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Disability in idiopathic inflammatory myopathies: questionnaire-based study

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Abstract

Idiopathic inflammatory myopathies (IIM) are progressive, debilitating diseases that can lead to severe impairment. The aim of the study was to evaluate the level of disability and compare it between different subtypes of IIM as well as to estimate clinical symptoms associated with greater risk of disability and distinguish the most troublesome activities in this group of patients. A online form concerning clinical symptoms, comorbidities and limitations in daily living was created and distributed to online support groups for patients with IIM. Health Assessment Questionnaire was used to estimate disability and physical limitations while visual analogue scales enabled to assess the intensity of clinical symptoms. 361 out of 377 responders were included for further evaluation. High prevalence of disability was observed in each subtype yet predominantly in patients with inclusion body myositis (IBM) as 51.43% of them fulfilled the criteria of severe to very severe disability. Level of disability correlated with muscle weakness, tolerance of physical activity and level of fatigue. 45.62% of responders in general required walking devices and 43.50% of participants declared using facilitating devices for maintaining hygiene. Patients with IIM encounter multitude physical limitations that can be partially compensated by usage of facilitating devices or aid of the caregivers. IBM seems to be the most disabling subtype.

Keywords Myositis · Disability · Health Assessment Questionnaire · Idiopathic inflammatory myopathy

Introduction

Idiopathic inflammatory myopathies (IIM) are rare, heterogeneous, autoimmune collagen vascular diseases [1]. Although commonly associated with muscle weakness and myalgia, they can also manifest with a variety of organ-specific symptoms [2, 3]. Due to progressive muscular atrophy, decreasing endurance, internal organ involvement and limited therapeutic options, the disease often leads to physical disability. Several subtypes of IIM have been distinguished including among others dermatomyositis (DM), polymyositis (PM), juvenile myositis (JDM) and inclusion body myositis (IBM) [1, 4]. The Health Assessment Questionnaire (HAQ) remains one of the most commonly used patient-reported outcome (PRO) tool [5, 6]. Its usefulness results from multidimensional character and universality, possibility to perform also by means of an online or phone survey, availability in many translations, besides it does not require supervision of the medical professional and provides information from the patients' perspective [5, 6]. It is recommended by scientific societies as a part of Core Set Measures [7]. Core Set Measures is a list of tests or questionnaires selected to provide a thorough and detailed evaluation of disease activity, disease damage, quality of life and clinical response in patients with IIM to meet the demands of scientific and clinical studies [5, 7]. The aim of the study was to assess the level of disability in different subtypes of IIM and to analyse the possible factors leading to progressive loss of physical function.

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Methods

In order to evaluate the prevalence of various symptoms and the degree of disability we constructed an online questionnaire which was distributed to English-speaking patients diagnosed with IIM by means of the social media such as online support groups and community forums. The questionnaire consisted of 40 questions concerning disease duration, subtype of myositis, comorbidities and concomitant symptoms. Intensity of myalgia and intensity of the feeling of muscle weakness as well as degree of fatigue and tolerance of physical activity were assessed in visual analogue scales (VAS 1–10 points range) where 1 indicated the lowest and 10 the highest intensity. Responders were also asked to fill the Short 2-page Health Assessment Questionnaire, also referred as The HAQ-DI. Inclusion criteria comprised of voluntary completion of the questionnaire, marking the diagnosis of PM/DM/IBM and answering the required questions. Patients with JDM ($n=9$, 2.39%) were excluded from the further evaluation, as the number of responders in this group was incomparably lower. Patients that marked answer "I don't know" ($n=7$; 1.86%) in a question concerning the subtype of myositis were also excluded from the study to eliminate the risk of inaccurate diagnosis. Patients that chose the diagnosis of DM/PM/IBM but declared concomitant diseases such as systemic lupus, scleroderma, Sjögren syndrome, rheumatoid arthritis, vasculitis as well as patients who declared diagnosis of mixed connective tissue disease (MCTD) were included in the subgroup of myositis with other connective tissue disorders (MwC). While evaluating the data we followed the instructions contained in The Health Assessment Questionnaire (HAQ) disability index (DI) of the clinical health assessment questionnaire (version 96.4) [8]. HAQ-DI includes 20 questions divided into eight functional categories such as dressing and grooming, arising, eating, walking, hygiene, reaching, gripping and daily activities [6]. In each category, patient has to answer two or three questions by choosing the statement that suits best his/her situation during the past week. Possible answers are scored, respectively, as 0 points for "without any difficulty", 1 point for "with some difficulty", 2 points if the answer is "with much difficulty" and 3 points for "unable to do". We obtained the final score for each of the 8 categories by selecting the highest score for the questions included in particular category. In order to count the Alternative Disability Index (ADI) we summed up the final scores of 8 categories and divided this sum by 8. In order to obtain the Standard Disability Index (SDI) we considered whether patient needed to use facilitating devices or help from another person to perform listed activities. The list of facilitating devices included equipment used for dressing such as button hook, zipper pull, shoe horn, special or built up chair for arising, built up or special utensils for eating, walking aids such as cane, walker, crutches or wheelchair, bathtub bar, long-handled appliances in bathroom and raised toilet seat for daily hygiene, long-handled appliances for reaching, aids for opening such as jar opener for jars previously opened. Patients were also asked to mark all of the categories in which they require help from another person. If final score for the category was 2 or 3 we left the score unchanged (even if patient used devices or needed help). If the final score for the category was 0 or 1 and the patient declared using devices or/and help from another person the score for that category was changed to 2 points. Final score of 0 reflects no disability, scores of 0 to 1 represent mild to moderate difficulty, between 1 and 2 moderate to severe disability while scores of 2–3 indicate severe to very severe disability [6, 8]. We correlated SDI and ADI scores with disease duration, feeling of muscle weakness and myalgia as well as fatigue and tolerance of physical activity declared by patients.

Statistical analysis

Data were analysed with STATISTICA 13.1 software. The normality of data was tested by Shapiro–Wilk test. Mean values of SDI and ADI were evaluated for each IIM subtype and compared between groups with Kruskal–Wallis ANOVA on ranks with post hoc tests. Number and percentage of responders who needed to use each type of facilitating devices were compared with χ^2 Test. Kruskal–Wallis statistic with post hoc Dunn's Multiple Comparison Test was used to compare intensity of myalgia, feeling of muscle weakness as well as tolerance of physical activity and degree of fatigue between groups of patients with different IIM subtype. Relation between disease duration and mean values of SDI and ADI was assessed with ANOVA Kruskal–Wallis on ranks with post hoc analysis. To evaluate the influence of myalgia, feeling of muscle weakness, tolerance of physical activity and fatigue on SDI and ADI scores linear regression was performed.

Results

Our survey was completed by 377 patients; 361 of them were included for further evaluation. Most of the responders were diagnosed with IBM (37.14%), 32.63% of patients with DM, 15.92% with PM and 10.08% with MwC. The patient characteristics are presented in Table 1.

Patients with IBM were found to be more disabled as compared to PM, DM and MwC. ADI in IBM patients was significantly higher as compared to DM (mean 1.9 ± 0.80 vs mean 0.77 ± 0.61; $p < 0.001$), as compared to PM (mean

Table 1 Age and sex structure, disease duration and comorbidities in patients with particular subtype of idiopathic inflammatory myopathies

Number of patients	Total	DM	PM	IBM	MwC	*p*
	377	123	60	140	38	
Female/male	68.17%/31.83%	88.62%/11.38%	78.33%/21.67%	37.86%/62.14%	86.84%/13.16%	IBM vs DM < 0.001 IBM vs PM < 0.001 IBM vs MwC < 0.001
Age (years)						
< 18	7(1.85%)	4(3.25%)	1(1.67%)	0	0	IBM vs DM < 0.001 IBM vs PM < 0.001 IBM vs MwC < 0.001
18–25	13(3.44%)	6(4.88%)	3(5.00%)	0	2(5.26%)	
26–40	49(13.00%)	30(24.39%)	6(10.00%)	1(0.71%)	6(15.79%)	
41–65	179(47.48%)	70(56.91%)	34(56.67%)	48(34.28%)	24(63.16%)	
66–75	92(24.40%)	11(8.94%)	14(23.33%)	59(42.14%)	5(13.16%)	
> 76	37(9.81%)	2(1.63%)	2(3.33%)	32(22.85%)	1(2.63%)	
Disease duration						
More than 5 years	176(46.68%)	40(32.52%)	32(53.33%)	76(54.29%)	21(55.26%)	IBM vs DM < 0.001
3–5 years	69(18.30%)	25(20.32%)	8(13.33%)	30(21.43%)	4(10.52%)	
1–3 years	81(21.48%)	35(28.45%)	11(18.33%)	27(19.29%)	5(13.16%)	
6–12 months	23(6.11%)	12(9.76%)	4(6.67%)	3(2.14%)	3(7.89%)	
3–6 months	15(3.98%)	8(6.50%)	2(3.33%)	3(2.14%)	1(2.63%)	
Less than 3 months	11(2.92%)	3(2.43%)	3(5.00%)	1(0.71%)	4(10.52%)	
Concomitant disorders						
Hypertension	120 (31.83%)	28 (22.76%)	19 (31.67%)	58 (41.43%)	10 (26.32%)	IBM vd DM $p = 0.013$
Reflux disease	98 (25.99%)	30 (24.39%)	18 (30.00%)	37 (26.43%)	11 (28.95%)	ns
Hypothyroidism	60 (15.92%)	23 (18.70%)	12 (20.00%)	17 (12.14%)	4 (10.53%)	ns
Diabetes mellitus type 2	43 (11.41%)	10 (8.13%)	8 (13.33%)	20 (14.29%)	5 (13.16%)	ns
Asthma	35 (9.28%)	16 (13.01%)	6 (10.00%)	10 (7.14%)	1 (2.63%)	ns
Arrhythmia	34 (9.02%)	8 (6.50%)	5 (8.33%)	17 (12.14%)	3 (7.89%)	ns
Cancer	30 (7.96%)	7 (5.69%)	3 (5.00%)	15 (10.71%)	4 (10.53%)	ns
Coronary artery disease/ ischemic heart disease	20 (5.31%)	6 (4.88%)	4 (6.67%)	10 (7.14%)	0 (0.00%)	ns
Chronic obstructive pulmonary disease	16 (4.24%)	4 (3.25%)	3 (5.00%)	9 (6.43%)	0 (0.00%)	ns
Heart failure	11 (2.91%)	3 (2.44%)	6 (10.00%)	2 (1.43%)	0 (0.00%)	ns
Venous thrombosis/pulmonary embolism	11 (2.92%)	5 (4.07%)	3 (5.00%)	3 (2.14%)	0 (0.00%)	ns
Interstitial lung disease	9 (2.39%)	5 (4.07%)	1 (1.67%)	0 (0.00%)	3 (7.89%)	ns
Hyperthyroidism	6 (1.59%)	3 (2.44%)	1 (1.67%)	2 (1.43%)	0 (0.00%)	ns
Osteoarthritis	5 (1.33%)	2 (1.63%)	1 (1.67%)	2 (1.43%)	0 (0.00%)	ns
Pulmonary hypertension	5 (1.33%)	1 (0.81%)	1 (1.67%)	0 (0.00%)	3 (7.89%)	ns
Diabetes mellitus type 1	4 (1.06%)	1 (0.81%)	2 (3.33%)	1 (0.74%)	0 (0.00%)	ns
Depression, anxiety, post-traumatic stress disorder	4 (1.06%)	4 (3.25%)	0 (0.00%)	0 (0.00%)	0 (0.00%)	ns
Osteoporosis	3 (0.80%)	1 (0.81%)	0 (0.00%)	1 (0.71%)	1 (2.63%)	ns

DM dermatomyositis, *PM* polymyositis, *IBM* inclusion body myositis, *MwC* myositis with other connective tissue disorders, *ns* not statistically significant

1.9 ± 0.80 vs mean 1.20 ± 0.72; $p < 0.001$) and MwC (mean 1.9 ± 0.80 vs mean 1.08 ± 0.80 $p < 0.001$) (Fig. 1a). Similarly, SDI score was significantly higher in IBM patients as compared to DM (mean 2.01 ± 0.76 vs mean 0.92 ± 0.69; $p < 0.001$), as compared to PM (mean 2.01 ± 0.76 vs mean 1.33 ± 0.73; $p < 0.001$) and MwC (mean 2.01 ± 0.76 vs mean 1.24 ± 0.82; $p < 0.001$) (Fig. 1b). Depending on SDI scores we assessed the degree of disability for each patient, following the instructions as explained in the "Methods" section. More than half of the patients diagnosed with IBM fulfilled the criteria of severe to very severe disability, while 87.41% of patients with IBM and 65% of patients with PM

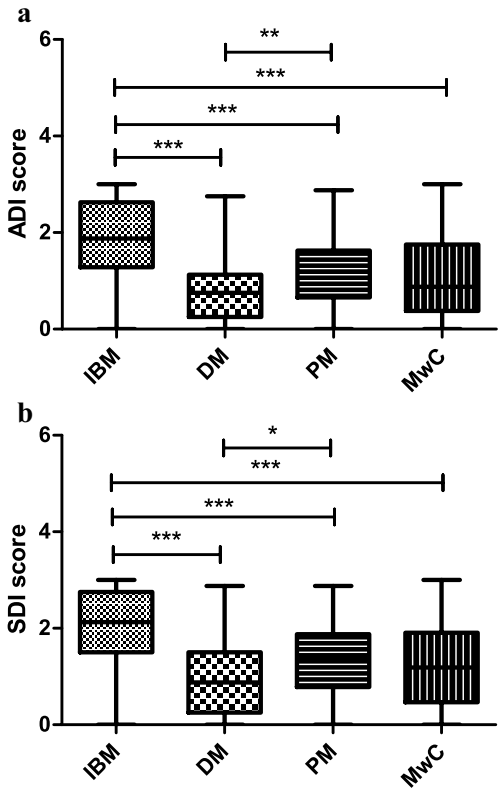

Fig. 1 ADI and SDI depending on the diagnosis. The figure presents mean values of standard and alternative disability indexes assessed with Health Assessment Questionnaire and illustrates statistically important differences between groups. *ADI* alternative disability index, *SDI* standard disability index, *IBM* inclusion body myositis, *DM* dermatomyositis, *PM* polymyositis, *MwC* myositis with other connective tissue disorders, ***$p < 0.001$; **$p < 0.01$; *$p < 0.05$

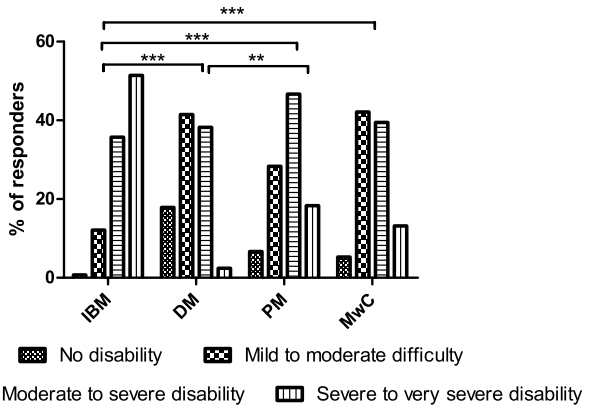

Fig. 2 Severity of disability depending on the diagnosis. The figure presents percentage of patients with different subtypes of myositis who suffer from particular severity of disability evaluated with Health Assessment Questionnaire as well as illustrates statistically important differences between groups. *IBM* inclusion body myositis, *DM* dermatomyositis, *PM* polymyositis, *MwC* myositis with other connective tissue disorders, ***$p < 0.001$; **$p < 0.01$

experienced at least moderate limitations. On the contrary, patients with DM presented rather lower degrees of disability, as almost 60% of them fulfilled the criteria of mild to moderate or no disability while moderate or worse limitations were twice as rare as in IBM group. Vast majority of patients with MwC presented either mild to moderate difficulties or moderate to severe disability (Fig. 2).

Patients with IIM most frequently required walking devices (45.62% of IIM patients), and 43.50% of responders declared using facilitating devices for hygiene activities. Aids to facilitate arising, gripping/opening and reaching were also frequently exploited, respectively, by 28.38, 24.67 and 19.36% of patients. Devices designed for easier dressing and eating were rarely required (respectively, by 10.61% and 7.16% of patients with IIM).

58.81% of patients declared they need aid from other people while gripping and opening things. Almost half (48.81%) of the patients depended on caregivers with activities such as running errands and shop, getting in and out of the car or chores such as vacuuming, housework or light gardening. Approximately 1/3 of IIM patients required help with reaching objects, 19.89% needed assistance with dressing and 17.51% with arising. Help with walking, maintaining hygiene and eating was required by, respectively, 16.18, 10.08 and 5.57% of IIM patients. Detailed data concerning the need to use facilitating devices and aid from caregivers are presented in Table 2.

Patients with IBM presented lower mean values of pain related to myositis (assessed by VAS) as compared to DM (mean 21.96 ± 26.58 vs mean 36.94 ± 27.62; $p < 0.001$), as compared to PM (mean 21.96 ± 26.58 vs mean 38.87 ± 28.65; $p < 0.001$) as well as to MwC (mean 21.96 ± 26.58 vs mean 37.97 ± 32.47; $p < 0.05$).

Majority of patients with PM, DM and MwC declared muscle pain for most days in a week, while myalgia was not as frequent in patients with IBM. Mean value of myalgia, assessed in 1–10 scale was the lowest for patients with IBM as compared to DM (mean 2.5 ± 1.82 vs mean 4.01 ± 2.05; $p < 0.001$), as compared to PM (mean 2.5 ± 1.82 vs mean 3.92 ± 2.20; $p < 0.001$) and MwC (mean 2.5 ± 1.82 vs mean 4.03 ± 2.23; $p < 0.001$) (Fig. 3a). Majority of IIM patients declared a feeling of muscle weakness for most days in a week. Feeling of muscle weakness, evaluated in 1–10 scale was the highest in patients with IBM (6.97 ± 1.92) and the lowest in patients with DM (4.41 ± 2.16). Patients with PM and MwC obtained mean values of, respectively, 5.57 ± 2.36 and 5.81 ± 1.98. We found statistically significant differences between IIM subtypes ($p < 0.001$ between IBM and DM and between IBM and PM; $p < 0.05$ between IBM and MwC, DM and PM, DM and MwC) (Fig. 3b).

Table 2 Usage of facilitating devices and aid of caregivers in patients with different subtypes of myositis

	IBM $n=140$	DM $n=123$	PM $n=60$	MwC $n=38$	p
Facilitating devices					
Dressing and grooming devices	30 (21.43%)	4 (3.25%)	2 (3.33%)	3 (7.89%)	ns
Arising devices	83 (59.29%)	7 (5.69%)	11 (18.33%)	5 (13.16%)	IBM vs DM < 0.001 IBM vs PM < 0.001 IBM vs MwC < 0.001
Eating devices	21 (15.00%)	3 (2.44%)	1 (1.67%)	2 (5.26%)	ns
Walking devices	110 (78.57%)	23 (18.70%)	23 (38.33%)	11 (28.95%)	IBM vs DM < 0.001 IBM vs PM < 0.001 IBM vs MwC < 0.001
Hygiene devices	98 (70.00%)	29 (23.58%)	18 (30%)	13 (34.21%)	IBM vs DM < 0.001 IBM vs PM < 0.001 IBM vs MwC < 0.001
Reaching devices	49 (35.00%)	11 (8.94%)	7 (11.67%)	4 (10.53%)	IBM vs DM < 0.001
Gripping and opening devices	50 (35.71%)	21 (17.07%)	6 (10.00%)	13 (34.21%)	IBM vs PM < 0.001
Aid needed from caregivers					
Dressing	41 (29.29%)	13 (10.57%)	10 (16.67%)	7 (18.42%)	IBM vs DM < 0.001 IBM vs PM < 0.05
Arising	39 (27.86%)	8 (6.50%)	12 (20.00%)	5 (13.16%)	IBM vs DM < 0.001
Eating	17 (12.14%)	1 (0.81%)	0 (0.00%)	2 (5.26%)	IBM vs DM < 0.001 IBM vs PM < 0.005
Walking	32 (22.86%)	12 (9.76%)	10 (16.67%)	6 (15.79%)	IBM vs DM < 0.005
Hygiene	21 (15.00%)	6 (4.88%)	6 (10.00%)	3 (7.89%)	IBM vs DM < 0.01
Reaching	56 (40.00%)	33 (26.83%)	22 (36.67%)	11 (28.95%)	IBM vs DM < 0.02 IBM vs MwC < 0.001
Gripping and opening	94 (67.14%)	53 (43.09%)	28 (46.67%)	19 (50.00%)	IBM vs DM < 0.001 IBM vs PM < 0.006 IBM vs MwC < 0.05
Errands and chores	84 (60.00%)	43 (34.96%)	29 (48.33%)	20 (52.63%)	IBM vs DM < 0.001

IBM inclusion body myositis, *DM* dermatomyositis, *PM* polymyositis, *MwC* myositis with other connective tissue disorders, *ns* not statistically significant

On average, patients suffering from different subtypes of myositis presented a similar, decreased tolerance of physical activity assessed with VAS (mean values, respectively, 6.90 ± 2.33 for IBM, 6.84 ± 2.19 for DM, 6.97 ± 2.67 for PM and 6.63 ± 2.26 for MwC; $p = 0.7592$).

Study participants presented increased degree of fatigue, which was significantly more profound in DM patients as compared to IBM (mean 6.70 ± 1.80 vs mean 5.93 ± 2.08; $p < 0.05$). Mean value observed in PM patients was 6.43 ± 2.29 and in MwC 6.58 ± 2.21; however, differences between remaining subtypes were insignificant.

Patients with disease duration longer than 5 years had significantly higher indexes of disability (both ADI and SDI) as compared to patients with shorter disease duration ($p = 0.0002$). In post hoc analysis, statistically significant difference was found between the group of patients with disease duration of more than 5 years and the group of patients with disease duration of 1–3 years ($p = 0.0064$) as well as between the group of patients with disease duration of more than 5 years and the group of patients with disease duration of 6–12 months ($p < 0.05$ for ADI, $p < 0.05$ for SDI).

Moreover, we demonstrated a statistically significant linear correlation between ADI score and declared feeling of muscle weakness ($p < 0.001$, $r^2 = 0.4774$) (Fig. 4 in supplementary data). No correlation was stated between ADI and degree of fatigue ($p < 0.05$, $r^2 = 0.028$), ADI and tolerance of physical activity ($p < 0.001$, $r^2 = 0.144$), SDI and declared intensity of fatigue ($p < 0.001$, $r^2 = 0.031$) or between SDI and tolerance of physical activity ($p < 0.001$, $r^2 = 0.113$).

Discussion

Sporadic IBM is considered as the most distinctive and severe out of IIM subtypes, usually associated with poorer prognosis and greater risk of disability, although the lifespan is comparable to general population [1, 9]. Our results indicate that patients with IBM are much more disabled as compared to other subtypes of IIM. It is in line with the

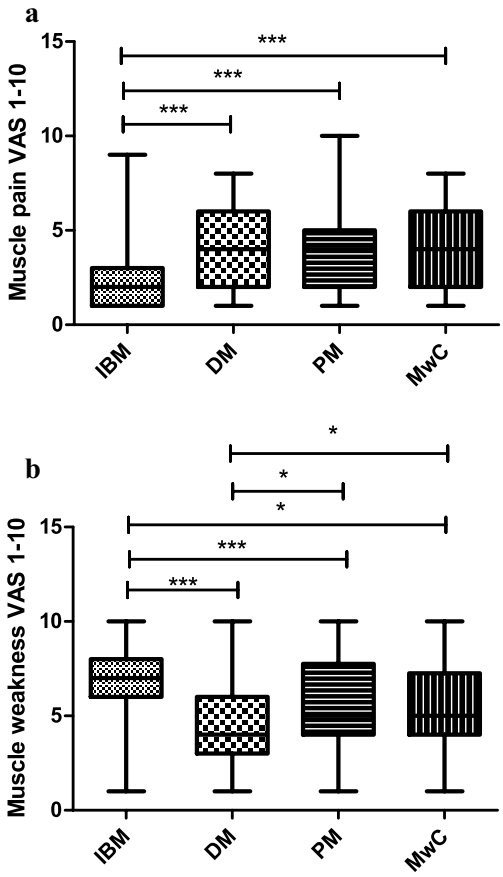

Fig. 3 Actual muscle pain and feeling of muscle weakness in scale from 1 to 10 depending on the diagnosis. The figure presents mean values of declared myalgia (**a**) and muscle weakness (**b**), assessed in scale from 1 to 10 and illustrates statistically significant differences between groups of patients with particular subtype of myositis. *VAS* visual analogue scale, *IBM* inclusion body myositis, *DM* dermatomyositis, *PM* polymyositis, *MwC* myositis with other connective tissue disorders, ***$p < 0.001$; **$p < 0.01$; *$p < 0.05$

study based on population enrolled in EuroMyositis Registry which displayed that patients with IBM, dysphagia and cardiac involvement obtained higher HAQ-DI scores [1]. In our study walking aids were the most frequently used facilitating devices. Noteworthy, we showed that such devices were mostly needed by patients with IBM. It corresponds to observations previously stated by other researchers. In a long-term study on 136 patients, walking difficulties affected 75.73% of patients with sporadic IBM, more than 1/3 of them declared using wheelchair and the median period between first symptoms and the need of wheelchair usage was estimated as 14 years [10]. In a 12-year follow-up of a Dutch cohort, muscle strength decreased significantly during observation period, forcing all of the survived patients to use wheelchair [9, 10]. In a study by Cox et al. the average period between the first symptoms of IIM and the need of using walking stick lasted 11 ± 5 years while wheelchair was necessary after approximately 16 ± 4 years [9]. In another long-term observation, 82% of IBM survivals needed to use assistive device; faster progression of disablement was observed in individuals with late-onset disease [11]. Since previous studies usually concentrated on assessing degree of disability in general or only walking difficulties, we did not manage to find precise data covering the need of using other facilitating devices. Little was known about usage of facilitating devices by patients with subtypes of myositis other than IBM. In our survey, we displayed that patients with IBM encounter more difficulties with daily duties as they needed to use devices for arising, eating or dressing more frequently than responders affected by other subtypes.

However, all IIM subtypes can lead to progressive disability. Bronner et al. demonstrated that almost a quarter of patients with PM and DM fulfilled the criteria of considerable disability (measured by Rankin score), while only 34% of patients were not affected or only slightly disabled [12]. Regardless of the IIM subtype, in a follow-up of 62 myositis patients vast majority experienced difficulties with proceeding daily duties and more than half of them depended on help from other people [13]. At the end-point of the study disability was observed in 68% of examined patients [13].

In our study, moderate, severe or very severe disability affected 40.65% of DM patients and even 65% of patients with PM. We noted higher median rates of HAQ-DI scores in both DM and PM group than it was previously reported in literature. The median HAQ-DI score observed in Hungarian cohort of PM and DM patients was 0.875 with majority of them fulfilling the criteria of mild or moderate disability; however, HAQ-DI scores were incomparably higher if the course of the disease was relapsing-remitting or chronic-progressive [3]. Analysis of possible risk factors showed that females, patients with a polyphasic and chronic course of the disease as well as with concomitant osteoporosis or longer duration of myositis could be associated with greater disability [3]. In our study comorbidities had no statistically significant impact on the limitations and the necessity to use assistive devices.

Surprisingly, in a single Chinese follow-up majority of patients with IIM developed no disability at all [14]. Those promising outcomes could indicate the impact of some yet unknown factors in developing disability that are potentially possible to avoid. Lower mean rate of HAQ score (0.6) in patients with IIM was also obtained in a smaller study by Neri et al.; however, the values were higher in individuals with moderate to severe muscle weakness [15]. This observation is in line with our analysis, as we found a statistically significant correlation between the feeling of muscle weakness and ADI. Therefore, significantly higher HAQ-DI scores in patients with IBM could result from greater intensity of declared muscle weakness rather than subtype of IIM itself. In our study patients with IBM were statistically

older than patients with other subtypes, what might have an impact on higher level of disability. We found patients with PM to be more disabled than patients with DM. As cutaneous symptoms usually accelerate the correct diagnosis, it can be assumed that patients with DM were diagnosed and treated earlier than patients with PM, yet the question about the duration period from first symptoms to diagnosis was not included in our survey. We noted that patients with PM presented higher subjective muscle weakness (4.41 ± 2.16 in DM vs 5.57 ± 2.36 in PM) what along with the correlation stated between ADI and declared feeling of muscle weakness could explain greater disability observed in PM patients as compared to DM.

Interestingly, we found that patients with IBM presented the lowest level of fatigue, but still obtained the highest HAQ-scores. Fatigue is now considered to have a multifactorial aetiology including inflammatory processes [16]. Pathogenesis of IBM, yet not clearly stated, undoubtedly differs from other subtypes of IIM, as apart from inflammation also muscle degeneration contribute to clinical outcome [17]. Unclear role of immune-mediated reactions may explain to some extent lower degrees of fatigue, observed in IBM patients as compared to other subtypes.

Study limitations and advantages

The main disadvantage of our research is the inability to verify the diagnoses as the study was questionnaire-based internet-administered survey with self-reported diagnosis. As the survey was in a form of an online questionnaire it was impossible to verify the diagnoses by means of medical examination. The risk of bias was limited by selecting community forums and support groups that are restricted only to patients with myositis and no other musculoskeletal disorders. In addition we included the answer "I don't know" to a question about subtype of the disease in order to exclude patients unsure about their subtype of myositis.

As the questionnaire was answered by patients from different continents and countries the results might have been influenced by cultural and ethnic differences, as well as discrepancies in the IIM classification criteria, diagnosis and therapy in different regions. Since the questionnaire was posted online we are not able to exclude the possibility that it was filled by individuals with weaker knowledge of English language, which could result in inaccurate understanding of the content of the survey. Besides, we are not able to verify how many people have seen the questionnaire and how many of those who have seen it were diagnosed with IIM. Therefore, we are not able to provide a response rate. Moreover, the disease activity of the individuals was not established; hence observed differences might be a result of different inflammatory activity. Reliable evaluation of disease activity requires a physical examination and/or additional tests (according to Myositis Disease Activity Assessment Tool) and, therefore, cannot be stated based only on self-reported symptoms. HAQ, when performed as an isolated tool, does not cover all areas included in the International Classification of Functioning, Disability and Health. Although it provides a detailed assessment in the field of activity limitation and to some extent participation restriction it does not assess impairment resulting from body functions and structures and also neglects the importance of environmental and personal factors [18]. Moreover, it does not include social or mental health as well as sensory organ impairment [6]. Researchers highlight also that HAQ does not discriminate between disease activity and disease damage, what could impede the evaluation in patients with moderate to severe damage, especially if they present irreversible musculoskeletal lesions [5]. In addition, it was validated so far for the most common subtypes of IIM including DM and PM, but further validation is still needed for patients with IBM [5, 7].

The main advantage of our research is that the study group is diverse and numerous, enabling to state comparisons and illustrate both the similarities and discrepancies between IIM subtypes. In addition, we evaluated the degree of disability using the HAQ, which is recommended in IIM patients by scientific societies and displays the limitations caused by IIM from the patients' perspective. Patient-oriented approach ensures better understanding of the disease by focusing on aspects that are truly vital for the patients. Noticing patients' perspective is necessary to improve their quality of life, which remains one of the most important goals in the therapeutic process of individuals with chronic diseases. In our study we compared patients with DM, PM, IBM and MwC, while most previous studies limited the study groups only to one or two subtypes of IIM. To our knowledge, usage of facilitating devices has never been studied as detailed as in our study.

Conclusions

Patients with IBM suffer most from progressing disability as compared to other subtypes of myositis despite relatively low level of myalgia and fatigue. Feeling of muscle weakness is the highest in IBM patients and affects significantly everyday life, resulting in poorer functioning and progressive loss of independence, reflected by the widespread need to use special utensils or aid from other people.

Noteworthy, although many of IIM patients used facilitating devices during activities such as walking, hygiene or arising, they rarely required additional aid from caregivers while performing those actions, what indicates the invaluable impact of such devices on maintaining independence. Our results reflect the urgency to search for novel methods of treatment and rehabilitation as well as the need to invent

more suitable aids for this group of patients in order to maintain the quality of life and physical functioning at the highest possible level.

Author contributions AHO was responsible for the design of the work, data acquisition, drafting the work, approval of the final version to be published and agreed to be accountable for all aspects of the work in ensuring that questions related to the accuracy or integrity of any part of the work are appropriately investigated and resolved. OEB was responsible for analysis and interpretation of data for the work, drafting the work, approval of the final version to be published and agreed to be accountable for all aspects of the work in ensuring that questions related to the accuracy or integrity of any part of the work are appropriately investigated and resolved. JSM was responsible for the conception of the work, interpretation of the data for the work, critical revising of the article for important intellectual content, approval of the final version to be published and agreed to be accountable for all aspects of the work in ensuring that questions related to the accuracy or integrity of any part of the work are appropriately investigated and resolved.

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Compliance with ethical standards

Conflict of interest Authors declare no conflict of interest.

Ethical approval All procedures performed in studies involving human participants were in accordance with the ethical standards of the institutional and/or national research committee and with the 1964 Helsinki declaration and its later amendments or comparable ethical standards.

Human and animal rights statement This article does not contain any studies with animals performed by any of the authors.

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