Table 1. ANCA in EGPA cohort (n = 73). BPI = bactericidal permeability-increasing protein.

IFT / ELISA	No. of patients (%)
P-ANCA	11 (15.1)
C-ANCA	5 (6.8)
MPO-ANCA	8 (10.9)
PR3-ANCA	2 (2.7)
BPI-ANCA	1 (1.4)
PTX3-ANCA	5 (6.8)
OLM4-ANCA	2 (2.7)

Conclusion: We report on the detection of PTX3-, BPI- and OLM4-ANCA in addition to MPO- and PR3-ANCA in EGPA. OLM4-ANCA has been reported in 2 patients with non-vasculitic inflammatory symptoms previously [5]. Herein, detection of OLM4-ANCA in EGPA is reported for the first time. Our study shows that the presence of ANCA with various specificities other than MPO and PR3 contribute to a higher prevalence of ANCA in EGPA. Moreover, clinical manifestations differ between ANCA-negative EGPA and ANCA-positive EGPA, and between patients with different ANCA-specificities.

REFERENCES:

[1] Schönermarck U, et al. Prevalence and spectrum of rheumatic diseases associated with proteinase 3-antineutrophil cytoplasmic antibodies (ANCA) and myeloperoxidase-ANCA. Rheumatology 2001;40:178-84.
[2] Bremer P, et al. Getting rid of MPO-ANCA: a matter of disease subtype. Rheumatology 2013:752-4.
[3]Comarmond C, et al. Eosinophilic granulomatosis with polyangiitis (Churg-Strauss): clinical characteristics and long-term followup of the 383 patients enrolled in the French Vasculitis Study Group cohort. Arthritis Rheum 2013;65:270-81.
[4] Padoan R, et al. IgG anti-Pentraxin 3 antibodies are a novel biomarker of ANCA-associated vasculitis and better identify patients with eosinophilic granulomatosis with polyangiitis. J Autoimmun 2021;124:102725.
[5] Amirbeagi F, et al. Olfactomedin-4 autoantibodies give unusual c-ANCA staining patterns with reactivity to a subpopulation of neutrophils. J Leukoc Biol 2015;97:181-9.

Disclosure of Interests: None declared

DOI: 10.1136/annrheumdis-2022-eular.3083

POS0830 SYSTEMATIC LITERATURE REVIEW INFORMING THE 2022 UPDATE OF THE EULAR RECOMMENDATIONS FOR THE MANAGEMENT OF ANCA-ASSOCIATED VASCULITIS: FOCUS ON TREATMENT STRATEGIES

J. Schirmer[1], B. Sanchez-Alamo[2], S. Monti[3], B. Hellmich[4], D. Jayne[5]. [1]*University Medical Center Schleswig-Holstein, Clinic for Internal Medicine I, Section for Rheumatology, Kiel, Germany;* [2]*Lund University, Skåne University Hospital, Nephrology, Lund, Sweden;* [3]*University of Pavia, Fondazione IRCCS Policlinico S. Matteo, Rheumatology, Pavia, Italy;* [4]*University of Tübingen, Medius Kliniken, Internal Medicine, Rheumatology and Immunology, Kirchheim-Teck, Germany;* [5]*University of Cambridge, Addenbrooke's Hospital, Department of Medicine, Cambridge, United Kingdom*

Background: The 2008 and 2016 European Alliance of Associations for Rheumatology (EULAR) recommendations for the management of ANCA-associated vasculitis (AAV)[1,2] have supported clinicians with comprehensive recommendations for the treatment of patients with AAV in daily practice. During the past 5 years, the publication of several high-impact randomized-controlled studies have further improved the standard of care of AAV.

Objectives: The aim of this systematic review was to collect evidence supporting the 2022 update the AAV management recommendations.

Methods: The recommendations were developed based on an evidence-based approach as outlined in the 2014 EULAR standardized operating procedures (SOP)[3]. Areas of interest were adopted from the 2016 recommendations and updated by identifying additional topics through a Delphi process. Key questions were framed in the PICO (Population, Intervention, Comparator, Outcome) format and a search strategy consisting of keywords identifying treatment-related topics of interest was created based on the PICO questions. Aspects of drug treatment and other therapeutic interventions in AAV were included in the search, with a focus on remission induction, maintenance treatment and steroid sparing protocols. Outcomes such as survival, remission/relapses, infectious complications and malignancies were also covered. PubMed (Medline), Embase and the Cochrane Library databases were searched for articles providing data on the search questions. Abstracts of the annual meetings of EULAR, ACR, ERA-EDTA, ASN and the Vasculitis and ANCA Workshops were also screened, but restricted to randomized controlled clinical trials (RCTs). After deduplication publications were sorted by title and abstract first. There was full text review for articles eligible after title/abstract screening. The data were extracted from included articles and grouped according to the PICO questions. Data extraction results were collected in evidence tables.

The Cochrane revised tool for assessing risk of bias for RCTs (RoB2), ROBINS-1 for observational studies and AMSTAR II for meta-analyses were used for bias assessment. Evidence was categorized based on the GRADE system as per EULAR SOP[3].

Results: Based on the results of the Delphi, 11 topics related to therapeutic interventions were identified that were transformed into PICO questions (Table 1). Other items that received lower scores were added in the format of subquestions. Based on these research questions, search strings for the SLR were created.

Table 1. Key topics of interest for treatment strategies identified in the Delphi exercise grouped according to the PICO format

Patients	Intervention & Comparators	Outcome
Diagnosis	Cyclophosphamide	Disease-related outcomes
Granulomatosis with Polyangiitis	Rituximab	Treatment-related adverse events
Microscopic Polyangiitis	Mycophenolate	
Eosinophilic Granulomatosis with Polyangiitis	Methotrexate	
Disease severity	Azathioprine	
New-onset disease	Glucocorticoids	
Relapsing disease	Avacopan	
Organ- or life-threatening disease	Mepolizumab	
Not organ- or life-threatening disease	Plasma exchange	

The SLR was still ongoing at the time this abstract has been written and results of the SLR will be presented at the meeting.

Conclusion: This SLR identified recent developments affecting key areas of AAV treatment, that provide systematic evidence to inform the 2022 update of the EULAR recommendations for the management of AAV, which will also be presented at this meeting.

REFERENCES:

[1] Mukhtyar C, et al. EULAR Recommendations for the management of primary small and medium vessel vasculitis. Ann Rheum Dis. 2008;68:310-317.
[2] Yates M et al. EULAR/ERA-EDTA recommendations for the management of ANCA-associated vasculitis. Ann Rheum Dis. 2016 Sep;75(9):1583-94
[3] van der Heijde D et al. 2014 Update of the EULAR standardised operating procedures for EULAR-endorsed recommendations. Ann Rheum Dis. 2015 Jan;74(1):8-13.

Acknowledgements: The project is funded by EULAR.

Disclosure of Interests: Jan Schirmer: None declared, Beatriz Sanchez-Alamo: None declared, Sara Monti: None declared, Bernhard Hellmich Speakers bureau: Abbvie, BMS, Chugai, GSK, MSD, Novartis, Pfizer, Roche, Vifor, Consultant of: Boehringer, BMS, Chugai, GSK, InflaRx, Novartis, Roche, Vifor, David Jayne Speakers bureau: Vifor, Consultant of: Astra-Zeneca, Boehringer, BMS, Chemocentryx, Chugai, GSK, Novartis, Roche

DOI: 10.1136/annrheumdis-2022-eular.3156

POS0831 IGA VASCULITIS IN ADULTS, PEDIATRICS AND NON-VASCULITIC IGA NEPHROPATHY

S. Levanon[1], V. Gotlieb[2], Y. Kraus[3], I. Novofastovski[1], S. Brikman[1], A. Fawaz[1], M. Aghbariyya[1], Y. Butbul[4], A. Balbir-Gurman[5], R. Mader[1], A. Bieber[1]. [1]*Emek Medical Center, Rheumatology, Afula, Israel;* [2]*Emek Medical Center, Pediatric Rheumatology Service, Afula, Israel;* [3]*Emek Medical Center, Pathology, Afula, Israel;* [4]*The Ruth Rappaport Children's Hospital, Pediatric Rheumatology Service, Haifa, Israel;* [5]*Rambam Health Care Campus, Rheumatology, Haifa, Israel*

Background: IgA vasculitis (IgAV) has been extensively studied in children, while its natural history remains poorly studied in adults. Sparse data comparing childhood and adult-onset disease has shown significant differences in their clinical presentation, especially in the severity of renal involvement, which accounts for the major long-term morbidity. IgAV shares similar renal histologic features with IgA nephropathy (IgAN), while clinically IgAN is a chronic kidney disease which may lead to end stage renal disease and dialysis. The extent of kidney injury among adults with IgAV is still a matter of uncertainty.

Objectives: We aimed to evaluate clinical manifestations, laboratory data, treatment patterns and long-term outcomes of pediatric and adult-onset IgAV in comparison to IgAN.

Methods: This retrospective collaborative study examined medical records of adults and children with IgAV and IgAN adult patients admitted to rheumatology clinic and in hospital pediatric departments in a 13-year period (2007-2019). Diagnosis of adults with IgAV relied on the Ankara criteria and was confirmed by a consistent skin biopsy with positive IgA staining by immunofluorescence. Children with IgAV were included in our study on a clinical basis. All IgAN patients had a kidney biopsy proven disease. We analyzed and compared frequencies of clinical manifestations, laboratory findings, treatment regimens and long-term outcomes at one year follow-up. Finally, we assessed long term outcomes, such as time to dialysis and all-cause mortality, till the end of the follow-up time.