REBELLIOUS MEDIC · RHEUMATOLOGY ALGORITHM SERIES

ANCA-Associated Vasculitis

Remission induction, maintenance, relapse, and refractory-disease management for GPA/MPA and EGPA, condensed from 41 recommendations and 10 position statements.

2021 ACR/VF Guideline
Guideline for the Management of ANCA-Associated Vasculitis
Chung SA, et al. · Arthritis Care Res 2021;73:1088-1105

GRADE — Cond. For · Cond. Against · Ungraded (UPS)

Virtually all graded recommendations are conditional, low-quality evidence. GCs = glucocorticoids.

1 GPA/MPA — Severe Disease Induction

INDUCTION AGENT

- Cond. — **Rituximab** preferred over cyclophosphamide for remission induction.
- UPS — **IV pulse or high-dose oral GCs** as initial therapy.

PLASMA EXCHANGE & DOSING

- Cond. — **Against** routine addition of plasma exchange for active glomerulonephritis or alveolar hemorrhage.
- Cond. — **Reduced-dose GC** regimen preferred over standard-dose for induction.

2 GPA/MPA — Nonsevere Induction & Maintenance

INDUCTION & AGENT CHOICE

- Cond. — **Nonsevere induction** — methotrexate + GCs preferred over cyclophosphamide, rituximab, azathioprine, mycophenolate, or TMP/SMX; over GCs alone.
- Cond. — **Maintenance after severe disease** — rituximab preferred over methotrexate/azathioprine.
- Cond. — Methotrexate/azathioprine preferred over mycophenolate or leflunomide for maintenance.

DOSING & DURATION

- Cond. — **Rituximab re-dosing** — scheduled re-dosing preferred over titrating by ANCA titer or CD19+ B-cell counts.
- Cond. — **Against** adding TMP/SMX to other maintenance therapy in GPA.
- UPS — **Duration** of non-GC and GC maintenance guided by clinical condition and patient values/preferences.

3 GPA/MPA — Relapse, Refractory & Other

RELAPSE & REFRACTORY

- Cond. — **Severe relapse** — rituximab preferred over cyclophosphamide if not on rituximab maintenance; switch to cyclophosphamide if relapse occurs on rituximab maintenance.
- Cond. — **Refractory disease** — switch between rituximab/cyclophosphamide rather than combining; add IVIG if refractory to induction or other agents unavailable.
- Cond. — **Against** dosing therapy by ANCA titer alone.

AIRWAY & PROPHYLAXIS

- UPS — **Sinonasal/airway lesions** — nasal rinses/topical therapy; reconstructive surgery for septal defects if desired.
- Cond. — **Immunosuppression** preferred over surgery alone for subglottic stenosis or mass lesions.
- UPS — **PJP prophylaxis** with rituximab/cyclophosphamide; evaluate renal transplant if ESKD achieves remission.

4 EGPA — Remission Induction

SEVERE DISEASE

- UPS — **IV pulse or high-dose oral GCs**; cyclophosphamide or rituximab as induction options.
- Cond. — **Cyclophosphamide or rituximab** preferred over mepolizumab.

NONSEVERE DISEASE

- Cond. — **Mepolizumab + GCs** preferred over methotrexate/azathioprine/mycophenolate + GCs.
- Cond. — Methotrexate/azathioprine/mycophenolate + GCs preferred over cyclophosphamide/rituximab + GCs or GCs alone.

5 EGPA — Maintenance & Relapse

- Cond. — **Maintenance** — methotrexate/azathioprine/mycophenolate preferred over rituximab or mepolizumab after cyclophosphamide induction.
- Cond. — **Severe relapse** — rituximab preferred over cyclophosphamide (or over switching agents if already on rituximab) for re-induction.
- Cond. — **Nonsevere relapse** — add mepolizumab preferred over switching agents, adding csDMARDs, or adding omalizumab (high IgE).

6 EGPA — Other Considerations

LEUKOTRIENE INHIBITORS & WORKUP

- UPS — **Leukotriene inhibitors** — continue rather than discontinue in newly diagnosed EGPA; not contraindicated with active asthma/sinonasal disease.
- UPS — **Workup** — echocardiogram at diagnosis; use the Five-Factor Score to guide therapy intensity.

SINONASAL & PROPHYLAXIS

- UPS — **Sinonasal disease** — nasal rinses and topical therapy may be considered.
- UPS — **Infection prophylaxis** — PJP prophylaxis with cyclophosphamide or rituximab.

Chung SA, Langford CA, Maz M, et al. 2021 American College of Rheumatology/Vasculitis Foundation Guideline for the Management of Antineutrophil Cytoplasmic Antibody-Associated Vasculitis. *Arthritis Care Res.* 2021;73(8):1088-1105.