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Illness Perceptions and Fatigue in Systemic Vasculitis

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Abstract

Objective—To compare illness perceptions among patients with different forms of vasculitis, identify risk factors for negative illness perceptions, and determine the association between illness perceptions and fatigue.

Methods—Participants were recruited from an online registry in vasculitis to complete the revised Illness Perception Questionnaire (IPQ-R). Mean scores on each IPQ-R dimension were compared across types of vasculitis. Cluster analysis and stepwise regression identified predictors of negative illness perception. Fatigue was measured using the general subscale of the Multidimensional Fatigue Inventory (MFI). Patient-reported measures of disease activity and IPQ-R dimensions were assessed in relation to MFI scores using linear regression in sequential, additive models with model-fit comparisons.

Results—692 participants with 9 forms of vasculitis completed the IPQ-R. For 6 out of 8 IPQ-R dimensions, there were no significant differences in mean scores between the different vasculitides. Scores in *identity* and *cyclical* dimensions were significantly higher in Behçet's disease compared to other types of vasculitis (13.5 vs 10.7; 4.0 vs 3.2, $p<0.05$). Younger age (OR=1.04; 95%CI 1.02–1.06), depression (OR=4.94; 95%CI 2.90–8.41), active disease status (OR=2.05; 95%CI 1.27–3.29), and poor overall health (OR=3.92; 95%CI 0.88–17.56) were associated with negative illness perceptions. Sequential models demonstrated that IPQ-R dimensions explained an equivalent proportion of variability in fatigue scores compared to measures of disease activity.

Conclusion—Illness perceptions are similar across different types of vasculitis, and younger age is a risk factor for negative illness perceptions. Illness perceptions explain differences in fatigue scores beyond what can be explained by measures of disease activity.

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Keywords

vasculitis; illness perceptions; fatigue

The idiopathic vasculitides are a group of rare, systemic diseases involving inflammation of arteries and other tissue with resulting organ- and life-threatening disease courses. The different forms of idiopathic vasculitis are typically classified based upon the predominant size of the arteries affected in each condition, including small vessel vasculitis [e.g. granulomatosis with polyangiitis (GPA, Wegener's)]; medium vessel vasculitis [e.g. polyarteritis nodosa (PAN)]; and large vessel vasculitis [e.g. giant cell arteritis (GCA)]. Although patients with each type of vasculitis manage disease-specific aspects of illness, there are substantial disease and treatment burdens common to patients with vasculitis. For each type of idiopathic vasculitis, the disease course is often chronic, relapse is common and unpredictable, organ and tissue damage can accrue over time, new symptoms can occur late into the disease course, and treatment is often associated with toxicity and serious side effects.

Illness perceptions, also called illness representations, are the organized beliefs that patients have about their illness. Based upon Leventhal's Self Regulatory Model (1), it is theorized that patients consistently structure illness perceptions into common and measurable dimensions. Such dimensions include: beliefs about the symptoms that belong to the disease; the perception of the course the disease has and will have; beliefs on the impact of the illness on functioning and emotional well-being; beliefs on the degree of personal and treatment-based controllability of the disease; and the overall understanding of the condition.

Illness perceptions have been found to be important determinants of health-related behavior and have been associated with important outcomes, such as treatment adherence and functional recovery, across a spectrum of diseases (2). Despite the substantial burdens of disease and treatment patients with vasculitis incur, illness perceptions have not been comprehensively assessed in systemic vasculitis. Understanding illness perceptions of patients with diverse forms of vasculitis may provide insight into the similarities and differences in the cognitive representations of disease and, as seen in other chronic diseases, may help explain important health-related behaviors and outcomes.

Fatigue is a highly prevalent and an increasingly-recognized important manifestation of vasculitis, yet the mechanisms underlying fatigue are poorly understood. One study reported a finding of fatigue with a significant effect on life in approximately half of patients with vasculitis (3). In another study, fatigue was the most commonly reported symptom in a cohort of patients with vasculitis and was a principal contributor to impaired quality of life (4). As seen in other chronic diseases (5), fatigue is not well correlated with disease duration or with *physician-derived* measures of disease activity in vasculitis (3), suggesting that fatigue may be driven by psycho-social measures in response to disease. Further, illness perceptions may be associated with fatigue in vasculitis as has been demonstrated in other diseases (6). Interestingly, fatigue is considered one of the most important disease burdens by patients with vasculitis and is often rated as more burdensome than most manifestations of vasculitis that are associated with organ damage (7). This finding highlights potential

different “models of illness” between patients with vasculitis and health professionals and suggests the importance of understanding illness from patients’ perspectives.

The objectives of this study were 1) to compare illness perceptions of patients with a variety of forms of vasculitis; 2) to determine clinical and demographic variables associated with “negative illness perceptions” for vasculitis, defined as less-desirable perceptions about illness; and 3) to assess the association between illness perceptions and fatigue in vasculitis.

PATIENTS AND METHODS

Study Sample

Participants were recruited online within the Vasculitis Clinical Research Consortium (VCRC) Contact Registry to complete an online questionnaire (8). The VCRC Contact Registry is an international resource which can be used to conduct online clinical research in vasculitis (http://rarediseasesnetwork.org/vcrc/registry). To join the registry, participants self-identify as having a particular form of vasculitis. For this study, participants were excluded if they were <18 years of age.

Data Elements

Demographic information including age, sex, race, ethnicity, highest education level, and annual income was recorded. Participants were asked if they ever experienced depressive symptoms or sleep disturbance since the onset of vasculitis, and respondents were categorized based on affirmative or negative responses to each of these questions. Disease characteristics were assessed per self-report and included disease status (active versus remission), disease duration (continuous variable), disease severity (severe disease defined categorically as history of at least 1 pre-defined organ or life-threatening symptom), and remission duration (less than 1 year, greater than 1 year).

Assessment of Illness Perceptions

All participants completed the revised Illness Perception Questionnaire (IPQ-R) (9). The IPQ-R was devised to assess cognitive representations or beliefs about any illness and has been determined to have good internal reliability, discrimination, and predictive validity. The IPQ-R has been used to study numerous patient populations including those with systemic sclerosis (10), renal disease (11), type II diabetes (12), cystic fibrosis (13), and multiple sclerosis (14) and can be modified for use in a particular disease of interest. Eight dimensions of illness perception were assessed within the IPQ-R: *identity* (22 items), *timeline-acute/chronic* (6 items), *timeline-cyclical* (4 items), *consequences* (6 items), *personal control* (6 items), *treatment control* (5 items), *emotional representations* (6 items), *and illness coherence* (5 items). High scores on the *identity*, *timeline-acute/chronic*, *consequences*, and *timeline-cyclical* dimensions of the IPQ-R represent strongly held beliefs about the number of symptoms attributed to the illness, the chronicity of the condition, the negative consequences of the illness, and the cyclical nature of the condition. High scores on the *personal control*, *treatment control* and *illness coherence* dimensions, represent positive beliefs about the controllability of the illness and increased understanding of the condition. *Identity* was assessed on a 0–22 point scale. All other dimensions were assessed on a 5 point

Likert scale (1 -strongly disagree, 2 - disagree, 3 - neither agree or disagree; 4 - agree; 5 - strongly agree).

Assessment of Fatigue

Fatigue was assessed using the Multidimensional Fatigue Inventory (MFI-20). The MFI-20 has been widely used for measuring fatigue in a variety of diseases (15). Five domains of fatigue are assessed in the MFI-20; however, the "general fatigue" domain has been shown to best discriminate between patients with vasculitis and healthy controls (16) and was therefore used in this study. The "general fatigue" domain of the MFI-20 consists of 4 questions scored on a 5-point Likert scale. An MFI-20 score of ≥13 on the general fatigue domain has been used as a threshold to define significant fatigue, based on the 95th percentile of the score in a healthy group (17).

Analytic Methods

Internal reliability for each dimension of the IPQ-R, except the *identity* subscale, was calculated using Cronbach's alpha per IPQ-R guidelines (9). Correlation between mean scores for each IPQ-R dimension and for fatigue scores on the MFI general subscale was calculated using Pearson's correlation coefficient. Mean scores for each IPQ-R dimension were compared across types of vasculitis for those types of vasculitis in which at least 30 participants were recruited using one-way analysis of variance (ANOVA) with post-hoc Scheffe tests. Mean IPQ-R dimension scores in vasculitis were also compared to literature-reported IPQ-R scores in a variety of chronic diseases including diabetes mellitus, hypertension, osteoarthritis, and systemic lupus erythematosis.

K-means cluster analysis of IPQ-R scores across each dimension was used to define a group of participants with negative illness perceptions. Distance between clusters was calculated using Euclidian distances with the cluster centers based on least-squares estimation (PROC FASTCLUS in SAS statistical software version 9.3). Stepwise logistic regression ($p<0.1$ selection criteria) was used to identify clinical and demographic variables associated with negative illness perceptions.

Unless otherwise noted, p-values <0.05 were used to define statistical significance. All statistical analyses were done using SAS 9.3 (SAS Institute, Cary, NC).

RESULTS

Participant characteristics

A summary of participant characteristics are listed in Table 1. Six hundred and ninety two participants with 9 types of vasculitis completed the study. The majority of participants were female (70%), Caucasian (89%), born in the USA (68%), college graduates (68%), and > 50 years of age. The distribution of demographic characteristics by type of vasculitis was consistent with known distributions for these diseases. For example, all participants reporting a diagnosis of giant cell arteritis were > 50 years of age, the highest proportion of female participants was seen in Takayasu's arteritis, and participants with Behçet's disease and polyarteritis nodosa were more likely to be born outside of the USA.

A substantial number of participants reported a history of depression (47%) and sleep disturbance (56%) since the onset of their vasculitis. Most participants reported several years disease duration at time of study participation (median 7.4 years, range 0 – 41 years). Disease remission was reported in 45% of participants, and among those in remission, 56% reported > 1 year sustained remission. A history of severe disease was reported in 46% of participants.

IPQ-R Dimension Scores

Correlations among the illness perception dimensions and the internal reliability scores for each dimension are listed in Table 2. Significant correlation was observed between most of the illness perception dimensions. Cronbach's alpha scores indicated good to excellent internal reliability for each IPQ-R dimension (range 0.83 – 0.90) with the exception of *treatment control* which had acceptable reliability (0.67). The mean IPQ-R dimension scores are listed by type of vasculitis in Table 3. On the *identity* dimension, participants on average experienced 10.7 (± 5.1) out of a possible 22 symptoms, which were directly attributed to vasculitis. High scores in the *timeline-acute/chronic* and *consequences* dimensions across all types of vasculitis indicated beliefs that the disease will last a long time and has negatively impacted ability to function. There were no statistically significant differences in mean scores by type of vasculitis for *the timeline-acute/chronic, consequences, personal and treatment control, emotional representations,* and *illness coherence* dimensions. Mean dimension scores in *identity* and *timeline-cyclical* dimensions were significantly higher in Behçet's disease compared to some of the other types of vasculitis indicating that participants with Behçet's disease attributed more symptoms to their illness and more strongly endorsed a cyclical component to illness. Although there were too few participants with central nervous system vasculitis for comparative analyses, these participants reported the highest scores in the *consequences* and *emotional representations* dimensions and the lowest scores in the *personal control, treatment control,* and *illness coherence* dimensions, reflecting a high degree of perceived psychological burden of illness in this type of vasculitis.

Compared to literature-reported IPQ-R dimension scores in other chronic diseases (Table 4), higher scores in the *consequences* and *emotional representations* dimensions were seen in people with vasculitis than people with diabetes mellitus (18), hypertension (19), and osteoarthritis (20), indicating stronger beliefs that vasculitis negatively impacts ability to function and emotional well-being. Scores in the *consequences* and *emotional representations* dimensions were similar between vasculitis and systemic sclerosis (10).

Negative Illness Perceptions

K-means cluster analysis separated participants into 3 groups [group 1 = 169 (25%) participants; group 2 = 192 (29%) participants; group 3 = 309 (46%) participants]. Analysis of mean IPQ-R dimension scores by clustered group demonstrated differences between the groups. Mean IPQ-R scores > 3 in the *consequences* and emotional *representations* dimensions represent less-desirable illness perceptions because these scores indicate agreement that the illness has had negative impact on function and emotional well-being. Mean IPQ-R scores < 3 in the *personal control, treatment control,* and *illness coherence*

dimensions also represent less-desirable illness perceptions because these scores indicate beliefs that the illness is not controllable or understandable. In group 1, mean scores for the consequences and emotional representations dimensions were > 3, and mean scores for the personal control, treatment control, and illness coherence dimensions were <3, indicating less desirable illness perceptions across all of these dimensions. Consequently, group 1 was defined as having "negative illness perceptions". For group 2, mean scores for the consequences and emotional representations dimensions were <3 and mean scores for the personal control, treatment control, and illness coherence dimensions were > 3, indicating that group 2 demonstrated "positive illness perceptions" across all of these dimensions. In group 3, which contained the majority of participants, mean scores for the consequences and emotional representations dimensions were < 3 indicating less desirable illness perceptions, but mean scores for the personal control, treatment control, and illness coherence dimensions were >3, indicating desirable illness perceptions for these dimensions. Group 3 was defined as having "mixed illness perceptions."

Stepwise logistic regression was performed to determine risk factors associated with inclusion into the "negative illness perceptions" group. For every year of younger age, the odds of inclusion into the negative illness perceptions group increased by 4% (OR=1.04; 95%CI 1.02–1.06). A history of depression (OR=4.94; 95%CI 2.90–8.41), active disease status (OR=2.05; 95%CI 1.27–3.29), and poor overall health (OR=3.92; 95%CI 0.88–17.56) were also associated with negative illness perceptions. Sex, race, education, annual income, place of birth, disease duration, disease severity, and remission duration were not significantly associated with negative illness perceptions.

Fatigue and Illness Perceptions

The median MFI general subscale scores are listed by type of vasculitis in Table 5. The majority of participants, across all types of vasculitis, scored ≥13 on the MFI, the threshold used to define significant fatigue. There was variability in fatigue scores (interquartile range = 8–18), and the range of results encompassed the entire scale of possible scores. Cronbach's alpha for the MFI general subscale was 0.87, and fatigue scores significantly correlated with each IPQ-R dimension (see Table 2).

The results from 3 sequential linear regression models using MFI scores as the outcome measure are displayed in Table 6. In the first step, disease activity and remission duration were significantly associated with the outcome and the overall model explained 18% of the variability in MFI scores. In step 2, with the addition of clinical and demographic variables, age, race, history of depression, and history of sleep disturbance were significantly associated with the outcome and this model explained an additional 8% of the variability in MFI scores. IPQ-R dimension scores were added in step 3 and all of the IPQ-R dimensions with the exception of *timeline-acute/chronic* were significantly associated with the outcome. The full model which included the IPQ-R dimensions explained an additional 18% of variability in MFI scores with an overall R^2 value of 0.44. These results indicate that IPQ-R dimensions are significantly associated with fatigue in vasculitis and explain variability of fatigue scores in vasculitis beyond measures of disease activity and other clinical and demographic characteristics.

DISCUSSION

This study examined beliefs about illness in 692 participants with 9 forms of systemic vasculitis. Participant-held beliefs about vasculitis were strikingly similar across the different forms of vasculitis, except stronger beliefs about the cyclic nature of illness and a greater number of symptoms attributed to illness were reported among patients with Behçet's disease. Currently, efforts are underway to develop patient-reported outcomes in vasculitis (21). Given the similarity of illness perceptions and the high prevalence of fatigue across many different forms of vasculitis, these data suggest that different types of vasculitis can reasonably be combined when studying illness perceptions and fatigue as a patient-reported outcome. Compared to other chronic illnesses like diabetes mellitus and osteoarthritis, participants with vasculitis reported stronger beliefs about the negative impact of disease on ability to function and emotional well-being. Despite substantial progress that has transformed vasculitis from a frequently fatal disease into a chronic illness, the illness perceptions of participants with vasculitis highlight a need for further improvements in treatment paradigms including a need for potential interventions that target patient beliefs.

Negative illness perceptions were observed in 25% of study participants. Younger age, a history of depression, active disease status, and poor overall health were risk factors for negative illness perceptions. Of these, younger age is perhaps the most intriguing finding. Vasculitis can affect people of all ages, and clinicians caring for patients with vasculitis should be mindful of the heightened psychological burden of disease perceived by younger patients. The association between negative illness perceptions and younger age may reflect differences in the meaning of health problems at different phases of life (22). There are few published studies examining the relationship between illness perceptions and age in other rheumatic diseases. In systemic sclerosis, illness perceptions were not significantly associated with age with the exception that younger patients reported more *personal control* over illness (10). In gout, younger age was associated with belief in stronger negative emotional impact of disease but age was not associated with other dimensions of illness perception (23).

Fatigue is a major symptom reported by patients with all forms of systemic vasculitis, yet the mechanisms underlying fatigue are poorly understood. Physicians and patients with vasculitis may differ in perspective about the relationship between fatigue and active disease in vasculitis (7). *Physician-derived* measures of disease activity do not correlate with fatigue (3). In this study, *patient-reported* measures of disease activity and remission-duration were significantly associated with fatigue scores, suggesting that patients consider fatigue a manifestation of active vasculitis. Illness perceptions significantly explained variability in fatigue scores beyond what could be explained by measures of disease characteristics and depression. These data suggest that i) fatigue is a major domain of illness that is only partially related to disease activity as currently assessed; ii) illness perceptions may have a causal and modifiable role in fatigue; and iii) the mechanisms underlying fatigue in vasculitis are complex and multifactorial. Illness perceptions have been associated with fatigue in other chronic illnesses. In chronic fatigue syndrome, illness perceptions were stronger predictors of fatigue score than mood scores (24). In patients after acute myocardial infarction, declining beliefs in the controllability of illness predicted development of chronic

fatigue and a lowered quality of life (25). In multiple sclerosis, illness perceptions were strongly predictive of fatigue (9) and a randomized controlled trial of cognitive behavioral therapy designed to change cognitive factors contributing to fatigue, such as illness perceptions about symptom identity and controllability of illness, demonstrated clinically significant and sustained reductions in fatigue (26).

There are some important strengths of this study to highlight. This study is the first of its kind to systematically assess illness perceptions in idiopathic vasculitis, a group of illnesses characterized by a high degree of disease and treatment burden. The results provide unique insight into vasculitis from the patient's perspective. Further, a substantial number of participants, representing 9 different types of vasculitis, completed the study, and rarer subtypes of vascuilitis such as polyarteritis nodosa, Behçet's disease, and central nervous system vasculitis were represented. Additionally, the complete version of the IPQ-R, rather than a shortened version known as the Brief Illness Perception Questionnaire (27), was used to comprehensively assess illness perceptions, and excellent internal reliability scores for the various illness perception dimensions support the validity of the IPQ-R for use in vasculitis.

Some potential limitations to this study should be noted. This was an online study in which participants self-reported diagnosis. However, the distribution of demographic characteristics across each type of vasculitis was consistent with known distributions for these diseases, lending face validity to the accuracy of self-reported diagnosis. Selection bias is an issue inherent to most online studies as the study population may not be representative of the general population. In this study, patients were more likely to be female, well-educated, and affluent which limits the generalizability of the findings. Due to the cross-sectional study design, changes in illness perceptions over time in relationship to fatigue and other outcomes such as depression could not be assessed. Variables such as depression and sleep disturbance were assessed per self-report without the use of validated measures and analyzed as categorical variables leading to the possibility of residual confounding. Understanding differences in perspective between patients and health care providers could highlight aspects of vasculitis which require further understanding and could help prioritize future research efforts in accordance with patient-specified needs. Given the online nature of the study, it was not possible to directly compare *patient-reported* measures of disease activity with similar *physician-derived* measures in vasculitis. Such a comparison might highlight the importance of illness perceptions as a means to further delineate ideological differences in disease understanding between patients and physicians.

In a variety of chronic diseases, illness perceptions are known to be important determinants of health related behavior(28) and are associated with important health-related outcomes including treatment adherence and functional recovery (29). In this study, illness perceptions highlighted a group of participants with vasculitis who displayed negative illness perceptions and provided novel insight into the complex, and highly prevalent, complaint of fatigue in systemic vasculitis. Given the potential for a myriad of poor health outcomes and function impairments in vasculitis, illness perceptions may provide an understanding of the patient-driven factors that contribute to these outcomes. Future work could focus on the illness perceptions of patients with vasculitis in relation to medication adherence, the doctor-patient relationship, relapse rates and survival, utilization of health care, and quality of life.

Efforts to modify maladaptive perceptions of illness in relationship to these outcomes should also be explored.

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INNOVATIONS AND SIGNIFICANCE

- Illness perceptions, or patient-held beliefs about illness, are strikingly similar across different types of vasculitis
- Younger age, a history of depression, active disease status, and poor overall health are risk factors for negative illness perceptions in vasculitis
- Fatigue is highly prevalent in vasculitis, strongly associated with illness perceptions, and only partially related to disease activity as it is currently assessed

Table 1

Participant Characteristics

		CLINICAL AND DEMOGRAPHIC CHARACTERISTICS								DISEASE CHARACTERISTICS			
Type of Vasculitis	**No.**	**Age - Years (median, range)**	**Sex (% female)**	**Race (% white)**	**Education (% college graduate)**	**Annual Income (% >75K)**	**Place of Birth (% USA)**	**Depression (% yes)**	**Sleep Disturbance (% yes)**	**Disease Duration - Years (median, range)**	**Disease Activity (% remission)**	**Remission Duration (% >1 year)**	**Disease Extent (% severe)**
Behçet's Disease	48	41.6 (22–71)	88	90	57	42	59	59	66	11.6 (2.5–26)	15	26	48
CNS Vasculitis	12	48.6 (23–65)	55	83	67	36	73	45	52	4.4 (0–16)	32	52	60
Eosinophilic granulomatosis with polyangiitis (Churg-Strauss)	121	55.0 (25–80)	69	84	74	43	64	82	64	7.2 (0–37)	17	63	67
Giant Cell Arteritis	32	67.4 (51–78)	85	75	65	44	62	55	62	3.9 (0–15)	53	50	19
Granulomatosis w/Polyangiitis	332	54.8 (26–86)	63	92	63	50	72	43	54	8.2 (0–41)	53	62	63
IgA Vasculitis (Henoch-Schönlein)	12	43.8 (24–69)	67	92	58	45	75	25	30	2.7 (0–12)	33	41	17
Microscopic Polyangiitis	42	57.7 (25–76)	82	90	79	50	69	50	59	6.7 (0.5–27)	59	66	60
Polyarteritis Nodosa	36	49.7 (18–76)	66	81	44	28	41	52	77	5.1 (0–18)	31	59	44
Takayasu's Arteritis	57	50.2 (26–72)	94	89	80	42	73	43	53	8.6 (0–20)	51	60	25
Total	692	53.8 (18–86)	70	89	66	38	68	47	56	7.4 (0–41)	45	56	46

Table 2

Correlation Between IPQ-R Dimensions and Fatigue Scores

	1	2	3	4	5	6	7	8	9
1. Identity	1.00								
2. Timeline – acute/ chronic (α=0.83)	.24**	1.00							
3. Timeline – cyclical (α=0.84)	.48**	.22**	1.00						
4. Consequences (α=0.83)	.52**	.44**	.48**	1.00					
5. Personal control (α=0.83)	−.10*	−.17**	−.14**	−.14**	1.00				
6. Treatment control (α=0.68)	−.17**	−.38**	−.26**	−.28**	.46**	1.00			
7. Emotional representations (α=0.89)	.40**	.21**	.41*	.51**	−.26**	−.32**	1.00		
8. Illness coherence (α=0.90)	−.16**	.03	−.24**	−.19**	.20**	.22**	−.35**	1.00	
9. Fatigue (α=0.87)	.43**	.32**	.42**	.54**	−.24**	−.29**	.43**	−.11*	1.00

α= Cronbach's alpha.

** p<0.001,

* p<0.01

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Table 3

IPQ-R dimension mean scores (standard deviation) in different forms of vasculitis

	Total	BD	CNSV	EGPA	GCA	GPA	IgAV	MPA	PAN	TAK
Identity	10.7 (5.1)	13.5 [ab] (5.3)	11.7 (4.5)	10.5 (4.9)	8.7 [a] (4.7)	10.9 (5.2)	5.7 (3.0)	10.9 (5.3)	10.9 (4.0)	9.2 [b] (5.0)
Timeline – acute/chronic	4.1 (0.8)	4.2 (0.6)	4.3 (0.9)	4.3 (0.8)	3.7 (0.7)	4.1 (0.8)	3.3 (1.0)	4.0 (0.9)	4.2 (0.7)	3.9 (0.9)
Timeline – cyclical	3.2 (1.0)	4.0 [abcde] (0.6)	3.4 (1.1)	3.1 a (0.9)	3.0 [b] (0.9)	3.1 [c] (1.0)	3.0 (1.0)	3.1 [d] (1.1)	3.6 (0.9)	3.2 [e] (0.8)
Consequences	3.8 (0.8)	4.0 (0.7)	4.6 (0.4)	3.9 (0.8)	3.7 (0.8)	3.8 (0.8)	3.1 (0.8)	3.7 (0.9)	4.0 (0.8)	3.9 (0.8)
Personal control	3.3 (0.8)	3.1 (0.7)	2.8 (0.8)	3.3 (0.9)	3.0 (0.8)	3.3 (0.8)	3.2 (0.8)	3.4 (0.7)	3.2 (0.8)	3.4 (0.7)
Treatment control	3.3 (0.7)	3.0 (0.7)	3.0 (1.0)	3.2 (0.7)	3.5 (0.7)	3.4 (0.7)	3.3 (0.5)	3.4 (0.7)	3.2 (0.8)	3.4 (0.7)
Emotional representations	3.1 (1.0)	3.4 (0.8)	3.7 (0.9)	3.2 (1.0)	3.2 (1.0)	3.0 (0.9)	2.8 (0.9)	3.2 (1.0)	3.5 (0.9)	3.0 (0.9)
Illness coherence	3.4 (1.0)	3.2 (1.0)	3.0 (1.1)	3.4 (1.0)	3.3 (1.2)	3.5 (0.9)	3.1 (1.1)	3.3 (1.0)	3.2 (1.0)	3.4 (0.9)

ANOVA was performed to compare means scores across different types of vasculitis types where >30 participants were recruited into the study (i.e. CNSV, IgAV were excluded from comparative analyses due to small sample size). Note [a,b,c,d,e] denotes row-wise pairs of groups different at 0.05 level Scheffe test. Identity is 0–22 point scale, all other dimensions are scaled 1–5. BD = Behçet's disease; CNSV = central nervous system vasculitis; EGPA = eosinophilic granulomatosis with polyangiitis (Churg-Strauss); GCA = giant cell arteritis; GPA = granulomatosis with polyangiitis (Wegener's); IgAV = IgA vasculitis (Henoch-Schönlein); MPA = microscopic polyangiitis; PAN = polyarteritis nodosa; TAK = Takayasu's arteritis.

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Table 4

Mean IPQ-R Dimension Scores in Vasculitis and Other Chronic Diseases

	Vasculitis*	Diabetes Mellitus[18]	Hypertension[19]	Osteoarthritis[20]	Systemic Sclerosis [10]
	n = 692	n = 164	n = 514	n = 241	n = 49
*Identity***	10.7 (±5.1)	NC	NC	NC	NC
Timeline	4.1 (±0.8)	3.9 (±0.8)	3.6 (±0.4)	4.2 (±0.8)	4.2 (±0.6)
Cyclical	3.2 (±1.0)	2.8 (±0.7)	3.2 (±0.8)	3.6 (±0.8)	3.5 (±1.0)
Consequences	3.8 (±0.8)	2.9 (±0.6)	2.6 (±0.6)	2.8 (±0.8)	3.8 (±0.8)
Emotional repr.	3.1 (±1.0)	2.0 (±0.6)	2.6 (±0.7)	2.4 (±0.8)	3.1 (±1.0)
Personal control	3.3 (±0.8)	3.9 (±0.6)	3.5 (±0.7)	3.1 (±0.6)	2.9 (±0.7)
Treatment control	3.3 (±0.7)	4.6 (±0.9)	3.5 (±0.6)	2.8 (±0.6)	3.0 (±0.6)
Illness coherence	3.4 (±1.0)	3.5 (±0.9)	NR	3.6 (±0.8)	3.1 (±1.0)

*Vasculitis types include Behçet's disease (n=48), central nervous system vasculitis (n=12), eosinophilic granulomatosis with polyangiitis (Churg-Strauss) (n=121), giant cell arteritis (n=32), granulomatosis with polyangiitis (Wegener's) (n=332), IgA vasculitis (Henoch-Schönlein) (n=12), microscopic polyangiitis (n=42), polyarteritis nodosa (n=36), and Takayasu's arteritis (n=57).

**Identity was scored on scale from 0–22. All other dimensions were scored on a scale from 1–5. NC = not comparable across diseases due to disease-specific modifications to IPQ-R. NR = not reported.

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Table 5

Multidimensional Fatigue Inventory (MFI) General Subscale Scores in Vasculitis

Type of Vasculitis	MFI Score (median, range)	% MFI Score ≥ 13*
Behçet's Disease	17.0 (7–20)	86
Central Nervous System Vasculitis	16.0 (4–20)	83
Eosinophilic Granulomatosis with Polyangiitis (Churg-Strauss)	16.1 (4–20)	76
Giant Cell Arteritis	16.0 (4–20)	82
Granulomatosis w/ Polyangiitis (Wegener's)	15.0 (4–20)	72
IgA Vasculitis (Henoch- Schönlein)	14.5 (4–20)	58
Microscopic Polyangiitis	15.5 (4–20)	74
Polyarteritis Nodosa	16.0 (8–20)	84
Takayasu's Arteritis	16.0 (4–20)	85
Total	16.0 (4–20)	76

* Threshold to define significant fatigue (17).

Table 6

Sequential Multiple Linear Regression Models to Assess Variability in MFI Scores

Step One: Disease Characteristics		**Step Two: Add Clinical/Demographic Variables**		**Step Three: Add IPQ-R Dimensions**	
Disease Activity *(active vs remission)*	β = 1.01**	Age *(per year)*	β = 0.01**	Identity *(more symptoms)*	β = 0.07*
Disease Extent *(severe vs not)*	β = 0.33	Sex *(male vs female)*	β = 0.43	Timeline -acute/chronic *(more chronic)*	β = 0.23
Disease Duration *(continuous)*	β = −0.03	Race *(other vs white)*	β = 2.28*	Timeline-cyclical *(more cyclical)*	β = 0.43 *
Shorter Remission Duration *(<1 year, ≥ 1year)*	β = 1.03**	Depression *(yes vs. no)*	β = 1.63**	Consequences *(negative impact)*	β = 1.23**
		Sleep Disturbed *(yes vs. no)*	β = 0.81 *	Personal control *(more control)*	β = −0.42*
				Treatment control *(more control)*	β = −0.73*
				Emotional representations *(negative impact)*	β = 0.38*
				Coherence *(more understanding)*	β = 0.40*
	F = 24.16** Adjusted R^2 = 0.18		F = 15.03** Adjusted R^2 = 0.26 R^2 change = 0.08		F = 16.03** Adjusted R^2 = 0.44 R^2 change = 0.18