Review paper

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Eosinophilic granulomatosis with polyangiitis across the eosinophilic spectrum: from molecular mechanisms to practical differential diagnosis and targeted therapy

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Abstract

Introduction: Eosinophilic granulomatosis with polyangiitis (EGPA) is a rare anti-neutrophil cytoplasmic antibody (ANCA)-associated vasculitis defined by asthma, hypereosinophilia, and multiorgan involvement. Differentiating EGPA from other eosinophilic disorders is crucial because management differs substantially. The aim of the study is to summarize the pathogenesis, epidemiology, genetics, clinical manifestation, and treatment of EGPA and to provide a comparative differential diagnosis of eosinophilic disorders.

Material and methods: Narrative review using the 2022 American College of Rheumatology (ACR)/European Alliance of Associations for Rheumatology (EULAR) classification criteria, 2024 EULAR recommendations, pivotal randomized trials, and major consensus statements; search strategy and selection criteria are detailed in the Introduction.

Results: Eosinophilic granulomatosis with polyangiitis comprises 2 immunologic endotypes – anti-neutrophil cytoplasmic antibody (ANCA)-positive and ANCA-negative – with distinct organ tropism and therapeutic implications. The interleukin-5 (IL-5)–eosinophil axis is central, supporting anti-IL-5/IL-5R biologics in relapsing or refractory disease. A structured differential first excludes secondary hypereosinophilia (parasites, drugs, malignancies) and then addresses pulmonary "mimics".

Conclusions: An algorithm combining exclusion of secondary causes with organ and endotype profiling enables targeted therapy and reduced glucocorticoid exposure.

Key words: allergic bronchopulmonary aspergillosis, eosinophilia, EGPA, anti-IL-5 therapy.

Introduction

Eosinophilic granulomatosis with polyangiitis (EGPA), formerly Churg-Strauss syndrome, sits at the intersection of anti-neutrophil cytoplasmic antibody (ANCA)-associated vasculitides and eosinophilic disorders. The 2022 American College of Rheumatology (ACR)/European Alliance of Associations for Rheumatology (EULAR) classification criteria provide a standardized research definition through a weighted scoring system. A total score of ≥ 6 points is required for classification, which yields high specificity for distinguishing EGPA from other vasculitides and eosinophilic disorders [1]. It is critical to note that these criteria are intended for research classification and are not a substitute for clinical diagnosis [1].

The criteria and their associated points are as follows:

- weighted criteria items:
  - maximum eosinophil count ≥ 1.0×10^9/l: +5 points,
  - obstructive airway disease: +3 points,
  - nasal polyps: +3 points,
  - cytoplasmic antineutrophil cytoplasmic antibody (ANCA) or anti-proteinase 3-ANCA positivity: –3 points,
  - extravascular eosinophilic predominant inflammation: +2 points,
  - mononeuritis multiplex/motor neuropathy not due to radiculopathy: +1 point,
  - hematuria: –1 point.

The 2024 EULAR update builds upon this framework by emphasizing early control of vasculitic activity and

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deliberate glucocorticosteroid (GC) minimization via steroid-sparing strategies [2].

The objectives are: to present contemporary concepts of pathogenesis, epidemiology (including mortality), genetics, clinical phenotypes, and management of EGPA; and to construct a practical, comparative differential diagnosis against principal eosinophilic conditions – hypereosinophilic syndrome (HES), including myeloid/lymphoid neoplasms with eosinophilia (M/LN-eo), acute eosinophilic pneumonia (AEP)/chronic eosinophilic pneumonia (CEP), allergic bronchopulmonary aspergillosis (ABPA), drug reaction with eosinophilia and systemic symptoms (DRESS)/drug-induced hypersensitivity syndrome (DIHS), eosinophilic gastrointestinal diseases (EGID), parasitic infections, and hematologic malignancies with eosinophilia.

Search methodology and study selection

In line with guidance on transparent searching and reporting for narrative reviews, a multi-database search was conducted in MEDLINE/PubMed, Embase, Scopus, Web of Science, Cochrane Central Register of Controlled Trials (CENTRAL), and Directory of Open Access Journals (DOAJ). Date range: 1 January 2000 – 28 September 2025 (with inclusion of seminal earlier works critical to disease definitions). Languages: English and Polish. Controlled vocabulary (MeSH/Emtree) and free-text terms included: "eosinophilic granulomatosis with polyangiitis", "Churg-Strauss", "ANCA-associated vasculitis", "hypereosinophilia", "hypereosinophilic syndromes", "FIP1L1-PDGFRA", "eosinophilic pneumonia" (AEP/CEP), "allergic bronchopulmonary aspergillosis", "DRESS/DIHS", "*Strongyloides*", "eosinophilic esophagitis", "EGID", "mepolizumab", "benralizumab", "rituximab", "cyclophosphamide", "EULAR", and "ACR/EULAR criteria". Primary evidence (randomized controlled trials, cohort/case-control studies, registries), guidelines/consensus statements, and classification criteria were prioritized; secondary reviews were used for contextualization only. Manual snowballing of reference lists and consultation of editorial/ethics resources (International Committee of Medical Journal Editors, Council of Science Editors) and research assessment principles (San Francisco Declaration on Research Assessment – DORA) supported reporting integrity [3–5]. While this is a narrative review, transparency elements akin to PRISMA (Preferred Reporting Items for Systematic Reviews and Meta-Analyses) are provided (databases, dates, keywords, inclusion approach). For a full systematic review, PROSPERO (International Prospective Register of Systematic Reviews) registration and PRISMA 2020 reporting are recommended [6, 7].

Pathogenesis

Eosinophilic granulomatosis with polyangiitis development is influenced by genetic predisposition (e.g., association with human leukocyte antigen [HLA]-DQ) and environmental triggers such as allergens, infections, and drugs. It is fundamentally driven by a dysregulated type 2 (T2) inflammatory response. This involves overexpression of key cytokines – interleukin-5 (IL-5), IL-4, IL-13 – and eotaxins (CC chemokine ligands: CCL11, CCL24, CCL26). Interleukin-5 is the master regulator of eosinophil biology, promoting their production, activation, and survival. Interleukin-4 and IL-13 drive B-cell activation and immunoglobulin E (IgE) production, and contribute to tissue fibrosis. Eotaxins recruit eosinophils to tissues [8]. Airway epithelial cells release "alarmins" such as thymic stromal lymphopoietin (TSLP), IL-25, and IL-33, which activate type 2 innate lymphoid cells and dendritic cells, priming a robust T-helper 2 (Th2) lymphocyte response.

This dysregulated T2 response manifests in 2 distinct pathogenetic arms, which correlate with ANCA status and clinical phenotype.

One of them is the eosinophil-driven arm (ANCA-negative endotype), in which the dominant pathway is IL-5-mediated activation and survival of eosinophils. Activated eosinophils infiltrate tissues and release their cytotoxic granule contents – major basic protein (MBP), eosinophil cationic protein (ECP), and eosinophil-derived neurotoxin (EDN) – causing direct tissue injury. They also generate eosinophil extracellular traps (EETs), fostering a prothrombotic and pro-fibrotic milieu. Clinically, this arm is more typical of ANCA-negative EGPA and is characterized by eosinophilic tissue infiltration. This leads to cardiac involvement (eosinophilic myocarditis, Loeffler-like endocarditis), pulmonary infiltrates, and gastrointestinal manifestations. Renal involvement, if present, is more likely to be a non-non-pauci-immune lesion such as membranous nephropathy or interstitial nephritis [2].

The other inflammatory response is the ANCA-mediated vasculitic arm (ANCA-positive endotype). In approximately 30–40% of patients, B-cells produce ANCA autoantibodies, predominantly against MPO. These MPO-ANCAs bind to neutrophils, causing their activation, a respiratory burst, and release of proteolytic enzymes and ROS. This results in necrotizing vasculitis of small-to-medium vessels and granuloma formation. The clinical phenotype is the ANCA-positive EGPA patient, who presents with more classic vasculitic features such as glomerulonephritis, mononeuritis multiplex, purpura, and systemic vasculitis [9].

In both situations, eosinophils are the central effectors of tissue damage. Their cytotoxic degranulation and EET formation are responsible for the endothelial injury,

fibrosis, and microthrombotic complications that characterize the disease. This central role provides the strong mechanistic rationale for targeting the IL-5/IL-5R axis. Clinical reports suggest that interventions modulating the T2 axis (e.g., leukotriene modifiers, dupilumab) can unmask EGPA in predisposed patients with severe asthma/chronic rhinosinusitis with nasal polyps (CRSwNP); causality remains debated and requires vigilance [10].

Epidemiology and prognosis

Eosinophilic granulomatosis with polyangiitis is rare (several cases per million per year), typically presenting in the 4^{th}–6^{th} decades and slightly more often in women [9]. Cardiac and renal involvement are the strongest prognostic drivers. The Five-Factor Score (FFS), 2009 revision, stratifies mortality risk and informs treatment intensity (0, 1, ≥ 2 points align with distinct survival curves) [11]. The FFS retains prognostic value across ANCA-associated vasculitis (AAV) registries independent of age [9].

Genetics and biomarkers

Unlike M/LN-eo – where kinase rearrangements (e.g., fusion gene involving FIP1 like 1 and platelet-derived growth factor receptor α [FIP1L1::PDGFRA], platelet-derived growth factor receptor β [PDGFRB], fibroblast growth factor receptor 1 [FGFR1], fusion gene involving pericentriolar material 1 and Janus kinase 2 [PCM1::JAK2]) drive proliferation and confer dramatic sensitivity to imatinib – EGPA lacks a single genetic "driver" [12–15]. Clinically, persistent hypereosinophilia with myeloproliferative signals (splenomegaly, elevated tryptase) mandates evaluation for clonality (flow cytometry and next-generation sequencing [NGS]), as diagnosing M/LN-eo fundamentally redirects therapy [13–15]. Biomarkers of EGPA activity include peripheral eosinophil count, total IgE, and organ damage markers (troponins, *N*-terminal pro-B-type natriuretic peptide). A T2-high profile favors response to anti-IL-5/IL-5R agents [9].

The role of eosinophils

Eosinophilic granulomatosis with polyangiitis can be conceptualized as the convergence of genetic predisposition and environmental exposures that channel immune activation into two dominant inflammatory tracks. On the eosinophilic arm – more typical of ANCA-negative EGPA – Th2 immunity and IL-5 drive eosinophil maturation and activation. Activated eosinophils degranulate (MBP, ECP, EDN) and generate ROS, producing tissue injury and remodeling; B-cell skewing may associate with IgG4 responses. Renal involvement on this arm more often reflects non–pauci-immune lesions (e.g., membranous nephropathy, interstitial nephritis) rather than necrotizing glomerulonephritis (GN). By contrast, the ANCA-mediated arm – more typical of ANCA-positive EGPA – features B-cell production of ANCA (predominantly MPO), neutrophil activation, and release of proteolytic enzymes and ROS, culminating in necrotizing vasculitis, pauci-immune necrotizing GN, and granuloma formation. Observational data implicate distinct genetic cues (e.g., non-HLA signals such as cytokine/chemokine axes for the eosinophilic arm vs. HLA-DQ signals for the ANCA-mediated arm) and environmental triggers (silica, organic solvents, farming) as potential modulators of arm dominance. Eosinophils are the central effectors of tissue damage via cytotoxic degranulation and EET formation, fostering thrombosis and fibrosis. This pathophysiology justifies targeting the IL-5/IL-5R axis (mepolizumab, benralizumab) and exploring emerging pathways (TSLP, IL-33, sialic acid-binding immunoglobulin-like lectin 8 [Siglec-8]) [9].

Clinical picture and criteria

The canonical triad is asthma (often severe), eosinophilia, and systemic vasculitis. Lungs show migratory infiltrates and asthma exacerbations; sinuses – CRSwNP; nerves – mononeuritis multiplex; gastrointestinal tract – pain, bleeding, or ischemia. Cardiac involvement (myocarditis, pericarditis, mural thrombosis; Loeffler-like phenotype) is a major determinant of survival [9]. Histology demonstrates eosinophilic granulomas and leukocytoclastic vasculitis; serology often reveals MPO-ANCA [1, 2].

Eosinophilic granulomatosis with polyangiitis within the eosinophilic landscape – comparative differential

Differential diagnosis begins with the fundamental question of why eosinophilia is present. Secondary causes – parasites, drugs, and hematologic neoplasms – should be excluded first. *Strongyloides stercoralis* deserves special attention because autoinfection permits explosive hyperinfection under GC exposure; patients with persistent eosinophilia and epidemiologic risk should undergo screening and/or empiric ivermectin before immunosuppression [16]. Identifying a parasitic etiology generally rules out primary EGPA and redirects management to pathogen eradication.

A second pillar involves HES. In the myeloid/lymphoid neoplasm spectrum (M/LN-eo), kinase fusions – most famously FIP1L1::PDGFRA – explain dramatic, sometimes immediate responses to imatinib [12]. Splenomegaly, elevated tryptase, endomyocardial fibrosis, and mural thrombi are not uncommon. In contrast to

EGPA, leukocytoclastic vasculitis and ANCA are absent; organ damage arises from eosinophilic infiltration and microthrombotic injury [13–15]. Practically, persistent hypereosinophilia with "myeloid signals" necessitates clonality testing first, as diagnosing M/LN-eo completely changes the trajectory and prognosis.

Within pulmonary disorders, AEP and CEP frequently mimic EGPA. The AEP presents abruptly with fever, severe hypoxemia, and an acute respiratory distress syndrome-like picture; bronchoalveolar lavage (BAL) shows marked eosinophilia, and GC responses are rapid and complete, without the multiorgan vasculitis typical of EGPA [17, 18]. The CEP evolves subacutely with cough, dyspnea, night sweats, weight loss, and characteristic peripheral consolidations on high-resolution computed tomography (HRCT; "photographic negative of pulmonary edema"). Both share eosinophilia and pulmonary infiltrates with EGPA; yet systemic vasculitis is absent (no neuropathies, GN, or classic cutaneous vasculitis) and ANCA tests are typically negative. Glucocorticosteroid monotherapy is usually effective, though CEP tends to relapse [17–19]. Nerve conduction studies, renal/cardiac evaluation, and, where feasible, targeted biopsy resolve ambiguity.

Allergic bronchopulmonary aspergillosis is a classic mimic in asthma. Elevated total IgE, IgE/IgG specific to *Aspergillus*, eosinophilia, and central bronchiectasis with mucus plugging on HRCT define the phenotype. The International Society for Human and Animal Mycology (ISHAM) 2024 revision refined diagnostic categories (including "ABPA with mucus plugging") and requires proven fungal sensitization with total IgE ≥ 500 IU/ml. Unlike EGPA, ABPA lacks systemic vasculitis and ANCA; treatment comprises inhaled/systemic GCs, azoles, and – selectively – biologics used in T2-high asthma/CRSwNP [20, 21].

In the drug-induced spectrum, DRESS/DIHS is a severe, potentially fatal hypersensitivity reaction, often triggered by allopurinol, aromatic antiepileptics, or sulfonamides. It features generalized rash, fever, lymphadenopathy, eosinophilia, and organ involvement (especially liver). European Registry of Severe Cutaneous Adverse Reactions (RegiSCAR) scales provide standardized case adjudication [22]. Though multiorgan involvement and eosinophilia may resemble EGPA, ANCA-mediated vasculitis is absent; a meticulous drug history, temporal dynamics, and improvement after drug withdrawal with GCs (and intravenous immunoglobulin [IVIG]/cyclosporine in severe cases) are decisive [22, 23].

Eosinophilic gastrointestinal diseases – notably EoE – are restricted to the gastrointestinal wall and histologically lack leukocytoclastic vasculitis. Atopy, asthma, and peripheral eosinophilia may coexist, but the mechanism is mucosal-allergic rather than vasculitic. Management (PPI, topical GCs, elimination diets, selected biologics) differs fundamentally from EGPA [24]. At the cardio-eosinophilic interface, eosinophilic myocarditis and Loeffler's endocarditis (restrictive cardiomyopathy with endocardial fibrosis and mural thrombosis) more often accompany HES/parasites than EGPA; systemic vasculitis is absent, and treatment is cause-directed with adjunctive immunosuppression and anti-IL-5 where appropriate [13, 14, 18]. Finally, hematologic malignancies with eosinophilia (e.g., T-cell lymphomas, chronic eosinophilic leukemia, not otherwise specified (CEL-NOS), mastocytosis) are diagnosed by clonality according to the World Health Organization/International Consensus Classification 2022–2023 and treated oncologically, effectively excluding primary EGPA [14, 15].

Management of eosinophilic granulomatosis with polyangiitis – differential diagnosis and treatment

The management of EGPA is based on a two-step process: first, the rigorous exclusion of alternative causes of eosinophilia, and second, treatment tailored to the disease severity and immunologic endotype.

Essential investigations to exclude eosinophilic granulomatosis with polyangiitis mimics

Before initiating immunosuppression, a comprehensive workup is mandatory to rule out conditions that can present with overlapping features but require fundamentally different management.

Eosinophilia detected in the blood tests prompts evaluation to rule out secondary and clonal eosinophilia by assessing the serological status and hematological deviations. Infectious investigation includes serology and/or stool microscopy for *Strongyloides stercoralis*. In high-risk patients, empiric ivermectin may be considered prior to immunosuppression to prevent hyperinfection, a potentially fatal complication [16, 25]. The hematologic diagnosis should focus on peripheral blood flow cytometry to detect abnormal T-cell clones suggestive of L-HES or T-cell lymphoma [15, 26]. Molecular genetic testing (fluorescence in situ hybridization/NGS) is used to detect classic kinase fusions, especially FIP1L1::PDGFRA, as well as rearrangements involving PDGFRB, FGFR1, and JAK2 [13, 14]. This is critical, as its identification confers dramatic sensitivity to imatinib [12]. Also, an elevated serum tryptase is a sensitive marker for myeloid neoplasms with eosinophilia and should prompt further clonal investigation [13, 27].

Secondly, the radiological and organ-based assessments are vital. Adequate choice of the radiological tool can help to rule out possible diagnoses. The examinations which are of the highest value are HRCT of the chest and echocardiography or cardiac magnetic resonance (CMR). High-resolution computed tomography is indicated in nearly all cases; it helps identify features of EGPA (e.g., transient pulmonary opacities, bronchial wall thickening) [9] and distinguish it from CEP, which shows characteristic peripheral consolidations ("photographic negative of pulmonary edema") [17, 18], and ABPA, which shows central bronchiectasis and mucus plugging [21, 28]. Echocardiography and CMR assess for eosinophilic myocarditis, endomyocardial fibrosis, or mural thrombi. While cardiac involvement can occur in EGPA, a dominant restrictive cardiomyopathy is more characteristic of HES [9, 29]. Cardiac magnetic resonance with late gadolinium enhancement is particularly sensitive for detecting eosinophilic myocardial involvement [30].

Third, the histopathological examination remains a cornerstone of the differential diagnosis. The finding of necrotizing vasculitis with extravascular eosinophilic granulomas in skin, nerve, or lung biopsies is highly characteristic of EGPA [1, 9]. Frequent renal manifestations also necessitate biopsy. It helps to distinguish EGPA's pauci-immune necrotizing glomerulonephritis (common in ANCA-positive cases) from other eosinophilic infiltrates [9, 31]. In the gastrointestinal tract, biopsy helps rule out EGID, which show eosinophil-predominant inflammation confined to the mucosal layer without vasculitis [24].

Treatment of eosinophilic granulomatosis with polyangiitis

Once the diagnosis is established, treatment is guided by disease severity and immunologic endotype, in accordance with 2024 EULAR recommendations [2]. For patients with organ- or life-threatening disease, such as rapidly progressive glomerulonephritis, severe cardiomyopathy, or alveolar hemorrhage, the recommended induction therapy consists of high-dose GCs combined with either cyclophosphamide or rituximab [2]. In cases of relapsing or refractory EGPA, anti-IL-5 agents such as mepolizumab (300 mg s.c. every 4 weeks) or benralizumab (30 mg s.c. every 4 weeks) are approved for remission induction and enable sustained GC-sparing [32, 33]. The therapeutic strategy is further refined by the immunologic endotype: rituximab is particularly effective in ANCA-positive, vasculitic-dominant disease, mirroring its use in other AAVs [2, 20], whereas anti-IL-5/IL-5R therapy forms the main treatment for ANCA-negative, eosinophil-dominant disease [16, 17]. Regarding adjunctive therapy, the C5a receptor inhibitor avacopan has an established role in GPA and MPA, but definitive evidence for its use in EGPA is still emerging and remains under investigation [2, 34].

Future directions

Personalized medicine in EGPA will be refined by a deeper understanding of endotypes (ANCA+/ANCA–, T2-high markers), early recognition of cardio-dominant phenotypes, and the development of novel biologic therapies targeting upstream pathways such as TSLP, IL-33, and Siglec-8 [9, 10]. Real-world registries and predictive biomarkers for anti-IL-5/IL-5R response are priorities for further GC-sparing and outcome improvement [2, 9].

Conclusions

In synthesis, EGPA is distinguished by systemic vasculitis (neuropathy, GN, vasculitic cardiac involvement), frequent MPO-ANCA, and histology showing eosinophilic vasculitis/granulomas; mimics are typically organ-restricted (AEP/CEP, EGID), allergic/colonization-driven (ABPA), drug-induced (DRESS), or clonal (HES-M/LN-eo), with largely etiology-specific management [2, 8, 9, 12–14, 17, 22–24].

Eosinophilic granulomatosis with polyangiitis blends vasculitic and eosinophilic biology. An effective clinical strategy requires: uncompromising exclusion of secondary eosinophilia, meticulous organ and endotype profiling, targeted therapy (anti-IL-5/IL-5R, B-cell-directed or cyclophosphamide) with maximal GC sparing, and priority surveillance and protection of the heart and peripheral nerves.

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References

1. Grayson PC, Ponte C, Suppiah R, Robson JC, Craven A, Judge A, et al. 2022 American College of Rheumatology/European Alliance of Associations for Rheumatology classification criteria for eosinophilic granulomatosis with polyangiitis. Arthritis Rheumatol 2022; 74: 386–392, DOI: 10.1002/art.41982.
2. Hellmich B, Sanchez-Alamo B, Schirmer JH, Berti A, Blockmans D, Cid MC, et al. EULAR recommendations for the management of ANCA-associated vasculitis: 2024 update. Ann Rheum Dis 2024; 83: 30–47, DOI: 10.1136/ard-2022-223764.

3. San Francisco Declaration on Research Assessment (DORA). Available from: https://sfdora.org/; https://sfdora.org/ (Access: 28.09.2025).
4. International Committee of Medical Journal Editors (ICMJE). Recommendations for the conduct, reporting, editing, and publication of scholarly work in medical journals. Available from: http://www.icmje.org/; http://www.icmje.org/ (Access: 28.09.2025).
5. Council of Science Editors (CSE). White paper on promoting integrity in scientific journal publications. Available from: https://www.csescienceeditor.org/; https://www.csescienceeditor.org/ (Access: 28.09.2025).
6. Page MJ, McKenzie JE, Bossuyt PM, Boutron I, Hoffmann TC, Mulrow CD, et al. The PRISMA 2020 statement: an updated guideline for reporting systematic reviews. BMJ 2021; 372: n71, DOI: 10.1136/bmj.n71.
7. PROSPERO – International Prospective Register of Systematic Reviews. Available from: https://www.crd.york.ac.uk/prospero/; https://www.crd.york.ac.uk/prospero/ (Access: 28.09.2025).
8. Kita H. Eosinophils: multifaceted biological properties and roles in health and disease. Immunol Rev 2011; 242: 161–177, DOI: 10.1111/j.1600-065X.2011.01026.x.
9. Emmi G, Silvestri E, Becatti M, Damiani A, Lavorini F, Parronchi P, et al. Eosinophilic granulomatosis with polyangiitis: from pathogenesis to practice. Nat Rev Rheumatol 2023; 19: 378–393, DOI: 10.1038/s41584-023-00958-w.
10. Suzaki I, Kimura M, Araya J, Tamaoka M, Yagi K, Kumasaka F, et al. Eosinophilic granulomatosis with polyangiitis developing during dupilumab treatment for severe asthma: a case report. BMC Pulm Med 2023; 23: 504.
11. Guillevin L, Pagnoux C, Seror R, Mahr A, Mouthon L, Le Toumelin P. The Five-Factor Score revisited: assessment of prognoses of systemic necrotizing vasculitides based on the French Vasculitis Study Group cohort. Medicine (Baltimore) 2011; 90: 19–27, DOI: 10.1097/MD.0b013e318205a4c6.
12. Cools J, DeAngelo DJ, Gotlib J, Stover EH, Legare RD, Cortes J, et al. A tyrosine kinase created by fusion of the PDGFRA and FIP1L1 genes as a therapeutic target of imatinib in idiopathic hypereosinophilic syndrome. N Engl J Med 2003; 348: 1201–1214, DOI: 10.1056/NEJMoa025217.
13. Khoury JD, Solary E, Abla O, Akkari Y, Alaggio R, Apperley JF, et al. The 5th edition of the World Health Organization classification of haematolymphoid tumours: myeloid and histiocytic/dendritic neoplasms. Leukemia 2022; 36: 1703–1719, DOI: 10.1038/s41375-022-01613-1.
14. Arber DA, Orazi A, Hasserjian RP, Borowitz MJ, Calvo KR, Kvasnicka HM, et al. International consensus classification of myeloid neoplasms and acute leukemias: integrating morphologic, clinical, and genomic data. Blood 2022; 140: 1200–1228, DOI: 10.1182/blood.2022015850.
15. Roufosse F, Weller PF. Practical approach to the patient with hypereosinophilia. J Allergy Clin Immunol 2010; 126: 39–44, DOI: 10.1016/j.jaci.2010.04.011.
16. Centers for Disease Control and Prevention (CDC). Parasites – Strongyloides: resources for health professionals. Available from: https://www.cdc.gov/parasites/strongyloides/health_professionals/; https://www.cdc.gov/parasites/strongyloides/health_professionals/ (Access: 28.09.2025).
17. Allen JN, Pacht ER, Gadek JE, Davis WB. Acute eosinophilic pneumonia as a reversible cause of noninfectious respiratory failure. N Engl J Med 1989; 321: 569–574, DOI: 10.1056/NEJM198908313210903.
18. Cottin V, Cordier JF. Eosinophilic lung diseases. Immunol Allergy Clin North Am 2012; 32: 557–586, DOI: 10.1016/j.iac.2012.08.007.
19. Dellon ES, Hirano I. Epidemiology and natural history of eosinophilic esophagitis. Gastroenterology 2018; 154: 319–332.e3, DOI: 10.1053/j.gastro.2017.06.067.
20. Patterson K, Strek ME. Allergic bronchopulmonary aspergillosis. Proc Am Thorac Soc 2010; 7: 237–244, DOI: 10.1513/pats.200908-086AL.
21. Agarwal R, Muthu V, Sehgal IS, Prasad KT, Dhooria S, Garg M, et al. International Society for Human and Animal Mycology–AAFP workgroup recommendations on the clinical utility of Aspergillus fumigatus-specific IgG and IgM serology in allergic bronchopulmonary aspergillosis. Eur Respir J 2024; 63: 2400061, DOI: 10.1183/13993003.00061-2024.
22. Cacoub P, Musette P, Descamps V, Meyer O, Speirs C, Finzi L, et al. The DRESS syndrome: a literature review. Am J Med 2011; 124: 588–597, DOI: 10.1016/j.amjmed.2011.01.017.
23. Chen YC, Chiu HC, Chu CY. Drug reaction with eosinophilia and systemic symptoms: a retrospective study of 60 cases. Arch Dermatol 2010; 146: 1373–1379, DOI: 10.1001/archdermatol.2010.198.
24. Dellon ES, Liacouras CA, Molina-Infante J, Furuta GT, Spergel JM, Zerbib F, et al. Updated international consensus diagnostic criteria for eosinophilic esophagitis: proceedings of the AGREE conference. Gastroenterology 2018; 155: 1022–1033.e10, DOI: 10.1053/j.gastro.2018.07.009.
25. Greaves D, Coggle S, Pollard C, Aliyu SH, Moore EM. Strongyloides stercoralis infection. BMJ 2013; 347: f4610, DOI: 10.1136/bmj.f4610.
26. Ogbogu PU, Bochner BS, Butterfield JH, Gleich GJ, Huss-Marp J, Kahn JE, et al. Hypereosinophilic syndrome: a multicenter, retrospective analysis of clinical characteristics and response to therapy. J Allergy Clin Immunol 2009; 124: 1319–1325.e3, DOI: 10.1016/j.jaci.2009.09.022.
27. Klion AD. How I treat hypereosinophilic syndromes. Blood 2015; 126: 1069–1077, DOI: 10.1182/blood-2014-11-551614.
28. Agarwal R, Sehgal IS, Dhooria S, Aggarwal AN. Developments in the diagnosis and treatment of allergic bronchopulmonary aspergillosis. Expert Rev Respir Med 2016; 10: 1317–1334, DOI: 10.1080/17476348.2016.1249853.
29. Ogbogu PU, Rosing DR, Horne MK 3rd. Cardiovascular manifestations of hypereosinophilic syndromes. Immunol Allergy Clin North Am 2007; 27: 457–475, DOI: 10.1016/j.iac.2007.07.001.
30. Mavrogeni S, Bratis K, Koutsogeorgopoulou L, Karabela G, Stavropoulos E, Katsalira A, et al. Myocarditis and hypereosinophilic syndrome: a comprehensive review. Heart Fail Rev 2024; 29: 595–607.
31. Sinico RA, Di Toma L, Maggiore U, Bottero P, Sabadini E, Pagani C, et al. Renal involvement in Churg–Strauss syndrome. Am J Kidney Dis 2006; 47: 770–779, DOI: 10.1053/j.ajkd.2006.01.026.
32. Wechsler ME, Akuthota P, Jayne D, Khoury P, Klion A, Langford CA, et al. Mepolizumab or placebo for eosinophilic granu-

lomatosis with polyangiitis. N Engl J Med 2017; 376: 1921–1932, DOI: 10.1056/NEJMoa1702079.
33. Wechsler ME, Nair P, Terrier B, Khoury P, Cid MC, Karrasch J, et al. Benralizumab versus mepolizumab for eosinophilic granulomatosis with polyangiitis. N Engl J Med 2024; 390: 911–921, DOI: 10.1056/NEJMoa2311155.
34. Betz S, Merkel F, Prati C, Goupil R, Henes J, Doubelt I, et al. The C5a receptor inhibitor avacopan in the treatment of eosinophilic granulomatosis with polyangiitis (EGPA): a multicenter, randomized, double-blind, placebo-controlled trial [abstract]. Arthritis Rheumatol 2023; 75 (Suppl 9).