Rheumatology 2009;48:134–139
Advance Access publication 11 December 2008
doi:10.1093/rheumatology/ken441

Distribution and severity of weakness among patients with polymyositis, dermatomyositis and juvenile dermatomyositis

M. O. Harris-Love[1,2,3], J. A. Shrader[3], D. Koziol[4], N. Pahlajani[5], M. Jain[3], M. Smith[3], H. L. Cintas[3], C. L. McGarvey[3], L. James-Newton[2], A. Pokrovnichka[5], B. Moini[5], I. Cabalar[5], D. J. Lovell[6], R. Wesley[4], P. H. Plotz[5], F. W. Miller[2], J. E. Hicks[3] and L. G. Rider[2]

Objective. To describe the distribution and severity of muscle weakness using manual muscle testing (MMT) in 172 patients with PM, DM and juvenile DM (JDM). The secondary objectives included characterizing individual muscle group weakness and determining associations of weakness with functional status and myositis characteristics in this large cohort of patients with myositis.
Methods. Strength was assessed for 13 muscle groups using the 10-point MMT and expressed as a total score, subscores based on functional and anatomical regions, and grades for individual muscle groups. Patient characteristics and secondary outcomes, such as clinical course, muscle enzymes, corticosteroid dosage and functional status were evaluated for association with strength using univariate and multivariate analyses.
Results. A gradient of proximal weakness was seen, with PM weakest, DM intermediate and JDM strongest among the three myositis clinical groups ($P \leq 0.05$). Hip flexors, hip extensors, hip abductors, neck flexors and shoulder abductors were the muscle groups with the greatest weakness among all three clinical groups. Muscle groups were affected symmetrically.
Conclusions. Axial and proximal muscle impairment was reflected in the five weakest muscles shared by our cohort of myositis patients. However, differences in the pattern of weakness were observed among all three clinical groups. Our findings suggest a greater severity of proximal weakness in PM in comparison with DM.

Key words: Myositis, Manual muscle test, Strength, Rehabilitation.

Introduction

The idiopathic inflammatory myopathies (IIMs) result in chronic skeletal muscle inflammation and weakness [1]. PM, DM, juvenile DM (JDM) and IBM are the major subtypes of IIM, differentiated by clinical, histopathological and immunological features [1, 2]. Therapy consists of anti-inflammatory and immunosuppressive agents [2].

The impaired force-generating capacity of skeletal muscle secondary to chronic muscle inflammation and residual muscle atrophy and scarring in IIM contribute to functional limitation, disability and decreased health-related quality of life [3, 4]. Peak muscle force is most relevant in the assessment of IIM disease activity and treatment responses [5], most commonly measured by the manual muscle test (MMT). The International Myositis Assessment and Clinical Studies Group (IMACS) and Pediatric Clinical Trials Organization (PRINTO) identified muscle strength assessment as a top core set measure, along with physician global disease activity in myositis patients [5, 6].

Most myositis therapeutic trials conducted over the past two decades incorporated the MMT as a primary outcome measure [5, 7], but differed widely in its use. Trials featured summed MMT scores derived from 4 to 20 muscle groups [8] and have incorporated different MMT grading scales, including the Medical Research Council 5-point scale and an expanded 10-point scale [8]. In addition, previous IIM natural history and clinical outcome studies have not characterized weakness of individual muscle groups [9–26].

The primary aim of this investigation is to describe the distribution and severity of muscle weakness measured with MMT in 172 patients with PM, DM and JDM enrolled in natural history studies. Secondary aims include characterizing individual muscle group weakness and determining associations of muscle weakness with other myositis illness characteristics in this large cohort of patients with myositis.

Patients and methods

Patients

We utilized a cross-sectional, retrospective design with a 172 patient sample of probable or definite PM, DM or JDM confirmed by Bohan and Peter criteria of IIM patients seen at the National Institutes of Health (NIH) Clinical Center from 1994 to 2005 [27]. Included subjects were $\geq$5 yrs of age, and examined by an adult or paediatric rheumatologist and a physical therapist, receiving a standardized MMT evaluation. Sixty-five had PM, 50 DM and 57 JDM. Six with PM, 6 with DM and 3 with JDM were diagnosed with myositis and another autoimmune disease (systemic lupus, scleroderma, SS or JRA). Of those with JDM, four patients were 4.6–6 yrs of age and five participants were between 7 and 8 yrs of age; five were >18 yrs of age, with a maximum age of 34.9 yrs. Patients and parents of juvenile patients provided written informed consent and were enrolled in NIH institutional review board approved studies of myositis natural history. Only patients with a complete MMT by a physical therapist were eligible.

Manual muscle testing

Physical therapists experienced in examining IIM patients administered the MMT using standardized instructions, positioning and stabilization techniques established by Kendall *et al*. [28].

[1]George Washington University, School of Medicine and Health Sciences, Washington DC, [2]Environmental Autoimmunity Group, Office of Clinical Research, National Institute of Environmental Health Sciences, [3]Rehabilitation Medicine Department (RMD), [4]Biostatistics and Clinical Epidemiology Service, Clinical Center, [5]National Institute of Arthritis and Musculoskeletal and Skin Diseases, National Institutes of Health (NIH), Department of Health and Human Services, Bethesda, MD and [6]Division of Rheumatology, Cincinnati Children's Hospital Medical Center, Cincinnati, OH, USA.

Submitted 30 April 2008; revised version accepted 22 October 2008.

Correspondence to: L. G. Rider, Environmental Autoimmunity Group, NIEHS, NIH; CRC 4-2352, MSC 1301, 10 Center Drive, Bethesda, MD 20892-1301, USA. E-mail: riderl@mail.nih.gov

Examiners attended a MMT workshop by Florence Kendall, or viewed seminar videotapes and received instruction from a Kendall-trained physical therapist, prior to assessing patients in the study. The total MMT score consisted of 13 muscle groups (2 unilateral and 11 bilateral), with a maximum (max) value of 240. The 13 muscle groups included in the MMT examination represent the common muscles tested among all clinical groups included in our cohort (see Supplementary Table 1, available as supplementary data at *Rheumatology* Online). Our inclusion of these 13 muscle groups reflects the variation of end-point measures in previous studies [8], and attempts to comprehensively describe the clinical presentation of the participants.

The MMT score was organized into regional anatomic groups: axial, proximal and distal. In addition, the MMT score was organized into functional anatomic groups: upper extremity and lower extremity (see Supplementary Table 1, available as supplementary data at *Rheumatology* Online). The Kendall 10-point MMT was selected because inter-rater reliability was established in myositis [28] and this MMT grading scale, as well as variations, has been used as a primary outcome measure in IIM therapeutic trials [29–32]. Pediatric physical therapists administered the MMT examination to all children with JDM. These therapists were instructed to contact the primary investigator or make a notation in the medical record if subject cooperation or behavioural factors confounded the MMT results.

We classified the degree of weakness for individual muscles into four distinct strata: severe, moderate, mild and none. For each muscle group, MMT was graded 0–3 (severe weakness), 4–6 (moderate weakness), 7–9 (mild weakness) and 10 (no detectable weakness). We implemented this system based on movement limitations derived from MMT grading criteria to stratify muscle weakness based on the patient's ability to move a body segment against gravity and withstand examiner-applied manual resistance. Grades 7–9 indicate the ability to move against gravity into the testing position and hold against varying degrees of applied manual resistance. Furthermore, Grades 7–9 should be discernable from Grade 10, which indicates no detectable strength impairment. Grades 4–6 indicate considerable difficulty in placing a limb in the testing position against gravity and sustaining the testing position against gravity or minimal manual resistance. Grades 0–3 primarily reflect the inability to move the limb beyond partial range of motion (<50% of passive range of motion) against gravity and the need for gravity-minimized testing techniques.

Secondary outcomes

Demographic and clinical data included age, BMI, serum levels of creatine kinase (CK; upper limit of normal value, 252 U/l) and lactate dehydrogenase (LDH; upper limit of normal value, 226 U/l), corticosteroid dosage (mg/kg of body weight) and physician global assessment of disease activity (MD Global Activity) using a 10-cm visual analogue scale [33]. MD Global Activity was completed for 35 of 65 patients with PM, 18 of 50 patients with DM and 55 of 57 patients with JDM. Age at onset was based on the first occurrence of IIM symptoms. Delay to diagnosis (months) was calculated from the onset date to the date of IIM diagnosis. Disease duration (months) was calculated from diagnosis date to examination date at our facility. Functional status was assessed with the modified Convery activities of daily living (ADL) scale [34] derived from the Convery assessment [35], administered to 30 of 65 patients with PM and 15 of 50 patients with DM by a single physiatrist. The Childhood Myositis Assessment Scale (CMAS) was used to observe muscle function and endurance in 53 of 57 JDM patients [36]. Forty-two parents of 57 patients with JDM who were <18 yrs of age completed the Childhood Health Assessment Questionnaire (CHAQ) to assess physical dysfunction [37].

Statistical analysis

Data were analysed using JMP version 6.0 statistical analysis software (SAS Institute, Cary, NC, USA), StatXact version 4.0.1 (Cytel Software Corporation, Cambridge, MA, USA) and Stata Statistical Software, Release 10 (StataCorp, College Station, TX, USA). Analysis of variance with Tukey–Kramer *post hoc* tests was used to determine differences among the groups regarding interval data for patient characteristics, clinical course and summed MMT scores. Chi-square or exact tests were used for categorical data.

Because MMT scores were ordinal, they were summarized by median and interquartile range for each individual muscle. MMT total score and MMT subscores based on functional and anatomical regions, calculated from bilateral summed data, were summarized as means and standard deviations. Parametric statistics were used for these summed MMT scores since they approximated interval data [38]. Summed MMT data were also expressed as a percentage of the maximum possible score to allow for comparisons among MMT regional subscores. Because paired analyses did not reveal differences between right and left individual MMT grades across all groups, right-sided data were arbitrarily selected for the analysis and depiction of the MMT grades for individual muscle groups. The Kruskal–Wallis test was used to determine differences in individual MMT grades among the three clinical groups; if significant, the Wilcoxon rank sum test was used for *post hoc* pairwise comparisons, using Holm's adjustment. To compare subscores of different muscle groups (e.g. proximal *vs* distal and upper *vs* lower), for each patient a difference between the two subscore groups was calculated using percentages of maximum scores, and tested for equality to zero using the one-sample *t*-test. For comparing ordered categorical data among the three clinical groups, the Kruskal–Wallis nonparametric test for singly ordered contingency tables was used.

Univariate and multivariate linear regression models were used to determine variables associated with muscle strength. An *a priori* decision was made to use the proximal MMT score as the dependent variable based on the prominent degree of proximal weakness. The coefficient of determination (R^2) was used to estimate the strength of the regression model. The predictor variables for the regression models were: age, BMI, CK and LDH serum levels, corticosteroid dosage, disease duration, physician global assessment of disease activity, and CMAS and CHAQ for the JDM group and modified Convery ADL scale for PM and DM groups. For all analyses, a two-tailed *P*-value of <0.05 was considered significant.

Results

Patient characteristics and clinical course

A comparison of patient demographics and clinical characteristics by clinical subgroup is presented in Table 1. No significant differences between patients with PM or DM were detected for age, gender, ethnicity, age of onset, delay in diagnosis, corticosteroid dosage and serum CK and LDH levels. Disease duration was significantly greater for PM compared with DM patient groups ($P \leq 0.05$). The female to male ratio exceeded 3 : 1 for both PM and DM groups. JDM patients had younger age at onset, lower BMI, higher corticosteroid dosages (mg/kg) and lower CK levels than the PM or DM groups ($P \leq 0.05$).

Weakness among the IIM clinical groups

The total MMT score was 180.3 ± 27.3 (75% potential max total score of 240) in PM, 191.8 ± 22.6 (80% of the potential max total score) in DM and 202.0 ± 29.9 (84% of the potential max total score) in JDM (Table 2). Patients with PM were weaker than JDM based on the total MMT score ($P < 0.05$). Using subgroups of muscles based on anatomical and functional regions

(see Supplementary Table 1, available as supplementary data at *Rheumatology* Online), patients with PM were significantly weaker than the DM and JDM patient groups when comparing the lower extremity and proximal MMT scores ($P \leq 0.05$; Table 2). Upper extremity and distal MMT subscores (85–87% and 89% of max potential score, respectively) were relatively higher than the axial MMT subscores for all clinical groups (77% of max potential score). Paired analyses revealed that proximal region muscle groups were weaker than the distal region, and lower extremity groups were weaker than upper extremity groups within each IIM clinical group ($P \leq 0.001$; Table 2).

TABLE 1. Comparison of subject characteristics by clinical group

	PM ($n=65$)	DM ($n=50$)	JDM ($n=57$)
Age, yrs	46.3 ± 11.8[a]	42.9 ± 12.6[a]	12.8 ± 5.6[a]
Gender, female, %	80.0	78.0	70.2
BMI, kg/m^2	28.5 ± 6.8[a]	28.1 ± 6.6[a]	21.3 ± 4.5[a]
Ethnicity			
Caucasian, %	67.7	66.0	75.4
African American, %	23.1	24.0	8.8
Hispanic, %	4.6	4.0	1.8
Other, %	4.6	6.0	14.0
Age of onset, yrs	41.7 ± 12.1[a]	40.7 ± 13.3[a]	8.2 ± 4.3[a]
Disease duration, months	55.3 ± 60.0[c]	30.3 ± 24.2[c]	41.7 ± 61.2
Delay in diagnosis, months	7.4 ± 10.6	10.4 ± 14.8	14.0 ± 23.8
Corticosteroid dosage, mg/kg/day	0.27 ± 0.39[a]	0.26 ± 0.22[a]	0.75 ± 1.01[a]
LDH, U/l	383 ± 189[b]	342 ± 226	251 ± 139[b]
CK, U/l	1579 ± 1766[a]	1095 ± 2335[a]	200 ± 416[a]

Non-percentage values are expressed as means ± S.D. [a]JDM significantly different from PM or DM at the ≤0.05 level by Tukey–Kramer Honestly Significant Difference. [b]JDM significantly different from PM at the ≤0.05 level by Tukey–Kramer Honestly Significant Difference. [c]PM significantly different from DM at the ≤0.05 level by Tukey–Kramer Honestly Significant Difference. (Percentage values in columns may not equal 100% due to rounding.).

Individual muscle group weakness

We conducted pairwise comparisons of the right and left MMT grades of each bilateral muscle group. No significant asymmetry of weakness was detected in the PM, DM or JDM clinical groups ($P = 0.12$–1.0). The PM group displayed more weakness in four muscle groups in comparison with DM. These muscle groups were the hip flexors, hip abductors, knee extensors and ankle plantar flexors (hip abductors, $P \leq 0.01$; all other groups, $P \leq 0.05$; Table 3).

In DM, the wrist flexors were significantly weaker than in both PM and JDM ($P \leq 0.05$). Over twice the number of patients with DM had a MMT grade of ≤6 for wrist flexors compared with patients with PM and JDM (Table 4).

Patients with JDM had the least overall weakness among the three clinical groups. At least 88% of the patients with JDM had MMT grades for neck extensors, shoulder elevators, elbow flexors, wrist flexors, wrist extensors, knee extensors, ankle dorsiflexors and plantar flexors that were in the 'mild' or 'none' strata for weakness (Table 4). Four muscle groups displayed appreciable weakness in our JDM cohort (median grade < 9 out of 10): neck flexors, shoulder abductors, hip flexors and hip extensors (Table 3). In each of these muscle groups, at least 20% of the patients had MMT grades in the 'moderate' to 'severe' strata of weakness (Table 4).

The five weakest muscle groups for all three IIM clinical groups were the hip flexors, hip extensors, hip abductors, neck flexors and shoulder abductors (Tables 3 and 4). The severity of weakness among these muscle groups varied within each clinical group. Neck flexors, the weakest muscle group in JDM, demonstrated moderate to severe weakness (MMT grade ≤6 out of 10) in 49% of the patients (Table 4). Neck flexors were only the third and fourth weakest muscle group for DM and PM. Muscle groups

TABLE 2. Comparison of MMT subscores and total MMT score by clinical group

MMT subscore	PM ($n=65$)	DM ($n=50$)	JDM ($n=57$)	Maximum possible MMT score
Proximal[a], *n* (%)	94.1 ± 20.8[b] (67.2 ± 14.9)	105.5 ± 16.4[b] (75.4 ± 11.7)	115.5 ± 19.9[b] (82.5 ± 14.2)	140
Distal[a], *n* (%)	70.8 ± 8.0 (88.5 ± 10.0)	70.8 ± 6.8 (88.5 ± 8.6)	71.0 ± 9.3 (88.8 ± 11.6)	80
Axial, *n* (%)	15.4 ± 3.4 (77.1 ± 17.1)	15.4 ± 2.9 (77.2 ± 14.7)	15.5 ± 3.2 (77.5 ± 16.2)	20
Upper extremity[c], *n* (%)	85.4 ± 11.0 (85.4 ± 11.0)	85.0 ± 10.1 (85.0 ± 10.1)	86.7 ± 11.5 (86.7 ± 11.5)	100
Lower extremity[c], *n* (%)	79.5 ± 17.8[b] (66.2 ± 14.8)	91.3 ± 13.5[b] (76.1 ± 11.3)	99.8 ± 16.8[b] (83.3 ± 14.0)	120
Total MMT, *n* (%)	180.3 ± 27.3[d] (75.2 ± 11.4)	191.8 ± 22.6 (79.9 ± 9.4)	202.0 ± 29.9[b] (84.2 ± 12.5)	240

Values are expressed as means ± S.D. (percentage of maximum possible MMT score ± S.D.). [a]Difference between distal subscore and proximal subscore is significantly different within each clinical group, $P \leq 0.001$, using one-sample *t*-test. [b]Each clinical group is significantly different from the other two groups at the $P \leq 0.05$ level by Tukey–Kramer Honestly Significant Difference. [c]Difference between upper and lower extremity subscore is significantly different within each clinical group, $P \leq 0.001$, using one-sample *t*-test. [d]PM is significantly different from JDM at the $P \leq 0.05$ level by Tukey–Kramer Honestly Significant Difference. All comparisons are based on the percentage of the maximum achievable subscore.

TABLE 3. Comparison of individual MMT grades by clinical group

	PM ($n=65$)		DM ($n=50$)		JDM ($n=57$)	
Muscle groups	Median	[25%, 75%]	Median	[25%, 75%]	Median	[25%, 75%]
Neck flexors	7.0	[6.0, 9.0]	7.0	[5.0, 8.0]	7.0	[6.0, 8.5]
Neck extensors	10.0	[7.5, 10.0]	9.0	[8.0, 10.0]	9.0	[8.0, 10.0]
Shoulder elevators	10.0	[9.0, 10.0]	10.0	[10.0, 10.0]	10.0	[9.0, 10.0]
Shoulder abductors	7.0	[6.0, 8.0]	8.0	[7.0, 9.0]	8.0	[7.0, 10.0]
Elbow flexors	9.0	[7.0, 10.0]	9.5	[7.0, 10.0]	9.0	[7.0, 10.0]
Wrist flexors[a,b]	10.0	[8.0, 10.0]	8.0	[7.0, 10.0]	9.0	[8.0, 10.0]
Wrist extensors	9.0	[8.0, 10.0]	9.0	[7.8, 10.0]	9.0	[8.0, 10.0]
Hip flexors[a,c,d]	4.0	[3.0, 7.0]	6.0	[3.0, 7.3]	7.0	[6.0, 9.0]
Hip extensors[c,d]	4.0	[3.0, 6.5]	6.0	[3.0, 7.0]	8.0	[6.5, 9.5]
Hip abductors[b,c,e]	5.0	[3.5, 7.5]	7.0	[5.8, 8.3]	9.0	[7.0, 10.0]
Knee extensors[a,b,c]	8.0	[7.0, 9.5]	9.0	[7.8, 10]	10.0	[9.0, 10.0]
Ankle dorsiflexors	9.0	[8.0, 10.0]	10.0	[9.0, 10.0]	10.0	[8.0, 10.0]
Ankle plantar flexors[a]	9.0	[8.0, 10.0]	10.0	[9.0, 10.0]	10.0	[9.0, 10.0]

The strength of muscle groups using MMT was compared among clinical groups using the Kruskal–Wallis test; if significant, *post hoc* pairwise comparisons were done with Wilcoxon rank sum test and resulting *P*-values were adjusted by Holm's method. [a]PM is significantly different from DM, $P \leq 0.05$. [b]DM is significantly different from JDM, *P*-value ≤0.05. [c]PM is significantly different from JDM, *P*-value ≤0.01. [d]DM is significantly different from JDM. *P*-value ≤0.01. [e]DM is significantly different from PM. $P \leq 0.01$.

TABLE 4. Strata of weakness[a] by clinical group

	PM ($n=65$) (percentage of subjects)				*DM* ($n=50$) (percentage of subjects)				*JDM* ($n=57$) (percentage of subjects)			
Muscle groups	None	Mild	Moderate	Severe	None	Mild	Moderate	Severe	None	Mild	Moderate	Severe
Neck flexors	11	46	35	8	8	52	34	6	14	37	44	5
Neck extensors	51	35	12	2	42	44	12	2	44	47	9	0
Shoulder elevators	71	20	8	2	82	16	0	2	67	28	5	0
Shoulder abductors	15	55	17	12	16	62	22	0	30	47	21	2
Elbow flexors	42	45	14	0	50	34	16	0	40	47	12	0
Wrist flexors[b]	52	42	6	0	30	52	16	2	46	47	5	2
Wrist extensors	46	48	6	0	26	68	6	0	44	47	9	0
Hip flexors[c]	2	25	40	34	4	34	36	26	14	51	30	5
Hip extensors[c]	2	23	29	46	2	26	44	28	25	51	16	9
Hip abductors[c]	9	26	40	25	14	56	22	8	28	49	18	5
Knee extensors[c]	25	52	18	5	40	52	8	0	63	28	9	0
Ankle dorsiflexors	48	48	2	3	58	40	2	0	58	40	2	0
Ankle plantar flexors[b]	45	45	9	2	66	34	0	0	61	32	7	0

[a]Strata of weakness are based on the MMT grading criteria: no detectable weakness = 10 out of 10, ability to move against gravity into the testing position and hold against maximal manual resistance; mild weakness = 7–9 out of 10, ability to move against gravity into the testing position and hold against varying degrees of submaximal manual resistance; moderate weakness = 4–6 out of 10, inability to sustain the testing position against gravity (4), inability to withstand minimal manual resistance in the testing position (5) or ability to withstand minimal manual resistance (6); severe weakness = 0–3 out of 10, inability to move limb beyond partial range of motion (<50% of partial range of motion) against gravity. [b]Distribution among clinical groups differs, $P<0.05$, Kruskal–Wallis non-parametric rank test for contingency table data. [c]Distribution among clinical groups differs, $P<0.001$, Kruskal–Wallis non-parametric rank test for contingency table data. (Values in rows may not equal 100% due to rounding; unilateral data were symmetrical, therefore only values from right side are included.)

associated with the hips (flexors, extensors and abductors) were the weakest muscle groups for both the PM and DM groups. MMT grades of <10 were observed for the three hip muscle groups by 86–98% of patients with PM or DM (Table 4).

Associations of myositis characteristics with MMT scores

We examined the association of measures of functional performance, myositis disease activity and demographic factors with the proximal MMT score. Mean modified Convery ADL scale scores were 51 ± 19.5 out of 91 for 30 of 65 patients with PM and 57 ± 14.3 for 15 of 50 patients with DM, indicating moderate dysfunction in PM and mild dysfunction in DM. Performance on the modified Convery ADL scale explained 59% ($P < 0.0001$) and 48% ($P < 0.005$) of the variance of the proximal MMT subscore in PM or DM in univariate linear regression analyses. Scores of 40 ± 11.0 out of possible 52 for the CMAS (in 53 of 57 patients) and 0.86 ± 0.91 out of 3.0 for the CHAQ (in 42 of 57 patients) indicated mild to moderate physical dysfunction in JDM [37]. Univariate linear regression analysis confirmed that the CMAS and CHAQ explained 62% ($P < 0.0001$) and 55% ($P < 0.0001$), respectively of the variance in the proximal MMT subscore in JDM.

None of the patient characteristics or myositis activity measures, such as age, disease duration, prednisone dose, CK and LDH serum levels, and BMI had a strong association with proximal MMT subscore. Other than functional performance measures, the best association with MMT was MD Global Activity ($R^2 = 0.20$–0.21, $P < 0.01$) in the PM and JDM groups. Patient age (PM) and LDH serum levels (PM and JDM) were significant, but had a low strength of association with proximal MMT subscores ($R^2 = 0.07$–0.12, $P \leq 0.03$). Multivariate linear regression did not significantly improve upon the univariate models in determining associations with strength.

Discussion

This cross-sectional study focused on force-generating capabilities of muscle in IIM measured with MMT. Our findings provided a data-driven description of weakness patterns in IIM and provided additional insights into associated muscle dysfunction.

The eleven bilateral muscle groups included in this study were found to be symmetric in degree of weakness for IIM clinical groups. We elected to organize the MMT subscores into regional and functional anatomic muscle groups to better understand the patterns of muscle weakness in IIM. While the traditional organization of MMT subscores into regional anatomic groups is used in the clinical diagnosis of IIM, these subgroups do not always reflect a functional organization of muscle groups within the context of basic and instrumental activities of daily living. We suggest that this functional organization of MMT scores may provide a construct for improving our understanding of strength function relationships in future studies (e.g. lower extremity strength influence on walking speed or chair rise ability). The severity of lower extremity and proximal muscle group weakness exceeded that of the upper extremity, axial and distal muscle groups, consistent with clinical insights of other investigators [1, 39]. However, our findings suggest that only the proximal and lower extremity MMT subscores were significantly different across all patient groups. Identifying and monitoring lower extremity weakness in patients with IIM is of clinical importance since impairments of these functional muscle groups have been associated with increased risk of falls and nursing home placement for older adults [40].

While the proximal pattern of weakness in IIM is well known, less attention has been given to variations in the patterns of weakness among different IIM clinical groups. Although distal MMT subscores were higher for all patients, it may be a distinguishing feature for some IIM clinical groups. Our findings revealed significantly lower MMT grades for the wrist flexors in patients with DM and the ankle plantar flexors in patients with PM.

Individual muscle groups were stratified based on the 10-point MMT grading criteria (Table 4). This allowed us to make distinctions among clinical groups and provide a clinical context for the MMT results. The proportion of individual MMT grades in the 'moderate' and 'severe' range revealed that PM, DM and JDM share the five weakest muscle groups from both the axial and proximal regions: neck flexors, shoulder abductors, hip flexors, hip extensors and hip abductors. These findings provide data to further refine MMT use as a core set measure [5] and clinical guidance regarding the targets of exercise interventions.

Improving our understanding of the distribution and severity of weakness in IIM is important, and raises a critical question: how does one interpret MMT scores in the context of functional performance? Strength demands of walking or rising from a chair require lower extremity muscle groups to be capable of not only moving the limbs against gravity, but also supporting and moving the body weight. Eriksrud and Bohannon [41] reported that elderly subjects unable to complete the sit-to-stand task attained 64% of the maximum summed MMT grades for the knee

extensors, whereas subjects with 89% of the maximum summed MMT grades could complete the task. Siegel *et al*. [42] showed the number of lower extremity muscles with an MMT grade of <7 out of 10 predicted walking speed in patients with juvenile IIM. Our JDM patients were relatively high functioning and did not exhibit any lower extremity median MMT grades <7 out of 10. However, our patients with DM or PM exhibited median MMT grades of ≤7 or ≤5 out of 10, respectively, for the hip muscle groups, indicating considerable weakness. The increased proximal weakness observed at all three hip muscles in patients with PM compared with DM may reflect a critical threshold of function and may partially explain why our patients with PM scored lower on functional performance measures.

Although this is the largest study of specific patterns of weakness in IIM clinical groups using a standardized approach to assessing strength, there are limitations. This study included MMT data obtained from patients over a wide range in age, including four participants with JDM between 4.6 and 7 yrs of age and five patients between 7 and 8 yrs of age. Some investigators [28, 43] have cautioned against administering the MMT to patients <5 or 6 yrs of age. Complications encountered in the strength assessment of very young subjects may be related to levels of attention, motivation and comprehension during testing [43]. In a study featuring 825 children with myelomeningocoel, McDonald and colleagues [44] reported that serial MMT examinations yielded stable results in participants older than 5 yrs of age. Our investigation featured only three subjects aged 5 yrs old or younger. In addition, our paediatric therapists did not encounter any patients in this study in which MMT findings were affected by subject cooperation or behaviour.

We found significant association among the functional measures and the proximal MMT subscore, but our regression analyses did not provide any additional insights regarding the relationship of clinical course variables with the magnitude of weakness. It should be noted, however, that disease duration was greater in patients with PM compared with DM, which may have had an influence on the greater degree of proximal weakness seen in this clinical group. Additional study will be needed to better understand the relationship among weakness, functional performance and extra-musculoskeletal disease features. Also, different physical therapists tested the adult and paediatric patients. Nonetheless, we believe that use of multiple physical therapists had minimal impact on results based on prior examiner training and our observation of good inter-rater reliability of summed MMT scores [45].

It is important to note that a general limitation of MMT is that it is unable to distinguish between strength impairments that result from disease activity *vs* disease damage [8]. IMACS and PRINTO have recognized the constraints of MMT as a primary therapeutic trial outcome and have recommended incorporating a physician global disease activity assessment to augment this core set measure [5, 6]. Finally, our reported findings regarding the distribution and severity of muscle weakness in our cohort of patients with IIM are constrained by our selection of muscle groups to include in the MMT. The results of our study do not suggest that other muscle groups not included in our MMT do not contribute to the functional limitations and disability observed in this patient population or that our entire group of muscles tested is recommended for inclusion in routine clinical testing or clinical trials.

In conclusion, our study provides the most detailed account of weakness in a cohort of patients with IIM. These findings suggest that PM is associated with greater proximal and lower extremity weakness than DM and JDM, and muscle groups are affected symmetrically across IIM groups. In addition, we have identified the five weakest muscle groups across IIM clinical groups, which may serve to influence the selection of end-points in future therapeutic trials and the targets of exercise intervention.

Rheumatology key messages

- The magnitude of weakness differs among IIM clinical groups, with PM weakest, adult DM intermediate and JDM strongest.
- Hip flexors, hip extensors, hip abductors, neck flexors and shoulder abductors are the muscle groups with the greatest weakness among all three clinical groups.

Acknowledgements

The authors thank Dr Maria Villalba and Dr Elizabeth Adams for their contributions to this work. We also thank Ms Earllaine Croarkin and Dr Mark Gourley for their critical reading of the manuscript. The opinions and information contained in this article are those of the authors and do not necessarily reflect those of the National Institutes of Health or the United States Public Health Service.

Funding: This work was supported in part by the Intramural Research Program of the National Institutes of Health, National Institute of Environmental Health Sciences, National Institute of Arthritis and Musculoskeletal and Skin Diseases, Rehabilitation Medicine Department and the NIH Clinical Center.

Disclosure statement: The authors have declared no conflicts of interest.

Supplementary data

Supplementary data are available at *Rheumatology* Online.

References

1 Dalakas MC, Hohlfeld R. Polymyositis and dermatomyositis. Lancet 2003;362:971–82.
2 Christopher-Stine L, Plotz PH. Adult inflammatory myopathies. Best Pract Res Clin Rheumatol 2004;18:331–44.
3 Harris-Love MO. Physical activity and disablement in the idiopathic inflammatory myopathies. Curr Opin Rheumatol 2003;15:679–90.
4 Stoll T, Bruhlmann P, Stucki G, Seifert B, Michel BA. Muscle strength assessment in polymyositis and dermatomyositis evaluation of the reliability and clinical use of a new, quantitative, easily applicable method. J Rheumatol 1995;22:473–7.
5 Rider LG, Giannini EH, Harris-Love M *et al.* Defining clinical improvement in adult and juvenile myositis. J Rheumatol 2003;30:603–17.
6 Ruperto N, Ravelli A, Murray KJ *et al.* Preliminary core sets of measures for disease activity and damage assessment in juvenile systemic lupus erythematosus and juvenile dermatomyositis. Rheumatology 2003;42:1452–9.
7 Oddis CV, Rider LG, Reed AM *et al.* International consensus guidelines for trials of therapies in the idiopathic inflammatory myopathies. Arthritis Rheum 2005;52:2607–15.
8 Miller FW, Rider LG, Chung YL *et al.* Proposed preliminary core set measures for disease outcome assessment in adult and juvenile idiopathic inflammatory myopathies. Rheumatology 2001;40:1262–73.
9 Airio A, Kautiainen H, Hakala M. Prognosis and mortality of polymyositis and dermatomyositis patients. Clin Rheumatol 2006;25:234–9.
10 Torres C, Belmonte R, Carmona L *et al.* Survival, mortality and causes of death in inflammatory myopathies. Autoimmunity 2006;39:205–15.
11 Sultan SM, Ioannou Y, Moss K, Isenberg DA. Outcome in patients with idiopathic inflammatory myositis: morbidity and mortality. Rheumatology 2002;41:22–6.
12 Rios G. Retrospective review of the clinical manifestations and outcomes in Puerto Ricans with idiopathic inflammatory myopathies. J Clin Rheumatol 2005;11:153–6.
13 Selva-O'Callaghan A, Labrador-Horrillo M, Munoz-Gall X *et al.* Polymyositis/dermatomyositis-associated lung disease: analysis of a series of 81 patients. Lupus 2005;14:534–42.
14 Marie I, Hachulla E, Hatron PY *et al.* Polymyositis and dermatomyositis: short term and longterm outcome, and predictive factors of prognosis. J Rheumatol 2001;28:2230–7.
15 Huber AM, Lang B, LeBlanc CMA *et al.* Medium- and long-term functional outcomes in a multicenter cohort of children with juvenile dermatomyositis. Arthritis Rheum 2000;43:541–9.
16 Ponyi A, Borgulya G, Constantin T, Vancsa A, Gergely L, Danko K. Functional outcome and quality of life in adult patients with idiopathic inflammatory myositis. Rheumatology 2005;44:83–8.
17 Maugars YM, Berthelot JM, Abbas AA, Mussini JM, Nguyen JM, Prost AM. Long-term prognosis of 69 patients with dermatomyositis or polymyositis. Clin Exp Rheumatol 1996;14:263–74.
18 Benbassat J, Gefel D, Larholt K, Sukenik S, Morgenstern V, Zlotnick A. Prognostic factors in polymyositis/dermatomyositis. A computer-assisted analysis of ninety-two cases. Arthritis Rheum 1985;28:249–55.

19 Danko K, Ponyi A, Constantin T, Borgulya G, Szegedi G. Long-term survival of patients with idiopathic inflammatory myopathies according to clinical features: a longitudinal study of 162 cases. Medicine 2004;83:35–42.
20 Constantin T, Ponyi A, Orban I *et al.* National registry of patients with juvenile idiopathic inflammatory myopathies in Hungary - clinical characteristics and disease course of 44 patients with juvenile dermatomyositis. Autoimmunity 2006;39:223–32.
21 Troyanov Y, Targoff IN, Tremblay JL, Goulet JR, Raymond Y, Senecal JL. Novel classification of idiopathic inflammatory myopathies based on overlap syndrome features and autoantibodies: analysis of 100 French Canadian patients. Medicine 2005;84:231–49.
22 Lynn SJ, Sawyers SM, Moller PW, O'Donnell JL, Chapman PT. Adult-onset inflammatory myopathy: North Canterbury experience 1989-2001. Intern Med J 2005;35:170–3.
23 Chwalinska-Sadowska H, Maldykowa H. Polymyositis-dermatomyositis: a 25-year follow-up of 50 patients (analysis of clinical symptoms and signs and results of laboratory tests). Mater Med Pol 1990;22:205–12.
24 Bronner IM, van der Meulen MF, de Visser M *et al.* Long-term outcome in polymyositis and dermatomyositis. Ann Rheum Dis 2006;65:1456–61.
25 Casademont J, Grau JM, Masanes F, Herrero C, Urbano-Marquez A. Analysis of the outcome of idiopathic inflammatory myopathies with particular emphasis on muscle capillary damage. Scand J Rheumatol 1993;22:292–8.
26 Drouet B, LeLoet X, Vittecoq O *et al.* A study of long-term survival, functional outcome and quality of life in patients with polymyositis or dermatomyositis. Rev Rhum 1996;63:321–30.
27 Bohan A, Peter JB. Polymyositis and dermatomyositis (second of two parts). N Engl J Med 1975;292:403–7.
28 Kendall FP, McCreary EK, Provance PG, Rodgers MM, Romani WA. Muscles: testing and function, 5th edn. Baltimore: Williams & Wilkins, 2005.
29 Dalakas MC, Sonies B, Dambrosia J, Sekul E, Cupler E, Sivakumar K. Treatment of inclusion-body myositis with IVIG: a double-blind, placebo-controlled study. Neurology 1997;48:712–6.
30 Mastaglia FL, Phillips BA, Zilko PJ. Immunoglobulin therapy in inflammatory myopathies. J Neurol Neurosurg Psychiatry 1998;65:107–10.
31 Adams EM, Pucino F, Yarboro C *et al.* A pilot study: use of fludarabine for refractory dermatomyositis and polymyositis, and examination of endpoint measures. J Rheumatol 1999;26:352–60.
32 Cabalar I, Villalba ML, Sherman J *et al.* A pilot study of the effect of methimazole, a drug that down-regulates MHC class I, on dermatomyositis and polymyositis. Arthritis Rheum 2001;44:S353.
33 Rider LG, Feldman BM, Perez MD *et al.* Development of validated disease activity and damage indices for the juvenile idiopathic inflammatory myopathies: I. Physician, parent, and patient global assessments. Juvenile dermatomyositis disease activity collaborative study group. Arthritis Rheum 1997;40:1976–83.
34 Convery FR, Minteer MA, Amiel D, Connett KL. Polyarticular disability: a functional assessment. Arch Phys Med Rehabil 1977;58:494–9.
35 Pincus T, Summey JA, Soraci SA Jr, Wallston KA, Hummon NP. Assessment of patient satisfaction in activities of daily living using a modified Stanford health assessment questionnaire. Arthritis Rheum 1983;26:1346–53.
36 Lovell DJ, Lindsley C, Rennebohm R *et al.* Development of validated disease activity and damage indices for the assessment of the juvenile idiopathic inflammatory myopathies: II. The Childhood Myositis Assessment Scale (CMAS): a quantitative tool for the evaluation of muscle function. Arthritis Rheum 1999;42:2213–9.
37 Huber AM, Hicks JE, Lachenbruch PA *et al.* Validation of the childhood health assessment questionnaire in the juvenile idiopathic myopathies. J Rheumatol 2001;28:1106–11.
38 Portney LG, Watkins MP. Foundations of clinical research: applications to practice. Connecticut: Appleton & Lange, 1993. 2nd edition, p. 52–60.
39 Pachman LM. Juvenile dermatomyositis and other inflammatory myopathies. In: Jones HR, De Vivo DC, Darras BT, eds. Neuromuscular disorders of infancy, childhood, and adolescence: a clinician's approach. Boston: Butterworth-Heinemann, 2003:901–37.
40 Brown M, Sinacore DR, Host HH. The relationship of strength to function in the older adult. J Gerontol Ser A-Biol Sci Med Sci 1995;50:55–9.
41 Eriksrud O, Bohannon RW. Relationship of knee extension force to independence in sit-to-stand performance in patients receiving acute rehabilitation. Phys Ther 2003;83:544–51.
42 Siegel KL, Hicks JE, Koziol DE, Gerber LH, Rider LG. Walking ability and its relationship to lower-extremity muscle strength in children with idiopathic inflammatory myopathies. Arch Phys Med Rehabil 2004;85:767–71.
43 Hinderer KA, Hinderer SR. Muscle strength development and assessment in children and adolescents. In: Harms-Ringdahl K, ed. Muscle strength. Edinburgh: Churchill Livingstone, 1993:93–140.
44 McDonald CM, Jaffe KM, Shurtleff DB. Assessment of muscle strength in children with meningomyelocele: accuracy and stability of measurements over time. Arch Phys Med Rehabil 1986;67:855–61.
45 Jain M, Smith M, Cintas H *et al.* Intra-rater and inter-rater reliability of the 10-point manual muscle test (MMT) of strength in children with juvenile idiopathic inflammatory myopathies (JIIM). Phys Occup Ther Pediatr 2006;26:5–17.