

PRINTO Trial (n=139)	24	0.0 (0.0 – 4.0)	0.0 [0.0 - 11.2]	7	4.0 (2.0 – 5.0)*	40.0 [32.5 – 40.0]*	22	5.0 (2.0 – 6.0)*	53.7 [47.5 – 60.0]* [†]	86	6.0 (5.0 – 6.0)* ^{†‡}	92.5 [87.5 - 97.5]* ^{†‡}
Juvenile Consensus Profiles (n=241)	59	1.0 (0.0 – 4.0)	7.5 [0.0 – 15.0]	27	4.0 (2.0 – 5.0)*	37.5 [32.5 – 40.0]*	50	5.0 (3.0 – 6.0)* [†]	55.0 [52.5 - 62.5]* [†]	105	6.0 (4.0 – 6.0)* ^{†‡}	90.0 [77.5 – 95.0]* ^{†‡}
All PRINTO JDM Data (n=380)	83	1.0 (0.0 – 4.0)	7.5 [0.0 – 15.0]	34	4.0 (2.0 – 5.0)*	37.5 [32.5 – 40.0]*	72	5.0 (2.0 – 6.0)* [†]	55.0 [50.0 - 61.2]* [†]	191	6.0 (4.0 – 6.0)* ^{†‡}	92.5 [82.5 - 97.5]* ^{†‡}

* Statistically significant difference (p-value < 0.006) from the No Improvement category

[†] Statistically significant difference (p-value < 0.006) from the Minimal Improvement category

[‡] Statistically significant difference (p-value < 0.006) from the Moderate Improvement category

[§] The Rituximab in Myositis Trial did not use PRINTO CSM.

Abbreviations: IMACS: xxx; PRINTO: xxx; CSM: Core Set Measure, TIS: Total Improvement Score, IQR: interquartile range, JDM: Juvenile Dermatomyositis.

Juvenile Consensus Profiles	-0.5 [(-13.0) – 11.0]	6.0 [(-2.0) – 24.0]	24.0 [8.0 – 38.0]* [†]	53.0 [34.0 – 69.0]** ^{††}
All IMACS JDM Datasets	0.0 [(-10.0) – 12.0]	10.0 [0.0 – 24.0]*	24.5 [10.0 – 38.0]* [†]	50.0 [37.0 – 68.0]** ^{††}

Manual Muscle Testing

Rituximab in Myositis Trial	3.0 [(-1.0) – 10.0]	5.5 [(-2.0) – 15.0]	6.0 [5.0 – 14.0]	21.5 [16.0 – 25.0]*
PRINTO Trial	0.0 [(-5.0) – 0.0]	5.0 [0.0 – 10.0]	20.0 [13.0 – 28.0]* [†]	43.0 [26.0 – 58.0]** ^{††}
Juvenile Consensus Profiles	-3.0 [(-9.0) – 1.0]	5.0 [3.0 – 10.0]*	16.0 [8.0 – 25.0]* [†]	40.0 [26.0 – 51.0]** ^{††}
All IMACS JDM Datasets	0.0 [(-8.0) – 3.0]	5.0 [0.5 – 10.0]*	16.0 [8.0 – 25.0]* [†]	39.0 [26.0 – 53.0]** ^{††}

Childhood Health Assessment Questionnaire

Rituximab in Myositis Trial	0.0 [(-4.0) – 4.0]	8.5 [(-4.0) – 21.0]	17.0 [8.0 – 21.0]*	21.0 [8.0 – 37.5]*
PRINTO Trial	0.0 [(-16.0) – 0.0]	13.0 [0.0 – 21.0]	29.0 [13.0 – 58.0]* [†]	63.0 [42.0 – 79.0]** ^{††}
Juvenile Consensus Profiles	0.0 [(-4.0) – 8.0]	8.0 [0.0 – 17.0]*	25.0 [10.0 – 33.0]* [†]	54.0 [33.0 – 75.0]** ^{††}
All IMACS JDM Datasets	0.0 [(-4.0) – 4.0]	8.0 [0.0 – 21.0]*	25.0 [8.0 – 38.0]* [†]	54.0 [33.0 – 75.0]** ^{††}

Extramuscular Global Disease Activity

Rituximab in Myositis Trial	-3.5 [(-13.0) – 1.0]	10.5 [5.0 – 14.5]*	19.5 [7.0 – 26.0]*	31.5 [27.5 – 36.5]* [†]
PRINTO Trial	0.0 [(-15.0) – 0.0]	10.0 [0.0 – 20.0]	6.0 [0.0 – 30.0]*	30.0 [10.0 – 50.0]*
Juvenile Consensus Profiles	-1.5 [(-12.0) – 2.0]	7.0 [0.0 – 18.0]*	9.0 [0.0 – 16.0]*	28.0 [10.0 – 46.0]** ^{††}
All IMACS JDM Datasets	(-1.0) [(-13.0) – 0.0]	9.5 [0.0 – 18.5]*	10.0 [0.0 – 25.0]*	30.0 [10.0 – 48.0]** ^{††}

Muscle enzyme

Rituximab in Myositis Trial	1.5 [(-3.0) – 6.0]	2.5 [0.0 – 7.0]	1.0 [(-1.0) – 5.0]	6.5 [(-1.0) – 13.0]
PRINTO Trial	0.0 [0.0 – 36.0]	27.0 [18.0 – 75.0]	18.0 [5.0 – 38.0]	84.5 [25.0 – 175.0]* [†]
Juvenile Consensus Profiles	6.0 [(-1.0) – 16.0]	8.0 [2.0 – 21.0]	13.0 [4.0 – 37.0]*	39.0 [17.0 – 85.0]** ^{††}
All IMACS JDM Datasets	4.5 [0.0 – 14.0]	11.0 [2.0 – 23.5]*	12.5 [3.0 – 31.0]*	45.0 [19.0 – 111.0]** ^{††}

PRINTO Core Set Measures	No Improvement	Minimal Improvement	Moderate Improvement	Major Improvement
	Median Absolute % Change [IQR]			

Physician Global Disease Activity

PRINTO Trial	0.0 [0.0 – 10.0]	15.0 [0.0 – 25.0]	29.5 [14.0 – 42.0]*	58.5 [40.0 – 70.0]** ^{††}
Juvenile Consensus Profiles	2.0 [(-15.0) – 9.0]	20.0 [14.0 – 28.0]*	25.5 [16.0 – 37.0]*	47.0 [33.0 – 61.0]** ^{††}
All PRINTO JDM Datasets	0.0 [(-11.0) – 9.0]	19.5 [13.0 – 27.0]*	26.0 [15.0 – 38.5]*	50.0 [38.0 – 68.0]** ^{††}

Parent Global Disease Activity

PRINTO Trial	0.0 [(-5.0) – 5.0]	2.0 [(-20.0) – 20.0]	20.5 [14.0 – 30.0]*	50.0 [35.0 – 65.0]** ^{††}
Juvenile Consensus Profiles	-2.0 [(-13.0) – 6.0]	5.0 [1.0 – 24.0]*	18.5 [4.0 – 32.0]*	50.0 [32.0 – 69.0]** ^{††}
All PRINTO JDM Datasets	0.0 [(-13.0) – 6.0]	4.0 [0.0 – 20.0]*	20.0 [8.5 – 30.5]*	50.0 [32.0 – 66.0]** ^{††}

Childhood Myositis Assessment Scale

PRINTO Trial	0.0 [(-3.0) – 0.0]	10.0 [8.0 – 19.0]*	15.0 [4.0 – 21.0]*	52.0 [33.0 – 67.0]** ^{††}
Juvenile Consensus Profiles	-2.0 [(-13.0) – 2.0]	8.0 [0.0 – 10.0]*	13.0 [10.0 – 21.0]* [†]	44.0 [29.0 – 58.0]** ^{††}
All PRINTO JDM Datasets	0.0 [(-12.0) – 0.0]	8.0 [4.0 – 10.0]*	14.0 [7.0 – 21.0]* [†]	46.0 [31.0 – 63.0]** ^{††}

Childhood Health Assessment Questionnaire

PRINTO Trial	0.0 [(-16.5) – 0.0]	13.0 [0.0 – 27.0]	19.0 [4.0 – 38.0]*	58.0 [42.0 – 75.0]** ^{††}
Juvenile Consensus Profiles	0.0 [(-8.0) – 0.0]	0.0 [0.0 – 13.0]	15.0 [4.0 – 29.0]* [†]	54.0 [33.0 – 71.0]** ^{††}

Juvenile Consensus Profiles	3.2 [0.7 - 5.2]	2.6 [0.9 - 4.4]	4.0 [2.5 - 6.1]	6.6 [5.0 - 8.4] [†]
All IMACS JDM Datasets	4.8 [1.5 - 5.9]	3.5 [1.6 - 5.5]	4.8 [2.7 - 6.6]	6.5 [5.0 - 8.0] ^{*††}
Manual Muscle Testing (0-80)				
Rituximab in Myositis Trial	62.0 [59.0 - 65.0]	63.5 [61.0 - 66.0]	64.0 [57.0 - 75.0]	60.0 [53.5 - 61.0]
PRINTO Trial	47.5 [43.0 - 58.0]	57.0 [49.0 - 61.0]	51.0 [44.0 - 61.0]	37.5 [25.0 - 52.0] ^{*††}
Juvenile Consensus Profiles	62.5 [50.0 - 74.0]	66.0 [54.0 - 73.0]	59.0 [48.0 - 66.0]	39.0 [26.0 - 50.0] ^{*††}
All IMACS JDM Datasets	61.0 [47.0 - 70.0]	63.5 [52.5 - 71.0]	57.0 [47.0 - 66.0] [†]	39.0 [26.0 - 54.0] ^{*††}
Childhood Health Assessment Questionnaire (0-3)				
Rituximab in Myositis Trial	0.9 [0.3 - 1.2]	1.0 [0.5 - 1.6]	1.1 [0.3 - 1.8]	1.4 [0.6 - 2.0]
PRINTO Trial	1.7 [1.3 - 2.5]	1.6 [0.6 - 2.5]	1.6 [0.7 - 2.2]	2.0 [1.5 - 2.6]
Juvenile Consensus Profiles	0.7 [0.2 - 1.7]	0.9 [0.2 - 1.4]	1.1 [0.7 - 1.7]	2.2 [1.5 - 2.6] ^{*††}
All IMACS JDM Datasets	1.0 [0.4 - 1.7]	1.1 [0.4 - 1.6]	1.3 [0.7 - 1.8]	2.1 [1.5 - 2.6] ^{*††}
Extramuscular Global Disease Activity (0-10)				
Rituximab in Myositis Trial	2.4 [1.4 - 3.5]	3.7 [2.2 - 5.2]	3.3 [1.8 - 5.0]	3.8 [3.2 - 5.1]
PRINTO Trial	3.0 [0.7 - 4.0]	3.0 [2.0 - 5.0]	1.8 [0.3 - 5.0]	4.0 [1.5 - 6.0]
Juvenile Consensus Profiles	0.6 [0.0 - 1.4]	1.3 [0.0 - 2.3]	1.7 [0.2 - 3.7]	3.5 [1.1 - 5.0] ^{*††}
All IMACS JDM Datasets	1.1 [0.1 - 3.0]	2.0 [0.4 - 3.5]	1.8 [0.4 - 4.2]	3.6 [1.4 - 5.0] ^{*††}
Muscle enzyme				
Rituximab in Myositis Trial	1.1 [0.9 - 2.1]	1.1 [0.9 - 1.3]	1.8 [1.0 - 2.5]	1.3 [1.1 - 2.3]
PRINTO Trial	4.8 [2.4 - 20.8]	6.2 [1.9 - 18.2]	3.2 [1.5 - 12.9]	13.3 [3.3 - 29.3] [†]
Juvenile Consensus Profiles	1.3 [0.9 - 2.6]	1.4 [1.1 - 2.0]	1.8 [1.2 - 4.5]	4.8 [2.0 - 14.4] ^{*††}
All IMACS JDM Datasets	1.6 [0.9 - 3.9]	1.4 [1.1 - 2.6]	2.0 [1.3 - 5.4]	5.6 [2.3 - 20.3] ^{*††}
PRINTO Core Set Measures (Range)	No Improvement	Minimal Improvement	Moderate Improvement	Major Improvement
	Median Baseline Value [IQR]			
Physician Global Disease Activity (0-10)				
PRINTO Trial	6.3 [4.0 - 7.5]	4.5 [4.0 - 6.0]	5.0 [4.0 - 7.0]	7.0 [6.0 - 8.0] [†]
Juvenile Consensus Profiles	3.0 [1.6 - 5.0]	2.8 [2.0 - 5.0]	4.1 [2.6 - 5.5]	5.8 [4.6 - 7.3] ^{*††}
All PRINTO JDM Datasets	4.0 [2.0 - 6.0]	3.1 [2.3 - 5.0]	4.8 [3.1 - 6.0]	6.2 [5.0 - 8.0] ^{*††}
Parent Global Disease Activity (0-10)				
PRINTO Trial	5.8 [5.0 - 7.0]	5.0 [3.5 - 6.0]	4.1 [2.1 - 5.0]	6.0 [5.0 - 8.0] [†]
Juvenile Consensus Profiles	3.0 [0.7 - 5.1]	1.2 [0.2 - 4.5]	3.7 [1.6 - 5.9]	6.2 [4.5 - 7.8] ^{*††}
All PRINTO JDM Datasets	4.2 [1.5 - 5.5]	2.4 [0.2 - 5.0]	3.8 [2.0 - 5.5]	6.0 [4.8 - 8.0] ^{*††}
Childhood Myositis Assessment Scale (0-52)				
PRINTO Trial	20.5 [10.5 - 29.5]	37.0 [19.0 - 39.0]	36.0 [25.0 - 43.0] [*]	16.0 [10.0 - 29.0] ^{††}
Juvenile Consensus Profiles	41.0 [28.0 - 48.0]	44.0 [36.0 - 48.0]	35.5 [27.0 - 42.0] [†]	21.0 [10.0 - 32.0] ^{*††}
All PRINTO JDM Datasets	38.0 [20.0 - 47.0]	41.0 [35.0 - 47.0]	36.0 [27.0 - 42.0] [†]	19.0 [10.0 - 30.0] ^{*††}
Childhood Health Assessment Questionnaire (0-3)				
PRINTO Trial	1.8 [1.3 - 2.5]	1.1 [0.1 - 1.6]	1.0 [0.5 - 2.2]	1.9 [1.5 - 2.6] ^{††}
Juvenile Consensus Profiles	0.7 [0.1 - 1.5]	0.5 [0.0 - 1.1]	1.2 [0.6 - 1.6] ^{*†}	2.1 [1.5 - 2.6] ^{*††}
All PRINTO JDM Datasets	1.0 [0.2 - 1.7]	0.5 [0.1 - 1.1]	1.1 [0.6 - 1.7] [†]	2.0 [1.5 - 2.6] ^{*††}
Disease Activity Score (0-20)				
PRINTO Trial	13.0 [11.5 - 14.5]	11.0 [11.0 - 15.0]	11.0 [10.0 - 13.0]	14.0 [12.0 - 15.0] [†]

Juvenile Consensus Profiles	0.0 [0.0 - 14.3]	5.9 [0.0 - 15.4]	11.1 [4.6 - 14.8]	10.5 [9.4 - 12.1]*
All IMACS JDM Datasets	10.0 [0.0 - 14.3]	7.1 [0.0 - 15.4]	11.1 [4.4 - 14.8]	10.5 [9.4 - 12.5]*
Manual Muscle Testing (Expected contribution 32.5%)				
Rituximab in Myositis Trial	44.4 [0.0 - 88.9]	24.3 [0.0 - 49.0]	20.5 [19.1 - 34.8]	31.0 [27.1 - 35.5]
PRINTO Trial	32.5 [0.0 - 32.5]	26.7 [0.0 - 30.8]	40.7 [29.6 - 45.8]	35.1 [32.5 - 39.4]*
Juvenile Consensus Profiles	0.0 [0.0 - 32.5]*	26.7 [23.5 - 33.3]*	38.1 [20.0 - 48.2]	35.1 [32.5 - 39.4]*
All IMACS JDM Datasets	0.0 [0.0 - 36.4]*	25.8 [0.0 - 32.1]*	37.2 [20.0 - 47.8]	35.1 [32.5 - 39.4]*
Childhood Health Assessment Questionnaire (Expected contribution 10%)				
Rituximab in Myositis Trial	0.0 [0.0 - 10.0]	7.1 [0.0 - 19.4]	12.3 [8.7 - 14.8]	10.0 [6.9 - 11.6]
PRINTO Trial	0.0 [0.0 - 10.0]*	12.5 [0.0 - 17.7]	14.8 [8.3 - 15.8]	10.8 [10.0 - 12.1]*
Juvenile Consensus Profiles	0.0 [0.0 - 18.2]	11.8 [0.0 - 23.1]	11.5 [9.5 - 13.6]*	10.5 [10.0 - 11.8]*
All IMACS JDM Datasets	0.0 [0.0 - 10.0]*	11.8 [0.0 - 19.4]	12.0 [9.1 - 14.8]*	10.7 [10.0 - 11.8]*
Extramuscular Global Disease Activity (Expected contribution 20%)				
Rituximab in Myositis Trial	0.0 [0.0 - 0.0]*	19.4 [8.8 - 27.2]	23.3 [15.0 - 26.3]	20.0 [19.4 - 21.1]
PRINTO Trial	20.0 [0.0 - 20.0]	20.0 [0.0 - 35.3]	11.1 [0.0 - 25.0]	15.8 [9.1 - 20.0]*
Juvenile Consensus Profiles	0.0 [0.0 - 20.0]*	18.8 [0.0 - 29.4]	11.1 [0.0 - 23.1]*	16.2 [9.4 - 20.7]*
All IMACS JDM Datasets	0.0 [0.0 - 20.0]*	18.8 [0.0 - 29.4]	11.8 [0.0 - 25.0]*	16.7 [9.4 - 20.5]*
Muscle Enzyme (Expected contribution 7.5%)				
Rituximab in Myositis Trial	0.0 [0.0 - 11.1]	0.0 [0.0 - 6.3]	0.0 [0.0 - 0.0]*	3.3 [0.0 - 5.2]
PRINTO Trial	7.5 [7.5 - 33.3]	17.7 [11.8 - 23.1]	7.7 [0.0 - 11.5]	8.1 [7.1 - 9.4]*
Juvenile Consensus Profiles	9.1 [0.0 - 42.9]	5.9 [0.0 - 11.8]	5.6 [0.0 - 11.1]	7.9 [5.6 - 9.1]
All IMACS JDM Datasets	7.5 [0.0 - 33.3]	6.3 [0.0 - 15.4]	4.7 [0.0 - 11.1]	7.9 [6.3 - 9.1]*
PRINTO Core Set Measures by Study (% Expected contribution)				
	No Improvement	Minimal Improvement	Moderate Improvement	Major Improvement
Physician Global Disease Activity (Expected contribution 20%)				
PRINTO Trial	20.0 [20.0 - 27.3]	23.1 [0.0 - 37.5]	30.4 [16.7 - 36.8]	20.5 [20.0 - 21.6]*
Juvenile Consensus Profiles	20.0 [0.0 - 50.0]	40.0 [20.0 - 46.2]	28.0 [22.2 - 33.3]*	21.1 [20.0 - 22.6]*
All PRINTO JDM Datasets	20.0 [0.0 - 37.5]	37.5 [18.8 - 43.8]	29.8 [19.4 - 34.1]*	20.7 [20.0 - 22.2]*
Parent Global Disease Activity (Expected contribution 10%)				
PRINTO Trial	10.0 [10.0 - 11.3]	0.0 [0.0 - 12.5]	9.3 [5.3 - 12.5]	10.0 [8.1 - 10.5]
Juvenile Consensus Profiles	10.0 [0.0 - 10.0]	0.0 [0.0 - 16.7]	8.5 [0.0 - 13.6]	10.0 [8.8 - 10.8]
All PRINTO JDM Datasets	10.0 [0.0 - 10.0]	0.0 [0.0 - 13.3]	8.9 [4.1 - 13.0]	10.0 [8.3 - 10.7]
Childhood Myositis Assessment Scale (Expected contribution 32.5%)				
PRINTO Trial	32.5 [0.0 - 32.5]	30.8 [25.0 - 64.7]	33.3 [15.4 - 42.3]	33.3 [32.5 - 35.1]*
Juvenile Consensus Profiles	32.5 [0.0 - 32.5]	25.0 [0.0 - 33.3]	33.3 [20.0 - 42.3]	34.2 [32.5 - 37.1]*
All PRINTO JDM Datasets	32.5 [0.0 - 32.5]*	25.8 [23.5 - 33.3]	33.3 [19.1 - 42.3]	34.2 [32.5 - 36.1]*
Childhood Health Assessment Questionnaire (Expected contribution 10%)				
PRINTO Trial	10.0 [0.0 - 10.0]	12.5 [0.0 - 23.1]	13.3 [0.0 - 16.7]	10.3 [10.0 - 10.8]*
Juvenile Consensus Profiles	0.0 [0.0 - 10.0]*	0.0 [0.0 - 16.7]	11.1 [0.0 - 13.6]	10.3 [10.0 - 11.1]*
All PRINTO JDM Datasets	0.0 [0.0 - 10.0]*	0.0 [0.0 - 17.7]	11.3 [0.0 - 14.3]	10.3 [10.0 - 10.8]*
Disease Activity Score (Expected contribution 20%)				
PRINTO Trial	20.0 [0.0 - 20.0]	0.0 [0.0 - 31.3]	22.0 [0.0 - 30.8]	20.5 [20.0 - 21.6]*
Juvenile Consensus Profiles	0.0 [0.0 - 20.0]*	25.0 [18.8 - 40.0]	23.8 [18.5 - 27.3]	20.0 [16.7 - 21.1]
All PRINTO JDM Datasets	20.0 [0.0 - 20.0]*	23.1 [0.0 - 35.3]	22.9 [15.8 - 27.3]	20.0 [17.7 - 21.1]
Child Health Questionnaire Parent Form EQ - Physical Summary Score (Expected contribution 7.5%)				

Physician Global Disease Activity	0.2 (0.04)	<0.0001	0.3 (0.05)	<0.0001	N/A	N/A	0.2 (0.05)	<0.0001
Parent Global Disease Activity	0.1 (0.03)	<0.0001	0.1 (0.04)	0.003	N/A	N/A	0.1 (0.03)	0.005
Childhood Myositis Assessment Scale	0.3 (0.04)	<0.0001	0.2 (0.06)	0.000	N/A	N/A	0.4 (0.05)	<0.0001
Childhood Health Assessment Questionnaire	0.1 (0.03)	0.006	0.1 (0.05)	0.03	N/A	N/A	0.1 (0.05)	0.1
Disease Activity Score	0.4 (0.04)	<0.0001	0.4 (0.07)	<0.0001	N/A	N/A	0.4 (0.05)	<0.0001
Child Health Questionnaire-Parent Form 50 - Physical Summary Score	0.2 (0.04)	<0.0001	0.3 (0.05)	<0.0001	N/A	N/A	0.2 (0.07)	0.005

*Absolute percentage change in CSM was used in each regression model.

Abbreviations: See Supplementary Table S1. SE: Standard Error.

(N=273)	15/68 (22.7)	35/68 (51.5)	2.0 (0.0 – 6.0)	10/55 (41.0)	4/55 (18.2)	1.0 (0.0 – 2.0)	15/61 (24.6)	1/61 (1.6)	0.0 (0.0 – 2.0)	7/107 (6.5)	0/107 (0.0)	0.0 (0.0 – 1.0)
All IMACS JDM Datasets (N=457)	27/102 (26.5)	47/102 (46.1)	1.0 (0.0 – 6.0)	26/60 (43.3)	7/60 (11.7)	1.0 (0.0 – 2.0)	25/106 (23.6)	5/106 (4.7)	0.0 (0.0 – 2.0)	12/189 (6.3)	0/189 (0.0)	0.0 (0.0 – 1.0)

B. Frequency and Degree of Worsening in CSM by MRC Categories

Core Set Measures by Study	Myositis Response Criteria Categories							
	No Improvement		Minimal Improvement		Moderate Improvement		Major Improvement	
	Number of subjects n/N (%)	Median abs% change of worsening [IQR]	Number of subjects n/N (%)	Median abs % change of worsening [IQR]	Number of subjects n/N (%)	Median abs % change of worsening [IQR]	Number of subjects n/N (%)	Median abs % change of worsening [IQR]

Physician Global Disease Activity

Rituximab in Myositis Trial	3/18 (16.7)	-9.0 [-13.0, -7.0]	0/8 (0.0)	N/A	1/14 (7.1)	-7.0 [-7.0, -7.0]	0/8 (0.0)	N/A
PRINTO Trial	4/18 (22.2)	-17.0 [-32.5, -12.5]	1/13 (7.7)	-20.0 [-20.0, -20.0]	1/31 (3.2)	-40.0 [-40.0, -40.0]	0/74 (0.0)	N/A
Juvenile Consensus Profiles	24/66 (36.4)	-22.0 [-31.5, -10.0]	0/39 (0.0)	N/A	0/61 (0.0)	N/A	0/107 (0.0)	N/A
All IMACS JDM Datasets	31/102 (30.4)	-20.0 [-30.0, -10.0]	1/60 (1.7)	-20.0 [-20.0, -20.0]	2/106 (1.9)	-23.5 [-40.0, -7.0]	0/189 (0.0)	N/A

Parent Global Disease Activity

Rituximab in Myositis Trial	3/18 (16.7)	-8.0 [-16.0, -7.0]	2/8 (25.0)	-18.5 [-25.0, -12.0]	1/14 (7.1)	-8.0 [-8.0, -8.0]	0/8 (0.0)	N/A
PRINTO Trial	5/18 (27.8)	-20.0 [-45.0, -14.0]	1/13 (7.7)	-52.0 [-52.0, -52.0]	2/31 (6.4)	-25.0 [-30.0, -20.0]	1/74 (1.3)	-59.0 [-59.0, -59.0]
Juvenile Consensus Profiles	24/66 (36.4)	-22.5 [-36.0, -11.5]	8/39 (20.5)	-17.5 [-31.0, -7.5]	6/61 (9.8)	-10.0 [-43.0, -8.0]	2/107 (1.9)	-10.0 [-14.0, -6.0]
All IMACS JDM Datasets	32/102 (31.4)	-18.5 [-36.0, -10.5]	11/60 (18.3)	-24.0 [-36.0, -8.0]	9/106 (8.5)	-10.0 [-30.0, -8.0]	3/189 (1.6)	-14.0 [-59.0, -6.0]

Manual Muscle Testing

Rituximab in Myositis Trial	3/18 (16.7)	-16.0 [-20.0, -8.0]	2/8 (25.0)	-6.0 [-8.0, -4.0]	0/14 (0.0)	N/A	0/8 (0.0)	N/A
PRINTO Trial	5/18 (27.8)	-20.0 [-33.0, -10.0]	1/13 (7.7)	-10.0 [-10.0, -10.0]	2/31 (6.4)	-29.0 [-43.0, -15.0]	0/74 (0.0)	N/A
Juvenile Consensus Profiles	37/66 (56.1)	-8.0 [-11.0, -5.0]	7/39 (17.9)	-30.0 [-54.0, -6.0]	3/61 (4.9)	-6.0 [-15.0, -3.0]	0/107 (0.0)	N/A
All IMACS JDM Datasets	45/102 (44.1)	-9.0 [-14.0, -5.0]	10/60 (16.7)	-9.0 [-34.0, -6.0]	5/106 (4.7)	-15.0 [-15.0, -6.0]	0/189 (0.0)	N/A

Childhood Health Assessment Questionnaire

Rituximab in Myositis Trial	4/18 (22.2)	-13.0 [-23.0, -10.5]	1/8 (12.5)	-33.0 [-33.0, -33.0]	0/14 (0.0)	N/A	0/8 (0.0)	N/A
PRINTO Trial	5/18 (27.8)	-25.0 [-29.0, -17.0]	3/13 (23.1)	-17.0 [-67.0, -8.0]	2/31 (6.4)	-17.0 [-21.0, -13.0]	0/74 (0.0)	N/A
Juvenile Consensus Profiles	16/66 (24.2)	-21.0 [-29.0, -14.5]	4/39 (10.3)	-16.5 [-23.0, -10.0]	2/61 (3.3)	-18.5 [-29.0, -8.0]	1/107 (0.9)	-13.0 [-13.0, -13.0]
All IMACS JDM Datasets	25/102 (24.5)	-21.0 [-29.0, -13.0]	8/60 (13.3)	-19.0 [-29.0, -10.0]	4/106 (3.8)	-17.0 [-25.0, -10.5]	1/189 (0.5)	-13.0 [-13.0, -13.0]

Extramuscular Global Disease Activity

Rituximab in Myositis Trial	7/18 (38.9)	-15.0 [-20.0, -11.0]	0/8 (0.0)	N/A	0/14 (0.0)	N/A	0/8 (0.0)	N/A
PRINTO Trial	5/18 (27.8)	-28.0 [-30.0, -20.0]	3/13 (23.1)	-15.0 [-40.0, -10.0]	3/31 (9.7)	-20.0 [-40.0, -7.0]	4/74 (5.4)	-20.0 [-30.0, -19.5]
Juvenile Consensus Profiles	25/66 (37.9)	-25.0 [-33.0, -11.0]	4/39 (10.3)	-9.5 [-13.0, -6.5]	4/61 (6.6)	-28.0 [-50.0, -13.5]	1/107 (0.9)	-18.0 [-18.0, -18.0]
All IMACS JDM Datasets	37/102 (36.3)	-20.0 [-33.0, -12.0]	7/60 (11.7)	-12.0 [-15.0, -7.0]	7/106 (6.6)	-20.0 [-40.0, -10.0]	5/189 (2.6)	-20.0 [-20.0, -19.0]

Muscle Enzyme

Rituximab in Myositis Trial	4/18 (22.2)	-7.0 [-9.5, -6.0]	1/8 (12.5)	-11.0 [-11.0, -11.0]	3/14 (21.4)	-11.0 [-23.0, -9.0]	0/8 (0.0)	N/A
PRINTO Trial	1/18 (5.6)	-253.0 [-253.0, -253.0]	1/13 (7.7)	-15.0 [-15.0, -15.0]	3/31 (9.7)	-9.0 [-16.0, -7.0]	0/74 (0.0)	N/A

Core Set Measures by Study	Myositis Response Criteria Categories							
	No Improvement		Minimal Improvement		Moderate Improvement		Major Improvement	
	Number of subjects n/N (%)	Median abs% change of worsening [IQR]	Number of subjects n/N (%)	Median abs % change of worsening [IQR]	Number of subjects n/N (%)	Median abs % change ² of worsening [IQR]	Number of subjects n/N (%)	Median abs % change of worsening [IQR]
Physician Global Disease Activity								
PRINTO Trial	5/24 (20.8)	-20.0 [-20.0, -14.0]	1/7 (14.3)	-40.0 [-40.0, -40.0]	0/22 (0.0)	N/A	0/86 (0.0)	N/A
Juvenile Consensus Profiles	21/59 (35.6)	-23.0 [-33.0, -11.0]	0/27 (0.0)	N/A	0/50 (0.0)	N/A	0/105 (0.0)	N/A
All PRINTO JDM Datasets	26/83 (31.3)	-22.0 [-33.0, -11.0]	1/34 (2.9)	-40.0 [-40.0, -40.0]	0/72 (0.0)	N/A	0/191 (0.0)	N/A
Parent Global Disease Activity								
PRINTO Trial	6/24 (25.0)	-32.5 [-52.0, -14.0]	2/7 (28.6)	-25.0 [-30.0, -20.0]	0/22 (0.0)	N/A	1/86 (1.2)	-59.0 [-59.0, -59.0]
Juvenile Consensus Profiles	24/59 (40.7)	-20.0 [-30.5, -11.0]	3/27 (11.1)	-8.0 [-9.0, -8.0]	7/50 (14.0)	-24.0 [-56.0, -10.0]	1/105 (0.9)	-7.0 [-7.0, -7.0]
All PRINTO JDM Datasets	30/83 (36.1)	-21.0 [-41.0, -11.0]	5/34 (14.7)	-9.0 [-20.0, -8.0]	7/72 (9.7)	-24.0 [-56.0, -10.0]	2/191 (1.1)	-33.0 [-59.0, -7.0]
Childhood Myositis Assessment Scale								
PRINTO Trial	6/24 (25.0)	-14.0 [-25.0, -12.0]	0/7 (0.0)	N/A	2/22 (9.0)	-7.0 [-8.0, -6.0]	0/86 (0.0)	N/A
Juvenile Consensus Profiles	24/59 (40.7)	-13.0 [-15.0, -9.0]	4/27 (14.8)	-12.5 [-23.0, -7.0]	1/50 (2.0)	-13.0 [-13.0, -13.0]	0/105 (0.0)	N/A
All PRINTO JDM Datasets	30/83 (36.1)	-13.0 [-15.0, -10.0]	4/34 (11.8)	-12.5 [-23.0, -7.0]	3/72 (4.2)	-8.0 [-13.0, -6.0]	0/191 (0.0)	N/A
Childhood Health Assessment Questionnaire								
PRINTO Trial	8/24 (33.3)	-21.0 [-41.5, -16.5]	1/7 (14.3)	-13.0 [-13.0, -13.0]	1/22 (4.5)	-21.0 [-21.0, -21.0]	0/86 (0.0)	N/A
Juvenile Consensus Profiles	17/59 (28.8)	-21.0 [-29.0, -16.0]	3/27 (11.1)	-21.0 [-25.0, -13.0]	4/50 (8.0)	-10.0 [-20.5, -8.0]	0/105 (0.0)	N/A
All PRINTO JDM Datasets	25/83 (30.1)	-21.0 [-29.0, -16.0]	4/34 (11.8)	-17.0 [-23.0, -13.0]	5/72 (6.9)	-12.0 [-21.0, -8.0]	0/191 (0.0)	N/A
Disease Activity Score								
PRINTO Trial	4/24 (16.7)	-15.0 [-20.0, -10.0]	0/7 (0.0)	N/A	1/22 (4.5)	-15.0 [-15.0, -15.0]	0/86 (0.0)	N/A
Juvenile Consensus Profiles	17/59 (28.8)	-20.0 [-25.0, -10.0]	0/27 (0.0)	N/A	2/50 (4.0)	-30.0 [-35.0, -25.0]	1/105 (0.9)	-10.0 [-10.0, -10.0]
All PRINTO JDM Datasets	21/83 (25.3)	-20.0 [-25.0, -10.0]	0/34 (0.0)	N/A	3/72 (4.2)	-25.0 [-35.0, -15.0]	1/191 (0.5)	-10.0 [-10.0, -10.0]
Child Health Questionnaire-Parent Form 50 - Physical Summary Score								
PRINTO Trial	1/24 (4.2)	-10.0 [-10.0, -10.0]	2/7 (28.6)	-11.0 [-13.0, -9.0]	1/22 (4.5)	-18.0 [-18.0, -18.0]	2/86 (2.3)	-24.5 [-34.0, -15.0]
Juvenile Consensus Profiles	17/59 (28.8)	-15.0 [-19.0, -9.0]	4/27 (14.8)	-12.5 [-19.0, -10.5]	6/50 (12.0)	-13.5 [-19.0, -8.0]	1/105 (0.9)	-13.0 [-13.0, -13.0]
All PRINTO JDM Datasets	18/83 (21.7)	-14.5 [-19.0, -9.0]	6/34 (17.6)	-12.0 [-14.0, -10.0]	7/72 (9.7)	-16.0 [-19.0, -8.0]	3/191 (1.6)	-15.0 [-34.0, -13.0]

Abbreviations: JDM: Juvenile Dermatomyositis, N/A: Not Available.

All IMACS JDM Datasets	53/60 (88.3%)	106/106 (100.0%)*	189/189 (100.0%)*
No Contribution from any Muscle Related Measure			
PRINTO Trial	0/13 (0.0%)	0/31 (0.0%)	0/74 (0.0%)
Rituximab in Myositis Trial	2/8 (25.0%)	0/14 (0.0%)	0/8 (0.0%)
Juvenile Consensus Profiles	5/39 (12.8%)	0/61 (0.0%)*	0/107 (0.0%)*
All IMACS JDM Datasets	7/60 (11.7%)	0/106 (0.0%)*	0/189 (0.0%)*

PRINTO Core Set Measures

Childhood Myositis Assessment Scale (CMAS) Contributing to TIS			
PRINTO Trial	7/7 (100.0%)	17/22 (77.2%)	86/86 (100.0%) [†]
Juvenile Consensus Profiles	19/27 (70.4%)	48/50 (96.0%)*	105/105 (100.0%)*
All PRINTO JDM Datasets	26/34 (76.5%)	65/72 (90.3%)	191/191 (100.0%)* [†]

CMAS, Childhood Health Assessment Questionnaire, or Child Health Questionnaire-Parent Form 50 - Physical Summary Score Contributing to TIS			
PRINTO Trial	7/7 (100.0%)	22/22 (100.0%)	86/86 (100.0%)
Juvenile Consensus Profiles	22/27 (81.5%)	50/50 (100.0%)*	105/105 (100.0%)*
All PRINTO JDM Datasets	29/34 (85.3%)	72/72 (100.0%)*	191/191 (100.0%)*

No Contribution from CMAS, Childhood Health Assessment Questionnaire, or Child Health Questionnaire-Parent Form 50 - Physical Summary Score Contributing to TIS			
PRINTO Trial	0/7 (0.0%)	0/22 (0.0%)	0/86 (0.0%)
Juvenile Consensus Profiles	5/27 (18.5%)	0/50 (0.0%)*	0/105 (0.0%)*
All PRINTO JDM Datasets	5/34 (14.7%)	0/72 (0.0%)*	0/191 (0.0%)*

Frequency of Patient Reported Outcome Measure Contribution to TIS

IMACS Core Set Measures			
Parent Global Disease Activity Contributing to TIS			
PRINTO Trial	10/13 (76.9%)	26/31 (83.8%)	73/74 (98.6%)* [†]
Rituximab in Myositis Trial	5/8 (62.5%)	13/14 (92.8%)	8/8 (100.0%)
Juvenile Consensus Profiles	20/39 (51.3%)	48/61 (78.6%)*	103/107 (96.2%)* [†]
All JDM datasets	35/60 (58.3%)	87/106 (82.0%)*	184/189 (97.3%)* [†]

Childhood Health Assessment Questionnaire Contributing to TIS			
PRINTO Trial	7/13 (53.8%)	26/31 (83.8%)	72/74 (97.3%)*
Rituximab in Myositis Trial	4/8 (50.0%)	11/14 (78.5%)	7/8 (87.5%)
Juvenile Consensus Profiles	20/39 (51.3%)	50/61 (81.9%)*	102/107 (95.3%)* [†]
All JDM datasets	31/60 (51.7%)	87/106 (82.0%)*	181/189 (95.7%)* [†]

Parent Global Disease Activity or Childhood Health Assessment Questionnaire Contributing to TIS			
PRINTO Trial	11/13 (84.6%)	29/31 (93.5%)	74/74 (100.0%)
Rituximab in Myositis Trial	5/8 (62.5%)	14/14 (100.0%)	8/8 (100.0%)
Juvenile Consensus Profiles	30/39 (76.9%)	54/61 (88.5%)	106/107 (99.0%)* [†]
All JDM datasets	46/60 (76.7%)	97/106 (91.5%)*	188/189 (99.4%)* [†]

PRINTO Core Set Measures

Parent Global Disease Activity Contributing to TIS			
PRINTO Trial	3/7 (42.8%)	20/22 (90.9%)	83/86 (96.5%)*
Juvenile Consensus Profiles	13/27 (48.2%)	37/50 (74.0%)	101/105 (96.1%)* [†]
All JDM datasets	16/34 (47.1%)	57/72 (78.2%)*	184/191 (96.3%)* [†]

Childhood Health Assessment Questionnaire Contributing to TIS

Abbreviations: TIS: Total Improvement Score, JDM: Juvenile Dermatomyositis, CHQ-PF50 PhS: Child Health Questionnaire-Parent Form 50 - Physical Summary Score. Spearman correlation coefficients shown.