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# What matters for patients with vasculitis?

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## In this issue

## Summary

Advances in clinical care for patients with vasculitis have improved survival rates and created new challenges related to the ongoing management of chronic disease. Lack of curative therapies, burden of disease, treatment-related side effects, and fear of relapse contribute to patient-perceived reduction in quality of life. Patient-held beliefs about disease and priorities may differ substantially from the beliefs of their health care providers, and research paradigms are shifting to reflect more emphasis on understanding vasculitis from the patient's perspective. Efforts are ongoing to develop disease outcome measures in vasculitis that better represent the patient experience. Health care providers who care for patients with vasculitis should be sensitive to the substantial burdens of disease commonly experienced by patients living with the disease and should strive to provide comprehensive care directed towards the medical and biopsychological needs of these patients.

The systemic idiopathic vasculitides are a group of rare diseases characterized by inflammation and necrosis of blood vessel walls with potential resultant organ- and life-threatening outcomes. The clinical spectrum of disease is often dependent on the size, number, and site of involved blood vessels [1]. Onset of vasculitis occurs throughout the age spectrum. Certain types of vasculitis, such as Takayasu's arteritis, present in childhood or early adulthood. Other types of vasculitis, such as giant cell arteritis, are exclusive to later life.

Advances in therapy have transformed vasculitis from a frequently fatal disease into a chronic illness. Physician-based measures for disease assessment have facilitated the conduct of successful clinical trials that have identified effective medical therapies and led to improved survival rates [2,3]. Despite these advances, patients with vasculitis continue to manage substantial burdens of illness. Treatment with potentially toxic medications, including glucocorticoids and other immune suppressants such as cyclophosphamide, is generally required to prevent fatal outcomes in severe cases. Although it is possible for most patients with vasculitis to achieve remission with therapy, the majority of patients experience disease relapse despite expert care, and relapses can be fatal. Across different types of vasculitis, the disease course is often chronic, relapse is common and

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unpredictable, organ and tissue damage can accrue over time, new symptoms can occur late into the disease course, and the burdens of disease and treatment-related side effects can significantly impair psychological well-being and quality of life.

The objective of this review is to describe disease burdens that are common to patients living with vasculitis. Research focused upon understanding vasculitis from the patient's perspective, prioritizing issues relevant to patients, and development of patient-centered outcome measures in vasculitis is discussed.

## Material and methods

A search of publications related to vasculitis and the burden of illness was conducted using the Medline (PubMed) library. The following search terms were used: vasculitis, granulomatosis with polyangiitis (Wegener's), microscopic polyangiitis, eosinophilic granulomatosis with polyangiitis (Churg-Strauss), polyarteritis nodosa, Takayasu's arteritis, giant cell arteritis, burden of illness, quality of life, health related quality of life, outcome, patient reported, self-reported, illness perceptions, patient education, employment, disability, psychological impact, physical impact, fatigue, depression, fibromyalgia, anxiety, unmet needs, sleep, reproductive health, pregnancy, fertility, genetics, medication adherence, and pain. Bibliographies of existing publications were reviewed. All abstracts were manually reviewed to identify studies of interest with focus upon prospective interventional or observational studies conducted within the last five years in larger study populations.

## Illness perceptions and educational needs

Illness perceptions are the organized beliefs that patients have about their illness [4]. The validated revised Illness Perception Questionnaire (IPQ-R) [5] has been used to study patient-held beliefs about the symptoms that belong to vasculitis, the perception of the course of vasculitis, the overall understanding of the condition, and the degree of personal and treatment-based controllability of the disease. Among 692 participants representing 9 types of vasculitis, illness perceptions were strikingly similar across different forms of vasculitis and reflected strongly held beliefs about the substantial negative impact of disease on ability to function and emotional well-being [6]. Younger age, history of depression, active disease status, and poor overall health risk were risk factors for higher perceived burden of illness. Patient-held beliefs about cause of systemic vasculitis are highly variable, but altered immunity and stress are commonly agreed-upon causal beliefs [7]. Patients who hold certain causal beliefs, such as a belief that a change in the weather triggers relapse, tend also to report a higher degree of functional impairment, fatigue, and confusion about the condition. The majority of patients (90%) have specific beliefs about what caused their vasculitis and find discussions with their healthcare provider about their beliefs to be beneficial [7].

Patient educational and self-management programs can influence patient self-efficacy and health outcomes [8]. In a cohort of patients with vasculitis from the United Kingdom or the United States, information on diagnosis, prognosis, investigations, treatment and side effects was rated as extremely important [9]. Information on patient support groups and psychosocial care was rated as less important, and there were no major differences in rating of needs based on available clinical or demographic information. Patients with chronic conditions like vasculitis are encouraged to self-manage as much as possible and lack of effective patient education is a potential barrier to effective self-management [10]. Patients with vasculitis prefer to receive educational information about their disease from a knowledgeable healthcare professional with supporting written material rather than receiving information from the Internet or in a group setting [9,11]; however, most patient education materials for rheumatologic diseases are written at readability and suitability levels that do not support patient health literacy [12].

## Quality of life, fatigue, and patient outcome measures

Several studies have examined health-related quality of life (HRQoL) in vasculitis, with particular focus upon the ANCA-associated vasculitides [13]. Despite advances in clinical management of vasculitis, HRQoL remains substantially reduced during both active disease and remission [14–19]. Approximately 20–30% of patients with vasculitis of working age report significant work disability [20–22]. Fatigue, depression, severe disease-related damage, and being overweight have been independently associated with risk for unemployment [20]. Over half of patients with vasculitis report that the circumstances of living with a rare chronic illness have negatively impacted friendships and social participation [23]. Interventions designed to help patients cope with the impact of vasculitis on HRQoL are needed.

Patients with vasculitis frequently cite fatigue as one of the most burdensome symptoms of disease with significant negative impact on quality of life [24–26]. Despite a reported prevalence of severe fatigue in 2 out of 3 patients with vasculitis [6,27], the mechanisms underlying fatigue are poorly understood. Similar to other chronic illnesses, fatigue does not strongly correlate with disease duration or validated measures of disease activity, suggesting that psychosocial measures may in part mediate fatigue [6,28]. Fatigue has been associated with numerous clinical and biopsychological factors, including disease- and medication-related effects [29], patient-held illness perceptions [6], findings on functional magnetic resonance imaging [30], androgen deficiency [31], sleep disturbance [27,28], pain [27,29], female gender [27], dysfunctional coping strategies [27], and depression [28]. In other forms of chronic illness, non-pharmacological interventions [32] and cognitive