

Supplementary file 1

Methods

The recommendations were drafted according to the 2014 update of the EULAR standardised operating procedures (SOPs) for the development of EULAR-endorsed recommendations and the updated version of the Appraisal of Guidelines for Research & Evaluation (AGREE II) instrument, where applicable.^{3 4} After approval by the EULAR Executive Committee, the Convenor (BH) and the methodologist (RL) assembled a task force including 24 members. The group consisted of 19 clinical experts (including rheumatologists, internists, immunologists, a neurologist, a neuro-ophthalmologist, and an epidemiologist), from 11 European countries, India and China, two fellows (AA, SM), one health professional and two patients affected by the diseases under study. Three members of the task force (SM, CP, CD) were EMEUNET members.

Based on results of a Delphi survey among the task force, we defined eight key research questions addressing the management and treatment of LVV. As the original systematic literature review (SLR) for the 2008 recommendations dated back 10 years ago with different methodology (focus, bias assessment etc.), it was decided to conduct two completely new SLRs without time limits, focusing on general management and treatment respectively (Table 1).

Table S1. Topics for the Systematic Literature Search

SLR 1: General Management	SLR 2: Treatment
<ul style="list-style-type: none"> • diagnosis: recognition, referral criteria, fast-track diagnosis, role of imaging for diagnosis, role of biopsy for diagnosis, interdisciplinary work-up, considerations for sub-types of disease such as cranial/ischaemic/large vessel, isolated aortitis, IgG4-related disease, LVV disease in other vasculitides • prognostic and therapeutic implications of disease phenotypes: cranial vs extra-cranial, isolated aortitis, other forms including IgG-4 related disease, imaging, other biomarkers, comorbidities and complications, disease damage versus activity • long-term follow-up of patients: clinical assessments and frequency, imaging, patient-reported outcomes, physical therapies and management of complications • patient education and other aspects of patients-centered care 	<ul style="list-style-type: none"> • drug therapy: dosing, length of therapy, outcome and treatment-related side effects for the following drugs: glucocorticoids, methotrexate and other non-biologic immunosuppressive agents, tocilizumab and other biologic agents; • specific treatment of organ complications: loss of vision and stroke), relapsing, refractory, glucocorticoid-dependent disease • revascularisation procedures: indications for referral, management of aneurysms and/or vessel stenosis • adjunctive therapies and prophylaxis: aspirin, cardiovascular and cerebrovascular disease, infections, vaccination, osteoporosis

The research strategy was framed according to the Population, Intervention, Comparator, Outcome (PICO) format. Both SLRs included the same population terms (including GCA, TAK and other forms of LVV), focusing on different interventions and outcomes. Given the wide area of

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topics covered by each SLR, a comparator did not apply to our search strategies and was excluded. The two fellows (SM and AA) performed the SLR with the guidance of two experienced librarians and the methodologist (RL) by exploiting the following databases: MEDLINE, EMBASE and Cochrane CENTRAL. Evidence was gathered from inception of each database until the 31st of December 2017. Eligible studies were all full research articles of any study design, without language restrictions, but excluding single-case-reports and reviews. Summary of findings (SoF) tables were created. Following the 2014 EULAR standard operating procedures and 2017 additional guidance document for the development/update of EULAR recommendations, each article was assigned a level of evidence (LoE) according to the 2009 standards of the Oxford Centre for Evidence-Based Medicine and was systematically assessed for bias.³ The results of the two SLRs and a detailed outline of the search strategy, methods and obtained evidence are published separately (*NOTE TO THE EDITOR: references to be added when the accompanying manuscripts are finished*).

During a face-to-face meeting, the SoF-tables derived from the SLRs were presented and formed the basis for the generation of the recommendations. After discussion, task force members independently voted on each recommendation. Only the recommendations (here presented in italic font) were formally voted on during the face-to-face meeting, but not the subsequent paragraphs. Agreement of at least 75 % of the task force members was required in the first ballot, which was achieved for all recommendations. After the meeting, participants were asked via email to anonymously report their level of agreement on each recommendation and on the overarching principles on a scale of 0-10 (10 meaning full agreement). A research agenda was formulated from the gaps in the evidence and any controversial issues. The final manuscript was approved by the EULAR Executive Committee.