Efficacy in gMG

# OLE data support addition of 300mg/2mL Q4W in Ph. 3

PK levels approximately half of 300mg/2mL Q2W steady state resulted in robust reductions on MG-ADL & QMG

## Mean Change in PBO Patients' MG-ADL and QMG Score from RCT Baseline to OLE Week 6



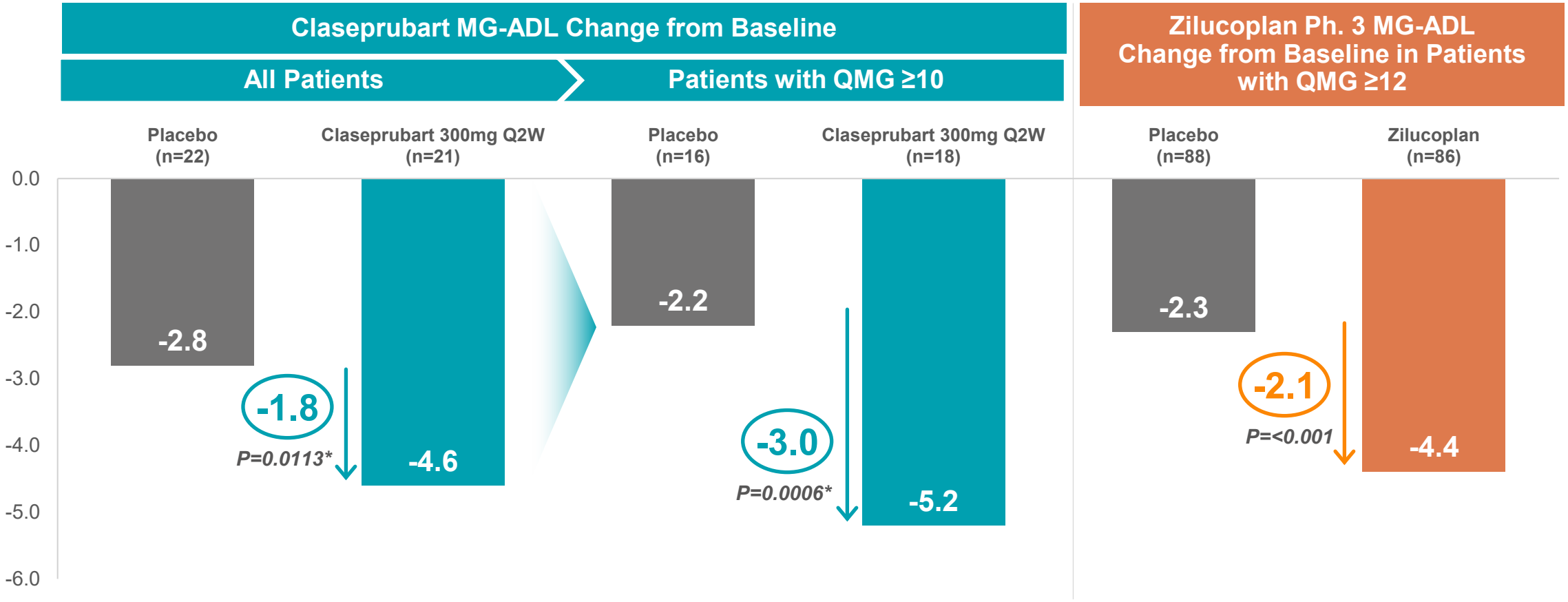

### PBO Patients Entering OLE Received 600mg/4mL Q2W w/ No Loading Dose

- PK of ~65 µg/mL at week 4 after only two 600mg/4mL doses is substantially lower than steady state seen with 300mg/2mL dosing of ~100-120 µg/mL
- Robust reductions in MG-ADL and QMG are achieved by week 4, after just two 600mg/4mL doses and remain stable in subsequent weeks
- Growing external evidence further supports that lower levels of complement inhibition (<90%) may be sufficient for efficacy in gMG[1]

The change from RCT baseline in MG-ADL and QMG were separately analyzed using a mixed effect model for repeated measures (MMRM) with randomized treatment group, visit, randomized treatment by visit interaction, stratification factors, and baseline measure included. All patients received claseprubart in OLE.

1. https://newsroom.regeneron.com/node/31216/pdf

# Adding QMG screening criteria in Ph. 3, similar to zilucoplan Ph. 3[1], may better control for placebo response



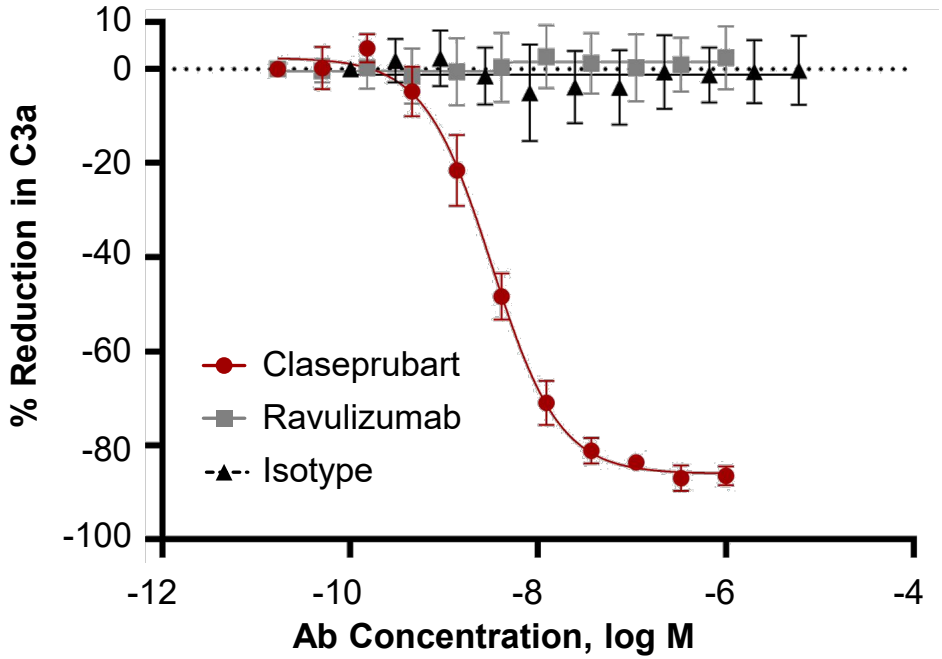

**Ph. 2 study did not include QMG inclusion criteria, similar to ravulizumab Ph. 3; post-hoc analysis of MaGic data demonstrates potentially best-in-class MG-ADL improvement in patients with QMG ≥10**

The change from baseline in ADL was analyzed using a mixed effect model for repeated measures (MMRM) with treatment group, visit, treatment by visit interaction, stratification factors, and baseline measure included.
*One-sided p-values are presented for comparisons of claseprubart vs placebo, with any p-value below 0.1 considered nominally statistically significant.
1. Zilucoplan Ph. 3 MG trial had screening criteria of QMG ≥ 12 and MG-ADL ≥ 6 (https://clinicaltrials.gov/study/NCT04115293)

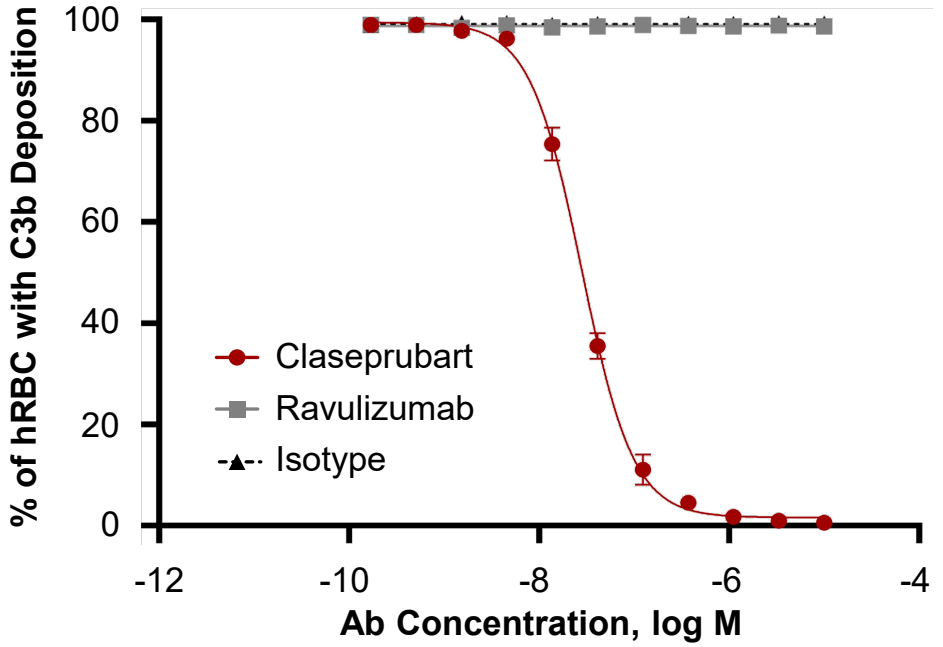
# Potential for improved efficacy vs. C5 inhibitors with claseprubart may be due to upstream inhibition

## Claseprubart Prevents the Creation of Pro-inflammatory Split Products C3a and C3b vs. Ravulizumab

### C3a Formation[1]




### C3b Deposition[2*]




**Upstream inhibition prevents the creation of pro-inflammatory C3a and C3b as well as MAC, potentially providing additional efficacy benefits for AChR+ gMG patients**

1. C3a Formation Assay: Human C3a ELISA specific to C3a-desArg with no cross-reactivity to C3 (N=3).
2. C3b Deposition Assay: Ab-sensitized hRBC triggered by complement-positive sera to deposit C3b on the hRBC surface, measured by flow cytometry (N=3).
*Enjaymo (sutimlimab) targets the C1s complement protein, which prevents C3b deposition on red blood cells, thereby stopping hemolysis and improving anemia in patients with cold agglutinin disease (Jager U, et al. Blood 2019;133:893–901).

# EMERGE is a global Ph. 3 trial evaluating Q4W and Q2W S.C. claseprubart in AChR+ gMG; top-line data 2H'28

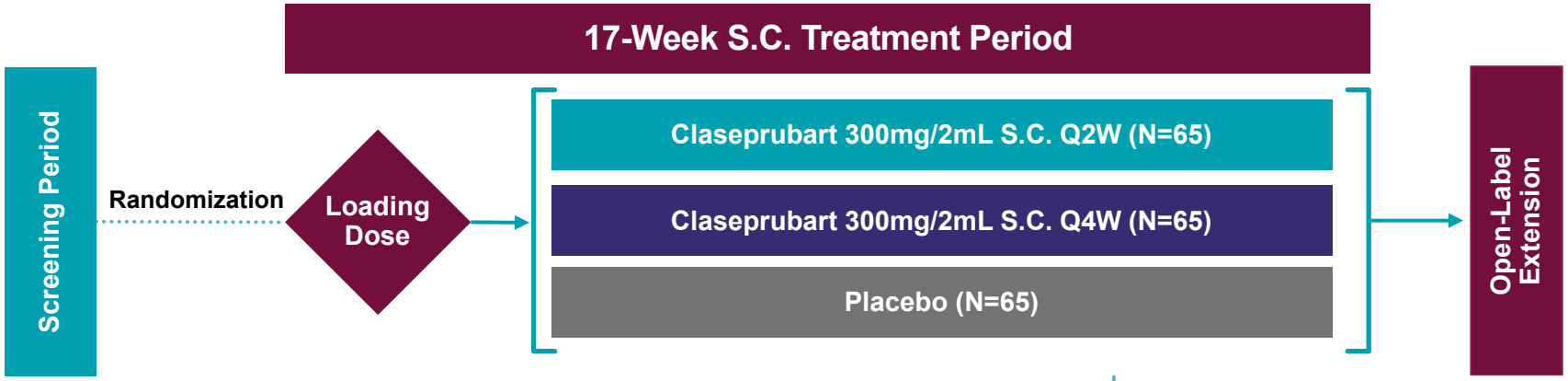
No ANA screening exclusion criteria or routine ANA testing during RCT or OLE, inclusion criteria of MG-ADL of ≥6 and QMG of ≥10, and 17-week S.C. treatment period per alignment with FDA

## Highlights

- **Design:** Male and female subjects randomized to receive either claseprubart or placebo for 17 weeks
- **Inclusion:** ≥18 years old with AChR antibody + gMG, **MG-ADL of ≥6 and QMG of ≥10**
- **Dosing:** I.V. Loading Dose followed by 300mg/2mL S.C. Q2W or **Q4W** starting Day 7
- **No ANA screening exclusion criteria or routine ANA testing** during the RCT or OLE

## Endpoints

- **Primary:** MG-ADL change from baseline
- **Secondary / Exploratory:** Efficacy (QMG, MSE, MGC, MG-QoL-15r)




**Potential to further enhance best-in-disease differentiation on efficacy and dosing convenience with QMG screening criteria and 300mg/2mL Q4W dosing**



EMERGE Trial: https://clinicaltrials.gov/study/NCT07647510

# Achieving this TPP could position claseprubart as a potential best-in-disease biologic and grow the gMG market



## C5 OR SUPERIOR EFFICACY
**(Ultomiris/Soliris/Zilbrysq)**

Rapid onset with similar or superior MG-ADL to FDA-approved C5 inhibitors with continuous, effective symptom control

**Targeting >2-point MG-ADL improvement vs. placebo**



## C1s SAFETY
**(Enjaymo)**

Comparable *safety* to FDA-approved C1s & Classical Pathway inhibitor, leaving the lectin and alternative pathways intact

**Targeting no Boxed Warning & REMS**



## AUTOINJECTOR CONVENIENCE
**(Dupixent)**

Comparable *convenience* to Dupixent with rapid, one-click, self-administered SHL-Molly autoinjector

**Targeting single 300mg/2mL self-administered S.C. Q2W or Q4W**

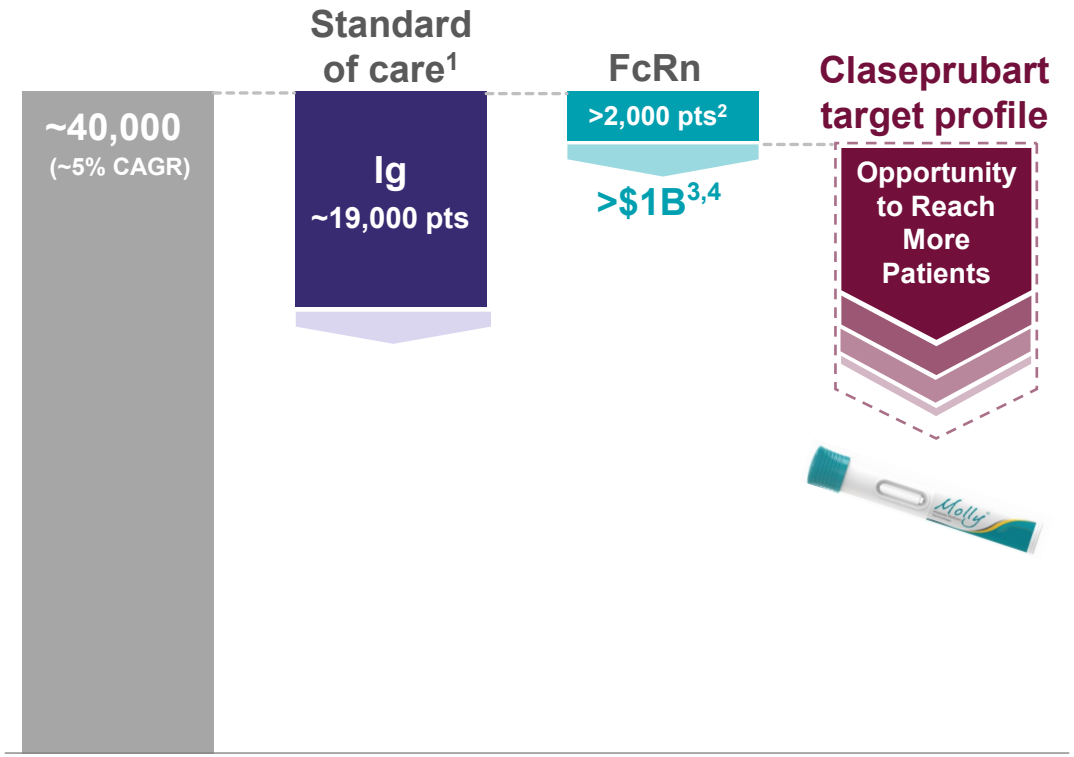
Claseprubart is an investigational agent that is not approved as a therapy in any indication in any jurisdiction worldwide. Safety and efficacy for claseprubart has not been evaluated in head-to-head comparative clinical studies.
Autoinjector for claseprubart administration is anticipated to be SHL Medical's Molly technology, patented or patent pending in the US, China, India, Japan, Korea, Taiwan and at the European Patent Office.

# Claseprubart: Opportunity to Change the Treatment Paradigm in Chronic Inflammatory Demyelinating Polyneuropathy

# The US CIDP market offers substantial growth potential given high unmet need and limitations of current standard of care

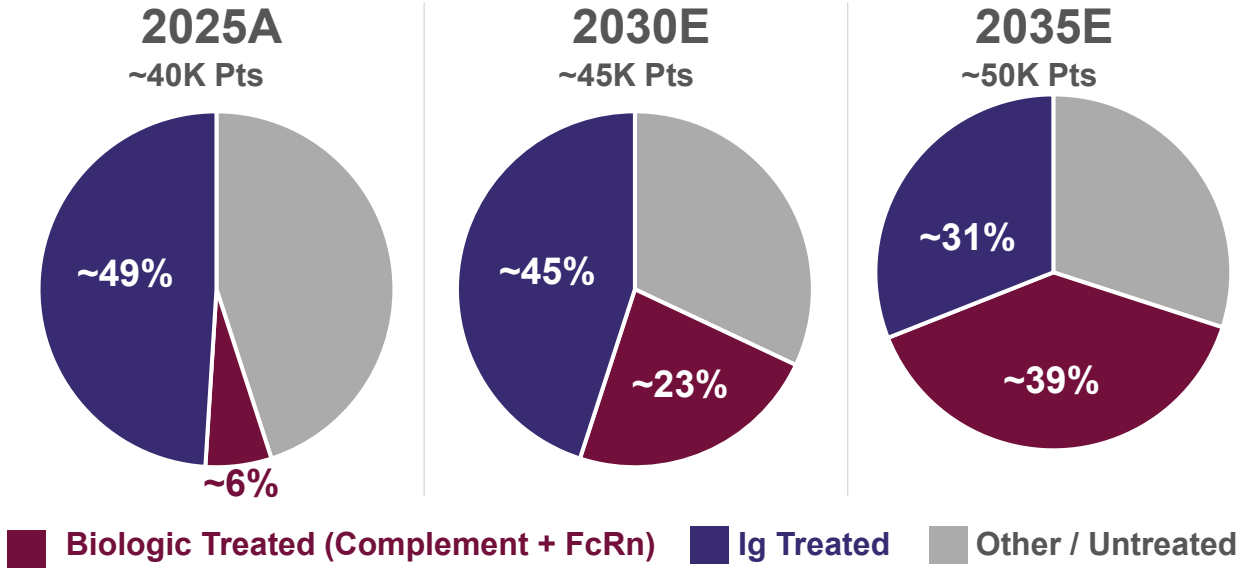
Opportunity for an active C1s inhibitor with the target profile of claseprubart to replace the standard of care

## Current US CIDP Market




## >$8B CIDP Opportunity for Complement in 2035[5]

### Estimated US biologic and Ig penetration[6]




### Estimated US biologic and Ig sales[6]

| 2025A | 2030E | 2035E |
|---|---|---|
| ~$4B | ~$10B | ~$16B |
| ~22K pts | ~31K pts | ~35K pts |

Note: Positioning of claseprubart's target profile is illustrative. 1. Komodo claims data 2013-2025, adjusted to account for 70% capture of real-world patient counts for biologic treated patients, adjusted to account for 27% misdiagnosed. 2. Argenx Vyvgart Hytrulo HCP website. 3. Fierce Pharma, CIDP Pricing. 4. Argenx 4Q 2025 Financial Results, Feb 26, 2026. 5. Based on Dianthus market research and estimates. 6. Based on EvaluatePharma (Jan '26), Immunoglobulin – Global Market Analysis, Fortune Business Insights, and Dianthus market research and estimates. Assumes Ig price per patient of ~$150,000 per year and average biologic net price per patient of ~$700,000 per year.

# Survey of US Neurologists supports potential transformative opportunity in CIDP

Claseprubart aims to differentiate and effectively address the significant unmet needs in the CIDP market

## Total Neurologists 80

| | |
|---|---|
| Neuromuscular Specialist | 81% |
| Generalist | 19% |
| Academic | 58% |
| Community based | 42% |

## Sample Demographics

**~13** years in active clinical practice (post-residency), on average

**~90%** of professional time spent providing direct patient care, on average

**~60** CIDP patients seen in the past 12 months, on average

**~79%**

of Neurologists strongly believe patients prefer treatments with **more consistent and sustained symptom control**

**~66%**

of Neurologists strongly believe there is high unmet **need for therapies with greater efficacy**

**~54%**

of Neurologists strongly prefer treatment options **without a Boxed Warning or REMS program**

**~75%**

of Neurologists strongly believe patients prefer therapies that are **more convenient and easier to administer**

Dianthus 2026 Neurologist CIDP and MMN quantitative survey (n=80, N = 65 Neuromuscular specialists, N = 15 General Neurologists), fielded Q1.
Data represents % of Neurologists selecting 7-9 on a 9-pt. agreement scale.

# Surveyed Neurologists want safer, more effective and convenient treatment options than IVIg for CIDP patients

Overall, surveyed Neurologists believe ~50% of patients on IVIg have partial or no response to treatment

| % of Surveyed Neurologists Who Strongly Believe… | |
|---|---|
|   | Significant unmet **need for effective therapies** for patients who don't respond to IVIg |
|   | Patients on IVIg commonly report **waning efficacy prior to their next dose** |
|   | IVIg imposes a **high treatment burden** due to frequent and time-consuming infusions |
|   | IVIg **boxed warning is concerning** due to potential risk of kidney failure or thrombosis |

Dianthus 2026 Neurologist CIDP and MMN quantitative survey (n=80, N = 65 Neuromuscular specialists, N = 15 General Neurologists), fielded Q1.
Data represents % of Neurologists selecting 7-9 on a 9-pt. agreement scale.

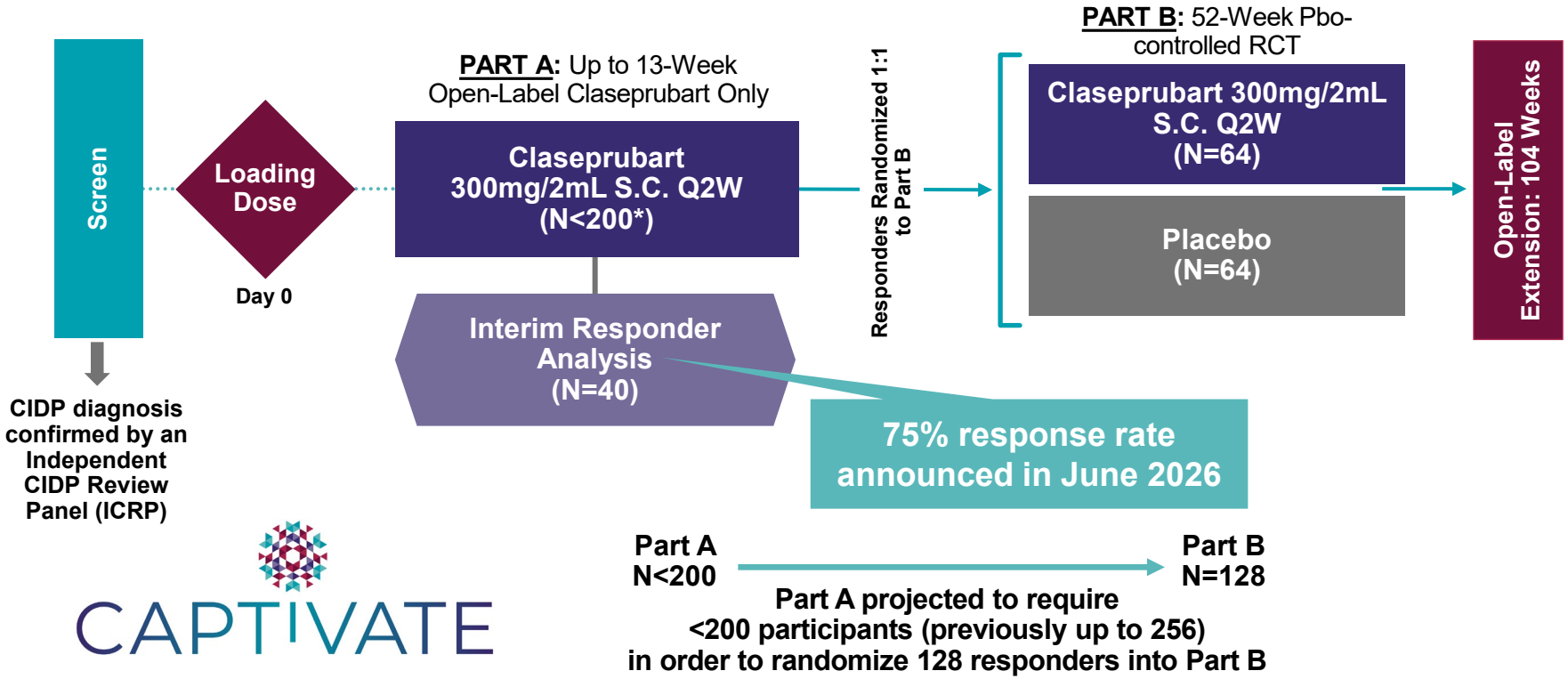
# Ph. 3 CAPTIVATE trial Part A top-line data expected by YE'27 and Part B top-line data by YE'28

### Highlights

- **Design:** All subjects receive claseprubart in Part A for up to 13 weeks. Only responders randomized to Part B for 52 weeks
- **Inclusion:** ≥18 years old with confirmed CIDP, including SoC-Refractory, SoC-Treated or SoC-Naïve
- **Dosing:** I.V. Loading Dose followed by 300mg/2mL S.C. Q2W in Part A; followed by 300mg/2mL or placebo in Part B
- **No ANA screening exclusion criteria or routine ANA testing** during the RCT or OLE

### Endpoints

- **Part A:** Response as measured as ≥1 point decrease (improvement) in adjusted INCAT score compared to Part A baseline
- **Part B Primary:** Efficacy (time to relapse) as measured as ≥1 point increase in adjusted INCAT




- **Single pivotal** two-part, randomized withdrawal, double-blind, placebo-controlled trial designed to **support BLA in adult patients with CIDP**
- Enrolling a **broad patient population including SoC-refractory patients**, in addition to SoC-Treated and SoC-Naïve patients
- **No requirement for IVIg withdrawal and disease worsening,** consistent with ongoing FcRn and complement CIDP studies
- **Only responders from Part A** randomized into the double-blind, placebo-controlled Part B

CAPTIVATE Trial: https://clinicaltrials.gov/study/NCT06858579
*Actual Part A enrollment is subject to Part A response rate.

# Significant differences between CAPTIVATE and ADHERE

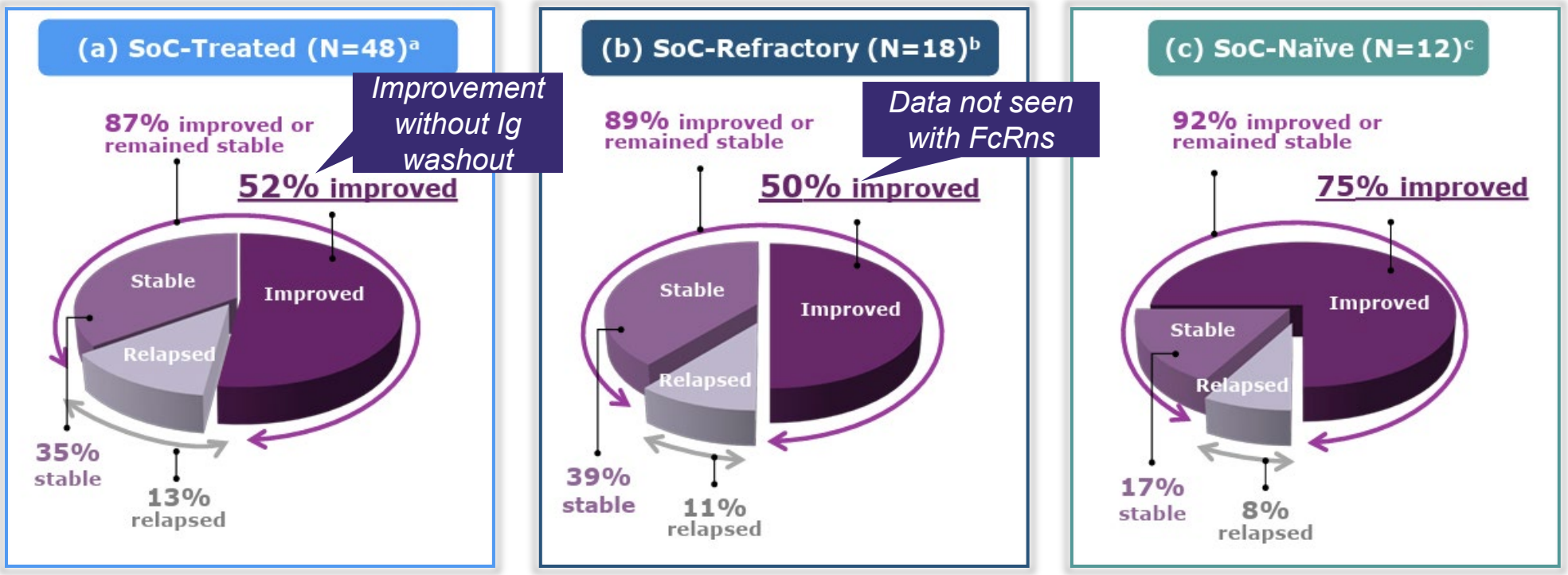
| Considerations | Efgartigimod (FcRn) S.C. QW | Claseprubart (aC1s) 300mg/2mL S.C. Q2W | Key Differentiators of CAPTIVATE |
|---|---|---|---|
| **Ph. 3 Study Populations** | adhere<br>**SoC-Treated**<br>**Off Treatment** | CAPTIVATE<br>**SoC-Treated**<br>**SoC-Naïve**<br>**SoC-Refractory** | **Evaluating claseprubart in SoC-Refractory CIDP patients**, in addition to a broader CIDP patient population including SoC-Treated and SoC-Naïve |
| **Require IVIg or SCIg Withdrawal and Relapse Prior to Enrolling in Part A of Study[1]** | **YES** | **NO** | **Immediate switch 7 days from last Ig dose to claseprubart**; consistent with other ongoing complement CIDP studies[3] |
| **Study Endpoints / Results** | • **Confirmed ECI[2]**<br>• **Ph. 3 Stage A results:**<br>–66.5% ECI (wk 12)<br>*~1/3 of pts* ***did not return*** *to pre-Ig washout baseline* | • **Switching Ig patients to claseprubart 7 days after last dose**<br>• **Aiming for ≥1-point adj. INCAT improvement OVER SoC/Ig in ≥50% of patients in Part A** | Part A designed to evaluate **clinically meaningful improvement over Ig after immediate switch 7 days after last dose** |

Source: Company filings, presentations and clinicaltrials.gov.
Ig refers to IVIg and SCIg.
Data are derived from different clinical trials conducted at different times, with differences in trial design and patient populations. As a result, cross-trial comparisons cannot be made, and no head-to-head clinical trials have been conducted.

1. ADHERE required discontinuation of IVIg or SCIg and evidence of clinically meaningful deterioration before dosing in Part A.
2. Defined as a clinical improvement on the parameters that the participant worsened in during run-in (≥4-point increase in I-RODS and/or ≥8-kPa increase in mean grip strength) or clinical improvement (≥1-point decrease) in INCAT.
3. Empasiprubart and riliprubart studies.

# Active C1s inhibition with riliprubart has demonstrated clinical proof-of-concept across broad patient groups

## Ph. 2 Riliprubart Data in Active C1s in CIDP[1] with High Volume, Weekly Dosing of 600mg/4mL[2]




**Claseprubart is being evaluated with a convenient, low volume dose of 300mg/2mL Q2W**

1. Riliprubart Phase 2 at PNS 2024.
2. Based on riliprubart patent filing (Pg 76).

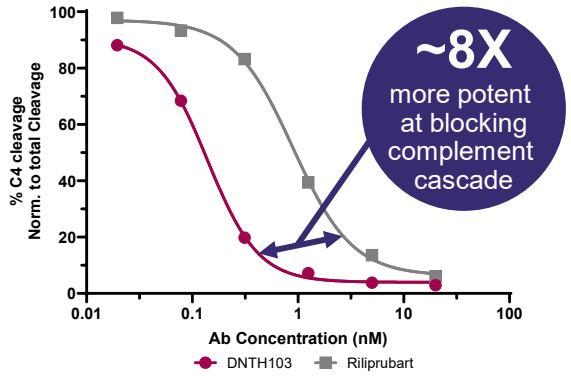
# Claseprubart *in vitro* affinity and potency

## Affinity assays

| | | DNTH103 | riliprubart | Fold Improvement |
|---|---|---|---|---|
| **Binding Affinity to human active C1s ($K_D$)[1]** | KinExa | **9pM** | 75pM | **~8X** |
| | SPR | **8pM** | 35pM | **~4X** |

## Enzymatic assay

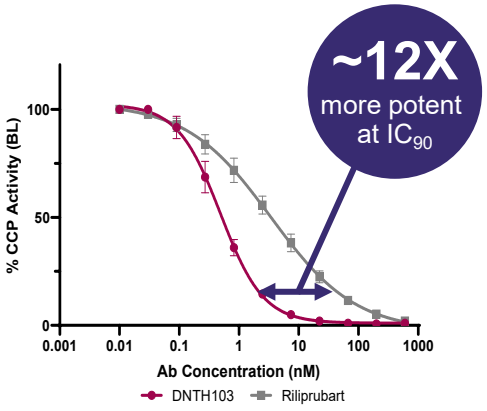
### C4 cleavage by human active C1s[2]




| | DNTH103 | riliprubart |
|---|---|---|
| $IC_{90}$ (nM) | **0.47** | 3.69 |

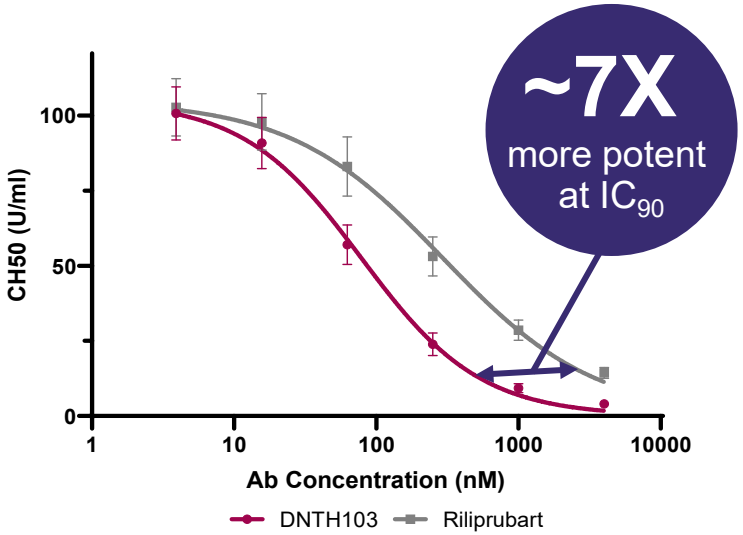
## Functional assays of Classical Pathway inhibition

### Wieslab Classical Pathway Assay in human serum[3]



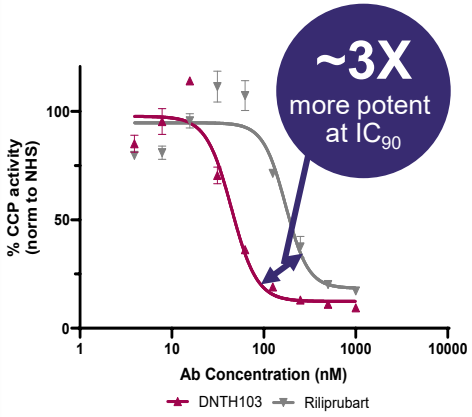

| | DNTH103 | riliprubart |
|---|---|---|
| $IC_{90}$ (µg/mL) | **0.45** | 5.4 |

### CH50 assay of RBC lysis in human serum[3]




| | DNTH103 | riliprubart |
|---|---|---|
| $IC_{90}$ (µg/mL) | **98** | 668 |

### Liposome lysis in human serum[3]




| | DNTH103 | riliprubart |
|---|---|---|
| $IC_{90}$ (µg/mL) | **14.7** | 39.8 |

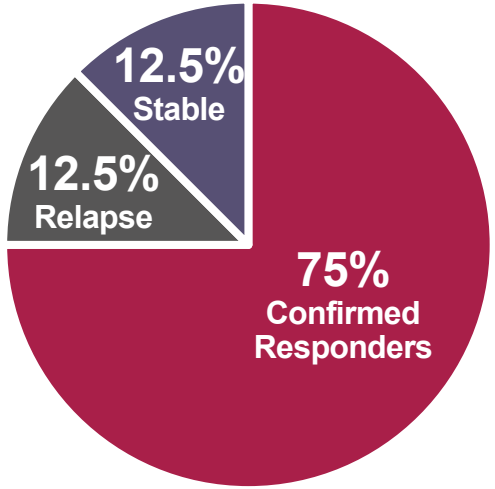
**Claseprubart has consistently higher affinity and stronger potency in multiple head-to-head *in vitro* experiments**

Note: Riliprubart is produced using sequence from patent WO2018071676A1

1. Data shown is dissociation constant ($K_D$) and the average of 3 different experiments performed at independent laboratories.
2. Data is quantitative analysis of active C1s protease inhibition of cleaved C4 fragments in the presence of claseprubart or riliprubart.
3. Data shown are the average of 3 experiments conducted for each of the functional assays (CH50 hemolysis, Wieslab and Liposome). CH50 and Wieslab were confirmed at independent laboratories.

# 75% response rate observed in Interim Responder Analysis from first 40 participants completing Part A of CAPTIVATE

## Breakdown of First 40 Participants Completing CAPTIVATE Part A




## Safety / Tolerability Update

Generally well tolerated with no related serious infections, no clinical symptoms of DIL, no related SAEs or discontinuations due to safety

## Summary of Change from Baseline at Last Evaluable Visit[1]

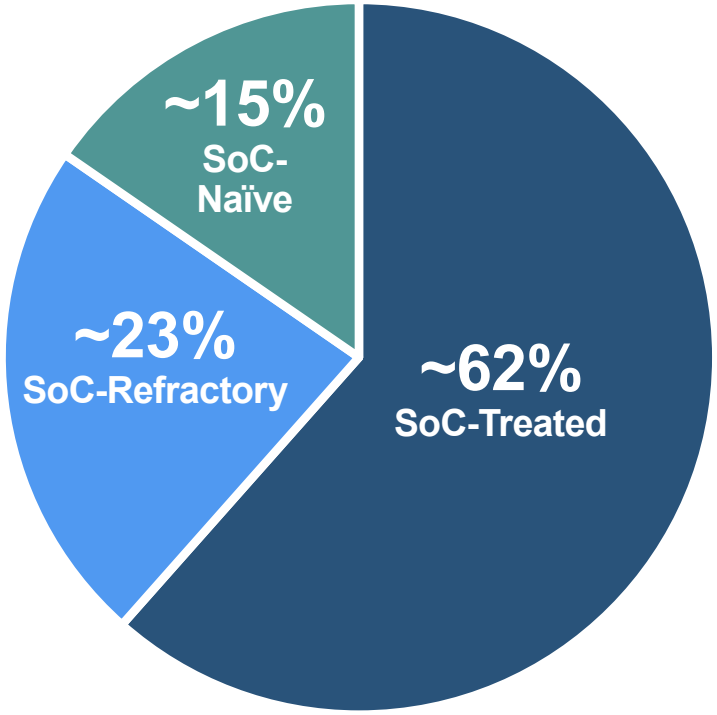
| | Confirmed Responders (N=30) | Non-Responders (N=10) |
|---|---|---|
| **INCAT**<br>*Decrease is improvement*<br>*mean (SD)* | **-1.6**<br>(0.89) | **0.7**<br>(1.06) |
| **Grip Strength**<br>*Increase is improvement*<br>*mean (SD)* | **15.9**<br>(12.78) | **-7.5**<br>(12.19) |
| **MRC-SS**<br>*Increase is improvement*<br>*mean (SD)* | **5.1**<br>(3.93) | **-2.3**<br>(8.82) |
| **I-RODS**<br>*Increase is improvement*<br>*mean (SD)* | **8.9**<br>(12.08) | **-3.0**<br>(10.69) |

**Consistent and clinically meaningful results across multiple efficacy measures from first 30 confirmed responders in Part A**

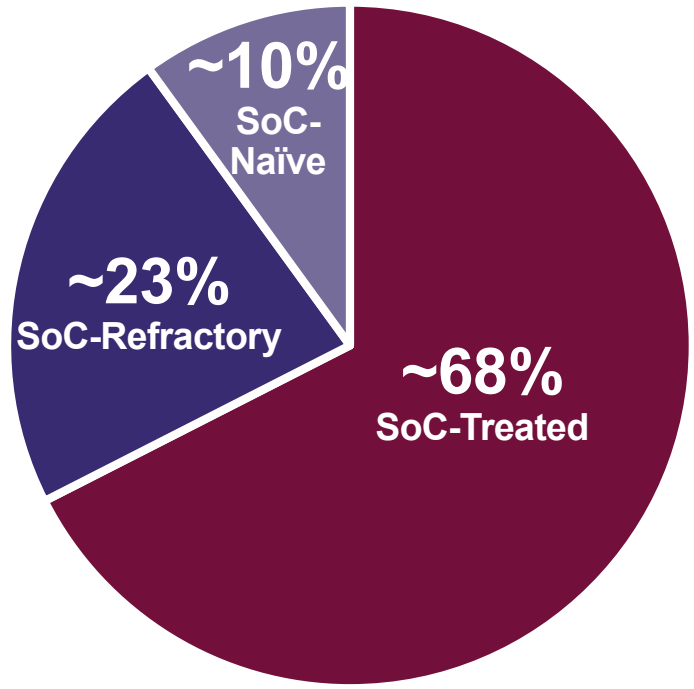
CAPTIVATE per protocol Interim Analysis data from first 40 participants to complete Part A. Non-Responders includes relapses and stable patients.
The summaries presented are preliminary. The study remains ongoing and the data are not final.
INCAT, Inflammatory Neuropathy Cause and Treatment; MRC-SS, Medical Research Council Sum Score; I-RODS, Inflammatory Rasch-built Overall Disability Scale.
1. Last Evaluable Visit: for confirmed responders signifies last visit prior to randomization in Part B; for stable patients signifies Week 13 visit (completion of Part A); for relapse patients signifies confirmed relapse visit prior to any rescue medication.

# CAPTIVATE Part A baseline patient group characteristics are similar to precedent aC1s Ph. 2 study[1]

**Riliprubart Ph. 2[1]**
**Baseline Patient Group Characteristics (N=78)**




**CAPTIVATE Part A[2]**
**Baseline Patient Group Characteristics (N=40)**




**Broad representation of patients across North America, Europe, and Asia in CAPTIVATE Part A**

Data from CAPTIVATE and riliprubart Ph. 2 are derived from different clinical trials conducted at different times, with differences in trial design and patient populations. As a result, cross-trial comparisons cannot be made, and no head-to-head clinical trials have been conducted.

1. Riliprubart Phase 2 at PNS 2024
2. Includes only first 40 participants who have completed Part A.

# Achieving this target profile could position claseprubart as a potential blockbuster treatment for CIDP



## EFFICACY
**(IVIg/SCIg)**

Improvement over SoC (i.e. Ig) with continuous, effective symptom control

**Targeting Best-in-Disease Efficacy**



## C1s SAFETY
**(Enjaymo)**

Comparable *safety* to FDA-approved C1s & Classical Pathway inhibitor, leaving the lectin and alternative pathways intact

**Targeting no Boxed Warning & REMS**



## AUTOINJECTOR CONVENIENCE
**(Dupixent)**

Most *convenient* therapy with self-administered SHL-Molly autoinjector

**Targeting single 300mg/2mL self-administered S.C. Q2W**

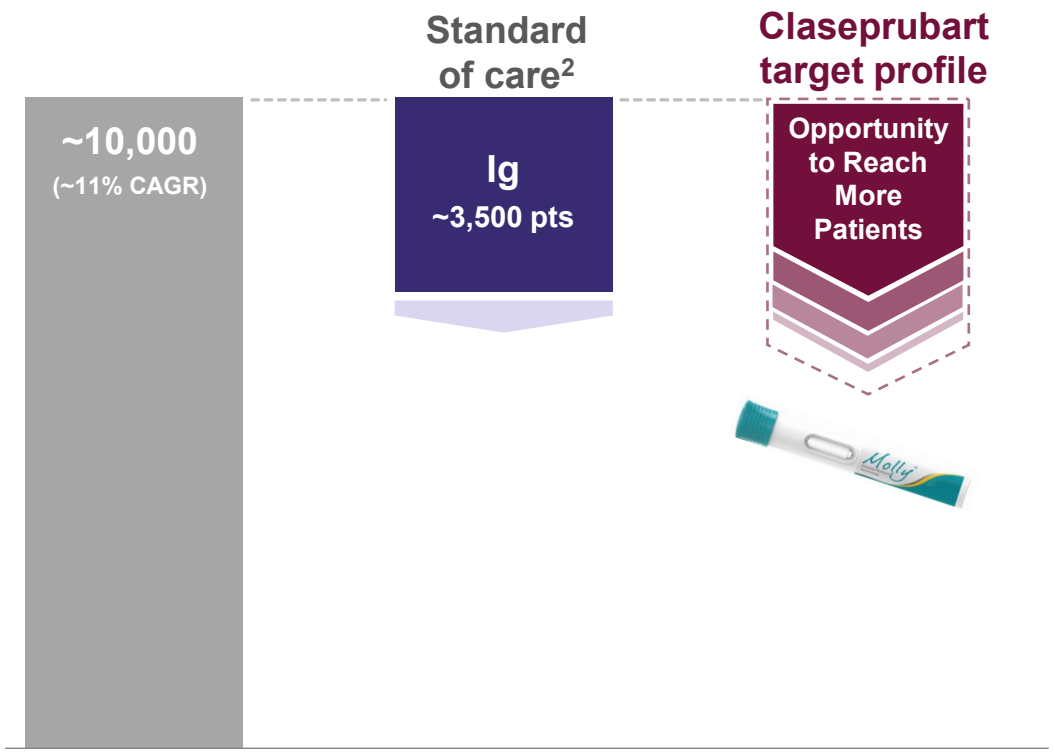
Claseprubart is an investigational agent that is not approved as a therapy in any indication in any jurisdiction worldwide. Safety and efficacy for claseprubart has not been evaluated in head-to-head comparative clinical studies.
Autoinjector for claseprubart administration is anticipated to be SHL Medical's Molly technology, patented or patent pending in the US, China, India, Japan, Korea, Taiwan and at the European Patent Office.

# Claseprubart: Opportunity to be Best-in-Disease in Multifocal Motor Neuropathy

# The US MMN market is a growing blockbuster market opportunity for claseprubart

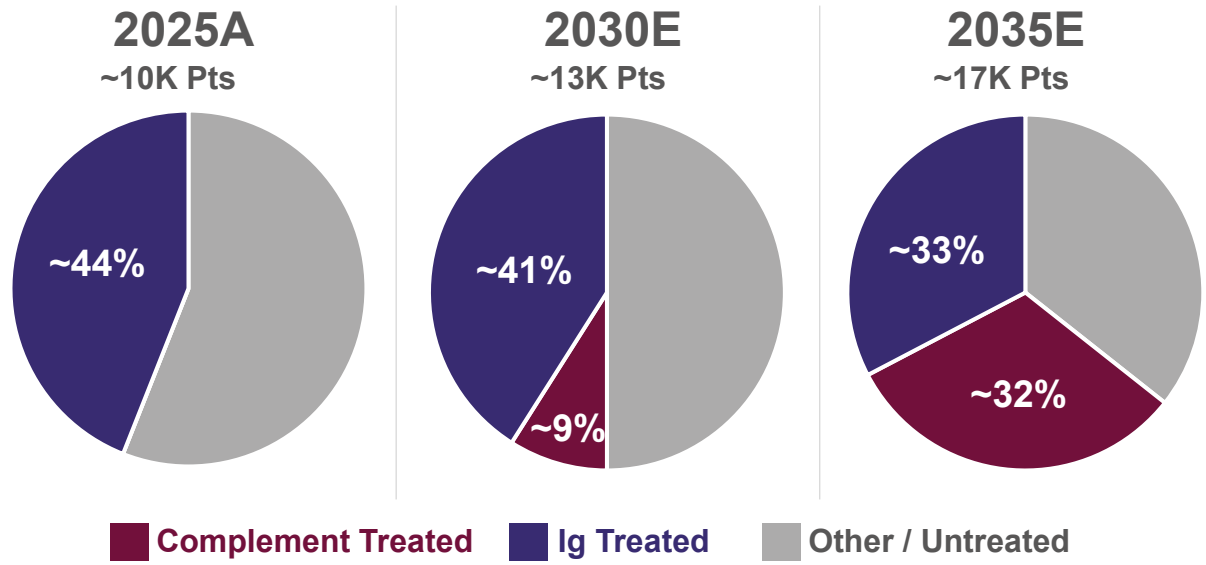
Opportunity for the target profile of claseprubart to become the new standard of care in MMN

## Current US MMN Market




## ~$4B MMN Opportunity for Complement in 2035[3]

### Estimated US complement and Ig penetration[4]




### Estimated US complement and Ig sales[4]

| <$1B | ~$2B | ~$5B |
|---|---|---|
| ~4K pts | ~7K pts | ~11K pts |

Note: Positioning of claseprubart's target profile is illustrative. 1. 2024 patients projected from 2023 count due to unreliable 2024 data from the Change Healthcare cyber-attack. 2. Komodo claims data 2013-2025, adjusted to account for 70% capture of real-world patient counts. 3. Dianthus market research and estimates. 4. Based on EvaluatePharma (Jan '26), Immunoglobulin – Global Market Analysis, Fortune Business Insights, and Dianthus market research and estimates. Assumes Ig price per patient of ~$150,000 per year and average biologic net price per patient of ~$700,000 per year.

# Survey of US Neurologists supports MMN underdiagnosis and large unmet need

## Claseprubart aims to offer a uniquely effective, safe, convenient Classical Pathway inhibitor

### Total Neurologists 80

| | |
|---|---|
| Neuromuscular Specialist | 81% |
| Generalist | 19% |
| Academic | 58% |
| Community based | 42% |

### Sample Demographics

**~13** years in active clinical practice (post-residency), on average

**~90%** of professional time spent providing direct patient care, on average

**~25** MMN patients seen in the past 12 months (median)

**~81%**
of Neurologists strongly believe that MMN is an **underdiagnosed condition**

**~73%**
of Neurologists strongly believe there is a high unmet need for **therapies with meaningful clinical improvement**



**~68%**
of Neurologists strongly believe that treatment options **without Boxed Warnings or REMS are preferred**

**~63%**
of Neurologists strongly believe there is a high unmet need **for therapies with a more favorable dosing schedule**

Dianthus 2026 Neurologist CIDP and MMN quantitative survey (n=80, N = 65 Neuromuscular specialists, N = 15 General Neurologists), fielded Q1.
Data represents % of Neurologists selecting 7-9 on a 9-pt. agreement scale.

# Over half of surveyed US Neurologists agree IVIg is a suboptimal treatment option for MMN patients

The lack of satisfaction with IVIg efficacy in MMN presents a significant opportunity for claseprubart

| % of Surveyed Neurologists Who Strongly Believe… | |
|---|---|
| 58% | Patients on IVIg often report **residual or unaddressed symptoms** |
| 51% | **Clinical efficacy of IVIg** is **incomplete, inconsistent, and/or short-lived** |
| 51% | Despite SoC, **1/3 of patients remain refractory** and face **risk of clinical deterioration** |
| 55% | Current **label warnings/risks** for IVIg are **important in their treatment decisions** |

Dianthus 2026 Neurologist CIDP and MMN quantitative survey (n=80, N = 65 Neuromuscular specialists, N = 15 General Neurologists), fielded Q1.
Data represents % of Neurologists selecting 7-9 on a 9-pt. agreement scale.

# Classical Pathway inhibition has demonstrated PoC in MMN

## Empasiprubart (an inhibitor of both the Classical and Lectin Pathways) Ph. 2 Data Demonstrating Efficacy Signals[1]




"We hypothesize that targeting the **classical complement pathway is a potential therapeutic approach in MMN**. We investigated the interaction of circulating anti-GM1 IgM from patients with MMN with complement in detail using iPSC-derived MNs. In this disease model for MMN, we evaluated the effects of ARGX-117, a novel monoclonal antibody that inhibits complement factor C2." - *Neurol Neuroimmunol Neuroinflamm. 2022 Jan; 9(1): e1107*

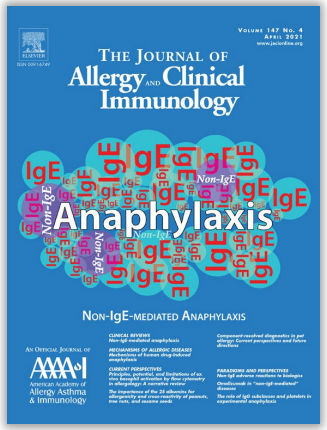
**Top-line Ph. 2 data from MoMeNtum trial of claseprubart, a potent & selective Classical Pathway inhibitor, expected in December 2026**

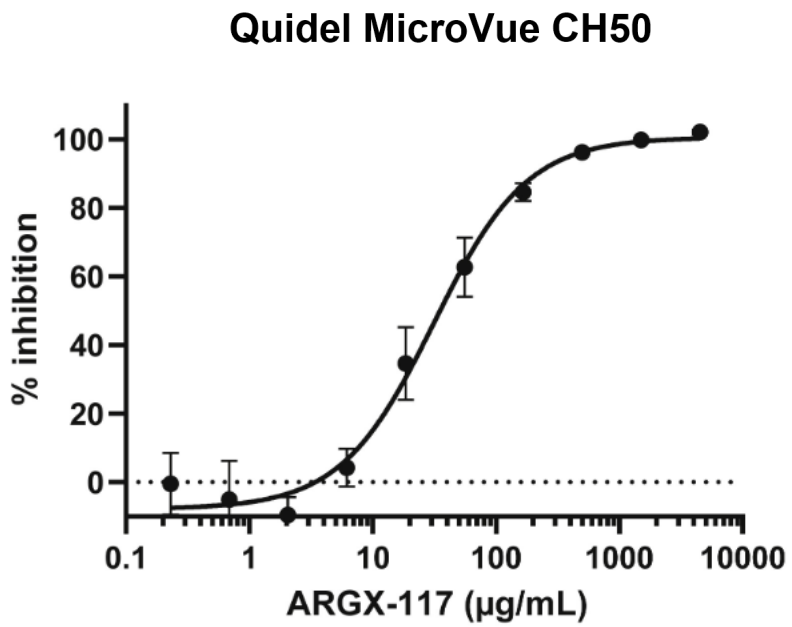


1. https://argenx.com/content/dam/argenx-corp/events-presentations/argenx_RnD_Day_2024_Slides.pdf/#page=126

# Claseprubart demonstrates superior Classical Pathway potency vs. empasiprubart

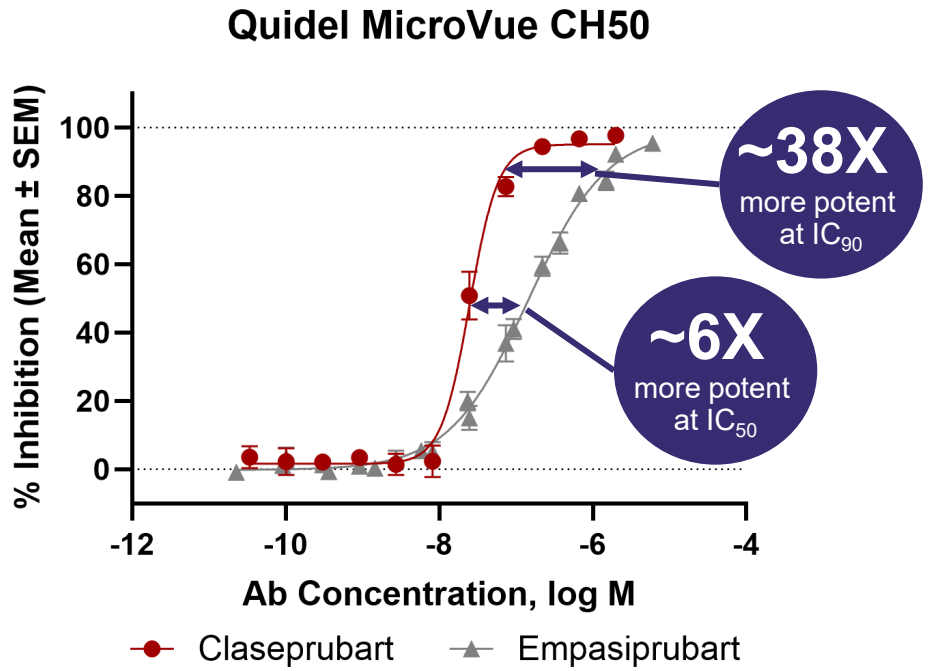
## Empasiprubart Published Classical Pathway Potency Data Using the Quidel MicroVue CH50[1]




"**ARGX-117 potently inhibited CP** and LP (half-maximal effective concentration [$EC_{50}$] = **30.5 ± 4.5** and 93.4 ± 10.4 µg/mL, respectively) in a concentration-dependent manner" – *Journal of Allergy and Clinical Immunology*

## Claseprubart Demonstrates Superior Classical Pathway Potency Head-to-head vs. Empasiprubart Using Same Assay




| | claseprubart | empasiprubart |
|---|---|---|
| $IC_{50}$ (µg/mL) | **3.8 ± 0.8** | 22.1 ± 5.7 |
| $IC_{90}$ (µg/mL) | **9.9 ± 2.5** | 375 ± 266 |

Claseprubart and empasiprubart are investigational agents that are not approved as therapies for MMN or any indication in any jurisdiction worldwide. Head-to-head data shown are the average of 3 experiments conducted for claseprubart and 8 experiments conducted for empasiprubart. Empasiprubart in the head-to-head experiment is produced using the sequence published in the IMGT database (DB card 12277). $EC_{50}$ and $IC_{50}$ can be considered as interchangeable for this analysis.

1. Journal of Allergy and Clinical Immunology, Volume 147, Issue 4, 1420 - 1429.e7

# MoMeNtum is a small PoC Ph. 2 in MMN primarily focused on safety, top-line data anticipated in December ’26

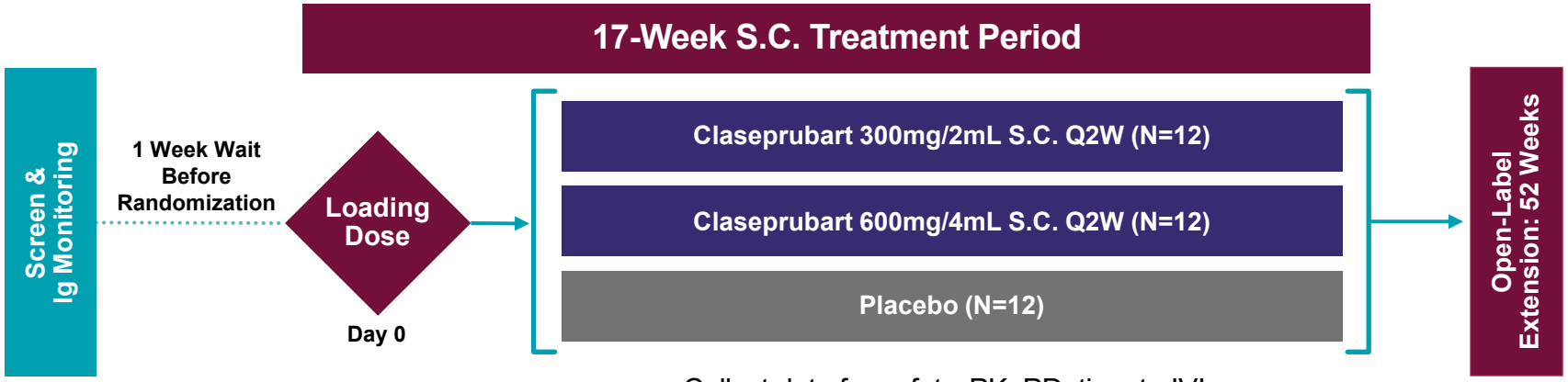
Efficacy endpoints, such as Grip Strength, are secondary endpoints

**Highlights**

- **Design:** 36 participants randomized to receive either claseprubart or placebo for 17 weeks
- **Inclusion:** ≥18 years old with MMN who are immunoglobulin responsive and dependent
- **Dosing:** I.V. Loading Dose followed by 300mg/2mL or 600mg/4mL S.C. Q2W starting Day 7
- **No ANA screening exclusion criteria or routine ANA testing** during the RCT or OLE

**Endpoints**

- **Primary:** Safety
- **Secondary:** Efficacy (time to IVIg retreatment, time to relapse, grip strength and other muscle strength and motor function measurements)




Collect data for safety, PK, PD, time to IVIg retreatment, time to relapse, grip strength and other muscle strength and motor function measurements

MOMENTUM

**Phase 2 MoMeNtum trial of claseprubart in MMN exceeded its original enrollment target of 36 patients; top-line results with 46 patients on track for December ’26**

MoMeNtum Trial: https://clinicaltrials.gov/study/NCT06537999

# Key difference between ARDA Ph. 2 and MoMeNtum Ph. 2 is how IVIg rescue was administered which makes cross-trial comparison challenging

| Key Difference between MoMeNtum and ARDA | Empasiprubart (C2) ARDA Trial | Claseprubart (aC1s) MoMeNtum Trial | Key Attributes of MoMeNtum |
|---|---|---|---|
| **Does Trial Allow Rescue Treatment Upon Patient Request *WITHOUT* Documented Clinical Deterioration?** | YES | **NO** | **MoMeNtum trial only allows rescue if** participant experienced **clinical deterioration** defined by MRC10 ≥2 points or GS decrease >30% |

**Grip Strength is the primary endpoint in the empasiprubart Phase 3 EMPASSION trial, IVIg rescue is not an endpoint in the study**



Source: Company filings, presentations and clinicaltrials.gov.

# Claseprubart has the potential to address the unmet need in the MMN market with its unique target product profile

| Considerations | Empasiprubart (C2)* | Claseprubart (active C1s)* | Key Differentiators of Claseprubart |
|---|---|---|---|
| **MMN is an IgM and Classical Pathway driven disease**[1] | Published classical pathway[3] $EC_{50}$ = 30.5 ±4.5 µg/mL using Quidel MicroVue CH50 | **Claseprubart has demonstrated potent inhibition of Classical Pathway in multiple assays** | **~38X more potent than empasiprubart** on $IC_{90}$ in head-to-head *in vitro* experiment using Quidel MicroVue CH50 |
| **Lectin pathway inhibition not required for efficacy in MMN** | Published lectin pathway[3] inhibition of $EC_{50}$ = 93.4 ±10.4 µg/mL | **Does not inhibit lectin pathway** | Claseprubart **preserves key bacterial killing role** of lectin pathway[2] and may avoid a boxed warning and REMS |
| **Patients prefer convenient therapies** | I.V. Q4W | **Targeting Q2W self-administration via 300mg/2mL S.C. autoinjector** | **More convenient** by targeting infrequent, low volume, self-administered S.C. autoinjector |

**Claseprubart has the potential to be the best-in-disease biologic treatment given its unique combination of classical pathway potency, preservation of the lectin pathway, convenience and potential for no Boxed Warning**

* Claseprubart and empasiprubart are investigational agents that are not approved as therapies for MMN or any indication in any jurisdiction worldwide. EC and IC can be considered as interchangeable for this analysis . Head-to-head data shown are the average of 3 experiments conducted for claseprubart and 8 experiments conducted for empasiprubart. Empasiprubart in the head-to-head experiment is produced using the sequence published in the IMGT database (DB card 12277).

1. Budding et al., (2021). Neurol Neuroimmunol Neuroinflamm.9(1):e1107; Vlam et al., (2015). Neurol Neuroimmunol Neuroinflamm. 2015;2(4):e119; 2. Ali et al., (2012). PLoS Pathog 8(7):e1002793 3. Journal of Allergy and Clinical Immunology, Volume 147, Issue 4, 1420 - 1429.e7.

# Achieving this target profile could position claseprubart as a potential blockbuster treatment for MMN



**EFFICACY**
**(IVIg/SCIg)**

Improvement over SoC (i.e. Ig) with continuous, effective symptom control

**Targeting Best-in-Disease Efficacy**



**C1s SAFETY**
**(Enjaymo)**

Comparable *safety* to FDA-approved C1s & Classical Pathway inhibitor, leaving the lectin and alternative pathways intact

**Targeting no Boxed Warning & REMS**



**AUTOINJECTOR CONVENIENCE**
**(Dupixent)**

Most *convenient* therapy with self-administered SHL-Molly autoinjector

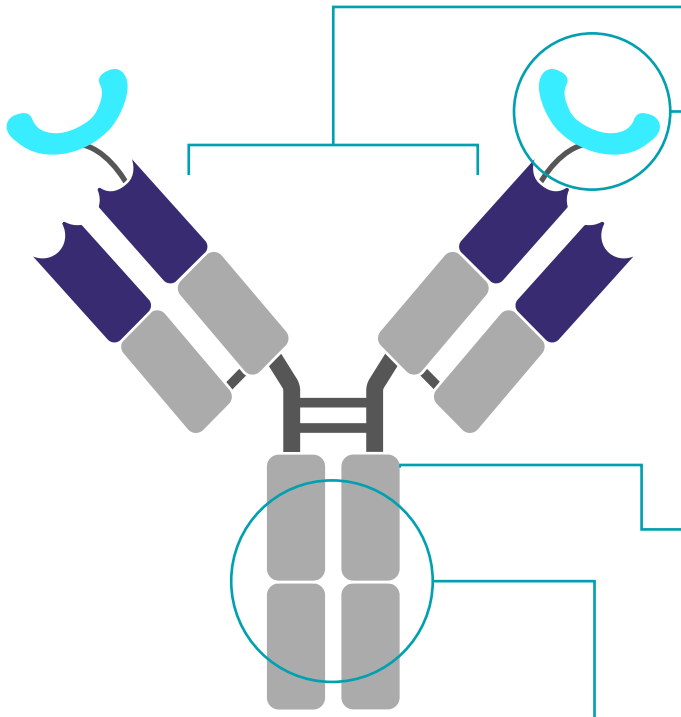
**Targeting single 300mg/2mL self-administered S.C. Q2W**

Claseprubart is an investigational agent that is not approved as a therapy in any indication in any jurisdiction worldwide. Safety and efficacy for claseprubart has not been evaluated in head-to-head comparative clinical studies.
Autoinjector for claseprubart administration is anticipated to be SHL Medical's Molly technology, patented or patent pending in the US, China, India, Japan, Korea, Taiwan and at the European Patent Office.

# DNTH212:
# Potential Best-in-Disease Bifunctional Fusion Protein for Multiple Autoimmune Indications

# DNTH212 is a bifunctional BDCA2 and BAFF/APRIL inhibitor targeting two validated pathways

- Inhibiting BDCA2 **reduces Type 1 interferon production from plasmacytoid dendritic cells (pDCs)**
- Single CRD2 domain of TACI designed to deliver **robust B cell modulation via BAFF/APRIL inhibition**



**DNTH212**
Humanized monoclonal antibody targeting **BDCA2** fused with **TACI-CRD2**

**Anti-BDCA2 Antibody**
Potent inhibition of Type 1 interferon

**TACI-CRD2 Domain**
Potent reduction of Ig levels via BAFF/APRIL inhibition

**Afucosylated Bifunctional Ab**
Enhanced ADCC on target cells leads to pDC depletion

**YTE Mutation on Fc**
Extended half-life

**DNTH212 targets both the innate and adaptive immune systems with complementary disease modifying mechanisms enabling potential best-in-class efficacy**



DNTH212 is an investigational agent that is not approved as a therapy in any indication in any jurisdiction worldwide.

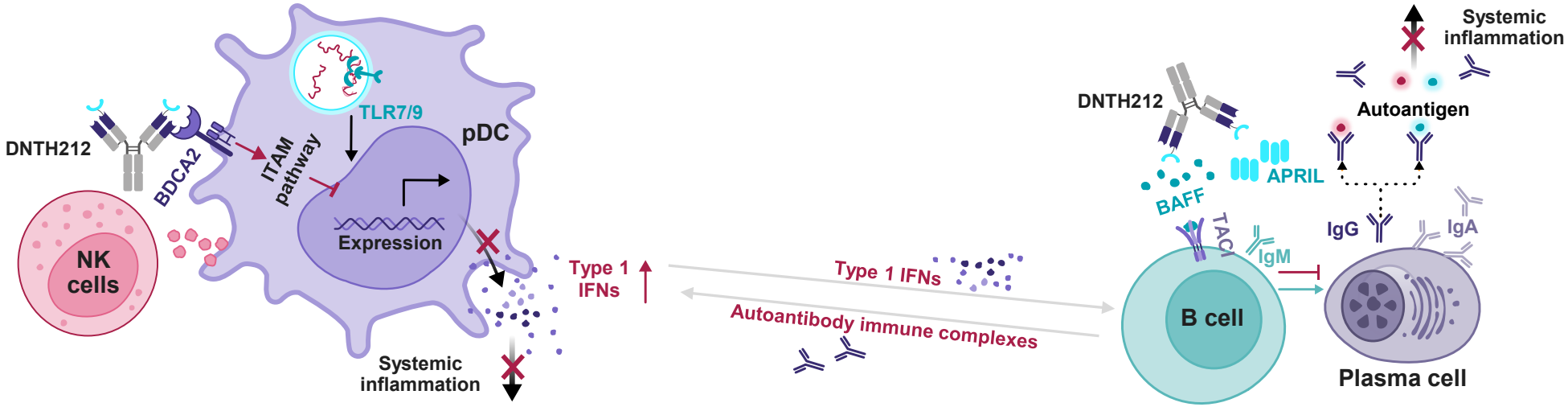
# Potential to drive superior clinical efficacy by targeting both the innate and adaptive immune systems

### Innate Immune System: Plasmacytoid Dendritic Cells (pDCs)

- Key cell type producing Type 1 interferon
- Type 1 interferon inhibition has been shown effective in multiple autoimmune diseases

### Adaptive Immune System: B Cells

- Generate autoantibodies, forming immune complexes that trigger inflammation and tissue damage
- Inhibiting BAFF/APRIL has been shown effective in multiple autoimmune diseases

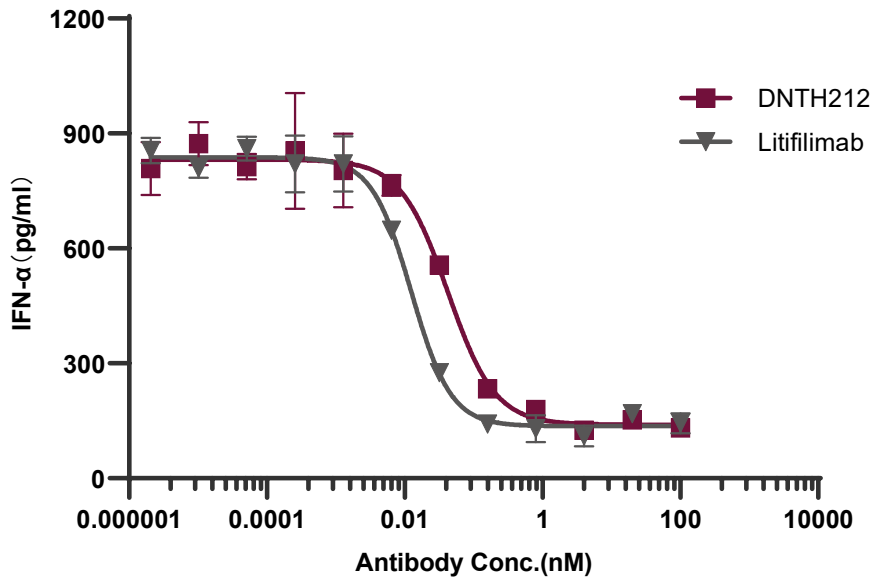



**Bifunctional approach addressing autoimmune diseases where both Type 1 interferon and B Cells are implicated has strong mechanistic rationale for potential best-in-class efficacy**

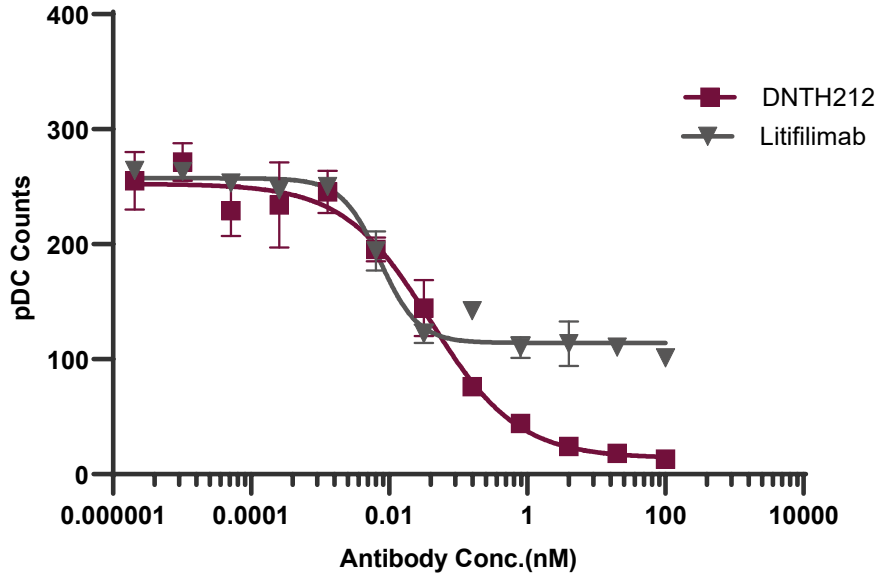

Panda SK, et al., Plasmacytoid dendritic cells in autoimmunity. Curr Opin Immunol. 2017 Feb;44:20-25.
Minaga K, et al., Plasmacytoid Dendritic Cells as a New Therapeutic Target for Autoimmune. Front Immunol. 2021 Jul 23;12:713779.

# DNTH212 achieves superior pDC depletion compared to litifilimab *in vitro*

## Comparable Suppression of Pro-Inflammatory Cytokine IFN-α[1]




## Deeper Depletion of pDC[2]

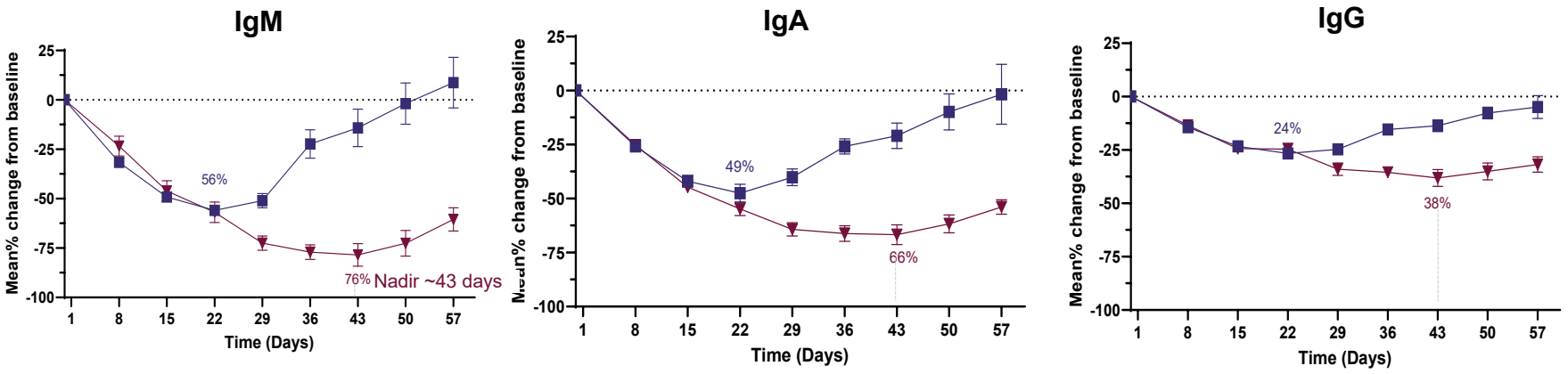



**pDC depletion removes a key cell type involved in Type 1 interferon production and activation of other immune cells which contribute to disease**

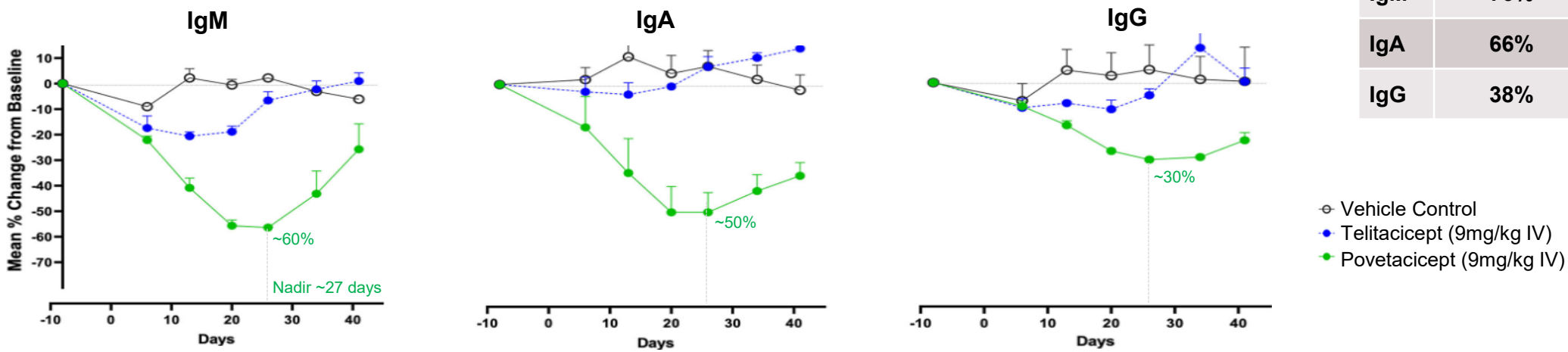
1. Method: Human PBMCs from a healthy donor were co-cultured with a TLR9 agonist and serially diluted antibodies for 24 hours. IFN-α release in the supernatant was measured using an HTRF kit.
2. Method: Human PBMCs from a healthy donor were co-cultured with serially diluted antibodies for 24 hours. pDC counts were assessed via flow cytometry.

# DNTH212 shows superior inhibition of IgM, IgA, and IgG compared to povetacicept following single dose in NHPs

S.C. DNTH212




IV povetacicept and telitacicept[1]




| Maximal Ig Inhibition | | |
|---|---|---|
| | DNTH212 10mg/kg | Povetacicept[1] 9mg/kg |
| IgM | 76% | ~60% |
| IgA | 66% | ~50% |
| IgG | 38% | ~30% |

**Deeper Ig reductions have potential to drive superior clinical efficacy while maintaining at least Q4W dosing**

Note: These data are derived from different studies at different points in time, with differences in methodology, design and populations. As a result, cross-trial comparisons cannot be made, and no head-to-head clinical trials of DNTH212 and other agents have been conducted.

1. Arthritis Rheumatol. 2023 Jul;75(7):1187-1202. Note: WT TACI (13-118) Fc:Telitacicept

# Priority indications with high unmet need and dual involvement of both innate and adaptive immune systems

## Prioritization Considerations

- Clinical evidence or biological rationale of both interferon / pDC and B-cell involvement
- Potential for improved efficacy from dual-mechanism approach
- High unmet need with current SoC and future treatments
- Well-established development pathways

## DNTH212 Priority Rheumatology Indications

**Sjögren's disease (SjD)**
~350,000 U.S. patients

**Systemic lupus erythematosus (SLE)**
~225,000 U.S. patients

**Dermatomyositis (DM)**
~50,000 U.S. patients

Targeting both innate and adaptive immune systems simultaneously may offer potential for meaningful differentiation

**Building a beachhead in rheumatology with clear clinical development and commercial synergies across priority indications**

Estimated U.S. patients per Dianthus meta-analysis and estimates.

# SLE: Strong BDCA2 and BAFF / APRIL validation with a proven development path support DNTH212 opportunity

**Disease Overview**

SLE is a systemic autoimmune disease affecting multiple organs such as the kidneys, skin, and joints and may lead to significant or life-threatening end-organ damage

**Supportive Evidence**

**BDCA2/Type 1 IFN (Innate)**

- Saphnelo approved
- Litifilimab Ph. 2 data

+

**BAFF/APRIL (Adaptive)**

- Benlysta approved
- Telitacicept Ph. 3 data (CN) (BAFF/APRIL)

**Mechanism Strength**

Proven skin and joint efficacy

Multi-organ responses (i.e. joints and kidney)

**DNTH212 Target Opportunity**

**Potential to more effectively address multiple disease manifestations with lower corticosteroid burden**

**~225K U.S. Patients**

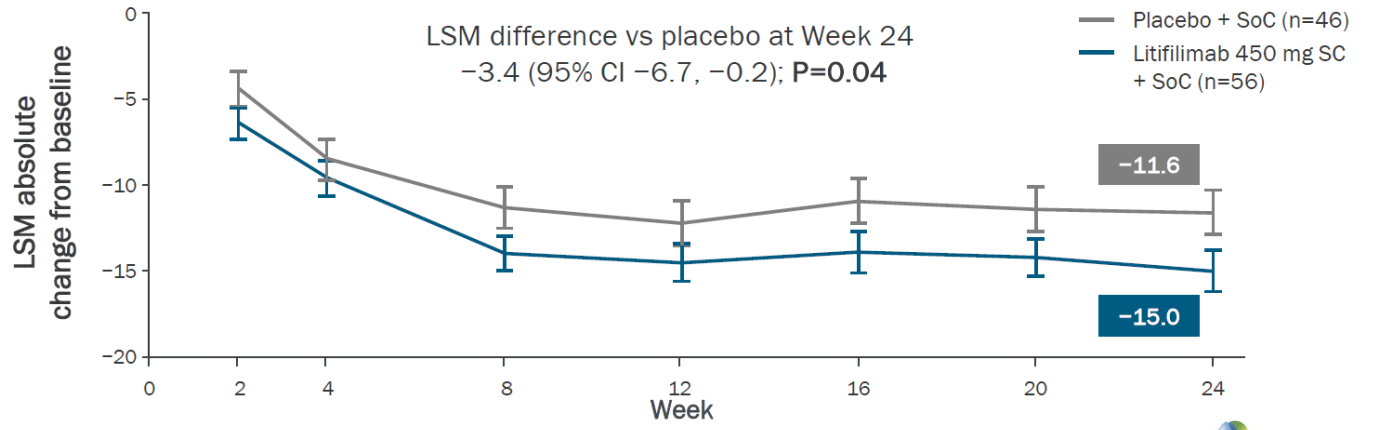
Source: Company filings, presentations and clinicaltrials.gov
Estimated U.S. patients per Dianthus meta-analysis and estimates. ClearView analysis

# SLE: Potential for dual mechanism to address key unmet needs despite approved treatments

## Litifilimab Ph. 2 data[1] and multiple approvals (Saphnelo, Benlysta) support dual-mechanism approach in SLE

*Improved Joint Activity: Litifilimab significantly reduced the mean total number of active joints vs placebo*

Total active joint count*† in patients with SLE and active skin disease and joint involvement (N=132‡) (primary endpoint at Week 24)[1]




Biogen

Benlysta (belimumab)

Saphnelo® (anifrolumab-fnia)

**Utilized concurrently with steroids**

## Significant Unmet Need

- SLE is a heterogenous disease - potential for dual-mechanism approach to impact multiple organs vs. single mechanism alone
- ~45% of SLE patients categorized as moderate-to-severe[2]

## DNTH212 Target Opportunity

**Potential to more effectively address multiple disease manifestations with lower corticosteroid burden**

1. Biogen Lupus Virtual Investor Seminar (2025)
2. Lupus Sci Med. 2025 Dec 23;12(2):e001766

# SjD: High unmet need for a single therapy that can address multiple disease manifestations

**Disease Overview**

Sjögren's destroys mucosal exocrine glands causing extreme dry eyes and mouth (sicca) and leads to debilitating systemic symptoms such as fatigue and joint pain

**Supportive Evidence**

**BDCA2/Type 1 IFN (Innate)**

- ✓ Evidence for glandular pDC infiltration

+

**BAFF/APRIL (Adaptive)**

- ✓ Telitacicept Ph. 3 data (CN) (BAFF/APRIL)
- ✓ Ianalumab Ph. 3 data (BAFF)

**Mechanism Strength**

Potential for improved patient reported outcomes, including systemic disease manifestations

Proven impact on primary endpoint (i.e. ESSDAI)

**DNTH212 Target Opportunity**

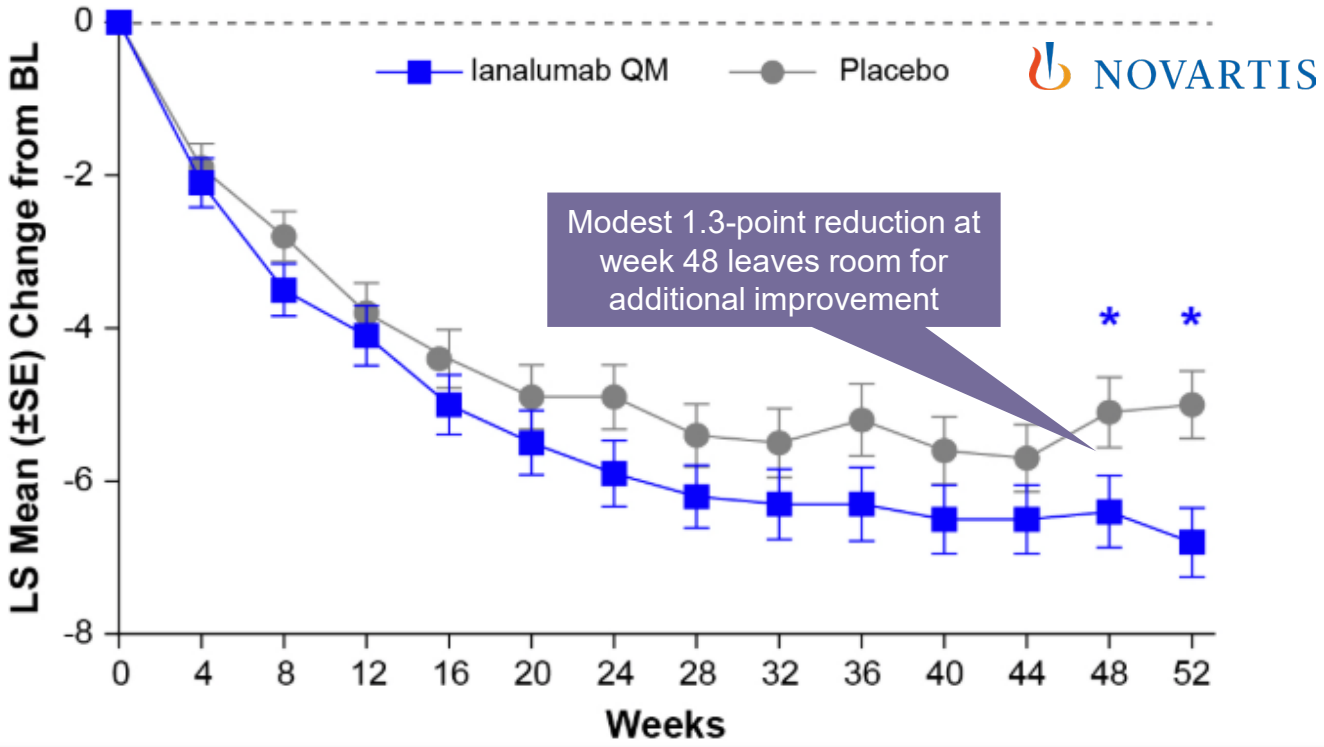
**More effectively address multiple disease manifestations including dry eye, dry mouth and other systemic symptoms (i.e. pain, fatigue)**

**~350K U.S. Patients**

Source: Company filings, presentations and clinicaltrials.gov.
Estimated U.S. patients per Dianthus meta-analysis and estimates.
ClearView analysis

# SjD: Potential for increased efficacy from dual-mechanism approach vs. targeting BAFF receptor only

## Ianalumab Ph. 3[1] highlights potential for B-cell approaches to lower ESSDAI




**"Additional secondary endpoints showed a trend towards improvement in both studies"** – Ianalumab ACR 2025[1]

## Significant Unmet Need

- Fatigue, pain and cognitive dysfunction are significant drivers of patient disability[2]
- 70% increased mortality compared with the general population due to infections, cancer and cardiovascular diseases[2]

## DNTH212 Target Opportunity

**More effectively address multiple disease manifestations including dry eye, dry mouth and other systemic symptoms (i.e. pain, fatigue)**

1. Ianalumab ACR 2025 abstract.
2. Pagnoni et al. J Rheum 2026, 53(2).

# DM: High unmet need for a single therapy that can more effectively address the disease

**Disease Overview**

DM causes progressive muscle weakness, painful rash, pruritus and interstitial lung disease; limited therapeutic options beyond corticosteroids (i.e. IVIg)

**Supportive Evidence**

**BDCA2/Type 1 IFN (Innate)**

- Dazukibart (IFN-β) Ph. 2 data
- Positive Saphnelo case reports

+

**BAFF/APRIL (Adaptive)**

- IVIg approved, Vyvgart Ph. 2
- Benlysta small studies / case reports

**Mechanism Strength**

Improvement in skin symptoms and TIS (Total Improvement Score)

Clear evidence of antibody driven pathology and improvement in TIS

**DNTH212 Target Opportunity**

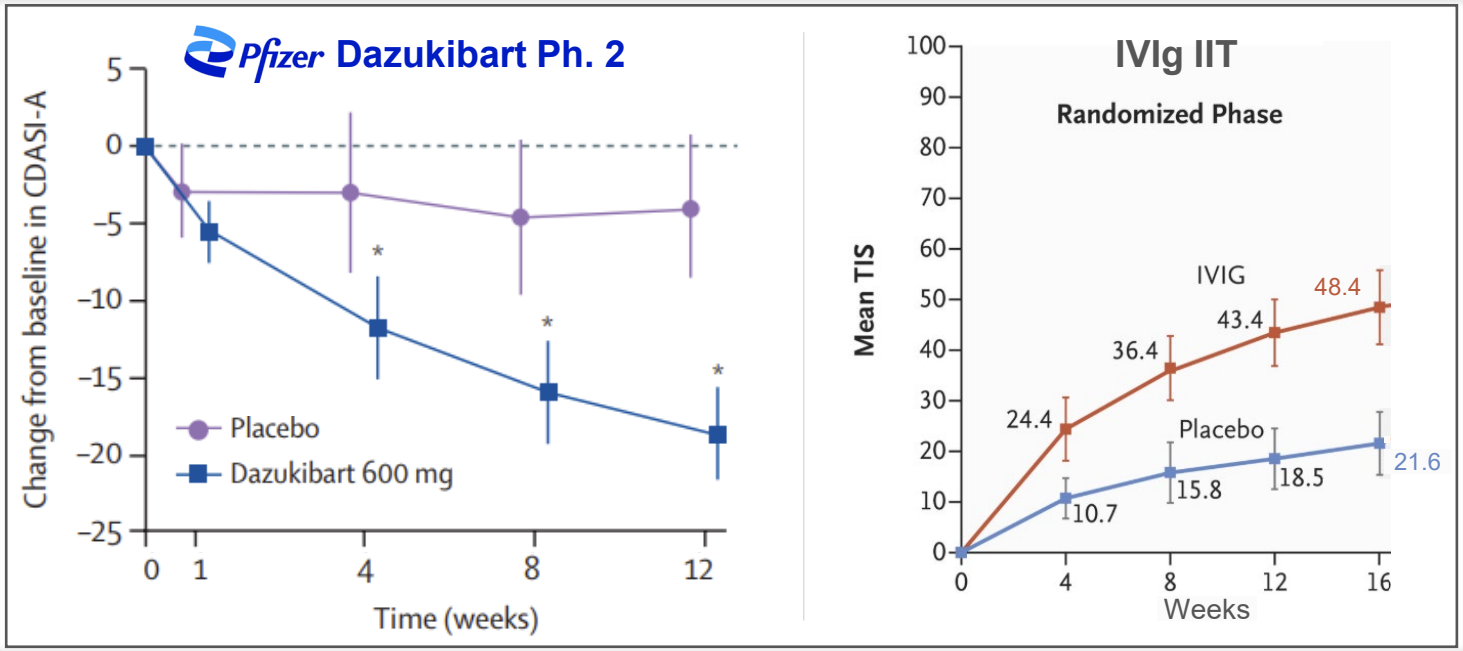
**More effectively address the disease and prevent damage over its course with lower corticosteroid burden**

**~50K U.S. Patients**

Source: Company filings, presentations and clinicaltrials.gov.
Estimated U.S. patients per Dianthus meta-analysis and estimates.
ClearView analysis
Dazukibart binds and neutralizes IFN-β. Saphnelo is a Type 1 receptor antagonist.

# DM: Dual-mechanism approach may more effectively address the disease

## Dazukibart Ph. 2 data[1] and IVIg[2] lay foundation for multi-organ efficacy in DM




"This study showed that IFNβ is an important mediator of disease activity in skin-predominant dermatomyositis" - Lancet 2025; 405; 137–46

## Significant Unmet Need

- Muscle weakness, fatigue and pain remain leading causes of morbidity[3]
- Limited therapeutic treatment options
- Majority of patients are managed with systemic steroids, which can carry long-term safety concerns[4]

## DNTH212 Target Opportunity

**More effectively address the disease and prevent damage over its course with lower corticosteroid burden**

1. Dazukibart Phase 2 publication.
2. Trial of Intravenous Immune Globulin in Dermatomyositis.
3. Christopher-Stine; BMC Rheumatology 2025.
4. George et al. RMD Open 2026.

# Preliminary Ph. 1 Part A healthy volunteer data expected in Q1'27; based upon data to date, Part B SLE dosing initiated

### Healthy Volunteers (Part A)

~46 HVs enrolled into seven cohorts:

- Treated (N= up to 6)
- Placebo (N= up to 2)

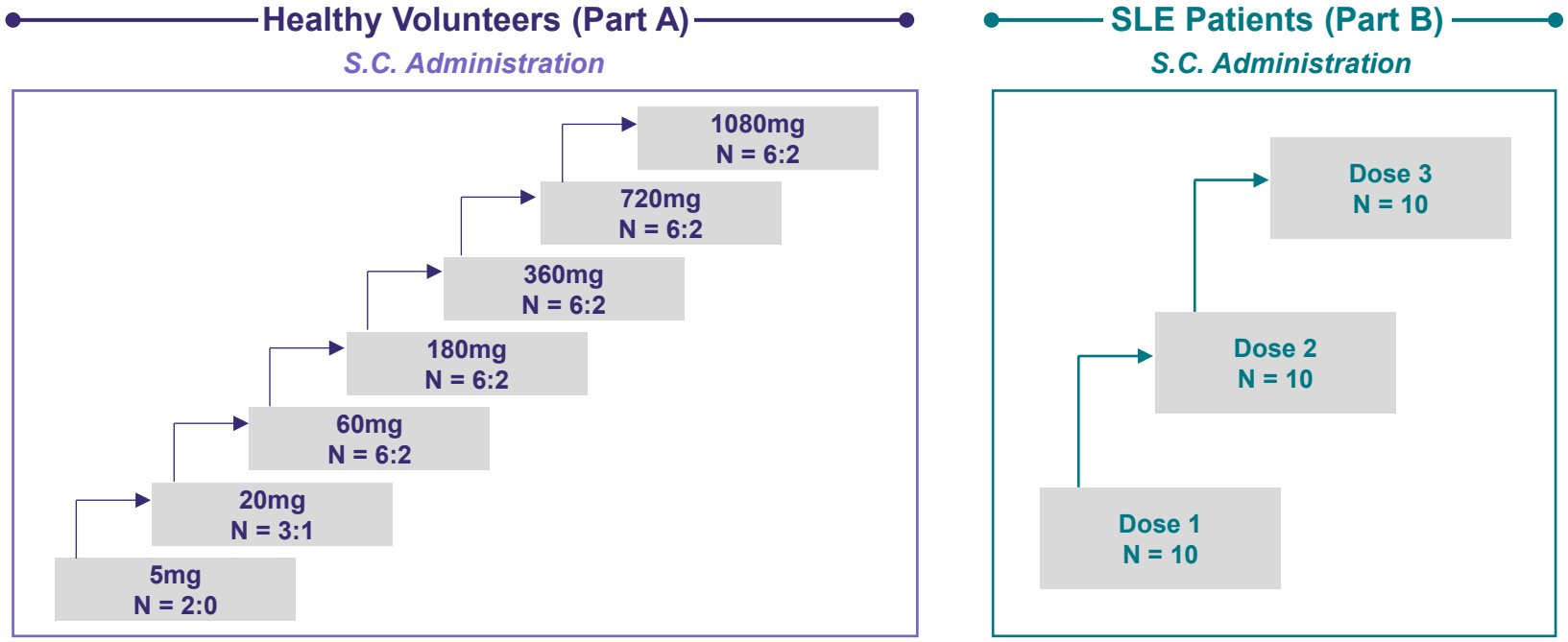
### SLE Patients (Part B)

~30 patients enrolled into three cohorts:

- Treated (N= up to 10)

### Key Parameters

- **Safety, PK, and PD as well as other biomarkers and preliminary efficacy**




**First SLE patient dosed in Part B in Oct. '26, with preliminary data expected 2H'27; Dianthus-led MAD study planned to initiate in Q2'27, with preliminary data expected 2H'27**

# DNTH212 TPP aims to deliver superior efficacy in a safe and well-tolerated therapy with patient friendly convenience



## EFFICACY

Bifunctional approach has potential for *superior* efficacy in various disease states versus only targeting innate or adaptive immune system



## SAFETY

Inhibiting Type 1 interferon or BAFF/APRIL has been generally safe and well tolerated

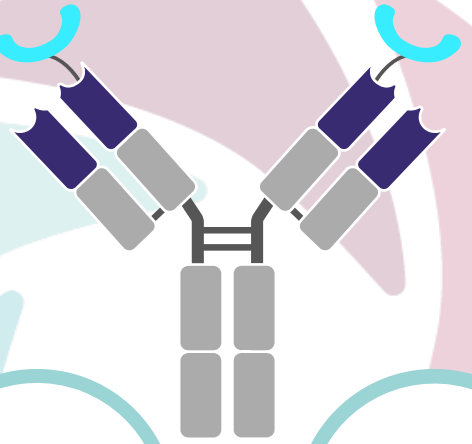


## CONVENIENCE

Targeting patient friendly S.C. self-administration with Q4W or less frequent dosing

**Achieving the TPP could position DNTH212 as a first-line biologic across a range of indications**

# Achieving DNTH212 TPP could position DNTH212 as a first-line, best-in-disease therapy across multiple indications



| Clinically Validated MoA | Superior Efficacy | Patient-friendly Convenience | Favorable Safety Potential | Pipeline-in-a-Product | Robust IP Protection |
|---|---|---|---|---|---|
| Type 1 interferon and BAFF/APRIL are known drivers of autoimmune disease | Targeting innate and adaptive immune systems has potential for superior efficacy | Potential S.C. self-administration Q4W or less frequent | Inhibiting Type 1 interferon or BAFF/APRIL has been generally safe and well tolerated | Broad opportunity across multiple diseases where Type 1 interferon and B-cells are implicated | Composition of matter patent expected to expire no earlier than 2044 |

DNTH212 is an investigational agent that is not approved as a therapy in any indication in any jurisdiction worldwide.

# DNTH312: Expanding our leadership position across severe autoimmune diseases

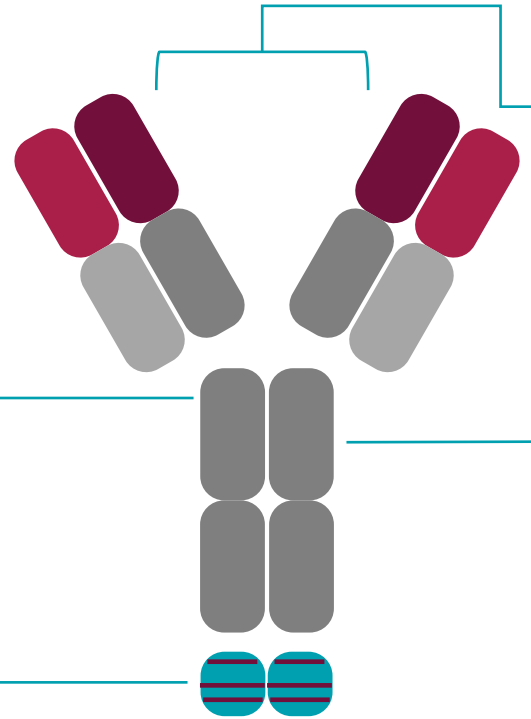
# DNTH312 is a first-in-class, next-generation bifunctional fusion protein combining claseprubart and TACI targeting two complementary, validated pathways

**IgG1 Fc**

No effector function (“silent”)

**TACI-CRD2 Domain**

High affinity resulting in potent inhibition of B cell activity via BAFF/APRIL



**Claseprubart**

Potent Classical Pathway inhibition via aC1s

**YTE Mutation on Fc**

Comparable ~22-day half-life to claseprubart in NHPs

**DNTH312 is designed to deliver robust Classical Pathway inhibition via aC1s and inhibition of B cell activity via BAFF/APRIL, targeting two disease modifying mechanisms with a potential for enhanced efficacy**



DNTH312 is an investigational agent that is not approved as a therapy in any indication in any jurisdiction worldwide.

# Goal to drive superior clinical efficacy by targeting both aC1s and BAFF/APRIL

### Upstream Classical Pathway or aC1s inhibition results in:

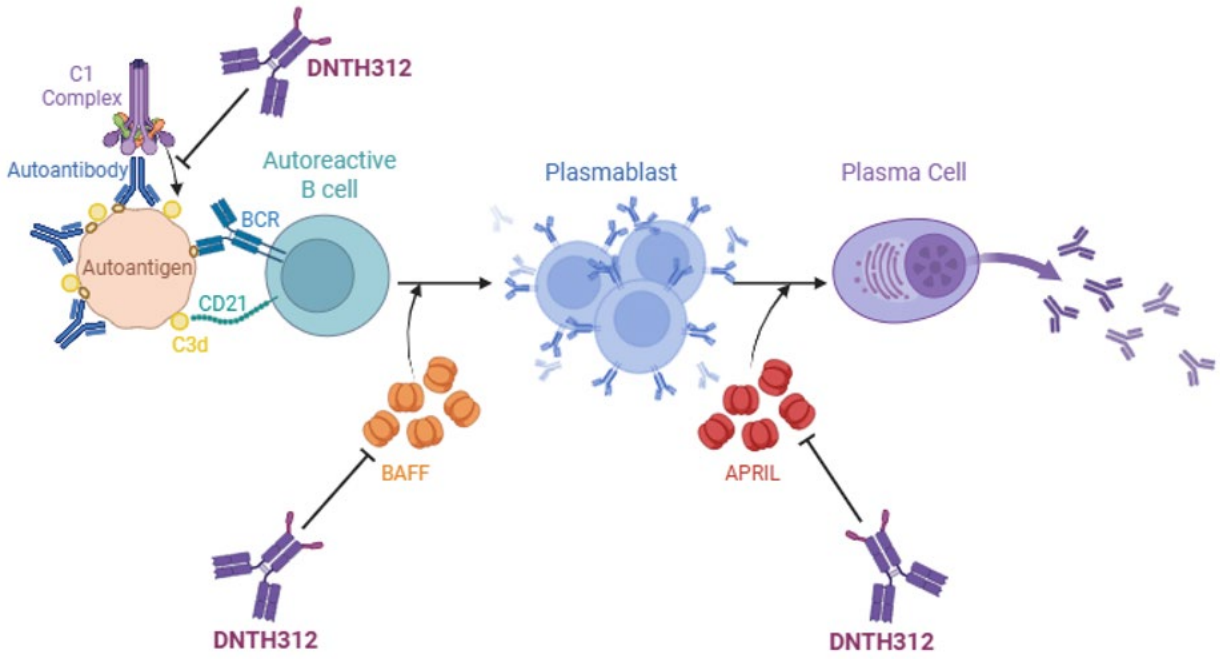
- Reduction of immune-complex-mediated tissue damage driven by MAC formation
- Reduction in recruitment and activation of inflammatory cells (including B cells) through inhibition of C3a and C5a split product formation
- Decrease of complement-dependent opsonization and cell destruction

**Example:** In MG, neuromuscular signaling may be preserved through inhibition of downstream inflammatory and tissue-destructive pathways

### BAFF/APRIL (B Cell) inhibition results in:

- Reduction in both short-lived and long-lived plasma cells responsible for the generation of autoantibodies
- Lower levels of autoantibodies limit the formation of immune complexes that trigger inflammation and tissue damage

**Example:** In MG, addressing an upstream driver may reduce the source of the antibodies that initiate complement activation




**Bifunctional approach addressing autoimmune diseases where both the Classical Pathway and B Cells are implicated has strong mechanistic rationale for potential best-in-class efficacy**

# DNTH312 builds on claseprubart best-in-class profile and expands Dianthus' leadership in neuromuscular markets

## Initial Focus: Grow Leadership Position in Neuromuscular Diseases

- ✓ Seeks to provide more benefit beyond claseprubart or BAFF/APRIL alone for patients with autoimmune neuromuscular diseases, such as Myasthenia Gravis
- ✓ Builds on Dianthus' established neuromuscular expertise with claseprubart, creating synergies in clinical development, regulatory, and commercial

## Future Opportunity: Expand into Other Therapeutic Areas

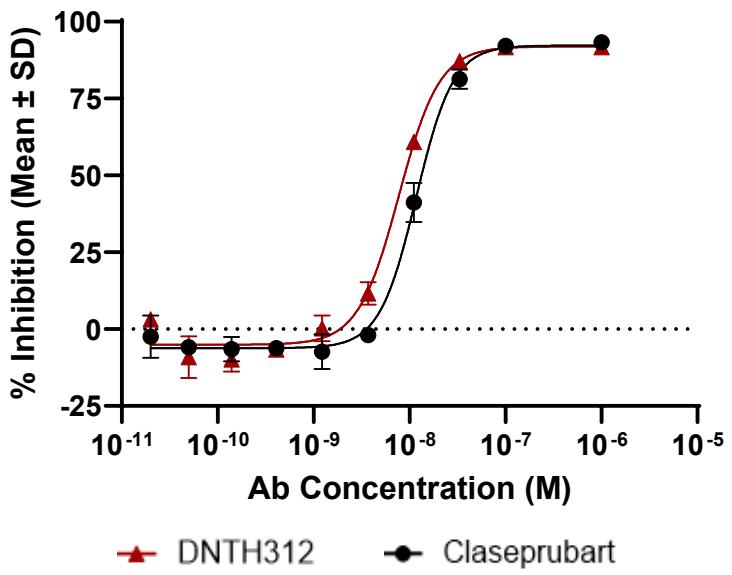
- ✓ Leverages dual MoA approach via validated pathways for new potential indications beyond either mechanism alone
- ✓ Builds on Dianthus' BAFF/APRIL expertise with potential for clinical development synergies with DNTH212

| Deeper Responses | Broader Symptom Control | Larger Target Populations |
|---|---|---|
| Target multiple sources of pathology that affect a single system and occur in a single patient | Target multiple sources of pathology that affect multiple different organ systems in more patients | Target additional diseases beyond neuromuscular conditions with heterogeneous pathology |

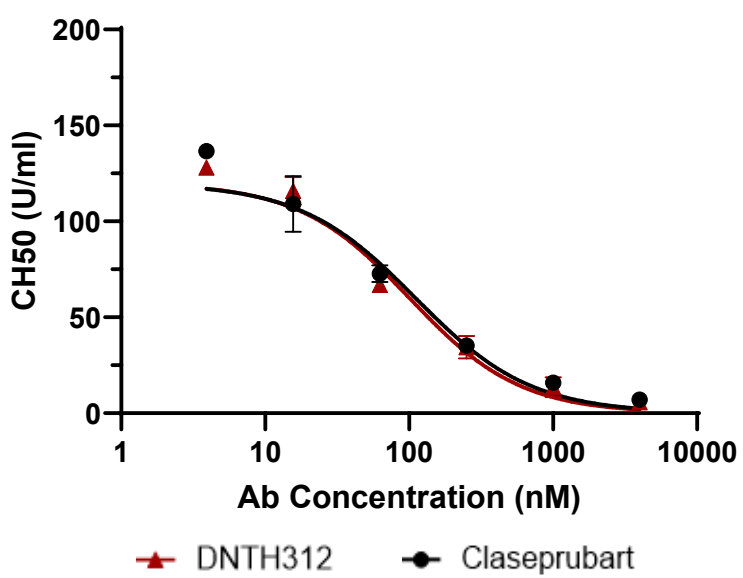
# DNTH312 demonstrates comparable *in vitro* potency and aC1s inhibition to potentially best-in-class claseprubart

## Functional Assays of Classical Pathway Inhibition

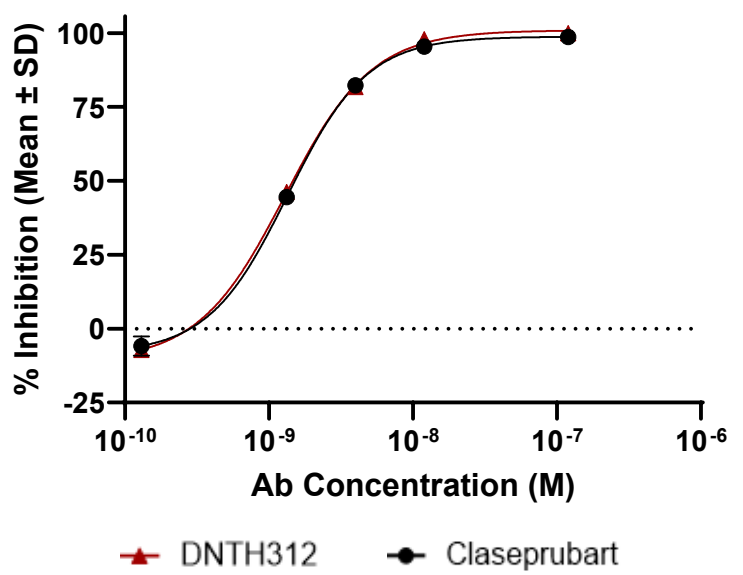
### Direct RBC Hemolysis in Human Serum




### CH50 Assay of RBC Lysis in Human Serum



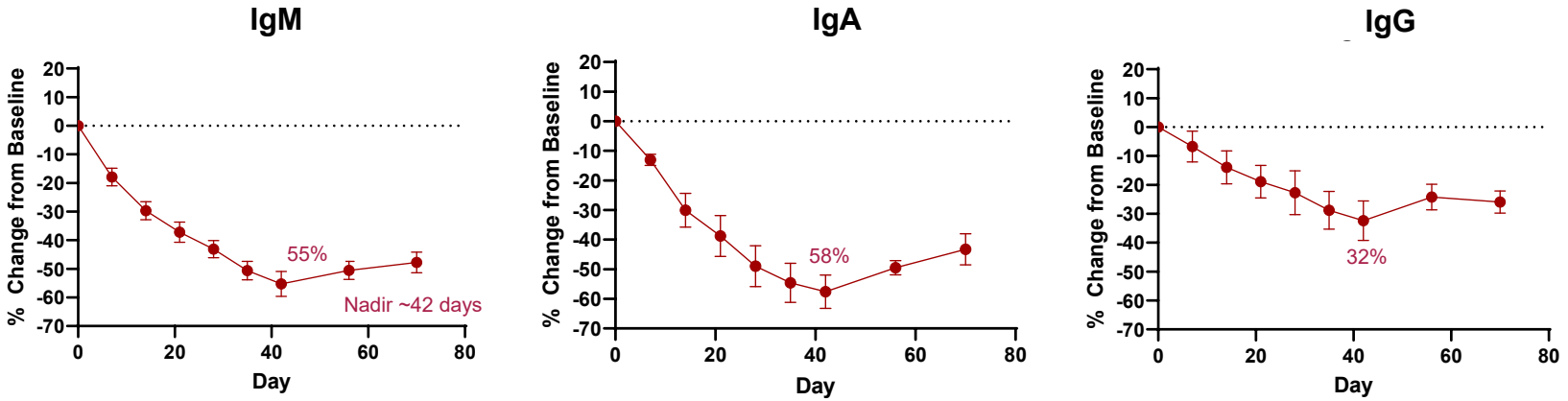

### Wieslab Classical Pathway Assay in Human Serum



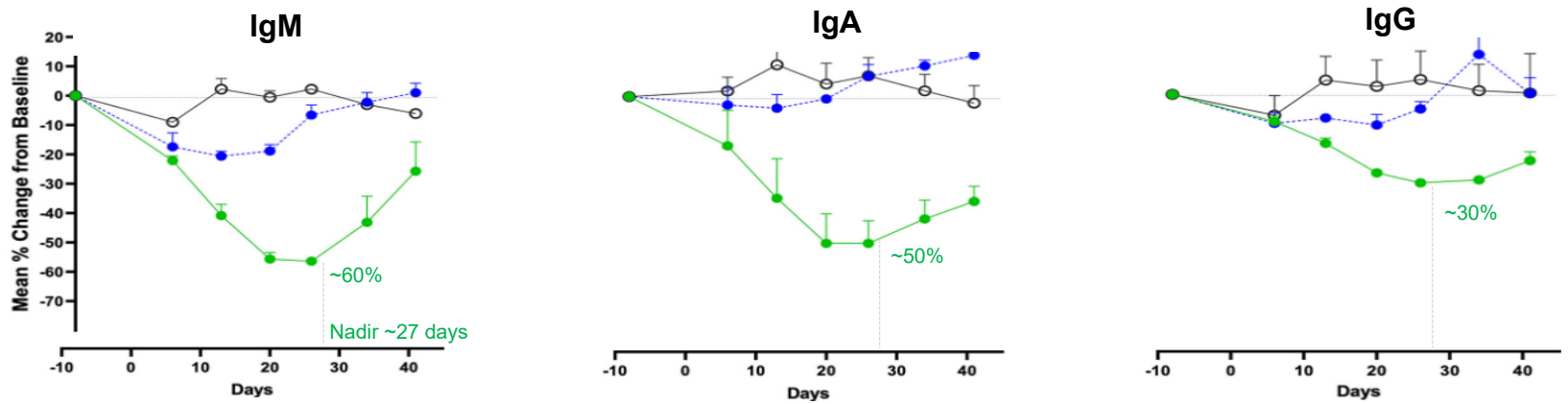

Note: Data shown are the average of 2-3 experiments conducted for each of the functional assays (Direct RBC Hemolysis, CH50, and Wieslab).

# DNTH312 shows similar depth of IgM, IgA, and IgG reduction vs. povetacicept following single dose in NHPs

**DNTH312**




**IV povetacicept and telitacicept[1]**




| | Maximal Ig Inhibition | |
|---|---|---|
| | DNTH312 10mg/kg | Povetacicept[1] 9mg/kg |
| IgM | 55% | ~60% |
| IgA | 58% | ~50% |
| IgG | 32% | ~30% |

| | NHP Half-life | |
|---|---|---|
| | DNTH312 | Povetacicept |
| T1/2 | ~22 days | ~5 days[2] |

**DNTH312 demonstrates similar depth of Ig reductions vs. povetacicept, with ~4X longer half-life**

Note: These data are derived from different studies at different points in time, with differences in methodology, design and populations. As a result, cross-trial comparisons cannot be made, and no head-to-head clinical trials of DNTH312 and other agents have been conducted. NHP: Non-human primate.

[1]Arthritis Rheumatol.2023 Jul;75(7):1187-1202. Note: WT TACI (13-118)-Fc:Telitacicept.

[2]Internally generated data; povetacicept is produced using sequence from patent WO2021226551A1.

# DNTH312 aims to be Phase 1 ready by YE'27

## Key Benefits of DNTH312

**Comparable NHP half-life to claseprubart**

DNTH312 has a similar NHP half-life (22 days) to claseprubart, which has an approximately 60-day half-life in humans

**Comparable *in vitro* aC1s inhibition and potency to claseprubart**

Demonstrated across multiple complement *in vitro* PD functional assays

**Comparable Ig reductions to povetacicept with a longer half-life in NHPs**

DNTH312 is targeting infrequent, S.C. self-administration

**Clinically validated MoAs combined have the potential for superior clinical efficacy**

Inhibition of the Classical Pathway or aC1s has been clinically validated in gMG, CIDP, MMN and more

BAFF/APRIL has been clinically validated in MG, SjD, SLE, IgAN, RA and more

**DNTH312 extends our neuromuscular leadership position with new IP protection expected through at least 2047, beyond claseprubart IP protection expected through at least 2043, excluding extensions**



DNTH312 is an investigational agent that is not approved as a therapy in any indication in any jurisdiction worldwide.

# Recap of Dianthus Leadership in Severe Autoimmune Diseases

# Building the leading autoimmune-focused biotech through potential best-in-disease innovation



## DNTH312: Next-generation Leadership

Leverage Core Competencies
to Expand Leadership
Across Severe Autoimmune



## DNTH212: Expansion into Rheumatology

- SjD
- SLE
- DM

U.S. Prevalence: ~625K



## Claseprubart: Foundation in Neurology

- EMERGE — Ph. 3 gMG
- CAPTIVATE — Ph. 3 CIDP
- MOMENTUM — Ph. 2 MMN

U.S. Prevalence: ~150K



Estimated U.S. patients per Dianthus meta-analysis and estimates.

# Advancing a leading autoimmune-focused biotech with two clinical-stage programs

| Program | Indication | Preclinical | Ph. 1 | Ph. 2 | Ph. 3 | Upcoming Milestones |
|---|---|---|---|---|---|---|
| Claseprubart *aC1s* | gMG* >100,000 U.S. Patients | | | | | • Ph. 3 top-line data expected in 2H'28 |
| | CIDP >40,000 U.S. Patients | | | | | • Ph. 3 Part A top-line data expected by YE'27<br>• Ph. 3 Part B top-line data expected by YE'28 |
| | MMN >10,000 U.S. Patients | | | | | • Ph. 2 top-line data expected in December '26<br>• Ph. 3 initiation planned in 2027 |
| DNTH212 *BDCA2 and BAFF/APRIL* | SjD, SLE, DM *~625,000 U.S. Patients (Combined)* | | *Healthy volunteers (Part A)* *SLE patients (Part B)* | | | • Ph. 1 HV preliminary data (Part A) expected Q1'27<br>• Ph. 1 MAD initiation expected Q2'27<br>• Ph. 1 SLE preliminary data (Part B) and preliminary MAD data expected in 2H'27 |
| DNTH312 *claseprubart and TACI* | Autoimmune Diseases | | | | | • Ph. 1 ready expected by YE'27 |

**Strong balance sheet with ~$1.2B[1] of cash & runway expected into 2030**
**~57.6M shares outstanding[2]**

* FDA Orphan Drug Designation received in May 2026 for claseprubart in Myasthenia Gravis.
1. Cash includes cash, cash equivalents and investments as of 9/30/26; 2. Includes 56.3M shares outstanding as of November 2, 2026 and assumes the exercise of all outstanding pre-funded warrants.

# Multiple internal and external catalysts expected across all three programs in 2027

| Program | Indication | 1H'27 | 2H'27 |
|---|---|---|---|
| **Claseprubart** *aC1s* | **gMG** | | **Claseprubart** Updated Ph. 3 EMERGE top-line guidance (YE'27) |
| | **CIDP** | **Riliprubart** (Sanofi; aC1s) Ph. 3 VITALIZE (H2H vs. IVIg) data and submission (2H'27); **Empasiprubart** (argenx; C2) Ph. 3 EMVIGORATE & EMNERGIZE data (2H'27) | **Claseprubart** Ph. 3 CAPTIVATE Part A top-line data (YE'27) |
| | **MMN** | **Claseprubart** Ph. 3 initiation planned (2027) | **Empasiprubart** (argenx; C2) potential approval (2027) |
| **DNTH212** *BDCA2 and BAFF/APRIL* | **SjD, SLE, DM** | **DNTH212** Ph. 1 Part A SAD HV preliminary data (Q1'27); **DNTH212** Dianthus-led Ph. 1 MAD study initiation (Q2'27); **Litifilimab** (Biogen; BDCA2) Ph. 3 CLE (Q2'27) | **DNTH212** Ph. 1 Part B SLE patient preliminary data and preliminary MAD data (2H'27); **Sibeprenlimab** (Otsuka; APRIL) Ph. 2 SjD data (2H'27) |
| **DNTH312** *Claseprubart and TACI* | **Autoimmune Diseases** | **Telitacicept** (Vor Bio; BAFF/APRIL) Ph. 3 UPSTREAM gMG top-line (1H'27) | **DNTH312** Ph. 1 ready (YE'27) |

Peer Milestone

SAD: single ascending dose; MAD: multiple ascending dose; HV: healthy volunteers; H2H: head-to-head; CLE: cutaneous lupus erythematosus; SjD: Sjögren's disease; gMG: generalized myasthenia gravis.
Source: Company filings, presentations, and clinicaltrials.gov.

# Appendix

# C1s is a clinically validated target in the Classical Complement Pathway with an FDA approved therapy

1

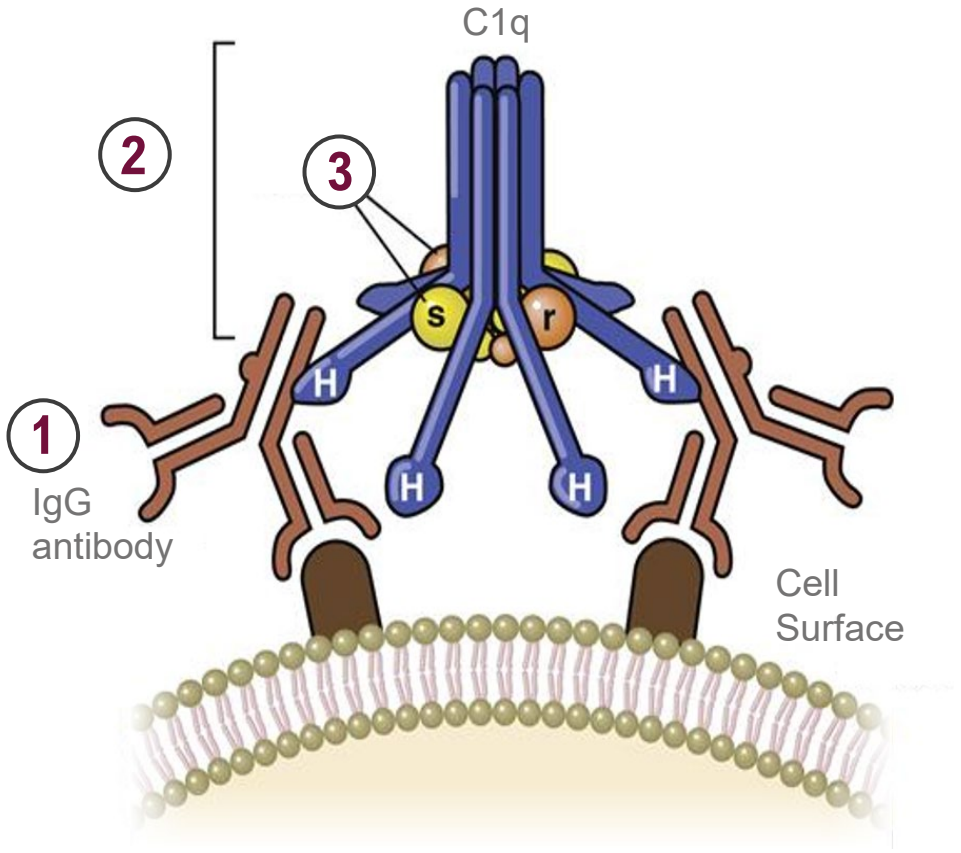
**Classical Pathway**
The only pathway activated by the presence of IgG and IgM, which bind to the **C1 complex**

2

**The C1 complex**
The initial component of the classical complement pathway consisting of C1q, C1r and C1s

3

**Active C1s**
A serine protease that executes catalytic function of the C1 complex, leading to MAC formation

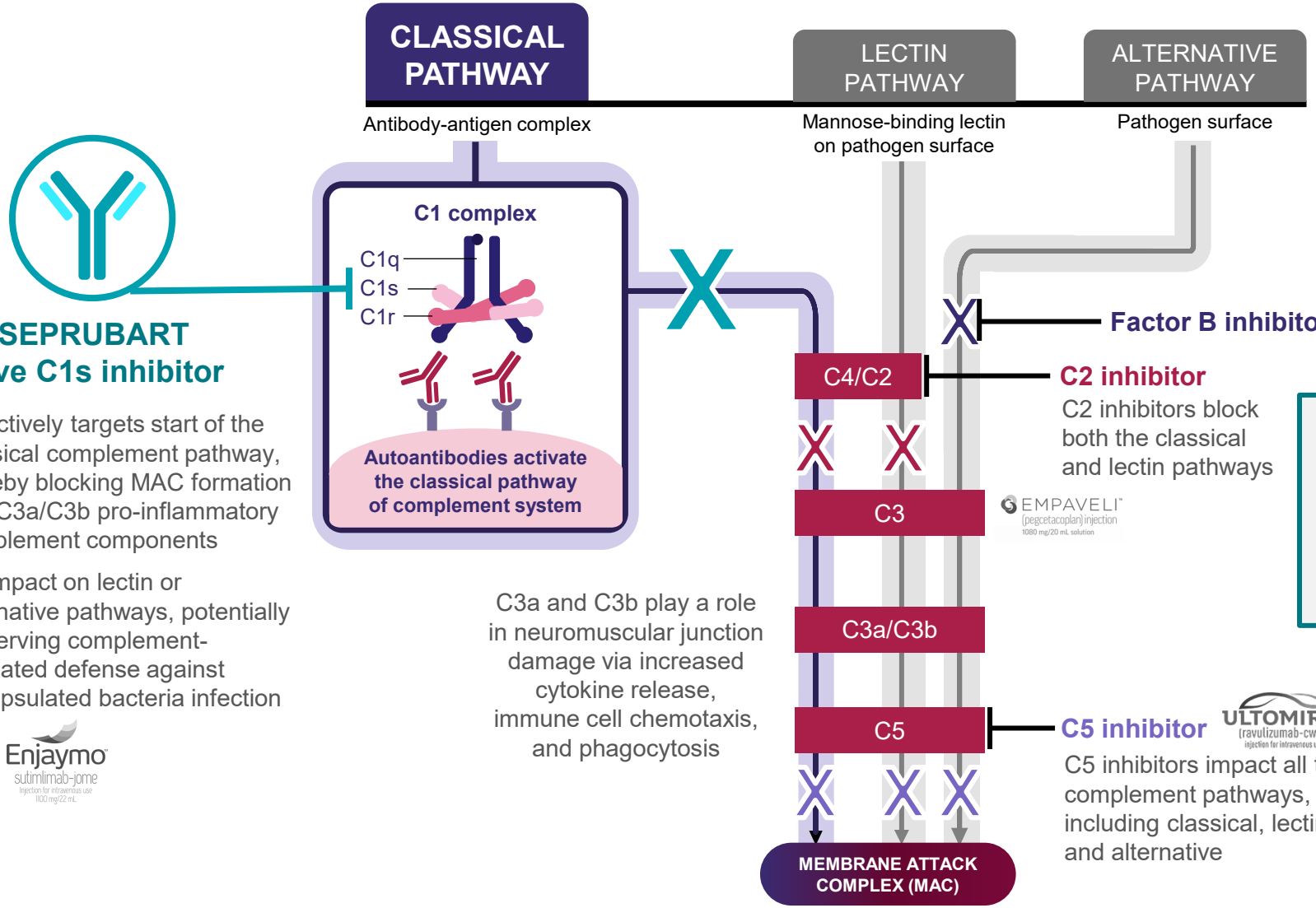



**C1s is the only target of the C1 complex with an FDA approved therapy**
Enjaymo, FDA approved in 2022 for CAD, is a C1s inhibitor but is not selective to the active form and dosed I.V. at 6,500-7,500mg every two weeks

**Active C1s inhibition has demonstrated clinical benefit in MG and CIDP**
Claseprubart MaGic and CAPTIVATE results provide clinical PoC for aC1s in neuromuscular diseases

Enjaymo information sourced from prescribing information.

# Claseprubart has potential to have a differentiated safety profile by inhibiting only the classical pathway




## Potential Safety Advantages of Active C1s Inhibition

Unlike approved C3, C5, and Factor B inhibitors, Enjaymo (approved C1s inhibitor) has **no FDA Boxed Warning and REMS, or prophylactic antibiotic requirement prior to vaccination**



Enjaymo® prescribing information, rev. 11/2024

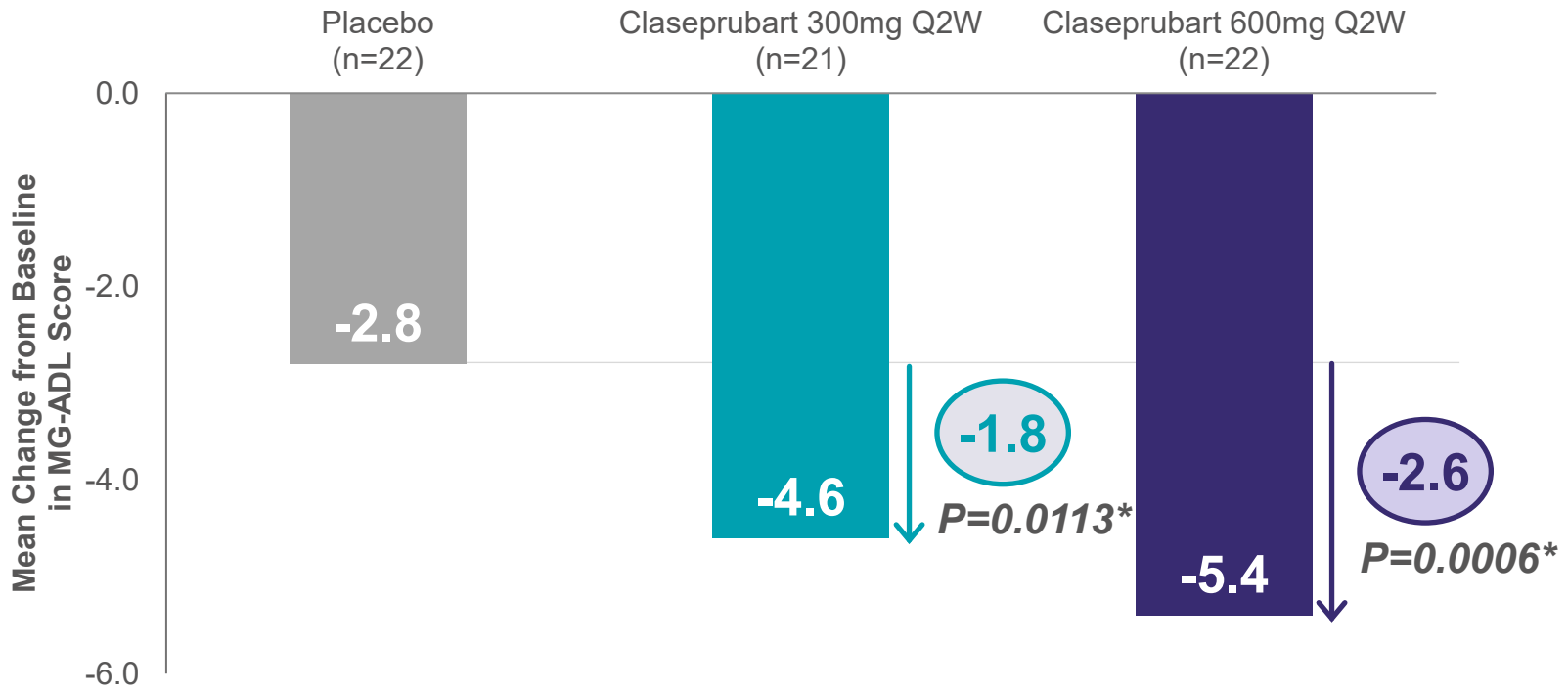
# Participant baseline characteristics were generally well balanced across arms

| AChR+ gMG participants | Placebo (N=22) | Claseprubart 300mg/2mL Q2W (N=21) | Claseprubart 600mg/4mL Q2W (N=22) |
|---|---|---|---|
| Age, mean (SD), years | 52.2 (16.5) | 57.1 (13.7) | 55.3 (12.0) |
| Male, n (%) | 13 (59%) | 14 (67%) | 10 (45%) |
| Weight, mean (SD), pounds | 195.0 (48.0) | 192.5 (35.5) | 179.0 (35.4) |
| Duration of disease, median (range), years | 7.7 (0.4 – 21.2) | 3.0 (0.5 – 22.1) | 7.6 (1.0 – 37.3) |
| MG-ADL score at baseline, mean (SD) | 8.5 (2.9) | 8.2 (2.2) | 8.4 (2.6) |
| QMG score at baseline, mean (SD) | 14.2 (5.8) | 12.2 (2.7) | 12.2 (3.6) |
| MG Composite score at baseline, mean (SD) | 15.0 (7.8) | 16.3 (4.5) | 16.0 (5.3) |
| MG-QoL-15r score at baseline, mean (SD) | 14.3 (7.0) | 15.4 (6.6) | 14.9 (5.9) |
| MGFA class at screening, n (%) | | | |
| II | 7 (32%) | 11 (52%) | 12 (55%) |
| III | 12 (55%) | 10 (48%) | 9 (41%) |
| IVa | 3 (14%) | 0 (0%) | 1 (5%) |
| Prior thymectomy, n (%) | 8 (36%) | 6 (29%) | 7 (32%) |
| Baseline corticosteroid use, n (%) | 19 (86%) | 17 (81%) | 20 (91%) |
| Number of ISTs at baseline, n (%) | | | |
| 1 | 11 (50%) | 10 (48%) | 12 (55%) |
| >1 | 11 (50%) | 11 (52%) | 10 (45%) |
| FcRn use in prior 24 months, n (%) | 0 (0%) | 1 (5%) | 0 (0%) |
| Prior complement use, n (%) | 0 (0%) | 0 (0%) | 0 (0%) |

gMG, generalized Myasthenia Gravis; SD, standard deviation; N (n), number; MG-ADL, Myasthenia Gravis Activities of Daily Living scale; QMG, Quantitative Myasthenia Gravis score; MGFA, Myasthenia Gravis Foundation of America clinical classification; IST, immunosuppressive therapy; MGC, Myasthenia Gravis Composite; MG-QOL-15r, Myasthenia Gravis quality of life 15 revised.

# Statistically significant improvement in MG-ADL score for both claseprubart arms vs. placebo at Week 13

## Mean Change in MG-ADL Score from Baseline at Week 13



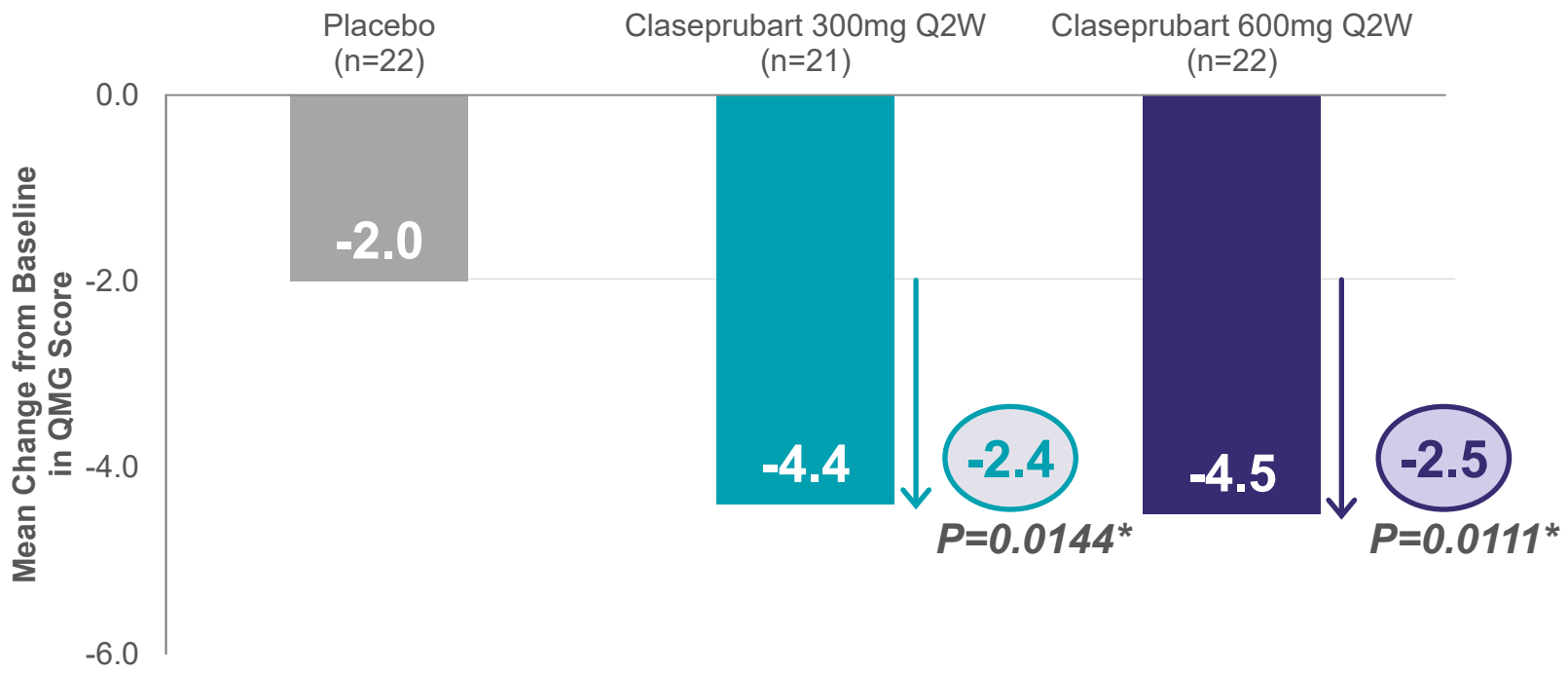

| P-value | 300mg/2mL | 600mg/4mL |
|---|---|---|
| One-sided | P=0.0113* | P=0.0006* |
| Two-sided | P=0.0227** | P=0.0013** |

**Statistically significant and clinically meaningful reductions in MG-ADL across both treatment arms**

The change from baseline in MG-ADL was analyzed using a mixed effect model for repeated measures (MMRM) with treatment group, visit, treatment by visit interaction, stratification factors, and baseline measure included.
*One-sided p-values are presented for comparisons of claseprubart vs placebo, with any p-value below 0.1 considered nominally statistically significant.
**Two-sided p-values are presented for comparisons of claseprubart vs placebo, with any p-value below 0.05 considered nominally statistically significant.

# Statistically significant improvement in QMG score for both claseprubart arms vs. placebo at Week 13

## Mean Change in QMG Score from Baseline at Week 13



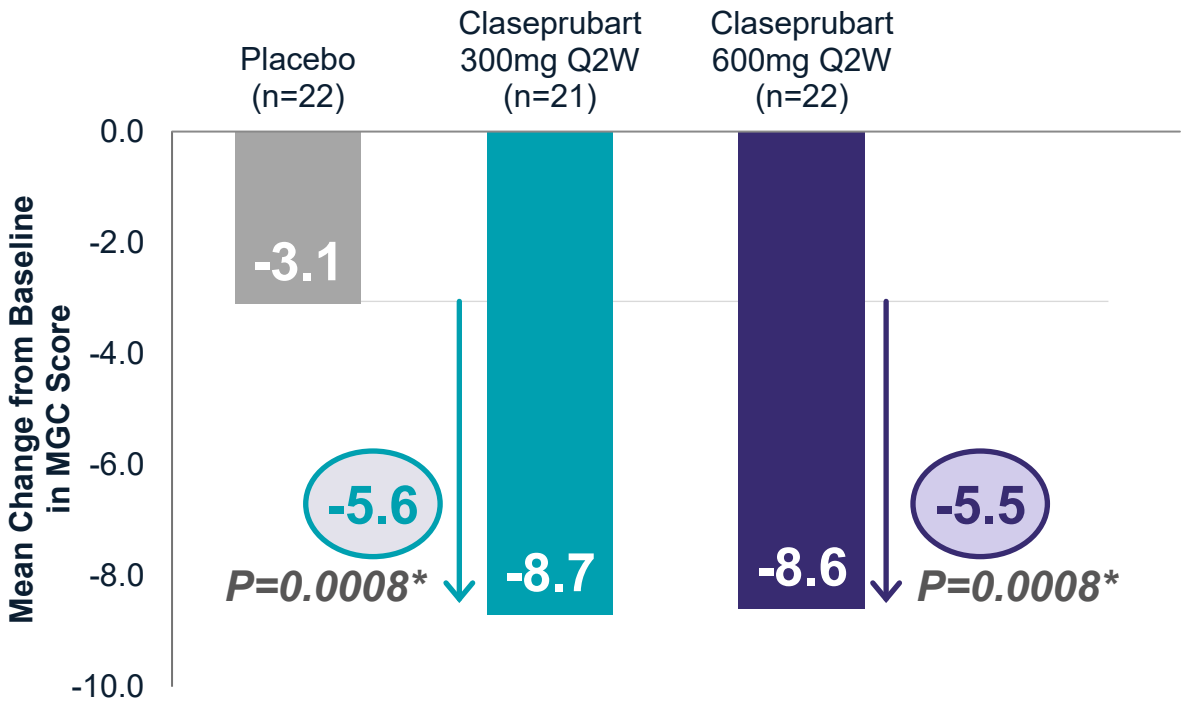

| P-value | 300mg/2mL | 600mg/4mL |
|---|---|---|
| One-sided | P=0.0144* | P=0.0111* |
| Two-sided | P=0.0288** | P=0.0222** |

**Statistically significant and clinically meaningful reductions in QMG across both treatment arms**

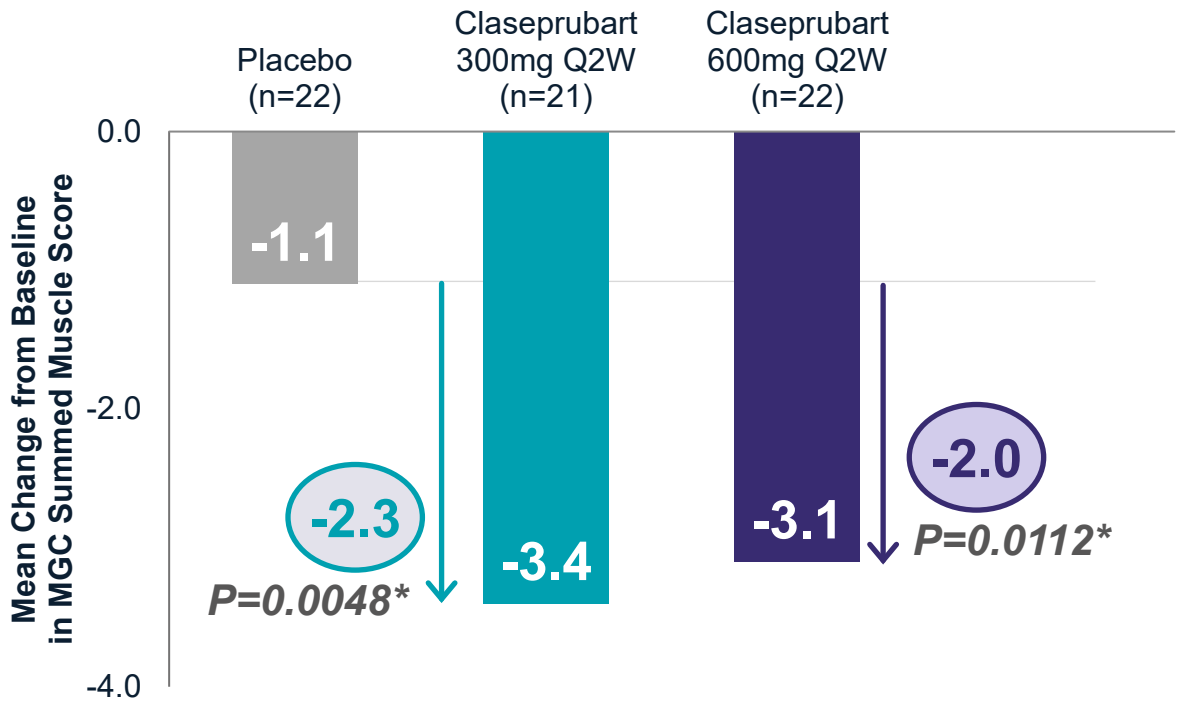
The change from baseline in QMG was analyzed using a mixed effect model for repeated measures (MMRM) with treatment group, visit, treatment by visit interaction, stratification factors, and baseline measure included.
*One-sided p-values are presented for comparisons of claseprubart vs placebo, with any p-value below 0.1 considered nominally statistically significant.
**Two-sided p-values are presented for comparisons of claseprubart vs placebo, with any p-value below 0.05 considered nominally statistically significant.

# Statistically significant improvement in MGC for both claseprubart arms vs. placebo at Week 13

## Mean Change in MGC Score from Baseline at Week 13




## Mean Change in MGC Summed Muscle Score from Baseline at Week 13 (post-hoc analysis)




The change from baseline in MGC and MGC Summed Muscle Score were separately analyzed using a mixed effect model for repeated measures (MMRM) with treatment group, visit, treatment by visit interaction, stratification factors, and baseline measure included.

*One-sided p-values are presented for comparisons of claseprubart vs placebo, with any p-value below 0.1 considered nominally statistically significant.

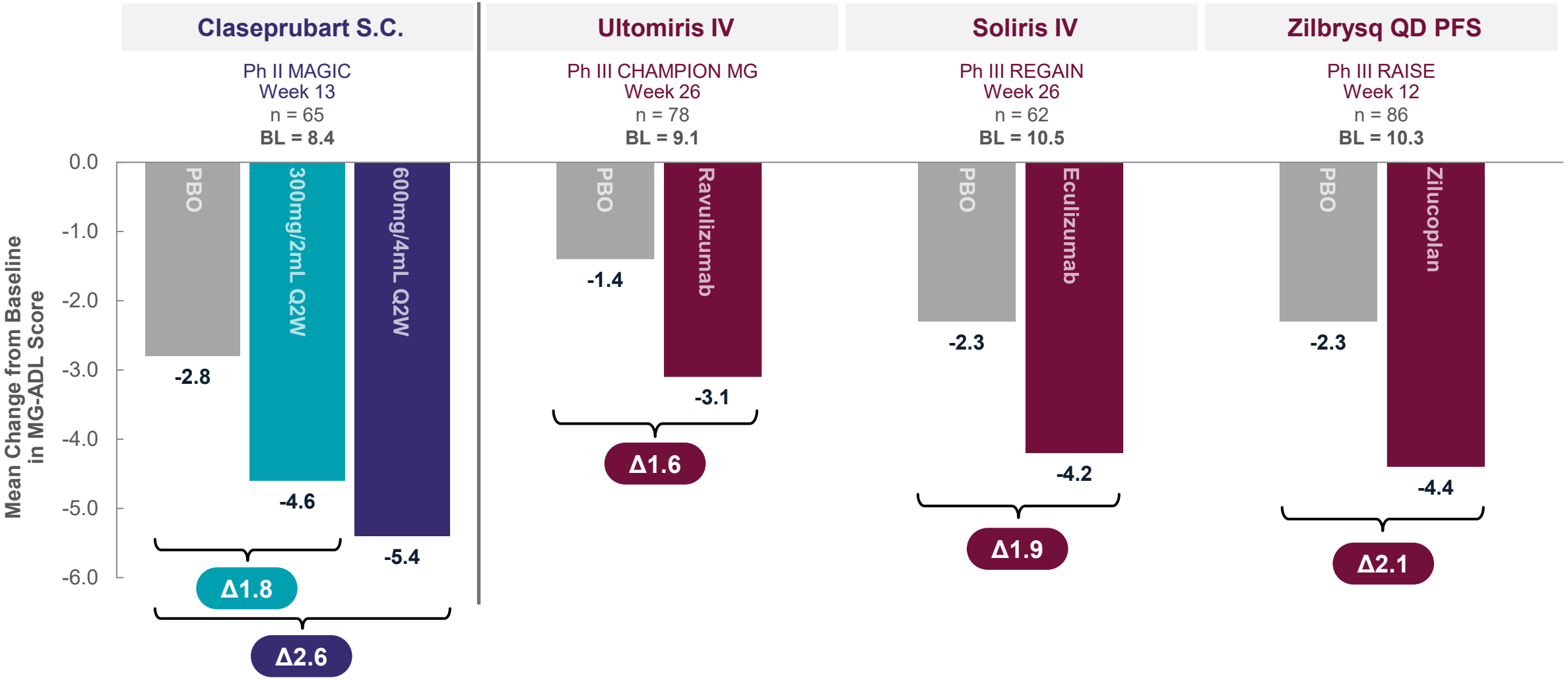
# Statistically significant improvement in MG-QoL-15r score for 300mg/2mL vs. placebo at Week 13

## Mean Change in MG-QoL-15r Score from Baseline at Week 13




The change from baseline in MG-QoL 15r was analyzed using a general linear model with treatment group, stratification factors, and baseline measure included.
*One-sided p-values are presented for comparisons of claseprubart vs placebo, with any p-value below 0.1 considered nominally statistically significant.

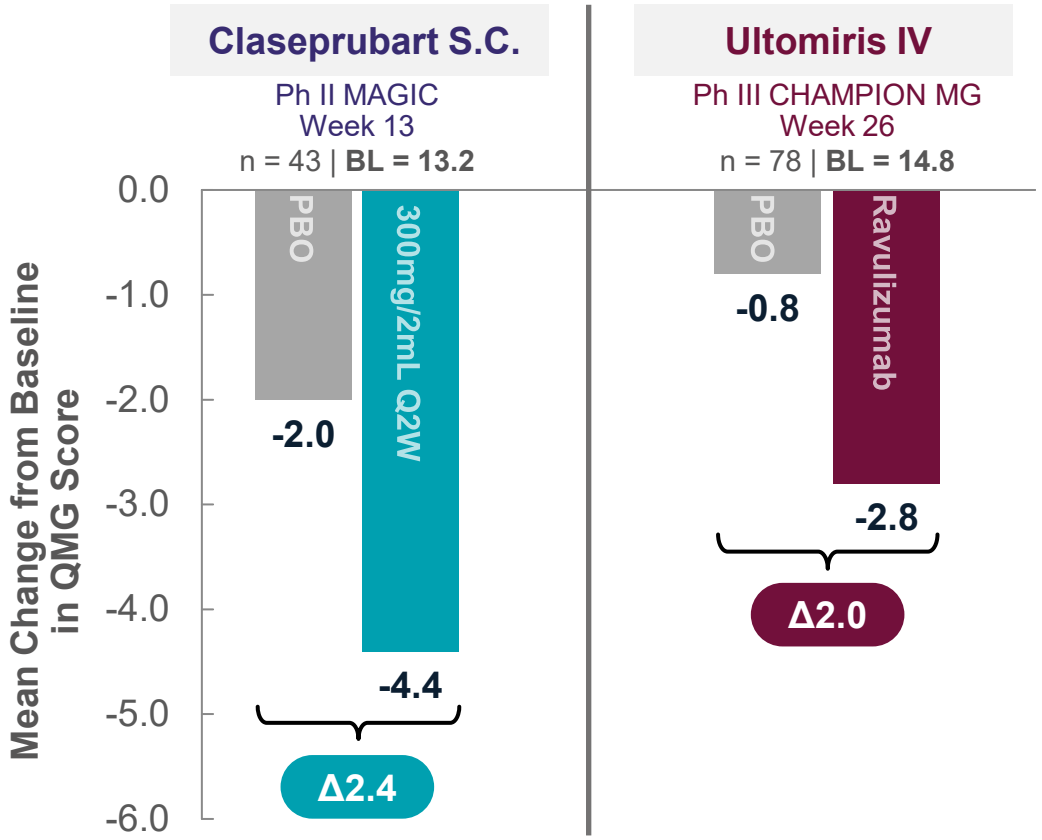
# Claseprubart demonstrated statistically significant and clinically meaningful improvements in MG-ADL



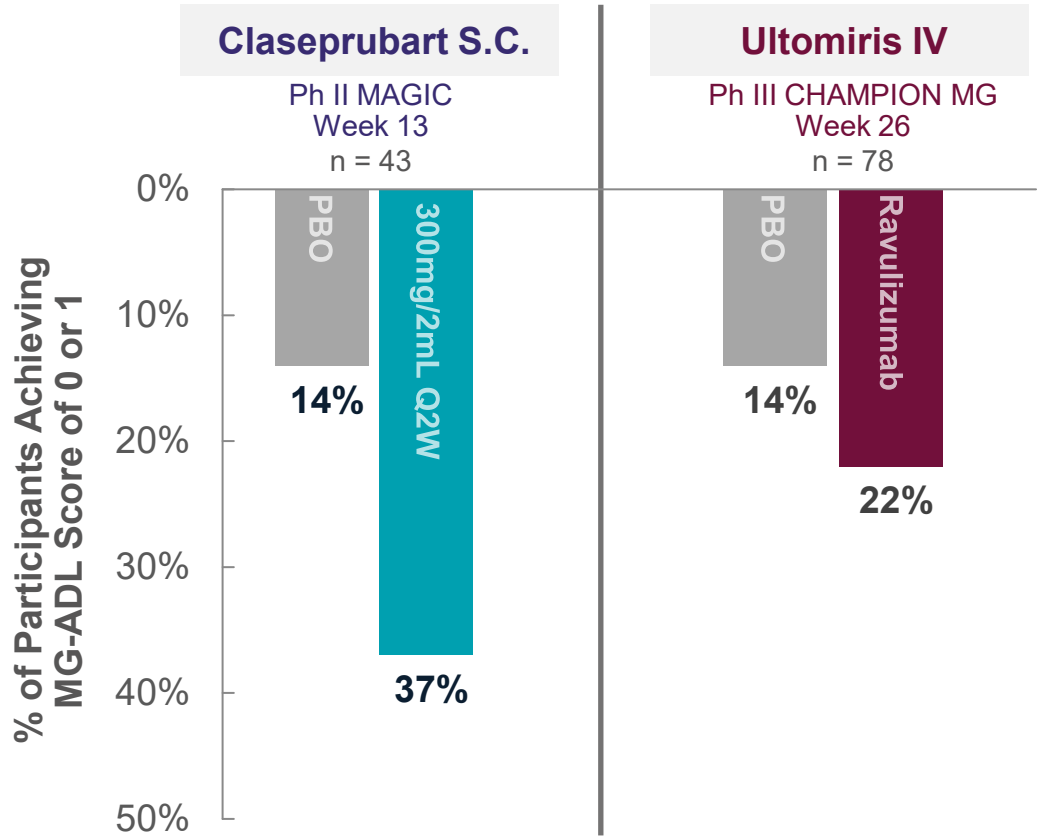

Note: For illustrative purposes only. Efficacy data are derived from different clinical trials conducted at different times, with differences in trial design and patient populations. As a result, cross-trial comparisons cannot be made, and no head-to-head clinical trials have been conducted. Statistical treatment of missing data may vary across studies shown. Source: Soliris (Ph3 REGAIN; 1200mg Q2W regimen; worst-rank ANCOVA). Ultomiris (Ph3 CHAMPION-MG; weight-based Q8W regimen with maintenance doses 3000-3600mg; MMRM ANCOVA with no imputation of missing data). Zilbrysq (Ph3 RAISE; 0.3mg/kg QD regimen, MMRM ANCOVA with no data censorship).

# Additional secondary efficacy measures support claseprubart potential as best-in-class complement inhibitor

## QMG Score




## Minimal Symptom Expression (MSE)




Note: For illustrative purposes only. Efficacy data are derived from different clinical trials conducted at different times, with differences in trial design and patient populations. As a result, cross-trial comparisons cannot be made, and no head-to-head clinical trials have been conducted. Statistical treatment of missing data may vary across studies shown. Source: Ultomiris (Ph3 CHAMPION-MG; weight-based Q8W regimen with maintenance doses 3000-3600mg; MMRM ANCOVA with no imputation of missing data).

# Unlike C5 inhibitors, Enjaymo® has no FDA Boxed Warning and REMS, or prophylactic antibiotic requirement prior to vaccination

ULTOMIRIS® (ravulizumab-cwvz) injection for intravenous use 300 mg/3 mL vial

C5 inhibitor

## ✗ Boxed Warning and REMS

**ULTOMIRIS® (ravulizumab-cwvz) injection, for intravenous or subcutaneous use**
**Initial U.S. Approval: 2018**

**WARNING: SERIOUS MENINGOCOCCAL INFECTIONS**
***See full prescribing information for complete boxed warning.***

**ULTOMIRIS increases the risk of serious and life-threatening infections caused by *Neisseria meningitidis*.**

- **Complete or update meningococcal vaccination at least 2 weeks prior to the first dose of ULTOMIRIS, unless the risks of delaying ULTOMIRIS outweigh the risks of developing a serious infection. Comply with the most current Advisory Committee on Immunization Practices (ACIP) recommendations for meningococcal vaccination in patients receiving a complement inhibitor. (5.1)**
- **Patients receiving ULTOMIRIS are at increased risk for invasive disease caused by *N. meningitidis*, even if they develop antibodies following vaccination. Monitor patients for early signs and symptoms of meningococcal infections and evaluate immediately if infection is suspected. (5.1)**

**ULTOMIRIS is available only through a restricted program called ULTOMIRIS and SOLIRIS REMS. (5.2)**

## ✗ Antibiotic Prophylaxis Required if not Fully Vaccinated

**2.2 Recommended Vaccination and Prophylaxis for Meningococcal Infection**

Vaccinate patients against meningococcal infection (serogroups A, C, W, Y and B) according to current ACIP recommendations at least 2 weeks prior to initiation of ULTOMIRIS *[see Warnings and Precautions (5.1)]*.

If urgent ULTOMIRIS therapy is indicated in a patient who is not up to date with meningococcal vaccines according to ACIP recommendations, provide the patient with antibacterial drug prophylaxis and administer these vaccines as soon as possible.

Healthcare providers who prescribe ULTOMIRIS must enroll in the ULTOMIRIS and SOLIRIS REMS *[see Warnings and Precautions (5.2)]*.

Enjaymo sutimlimab-jome Injection for intravenous use 1100 mg/22 mL

C1s inhibitor

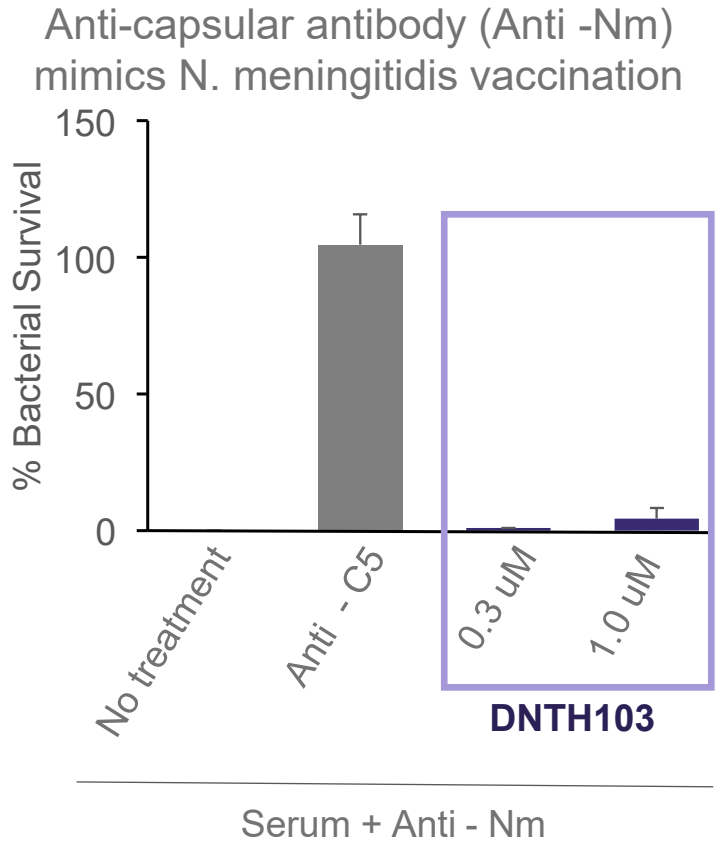
## ✓ No Boxed Warning or REMS

## ✓ No Antibiotic Prophylaxis Required

Source: Ultomiris prescribing information & Enjaymo prescribing information

# Claseprubart *in vitro* study indicates lower risk of *Neisseria meningitidis* infections

- Protection against infection is a critical function of the complement pathway
- **DNTH103 selectively inhibits** the **Classical Pathway**, leaving the alternative and lectin-activated defense pathways intact
- An *in vitro* assay measured **antibody-dependent complement-mediated killing of *N. meningitidis*** in the presence of **DNTH103** and **anti-C5 (ravulizumab*)**
- In this assay, **DNTH103 maintained bacterial killing**, potentially leading to a decreased risk of infection vs. C5 inhibitors



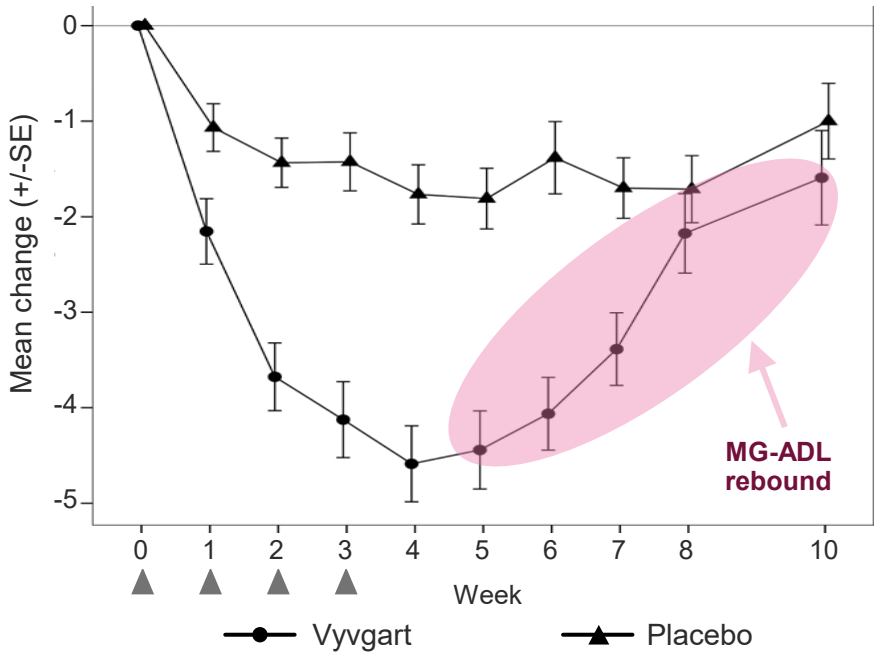

**Results further validate the potential differentiated safety profile for DNTH103 as a selective Classical Pathway inhibitor consistent with Enjaymo, an approved C1s inhibitor without an FDA Boxed Warning or REMS**

*Engineered using patent sequence.

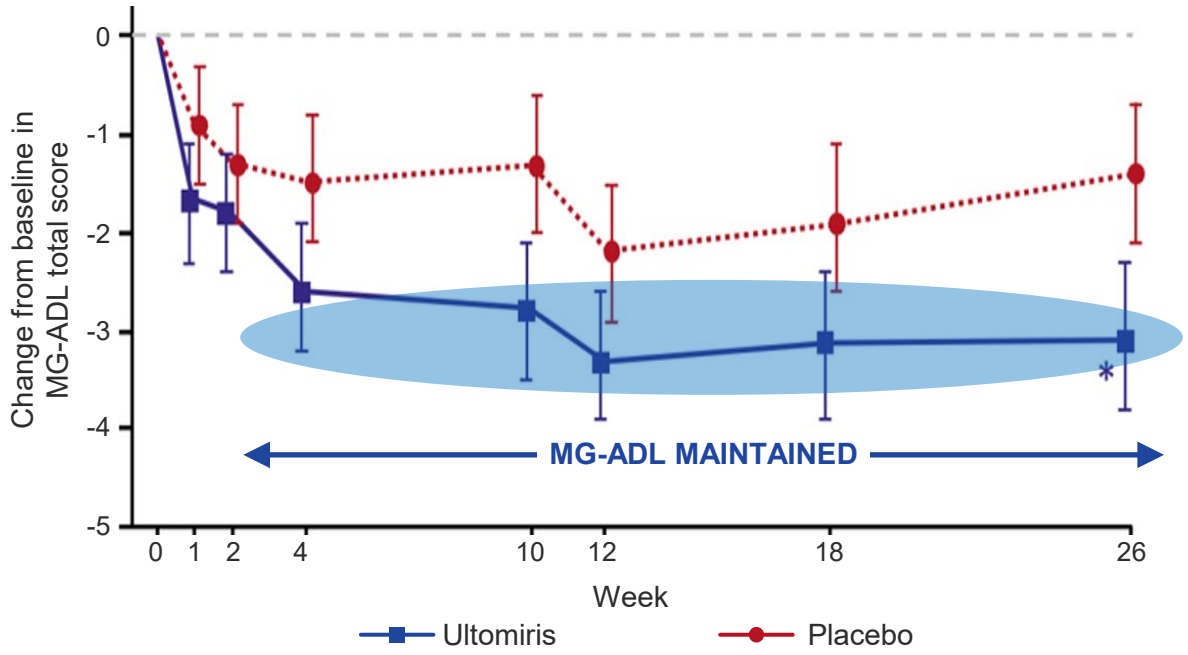
# Claseprubart aims to provide consistent symptom control with convenient S.C. Q2W dosing

- Chronic diseases like MG benefit from **consistent treatment and symptom control**
- When patients are required to take a drug holiday on FcRns, **MG-ADL scores immediately begin to rebound** as patients self-report symptoms and disease worsening
- Real-world evidence[1] suggests **sustained inhibition / treatment with complement is better over time**

## Vyvgart®: Cyclic Dosing Leads to MG-ADL Rebound




## Ultomiris®: Complement Inhibition Provides Consistent MG-ADL Symptom Control




▲ Indicates Vyvgart® dose administered

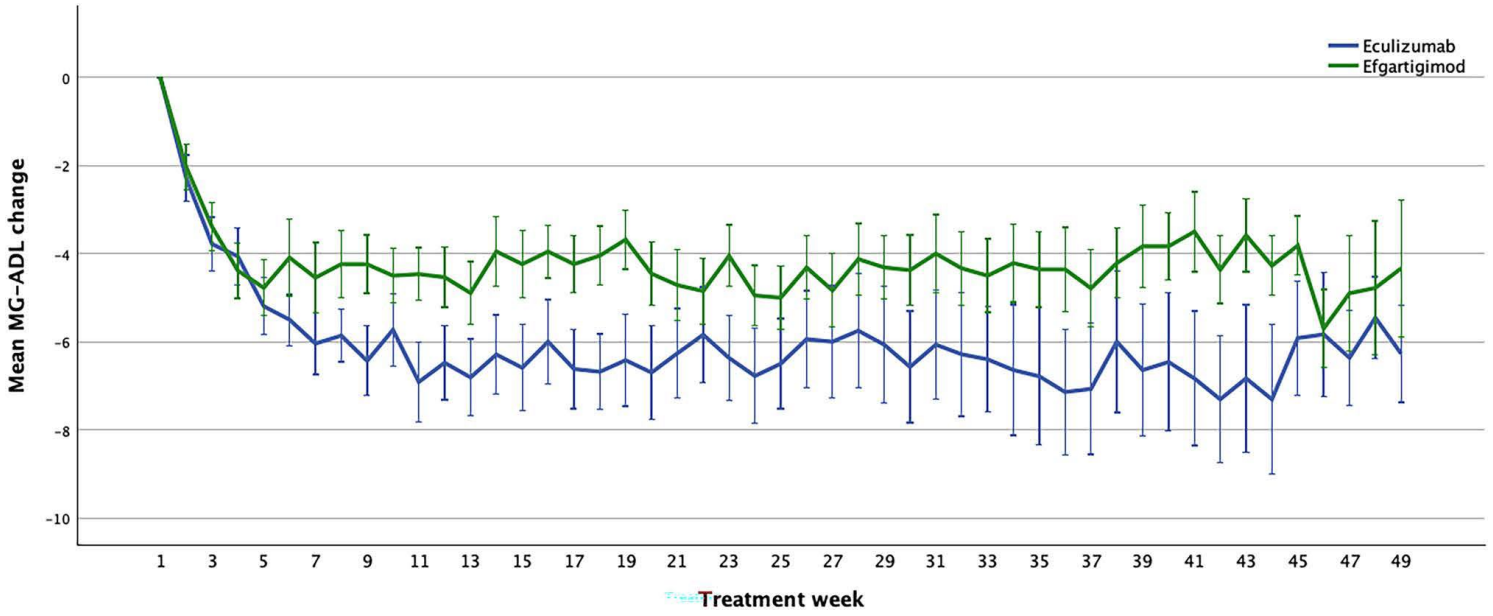
Source: Vyvgart® prescribing information and Ultomiris® prescribing information

1. Pane et al. A real-life experience with eculizumab and efgartigimod in generalized myasthenia gravis patients. *J Neurol* 271, 6209–6219 (2024)

# Real-world evidence demonstrates clinical advantages for complement vs. FcRns

## Real-world Evidence Indicates Sustained MG-ADL Reductions for Complement Inhibitors vs. FcRns, Among Other Clinical Benefits

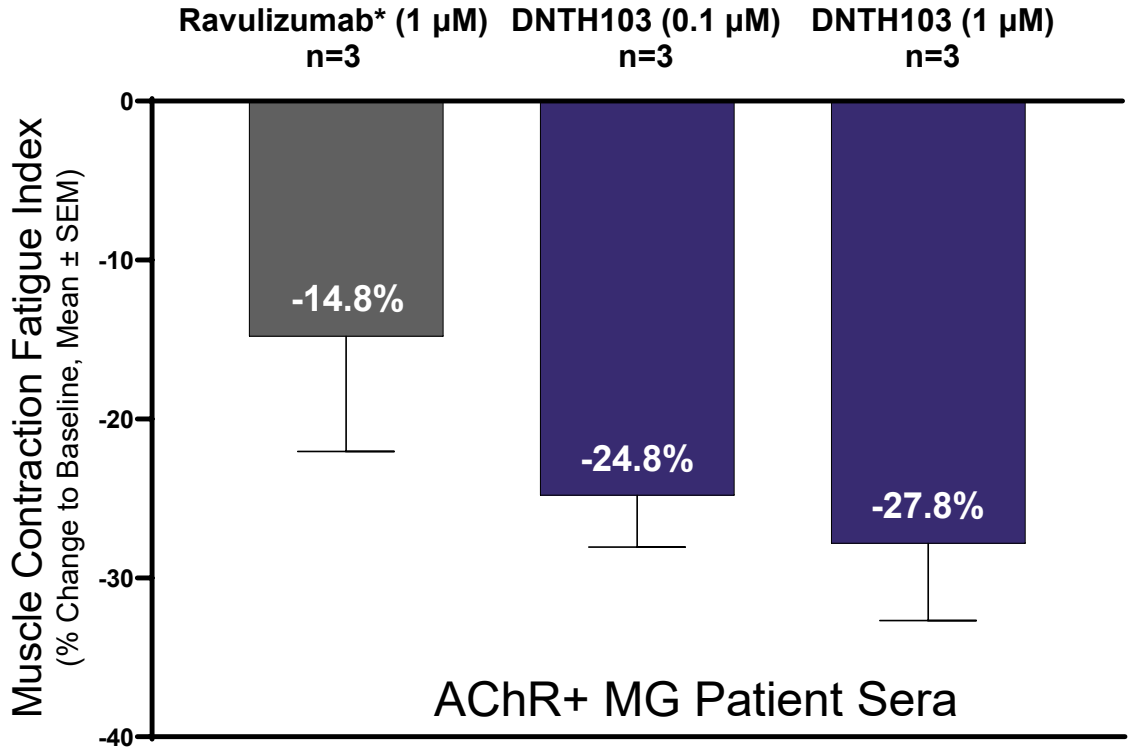
- **Patients on eculizumab showed a greater reduction in MG-ADL** over time than those on efgartigimod
- **Eculizumab showed statistically significant, deeper QMG reduction** and higher responder rate than efgartigimod
- **Eculizumab patients had a statistically significant greater reduction in rate of clinical events** (deteriorations, crisis or hospitalization) than efgartigimod
- **Patients on efgartigimod were more likely to suspend treatment** (p = 0.015 for AChR+ patients) with the “main reason for discontinuation was MG deterioration”
- **Eculizumab patients also had a statistically significant greater reduction in steroid dose** coming from a higher baseline dose vs. efgartigimod patients, but reaching a numerically lower dose during the treatment period




Source: Pane et al., J Neurol 271, 6209–6219 (2024)

# Claseprubart improves neurotransmission and muscle contraction in an AChR+ MG model

- **Serum from MG patients** used in a validated in vitro MG model[1,2,3]
- **Assessed improvement in neurotransmission and muscle contraction** of ravulizumab* and DNTH103, as measured by decrease in muscle contraction fatigue
- **Results confirm DNTH103 improved neurotransmission and muscle contraction**

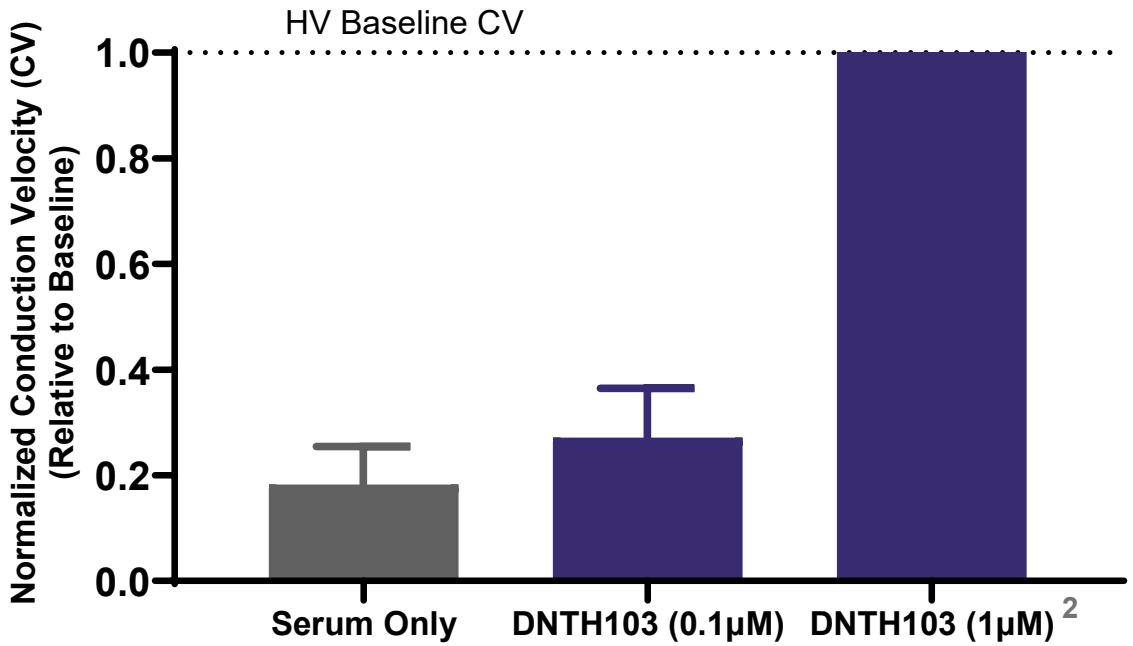



**Results provide further scientific rationale for DNTH103 in gMG**

1. Smith et al., Front Cell Dev Biol. 2021;9:745897; 2. Vila et al., Expert Opin Drug Discov. 2020;15(3):307-317; 3. Vila et al., Theranostics. 2019;9(5):1232-1246.
* Engineered using patent sequence

# Claseprubart restores neuronal conduction velocity in an *in vitro* CIDP model

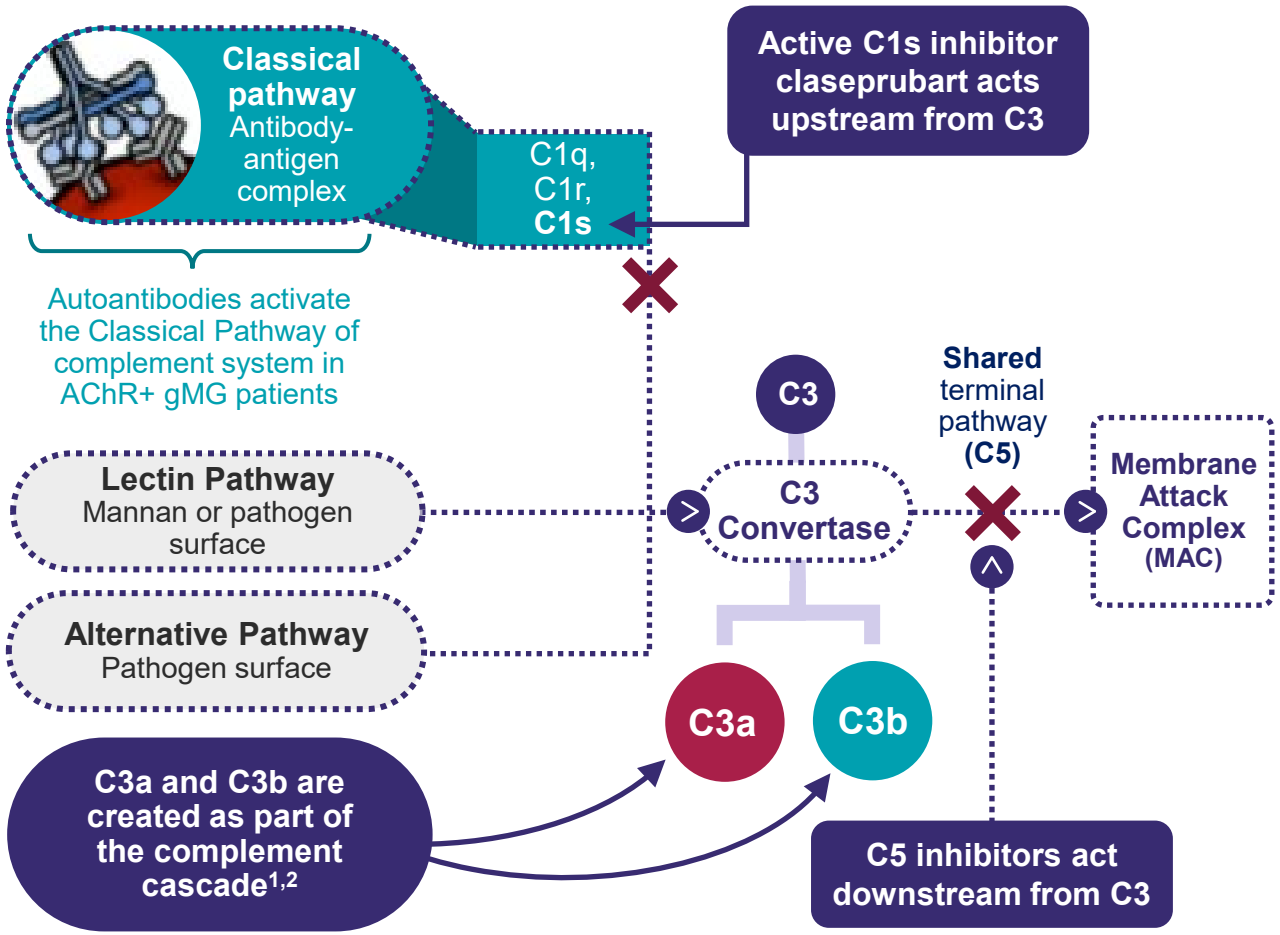
- **Serum from 3 CIDP patients** was evaluated in a commercially available *in vitro* CIDP model[1]
- **Assessed improvement in neuronal conduction velocity** of two doses of DNTH103 as compared to baseline conduction velocity determined in sera from healthy volunteers (N=3)
- **Results confirm DNTH103 completely restored conduction velocity** across the axons of human motor neurons in the presence of autoantibodies from CIDP patient sera




**Results provide further scientific rationale for DNTH103 in CIDP**

1 Rumsey et al., *Adv. Therap.*, 2022, 5(6): 2200030
2 Results for DNTH103 (1uM) include data from multiple conduction velocity recordings that exceed 1.0. For the purposes of this illustration, results are shown up to the baseline value.

# Pro-inflammatory split products C3a and C3b have potential negative effects on NMJ pathology beyond MAC in gMG




## C3a and C3b Are Elevated in AChR+ gMG – Both Cause Inflammatory Damage[1,2]

| C3a | C3b |
|---|---|
| • Anaphylatoxin[2,3]<br>• Initiates mast cell degranulation[4]<br>• Promotes neutrophil-mediated acute phase reactions[5] driven by pro-inflammatory cytokines[6] – particularly IL-6 in gMG[7]<br>• Has the potential to directly act on NMJ via smooth muscle contraction and endothelial permeability[8] | • Opsonin[3]<br>• Forms part of an amplification loop that perpetuates a cycle of complement activation[3]<br>• Facilitates integrin-mediated phagocytosis[3] |

Ab, antibody; gMG, generalized myasthenia gravis; IL, interleukin; MAC, membrane attack complex; NMJ, neuromuscular junction.

1. Stascheit F, et al. *Eur J Neurol* 2023;30:1409–16; 2. Iacomino N, et al. *Biomedicines* 2022;10; 3. Watanabe-Kusunoki K, Anders HJ. *J Autoimmun* 2024;145:103216; 4. Nilsson G, et al. *J Immunol* 1996;157:1693–8; 5. Riaz B, Sohn S. Cells 2023;12; 6. Wang Y, et al. J Neurol 2025;272:489; 7. Uzawa A, et al. J Neuroimmunol 2021;358:577634; 8. Drouin SM, et al. *J Immunol* 2001;166:2025–32; 2015;2:e119.

# Broad opportunity for DNTH212 across multiple diseases where Type 1 interferon and B Cells are implicated

## Indications with biological rationale and supportive clinical data

| | Biological Rationale | Clinical Evidence |
|---|---|---|
| **Primary Sjögren's Disease** *~350,000 U.S. Patients* | ✓ | • *B Cell:* ianalumab positive Ph. 3; telitacicept positive Ph. 3 |
| **Systemic Lupus Erythematosus** *~225,000 U.S. Patients* | ✓ | • *Type 1 interferon:* anifrolumab approved; litifilimab positive Ph. 2<br>• *B Cell:* belimumab approved; telitacicept approved (CN); ianalumab positive Ph. 2 |
| **Dermatomyositis** *~50,000 U.S. Patients* | ✓ | • *Type 1 interferon:* dazukibart positive Ph. 2 |
| **Cutaneous Lupus Erythematosus** *~300,000 U.S. Patients* | ✓ | • *Type 1 interferon:* litifilimab positive Ph. 2 |
| **Lupus Nephritis** *~120,000 U.S. Patients* | ✓ | • *B Cell:* belimumab approved |

## Indications with biological rationale

| | Biological Rationale |
|---|---|
| **Hidradenitis Suppurativa** *~330,000 U.S. Patients* | ✓ |
| **Scleroderma** *~75,000 U.S. Patients* | ✓ |
| **Pemphigus Vulgaris** *~32,000 U.S. Patients* | ✓ |

**Dianthus identified first three priority indications of Sjögren's Disease, Systemic Lupus Erythematosus, and Dermatomyositis to build a rheumatology franchise**

Type 1 interferon targeting Ph. 3 studies currently ongoing in: SLE, CLE, LN, Scleroderma, DM. B Cell (BAFF/APRIL) targeting Ph. 3 studies currently ongoing in: LN, SLE.
Estimated U.S. patients per Dianthus meta-analysis and estimates.

# Accomplished team of biotech industry veterans and scientists committed to bringing innovation to market

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*Select Autoimmune Drugs Developed by Dianthus Team*

CellCept, Soliris (eculizumab), Ultomiris (ravulizumab-cwvz), Lupkynis (voclosporin), Zilbrysq (zilucoplan) injection

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