Background - 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 - Pubmed literature search (2012-2017) was performed using key words of AAV plus Patient views, Patient information, Patient education, Patient burden, Patient experience, Patient reported outcomes, Health status, Patient perspectives, concordance, Patient experience, Patient reported experience, Quality of Life, employment, satisfaction, decision making.

Results - Literature search revealed an overall paucity of studies (under 30) relating directly to patient experience and patient reported outcomes. Key themes which emerged are;

1. Provision of information - 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. QoL - 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 and outcomes - 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 outcomes in clinical trials of new therapies for AAV.

Discussion - 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. 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.