Causal Attributions about Disease Onset and Relapse in Patients with Systemic Vasculitis

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ABSTRACT. ***Objective.*** Patients vary in their beliefs related to the cause of serious illness. The effect of these beliefs among patients with systemic vasculitis is not known. Our study aimed to describe causal attributions about disease onset and relapse in systemic vasculitis and to examine whether causal beliefs differ by type of vasculitis or are associated with negative health outcomes.

Methods. Patients with vasculitis were recruited to complete an online questionnaire. Categories of causal beliefs were assessed with the Revised Illness Perception Questionnaire (IPQ-R). Differences in beliefs about disease onset versus relapse were compared across different forms of vasculitis. Causal beliefs were assessed in association with several health outcomes including fatigue, functional impairments, and personal understanding of the condition.

Results. The questionnaire was completed by 692 patients representing 9 forms of vasculitis. The majority (90%) of patients had beliefs about the cause of their illness. Causal attributions were highly variable, but altered immunity and stress were the most commonly agreed-upon causal beliefs. Frequencies of causal beliefs were strikingly similar across different forms of vasculitis, with a few notable exceptions primarily in Behçet disease. Beliefs differed about causes of disease onset versus relapse. Specific beliefs about disease onset and relapse were weakly associated with fatigue, functional impairments, and understanding of the condition.

Conclusion. Patient beliefs related to the cause of systemic vasculitis are highly variable. Patterns of causal beliefs are associated with important negative health outcomes. Clinicians who care for patients with vasculitis should be mindful of these associations and consider asking about patients' causal beliefs. (First Release March 15 2014; J Rheumatol 2014;41:923–30; doi:10.3899/jrheum.131096)

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The primary systemic vasculitides are a group of rare, life-threatening illnesses of unknown cause. Inflammation of blood vessels characterizes vasculitis, and preferential involvement of small, medium, or large arteries helps distinguish among the many different forms of the disease[1]. Although symptoms and patterns of organ system involvement can differ by type of vasculitis, all patients with vasculitis must manage both substantial burdens of disease and treatment-related side effects, either of which can significantly impair psychological well-being and quality of life[2,3]. Vasculitis typically manifests with an initial acute and severe event that requires high-potency immunosuppressive therapy as a life-saving or organ-preserving measure. Despite advances in medical therapy to induce disease remission, the majority of patients with vasculitis will experience 1 or more disease relapses[4,5]. Relapses often result in more organ damage, can be fatal, and can occur at any point in the course of disease, even after several years of sustained remission[6,7].

Patient-held beliefs about the cause of their illness have been associated with emotional well-being, health behavior, quality of life, and disease understanding[8,9]. Although causal attributions have not been formally assessed in

systemic vasculitis, evidence in other life-threatening diseases suggests that patients are more likely to seek causal explanations for illnesses that are more severe or when the disease course is both unpredictable and uncontrollable[10]. Causal attribution theory, as proposed by Weiner, holds that causal beliefs are meaningful because they contribute to subsequent inferences patients will make about their actions in relation to the outcome of their illness[11]. Therefore, causal attributions can provide important insight into an individual's cognitive and behavioral motivation to become well and to engage in therapeutic and preventive health measures[12].

The primary objective of our study was to assess causal attributions about both disease onset and relapse in patients with 9 different forms of vasculitis. An additional study objective was to determine whether patients with vasculitis discussed causal attributions with a healthcare provider and the extent to which such discussions were desirable and helpful to the patient.

MATERIALS AND METHODS

Study sample. Patients were recruited from within the Vasculitis Clinical Research Consortium (VCRC) Contact Registry to complete an online questionnaire. The VCRC Contact Registry is an online resource with international reach that can be used to conduct clinical research in vasculitis (http://rarediseasesnetwork.org/vcrc/registry). To join the registry, patients self-identify as having a particular form of vasculitis. Physician-based data were not available for diagnostic confirmation. Patients were excluded if they were < 18 years of age. An initial survey was e-mailed to members of the VCRC Contact Registry in November 2011. Two separate reminder e-mails were sent 3 weeks apart, and study recruitment closed in January 2012. Ethics approval was provided by the University of South Florida Institutional Review Board.

Demographic information including age, sex, race, ethnicity, highest education level, and annual income was recorded. Disease characteristics were assessed per self-report and included disease status (active versus remission), disease duration (continuous variable), disease severity (defined categorically as history of at least 1 predefined organ or life-threatening symptom), and history of relapse.

Causal attributions assessment. All study patients were asked whether they had beliefs about what caused their vasculitis and whether they had discussed these beliefs with a healthcare provider. Respondents who discussed their beliefs with a healthcare provider were asked whether this discussion was helpful and whether the provider was respectful of their beliefs. Respondents who had not discussed their beliefs with a healthcare provider were asked whether discussion of beliefs would be helpful, if they would like to discuss their beliefs, and if they felt embarrassed to discuss these beliefs. Responses to these questions were coded as "yes" or "no".

The revised version of the Illness Perception Questionnaire (IPQ-R) was used to assess categories of causal attributions[13]. The IPQ-R was developed in accordance with the common sense model of illness perceptions, which asserts that patients consistently structure illness perceptions into common dimensions[14]. Nine dimensions of cognitive representations or beliefs about any illness, including a section on causal attributions, are assessed within the IPQ-R. The IPQ-R has been determined to have good internal reliability, discrimination, and predictive validity[13]. Patients were instructed to provide "your own views about the factors that caused your illness rather than what others including doctors and family members may have suggested to you." Patients were then asked to indicate level of agreement to 18 specified categories of causal beliefs on a scale of 1: strongly disagree; 2: disagree; 3: neither agree nor disagree; 4: agree; and 5: strongly agree.

The IPQ-R has been used in numerous patient populations and can be modified for a particular disease interest[13]. Given the frequent relapsing nature of vasculitis, the IPQ-R was modified for our study to assess causal beliefs about both disease onset and relapse. The same 18 causal belief items were assessed for disease onset and relapse, and an additional 3 belief items ("stopped taking my medications", "change in my medications", and "change in the weather") were only assessed for relapse. The additional belief items were chosen because patients with other rheumatic diseases frequently report these beliefs as causal for disease relapse[9]. Only patients who reported a history of at least 1 disease relapse were queried about beliefs related to relapse.

Outcome assessment. Fatigue is highly prevalent in vasculitis[15,16] and is rated by patients to be one of the most important burdens of disease[17]. The Multidimensional Fatigue Inventory (MFI-20) was used to quantify fatigue[18]. The MFI-20 has been used in a variety of diseases to assess 5 domains of fatigue. The "general fatigue" domain has been shown to best differentiate patients with vasculitis from healthy controls[19]. The "general fatigue" domain of the MFI-20 consists of 4 questions scored on a 5-point Likert scale. Higher scores on the MFI-20 indicate a greater degree of perceived fatigue.

Functional impairment. Three subscales of the Medical Outcomes Study General Health Survey (MOS) were used to measure functioning[20]. "Physical" functioning, defined by activities such as climbing stairs and walking 1 block, was assessed by responses to 6 items, each rated on a 3-point scale ("limited for more than 3 months", "limited for 3 months or less", "not limited at all"). "Role" functioning, defined by the ability to perform schoolwork, housework, or job-related tasks, was assessed by responses to 2 items, each rated on a 3-point scale ("limited for more than 3 months" to "not limited at all"). "Social" functioning was assessed by 1 item, which queried how often over the last month vasculitis has limited social activities (such as visiting with friends or close relatives), with responses recorded on a 6-point scale (ranging from "none of the time" to "all of the time"). Higher scores on the MOS subscales indicate worse levels of functioning.

Illness coherence. The "illness coherence" domain within the IPQ-R was used to assess each participant's perceived understanding of their condition[13]. Illness coherence was assessed by 5 items scored on a 5-point scale corresponding to level of agreement (ranging from 1: strongly disagree, to 5: strongly agree) in response to statements such as "my vasculitis doesn't make any sense to me" and "I have a clear picture or understanding of my vasculitis". Higher scores on the illness coherence domain represent increased perceived understanding of vasculitis.

Statistical analyses. Mean scores for each causal belief item on the IPQ-R were calculated for attributions related to disease onset and relapse. The percentage of respondents who indicated agreement (either "agree" or "strongly agree") to a particular belief item was calculated. Mean scores of each belief item assessed for disease onset and relapse were compared using paired t tests with a Bonferroni correction applied to adjust for multiple comparisons[21]. Because 18 items were compared, a p value < 0.003 was used to define the threshold for statistical significance in these comparisons. As recommended by the designers of the IPQ-R, factor analysis was performed to potentially reduce the number of causal belief items[13,22]. Principal components analysis with a varimax rotation was performed, and subsequent factor loadings for the individual items and their factors were described. Factor loading patterns for causal attributions related to disease onset and relapse were examined separately. Internal reliability for each factor was calculated using Cronbach's alpha coefficient. Differences in mean IPQ-R scores for individual causal belief items or factors were compared between different types of vasculitis using 1-way ANOVA with posthoc Scheffe tests. Only those types of vasculitis in which at least 30 patients completed the survey were included in these comparative analyses, and a p value of < 0.05 defined statistical significance. For

any identified differences in beliefs by type of vasculitis, adjustments for potential demographic and clinical confounders were performed using linear regression models. Correlations between causal belief items or factors and MFI-20 scores, MOS functional scores (physical, social, and role), and scores on the illness coherence domain of the IPQ-R were assessed using Pearson's correlation coefficient. A Bonferroni correction was applied to account for multiple outcome comparisons. Because 5 outcomes were evaluated, a p value < 0.01 was used to define statistical significance for these analyses.

RESULTS

Patient characteristics. Subject characteristics are displayed in Table 1. The survey was completed by 692 patients with 9 different types of vasculitis. Various types of small vessel vasculitis were represented [granulomatosis with polyangiitis (GPA), microscopic polyangiitis, eosinophilic granulomatosis with polyangiitis (EGPA, Churg-Strauss), IgA vasculitis (Henoch-Schonlein purpura)]. The medium vessel vasculitis that was represented was polyarteritis nodosa. Of the large vessel vasculitis types, the survey included giant cell arteritis and Takayasu arteritis. Other types of vasculitis with no clear predilection for a specific vessel size were also represented, including central nervous system vasculitis (CNS vasculitis) and Behçet disease.

Table 1. Participant characteristics.

Vasculitis Type	N (% of Cohort)
Behçet disease	48 (7)
Central nervous system vasculitis	12 (2)
Eosinophilic granulomatosis with polyangiitis	121 (17)
Giant cell arteritis	32 (5)
Granulomatosis with polyangiitis	332 (48)
IgA vasculitis	12 (2)
Microscopic polyangiitis	42 (6)
Polyarteritis nodosa	36 (5)
Takayasu arteritis	57 (8)
Total	692
Age, yrs, median (range)	54 (18–86)
Female sex (%)	70
Race (%)	
White	89
Asian	2
African American	2
Other	7
Ethnicity (% non-Hispanic)	97
Highest level of education (%)	
No high school diploma	2
High school graduate	32
College graduate	30
Advanced degree	36
Annual income (USD)	
Less than $25,000	10
$25,000–$74,999	36
More than $75,000	38
Prefer not to answer	16
Place of birth (%)	
United States	68
Europe	16
Latin America	10
Other	6
Disease duration, yrs, median (range)	7 (0–41)
Disease activity (% active)	55
Disease extent (% severe)	46
At least 1 relapse (% yes)	72

The majority of patients reported a diagnosis of GPA, and > 30 patients were recruited for each type of vasculitis, except CNS vasculitis and IgA vasculitis. The majority of patients were female (70%), white (89%), born in the United States (68%), college graduates (66%), and > 50 years of age. Most patients reported several years of disease duration at the time of study participation. Active disease was reported by 55% of patients, and a history of at least 1 severe disease manifestation was reported by 46% of patients. Most study patients (72%) reported at least 1 disease relapse.

Discussion of causal beliefs with healthcare provider. Of 692 study patients, 90% had beliefs about what caused their vasculitis. Sixty percent of those patients had discussed causal attributions with a healthcare provider. Of those who had discussed their beliefs, 59% found the discussion to be helpful and 75% felt that the healthcare provider was respectful of their beliefs. Of the 30% of patients who had causal beliefs but had not discussed their beliefs with a healthcare provider, 49% wanted to discuss their beliefs, 41% believed such a discussion would be helpful, and 16% felt embarrassed to discuss their beliefs. Patients who had discussions with their healthcare provider about causal beliefs had significantly higher mean scores on the illness coherence domain of the IPQ-R than those who had not discussed their beliefs (3.5 vs 3.2; $p < 0.01$), indicating a better perceived understanding of their condition.

Categories of causal beliefs. The distribution of responses for each causal belief item is displayed in Table 2. "Altered immunity" and "stress or worry" were the most commonly agreed-upon causal attributions for both disease onset and relapse, with the majority of respondents endorsing these beliefs. A "change in my medications" was frequently considered a cause of relapse. Responses to 9 out of 18 items significantly differed between causal beliefs related to disease onset and relapse. "Chance or bad luck" and "pollution in environment" were more strongly held causes of disease onset. "Diet or eating habits," "poor medical care," "my own behavior," "my mental attitude," "family problems," "overwork," and "my emotional state" were more strongly held causes of relapse. Patients who had conversations about causal beliefs with their healthcare provider were significantly less likely to attribute disease onset and relapse to chance compared to patients who had never discussed their beliefs ($p < 0.01$), but the frequencies of other causal attributions were similar.

Factor analysis of causal belief items. Factor analysis suggested a 5-factor loading of belief items for both disease onset and relapse. Two factors contained the same items for both disease onset and relapse. A psychological factor

Table 2. Categories of causal attributions in systemic vasculitis.

Causal Attribution	Disease Onset, n = 692 Mean (SD)	Disease Onset, n = 692 % Agree	Relapse, n = 498 Mean (SD)	Relapse, n = 498 % Agree	Disease Onset vs Relapse P value for Difference in Means
Altered immunity	3.6 (1.2)	62.3	3.6 (1.3)	64.4	0.350
Stress or worry	3.2 (1.3)	50.2	3.4 (1.4)	57.7	0.016
Chance or bad luck	2.9 (1.4)	38.4	2.6 (1.3)	30.4	**< 0.001**
Pollution in environment	3.0 (1.2)	35.8	2.0 (1.1)	12.2	**< 0.001**
A germ or virus	2.9 (1.2)	34.5	2.9 (1.3)	35.8	0.506
Overwork	2.5 (1.3)	27.0	2.7 (1.4)	35.6	**< 0.001**
Hereditary	2.3 (1.2)	19.9	2.2 (1.2)	16.4	0.008
My emotional state	2.2 (1.2)	18.7	2.5 (1.3)	27.7	**< 0.001**
Ageing	2.2 (1.1)	16.4	2.2 (1.2)	17.1	0.054
Family problems	2.1 (1.1)	15.3	2.5 (1.3)	27.9	**< 0.001**
My own behavior	2.2 (1.0)	11.8	2.3 (1.2)	20.0	**< 0.001**
Diet or eating habits	2.1 (1.0)	8.6	2.2 (1.1)	14.9	**0.001**
Poor medical care	1.9 (1.0)	8.4	2.7 (1.3)	29.5	**< 0.001**
My mental attitude	1.9 (1.0)	8.0	2.0 (1.1)	11.7	**0.002**
Accident or injury	1.7 (1.0)	6.9	1.8 (1.0)	8.9	0.323
My personality	1.8 (1.0)	5.8	1.9 (1.0)	9.5	0.004
Smoking	1.7 (0.9)	4.6	1.6 (0.8)	1.8	0.035
Alcohol	1.6 (0.8)	2.0	1.6 (0.8)	2.3	0.166
Change in my medications	NA	NA	3.2 (1.4)	50.5	NA
Stopped my medications	NA	NA	2.5 (1.5)	31.5	NA
Change in the weather	NA	NA	2.3 (1.3)	22.5	NA

Causal attributions are ordered by "Disease onset % agree" from highest to lowest. Mean scores assessed through IPQ-R on a scale of 1 – strongly disagree; 2 – disagree; 3 – neither agree or disagree; 4 – agree; 5 – strongly agree. For each item, "% agree" refers to percentage of participants who responded "agree" or "strongly agree". Comparisons between mean causal attribution scores related to "disease onset" versus "relapse" were made using paired t test. Bonferroni corrections to account for multiple comparisons were used to define a statistical significance threshold of $p < 0.003$ (0.05/18 items). Statistically significant p-values are shown in boldface. NA: not assessed; IPQ-R: revised Illness Perception Questionnaire.

contained the items "stress or worry," "my mental attitude," "family problems," "overwork," "my emotional state," and "my personality." All items loaded > 0.50 on the factor pattern matrix, and the Cronbach's alpha coefficient for this factor was 0.86 (disease onset) and 0.87 (relapse). A medical risk factor contained the items "diet or eating habits," "ageing," "alcohol," "smoking," and "accident or injury." All items loaded > 0.50 on the factor pattern matrix, and the Cronbach's alpha coefficient for this factor was 0.81 (disease onset) and 0.79 (relapse). A medication factor contained the items "change in my medications" and "stopped taking my medications." This factor was only assessed in relation to relapse, both items loaded with > 0.50 on the factor pattern matrix, and the resultant Cronbach's alpha coefficient was 0.65. No further reduction in items was suggested by the factor analyses, so all other beliefs were analyzed as individual items.

Causal attributions by type of vasculitis. The mean IPQ-R scores for causal belief items or factors are reported by type of vasculitis in Table 3. Causal attributions for disease-onset and relapse are displayed for those types of vasculitis with n > 30 patients. There were few differences in causal attributions by type of vasculitis. Attributing alterations in immunity as a cause of disease onset and relapse was the most strongly endorsed belief across all types of vasculitis. More patients with Behçet disease attributed disease onset and relapse to infectious and hereditary causes compared to patients with some other types of vasculitis, and these differences remained significant after adjustments for clinical and demographic characteristics. A change in weather and medications was also more strongly endorsed in Behçet disease as a cause of relapse; however, these differences did not persist after adjustment for age of the participant. Younger age was associated with ascribing relapse to medication factors ($r = 0.21$, $p < 0.001$) and to changes in weather ($r = 0.14$, < 0.001).

Correlation between causal attributions and outcomes. There were several significant, weak associations between causal belief items or factors and fatigue, function, and illness coherence (Table 4). Causal beliefs related to medical risk factors and chance or bad luck were consistently and significantly associated with lower illness coherence scores for disease onset and relapse attributions. Attributing disease onset to pollution in the environment was associated with impaired physical and social functioning. Attributing relapse to psychological and medication factors was associated with a higher level of reported fatigue and impairments in physical, role, and social functioning.

Table 3. Causal attributions by vasculitis type.

Causal Attribution	Beçhet Disease		Eosinophilic Granulomatosis with Polyangiitis		Giant Cell Arteritis		Granulomatosis with Polyangiitis		Microscopic Polyangiitis		Polyarteritis Nodosa		Takayasu Arteritis	
	O	R	O	R	O	R	O	R	O	R	O	R	O	R
Psychological factors	2.4 (1.0)	3.0 (1.0)	2.1 (0.9)	2.4 (1.0)	2.2 (0.8)	2.3 (1.0)	2.3 (0.9)	2.5 (1.0)	2.5 (1.0)	2.4 (1.1)	2.3 (0.9)	2.5 (1.1)	2.4 (0.8)	2.5 (0.8)
Medical risk factors	2.0 (0.9)	2.2 (0.8)	1.9 (0.7)	2.0 (0.7)	2.0 (0.7)	1.8 (0.6)	1.9 (0.6)	1.9 (0.7)	2.1 (0.8)	1.8 (0.7)	2.0 (0.7)	2.0 (0.9)	1.8 (0.6)	1.9 (0.7)
Pollution in environment	2.9 (1.3)	2.8 (1.3)	3.0 (1.2)	2.9 (1.3)	2.4 (1.4)	2.2 (1.3)	3.1 (1.1)	2.8 (1.2)	3.2 (1.3)	3.0 (1.5)	2.6 (1.3)	2.1 (1.2)	2.6 (1.1)	2.5 (1.1)
Infectious	**3.4 [a] (1.1)**	**3.3 [d] (1.4)**	**2.4 [abc] (1.1)**	2.7 (1.4)	2.6 (1.2)	2.4 (1.3)	**3.0 [b] (1.2)**	3.0 (1.3)	2.9 (1.4)	2.8 (1.5)	2.8 (1.5)	**1.9 [d] (1.3)**	**3.2 [c] (1.1)**	2.9 (1.2)
Hereditary	**3.0[ab] (1.4)**	**3.1 [cde] (1.5)**	**2.0 [a] (1.1)**	**2.0 [c] (1.1)**	**1.9 [b] (1.1)**	2.1 (1.3)	2.4 (1.2)	**2.2 [d] (1.2)**	2.4 (1.3)	2.1 (1.3)	**2.0 [e] (1.3)**	1.7 (1.0)	2.5 (1.3)	2.2 (1.2)
Chance or bad luck	2.7 (1.2)	2.5 (1.1)	2.8 (1.4)	2.8 (1.5)	2.1 (1.3)	1.8 (1.2)	2.9 (1.4)	2.5 (1.3)	2.9 (1.3)	2.2 (1.3)	3.0 (1.6)	2.9 (1.6)	2.8 (1.3)	2.7 (1.3)
Altered immunity	4.0 (1.0)	4.0 (1.1)	3.7 (1.1)	3.6 (1.3)	4.0 (1.1)	3.8 (1.5)	3.6 (1.1)	3.5 (1.2)	3.7 (1.2)	3.6 (1.6)	3.7 (1.1)	3.7 (1.2)	3.5 (1.2)	3.4 (1.3)
Medication factors	NA	**3.4 [a] (1.3)**	NA	3.1 (1.3)	NA	2.5 (1.0)	NA	2.8 (1.2)	NA	**2.3 [a] (1.2)**	NA	2.6 (1.2)	NA	2.9 (1.0)
Change in weather	NA	**3.1 [a] (1.3)**	NA	2.3 (1.3)	NA	**1.3 [a] (0.8)**	NA	2.4 (1.3)	NA	2.0 (1.2)	NA	2.4 (1.5)	NA	2.2 (1.3)

ANOVA was performed to compare means scores across different types of vasculitis types where ≥ 30 participants were recruited into the study (i.e., central nervous system vasculitis and IgA vasculitis were excluded from comparative analyses because of small sample size). Note [a,b,c,d,e] denotes row-wise pairs of groups different at 0.05 level Scheffe test (displayed in bold). Psychological, medical risk, and medication factors are composite items determined by factor analysis. All other causal attributions are single-item beliefs. Psychological factors: stress, my mental attitude, family problems, overwork, my emotional state, my personality ($\alpha = 0.86$). Medical risk factors: diet or eating habits, ageing, alcohol, smoking, accident or injury ($\alpha = 0.81$). Medication factors: change in my meds and stopped taking my meds ($\alpha = 0.65$). O: onset; R: relapse; NA: not assessed.

Table 4. Correlations between causal attributions, fatigue, functional impairment, and illness coherence.

Causal Attribution	Disease Onset					Relapse				
	Fatigue	Physical	Role	Social	Coherence	Fatigue	Physical	Role	Social	Coherence
Psychological factors	0.09 (0.03)	0.03 (0.39)	**0.12 (< 0.01)**	0.10 (0.01)	0.08 (0.04)	**0.17 (< 0.01)**	0.08 (0.09)	**0.13 (< 0.01)**	**0.17 (< 0.01)**	0.04 (0.31)
Medical risk factors	0.01 (0.90)	0.02 (0.57)	0.05 (0.19)	0.08 (0.85)	**0.20 (< 0.01)**	0.04 (0.45)	0.01 (0.78)	0.03 (0.56)	0.11 (0.03)	**0.14 (< 0.01)**
Pollution in environment	0.10 (0.01)	**0.12 (< 0.01)**	0.10 (0.01)	**0.13 (< 0.01)**	0.06 (0.14)	0.07 (0.14)	0.10 (0.04)	0.01 (0.91)	0.12 (0.02)	0.06 (0.22)
Infectious	0.03 (0.51)	0.03 (0.52)	0.05 (0.24)	0.08 (0.04)	0.06 (0.12)	–0.01 (0.89)	0.02 (0.61)	0.01 (0.91)	0.10 (0.04)	0.06 (0.22)
Hereditary	–0.04 (0.30)	0.01 (0.80)	–0.02 (0.68)	–0.01 (0.90)	0.03 (0.45)	–0.03 (0.59)	0.00 (0.95)	0.03 (0.57)	–0.01 (0.84)	0.03 (0.54)
Chance or bad luck	0.08 (0.04)	0.05 (0.23)	0.09 (0.02)	**0.11 (< 0.01)**	**0.19 (< 0.01)**	0.08 (0.08)	0.08 (0.08)	0.07 (0.16)	0.12 (0.01)	**0.15 (< 0.01)**
Altered immunity	0.03 (0.95)	0.06 (0.15)	0.04 (0.31)	0.02 (0.66)	0.02 (0.66)	0.03 (0.56)	0.05 (0.30)	0.02 (0.66)	0.08 (0.12)	**0.13 (< 0.01)**
Medication factors	NA	NA	NA	NA	NA	0.10 (0.04)	0.00 (0.97)	0.04 (0.44)	0.10 (0.05)	0.02 (0.62)
Change in weather	NA	NA	NA	NA	NA	**0.17 (< 0.01)**	**0.17 (< 0.01)**	**0.17 (< 0.01)**	**0.19 (< 0.01)**	0.04 (0.38)

Pearson's correlation coefficient with p values listed in parentheses. Bonferroni correction applied to define statistical significance at $p < 0.01$ (0.05 / 5 outcome items). Significant associations are shown in bold. NA: not assessed.

DISCUSSION

The vast majority (90%) of patients with vasculitis have beliefs about the cause of their illness. Most of those patients have discussed their beliefs with a healthcare provider and found these discussions to be helpful. About one-third of patients with vasculitis have never discussed causal attributions with a healthcare provider and a substantial number of these patients would like to have these conversations but may feel embarrassed to broach the topic. Specific causal attributions were highly variable among individuals with vasculitis, but the general frequencies of categorical beliefs were strikingly similar across the different vasculitides. In cases where there were differences in causal belief by type of vasculitis, age was often a confounder. Younger age, a common risk factor for medication noncompliance[23], was strongly associated with beliefs that medication factors, including alterations in medication regimen and medication noncompliance, were causal for relapse in vasculitis. Categories of causal attributions were weakly associated with deleterious outcomes including fatigue, functional impairments, and perceived lack of understanding of the condition.

Scientific understanding about the etiology of vasculitis is limited; however, an interaction between genetic and environmental factors leading to the development of autoimmunity likely underpins disease[24]. Accordingly, most patients with vasculitis acknowledged autoimmune factors as causal in the disease process. Of all types of vasculitis under study, the strongest scientific evidence for a hereditary component of disease has been reported in Behçet disease[25,26,27], and hereditary factors among patients with Behçet disease were indeed more commonly attributed as causal than in other forms of vasculitis. Few environmental exposures have been associated with vasculitis. Silica dust exposure has been associated with development of GPA and EGPA[28,29], which are 2 types of vasculitis with prominent airway involvement. Although not statistically significant, the highest disease onset causal attribution scores for environmental exposure were seen in patients with GPA and EGPA. Infectious causes have long been sought in vasculitis; however, apart from the role of viral hepatitis in a subset of patients with polyarteritis nodosa, no causal infectious agents have been identified for any type of vasculitis included in this study. There is no scientific evidence linking psychological factors or variations in weather patterns with disease onset or relapse in vasculitis, yet these causal beliefs were commonly held and significantly associated with negative outcomes such as fatigue and functional impairment.

Causal attribution theory posits that the structure of thinking, rather than the causal beliefs themselves, is more relevant for predicting behavioral motivation and emotional response to illness[11]. Perceived causes of success and failure share 3 common dimensions: locus of causality, stability, and controllability[12]. Locus of causality refers to whether the source of the belief is internal (e.g., stress or feeling tired) or external (e.g., pollution in the environment or bad luck). Stability indicates the changeable aspects of the belief over time, and controllability differentiates whether an individual has volitional ability to modify the source of a belief. When viewed from the perspective of causal dimensions, some interesting findings in this study emerge. A shift in locus of causality was observed for beliefs about disease onset versus relapse. Patients with vasculitis more strongly ascribed causal beliefs about disease onset to external factors (pollution in the environment, chance, or bad luck), yet causal beliefs about relapse were predominantly attributed to internal factors (diet or eating habits, my own behavior, my mental attitude, family problems, overwork, my emotional state). Additionally, patients who endorsed external over internal causal beliefs were more likely to report a poorer understanding of their condition.

When dimensions of causal attributions are considered in combination, as encouraged by causal attribution theory, predictable cognitive illness representations and behavioral consequences are further apparent[11]. For example, external, stable, and uncontrollable attributions preserve self-esteem but decrease expectations that behavioral modification can affect the course of illness and that future recovery is possible. Internal, unstable, and controllable attributions decrease self-esteem, but they also increase expectations for coping strategies and bolster feelings that behavioral modification can improve outcomes. Beliefs that a change in weather was causal for relapse (an external, unstable, and uncontrollable attribution) were most strongly and consistently associated with negative outcomes including higher levels of fatigue and impairment in physical, social, and role functioning. Beliefs that pollution in the environment (an external, stable, and uncontrollable attribution) was causal for disease onset were associated with impairments in physical and social functioning. Psychological factors (which tend to be internal, unstable, and controllable attributions) were also associated with worse fatigue and impairments in role and social functioning. According to the principles of attribution theory, out of all of the identified beliefs associated with negative outcomes in this study, psychological factors are most likely to motivate an individual's adaptive response to overturn or actively address the adverse outcome and could be a focus for behavioral intervention.

The categories of causal attributions commonly reported by patients with vasculitis are not unique and mirror descriptions of causal attributions in other rheumatic diseases. Patients with rheumatoid arthritis (RA)[9] and systemic lupus erythematosus[30] most frequently attribute disease onset to hereditary factors, autoimmune factors, and psychological stressors. Similarly, patients with RA commonly attribute relapse to psychological stressors and changes in weather, and categorical causal beliefs have also been modestly associated with a series of negative outcomes in RA including functional impairments, greater disease activity, poorer psychosocial adjustment, and illness unpredictability[9]. Conversely, causal attributions in vasculitis differ markedly from the structure of beliefs in patients afflicted by diseases with known strong causal associations, such as chronic obstructive pulmonary disease and lung cancer[31,32].

The results in our study have important implications for healthcare providers caring for patients with systemic vasculitis. According to the self-regulation model of illness proposed by Leventhal, *et al*[14], patients with vasculitis actively process information about their condition, evaluate attempts to moderate or cope with the effects of illness, and form representations of illness, including causal beliefs, based on their experiences. The majority of patients with vasculitis hold beliefs about the cause of their illness, are interested in discussing these beliefs with a healthcare provider, and generally find these discussions helpful. The findings in this report provide context for healthcare providers to engage in discussions about causal attributions with their patients. These discussions provide an oppor-

tunity to educate patients about causal beliefs commonly held by patients with vasculitis in relation to known scientific evidence about the etiology of these diseases. A patient's specific causal attributions may also provide useful clinical information. For example, beliefs about the role of psychological stressors may prompt further inquiry into a patient's perceived negative consequences of illness, understanding of disease, and behavioral motivation and may help to identify patients who could benefit from behavioral interventions such as stress management programs. Beliefs that medication factors cause relapse, particularly in younger patients, may prompt further inquiry into medication compliance. A belief that chance or bad luck is causal for disease onset may prompt further discussion about that individual's perceived understanding of the condition.

Our study has some potential limitations to consider. Correlations between specific causal attributions and health outcomes, while statistically significant, demonstrated only weak association (r = 0.12–0.19). Despite the low magnitude of correlation between specific causal beliefs and health outcomes, broader dimensions of causal attributions were consistently associated with negative health outcomes in accordance with the principles of causal attribution theory. Recruitment of patients online and the use of quantitative rather than qualitative methods to assess causal beliefs may limit the generalizability of the findings and restrict the potential types of causal beliefs identified. However, the frequencies of specific categorical beliefs and the patterns of association with health outcomes mirrored findings in other chronic rheumatic diseases, lessening concerns about selection bias and ascertainment bias. The cross-sectional nature of this study is another important limitation that precludes study of whether causal attributions predictably change over the disease course. The strengths of our study include a large cohort of a rare disease with a broad representation of many rare disease subtypes. Further, this is the first study of its kind to assess causal attributions about disease onset and relapse in systemic vasculitis, and the findings could be used to facilitate clinician-patient discussions about causal attributions in these diseases.

The majority of patients with vasculitis have beliefs about disease onset and relapse. These beliefs are highly variable and may provide important clinical information and opportunities for discussion that could strengthen therapeutic alliance. Similar to other chronic, relapsing, and life-threatening diseases, causal attributions in vasculitis are associated with important negative health outcomes and may predict health-related behaviors. Clinicians who care for patients with vasculitis should be mindful of these associations and consider asking about patients' causal beliefs.

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