

## INTRODUCTION AND OBJECTIVES

Anti-neutrophil cytoplasmic antibody associated vasculitis (AAV) causes a severe systemic vasculitis often involving lung and kidney. Treatment is with high dose glucocorticoids (GC) and immunosuppressants to achieve remission. AAV is a relapsing disease and patients are at risk from organ damage from vasculitis as well as adverse effects of treatment including those related to high dose GCs. Patients are confronted with the combination of a severe illness alongside difficult treatment decisions with both short and long term implications for their health.

This study aimed to examine the current literature relating to patient experience and quality of life (QoL) in AAV to identify themes and data gaps.

## METHODS

A Pubmed literature search (2012-2017) was performed to examine the recent published scientific evidence.

Key search words used were –

- (1) AAV **plus** each of the following as separate searches
- (2) Patient views, Patient information, Patient education, Patient burden, Patient experience, Patient reported outcomes, Health status, Patient perspectives, concordance, Patient reported experience, Quality of Life, employment, satisfaction, decision making.

Initial search strategy revealed that only 69 scientific publications (from over 900 relating to AAV in general) examined the research topic. These full publications were read to demonstrate that the relevance to the overall research objective was ranked as:

- (1) Moderate – 27
- (2) High 22
- (3) Very high – 20

A narrative review was performed of the higher ranking publications to reveal key themes relating to the patient experience in AAV

## RESULTS

Key themes demonstrated major challenges patients face plus significant evidence gaps.

- (1) **Patients have need for more information**– in contrast to other rare diseases and long term conditions there is little evidence regarding patient information needs and shared decision making. A single study found people with AAV seek information concerning their disease, treatment regimens and side effects and the results of investigations – with preference to receive from their physician. A linked survey of physicians found significant variation in the information given by physicians about therapy choices.
- (2) **Quality of life is low and fatigue is a major driver** - QoL was measured in particular during clinical trials comparing various treatment regimes. AAV patients have lower QoL compared with the general population and this influences their perception of future health. AAV patients identified fatigue and reduced activity as particular problems associated with heightened perception of exertion, depression and anxiety. The emphasis on fatigue links with the reduced ability to work observed consistently in studies from several countries.
- (3) **Patient experience needs more research and high dose glucocorticosteroids (GCs) have a major negative impact** - there was a lack of qualitative research studies examining patient views of living with AAV and/or the impact of treatment in comparison to the literature in other long term conditions.

One recent study examined patient views of high dose GCs and observed patients have views around the positive effectiveness to achieve remission but also that they are associated with important emotional, physical and social effects – patients have important views on balancing risks and benefits of GCs.

There are initiatives to improve measurement of AAV patient experience and the Outcome Measures in Rheumatology (OMERACT) Vasculitis Working Group has endorsed the PROMIS instruments for fatigue, physical functioning, and pain interference and the disease-specific AAV-PRO instrument. These will be invaluable tools for clinical trials of new therapies for AAV.

## CONCLUSIONS

This literature review of AAV patient experience has revealed important data gaps around provision of patient information, measurement of patient outcomes and impact of disease and therapy. Fatigue is a particular concern and high dose glucocorticosteroids

New instruments have been developed to examine patient reported outcomes in AAV but more data are required to further understand patient experience and their prioritized concerns.