REBELLIOUS MEDIC · RHEUMATOLOGY ALGORITHM SERIES

Polyarteritis Nodosa

Diagnostic testing, remission induction/maintenance, refractory disease, and DADA2 recognition for medium-vessel vasculitis.

2021 ACR/VF Guideline
Guideline for the Management of Polyarteritis Nodosa
Chung SA, et al. · Arthritis Care Res 2021;73:1061-1070

GRADE — Strong · Cond. For · Cond. Against

All recommendations conditional unless marked Strong (very low-quality evidence throughout). GCs = glucocorticoids; DADA2 = deficiency of adenosine deaminase 2.

1 Diagnostic Testing

- Cond. — **Suspected PAN** — abdominal vascular imaging to aid diagnosis and determine disease extent.
- Cond. — **Skin involvement** — deep-skin biopsy (reaching medium-sized dermal vessels) preferred over superficial punch biopsy.
- Cond. — **Peripheral neuropathy** — combined nerve + muscle biopsy preferred over nerve biopsy alone.
- Cond. — **Follow-up** — repeat abdominal imaging if prior severe abdominal involvement, now asymptomatic; serial neuro exams (not repeat EMG/NCS) to monitor motor neuropathy.

2 Treatment of Active Disease

- Cond. — **Severe, new diagnosis** — IV pulse GCs preferred over high-dose oral GCs; cyclophosphamide + high-dose GCs over GCs alone; cyclophosphamide + GCs over rituximab + GCs.
- Cond. — **Cyclophosphamide intolerant** — other non-GC immunosuppressive agent + GCs preferred over GCs alone.
- Cond. — **Nonsevere disease** — non-GC immunosuppressive agent + GCs preferred over GCs alone.
- Cond. — **Against** plasmapheresis added to cyclophosphamide + GCs.

3 Remission Maintenance & Refractory Disease

- Cond. — **After cyclophosphamide remission** — transition to another non-GC agent rather than continuing cyclophosphamide.
- Cond. — **On non-GC therapy in remission** — discontinue non-GC agent after 18 months preferred over indefinite treatment.
- Cond. — **Refractory to non-cyclophosphamide agent** — switch to cyclophosphamide preferred over increasing GCs alone.

GC duration not well established — guide by clinical condition, patient values, and preferences.

4 Other Considerations

Nerve/muscle involvement — physical therapy recommended.

- Strong — **DADA2** — TNF inhibitor preferred over GCs alone. Diagnosed via ADA2 sequencing or functional assay; consider in early-onset, familial, or lacunar-stroke PAN phenotypes.

Chung SA, Gorelik M, Langford CA, et al. 2021 American College of Rheumatology/Vasculitis Foundation Guideline for the Management of Polyarteritis Nodosa. *Arthritis Care Res.* 2021;73(8):1061-1070.