Perceptions of Patients, Caregivers, and Healthcare Providers of Idiopathic Inflammatory Myopathies: An International OMERACT Study

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ABSTRACT. Objective. Patient-reported outcome measures (PROM) that incorporate the patient perspective have not been well established in idiopathic inflammatory myopathies (IIM). As part of our goal to develop IIM-specific PROM, the Outcome Measures in Rheumatology (OMERACT) Myositis special interest group sought to determine which aspects of disease and its effects are important to patients and healthcare providers (HCP).

Methods. Based on a prior qualitative content analysis of focus groups, an initial list of 24 candidate domains was constructed. We subsequently conducted an international survey to identify the importance of each of the 24 domains to be assessed in clinical research. Patients with IIM, their caregivers, and HCP treating IIM completed the survey.

Results. In this survey, a total of 638 respondents completed the survey, consisting of 510 patients, 101 HCP, and 27 caregivers from 48 countries. Overall, patients were more likely to rank "fatigue," "cognitive impact," and "difficulty sleeping" higher compared with HCP, who ranked "joint symptoms," "lung symptoms," and "dysphagia" higher. Both patients and providers rated muscle symptoms as their top domain. In general, patients from different countries were in agreement on which domains were most important. One notable exception was that patients from Sweden and the Netherlands ranked lung symptoms significantly higher compared to other countries including the United States and Australia (mean weighted rankings of 2.86 and 2.04 vs 0.76 and 0.80, respectively; $p < 0.0001$).

Conclusion. Substantial differences exist in how IIM is perceived by patients compared to HCP, with different domains prioritized. In contrast, patients' ratings across the world were largely similar. (First Release September 15 2018; J Rheumatol 2019;46:106–11; doi:10.3899/jrheum.180353)

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Idiopathic inflammatory myopathies (IIM) affect muscle and extramuscular organs, resulting in significant limitation in activities of daily living and health-related quality of life[1,2,3,4]. However, outcome measures used in clinical studies for IIM are often based on the measurement of pathophysiologic manifestations of the disease such as muscle weakness, elevated muscle enzymes, and skin changes, whereas patients' perceptions of the disease's effect on their lives have not been systematically studied or well represented[3]. The Outcome Measures in Rheumatology (OMERACT) Myositis special interest group (SIG) was established to define a set of core domains and ultimately identify and/or develop instruments that reflect the symptoms and life effects that are experienced by people living with myositis[5,6]. OMERACT is the acronym for an international, organized network initiated in 1992 and aimed at improving outcome measurement in rheumatology. A domain, according to OMERACT, is a further specification of an aspect of health, for example, pain or physical function. Data-driven recommendations are prepared and updated by expert working groups, which consist of experts from multiple continents and include patients with the disease[7]. The Myositis SIG comprises 2 patient research partners with myositis, healthcare providers (HCP) with experience and expertise in IIM from 4 continents, and experts in quantitative and qualitative methodology.

Over several years, the Myositis SIG has performed a systematic review of the literature and identified gaps in the current patient-reported outcome measures (PROM) used in IIM research[8]. None of the existing PRO instruments have been developed following the currently recommended qualitative methodology outlined by OMERACT and other groups for domain identification and prioritization[9,10,11,12,13,14]. Therefore, our group conducted several focus groups in multiple countries to identify themes that were described by patients as relevant to their experience of myositis[15]. An initial modified Delphi was performed to allow patients to rate the importance of each domain as well as to write in any new domains. From these results, a list of 24 candidate domains was developed with the intention of surveying a large, international group of IIM patients to determine the most important domains to carry forward in the development of PROM, for use in research and clinical practice. We also chose to include other groups in this survey, including caregivers and HCP, to compare which domains are perceived as most important by different groups. The aim of this report is to describe the most important domains according to patients, and how these domains compare to other groups.

MATERIALS AND METHODS

Patients with adult polymyositis (PM), dermatomyositis (DM), antisynthetase syndrome (AS), or immune-mediated necrotizing myopathy (IMNM) were invited to participate in this survey. Exclusion criteria were patients with a diagnosis of inclusion body myositis or juvenile-onset myositis. The survey was distributed internationally through several listservs including the Global Conference on Myositis, The Myositis Association, the European Neuromuscular Centre, The Myositis Association of Australia, and the International Myositis Assessment and Clinical Studies Group. Through these listservs, the survey was distributed to patients, caregivers, HCP (clinicians and allied health professionals including physical and occupational therapists), and regulatory personnel (government advisory and industry employees). The survey was distributed using the Internet-based survey platform Qualtrics. The Johns Hopkins University Hospital institutional review board (IRB; NA_00098790), as well as each individual institution's IRB equivalent, approved this study. The completion of the survey served as informed consent to participate in this research study as outlined in the respective IRB protocols.

The full text of the survey can be found in the Supplementary Data (available from the authors on request). The survey was forward and backward translated into Swedish, Dutch, and Korean. These languages were chosen for translation because Sweden, the Netherlands, and South Korea were represented by members in the OMERACT SIG. In the survey, respondents were initially asked to select their top 10 domains from the list of 24. From this list of 10, they were next asked to select their top 5 and rank them in order of importance. A point system was used to weight each domain ranked in the respondents' top 5; for example, the top-ranked domain was weighted with 5 points, the second-ranked domain was weighted with 4 points, the third-ranked domain was weighted with 3 points, and so on. Based on this system, a weighted ranking score was assigned to each domain. Continuous variables were analyzed using ANOVA. Categorical variables were compared using chi-square tests or Fisher's exact test, as appropriate. $P < 0.05$ was considered to indicate statistical significance. All analyses were performed using SPSS version 21 (IBM Corp.).

RESULTS

A total of 876 individuals opened the survey, and there were 638 responses completed in their entirety from eligible participants. A total of 238 patients were excluded because they failed to complete the survey (n = 88) or they had conditions such as inclusion body myositis and juvenile dermatomyositis (n = 150), which were not part of our study. The exact denominator could not be determined, because recipients of the survey subsequently forwarded the anonymous link to other relevant interested parties. Thus, the survey response rate could not be calculated.

A total of 510 patients, 101 HCP, and 27 caregivers

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completed the survey. A single respondent identified as a healthcare authority. The average age of the respondents was 55 ± 13.5 years, and 73% were female. Of the HCP, 76 were physicians, 22 were physical/occupational therapists, and 3 identified as "other." Of the physicians, 45 were rheumatologists, 28 were neurologists, and 3 did not report their subspecialty area. The average duration of disease was 7.9 ± 13.7 years. Of the 510 patients, 238 self-identified as DM, 212 as PM, and 60 as "other," including AS and IMNM. Countries with the most participants included the United States (n = 387), the Netherlands (n = 52), Sweden (n = 50), Australia (n = 30), Canada (n = 26), Germany (n = 19), South Korea (n = 17), and the United Kingdom (n = 13). Respondents from 40 other countries also completed the survey in 1 of the 4 translated languages, including Argentina, Austria, Belgium, Brazil, Czech Republic, Denmark, El Salvador, Finland, the former Yugoslav Republic of Macedonia, France, Ghana, Greece, Guatemala, Hong Kong, Hungary, India, Indonesia, Ireland, Israel, Italy, Jamaica, Japan, Kazakhstan, Malaysia, Mexico, New Zealand, Pakistan, Poland, Republic of Moldova, Serbia, Slovakia, South Africa, Spain, Suriname, Switzerland, Trinidad and Tobago, Tunisia, United Arab Emirates, and Vietnam.

Of the 638 respondents who completed the survey, the most frequent domains selected as important included "muscle symptoms," "fatigue," "physical activity," "medication side effects," and "pain." A full list of all domains in rank order are presented in Table 1. When comparing patients with HCP, significant differences were observed. Patients were much more likely to select "fatigue," "emotional distress," "difficulty sleeping," and "dryness of eyes and/or mouth." In contrast, HCP were more likely to select "skin symptoms," "lung symptoms," "dysphagia," and "personal care" (p value for all comparisons < 0.005).

We next analyzed the results stratified by country of origin (Table 2). Given the small number of HCP responding from each country, we focused our analyses on patients. Overall, patients around the world ranked domains similarly; however, several exceptions were noted. Patients from Sweden and the Netherlands had higher proportions of patients who ranked lung symptoms highly (2.86 and 2.04, respectively) compared to other countries including the United States and Australia (0.76 and 0.8, respectively, p < 0.0001). Other domains that were statistically different between countries were fatigue (p = 0.005), medication side effects (p = 0.03), pain (p = 0.012), and cardiovascular symptoms (p = 0.003).

Upon examining responses by diagnosis, patients with DM ranked skin symptoms higher than patients with PM or other diagnoses (including IMNM or AS; p < 0.0001). Patients with non-DM and non-PM ranked lung symptoms significantly higher (p < 0.0001). Patients with PM ranked muscle symptoms (p = 0.005), difficulty with leisure activities (p = 0.017), and physical activities (p = 0.009) higher

Table 1. Rank ordering of domains selected by patients and HCP, based on proportion of respondents that selected each domain in their top 10.

	Domain	Total, n = 638	Patients, n = 510	HCP, n = 101	Caregivers, n = 27	p
1	Muscle symptoms	96.2	96.1	99.0	88.9	0.045
2	**Fatigue**	**82.4**	**85.9**	**66.3**	**77.8**	**< 0.0001**
3	Levels of physical activity	70.7	68.4	83.2	66.7	0.011
4	Medication side effects	65.8	66.7	59.4	74.1	0.243
5	Pain	59.9	59.4	59.4	70.4	0.524
6	Skin symptoms	54.9	51.6	72.3	51.9	0.001
7	**Lung symptoms**	**53.6**	**48.8**	**83.2**	**33.3**	**< 0.0001**
8	**Dysphagia**	**50.0**	**45.5**	**76.2**	**37.0**	**< 0.0001**
9	Increased risk of infection	43.9	44.9	34.7	59.3	0.043
10	Work ability	45.1	42.7	61.4	29.6	0.001
11	**Cognitive impact**	**37.9**	**41.8**	**14.9**	**51.9**	**< 0.0001**
12	**Joint symptoms**	**44.7**	**41.6**	**62.4**	**37.0**	**< 0.0001**
13	**Difficulty sleeping**	**34.6**	**39.0**	**9.9**	**44.4**	**< 0.0001**
14	Emotional distress	35.7	37.1	23.8	55.6	0.003
15	Gastrointestinal tract symptoms	34.3	36.5	22.8	37.0	0.029
16	**Dryness of eyes and/or mouth**	**27.3**	**31.6**	**5.0**	**29.6**	**< 0.0001**
17	Household activities	29.5	29.6	30.7	22.2	0.684
18	**Cardiovascular symptoms**	**30.1**	**27.8**	**45.5**	**14.8**	**< 0.0001**
19	Leisure activities	25.4	26.7	21.8	14.8	0.256
20	**Interaction***	**23.5**	**24.9**	**8.9**	**51.9**	**< 0.0001**
21	**Personal care**	**17.7**	**15.1**	**31.7**	**14.8**	**< 0.0001**
22	Relation and/or intimacy	14.1	14.9	7.9	22.2	0.085
23	Incontinence	11.0	12.7	3.0	7.4	0.013
24	Social gathering	11.6	10.8	16.8	7.4	0.175

Values are % unless otherwise specified. * Interaction with healthcare personnel and authorities. Values in bold face are statistically significant. HCP: healthcare providers.

Table 2. Weighted rank ordering of domains selected by patients based on proportion of respondents who selected each domain in their top 5 according to countries (based on Question 15).

	Domains	Total, n = 510	Australia, n = 25	Netherlands, n = 27	Sweden, n = 29	USA, n = 339	p
1	Muscle symptoms	3.45 ± 1.86	3.6 ± 1.96	3.22 ± 1.93	3.28 ± 2.05	3.47 ± 1.84	0.838
2	**Fatigue**	**1.98 ± 1.77**	**2.12 ± 1.83**	**2.85 ± 1.85**	**1.17 ± 1.58**	**1.97 ± 1.75**	**0.005**
3	Pain	1.32 ± 1.81	1.76 ± 2.09	0.81 ± 1.55	0.45 ± 0.95	1.4 ± 1.85	0.012
4	**Lung symptoms**	**0.99 ± 1.77**	**0.8 ± 1.55**	**2.04 ± 2.08**	**2.86 ± 2.18**	**0.76 ± 1.59**	**< 0.0001**
5	Skin symptoms	0.99 ± 1.65	0.8 ± 1.47	0.56 ± 1.09	0.59 ± 1.3	1.07 ± 1.72	0.187
6	Levels of physical activity	0.92 ± 1.39	1.2 ± 1.5	0.89 ± 1.25	0.45 ± 0.83	0.95 ± 1.43	0.214
7	Medication side effects	0.76 ± 1.35	1.2 ± 1.44	1.15 ± 1.54	1.14 ± 1.53	0.67 ± 1.3	0.033
8	Dysphagia	0.71 ± 1.46	0.36 ± 0.91	0.33 ± 1.21	0.86 ± 1.68	0.76 ± 1.48	0.275
9	Cognitive impact	0.59 ± 1.27	0.48 ± 1	0.3 ± 0.95	0.17 ± 0.47	0.65 ± 1.35	0.136
10	Increased risk of infection	0.45 ± 1.09	0.48 ± 1.23	0.41 ± 1.01	0.97 ± 1.4	0.41 ± 1.05	0.073
11	Difficulty sleeping	0.4 ± 1.02	0.28 ± 1.06	0.22 ± 0.8	0.03 ± 0.19	0.45 ± 1.06	0.122
12	Joint symptoms	0.39 ± 1.03	0.32 ± 0.9	0.37 ± 0.93	0.66 ± 1.2	0.37 ± 1.03	0.546
13	Gastrointestinal tract symptoms	0.34 ± 0.98	0.28 ± 0.74	0.41 ± 1.05	0.24 ± 0.99	0.35 ± 0.99	0.910
14	**Cardiovascular symptoms**	**0.29 ± 0.99**	**0.44 ± 1.39**	**0.48 ± 1.31**	**0.86 ± 1.62**	**0.21 ± 0.83**	**0.003**
15	Emotional distress	0.28 ± 0.88	0.24 ± 0.72	0.15 ± 0.53	0.45 ± 0.95	0.28 ± 0.9	0.631
16	Work ability	0.25 ± 0.81	0.08 ± 0.4	0.15 ± 0.36	0.21 ± 0.94	0.27 ± 0.85	0.591
17	Interaction*	0.22 ± 0.86	0.04 ± 0.2	0 ± 0	0.52 ± 1.06	0.23 ± 0.9	0.097
18	Dryness of eyes and/or mouth	0.16 ± 0.6	0.04 ± 0.2	0.04 ± 0.19	0.07 ± 0.37	0.19 ± 0.65	0.312
19	Incontinence	0.13 ± 0.61	0.12 ± 0.44	0 ± 0	0 ± 0	0.15 ± 0.67	0.385
20	Relation and/or intimacy	0.09 ± 0.49	0.16 ± 0.8	0.22 ± 0.97	0 ± 0	0.08 ± 0.42	0.317
21	Leisure activities	0.09 ± 0.44	0.2 ± 0.71	0.26 ± 0.86	0.03 ± 0.19	0.07 ± 0.38	0.085
22	Household activities	0.08 ± 0.42	0 ± 0	0 ± 0	0 ± 0	0.09 ± 0.47	0.358
23	Personal care	0.06 ± 0.43	0 ± 0	0 ± 0	0 ± 0	0.08 ± 0.48	0.535
24	Social gathering	0.05 ± 0.35	0 ± 0	0.15 ± 0.6	0 ± 0	0.04 ± 0.35	0.346

Values are mean ± SD. *Interaction with healthcare personnel and authorities. Values in bold face are statistically significant.

compared to patients with DM or other forms of myositis. Full details can be found in Table 3.

DISCUSSION

We show that there are substantial differences in how patients and HCP perceive IIM. Not surprisingly, most domains prioritized by patients reflect those symptoms and effects indicative of the lived experience of disease such as fatigue, cognition, and difficulty sleeping. In contrast, HCP selected domains that have traditionally been measured (i.e., skin, lung, and muscle symptoms). There was agreement between patients and HCP on several domains. The most prominent agreement involved muscle symptoms, but there was also agreement on medication side effects, joint symptoms, the ability to work, and cardiovascular symptoms. The observation that cognitive symptoms are rated so high by patients may be counterintuitive, given that IIM is predominantly a muscle disease. However, patients with rheumatoid arthritis have cognitive impairment, which has been associated with disease activity and immunity[16]. To our knowledge, our group was the first to describe the cognitive effect of myositis; mechanisms for cognitive impairment are unknown to date. Systemic inflammation could be a contributing factor, but poor sleep quality and fatigue could also be involved. Further, many patients may have interpreted cognitive symptoms to include depression and anxiety, which have been reported in patients with DM[17]. However, patients with PM, AS, and IMNM ranked cognitive symptoms similarly, suggesting that it is not only skin symptoms driving this domain selection.

Comparisons by country are particularly interesting, in that within such a heterogeneous disease as myositis and in the backdrop of different healthcare delivery systems and cultures, there exists remarkable agreement in how domains are prioritized. The prominent difference comparing 2 European countries (Sweden and the Netherlands) to other western countries (United States and Australia) was the notable exception. Sweden and the Netherlands rated lung symptoms significantly higher (2.86 and 2.04, respectively), compared to 0.76 and 0.8 for the United States and Australia. In a recent report of the EuroMyositis Registry, it was estimated that the prevalence of interstitial lung disease (ILD) was 30% as defined by chest radiograph/high-resolution computed tomography (HRCT) scan and pulmonary function test (PFT)[18]. This is higher than that reported in a US cohort of 11%, where ILD was defined similarly by HRCT and PFT[19]; thus geographic or genetic differences may explain these discrepant findings. There has not been robust research into the prevalence of pulmonary disease in other individual countries, but in discussions among authors of our study, the range of patients with ILD seen in our respective clinics varies from about 20–45%. However, it is unknown whether the patients responding to the survey were representative of these cohorts.

Table 3. Comparison between DM, PM, and other patients.

	Domains	Total, n = 510	DM, n = 238	PM, n = 212	Others†, n = 60	p
1	**Muscle symptoms**	**3.43 ± 1.88**	**3.34 ± 1.91**	**3.7 ± 1.75**	**2.85 ± 2.04**	**0.005**
2	Fatigue	1.89 ± 1.78	1.89 ± 1.76	1.85 ± 1.81	2.02 ± 1.76	0.823
3	Pain	1.33 ± 1.83	1.39 ± 1.8	1.27 ± 1.84	1.28 ± 1.97	0.752
4	**Lung symptoms**	**1.03 ± 1.78**	**0.83 ± 1.62**	**0.95 ± 1.73**	**2.1 ± 2.18**	**< 0.0001**
5	**Skin symptoms**	**0.96 ± 1.64**	**1.78 ± 1.95**	**0.25 ± 0.81**	**0.23 ± 0.79**	**< 0.0001**
6	**Levels of physical activity**	**0.91 ± 1.4**	**0.71 ± 1.23**	**1.11 ± 1.53**	**1 ± 1.45**	**0.009**
7	Medication side effects	0.79 ± 1.37	0.63 ± 1.21	0.96 ± 1.5	0.88 ± 1.43	0.033
8	Dysphagia	0.7 ± 1.45	0.79 ± 1.45	0.64 ± 1.45	0.58 ± 1.44	0.440
9	Cognitive impact	0.56 ± 1.25	0.58 ± 1.26	0.54 ± 1.22	0.58 ± 1.34	0.940
10	Increased risk of infection	0.47 ± 1.11	0.52 ± 1.17	0.38 ± 0.97	0.63 ± 1.28	0.218
11	Joint symptoms	0.41 ± 1.05	0.4 ± 1.06	0.4 ± 1.03	0.5 ± 1.11	0.786
12	Difficulty sleeping	0.4 ± 1.02	0.35 ± 0.99	0.48 ± 1.09	0.33 ± 0.9	0.380
13	Work ability	0.32 ± 0.91	0.23 ± 0.71	0.38 ± 1.01	0.45 ± 1.16	0.094
14	Gastrointestinal tract symptoms	0.3 ± 0.94	0.29 ± 0.99	0.34 ± 0.92	0.22 ± 0.78	0.654
15	Emotional distress	0.29 ± 0.88	0.26 ± 0.89	0.33 ± 0.9	0.23 ± 0.74	0.676
16	Cardiovascular symptoms	0.28 ± 0.97	0.26 ± 0.91	0.35 ± 1.1	0.13 ± 0.6	0.252
17	Interaction*	0.2 ± 0.82	0.18 ± 0.79	0.23 ± 0.88	0.22 ± 0.72	0.774
18	Dryness of eyes and/or mouth	0.18 ± 0.66	0.23 ± 0.79	0.13 ± 0.5	0.17 ± 0.56	0.307
19	Incontinence	0.11 ± 0.56	0.06 ± 0.43	0.18 ± 0.69	0.08 ± 0.53	0.057
20	Relation and/or intimacy	0.11 ± 0.53	0.1 ± 0.49	0.12 ± 0.52	0.13 ± 0.7	0.856
21	Leisure activities	0.09 ± 0.45	0.04 ± 0.33	0.16 ± 0.56	0.07 ± 0.36	0.017
22	Household activities	0.09 ± 0.44	0.06 ± 0.31	0.1 ± 0.45	0.18 ± 0.75	0.140
23	Personal care	0.07 ± 0.45	0.08 ± 0.45	0.06 ± 0.45	0.08 ± 0.42	0.919
24	Social gathering	0.05 ± 0.34	0.01 ± 0.13	0.09 ± 0.49	0.03 ± 0.18	0.037

Values are mean ± SD. † Others included necrotizing autoimmune myopathies and the antisynthetase syndrome patients. *Interaction with healthcare personnel and authorities. Values in bold face are statistically significant. DM: dermatomyositis; PM: polymyositis.

Upon comparing items by diagnosis, predictably, patients with DM ranked skin highly, whereas patients with PM rated muscle symptoms and difficulty with leisure activities and physical activity higher, suggesting more muscle involvement leading to decreased function. Patients who were non-PM and non-DM ("other") encompassed a combination of those with AS and IMNM. The high ranking of lung symptoms in this group is likely attributable to the higher prevalence of ILD associated with AS.

Our study has several strengths rooted in the large number of patients responding from many different countries, including traditionally non–English-speaking countries. The generalizability of survey responses is often limited by constraining the source population to predominantly English-speaking countries including the United Kingdom, Australia, the United States, and Canada. The distribution of a survey translated into 4 languages was done in an effort to include as many different patients as possible. The forward and backward translations by native speakers were intended to reduce any potential misinterpretation and eliminate idioms[20]. In addition, the large number of patient participants allows for comparisons between countries, providing insights into similarities (or differences) in how IIM is perceived in different parts of the world.

Our study is not without limitations. For one, some domains did not have explanations clarifying their intended meaning. This was in part purposeful as we wanted to leave this interpretation up to the survey respondent. However, an unintended consequence may be misinterpretation; for example, "lung symptoms" could be considered referring to cough, wheezing, or shortness of breath. In addition, while we translated the survey into 4 languages, individuals from many other countries who may have different native languages completed the survey. This may have led to misinterpretation. Also, while a large number of patient respondents were obtained, there exists the potential for respondent bias of individuals who are doing poorly (e.g., worse disease activity, emotional distress, treatment side effects). Other limitations include the lack of other HCP groups including pulmonologists, dermatologists, speech therapists, and nurses. Future research should include other organization listservs including these profession groups. Despite the absence of these subspecialists, however, both skin and lung symptoms were ranked highly.

Both patients and providers rated muscle symptoms as their top domain. In general, patients across the world appear to value the same domains. However, a number of differences were noted between patients and HCP. These data will allow for the education of HCP to better understand the life effect of patients with IIM. It is hoped that having these differences highlighted also will make it more likely that HCP will endorse and use the PROM that result from this work.

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