Development and Evaluation of the Dermatomyositis Outcomes for Muscle and Skin as an Outcome Measure in Dermatomyositis Clinical Trials

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The Total Improvement Score (TIS), which is used as the primary efficacy measure in dermatomyositis (DM) clinical trials, lacks a skin-specific measure. However, skin is a defining feature of DM. In this study, data were analyzed from the phase 3 trial of lenabasum in DM. Cutaneous Dermatomyositis Disease Area and Severity Index-Activity scores and all components of the TIS were collected at baseline and weeks 16, 28, 40, and 52. From these assessments, a composite outcome was developed, named Dermatomyositis Outcomes for Muscle and Skin, which includes certain components of the TIS and the Cutaneous Dermatomyositis Disease Area and Severity Index-Activity scores. The relative sensitivities of the TIS and Dermatomyositis Outcomes for Muscle and Skin to detect improvement in DM skin and muscle disease activity were analyzed. A total of 174 patients with DM were included, 82% were female, and 75% were White. Mean (SD) age was 51.9 (12.20) years. Treatment effect using the TIS ranged between 17.6 and 21.7 points for muscle and skin responders versus nonresponders across time points. The Dermatomyositis Outcomes for Muscle and Skin score displayed a statistically significantly greater treatment effect of 25.9−40.0 points for responders than for nonresponders, depending on the response assessed and the time point. Dermatomyositis Outcomes for Muscle and Skin is a more sensitive composite measure that reflects improvement from baseline in both skin and muscle disease activity, suggesting usefulness for use in future DM clinical trials.

Keywords: Autoimmune connective tissue disease, Clinic trials, Dermatomyositis, Outcome measures

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INTRODUCTION

Dermatomyositis (DM) is an autoimmune inflammatory myopathy that also includes skin symptoms, with heterogeneity in the amount of skin and muscle activity among those affected (Findlay et al, 2015). The mainstay of treatment in DM currently includes antimalarials, steroids, nonsteroidal immunosuppressives, and Igs. However, DM can be refractory to such standard-of-care treatments (Kurtzman and Vleugels, 2017), and this refractoriness suggests the need for new drug development and clinical trials to evaluate new drugs.

Current clinical trials in DM are largely focused on muscle improvement and may use the Total Improvement Score (TIS) as the primary efficacy measure (Aggarwal et al, 2017). The TIS is a validated composite measure scored on a scale of 1−100 and consists of 6 component measures. These include manual muscle testing (MMT) that scores muscle strength, Physician Global Assessment (PGA) and Patient Global Assessment (PtGA) that score overall disease activity, Health Assessment Questionnaire that scores functional disability, Extramuscular Global Assessment (EMGA) that scores all extramuscular disease activity, and serum muscle enzymes. The TIS assesses muscle weakness, extramuscular involvement, physical function, and global disease activity but lacks a skin-specific measure.

Skin involvement is a defining feature of DM, and assessment of skin disease activity would ideally be included in any composite endpoint designed to assess overall disease activity in DM. In addition, recent work has indicated that a large number of patients with DM can present with skin-predominant disease (Pandya et al, 2024) and that skin disease activity may be more refractory to treatment than muscle disease activity (Bhatt et al, 2024). Skin involvement is also a physically and emotionally burdensome aspect of the disease

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Abbreviations: CDASI-A, Cutaneous Dermatomyositis Disease Area and Severity Index-Activity; DM, dermatomyositis; DMOMS, Dermatomyositis Outcomes for Muscle and Skin; EMGA, Extramuscular Global assessment; MMT, manual muscle testing; PGA, Physician Global Assessment; PtGA, Patient Global Assessment; TIS, Total Improvement Score

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that is important from the patient perspective (Kleitsch et al, 2023), supporting the usefulness of including skin-specific measures in any primary composite efficacy endpoint designed to evaluate overall disease activity in DM. There is a need to develop composite outcome measures that include assessment of skin disease activity for use in future DM clinical trials.

The Cutaneous Dermatomyositis Area and Severity Index-Activity (CDASI-A) score is a validated DM skin–specific instrument that has been used in clinical trials as a primary or secondary efficacy endpoint. The CDASI-A assesses skin erythema, scale/crust/lichenification, and erosions/ulcerations in 15 anatomical locations, along with Gottron's sign/papules, periungual changes, and alopecia (Ahmed et al., 2020a; Anyanwu et al, 2015; Gaffney et al, 2019; Goreshi et al, 2012; Klein et al, 2008). The underlying hypothesis of this work was that a composite endpoint that includes CDASI-A in combination with outcome measures that measure muscle weakness and overall disease activity would better capture improvements in DM disease activity than the TIS, which does not include a skin-specific measure.

In this study, data were analyzed from the phase 3 trial of lenabasum in patients with DM with active muscle weakness and/or skin disease who were receiving background therapies, including immunosuppressives. CDASI-A scores and all components of the TIS were collected at baseline and weeks 16, 28, 40, and 52. From these assessments, a composite outcome was developed, named Dermatomyositis Outcomes for Muscle and Skin (DMOMS), which includes certain components of the TIS and the CDASI-A score. The relative sensitivities of the TIS and DMOMS to detect improvement in DM skin and muscle disease activity were compared.

RESULTS

Baseline patient demographics and disease characteristics

Data were available on 174 subjects with DM who enrolled in the phase 3 trial, all of whom met 1975 Bohan and Peter's classification criteria for probable or definite DM (Bohan and Peter, 1975a, 1975b) or the American College of Rheumatology/European League Against Rheumatism classification criteria for idiopathic inflammatory myopathy (Lundberg et al, 2017). Baseline demographics and disease characteristics are provided in Table 1. A total of 87% had classic DM, 10% had amyopathic DM, and 3% had juvenile-onset DM. A total of 75% of the patients self-identified as White, and 20% identified as Asian. The mean (SD) age was 51.9 (12.20) years, and median (interquartile range) disease duration was 4.9 (7.8) years. Mean (SD) baseline disease activity measures were MMT = 133.3 (15.50), CDASI-A = 23.4 (12.85), EMGA = 5.2 (1.82), PGA = 5.5 (1.67), PtGA = 5.1 (2.43), and Health Assessment Questionnaire = 0.8383 (0.71598).

Table 1. Baseline Subject Demographics and Disease Characteristics (N = 174)

Characteristic	Values
Age (y), mean (SD)	51.9 (12.20); range = 22–76
Sex, n (%)	Female: 142 (82)
	Male: 32 (18)
Race, n (%)	Asian: 35 (20)
	Black: 3 (2)
	White: 131 (75)
	Other: 5 (3)
Duration of disease (y), median (IQR)	4.9 (7.8); range = 0.08–31.72
DM subtype, n (%)	Amyopathic: 18 (10)
	Classic: 151 (87)
	Juvenile: 5 (3)
Disease measurement, mean (SD)	CDASI-A: 23.4 (12.85); range = 1–65
	EMGA: 5.2 (1.82); range = 0.5–9.7
	HAQ-DI: 0.8383 (0.71598); range = 0–2.63
	MMT-8: 133.3 (15.50); range = 86–150
	PGA: 5.5 (1.67); range = 2.3–9.4
	PtGA: 5.1 (2.43); range = 0–9.9

Abbreviations: CDASI-A, Cutaneous Dermatomyositis Disease Area and Severity Index-Activity; EMGA, Extramuscular Global Assessment; HAQ-DI, Health Assessment Questionnaire-Disability Index; MMT-8, Manual Muscle Testing 8; PGA, Physician Global Assessment; PtGA, Patient Global Assessment.

DMOMS composite measure and scoring

Derivation of components. The DMOMS instrument was developed using the TIS as a starting point (Table 2 and Supplementary Figures S1–S4). To develop the DMOMS instrument, elements of the TIS were assessed for redundancy and utility in measuring different disease aspects of DM.

Pearson's correlation of component measures of the TIS using baseline data showed that PGA and EMGA were redundant measures ($r = 0.7$) (Table 3). The PGA has been validated to measure disease activity in DM and correlates well with other objective disease measures (Rider et al, 2011). The PGA is included as a core measure for DM by the International Myositis Assessment and Clinical Studies Group and American College of Rheumatology/European League Against Rheumatism. The EMGA is another PGA score but has not been validated as a measure of disease activity in DM in a clinical trial setting. For these reasons, PGA was included as a component in DMOMS for assessment of clinical responsiveness from the physician's perspective, and EMGA was excluded.

The PtGA, similar to PGA, has been validated to measure overall disease activity in myositis and displays good inter-rater reliability (Rider et al, 2011). It was included in DMOMS to assess clinical benefit from the patient's perspective.

The CDASI-A is a reliable tool to measure skin disease in DM and was added as a skin-specific measure in DMOMS. CDASI-A has been fully validated in multiple studies; has excellent inter- and intrarater reliability; and correlates well with skin activity, QOL, and biomarkers of disease activity (Ahmed et al., 2020b, Anyanwu et al, 2015; Gaffney et al, 2019; Goreshi et al, 2012; Huard er al, 2017; Klein et al, 2008). The CDASI-A score at baseline was not redundant ($r < 0.7$) with MMT-8, PGA, or PtGA (Table 4). For these reasons, the CDASI-A was included as a component in the DMOMS score to assess skin disease activity.

The MMT is a valid, reliable, and consistent measure of muscle strength and is included as a core activity measure for DM by International Myositis Assessment and Clinical Studies Group and American College of Rheumatology/

Table 2. The Dermatomyositis Outcomes for Muscle and Skin Scoring Instrument

Measure	Level of Improvement	Improvement Score
Manual muscle testing or childhood myositis assessment scale (150-point maximum scale)	≤3-point improvement	0
	4–7-point improvement	10
	8–12-point improvement	20
	13–19-point improvement	27.5
	≥20-point improvement	32.5
Cutaneous dermatomyositis disease area and severity index (100-point maximum scale)	≤4-point improvement	0
	5–7-point improvement	10
	8–12-point improvement	20
	13–19-point improvement	27.5
	≥20-point improvement	32.5
Physician Global Assessment (0–10 scale)	≤0.5-point improvement	0
	0.6–1.5-point improvement	7.5
	1.6–2.5-point improvement	15
	2.6–4.0-point improvement	17.5
	≥4.1-point improvement	20
Patient or Parent Global Assessment (0–10 scale)	≤0.5-point improvement	0
	0.6–1.5-point improvement	4
	1.6–2.5-point improvement	7.5
	2.6–4.0-point improvement	11
	≥4.1-point improvement	15

European League Against Rheumatism (Rider et al, 2011). As a direct measure of muscle disease, MMT was included in the DMOMS tool.

The Health Assessment Questionnaire has been used to assess functional limitation in myositis, but it is a joint-specific tool that was originally developed for use in rheumatoid arthritis and has limited validity in DM (Rider et al, 2011). This component was removed as a component of DMOMS.

Up to 5% of patients with DM with documented muscle involvement have normal serum muscle enzyme levels throughout the course of disease (Bohan et al, 1977), and muscle enzymes are only indirect measures of muscle involvement that often do not reflect the degree of muscle disease. It has also been documented that up to 20% of patients with DM can have normal serum creatinine kinase levels and that other muscle enzymes (lactate dehydrogenase, aspartate aminotransferase, alanine amino transferase, and aldolase) are less sensitive and also may not be elevated (Malik et al, 2016). Consequently, muscle enzymes were removed in the DMOMS tool.

Table 3. Pearson's Correlation Coefficients for the Component Measures of TIS at Baseline

Comparison	Pearson's Correlation Coefficient
MMT versus PGA	−0.4
MMT versus PtGA	−0.3
MMT versus EMGA	0
MMT versus HAQ	−0.6
MMT versus muscle enzymes	−0.1
PGA versus PtGA	0.3
PGA versus EMGA	0.7
PGA versus HAQ	0.3
PGA versus muscle enzymes	0.1
PtGA versus EMGA	0.2
PtGA versus HAQ	0.5
PtGA versus muscle enzymes	−0.1
EMGA versus HAQ	0.0
EMGA versus muscle enzymes	−0.1
HAQ versus muscle enzymes	0.1

Abbreviations: EMGA, Extramuscular Global Assessment; HAQ, Health Assessment Questionnaire; MMT, manual muscle testing; PGA, Physician Global Assessment; PtGA, Patient Global Assessment; TIS, Total Improvement Score.

Derivation of scoring scale. The scoring scale used in DMOMS was adapted from the scoring method of TIS (Table 2 and Supplementary Figures S1–S4). The decision was made to weight MMT and CDASI-A equally because both muscle and skin diseases are cardinal features of DM. The TIS scoring uses absolute percentage changes (Aggarwal et al, 2017). With this approach, large changes in disease activity in MMT-8 or CDASI-A would have been needed for incremental improvements in overall DMOMS score. Instead, a point-based scale was chosen to score MMT-8 and CDASI-A to improve sensitivity to change in the overall score. The MMT score was weighted the same as in the TIS, and CDASI-A was weighted very similarly by points. The PtGA and PGA

Table 4. Pearson's Correlation Coefficients for the Component Measures of DMOMS at Baseline

Comparison	Pearson's Correlation Coefficient
CDASI versus MMT	0.2
CDASI versus PtGA	−0.02
CDASI versus PGA	0.2
MMT versus PtGA	−0.3
MMT versus PGA	−0.4
PtGA versus PGA	0.3

Abbreviations: CDASI, Cutaneous Dermatomyositis Disease Area and Severity Index; DMOMS, Dermatomyositis Outcomes for Muscle and Skin; EMGA, Extramuscular Global Assessment; HAQ, Health Assessment Questionnaire; MMT, manual muscle testing; PGA, Physician Global Assessment; PtGA, Patient Global Assessment; TIS, Total Improvement Score.

components were also scored on a point-based scale to standardize scoring of the tool. The PGA was weighted the same as in the TIS. The PtGA was assigned a 50% increase in weight compared with its weight in the TIS because it was considered important to capture patient input on clinical improvement.

Improvement in the TIS and DMOMS

The mean scores for the TIS and DMOMS at weeks 16, 28, 40, and 52 were compared for the overall patient group as well as subgroups of CDASI-A and MMT responders and nonresponders. CDASI-A responders included those with a ≥8-point improvement in scores between baseline and the time point, and MMT responders were those with a ≥10-point improvement in scores between baseline and the time point. The ability of TIS versus DMOMS score to reflect differences between skin and muscle responders and nonresponders over time was compared to address the sensitivity to change of the 2 composite outcome measures. Given that responders and nonresponders were defined by CDASI-A and MMT scores for the subgroup analysis, but these scores are also components of the overall DMOMS scoring system, the ability of TIS versus DMOMS score to reflect differences in the overall patient group over time was also compared, irrespective of response group.

Across all time points, DMOMS scores had a greater mean (range = 25.5−41.1 points) for the overall patient group than TIS (range = 22.6−35.7 points), with a statistically significant difference ($P < .05$) seen at week 40 (Figure 1).

At all time points after baseline, DMOMS scores had a greater mean for skin responders, ranging from 50.4 to 62.5 points, than mean TIS, which ranged from 38.6 to 46.7 points (Figure 2a). Differences between mean DMOMS scores and TIS were statistically significant ($P < .05$) at all time points after baseline. For skin nonresponders, the mean DMOMS scores ranged between 17.5 and 22.5 points, and mean TIS ranged from 17.5 to 26.2 points, depending on the time point (Figure 2b). None of the differences between mean DMOMS scores and TIS were statistically significant. Furthermore, DMOMS scores displayed treatment effects for responders versus nonresponders ranging from 31.9 to 40.0 points, depending on the time point, whereas the TIS displayed a treatment effect ranging from 18.5 to 21.1 points or about half that of DMOMS scores. The treatment effect of DMOMS was statistically significantly greater ($P < .05$) than the treatment effect of the TIS.

Similarly, for the muscle responders, the DMOMS scores had a greater mean (range = 47.8−65.3 points) than TIS (range = 39.6−48.8 points) at all time points after baseline, with most differences between mean DMOMS scores and mean TIS being statistically significant ($P < .05$) (Figure 3a). For muscle nonresponders, the mean DMOMS scores ranged between 21.9 and 32.2 points, and the mean TIS ranged from 19.9 to 30.7 points (Figure 3b). None of these differences between mean DMOMS scores and TIS were statistically significant. Furthermore, DMOMS scores displayed treatment effect for responders versus nonresponders ranging from 25.9 to 33.7 points, depending on the time point, whereas the TIS displayed a treatment effect ranging from 17.6 to 21.7 points. The treatment effect of DMOMS was statistically significantly greater ($P < .05$) than the treatment effect of the TIS.

DISCUSSION

Skin manifestations in DM have a significant impact on the patient's functionality and QOL (Goreshi et al, 2011; Robinson et al, 2015). When developing composite outcome measures for use as primary endpoints in DM clinical trials, outcomes that include a direct measure of skin activity may be more sensitive to overall disease activity than composite measures that do not, especially given that skin involvement is a defining characteristic of DM. This study developed a composite outcome, the DMOMS, and compared the responsiveness of the TIS and DMOMS outcomes with data obtained from the large, prospective phase 3 trial of lenabasum in DM. Similar to the TIS, DMOMS assigns points only when improvement is present.

The TIS is a composite measure of DM disease activity that is heavily weighted toward muscle and does not include a skin-specific measure. The TIS has been used as the primary efficacy endpoint in DM clinical trials (Aggarwal et al, 2022; Werth et al, 2022). The PGA and EMGA components of the TIS were found to be redundant. Treatment effect using the

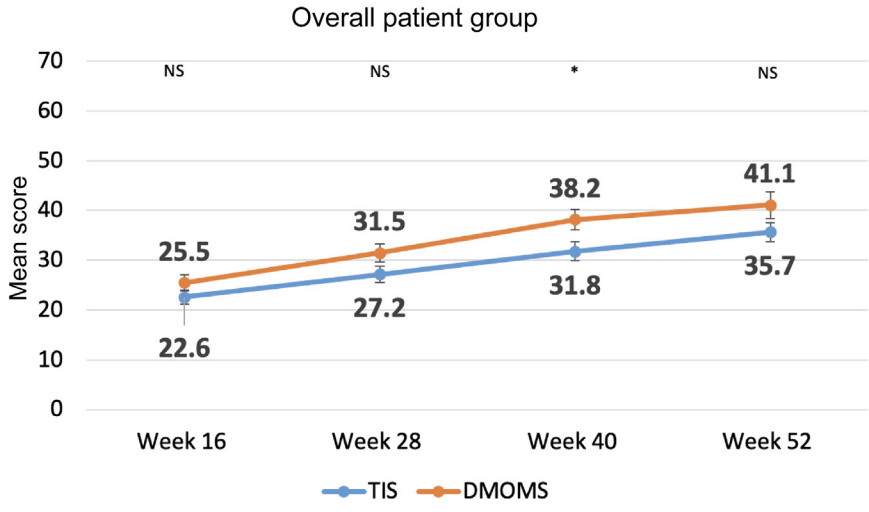

Figure 1. Improvement in TIS and DMOMS for all patients. Each data point represents the mean score ± SEM. *$P \leq .05$. DMOMS, Dermatomyositis Outcomes for Muscle and Skin; NS, not significant; TIS, Total Improvement Score.

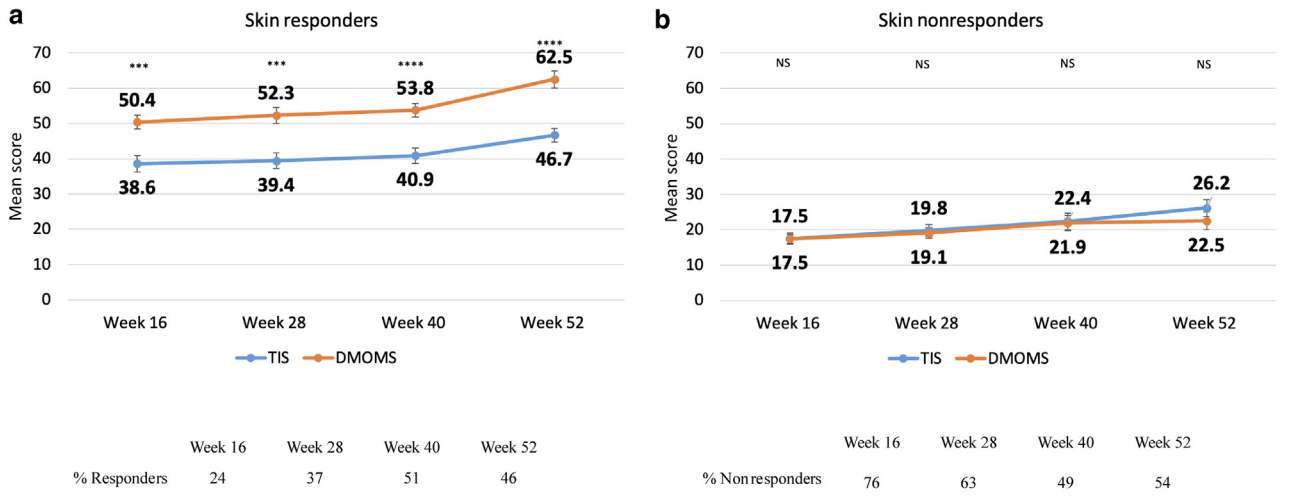

Figure 2. Improvement in TIS and DMOMS for CDASI-A. (**a**) Responders and (**b**) nonresponders. Each data point represents the mean score ± SEM. ***$P \leq .001$, and ****$P \leq .0001$. CDASI, Cutaneous Dermatomyositis Disease Area and Severity Index; DMOMS, Dermatomyositis Outcomes for Muscle and Skin; NS, not significant; TIS, Total Improvement Score.

TIS in the lenabasum study ranged between 17.6 and 21.7 points for muscle and skin responders versus nonresponders across time points.

DMOMS is a simpler composite outcome than TIS, with 4 versus 6 component measures. In addition, DMOMS lacks redundant components, includes CDASI-A scores that are weighted equally by points to MMT scores, and assigns greater weight to the PtGA to increase impact of clinical benefit as assessed by patients. The DMOMS score displayed a treatment effect of 25.9−40.0 points for responders versus nonresponders, depending on the response assessed and the time point after baseline. The DMOMS score measured 3−6 points greater improvement in the overall patient group than the TIS, on the basis of the time point assessed. The DMOMS score also consistently measured 12−16 points greater improvement in skin responders than the TIS and 7−13 points greater improvement in muscle responders, whereas scores in skin and muscle nonresponders were similar. Similarity in mean scores of nonresponders for DMOMS and the TIS for muscle and skin at each time point after baseline may reflect the design of both scoring systems to provide points only for improvement.

On the basis of these findings, DMOMS was more sensitive to improvement in both skin and muscle disease activity and identified a larger treatment effect. In fact, DMOMS scores identified up to twice the treatment effect as the TIS depending on the response assessed and the time point after baseline. In addition, the treatment effect of DMOMS was statistically significantly greater than that of the TIS. These findings suggest that DMOMS may be better suited than the TIS to detect improvement in DM clinical trials that include patients with all DM phenotypes and may even allow a smaller sample size, which would be important in DM, a rare disease.

It is also important to note that MMT and CDASI-A scores should negatively correlate, under the assumption that skin

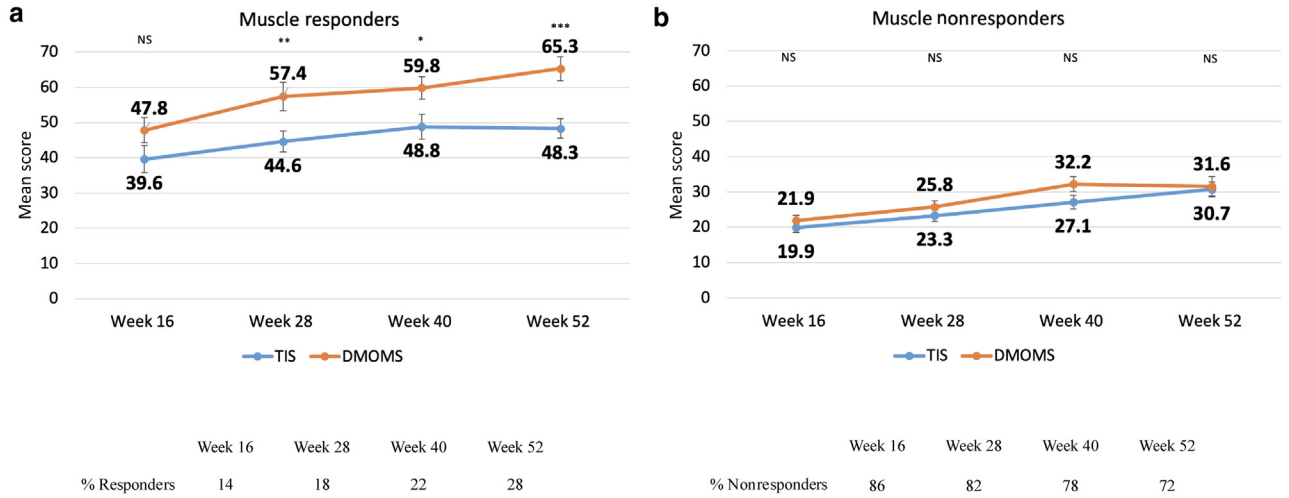

Figure 3. Improvement in TIS and DMOMS for MMT. (**a**) Responders and (**b**) nonresponders. Each data point represents the mean score ± SEM. *$P \leq .05$, **$P \leq .01$, ***$P \leq .001$. DMOMS, Dermatomyositis Outcomes for Muscle and Skin; NS, not significant; TIS, Total Improvement Score.

disease changes with muscle disease in DM. However, Pearson's correlation of CDASI-A and MMT scores at baseline showed that these measures do not correlate inversely, indicating that skin disease changes independently of muscle disease and that the MMT is not a reliable measure for reflecting changes in skin disease activity. Furthermore, Pearson's correlation of CDASI-A and PGA scores at baseline did not indicate any correlation, suggesting that physicians rate muscle disease more than skin disease in the PGA. Together, this further strengthens the need for a composite outcome measure such as DMOMS that independently measures skin disease improvement in DM.

A strength of this paper is that data were prospectively collected from a clinical trial in a large group of patients with heterogeneity in DM presentation. Because of the trial design, a limitation of this study is that data from fewer patients were available at weeks 40 and 52.

In conclusion, DMOMS is a composite outcome that is a more sensitive composite measure that reflects improvement from baseline in skin and muscle disease activity, suggesting potential usefulness for use in future DM clinical trials as a more sensitive option than the TIS.

MATERIALS AND METHODS

Data collection and analysis

Posthoc analysis of prospectively collected data from the lenabasum phase 3 trial in DM (ClinicalTrials.gov Identifier: NCT03813160) was conducted. All components of the TIS and CDASI-A scores were collected at baseline and at weeks 16, 28, 40, and 52. Patients were analyzed as a single group irrespective of treatment allocation because the study did not achieve statistical significance for its primary efficacy endpoint of the TIS at week 28. Patients that had $\geq$10-point improvement in MMT scores or $\geq$8-point improvement in CDASI-A scores at each time point compared with baseline were considered to be responders. These thresholds for each category of improvement were based on documented or reasonable values for minimal clinically important differences of these measures. In the setting of a clinical trial, the minimal clinically important difference for CDASI-A has been reported to be an improvement of 5.5−7.8 points (Pandya et al, 2023), so a conservative measure of an 8-point improvement was used. There is currently no documented minimal clinically important difference for MMT, so the generally accepted value of a 10-point improvement was used. These thresholds for each category of improvement were established prior to analyses.

To assess for redundancy in TIS, a Pearson's correlation was conducted on absolute values of all component measures at baseline, with $r \geq 0.7$ indicating redundancy. To assess efficacy of the TIS and the DMOMS scores in capturing skin and muscle disease improvement, a Student's *t*-test was used to compare mean scores at weeks 16, 28, 40, and 52 in the overall patient group and in both responders and nonresponders. The treatment effect of responders versus nonresponders for the TIS and the DMOMS across the time points was also assessed and compared using a Student's *t*-test.

ETHICS STATEMENT

This study was performed in accordance with the Declaration of Helsinki. This study completed secondary analysis of data collected from the phase 3 trial of lenabasum in dermatomyositis. The phase 3 trial was a multicenter study conducted in accordance with the Declaration of Helsinki and Good Clinical Practice and approved by Corbus Pharmaceuticals and central or local Institutional Review Board or Independent Ethics Committee at each site. Written informed consent was also obtained.

DATA AVAILABILITY STATEMENT

Requests for clinical trial data should be sent to VPW (werth@pennmedicine.upenn.edu).

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CONFLICT OF INTEREST

BW is a former employee and current shareholder of Corbus Pharmaceuticals. VPW has received research support from Corbus Pharmaceuticals. The remaining authors state no conflict of interest.

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AUTHOR CONTRIBUTIONS

Conceptualization: RP, JD, BW, VPW; Data Curation: RP, JD; Funding Acquisition: BW, VPW; Supervision: BW, VPW; Writing - Original Draft Preparation: RP; Writing - Reviewing and Editing: RP, JK, DL, BW, VPW

DECLARATION OF GENERATIVE ARTIFICIAL INTELLIGENCE (AI) OR LARGE LANGUAGE MODELS (LLMS)

The authors did not use AI/LLM in any part of the research process and/or manuscript preparation.

SUPPLEMENTARY MATERIAL

Supplementary material is linked to the online version of the paper at www.jidonline.org, and at https://doi.org/10.1016/j.xjidi.2024.100337.

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Cutaneous Dermatomyositis Disease Area and Severity Index (CDASI) *ver02*Select

the score in each anatomical location that describes the most severely affected dermatomyositis-associated skin lesion

Extent

	activity			damage		
Anatomical Location	Erythema	Scale	Erosion/ Ulceration	Poikiloderma (Dyspigmentation or Telangiectasia)	Calcinosis	Anatomical Location
	0-absent 1-pink; faint erythema 2-red 3-dark red	0-absent 1-scale 2-crust; lichenification	0-absent 1-present	0-absent 1-present	0-absent 1-present	
Scalp						Scalp
Malar Area						Malar Area
Periorbital						Periorbital
Rest of the face						Rest of the face
V-area neck (frontal)						V-area neck (frontal)
Posterior Neck						Posterior Neck
Upper Back & Shoulders						Upper Back & Shoulders
Rest of Back & Buttocks						Rest of Back & Buttocks
Abdomen						Abdomen
Lateral Upper Thigh						Lateral Upper Thigh
Rest of Leg & Feet						Rest of Leg & Feet
Arm						Arm
Mechanic's Hand						Mechanic's Hand
Dorsum of Hands (not over joints)						Dorsum of Hands (not over joints)
Gottron's – Not on Hands						Gottron's – Not on Hands

Gottron's – Hands

Examine patient's hands and double score if papules are present		Ulceration	Examine patient's hands and score if damage is present	
0-absent 1-pink; faint erythema 2-red erythema 3-dark red			0-absent 1-dyspigmentation 2-scarring	

Periungual

Periungual changes (examine)			
0-absent 1-pink; red erythema/microscopic telangiectasias 2-visible telangiectasias			

Alopecia

Recent Hair loss (within last 30 days as reported by patient)			
0-absent 1-present			

(For the activity score, please add up the scores of the left side, i.e. Erythema, Scale, Excoriation, Ulceration, Gottron's, Periungual, Alopecia

Total Activity Score

(For the damage score, add up the scores of the right side, i.e. Poikiloderma, Calcinosis)

Total Damage Score

Signature _____ Date____/____/____

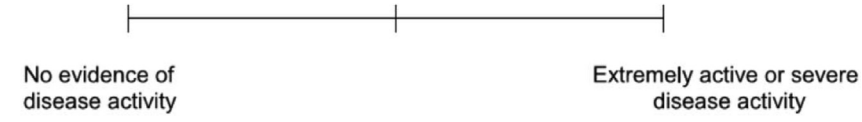
Supplementary Figure S1. Cutaneous Dermatomyositis Disease Area and Severity Index.

Muscle Groups	Right (0 – 10)	Left (0 – 10)	Axial (0 – 10)
Axial Muscles (0 – 10)			
Neck flexors	X	X	0-10
Proximal Muscles (0 – 100)			
Deltoid	0-10	0-10	X
Biceps	0-10	0-10	X
Gluteus maximus	0-10	0-10	X
Gluteus medius	0-10	0-10	X
Quadriceps	0-10	0-10	X
Distal Muscles (0 – 40)			
Wrist extensors	0-10	0-10	X
Ankle dorsiflexors	0-10	0-10	X
MMT- 8 score (0 – 150)	0-70	0-70	0-10

Supplementary Figure S2. Manual Muscle Testing – 8 items.

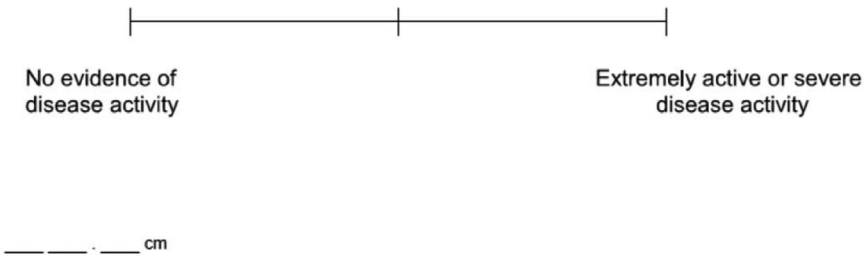
Key to Muscle Grading

	Function of the Muscle	Grade		
No Movement	No contractions felt in the muscle	0	0	Zero
	Tendon becomes prominent or feeble contraction felt in the muscle, but no visible movement of the part	T	1	Trace
Test Movement	**MOVEMENT IN HORIZONTAL PLANE**			
	Moves through partial range of motion	1	2-	Poor-
	Moves through complete range of motion	2	2	Poor
	ANTIGRAVITY POSITION / Moves through partial range of motion	3	2+	
Test Position	*Gradual* release from test position	4	3-	Fair-
	Holds test position (no added pressure)	5	3	Fair
	Holds test position against slight pressure	6	3+	Fair+
	Holds test position against slight to moderate pressure	7	4-	Good-
	Holds test position against moderate pressure	8	4	Good
	Holds test position against moderate to strong pressure	9	4+	Good+
	Holds test position against strong pressure	10	5	Normal



___ ___ . ___ cm

Supplementary Figure S3. Physician Global Assessment. Global (overall) disease activity at each study visit is rated by drawing a vertical mark on the 10-cm line shown in the figure according to the following scale: left end of line = no evidence of disease activity and right end of line = extremely active or severe disease activity, with midpoint at 5 cm.



Supplementary Figure S4. Patient Global Assessment. Global (overall) disease activity at each study visit is rated by drawing a vertical mark on the 10-cm line below according to the following scale: left end of line = no evidence of disease activity and right end of line = extremely active or severe disease activity, with midpoint at 5-cm.