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Efficacy and Safety of Nipocalimab in Participants With Idiopathic Inflammatory Myopathies: Results From the Phase 2 SPIREA Trial Through 52 Weeks

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Conflicts of Interest

RA received honoraria, consulting fees, speakers bureau payments, advisory/board fees, expert witness fees, or royalties from Alexion, ANI Pharmaceuticals, argenx, Artiva Biotherapeutics, AstraZeneca, Boehringer Ingelheim, Bristol Myers Squibb, Cabaletta Bio, Capstan Therapeutics, Century Therapeutics, CRISPR Therapeutics, CSL Behring, EMD Serono, Fate Therapeutics, Galapagos, GSK, Horizon Therapeutics, Immunovant, Jade Biosciences, Janux Therapeutics, Johnson & Johnson, Kiniksa Pharmaceuticals, Lilly, Meiji Seika Pharma, Nkarta, Novartis, Octapharma, OneFour Bio, Orna Therapeutics, Ouro Medicines, Pfizer, Priovant Therapeutics, and Sobi.

HC has nothing to disclose.

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HK has nothing to disclose.

MMD received consultancy fees, payment or honoraria for lectures, presentations, manuscript writing, or educational events, or participated on a Data Safety Monitoring Board or Advisory Board for the American Academy of Neurology, American Association of Neuromuscular and Electrodiagnostic Medicine, Abcuro, Amicus, Annexon Biosciences, argenx, Astellas Pharma, Alexion/AstraZeneca, Cabaletta Bio, Candid Therapeutics, Catalyst Pharmaceuticals, Covance/Labcorp/Fortrea, Creyon Bio, CSL Behring, Dianthus Therapeutics, Dyne Therapeutics, EMD Serono/Merck, Horizon/Immunovant, Immunoglobulin Society, Johnson & Johnson, Kriya Therapeutics, MedLink, Nuvig Therapeutics, Octapharma, Priovant Therapeutics, Ra Pharma/UCB Biopharma, Sanofi/Genzyme, Shire/Takeda, TREAT-NMD Advisory Committee for Therapeutics, UCB, and Vertex Pharmaceuticals; received research grants or support from Abcuro, Alexion/AstraZeneca, Alnylam Pharmaceuticals, Amicus, argenx, Biosensics, Bristol Myers Squibb, Cabaletta Bio, Catalyst Pharmaceuticals, CSL Behring, EMD Serono/Merck, FDA Office of Orphan Products Development (OOPD), GSK, Genentech, Grifols, Horizon/Immunovant, Mitsubishi Tanabe Pharma, Muscular Dystrophy Association, National Institutes of Health, Nkarta, Novartis, Octapharma, Orphazyme, Priovant Therapeutics, Ra Pharma/UCB Biopharma, Sanofi/Genzyme, Sarepta Therapeutics, Shire/Takeda, Spark Therapeutics, and The Myositis Association; received royalties for manuscript writing from Wolters Kluwer Health and from Elsevier and for discovery relating to health care from Zevra Therapeutics; and had roles with the Clinical Neuromuscular Pathology Specialty Exam, the Committee of United Council for Neurologic Subspecialties, the International Myositis Assessment & Clinical Studies Group, Kansas City Neurology Neurosurgery Society Neuromuscular Study Group, Myositis Clinical Trial Consortium, and Myositis International Health and Research Collaborative Alliance.

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Idiopathic Inflammatory Myopathies

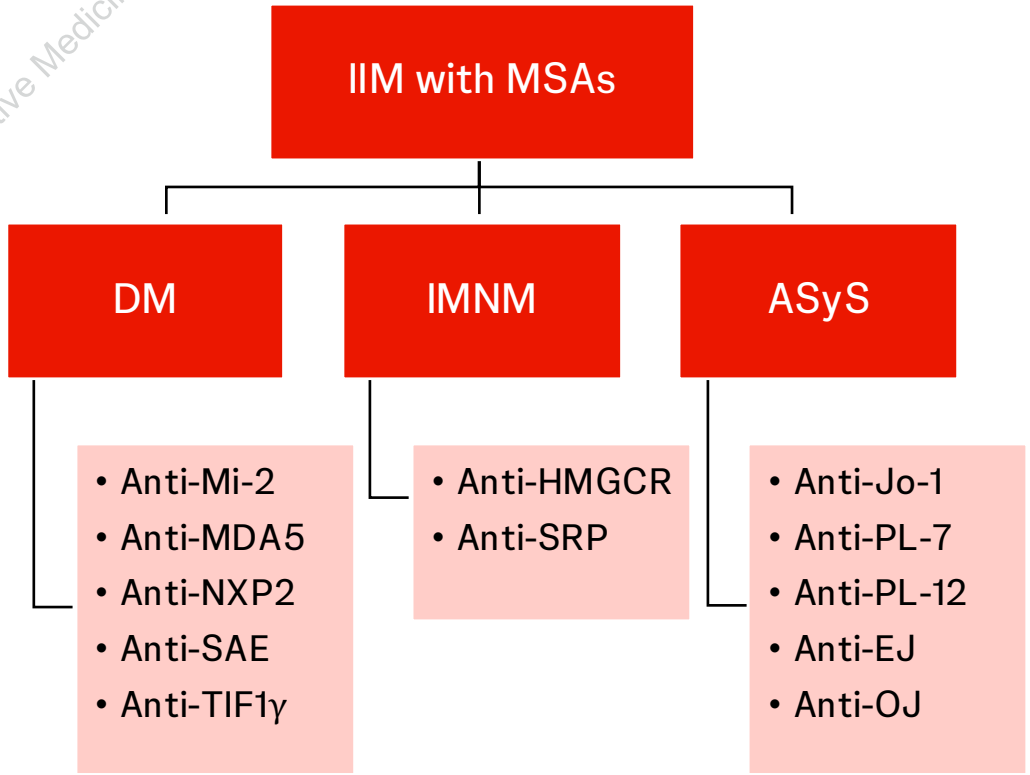
IIMs

- A group of rare, complex, heterogeneous autoimmune disorders that cause chronic muscle inflammation and affect multiple organ systems[1]
  - Major subtypes include DM, IMNM, and ASyS[2]
- Often characterized by the presence of pathogenic MSAs associated with unique clinical phenotypes[3]
- Patients with IIM have ~4 times higher mortality than the general population[4]

Treatment and unmet needs

- Treatment burden (side effects, productivity loss) adds to the impact[5,6]
- Existing therapies are suboptimal, with few approved options, and refractory disease[7]
- With corticosteroids and immunosuppressive medications, relapses were reported in 65% of adult patients with IIM[8]

IIM subtypes and associated MSAs[9,10]



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ASyS=antisynthetase syndrome, **DM**=dermatomyositis, **IIM**=Idiopathic inflammatory myopathy, **IMNM**=immune-mediated necrotizing myopathy, **MSA**=myositis-specific autoantibody.

1. Lundberg IE, et al. *Nat Rev Dis Primers*. 2021;2;7(1):87. 2. Oldroyd A, et al. *Clin Med (London)*. 2017;17(4):322-328. 3. Allameen NA, et al. *Nat Rev Rheumatol*. 2025;21(1):46-62. 4. Dobloug GC, et al. *Ann Rheum Dis*. 2018;77(1):40-47. 5. Xu A, et al. *Int J Rheum Dis*. 2021;4(6):809-814. 6. Loarce-Martos J, et al. *Clin Rheumatol*. 2021; 40(12):4977-4982. 7. Kocyigit M, et al. *J Neuromuscul Dis*. 2026;13(4):553-572. 8. Agarwal SK, et al. *Clin Rheumatol*. 2006; 25(4):476-481. 9. Raaphorst J, et al. *Lancet Neurol*. 2025;24(9):776-788. 10. Musai J, et al. *Curr Rheumatol Rep*. 2024;26(12):421-430.

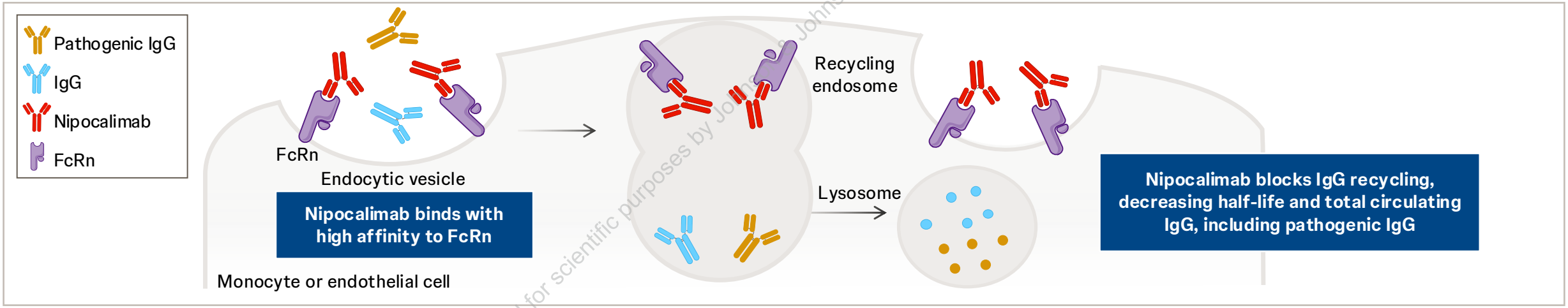
Nipocalimab: FcRn Blocker Decreases Serum IgG, Including Pathogenic IgG

FcRn:

- A recycling or transcytosis receptor that maintains IgG levels in circulation[1-3]
- Expressed in multiple cell types, including monocytes and endothelial cells[1-3]

Nipocalimab:

- A high-affinity, fully human, IgG1 FcRn blocker that selectively blocks IgG recycling[4]
- Decreases serum IgG, including pathogenic IgG, while preserving immune cell functions[4-7]
- Approved for gMG in the United States, Brazil, Japan, Europe, China, and Panama[8]
- Approved for wAIHA in the United States, Brazil, and Panama[9]



Objective: The SPIREA study aimed to evaluate the efficacy and safety of nipocalimab 15 mg/kg IV q2w versus placebo, in addition to standard-of-care immunosuppression, in adults with active IIM

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FcRn=neonatal crystallizable fragment receptor, **gMG**=generalized myasthenia gravis, **IgG**=immunoglobulin G, **IIM**=idiopathic inflammatory myopathy, **IV**=intravenous, **q2w**=every 2 weeks, **wAIHA**=warm autoimmune hemolytic anemia.

1. Blumberg LJ, et al. *Sci Adv*. 2019;5(12):eaax9586. 2. Roopenian DC, Akilesh S. *Nat Rev Immunol*. 2007;7(9):715-725. 3. Peter HH, et al. *J Allergy Clin Immunol*. 2020;146(3):479-491.e5. 4. Seth NP, et al. *MAbs*. 2025;17(1):2461191. 5. Ling LE, et al. *Clin Pharmacol Ther*. 2019;105(4):1031-1039. 6. Antozzi C, et al. *Lancet Neurol*. 2025;24(2):105-116. 7. Noaiseh G, et al. *Lancet*. 2025;406(10518):2435-2448. 8. IMAAVY™ (nipocalimab-aahu) injection, for intravenous use. Prescribing information. Janssen Biotech, Inc.; 2025. 9. FDA approves first drug for warm autoimmune hemolytic anemia. US Food and Drug Administration. Accessed September 25, 2026. https://www.fda.gov/drugs/news-events-human-drugs/fda-approves-first-drug-warm-autoimmune-hemolytic-anemia

SPIREA Study Design

A phase 2, multicenter, randomized (2:1), double-blind, placebo-controlled, parallel-group, proof-of-concept study (NCT05379634)

Key inclusion criteria

- Participants ≥18 years of age with active, moderate-to-severe, MSA-positive, probable or definite IIM meeting 2017 EULAR/ACR classification criteria (DM, IMNM, and ASyS subtypes) were included

Primary endpoint[a]

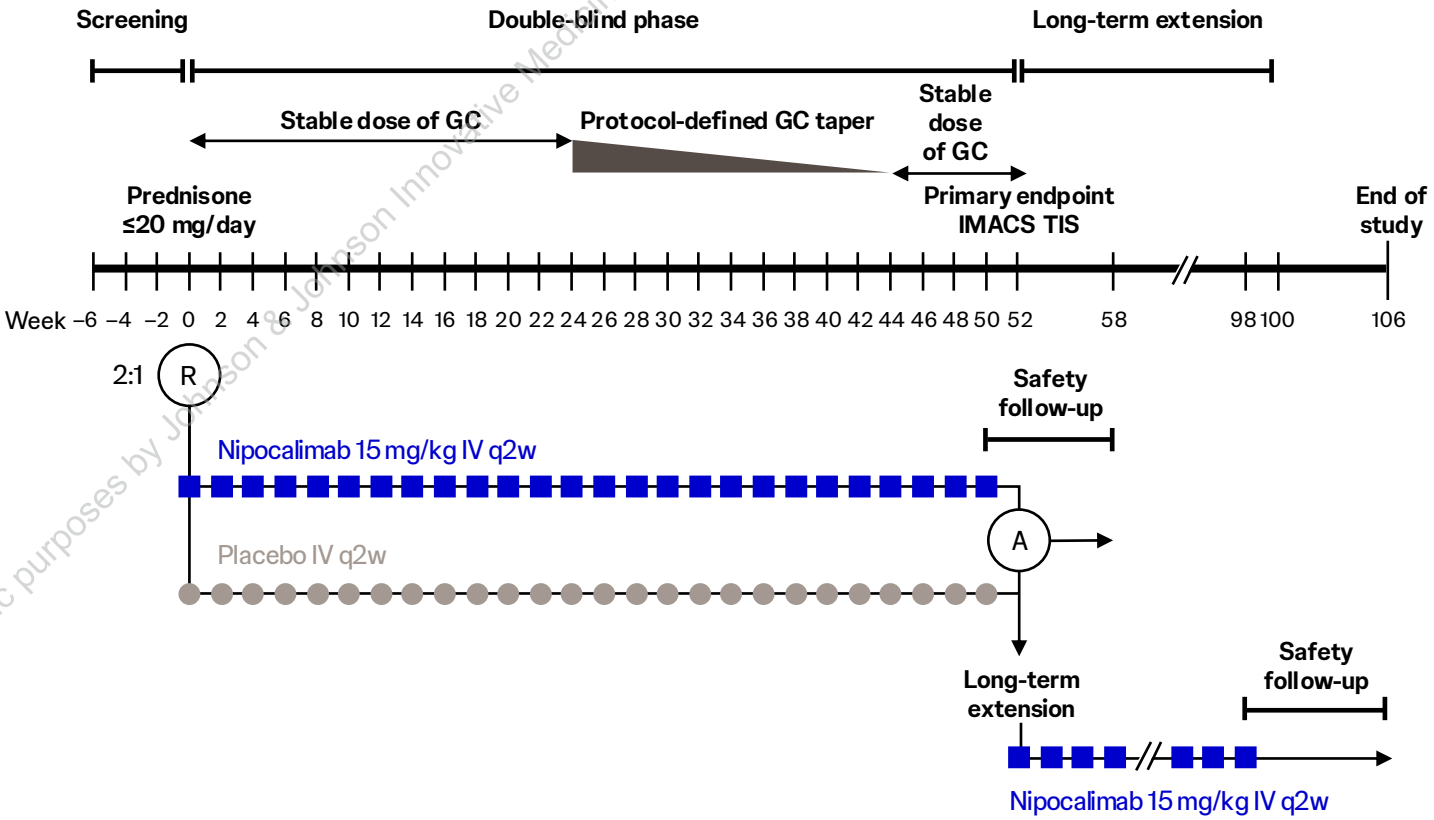
- Proportion achieving at least minimal improvement in IMACS TIS at Week 52 and on ≤5 mg/day prednisone (or equivalent) from Weeks 44 to 52

Key secondary endpoints

- At least minimal improvement in IMACS TIS at Week 24
- IMACS TIS at Week 52
- At least moderate improvement in IMACS TIS at Week 24
- Change from baseline in PROMIS-PF-20 total scores and MMT-8 at Week 52
- Achieving GC reduction to 5 mg/day of oral prednisone at Week 44 and maintaining that reduction through Week 52
- At least minimal improvement in IMACS TIS from Weeks 44 to 52 and on ≤5 mg/day prednisone (or equivalent) from Weeks 44 to 52

Other endpoints

- Change from baseline in muscle enzymes at Week 24 and Week 52



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[a]Primary endpoint tested at α=0.2.

A=assess eligibility for long-term extension, **ASyS**=antisynthetase syndrome, **DM**=dermatomyositis, **EULAR/ACR**=European Alliance of Associations for Rheumatology/American College of Rheumatology, **GC**=glucocorticoid, **IIM**=idiopathic inflammatory myopathy, **IMACS TIS**=International Myositis Assessment & Clinical Studies–Total Improvement Score, **IMNM**=immune-mediated necrotizing myopathy, **IV**=intravenous, **MMT-8**=Manual Muscle Testing of 8 muscle groups, **MSA**=myositis-specific autoantibody, **PROMIS-PF-20**=Patient-Reported Outcomes Measurement Information System–Physical Function 20-item short form, **q2w**=every 2 weeks, **R**=randomization.

Demographic and Baseline Characteristics

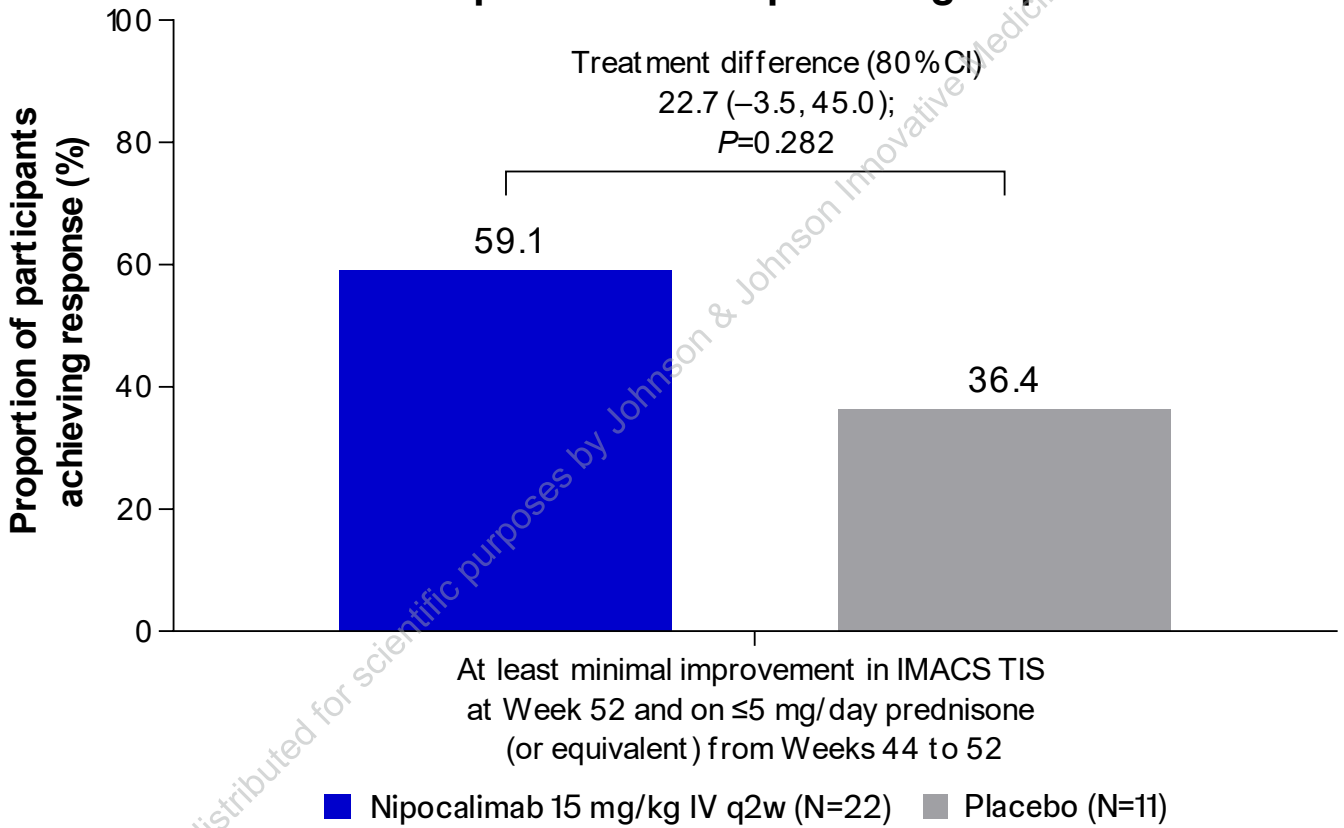
Baseline characteristics	Nipocalimab 15 mg/kg IV q2w (N=22)	Placebo (N=11)	Total (N=33)
Female, n (%)	15 (68.2)	5 (45.5)	20 (60.6)
Age (years), mean (SD)	52.7 (13.5)	52.2 (10.4)	52.5 (12.4)
Race, n (%)			
American Indian or Alaska Native	1 (4.5)	0	1 (3.0)
Asian	5 (22.7)	4 (36.4)	9 (27.3)
Black or African American	2 (9.1)	1 (9.1)	3 (9.1)
Not reported	3 (13.6)	0	3 (9.1)
White	11 (50.0)	6 (54.5)	17 (51.5)
Region, n (%)			
Asia Pacific	5 (22.7)	4 (36.4)	9 (27.3)
Europe	9 (40.9)	5 (45.5)	14 (42.4)
Latin America	1 (4.5)	2 (18.2)	3 (9.1)
North America	7 (31.8)	0	7 (21.2)
BMI (kg/m²), mean (SD)	27.97 (6.271)	25.42 (4.733)	27.12 (5.857)
IIM duration (years), mean (SD)	5.8 (6.3)	5.2 (8.2)	5.6 (6.9)
Age at onset (years), mean (SD)	47.3 (15.4)	47.5 (13.0)	47.3 (14.45)
Severe disease, n (%)	13 (59.1)	7 (63.6)	20 (60.6)
PROMIS-PF-20 T-Score (9.2-62.7), mean (SD)	33.7 (7.2)	36.2 (9.9)	34.6 (8.1)
MMT-8 (0-150), mean (SD)	121.8 (11.7)	121.3 (12.0)	121.6 (11.6)
Creatine kinase (U/L), mean (SD)	1679.4 (1514.9)	2010.2 (2440.5)	1789.7 (1841.9)
Taperable GCs (>5 mg/day prednisone), n (%)	12 (54.5)	8 (72.7)	20 (60.6)

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BMI=body mass index, **GC**=glucocorticoid, **IIM**=idiopathic inflammatory myopathy, **IV**=intravenous, **MMT-8**=Manual Muscle Testing of 8 muscle groups, **PROMIS-PF-20**=Patient-Reported Outcomes Measurement Information System–Physical Function 20-item short form, **q2w**=every 2 weeks, **SD**=standard deviation.

Primary Endpoint: At Least Minimal Improvement in IMACS TIS at Week 52 and on ≤5 mg/day Prednisone (or Equivalent) From Weeks 44 to 52

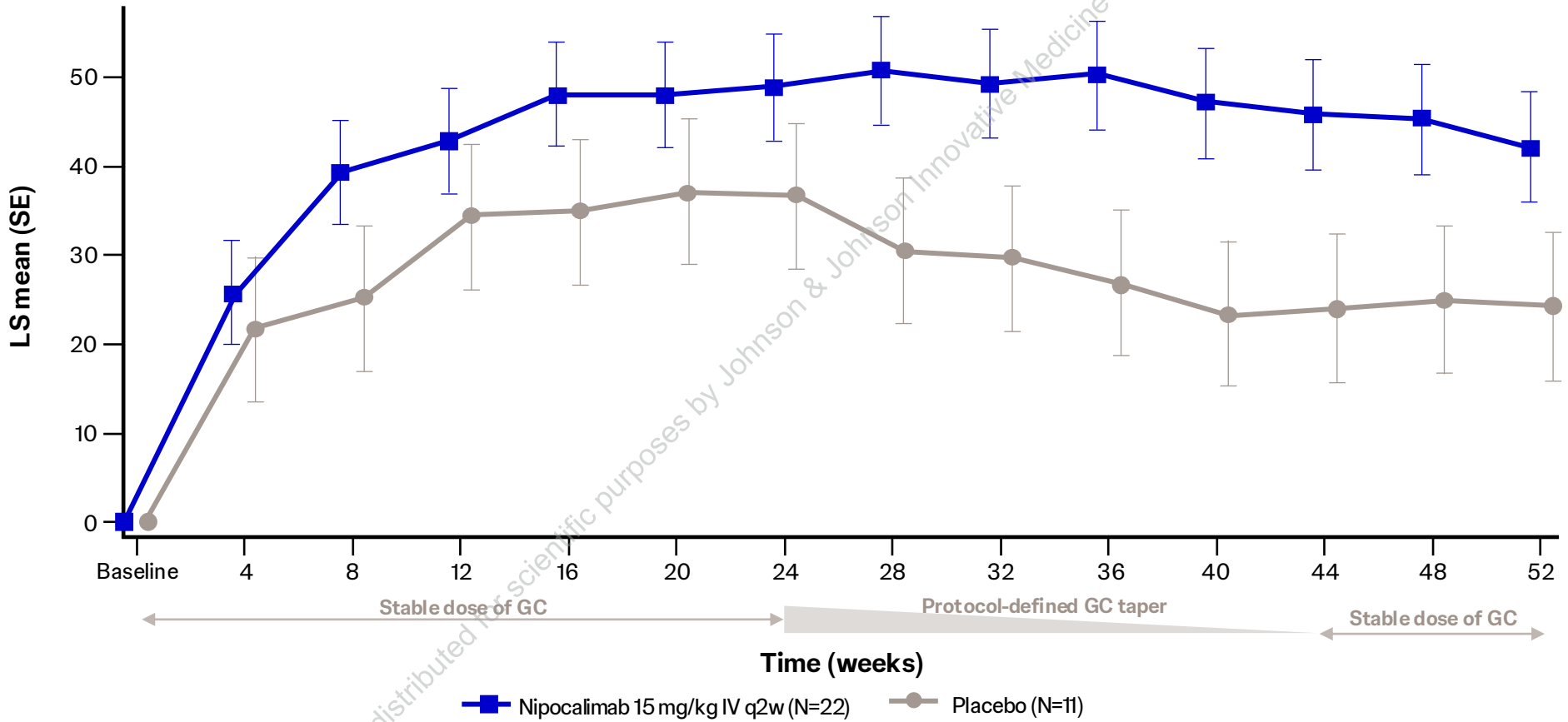
Nipocalimab group achieved a numerically greater, but nonsignificant, response of at least minimal improvement in IMACS TIS at Week 52 and on ≤5 mg/day prednisone (or equivalent) from Weeks 44 to 52 compared with the placebo group



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CI=confidence interval, **IMACS TIS**=International Myositis Assessment & Clinical Studies–Total Improvement Score, **q2w**=every 2 weeks.

IMACS TIS Over Time Through Week 52

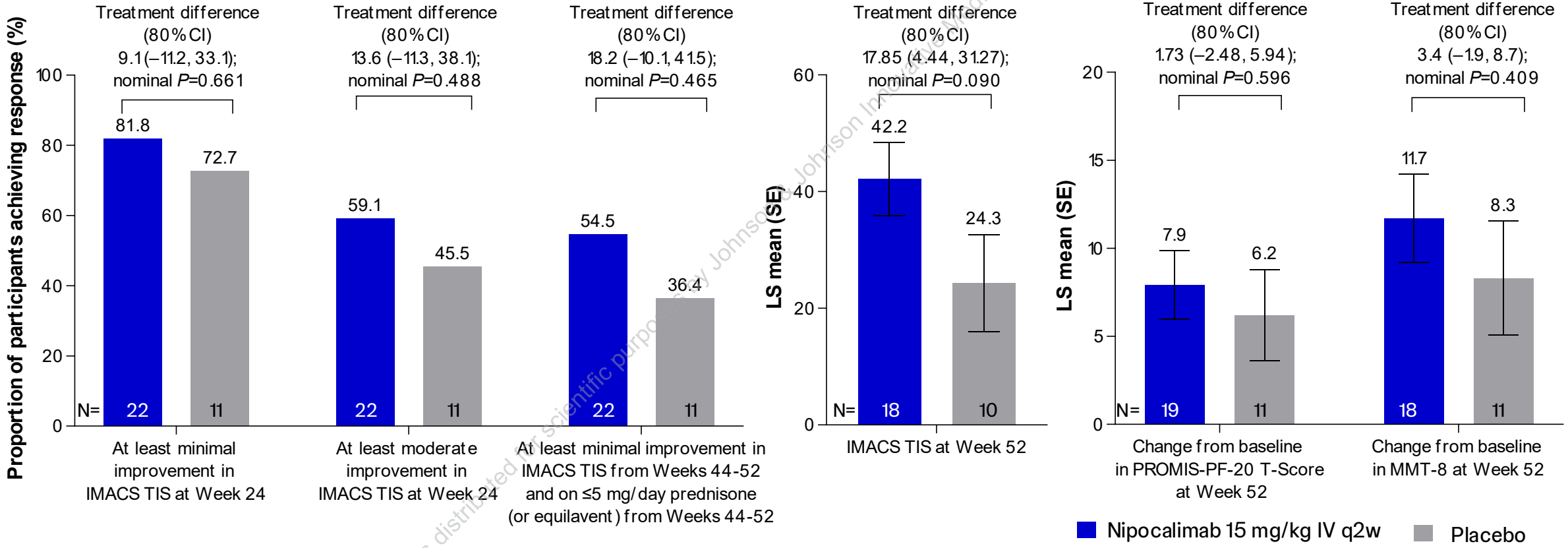


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IMACS TIS=International Myositis Assessment & Clinical Studies–Total Improvement Score, **IV**=intravenous, **LS**=least squares, **q2w**=every 2 weeks, **SE**=standard error.

Key Secondary Efficacy Endpoints

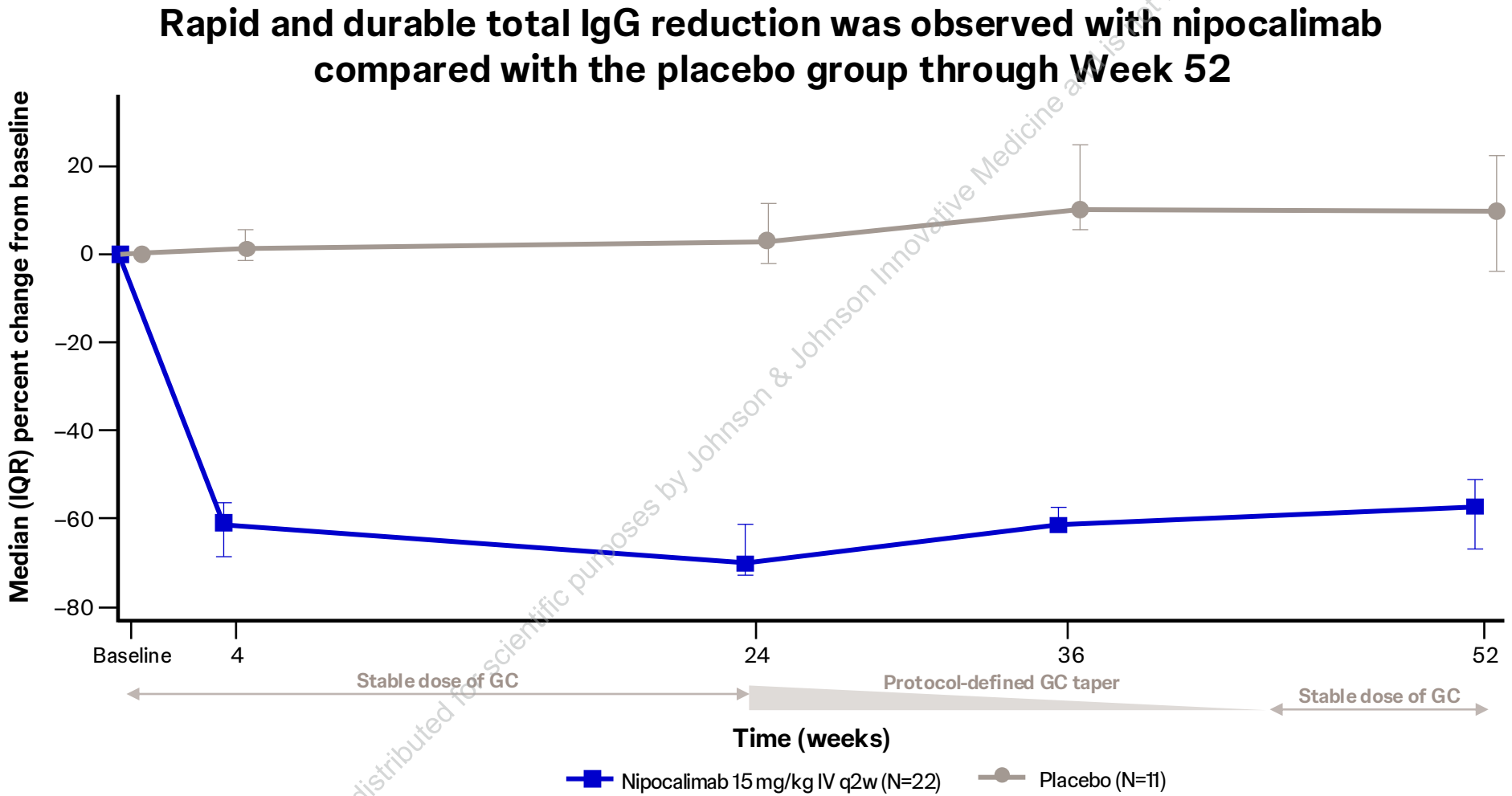
Participants treated with nipocalimab 15 mg/kg IV q2w demonstrated numerically greater improvements in key secondary endpoints compared with the placebo group



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CI=confidence interval, **IMACS TIS**=International Myositis Assessment & Clinical Studies–Total Improvement Score, **IV**=intravenous, **LS**=least squares, **MMT-8**=Manual Muscle Testing of 8 muscle groups, **PROMIS-PF-20**=Patient-Reported Outcomes Measurement Information System–Physical Function 20-item short form, **q2w**=every 2 weeks, **SE**=standard error.

Percent Change From Baseline in Total IgG Through Week 52

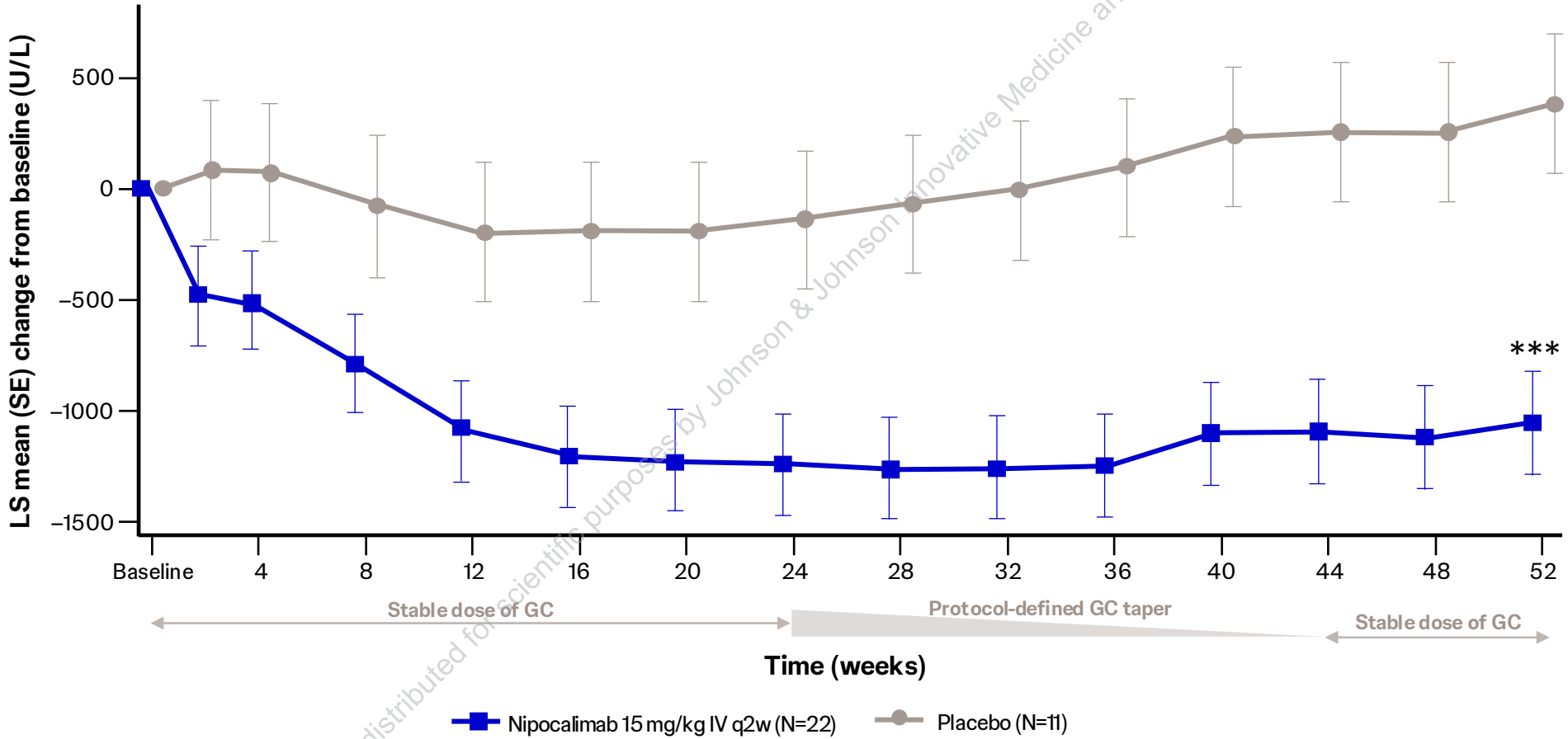


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IgG=immunoglobulin G, **IV**=intravenous, **IQR**=interquartile range, **q2w**=every 2 weeks.

Change From Baseline in Creatine Kinase Over Time Through Week 52

Greater reduction in creatine kinase was observed with nipocalimab versus placebo through Week 52



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***Nominal *P*<0.001 for nipocalimab vs placebo.
IV=intravenous, **LS**=least squares, **q2w**=every 2 weeks, **SE**=standard error.

Safety: Treatment-Emergent AEs

	Nipocalimab 15 mg/kg IV q2w (N=22)	Placebo (N=14)[a]
Any AE, n (%)	21 (95.5)	11 (78.6)
Mild	11 (50.0)	1 (7.1)
Moderate	6 (27.3)	7 (50.0)
Severe	4 (18.2)	3 (21.4)
SAE	4 (18.2)	3 (21.4)
AEs leading to permanent discontinuation of study treatment	1 (4.5)	2 (14.3)
AEs of special interest		
Severe infections or infections requiring IV anti-infective or operative/invasive intervention	3 (13.6)	0
Hypoalbuminemia	1 (4.5)	0
Deep vein thrombosis and/or pulmonary embolism	0	0
AEs of clinical interest		
Infusion reactions	1 (4.5)	1 (7.1)
Infusion-site reactions	0	0
Anaphylactic reactions or serum sickness reactions	0	0
Hyperlipidemia	2 (9.1)	2 (14.3)
Activation of latent virus	3 (13.6)	0
Opportunistic infections	0	0
Potential hypersensitivity reactions	1 (4.5)	1 (7.1)
MACE	0	0

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[a]The safety analysis set includes all participants dosed in the study, including 3 participants excluded from efficacy analyses due to ~~a site~~ data integrity issue.
AE=adverse event, **IV**=intravenous, **MACE**=major adverse cardiovascular event, **q2w**=every 2 weeks, **SAE**=serious adverse event.

Key Takeaways

A numerically greater but nonsignificant proportion of participants achieved the primary endpoint (at least minimal improvement in IMACS TIS at Week 52 and on ≤5 mg/day prednisone [or equivalent] from Weeks 44 to 52) with nipocalimab 15 mg/kg IV q2w compared with placebo

- ✓ Numerically higher improvements were also observed in secondary endpoints
- ✓ Participants treated with nipocalimab demonstrated rapid and durable IgG reduction through Week 52
- ✓ Nipocalimab was well tolerated with no new safety signals identified
- ✓ These preliminary data provide insights that FcRn blockade could potentially play a role in IIM

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IIM=idiopathic inflammatory myopathy, **IMACS TIS**=International Myositis Assessment & Clinical Studies–Total Improvement Score, **IV**=intravenous, **q2w**=every 2 weeks.

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