Eosinophilic granulomatosis with polyangiitis (EGPA)

Churg–Strauss syndrome

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Eosinophilic granulomatosis with polyangiitis (EGPA)
Churg–Strauss syndrome

- A rare anti- neutrophil cytoplasmic antibody (ANCA)-associated vasculitis, characterized by:
  - Asthma
  - Eosinophilia
  - Tissue eosinophilia, necrotizing vasculitis and eosinophil-rich granulomatous inflammation

- The diagnosis and management of EGPA are often challenging and require

An integrated, multidisciplinary approach

Emmi, G., E. *et al.* Evidence-Based Guideline for the diagnosis and management of eosinophilic granulomatosis with polyangiitis. *Nat Rev Rheumatol* **19**, 378–393 (2023).

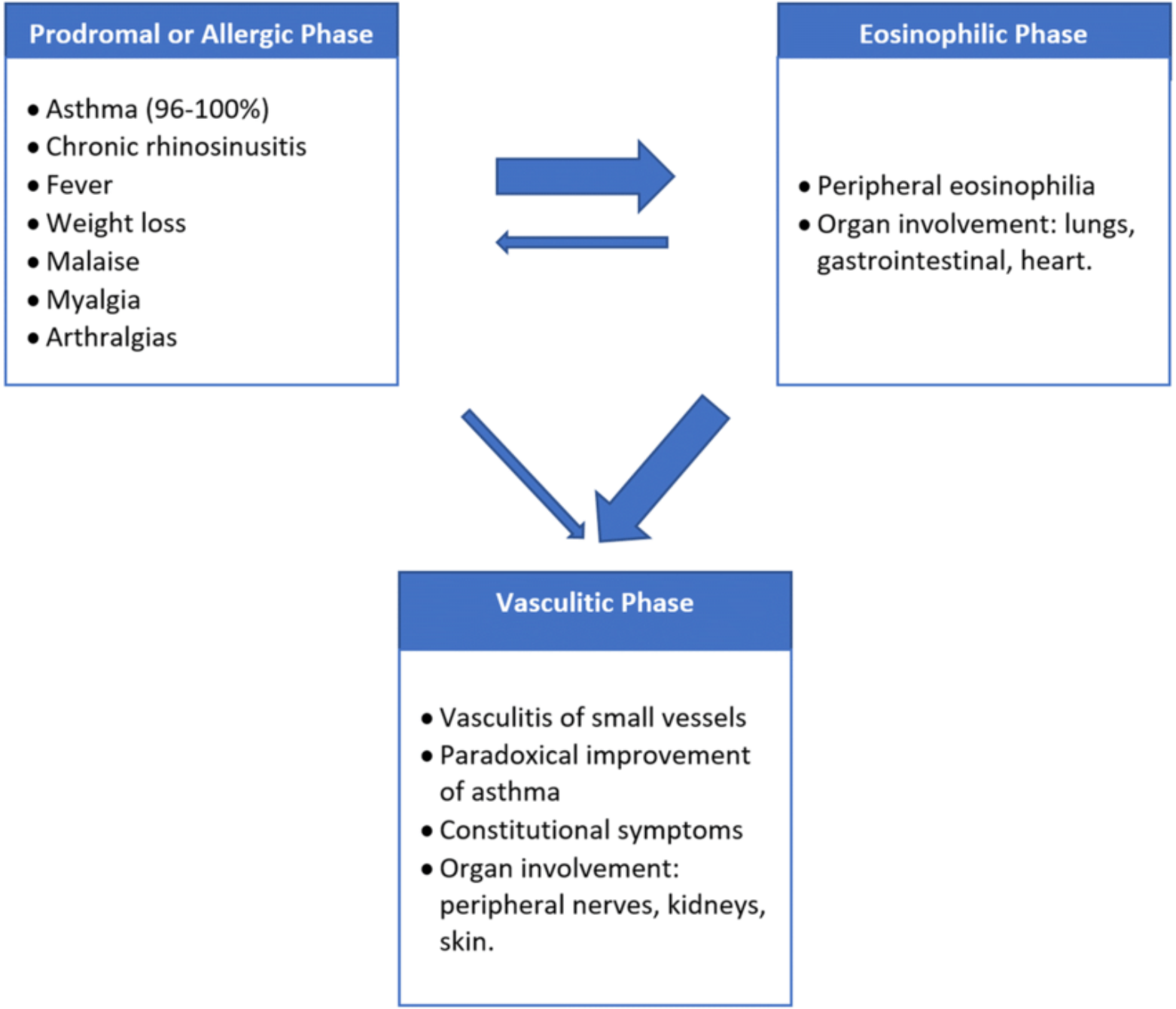
Eosinophilic granulomatosis with polyangiitis (EGPA)
Epidemiology

- The incidence : between 0.5 and 4.2 cases per million people per year
- The prevalence: between 10 and 14 cases per million globally
- Comparable in men and women
- The mean age at diagnosis: ~50 years
- Paediatric cases are extremely rare

Emmi, G., E. *et al.* Evidence-Based Guideline for the diagnosis and management of eosinophilic granulomatosis with polyangiitis.*Nat Rev Rheumatol* **19**, 378–393 (2023).

Three different phases

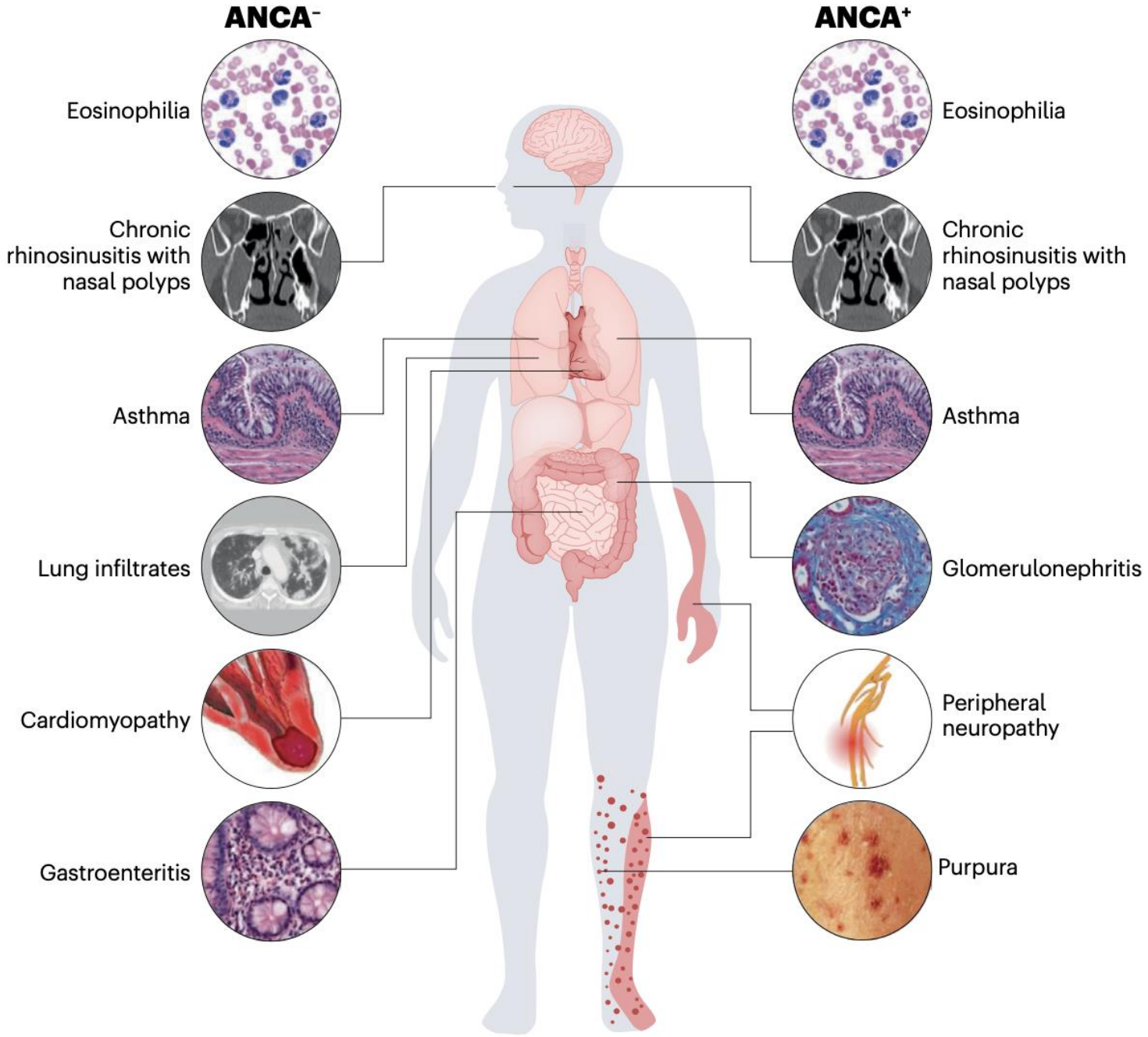
- These phases often overlap
- Do not necessarily develop in the aforementioned sequence
- Some patients do not manifest vasculitic complications



Vega Villanueva KL, Espinoza LR. Eosinophilic Vasculitis. Curr Rheumatol Rep. 2020 Jan 11;22(1):5.

The clinical phenotype of EGPA

- Quite heterogeneous
- Not straightforward diagnosis
- ANCA (against MPO) in ~40%
- Features of vasculitis more often in ANCA^{+} patients
- Eosinophilic features more frequent in ANCA^{-} patients



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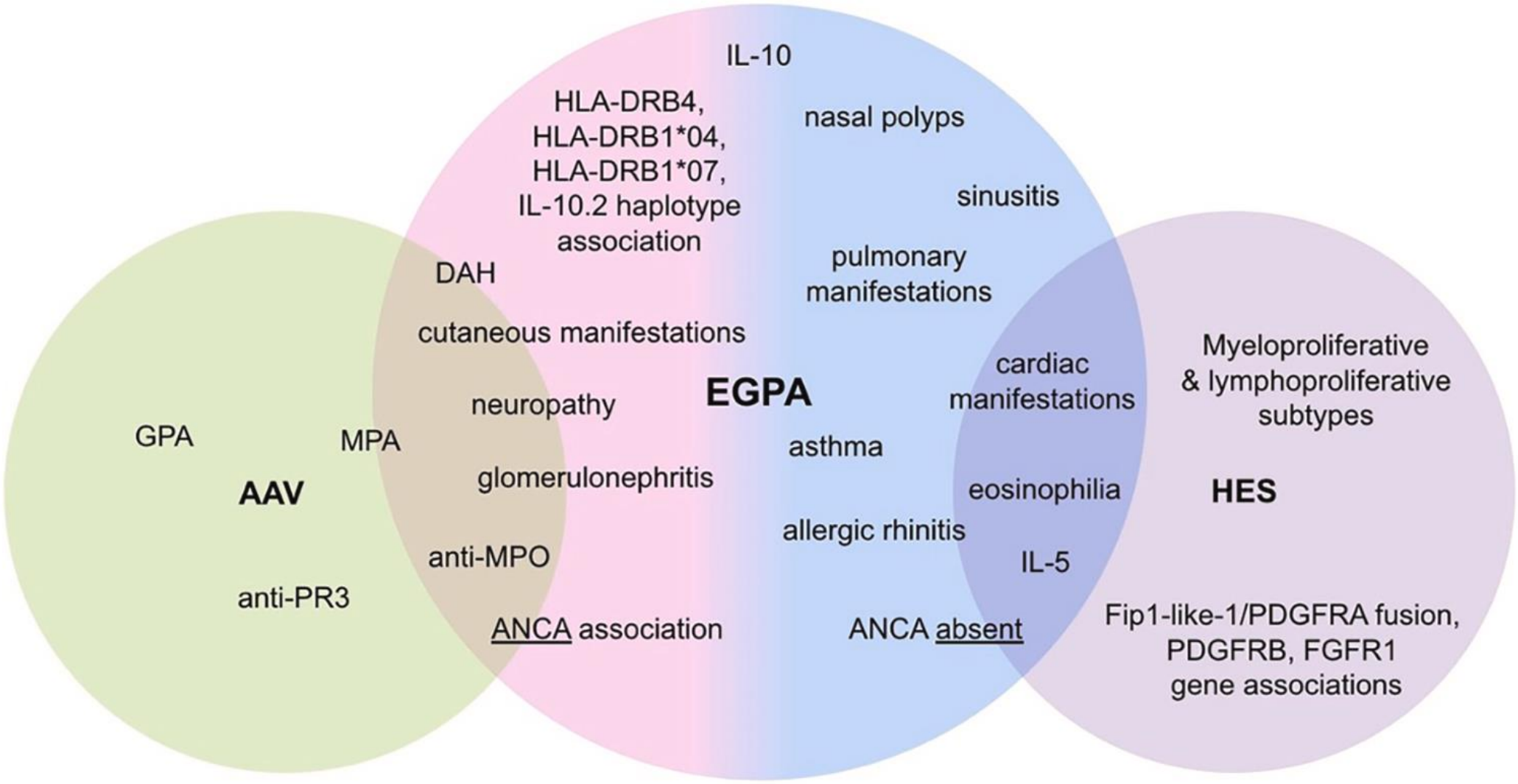
Overlapping syndromes with their association immunological and cliniclal manifestations



White J, Dubey S. Eosinophilic granulomatosis with polyangiitis: A review. Autoimmun Rev. 2023 Jan;22(1):103219.

REVIEW

MAYO CLINIC PROCEEDINGS

HES and EGPA: Two Sides of the Same Coin

Paneez Khoury, MD; Praveen Akuthota, MD; Namhee Kwon, MD, PhD;
Jonathan Steinfeld, MD; and Florence Roufosse, MD, PhD

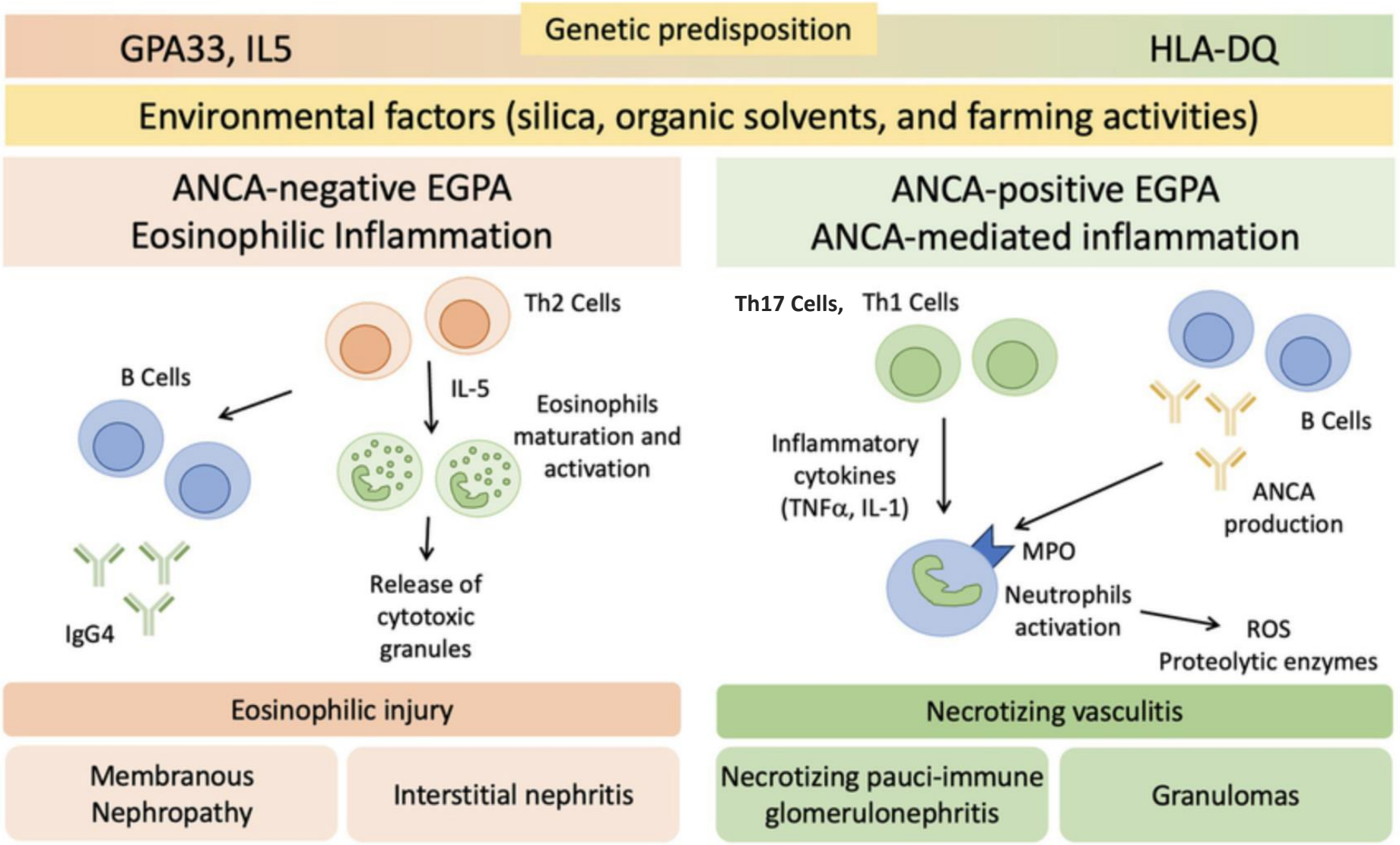
Khoury P, et al. HES and EGPA: Two Sides of the Same Coin. Mayo Clin Proc. 2023 Jul;98(7):1054-1070.

HES and EGPA: Two Sides of the Same Coin

- Differentiation between ANCA-negative EGPA and I-HES
  - No validated reliable biomarker
  - One recent study proposed
    - In patients with eosinophilia and asthma at diagnosis
      - Low serum CRP levels may be suggestive of I-HES
      - Mediastinal lymphadenopathy associated with EGPA

Khoury P, et al. HES and EGPA: Two Sides of the Same Coin. Mayo Clin Proc. 2023 Jul;98(7):1054-1070.

Pathogenesis of Renal Involvement in EGPA



cigarette smoking was associated with a lower risk.

Reggiani, et al. (2023). Renal involvement in eosinophilic granulomatosis with polyangiitis. Frontiers in Medicine. 10. 10.3389/fmed.2023.1244651.

Eosinophilic granulomatosis with polyangiitis (EGPA)
Prodromal features

- **Adult-onset asthma (**95–100%) **is a key manifestation**
  - Several years before of other systemic features
  - Typical seasonal variations are not observed, unlike classical asthma
  - Negative allergy investigations and sputum eosinophilia

- Chronic recurrent rhinosinusitis, and nasal polyposis (50%)
- Otitis media (serous and purulent)
- Progressive sensorineural hearing loss
- Unilateral facial palsy

- Eosinophilic single organ disease such as otitis media:
  - Differentiating from limited forms of EGPA can be very challenging.
  - Diagnosis is often made on histology

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Eosinophilic granulomatosis with polyangiitis (EGPA)
Pulmonary involvement

- Pulmonary infiltrates (40–70%)
  - The most typical manifestation
  - CXR:
    - Patchy, peripheral and migratory consolidation
  - Other lesions on HRCT
    - Ground-glass opacification, non-cavitating small centrilobular nodules, bronchial wall thickening and ‘tree-in-bud’ sign
- Diffuse alveolar haemorrhage (DAH) (3-4%)
- Pleural effusions
  - Secondary to eosinophilic pleurisy or eosinophilic cardiomyopathy-associated congestive cardiac failure

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Eosinophilic granulomatosis with polyangiitis (EGPA)
Cardiac involvement

- An adverse prognostic indicator, particularly in over 2/3 of ANCA^{-} patients
- One third of EGPA deaths and a 14% reduction in five-year survival, compared to those without heart disease
- A prognosticator of frequent **disease relapse**

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Eosinophilic granulomatosis with polyangiitis (EGPA)
Cardiac involvement

- Various cardiac structures can be involved
  - Cardiomyopathy
  - Eosinophilic coronaritis
  - Left ventricular dysfunction
  - Valvular insufficiencies
  - Pericardial effusions

- Of the patients with cardiac involvement, over half had (MRI) or histologically- confirmed **endomyocarditis**
  - The most severe manifestation given its potential fatal outcomes
  - Severe cardiac dysfunction and occasional intra-cardiac thrombi

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Eosinophilic granulomatosis with polyangiitis (EGPA)
Cardiac involvement

- Other features
  - Myocardial fibrosis
  - Restrictive, dilated or ischaemic cardiomyopathy
  - Constrictive or acute pericarditis (with pericardial effusions with potential tamponade)
  - Conduction defects
  - Ventricular or supraventricular dysrhythmias
  - Sudden death
- **Late myocardial gadolinium enhancement on cardiac MRI (cMRI)**
  - **Highly sensitive for detecting cardiac inflammation and fibrosis**
- cMRI abnormalities without clinical features of cardiac disease
  - In a high proportion of patients
  - The prognostic and clinical significance of this remains unclear

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Eosinophilic granulomatosis with polyangiitis (EGPA)
Gastrointestinal features (20–50%)

- An eosinophilic gastroenteritis
  - Non-specific abdominal pain, diarrhoea and haematochezia
  - In severe EGPA as a prelude to the vasculitic phase
  - Hard to diagnose
- The vasculitic process in small-to-medium vessels which are not abundant in the intestinal mucosa
  - Diagnostic challenges on endoscopic biopsy
  - Reports of histologically-confirmed mucosal necrotising vasculitides
- Endoscopic investigations
  - Large bowel mucosal ulcerations
- **Capsule endoscopy is more useful**
  - More frequently involvement of small bowel

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Eosinophilic granulomatosis with polyangiitis (EGPA)
Gastrointestinal features

- Gastrointestinal involvement
  - A poor prognostic indicator
  - Progression to life-threatening complications
    - Bowel perforation
    - Peritonitis
    - Pancreatitis and in some cases
    - Bowel ischaemia secondary to mesenteric vasculitis

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Eosinophilic granulomatosis with polyangiitis (EGPA)
Neurological features (50–75%)

- Peripheral neurological features during the early vasculitis phase of illness

- Severe and sensorimotor Peripheral neuropathy
  - Mononeuritis multiplex due to axonal injury
  - Asymmetrical or symmetrical polyneuropathies
  - Sensory deficits or neuropathic pain

- Sural nerve biopsies
  - Epineural necrotising vasculitis
    - Nerve ischaemic injury

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Eosinophilic granulomatosis with polyangiitis (EGPA)
Renal disease (25–30%)

- Less common and severe, compared with GPA and MPA
- The commonest renal presentation
  - Necrotising pauci-immune glomerulonephritis in $ANCA^{+}$ EGPA
- Less common and atypical presentations in $ANCA^{-}$ disease
  - Membranous nephropathy
  - Membranoproliferative glomerulonephritis
- Another common feature on renal histology
  - Interstitial nephritis with eosinophilic infiltration

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Eosinophilic granulomatosis with polyangiitis (EGPA)
Renal disease

- ANCA positivity was more frequent
  - In cases with renal involvement (75% vs. 25.7%) up to 84% in ANCA+ cases

- Clinical presentation ranging from urinary abnormalities to AKI and RPGN
  - The **need for renal biopsy** to confirm and characterize the specific kidney modifications

Reggiani Fet al. Renal involvement in eosinophilic granulomatosis with polyangiitis. Front Med (Lausanne). 2023 Sep 18;10:1244651.

Eosinophilic granulomatosis with polyangiitis (EGPA)
Renal disease

- Necrotizing and transmural arteritis of small and/or medium sized arteries at renal biopsy
  - A rare presentation in ANCA^{+} cases of EGPA
  - A worse prognostic significance

- Early renal involvement
  - Compared to anti-MPO^{+} MPA patients, in which is diagnosed at advanced stages
  - Probably determined by the severity of extra-renal symptoms, which lead to a rapid diagnosis of systemic vasculitis

Reggiani Fet al. Renal involvement in eosinophilic granulomatosis with polyangiitis. Front Med (Lausanne). 2023 Sep 18;10:1244651.

Eosinophilic granulomatosis with polyangiitis (EGPA)
Renal disease

- Overlap syndrome between AAV and IgG4-related disease
  - Both MN and IgG4+ plasma cell rich interstitial nephritis manifestations of IgG4-related disease

- Obstructive uropathy due to ureteral involvement

Reggiani Fet al. Renal involvement in eosinophilic granulomatosis with polyangiitis. Front Med (Lausanne). 2023 Sep 18;10:1244651.

Eosinophilic granulomatosis with polyangiitis (EGPA)
Cutaneous manifestation (50%)

- Mostly in the vasculitic phase
- Typically haemorrhagic lesions
  - **Palpable purpura**, ecchymoses, petechiae, haemorrhagic bullae

- Dermal or subcutaneous nodules and papules
  - Typically distributed over the scalp or bilateral extensor surfaces

- Urticaria
- Livedo reticularis
- Erythematous macules

White J, Dubey S. Eosinophilic granulomatosis with polyangiitis: A review. Autoimmun Rev. 2023 Jan;22(1):103219.

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CLASSIFICATION CRITERIA FOR **EOSINOPHILIC GRANULOMATOSIS WITH POLYANGIITIS**

CONSIDERATIONS WHEN APPLYING THESE CRITERIA

- **These classification criteria should be applied to classify a patient as having eosinophilic granulomatosis with polyangiitis when a diagnosis of small- or medium-vessel vasculitis has been made**
- **Alternate diagnoses mimicking vasculitis should be excluded prior to applying the criteria**

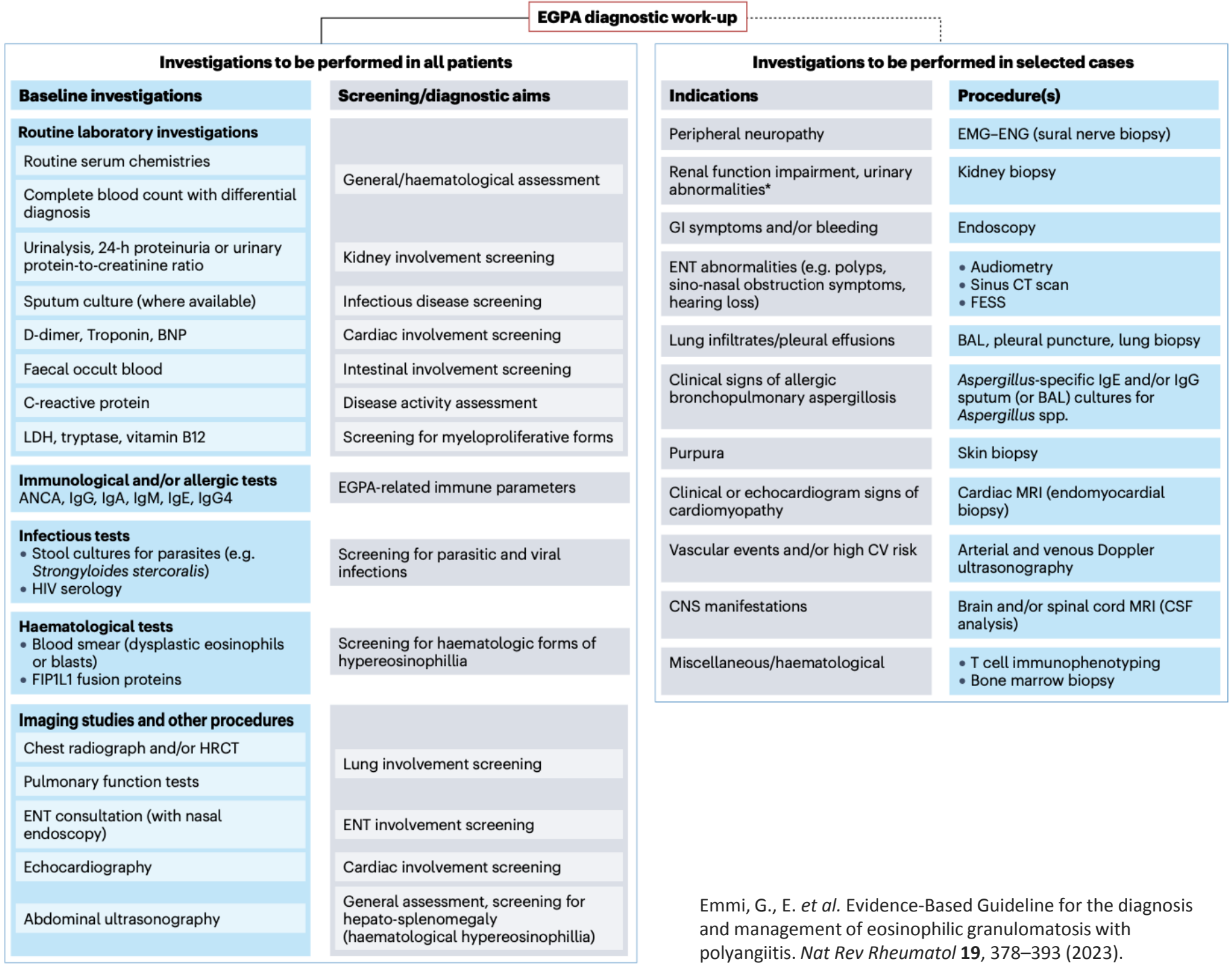
CLINICAL CRITERIA

Obstructive airway disease	+3
Nasal polyps	+3
Mononeuritis multiplex	+1

LABORATORY AND BIOPSY CRITERIA

Blood eosinophil count ≥ 1 x10^9/liter	+5
Extravascular eosinophilic-predominant inflammation on biopsy	+2
Positive test for cytoplasmic antineutrophil cytoplasmic antibodies (cANCA) or antiproteinase 3 (anti-PR3) antibodies	-3
Hematuria	-1

Sum the scores for 7 items, if present. A score of ≥ 6 is needed for classification of EOSINOPHILIC GRANULOMATOSIS WITH POLYANGIITIS.



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Investigations to be performed in all patients

Baseline investigations	Screening/diagnostic aims
Routine laboratory investigations	
Routine serum chemistries	General/haematological assessment
Complete blood count with differential diagnosis	
Urinalysis, 24-h proteinuria or urinary protein-to-creatinine ratio	Kidney involvement screening
Sputum culture (where available)	Infectious disease screening
D-dimer, Troponin, BNP	Cardiac involvement screening
Faecal occult blood	Intestinal involvement screening
C-reactive protein	Disease activity assessment
LDH, tryptase, vitamin B12	Screening for myeloproliferative forms

Investigations to be performed in all patients

Immunological and/or allergic tests ANCA, IgG, IgA, IgM, IgE, IgG4	EGPA-related immune parameters
Infectious tests • Stool cultures for parasites (e.g. *Strongyloides stercoralis*) • HIV serology	Screening for parasitic and viral infections
Haematological tests • Blood smear (dysplastic eosinophils or blasts) • FIP1L1 fusion proteins	Screening for haematologic forms of hypereosinophillia

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Investigations to be performed in all patients

Imaging studies and other procedures	
Chest radiograph and/or HRCT	Lung involvement screening
Pulmonary function tests	
ENT consultation (with nasal endoscopy)	ENT involvement screening
Echocardiography	Cardiac involvement screening
Abdominal ultrasonography	General assessment, screening for hepato-splenomegaly (haematological hypereosinophillia)

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Investigations to be performed in selected cases

Indications	Procedure(s)
Peripheral neuropathy	EMG–ENG (sural nerve biopsy)
Renal function impairment, urinary abnormalities*	Kidney biopsy
GI symptoms and/or bleeding	Endoscopy
ENT abnormalities (e.g. polyps, sino-nasal obstruction symptoms, hearing loss)	• Audiometry • Sinus CT scan • FESS
Lung infiltrates/pleural effusions	BAL, pleural puncture, lung biopsy
Clinical signs of allergic bronchopulmonary aspergillosis	*Aspergillus*-specific IgE and/or IgG sputum (or BAL) cultures for *Aspergillus* spp.
Purpura	Skin biopsy
Clinical or echocardiogram signs of cardiomyopathy	Cardiac MRI (endomyocardial biopsy)
Vascular events and/or high CV risk	Arterial and venous Doppler ultrasonography
CNS manifestations	Brain and/or spinal cord MRI (CSF analysis)
Miscellaneous/haematological	• T cell immunophenotyping • Bone marrow biopsy

Eosinophilic granulomatosis with polyangiitis (EGPA)
ANCA

- ANCA status could have prognostic implications
- Worse survival in ANCA-negative patients
  - Probably due the higher frequency of cardiac involvement
- More frequent relapses in ANCA-positive patients
- **ANCA status itself is not useful in the choice of treatment**

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Eosinophilic granulomatosis with polyangiitis (EGPA)
survival prediction

- **The Five-Factor Score (FFS)**

Original 1996 FFS	Revised 2011 FFS
Cardiac involvement	Age > 65 years
Gastrointestinal disease (bleeding, perforation, infarction, or pancreatitis)	Cardiac insufficiency
Renal insufficiency (plasma creatinine concentration >1.6 mg/dL [141 mmol/L])	Renal insufficiency (stabilized peak creatinine 1.7 mg/dL [150 micromol/L])
Proteinuria (>1 g/day)	Gastrointestinal involvement
Central nervous system involvement	Absence of ENT manifestations (presence is associated with a better prognosis)

The FFS score ranges from 0 to 2: a score of 0 is given when none of the factors is present, a score of 1 for 1 factor, and a score of 2 for 2 or more factors.

- **Peripheral neuropathy**
- **Other rare manifestations (for example, alveolar haemorrhage)**

Active severe EGPA

With life- or organ-threatening manifestations (e.g., alveolar hemorrhage, **renal involvement**, nervous system involvement, cardiac involvement, gastrointestinal involvement)

	2021 ACR/VF guidelines	2022 EULAR recommendations	2023 evidence-based guidelines
Remission Induction	Pulse IV or high-dose daily oral GCs + CYC or RTX	High-dose daily oral GCs + CYC (or RTX)	(Pulse IV followed by) high-dose daily oral GCs + CYC or RTX
Remission Maintenance	*Remission with CYC* Switch CYC to MTX, AZA or MMF *Remission with RTX* Consider RTX prosecution	Switch CYC to AZA, MTX, MEPO or RTX	GCs + RTX and/or MEPO and/or DMARDs
Relapse Treatment	*Severe disease relapse after remission with CYC or RTX* Pulse IV or high-dose daily oral GCs + RTX	*Severe disease relapse* High-dose daily oral GCs + RTX	*Severe disease relapse* (Pulse IV followed by) high-dose daily oral GCs + CYC or RTX

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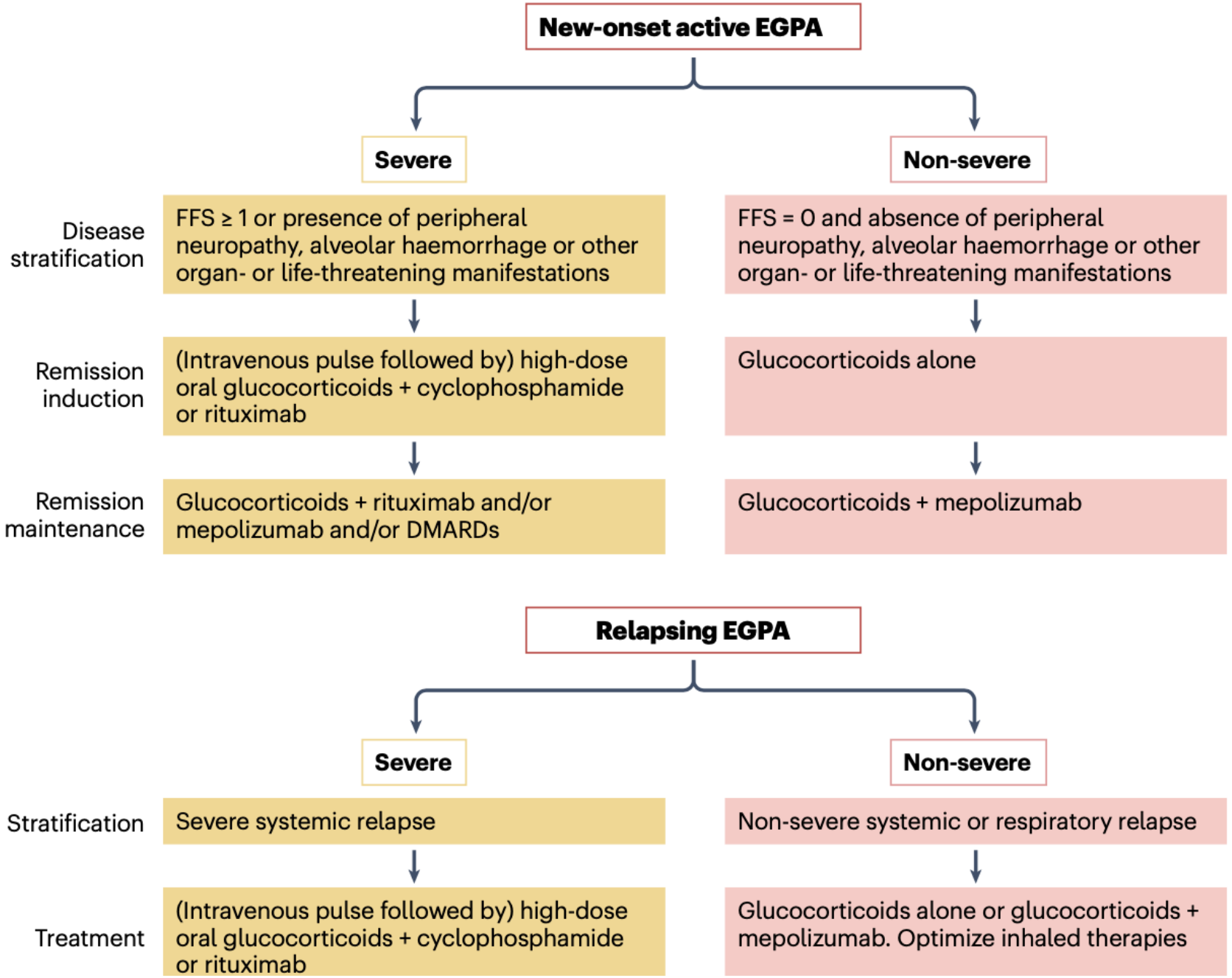

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Eosinophilic granulomatosis with polyangiitis (EGPA)
treatment

- The REOVAS trial
  - No significant differences in the rates of response to **rituximab** between ANCA^{+} and ANCA^{-} patients

- The MIRRA trial
  - No significant difference in response to **mepolizumab** between the two subgroups

- ANCA status should not influence treatment decisions
  - Even though it denotes differences in clinical phenotype and genetic backgrounds

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Eosinophilic granulomatosis with polyangiitis (EGPA)
treatment

- **A stepwise approach for respiratory relapses**
- Optimized topical therapies (for example, bronchodilators)
- Increment of the dose of oral glucocorticoids and short courses of high-dose glucocorticoids (0.5–1 mg/kg per day for 5–7 days)
- Mepolizumab

- **Relapsing ENT disease**
- Functional endoscopic sinus surgery for that does not adequately respond to the above approach

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Eosinophilic granulomatosis with polyangiitis (EGPA)
Refractory disease

- **Definition**
  - Unchanged or increased **disease activity after 4 weeks** of appropriate remission-induction therapy
  - The persistence or worsening of systemic manifestations should be distinguished from that of respiratory manifestations

- **EGPA can be defined as refractory only after addressing the following issues**:
  - Re-evaluation the primary diagnosis
  - Exclusion of other aetiologies such as infections or malignancies
  - Checking the appropriateness of the remission-induction treatment
  - Assessment of the patient's compliance with the remission-induction regimen
  - Distinguishing the persistently active manifestations from irreversible damage

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Eosinophilic granulomatosis with polyangiitis (EGPA)
Refractory disease

- Different therapeutic options including:
  - Other anti-IL-5 agents (benralizumab and reslizumab)
  - Plasma exchange
  - Intravenous immunoglobulin therapy
  - Anti-IgE agents
  - IFNα or mycophenolate mofetil in selected patients

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Eosinophilic granulomatosis with polyangiitis (EGPA)
Disease activity

- **No reliable biomarkers to measure disease activity in EGPA**

- **Assessment of disease activity by using validated clinical tools**
  - Detecting signs and symptoms of active disease
  - Appropriate imaging **or**
  - Functional studies (such as PFT, EMG–NCV and echocardiography) **and**
  - Routine laboratory tests

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Eosinophilic granulomatosis with polyangiitis (EGPA)
Disease activity

- The **eosinophil count** is routinely assessed
- Markedly high eosinophil counts at diagnosis and decreased during remission
- **Relapses can occur without an increase in the eosinophil count**
  - In a cohort study of 141 patients, the eosinophil count — as well as ESR and serum CRP and IgE concentrations — showed weak or no association with disease activity and disease flares.
  - Limited role of these parameters as longitudinal biomarkers

- Monitoring of **serum IgG4** concentration for the assessment of disease activity
  - It is controversial

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Eosinophilic granulomatosis with polyangiitis (EGPA)
Disease activity

- Serum ANCA monitoring is advisable in patients with MPO-ANCA positivity at disease onset
    - Persistence, rise or reappearance of ANCA might justify more frequent clinical assessment

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Eosinophilic granulomatosis with polyangiitis (EGPA)
Fallow up recommendation

- Routine monitoring of EGPA-related manifestations, particularly:
  - Lung function
  - Cardiovascular events
  - Neurological complications

- Long-term monitoring of comorbidities
  - (such as cancer, infections and osteoporosis)

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Eosinophilic granulomatosis with polyangiitis (EGPA)
Infection prophylaxis

- An increased risk of infections due to immunosuppressive therapy
- In all patients treated with cyclophosphamide and/or rituximab
  - Screening for major chronic infections (such as HBV and HIV) before initiating treatment
  - Prophylaxis against *Pneumocystis jirovecii* infection
    - Sulfamethoxazole–trimethoprim (800 mg–160 mg on alternate days or 400 mg–80 mg daily)
  - A negative effect of therapy on the humoral vaccine response and can lead to clinically relevant secondary hypogammaglobulinaemia
    - Timely vaccination according to current recommendations
    - Passive immunization if necessary
    - Monitoring of quantitative IgG serum concentrations

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Eosinophilic granulomatosis with polyangiitis (EGPA)
Cancer screening

- Age-appropriate cancer screening in all patients
- Regularly screening of cyclophosphamide- treated patients for:
  - Bladder cancer (for example, urine cytology examination)
  - Myeloid leukaemia (evaluation of peripheral blood cell counts and/or haematological examination)
  - Skin cancer (dermatological surveillance)

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Eosinophilic granulomatosis with polyangiitis (EGPA)
Osteoporosis

- Periodic assessment of bone density in all patients with EGPA
  - Especially those with a high cumulative glucocorticoid dose
  - In those with concomitant traditional risk factors for osteoporosis

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Eosinophilic granulomatosis with polyangiitis (EGPA)
Prognosis and long-term outcomes

- 96% overall 5-year survival rate in non-severe EGPA

- Vasculitis relapses
  - Over 40% of patients
  - The first two years post-diagnosis
  - Patients with anti-MPO antibodies and baseline eosinophils <3 × 109/l

- Other persistent symptoms (asthma and rhinosinusitis) occurred throughout ongoing follow-up
- Age ≥ 65 years, the only factor associated with a higher risk of death

- A larger American cohort with 354 patients and median follow-up of 7 years
  - Mortality of 4.0%
  - 12.6% patients were off all treatment
  - A prolonged clinical course and repeated relapses

White J, Dubey S. Eosinophilic granulomatosis with polyangiitis: A review. Autoimmun Rev. 2023 Jan;22(1):103219.


Thanks for your attention