

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

Core Set Measure*	Level of Improvement Based on absolute percentage change	Improvement Score
Physician Global Activity	Worsening to 5% improvement	0
	>5% to 15% improvement	7.5
	>15% to 25% improvement	15
	>25% to 40% improvement	17.5
	>40% improvement	20
Patient or Parent Global Activity	Worsening to 5% improvement	0
	>5% to 15% improvement	2.5
	>15% to 25% improvement	5
	>25% to 40% improvement	7.5
	>40% improvement	10
Manual Muscle Testing or Childhood Myositis Assessment Scale	Worsening to 2% improvement	0
	>2% to 10% improvement	10
	>10% to 20% improvement	20
	>20% to 30% improvement	27.5
	>30% improvement	32.5
(Childhood) Health Assessment Questionnaire	Worsening to 5% improvement	0
	>5% to 15% improvement	5
	>15% to 25% improvement	7.5
	>25% to 40% improvement	7.5
	>40% improvement	10
Enzyme (most abnormal) or CHQ-PhS	Worsening to 5% improvement	0
	>5% to 15% improvement	2.5
	>15% to 25% improvement	5
	>25% to 40% improvement	7.5
	>40% improvement	7.5
Extramuscular Activity or Disease Activity Score	Worsening to 5% improvement	0
	>5% to 15% improvement	7.5
	>15% to 25% improvement	12.5
	>25% to 40% improvement	15
	>40% improvement	20
	<b>Improvement category</b>	<b>Total improvement score†</b>
DM/PM thresholds	Minimal	≥ 20
	Moderate	≥ 40
	Major	≥ 60

JDM thresholds	Minimal	≥ 30
	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 ( $[(\text{final value} - \text{baseline value}) / \text{range}] \times 100$ ) 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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