2016 ACR/EULAR Criteria for Minimal, Moderate, and Major Clinical Response in Adult Dermatomyositis and Polymyositis and Juvenile Dermatomyositis

Physician Global Activity Physician Global Activity Series of 15% improvement 15. >5% to 15% improvement 15. >25% to 40% improvement 17.5 >40% improvement 20. Worsening to 5% improvement 2.0 Worsening to 5% improvement 2.5 >5% to 15% improvement 2.5 >5% to 15% improvement 5. >5% to 15% improvement 5. >25% to 40% improvement 5. >25% to 40% improvement 7.5 >40% improvement 10. Worsening to 2% improvement 10. Worsening to 2% improvement 10. Worsening to 2% improvement 20. >20% to 30% improvement 20. Assessment Scale 20% to 30% improvement 32.5 Worsening to 5% improvement 32.5 Worsening to 5% improvement 5. >30% improvement 7.5 >30% improvement 7.5 >25% to 15% improvement 7.5 >25% to 40% improvement 7.5 >25% to 40% improvement 7.5 >25% to 40% improvement 7.5 >25% to 15% improvement 10. Worsening to 5% improvement 7.5 >25% to 40% improvement 7.5 >25% to 15% improvement 7.5 >25% to 15% improvement 7.5 >25% to 40% improvement 7.5 >25% to 15% improvement 7.5 >25% to 40% improvement 7.5 >25% to 40% improvement 7.5 >25% to 15% improvement 7.5 >25% to 40% improvement 7.5 >25% to 15% improvement 7.5 >25% to 40% improvement 7.5 >25% to 15% improvement 7.5 >25% to 15% improvement 7.5 >25% to 15% improvement 7.5 >25% to 40% improvement	Core Set Measure*	Level of Improvement Based on absolute percentage change	Improvement Score
Physician Global Activity Physician Global Activity Patient or Parent Global Activity Patient or Parent Global Activity Manual Muscle Testing or Childhood Myositis Assessment Scale (Childhood) Health Assessment Questionnaire Enzyme (most abnormal) or CHQ-PhS Extramuscular Activity or Disease Activity Score Extramuscular Activity or Disease Activity Score Extramuscular Activity or Disease Activity Some DM/PM thresholds Patient or Parent Global Activity 10 Worsening to 5% improvement 2.5 2.5% to 40% improvement 2.5 2.5% to 10% improvement 2.0 2.0% to 30% improvement 2.0 2.0% to 30% improvement 2.10 2.20% to 30% improvement 2.25 2.20% to 30% improvement 2.25 2.20% to 40% improvement 2.25 2.25% to 40%	Physician Global Activity		0
Physician Global Activity		•	-
Patient or Parent Global Activity Solution Solut		·	15
Patient or Parent Global Activity			17.5
Patient or Parent Global Activity Sto 15% to 15% improvement 5			20
Patient or Parent Global Activity >15% to 25% improvement 5 >25% to 40% improvement 10 Worsening to 2% improvement 10 Manual Muscle Testing or Childhood Myositis >10% to 20% improvement 20 Assessment Scale >20% to 30% improvement 27.5 >30% improvement 32.5 Worsening to 5% improvement 5 Assessment Questionnaire >5% to 15% improvement 7.5 Questionnaire >25% to 40% improvement 7.5 >25% to 40% improvement 10 Worsening to 5% improvement 7.5 >40% improvement 10 Worsening to 5% improvement 7.5 >40% improvement 10 Worsening to 5% improvement 5 >25% to 15% improvement 5 >25% to 15% improvement 7.5 >40% improvement 7.5 >40% improvement 7.5 >40% improvement 7.5 >25% to 15% improvement 7.5 >40% improvement 12.5 >25% to 40% improvement 15 >40% improvement 20 Total improvement 30 Minimal ≥ 20 DM/PM thresholds Moderate ≥ 40 DM/PM thresholds 26% 20% Advance 20% 20%		Worsening to 5% improvement	0
Activity Solition Solition		>5% to 15% improvement	2.5
S25% to 40% improvement 7.5		>15% to 25% improvement	5
Manual Muscle Testing or Childhood Myositis Assessment Scale>2% to 10% improvement10Assessment Scale>20% to 30% improvement20>20% to 30% improvement27.5>30% improvement32.5(Childhood) Health Assessment Questionnaire>5% to 15% improvement525% to 40% improvement Questionnaire>15% to 25% improvement7.525% to 40% improvement Or CHQ-PhS>5% to 15% improvement025% to 40% improvement Or CHQ-PhS>5% to 15% improvement025% to 40% improvement Soft improvement Or CHQ-PhS>5% to 15% improvement7.525% to 40% improvement Soft improvement Soft improvement Soft improvement7.525% to 40% improvement Soft improvement Soft improvement Soft improvement7.525% to 15% improvement Soft improvement Soft improvement Soft improvement7.525% to 25% improvement Soft improvement Soft improvement Soft improvement2.025% to 40% improvement Soft improvement Soft improvement Soft improvement2.025% to 40% improvement Soft improvement S	Activity	>25% to 40% improvement	7.5
Manual Muscle Testing or Childhood Myositis Assessment Scale>2% to 10% improvement10Assessment Scale>20% to 30% improvement27.5>30% improvement32.5(Childhood) Health Assessment Questionnaire>5% to 15% improvement525% to 40% improvement Questionnaire>15% to 25% improvement7.525% to 40% improvement Or CHQ-PhS>5% to 15% improvement025% to 15% improvement Or CHQ-PhS>5% to 15% improvement025% to 40% improvement Score Or CHQ-PhS>5% to 15% improvement525% to 40% improvement Score Or CHQ-PhS>25% to 15% improvement7.525% to 15% improvement Score Or CHQ-PhS>5% to 15% improvement7.525% to 40% improvement Score Or CHQ-PhS>5% to 15% improvement7.525% to 15% improvement Score Or CHQ-PhS>5% to 15% improvement7.525% to 40% improvement Score Or CHQ-PhS>5% to 15% improvement7.525% to 15% improvement Score Or CHQ-PhS>5% to 15% improvement7.525% to 40% improvement Score Or CHQ-PhS>5% to 15% improvement7.525% to 40% improvement Score Or CHQ-PhS>5% to 15% improvement7.525% to 40% improvement Score Or CHQ-PhS>5% to 15% improvement7.525% to 40% improvement Score Or CHQ-PhS>5% to 15% improvement7.525% to 40% improvement Score Or CHQ-PhS>5% to 15% improvement7.525% to 40% improvement Score Or CHQ-PhS>5% to 15% improvement7.525% to 40% improvement Score Or CHQ-PhS>5% to 15% improvement7.5		>40% improvement	10
Childhood Myositis Assessment Scale>10% to 20% improvement >20% to 30% improvement20Assessment Scale>20% to 30% improvement27.530% improvement32.5(Childhood) Health Assessment 	Childhood Myositis	Worsening to 2% improvement	0
Assessment Scale >20% to 30% improvement >30% improvement 32.5 Worsening to 5% improvement 5 Assessment Questionnaire 25% to 15% improvement 25% to 25% improvement 25% to 25% improvement 25% to 40% improvement 25% to 40% improvement 25% to 15% improvement 25% to 25% improvement 25% to 15% improvement 25% to 25% improvement 25% to 25% improvement 25% to 40% improvement 20 Improvement category Minimal ≥ 20 DM/PM thresholds Moderate		>2% to 10% improvement	10
S20% to 30% improvement 27.5		>10% to 20% improvement	20
(Childhood) Health Assessment Questionnaire Assessment Questionnaire Enzyme (most abnormal) or CHQ-PhS Extramuscular Activity or Disease Activity Score Dimetals Activity Score Dimetals Activity Score Dimetals Activity Score Minimal DM/PM thresholds Worsening to 5% improvement >5% to 15% improvement >2.5 >10 >10 Worsening to 5% improvement 2.5 >5% to 15% improvement 2.5 >15% to 25% improvement 5 >25% to 40% improvement 7.5 >40% improvement 7.5 >15% to 25% improvement 7.5 >15% to 15% improvement 7.5 >15% to 25% improvement 7.5 >15% to 25% improvement 12.5 >25% to 40% improvement 15 >40% improvement 20 Total improvement score † Minimal ≥ 20 DM/PM thresholds Moderate ≥ 40		>20% to 30% improvement	27.5
(Childhood) Health Assessment Questionnaire Assessment Questionnaire >25% to 25% improvement >25% to 40% improvement -40% improvement -50% to 15% improvement -50% to 25% improvement -50% to 25% improvement -50% to 25% improvement -7.5 -40% improvement -7.5 -40% improvement -7.5 -7		>30% improvement	32.5
Assessment Questionnaire >25% to 40% improvement >40% improvement >40% improvement >5% to 15% improvement >5% to 15% improvement >5% to 15% improvement >10 >5% to 15% improvement >5% to 15% improvement >2.5 >15% to 25% improvement >40% improvement 7.5 >40% improvement 7.5 >40% improvement 7.5 >40% improvement 7.5 >5% to 15% improvement 7.5 >40% improvement 2.5 >40% improvement 7.5 >5% to 15% improvement 12.5 >15% to 25% improvement 20 Total improvement score † Minimal ≥ 20 DM/PM thresholds Moderate ≥ 40		Worsening to 5% improvement	0
Questionnaire >25% to 40% improvement >40% improvement 10 Worsening to 5% improvement >5% to 15% improvement >10 >5% to 15% improvement >10 >5% to 15% improvement >10 >5% to 15% improvement 5 >25% to 40% improvement 7.5 >40% improvement 7.5 >40% improvement 7.5 Worsening to 5% improvement >5% to 15% improvement 7.5 >40% improvement 7.5 >5% to 15% improvement 10 >5% to 15% improvement 20 15% to 25% improvement 20 Improvement category Minimal ≥ 20 DM/PM thresholds Moderate ≥ 40	Assessment	>5% to 15% improvement	5
Section 40% improvement 10		>15% to 25% improvement	7.5
Enzyme (most abnormal) or CHQ-PhS Enzyme (most abnormal) or CHQ-PhS >55% to 15% improvement		>25% to 40% improvement	7.5
Enzyme (most abnormal) or CHQ-PhS		>40% improvement	10
Finzyme (most abnormal) or CHQ-PhS >15% to 25% improvement 5 >25% to 40% improvement 7.5 >40% improvement 0 >5% to 15% improvement 7.5 >15% to 25% improvement 12.5 >15% to 25% improvement 12.5 >25% to 40% improvement 15 >40% improvement 20 Improvement category Total improvement score †		Worsening to 5% improvement	0
or CHQ-PhS >15% to 25% improvement >25% to 40% improvement 7.5 >40% improvement 7.5 Worsening to 5% improvement 0 >5% to 15% improvement 7.5 >15% to 25% improvement 12.5 >25% to 40% improvement 15 >40% improvement 15 >40% improvement 20 Improvement category Minimal ≥ 20 DM/PM thresholds Moderate ≥ 40		>5% to 15% improvement	2.5
>25% to 40% improvement7.5>40% improvement7.5Authorized Paramuscular Activity or Disease Activity Score>5% to 15% improvement7.5>15% to 25% improvement12.5>25% to 40% improvement15>40% improvement20Improvement categoryTotal improvement score†DM/PM thresholdsModerate≥ 40		>15% to 25% improvement	5
Extramuscular Activity or Disease Activity Score ->5% to 15% improvement		>25% to 40% improvement	7.5
Extramuscular Activity or Disease Activity Score >15% to 15% improvement 12.5 >15% to 25% improvement 15 >25% to 40% improvement 20 Improvement category Total improvement score † Minimal ≥ 20 DM/PM thresholds Moderate ≥ 40		>40% improvement	7.5
Extramuscular Activity or Disease Activity Score >15% to 25% improvement 15.5 >25% to 40% improvement 20 Total improvement score † Minimal ≥ 20 DM/PM thresholds Moderate ≥ 40	· · · · · · · · · · · · · · · · · · ·	Worsening to 5% improvement	0
Disease Activity Score >15% to 25% improvement >25% to 40% improvement 15 >40% improvement 20 Total improvement score† Minimal ≥ 20 DM/PM thresholds Moderate ≥ 40		>5% to 15% improvement	7.5
>25% to 40% improvement 15 >40% improvement 20 Improvement category $\frac{\text{Total improvement score}}{\text{score}^{\dagger}}$ DM/PM thresholds $\frac{1}{2}$ Moderate $\frac{1}{2}$ Moderate		>15% to 25% improvement	12.5
$\begin{array}{c ccccccccccccccccccccccccccccccccccc$		>25% to 40% improvement	15
$\begin{array}{c ccccccccccccccccccccccccccccccccccc$		>40% improvement	20
DM/PM thresholds Moderate ≥ 40		Improvement category	improvement
	DM/PM thresholds	Minimal	≥ 20
Maior > 60		Moderate	≥ 40
		Major	≥ 60

	Minimal	≥ 30
JDM thresholds	Moderate	≥ 45
	Major	≥ 70

Abbreviations: JDM, juvenile dermatomyositis; DM, dermatomyositis; PM, polymyositis; Enzyme, most abnormal serum muscle enzyme level among creatine kinase, aldolase, alanine aminotransferase, aspartate aminotransferase, and lactate dehydrogenase; CHQ-PhS, Physical Summary Score of the Child Health Questionnaire-Parent Form 50.

*Note that either all the IMACS or all the PRINTO core set measures may be used.

†The Total Improvement Score is the sum of all 6 improvement scores associated with the change in each core set measure. Note that the response criteria are also for use in combined adult DM/PM and JDM clinical trials.

How to calculate the Improvement Score:

The absolute percent change ([final value – baseline value] / range) X 100) is calculated for each core set measure. For muscle enzymes, the most abnormal enzyme at baseline is used. The enzyme range was calculated based on 90% range of enzymes from natural history data. For Adults, creatine kinase is 15 times the upper limit of normal, for aldolase is six times the upper limit of normal, and for lactate dehydrogenase, aspartate aminotransferase, and alanine aminotransferase is three times the upper limit of normal. For Juveniles, creatine kinase is 20 times the upper limit of normal, for aldolase is six times the upper limit of normal, and for lactate dehydrogenase, aspartate aminotransferase, and alanine aminotransferase is five times the upper limit of normal. The ranges for the other core set activity measures are based on the instrument scale used.

An Improvement Score is assigned for each core set measure based on the absolute percent change. These are totaled among the six IMACS or PRINTO core set measures. The thresholds for minimal, moderate, and major improvement are provided. The Total Improvement Scores may also be compared among treatment arms in a trial. A Total Improvement Score between 0 and 100 also corresponds to the degree of improvement, with higher scores corresponding to a greater degree of improvement.

From:

Aggarwal R, Rider LG, Ruperto N, Bayat N, Erman B, Feldman BM, Oddis CV, Amato AA, Chinoy H, Cooper RG, Dastmalchi M, Fiorentino D, Isenberg D, Katz JD, Mammen A, de Visser M, Ytterberg SR, Lundberg IE, Chung L, Danko K, García-De la Torre I, Song YW, Villa L, Rinaldi M, Rockette H, Lachenbruch PA, Miller FW, Vencovsky J; International Myositis Assessment and Clinical Studies Group and the Paediatric Rheumatology International Trials Organisation. 2016 American College of Rheumatology/European League Against Rheumatism Criteria for Minimal, Moderate, and Major Clinical Response in Adult Dermatomyositis and Polymyositis: An International Myositis Assessment and Clinical Studies

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Rider LG, Aggarwal R, Pistorio A, Bayat N, Erman B, Feldman BM, Huber AM, Cimaz R, Cuttica RJ, de Oliveira SK, Lindsley CB, Pilkington CA, Punaro M, Ravelli A, Reed AM, Rouster-Stevens K, van Royen-Kerkhof A, Dressler F, Magalhaes CS, Constantin T, Davidson JE, Magnusson B, Russo R, Villa L, Rinaldi M, Rockette H, Lachenbruch PA, Miller FW, Vencovsky J, Ruperto N; International Myositis Assessment and Clinical Studies Group and the Paediatric Rheumatology International Trials Organisation. 2016 American College of Rheumatology/European League Against Rheumatism Criteria for Minimal, Moderate, and Major Clinical Response in Juvenile Dermatomyositis: An International Myositis Assessment and Clinical Studies Group/Paediatric Rheumatology International Trials Organisation Collaborative Initiative. Arthritis Rheumatol. 2017 May;69(5):911-923. doi: 10.1002/art.40060. Epub 2017 Apr 6.

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Rider LG, Aggarwal R, Pistorio A, Bayat N, Erman B, Feldman BM, Huber AM, Cimaz R, Cuttica RJ, de Oliveira SK, Lindsley CB, Pilkington CA, Punaro M, Ravelli A, Reed AM, Rouster-Stevens K, van Royen-Kerkhof A, Dressler F, Saad Magalhaes C, Constantin T, Davidson JE, Magnusson B, Russo R, Villa L, Rinaldi M, Rockette H, Lachenbruch PA, Miller FW, Vencovsky J, Ruperto N; International Myositis Assessment and Clinical Studies Group and the Paediatric Rheumatology International Trials Organisation. 2016 American College of Rheumatology/European League Against Rheumatism Criteria for Minimal, Moderate, and Major Clinical Response in Juvenile Dermatomyositis: An International Myositis Assessment and Clinical Studies Group/Paediatric Rheumatology International Trials Organisation Collaborative Initiative. Ann Rheum Dis. 2017 May;76(5):782-791. doi: 10.1136/annrheumdis-2017-211401.