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DISABILITY IN PATIENTS WITH IDIOPATHIC INFLAMMATORY MYOPATHIES

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Abstract

Introduction. Idiopathic inflammatory myopathies (IIMs) are a heterogeneous group of disorders, characterized by chronic muscle weakness, low muscle endurance and by the presence of inflammatory cell infiltrates in muscle tissue, with organ damage and disability.
The objective of the study was to determine the degree of disability in a Moldavian cohort of patients with idiopathic inflammatory myopathies, by Rankin's scale.
Methods. We performed a cross-sectional study, from December 2015 to December 2018, in which were included patients with IIMs. Demographic and clinical data were collected using a special questionnaire, including employment status, upholding dose of corticosteroids. In order to estimate the degree of functional disability at the time of the research, we applied the modified Rankin's scale, with possible scores from 0-no disability to 5-totally dependent.
Results. 65 IIMs patients were enrolled in the study. Male to female ratio was 1:3.3, the patients' mean age was 50.2±11.7 years, the mean disease duration 95.2±6.89 months. According to modified Rankin's

Résumé

L'invalidité chez les patients aux myopathies inflammatoires idiopathiques

Introduction. Les myopathies inflammatoires idiopathiques (MII) sont un groupe hétérogène de troubles caractérisé par une faiblesse musculaire chronique, une faible endurance musculaire et des infiltrats de cellules inflammatoires dans les tissus musculaires, avec des lésions organiques et des conséquences multiples et invalidantes.
Le but de l'étude était de déterminer le degré d'invalidité chez les patients atteints de myopathies inflammatoires idiopathiques selon l'échelle de Rankin dans une cohorte moldave.
Méthodes. Nous avons effectué une étude transversale de décembre 2015 à décembre 2018, dans laquelle ont été inclus des patients avec des MII. Les données démographiques et cliniques ont été collectées à l'aide d'un questionnaire spécifique, indiquant notamment le statut d'emploi et la dose d'entretien de corticostéroïdes. Pour estimer le degré d'incapacité fonctionnelle et d'invalidité au moment de l'enquête, nous avons appliqué l'échelle de Rankin modifiée, avec des scores

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scale, the 1st degree was found in 20 cases (30.77%), the 2nd degree in 24 patients (36.92%), 11 patients (16.92%) had 3rd degree, 8 patients (12.31%) 4th degree, and 5th degree was determined in 2 patients (3.08%). Many variables were analysed and we determined that the dose of prednisolone-equivalent <10 mg (RR 2.2; 95% CI 1.34 to 3.6, p<0.01) and disease duration less than 2 years are protective factors (RR 0.4; 95% CI 0.2 to 0.9, p<0.05) for severe degree of disability (≥ 3).

Conclusion. Our study has shown that patients with idiopathic inflammatory myopathies have a mild disability in 67.69% of the cases, moderate in 16.92% and severe disability was found in 15.39% of the cases. The dose of prednisolone-equivalent <10 mg and disease duration less than 2 years were determined as protective factors against the severe degree of disability.

Keywords: idiopathic inflammatory myopathies, disability, modified Rankin's scale.

possibles commencement de 0 (incapacité nulle) à 5 points (dépendance totale).

Résultats. Dans l'étude, ont été inclus 65 patients avec MII, le rapport hommes / femmes a été de 1: 3,3, l'âge moyen des patients était de 50,2 ±11,7 ans et la durée moyenne de la maladie de 95,2±6,89 mois. Selon l'échelle de Rankin modifiée, le 1er degré a été identifié dans 20 cas (30,77%), le 2-ème degré chez 24 sujets (36,92%), 11 patients (16,92%) avaient le 3-ème degré, le 4ème - 8 patients (12,31%) et le 5-ème, invalidité sévère, chez 2 patients (3,08%). Nous avons analysé plusieurs variables et on a établi que le dosage de l'équivalent de la prednisolone <10 mg (RR 2.2, IC à 95% de 1,34 à 3,6, p≤0,01) et la durée de la maladie moins de 2 ans (RR 0,4; IC 95% 0,2 à 0,9, p≤0,05) sont des facteurs de protection pour un degré d'invalidité plus sévère (Rankin≥ 3 points).

Conclusion. Les patients atteints de myopathies inflammatoires idiopathiques présentaient une incapacité légère dans 67,69% des cas, une incapacité modérée en 16,92% des cas et une incapacité grave dans 15,39% des cas. Une dose équivalente à environ 10 mg d'équivalent de la prednisolone et une durée de la maladie inférieure à 2 ans ont été identifiées comme des facteurs de protection pour les degrés plus sévères d'invalidité.

Mots-clés: myopathies inflammatoires idiopathiques, invalidité, échelle modifiée de Rankin.

Introduction

The idiopathic inflammatory myopathies (IIMs) are a heterogeneous group of disorders, characterized by chronic muscle weakness, low muscle endurance, and inflammatory cell infiltrates in muscle tissue, that may lead to significant disability and reduced quality of life[1]. Although IIMs are regarded as treatable disorders, the prognosis is not well established[2-3]. Although in the literature it has been suggested that the mortality has been greatly reduced, many patients with myopathies continue to exhibit physical disability and multiple organ damage[4-6]. According to the World Health Organization, disability is defined as impairments, activity limitations, and participation restrictions. Disability is not only a health problem. It is a complex phenomenon, reflecting the interaction between the features of a person's body and features of the society in which he or she lives. Evidence suggests that people with disabilities face barriers to accessing the health and rehabilitation services they need in many settings[7]. There are few published data regarding disability in patients with IIMs. The first who applied the modified Rankin's scale in this type of patients was Bronner et al, in 2006[3]. In their research, the authors concluded that 34% of the patients had none or mild disability and that it was associated with male sex (OR 3.1; 95% CI 1.2 to 7.9). Disability assessment through modified Rankin's scale was also an objective set by Shu et al, in 2011; they determined that 83.4% of their patients had no disability[2]. In another study, the authors found a statistically significant correlation between Modified Rankin's Scale and the number of muscles with fat infiltration (r = 0.48; p = 0.03), meaning that the higher the number of muscles with fat infiltration, the worse was the clinical and functional outcome[8]. The assessment of disability in the Republic of Moldova is done by the National Commission for Evaluation of Disability and Work Capacity only according to criteria that cover 7 domains; however, these criteria are general and not disease-specific. The modified Rankin's scale is used as one of a set of measures to capture the disease burden for patients with IIMs, in addition to validated measures of disease activity, disease damage and quality of life[9-11,12].

The objective of the study was to determine the degree of disability in a Moldavian cohort of patients with IIMs, by Rankin's scale.

Materials and methods

We performed a cross-sectional study, from December 2015 to December 2018. The inclusion criteria were: adult patients with IIMs, disease duration more than 6 months, signed informed consent. The exclusion criteria were myopathies of other etiology. Demographic and clinical data were collected using a special questionnaire, including upholding dose of corticosteroids. The study group was divided into two subgroups, by disease duration: group 1 – less than 24 months and group 2, with duration more than 2 years. In order to estimate the degree of functional disability at the time of the research, we applied the modified Rankin's scale with possible scores 0 – no disability, 1 – without significant disability despite symptoms or symptoms did not interfere with their normal daily activity, 2 – mild disability or inability to perform all previous activities, but he can take care of himself without help, 3 – moderate disability, the symptoms significantly restrict the patient's usual activities and prevent him from having a completely independent life (but he can walk without help), 4 – moderately-severe disability, manifested through the inability to have an independent life (cannot walk without help, cannot handle personal needs without assistance), but does not require permanent care, 5- severe disability with bedridden, requiring permanent day and night care, totally dependent[13]. The professional situation was considered the state of employment at the time of assessment and categorized as follows – full time, part-time, unemployed, in training, housewife, disability retirement according to Moldavian disability criteria. In order to assess the work productivity, we used Work Productivity and Activity Impairment: General Health (WPAI:GH) questionnaire, that was administered only to employed subjects. The quality of life was determined using SF-8 with 2 domains: physical and mental. The study was performed according to the Declaration of Helsinki for human rights. The statistical processing of the data was done through the MedCalc v.1.2 and Excel program, the arithmetic mean and the standard deviation were calculated. The t-Student test was used to test the statistical difference. The correlation analysis was performed by Pearson's test and relative risk with 95 % confidence interval. Values of $p < 0.05$ were considered statistically significant.

Results

65 patients with idiopathic inflammatory myopathies who met the inclusion criteria for the study were enrolled. Demographic parameters are included in Table 1.

Table 1. The general characteristics of patients with IIMs.

Variables	*IIM patients, n=65*
Sex	
Female	50 (75.38 %)
Male	15 (24.62 %)
Mean age at the time of research, years	50.2±11.7 (range 25-78)
Mean disease duration, months	95.2±68.9 (range 6-324)
Marital status	
Married	60 (92.31 %)
Widower	2 (3.07 %)
Divorced	1 (1.53%)
Bachelor	2 (3.07%)
Educational years	
< 9	4 (6.15 %)
9-12	29 (44.62 %)
>12	32 (49.23 %)
Employee status	
Full-time	9 (13.85 %)
Part-time	7 (10.77 %)
Unemployed	7 (10.77 %)
In training	10 (15.38 %)
Housewife	6 (9.23 %)
Disability retirement	26 (40.00 %)
Dose of corticosteroids*	
<10 mg	55 (87.30%)
≥10 mg	8 (12.70 %)
Drug-induced remission	21 (32.31 %)
Remission without drugs	2 (3.07 %)

*dose of equivalent-prednisolone

Data presented in Table 1 show the prevalence of women, with a female to male ratio of 3.3:1. The mean age was 50.2 years, with a range from 25 to 78 years. The disease duration varied from 6 months to 324 months, in average 95 months. Regarding the matrimonial status of 65 patients, we found that at the time of the research 60 (92.31%) patients were married, 2 (3.07 %) subjects were bachelor, widows and divorced, living alone – 2 (3.07%) and 1 (1.53%), respectively. The analysis of educational years in patients with IIMs revealed the variation from 9 to 23 years, 4 (6.15 %) patients had 9 years of study, 29 (44.62%) – college, 32 (49.23%) with university studies, including 5 (7.69 %), with master degree. We analyzed the employee status in the IIM group and found that 9 (13.85%) of 65 patients were full-time employed, 7 (10.77%) part-time, 7 (10.77%) cases unemployed, 10 (15.38%) subjects in training, 6 (9.23 %) housewives and 26 (40.0%) had already disability retirement by Moldavian criteria and did not work at the time of the research. It was determined that 7 (10.77%) patients had monocyclic disease course, when the patient remained in remission (no detectable clinical or biochemical disease activity) after 24 months since diagnosis. In 42 (64.61%) cases,

it was found a polycyclic disease course, when the patient had recurrence of disease activity (determined by clinical or biochemical parameters), and 17 (24.62%) cases were chronically continuous, when there was persistent disease or continuation of drugs beyond 24 months after diagnosis. The basic treatment of IIMs included administration of corticosteroids (63 patients), methotrexate (10 patients), azathioprine (3 patients) and hydroxychloroquine (7 patients). Referring to corticosteroids' administration, the maintenance dosage varied from 5 to 60 mg prednisolone-equivalent per day, the mean dose was 10.26±9.42 mg. To be noted that in 87.3% of cases the dosage of prednisolone-equivalent was <10 mg/day. Disease remission induced by drugs was determined in 21 (32.31%) patients with IIMs and only 2 (3.07%) subjects had remission without drugs.

According to the data available in the literature, disability was found to be an important consequence of the disease, confirmed by the results of the Rankin's scale. In the study group, disability 0 degree (signifying the absence of symptoms) has not been identified. Twenty patients (30.77%) had 1st degree – without significant disability, symptoms or symptoms did not interfere with their normal daily activity. Mild disability or inability to perform all previous activities, but he/she can take care of himself without help (2nd degree), was identified in 24 (36.92%) subjects. Eleven (16.92%) patients had a moderate disability, the symptoms significantly restricted the patient's usual activities and prevent him from having a completely independent life (but can walk without help). Moderately-severe disability (4th degree) manifested through the inability to have an independent life (cannot walk without help, cannot handle personal needs without assistance), but does not require permanent care, was appreciated in 8 (12.31%) patients. Severe disability with bedridden, requiring permanent day and night care (5th degree), respectively, was determined in 2 (3.08%) patients. To be noted that degree 6, assigned to death, was not found at the time of examination. We were interested to analyse Rankin's disability depending on the duration of the disease (Figure 1).

The data presented in Fig. 1 reveal that patients with 1st degree prevailed in the group with disease duration less than 2 years, which was found in 37.5% versus 28.57 % in group II. It is noteworthy that, in the second group, patients with the second degree of disability predominated, in 38.78% of cases versus 31.25% in group I. 3rd degree disability was appreciated in 18.75% and 16.32% cases from group I and II, respectively. It should be noted that the 4th degree of disability was encountered in 6.25% of patients from the group I, compared to 14.29% of patients from the group with disease duration of more than 2 years. Severe disability, 5th degree, was determined in 6.25% of cases in the group with the duration of the illness under 2 years.

We continued the analysis of the obtained data and established that from the 9 patients employed full-time 6 (37.5%) of them had a 1st degree on Rankin's scale and 3 (18.75%) second degree. Of 7 IIM patients who were part-time employed, 4 (25.0%)

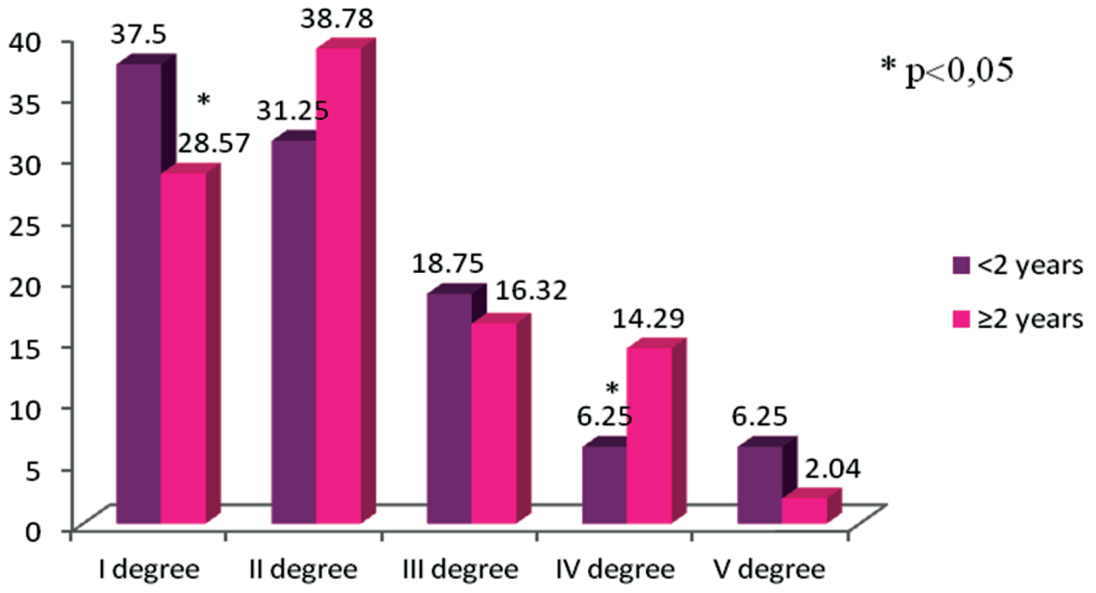

Fig. 1. Disability according to the Rankin's scale depending on the duration of the disease.

had a first degree, 5 (31.25%) second degree and in one case 3rd degree by Rankin. With regard to employed patients, we were interested to study work productivity through WPAI. Analyzed data revealed that patients were absent from work during the last week from 0 to 35 hours, representing 0-86.5% of working hours (absenteeism) with an average of 17.06±4.7%. At the same time, presenteeism – the loss of productivity in hours at work – was 26.47±9.3%, with varying ranges from 0 to 28 hours, which makes up 0-70% of cases. The loss of general productivity was 36.19±28.09 (i-v 1-93.25%) cases. Regarding the activity limitation outside the workplace, it was 28.82±22.33%, with a variation interval of 0 to 80% of cases. To be noted that, at the time of research, 26 (40.0 %) patients had disability retirement according to national disability criteria; of them, 2 (7.69%) had 1st degree, 7 (26.92%) second degree, 10 (38.46%) third degree, 6 (23.08%) 4th degree and one patient had 5th degree on the Rankin's scale.

The data presented in Fig. 2 demonstrate the distribution of the quality of life values in relation to the average values, with a wider distribution of the mental component values than the physical one. The mean physical component score was 41.69 and the mental 36.48, which represents a reduced quality of life in patients with myopathies, mostly through the physical component.

We analyzed multiple variables in order to establish correlation relationships and it was determined a strong correlation between disability degree and the upholding dose of prednisolone-equivalent ($r=0.76$, $p<0.001$) and moderate correlation with disease duration ($r=0.57$, $p<0.001$). Interested by these correlations, we analysed the relative risk and we determined that dose of prednisolone-equivalent <10 mg (RR 2.2; 95% CI 1.34 to 3.6, $p<0.01$), and disease duration less than 2 years are protective factors (RR 0.4; 95% CI 0.2 to 0.9, $p<0.05$) for severe degree of disability (≥ 3).

Discussion

According to our study, patients with IIMs have diverse degrees of disability according to modified Rankin's scale. In most of IIMs patients, we determined a second degree of disability, this corresponding with some data available in the literature. Bronner et al found that 34% of the IIMs patients had none or only a mild disability (0-I degree)[3]. An interesting finding was revealed by Shu et al; they stated that 84% of their patients had no disability and only 16% had diverse degrees on Rankin's scale[2]. According to Suzuki et al, after 2-years of treatment, 22 (27%) of 81 patients had modified Rankin's scale scores of 3–5[13]. A recent study done by Guimaraes et al found second degree on Rankin's scale in 33.33%,

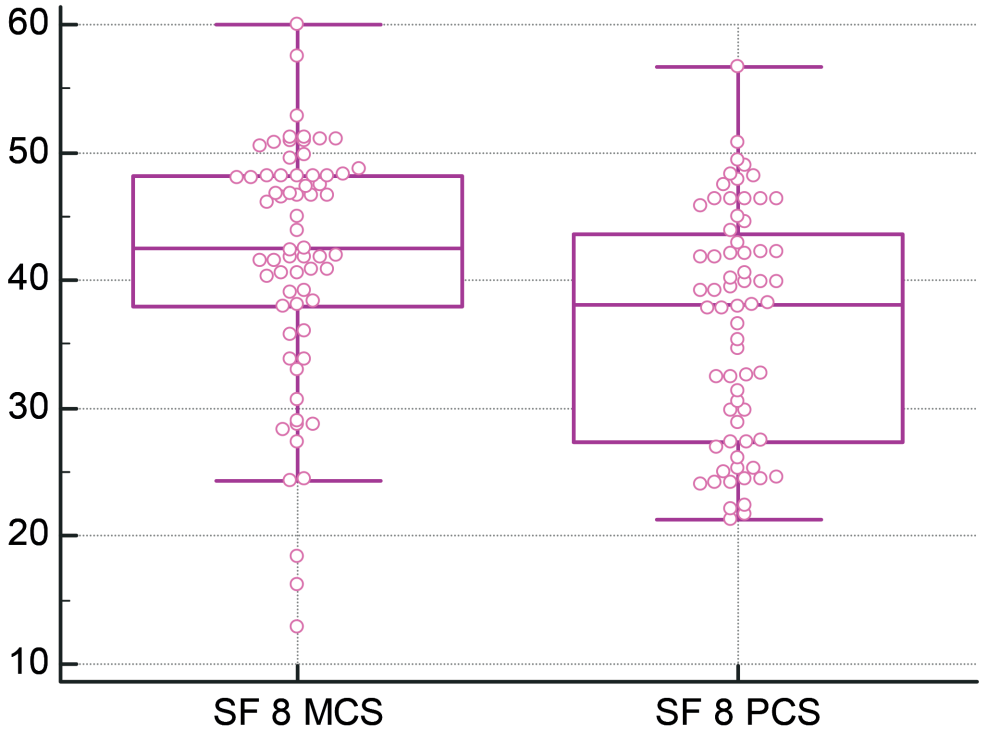

Fig. 2. Quality of life in patients with IIMs.

third degree in 41.67 % and fourth degree in 25% of patients, these data being different from the one identified in our cohort[8]. The degree of disability is related to another organ involvement[14,15].

Most of the patients from our study had a chronic continuous or polycyclic disease course. In the long term, 87.3% of the patients were still taking a low dosage of drugs, and 3.08 % of the subjects had significant disability. At long-term follow-up, 67.69 % of them had a mild disability and low quality of life.

Conclusion

Our study has shown that patients with IIMs have mild disability in 67.69% of the cases, moderate in 16.92% of the cases and severe disability was found in 15.39% of the cases. A dose of prednisolone-equivalent <10 mg and disease duration less than 2 years were determined as protective factors against a severe degree of disability.

Compliance with Ethics Requirements:

„The authors declare no conflict of interest regarding this article"

„The authors declare that all the procedures and experiments of this study respect the ethical standards in the Helsinki Declaration of 1975, as revised in 2008(5), as well as the national law. Informed consent was obtained from all the patients included in the study"

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