Updated: AAV treatment recommendations

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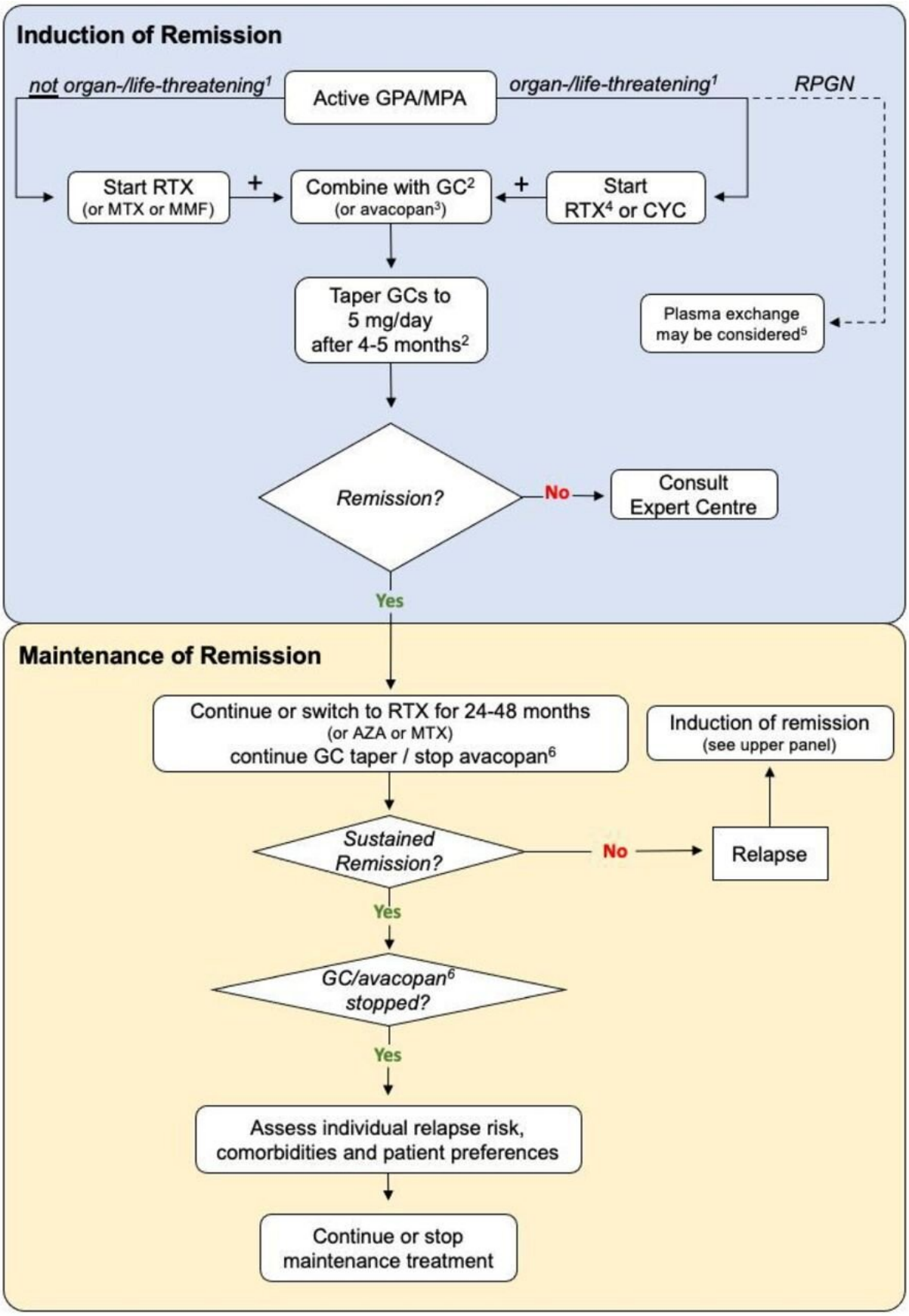
Induction of Remission
not organ-/life-threatening[1]
Active GPA/MPA
organ-/life-threatening[1]
RPGN
Start RTX
(or MTX or MMF)
+
Combine with GC[2]
(or avacopan[3])
+
Start
RTX[4] or CYC
Taper GCs to
5 mg/day
after 4-5 months[2]
Plasma exchange
may be considered[5]
Remission?
No
Consult
Expert Centre
Yes
Maintenance of Remission
Continue or switch to RTX for 24-48 months
(or AZA or MTX)
continue GC taper / stop avacopan[6]
Induction of remission
(see upper panel)
Sustained
Remission?
No
Relapse
Yes
GC/avacopan[6]
stopped?
Yes
Assess individual relapse risk,
comorbidities and patient preferences
Continue or stop
maintenance treatment

The 2022 EULAR algorithm for treatment of granulomatosis with polyangiitis (GPA) or microscopic polyangiitis (MPA). Dashed lines indicate supplementary action to consider. GC doses are provided as prednisolone equivalent. Credit: *Annals of the Rheumatic Diseases* (2023). DOI: 10.1136/ard-2022-223764

AAV is a multi-organ disease, which can be complex and may vary from person to person. Types of AAV include granulomatosis with polyangiitis (GPA), microscopic polyangiitis (MPA), and eosinophilic granulomatosis with polyangiitis (EGPA). This variety can make management challenging. Comorbidities, an individual's history, toxicities, medication availability and cost, and patient preferences should all be considered in the process of informed decision making.

EULAR first wrote recommendations for small and medium vessel vasculitis in 2009; this was subsequently updated in 2016 with more of a focus on AAV. The new 2022 update includes substantial alterations, including the introduction of overarching principles and new recommendations on ANCA-testing, glucocorticoid treatment, the use of agents with novel modes of action, and preventative measures against infection. In addition, new data has allowed the development of some separate recommendations for people with specific AAV subtypes—namely GPA, MPA, and EGPA.

The updated EULAR recommendations were developed by a multidisciplinary task force that included rheumatologists, internists, nephrologists, and methodologists, as well as patient representatives. The information is based on evidence collected from the literature and a survey among the task force members.

The new paper developed by EULAR and published in the March 2023 issue of the *Annals of the Rheumatic Diseases* includes 4 overarching principles and 17 recommendations. The principles say that people with AAV should be offered best care based on shared decision making between the patient and the physician.

People with AAV should have access to education so that they understand the impact of their disease, and be able to recognize key symptoms and complications. They also recommend that people with AAV be screened for treatment-related adverse effects and comorbidities, and offered appropriate preventative treatment or lifestyle advice.

Finally, the overarching principles stress that AAV are rare, heterogeneous, and potentially life-and organ-threatening diseases which require multidisciplinary management that includes vasculitis experts. The individual recommendations outline the various treatment options for people to induce and maintain remission, depending on their specific diagnosis and manifestations.

One additional key element of these updated recommendations is inclusion of definition of disease activity states. These can vary between different clinical trials, but EULAR recommend these consensus definitions to further understanding and make it easier to compare results. This includes definitions of active disease, remission, relapse, and refractory disease.

EULAR believes that these recommendations will help to streamline the management of people with AAV—while they are not intended to be used as a one-size-fits-all strategy, and may need to be used alongside other recommendations and treatment algorithms, depending on each person's disease, manifestations, and comorbidities. It is hoped these recommendations will be incorporated into everyday clinical practice to

effectively manage AAV and to improve quality of care.

More information: Bernhard Hellmich et al, EULAR recommendations for the management of ANCA-associated vasculitis: 2022 update, *Annals of the Rheumatic Diseases* (2023). DOI: 10.1136/ard-2022-223764

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