RHEUMATOLOGY

Rheumatology 2017;56:1439–1440
doi:10.1093/rheumatology/kew438
Advance Access publication 16 December 2016

Editorial

The global burden of anti-neutrophil cytoplasmic antibody vasculitis

High but unquantified

The study of the global burden of disease involves evaluation of disparities in health-care provision, in order to improve outcomes and optimize care provision. A recent British Society for Rheumatology workshop highlighted the urgent need for a collaborative approach into research into rare rheumatic conditions; indeed, this is also a priority of the UK National Health Service (NHS) strategy for rare diseases [1, 2].

The ANCA vasculitides (AAV) are a group of complex autoimmune conditions characterized by inflammation and necrosis of blood vessels, leading to end-organ dysfunction and failure. Overall, the conditions are rare, and this has impeded gathering of epidemiological data but also clinical trials and studies on the socio-economic consequences of AAV. The main types of AAV are granulomatosis with polyangiitis (GPA) and microscopic polyangiitis (MPA). PR3-ANCA is associated with GPA, and MPO-ANCA is associated with MPA.

In most populations, the overall annual incidence of AAV is 15–20 per million [3], with considerable variations in the proportions of patients with PR3-ANCA and MPO-ANCA vasculitis or GPA and MPA. In Southern Europe, Japan and China, MPO vasculitis or MPA is the predominant form of vasculitis, with the reverse in Northern Europe. The age at presentation is older in MPA (60–65 years) compared with GPA (45–55 years). The AAV are less common in some non-Caucasian populations; in the USA, AAV appears to be less common in African-Americans. UK data suggest that there is no difference between the Caucasian population and non-Caucasian populations, but this study did not include African-Americans. There is a paucity of data from the Indian subcontinent, Africa and Latin America, but AAV occurs in these populations. The clinical features of GPA and MPA are different, with GPA tending to have more retro-orbital granulomatous disease and MPA more severe renal disease. The global Diagnostic and classification of vasculitis study has provided the opportunity to look at clinical features of AAV across different ethnicities. The main differences appear to be in the distribution of ANCA specificity rather than clinical features. The AAV have a high mortality. In the UK general practice Clinical Practice Research Datalink database, the 1-year mortality of GPA is 13.6%, equal between sexes, but despite the size of the study it was not possible to analyse the influence of ethnicity on outcome. In a trial setting, the overall survival at 1 year is 88% and at 5 years 78%, and compared with the background population the mortality is increased 2.6-fold [4]. Age and renal function were the principal predictors of a poor outcome. A multi-ethnic study from the USA observed that failure to achieve complete remission was associated with female sex, black ethnicity, severity of renal disease and MPO ANCA [5]. This study has not been replicated in other multi-ethnic populations.

The socio-economic status of AAV patients has not yet been studied in detail, and whether AAV patients come disproportionately from poorer socio-economic groups is not known. There are no data on whether low socio-economic status is associated with higher disease activity, although this is likely; in GPA, social deprivation is associated with an increase in cardiovascular disease and ischaemic events [6].

Patients with AAV have significantly impaired quality of life both physically and emotionally, with lower health-related quality of life than the normal population and their spouses. Fatigue is a major determinant of poor health-related quality of life and is relatively unresponsive to immunosuppressive therapy [7]. There is significant financial impact, which may also reduce quality of life. Men with GPA have 2.6-fold increased risk of unemployment after diagnosis [8].

In the UK, only 16.8% of individuals with AAV have received education to degree level compared with 55% in the USA, although this could reflect different education systems [9]. In SLE, low educational status is associated with a worse outcome and higher disease activity and again, this is likely also to be true in AAV.

The AAV are rare conditions, and this poses problems for patient education. Patients have high educational needs, especially in the period after diagnosis [9], and many physicians in primary and secondary care have a poor understanding of the diseases. Owing to the multisystem nature of AAV, patients can often be seen by different medical and surgical specialists. Patients report that they can feel lost in the system and need help to navigate their way through often complex health-care set-ups. Social support networks, such as relatives and friends, also fail, because people have not heard of AAV or even the term vasculitis. The Internet and patient support groups then become key means of information provision. Low educational achievement makes patient

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education harder. Self-management has been highlighted as an important aspect of care for patients with long-term conditions within the NHS 5-year plan.

Calculation of both direct health costs and indirect costs is required to estimate the economic burden to society. The conditions are long term, with the potential for organ damage and high morbidity, and are associated with considerable health costs. The highest costs will be associated with development of end-stage renal disease and the need for dialysis or transplantation; these costs in the UK have been estimated to be £23 426 per patient per year [10]. Introduction of biological therapies, such as rituximab, increases the drug costs, but some of these might be offset by better disease control. The personal costs to the patient and family have not been established. This lack of data can have serious consequences. In the UK, poor-quality health economic data delayed the introduction of rituximab into routine clinical practice.

There is an urgent need to gain a better understanding of the non-medical determinants of outcome. The rarity of the diseases makes their study challenging but does not pose insuperable obstacles. Development of large-scale population-based studies permits study of the socio-economic determinants of disease occurrence and outcome. High-quality, detailed health economic studies are required to determine the direct and indirect costs to enable better resource allocation and to justify to health funders the introduction of novel biologic drugs. The vasculitis community is well networked and in a position to deliver, and it needs to rise to the challenge to help our patients.

Funding: No specific funding was received from any bodies in the public, commercial or not-for-profit sectors to carry out the work described in this manuscript.

Disclosure statement: The authors have declared no conflicts of interest.

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Accepted 25 October 2016

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