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EDITORIALS

Formation of the Australian and New Zealand Vasculitis Society (ANZVASC) to improve the care of patients with vasculitis in Australia and New Zealand

The term vasculitis encompasses a group of disorders that result in the inflammation of blood vessels. These disorders can lead to irreversible damage in many organ systems. In Australia and New Zealand, practitioners from numerous specialties including rheumatology, immunology, nephrology, respiratory medicine, dermatology, ophthalmology, neurology and ear nose and throat (ENT) surgery manage patients with vasculitis. Not surprisingly, the patient's experience of medical care can become fragmented.

Australia and New Zealand centres have been active in vasculitis research over many years. Antineutrophil cytoplasmic antibodies (ANCA) were first described in Melbourne in 1982 by Davies *et al.*, helping to define a major category of small vessel vasculitis – ANCA-associated vasculitis (AAV).[1,2] This discovery led to further local research into ANCA testing and input into international testing guidelines.[3] More recently, mechanistic studies have examined the loss of tolerance to ANCA antigens and the involvement of T cells in tissue injury,[4] as well as the relationship between human leukocyte antigen (HLA) and disease.[5] In addition, there is ongoing collaborative research into the contribution of genetic variation in the initiation and outcome of vasculitis.

Epidemiological studies of vasculitis in Australia and New Zealand have largely focussed on giant cell arteritis (GCA) and AAV. These studies face not only the common challenges of case definition and identification, but also the relatively low population density with fragmented healthcare provision within and across states and territories. Similar to other international studies, Australian and New Zealand studies have used single source registry cohorts,[6–9] with reported prevalence of up to 189 per million and incidence up to 13 per million when AAV subtypes are combined. The greater proportion of patients with granulomatosis with polyangiitis compared with microscopic polyangiitis (MPA) mirrors the patterns seen in European cohorts, in contrast to the reverse observation in Japanese cohorts. In AAV, such studies do not completely capture the burden of disease-related damage, treatment-related adverse effects and the impact of the high rate of relapse (up to 50% of patients at 5 years) on the individual, their family and the healthcare system. Australian and New Zealand estimates of biopsy-proven GCA incidence of 3.2 per 100 000 and 12.7 per 100 000, respectively, are lower than those in European and North American cohorts, where reported incidence has been greatest in Scandinavian populations[10,11] Although seasonal variations in GCA have been reported, this was not demonstrated in a cohort of over 2000 patients with biopsy-proven GCA from Australia and New Zealand.[12]

There are limited outcome data for Australian and New Zealand patients with vasculitis. Among dialysis and renal transplant patients, the Australian and New Zealand dialysis and transplant registry data have identified that patients with AAV who develop end-stage kidney disease (ESKD) have comparable dialysis outcomes to patients with other causes of ESKD. However, after kidney transplantation, AAV patients have poorer allograft survival and those patients with MPA have inferior overall survival.[13] The drivers of mortality identified in European trials of AAV include infection, disease activity and cardiovascular comorbidities.[14] Further studies are required to enable a better understanding of the impact of these rare diseases on mortality, morbidity, quality of life and healthcare utilisation in the Australia and New Zealand setting.

There have been significant contributions from Australia and New Zealand in several international, multicentre investigator-led clinical trials of AAV. These include PEXIVAS,[15] by far the largest randomised trial to date in AAV, which included 704 patients of which 104 were from Australia and New Zealand. This was made possible with support from the Australasian Kidney Trials Network and National Health and Medical Research Council (NHMRC) funding. Other major AAV trials with significant Australia and New Zealand contributions include RITUXVAS, MYCYC, RITAZAREM and the industry-led ADVOCATE study.[16–18] Several of these trials have led to changes in clinical practice and substantial improvements in outcomes for patients with AAV. Close links with international groups such as the European Vasculitis Society, Vasculitis Clinical Research Consortium and research centres in Cambridge and Oxford provide optimism for future collaborative research.

Individuals with rare diseases such as vasculitis face many unique challenges including the psychological, medical and social consequences of having a rare condition. People living with rare diseases in Australia and New Zealand frequently face diagnostic delays and often do not have the benefit of multi-disciplinary care. In a report from the UK, 93% of respondents reported seeing clinicians from multiple medical specialties as part of their routine treatment. Yet, less than one in five was able to see multiple specialists at a joint clinic.[19] Vasculitis patients and their families can feel isolated, under-supported and often face financial hardship as a consequence.[20] There is little published literature evaluating the clinical and cost-effectiveness of multidisciplinary vasculitis care clinics. Interdisciplinary care of patients with systemic lupus erythematosus, rheumatoid arthritis and psoriasis has been shown to be efficacious[21,22] and a vasculitis centre based management strategy for people with eosinophilic granulomatosis and polyangiitis may improve mortality to that of the general population.[23] Recent European recommendations for the management of AAV advocate the use of multidisciplinary care.[24] Furthermore, the National Strategic Action Plan for Rare Diseases released in February 2020 listed interdisciplinary clinics as an enabler for improved rare disease care in Australia.[25] However, the feasibility of such clinics in much of Australia and New Zealand is yet to be determined. Regardless, it is vital that the medical community advocates on behalf of patients with vasculitis for access to timely, evidence- and patient-based therapy. In addition, we need to invest more effort to establish patient and carer support networks.

In 2017, around 40 clinicians, clinician-scientists and a consumer representative across the disciplines of nephrology, rheumatology, clinical immunology and ENT agreed that vasculitis care and research in Australia and New Zealand would benefit from working together in a collaborative and multidisciplinary way. As a result, the Australian and New Zealand Vasculitis Society (ANZVASC) (www.anzvasculitis.org) was formed in late 2018. As a collaborative charitable society of clinicians, scientists and consumers interested in research, best practice clinical care and education in vasculitis, ANZVASC's activities aim to: (i) promote best practice clinical care for patients with vasculitis; (ii) facilitate vasculitis research activities; (iii) bring together members and other physicians and health professionals who have an interest in vasculitis; (iv) take a key role in educational meetings in the field of vasculitis; (v) establish a clinical quality registry and (vi) advocate for people who have vasculitis in Australia and New Zealand.

The inaugural ANZVASC meeting was held in Brisbane on 24 August 2019 with over 70 attendees drawn from a range of disciplines. In addition, a well attended patient question and answer session was held. The formation of ANZVASC is timely and we invite all consumers, scientists and clinicians with an interest in vasculitis to join this group. It is an exciting time for people with vasculitis, their families and clinicians who are interested in advancing the care of patients with vasculitis in Australia and New Zealand.

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