EOSINOPHILIC GRANULOMATOSIS WITH POLYANGIITIS (EGPA):

Diagnosis & Treatment Approaches



STEP

STEP

STEP

3

LUNG

DISEASES

Characteristics

- Moderate to severe asthma
- Peripheral blood eosinophilia (> 10% eosinophils)
- Pulmonary infiltrates
- Paranasal sinus abnormality
- Mononeuropathy or polyneuropathy
- Extravascular eosinophils/eosinophilic vasculitis of small- to medium-sized blood vessels
- Positive ANCA

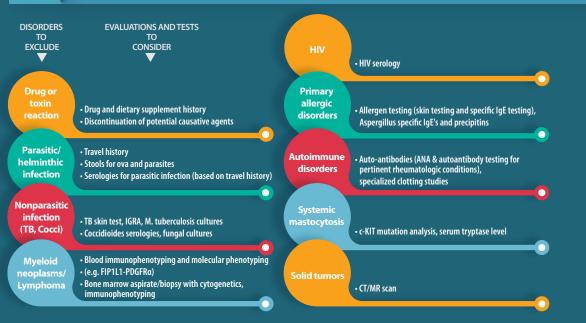


Basic evaluation

DETAILED HISTORY AND PHYSICAL EXAM

- ASK ABOUT EXPOSURES: Medications, dietary supplements, recreational drugs, toxins, workplace, travel
- SEARCH FOR EXTRAPULMONARY DISEASE: Nasal/sinus, ocular, skin, cardiac, gastrointestinal, neurologic, renal, vascular
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- Tailor according to history and physical:
- IMAGING/TESTING: CXR, chest CT scan, sinus CT scan, EMG/NCV, echocardiogram or cardiac MRI
- LABORATORY: CBC with differential, ESR, CRP, ANCA(MPO/PR3), vitamin B12, electrolytes, LFTs, total protein, urinalysis
 Serum tryptase level, serum Ig's (include IgE), Troponin
 Bone marrow aspirate and biopsy with reticulin and tryptase stains

Exclude major secondary causes of pulmonary eosinophilia



Exclude other diseases associated with eosinophilia

SYSTEMIC

DISEASES

- Asthma/eosinophilic bronchitis
 Allergic bronchopulmonary aspergillosis
- Bronchocentric granulomatosis
- Cryptogenic organizing pneumonia
- Hypersensitivity pneumonitis
- Idiopathic pulmonary fibrosis
- Pulmonary Langerhans cell histiocytosis
- Postradiation pneumonitis
- Chronic eosinophilic pneumonia

This reference aid was supported by an educational grant from GlaxoSmithKline.

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Rheumatoid arthritis

Sjögren syndrome

• Hypereosinophilic Syndrome

Sarcoidosis

Distinguish between the primary pulmonary eosinophilic disorders

	ACUTE EOSINOPHILIC PNEUMONIA	CHRONIC EOSINOPHILIC PNEUMONIA	HYPEREOSINOPHILIC SYNDROME	EOSINOPHILIC GRANULOMATOSIS WITH POLYANGIITIS ¹	
KEY FEATURES'	Acute febrile illness; hypoxemic respiratory failure; diffuse pulmonary infiltrates; BAL fluid eosinophils > 25%; previously healthy person	Subacute illness; eosinophilia in lung tissue and blood; bilateral pulmonary infiltrates may be peripheral in location	Sustained overproduction of eosinophils; marked peripheral blood eosinophilia; tissue eosinophilia; end- organ damage; includes myeloproliferative, lymphocytic, familial, and idiopathic variants	Necrotizing vasculitis of small- to medium- sized blood vessels; asthma; eosinophil-rich granulomatous inflammation of respiratory tract; involves multiple organs	
Onset	Acute (days)	Indolent (weeks/months)	Indolent (months/years)	Indolent (months/years)	
Pulmonary infiltrates	Diffuse	Peripheral	Patchy	Patchy	
Fulminant respiratory failure	Frequently Occurs	Rarely Occurs	Rarely Occurs	Rarely Occurs	
Asthma/allergy history	Rarely Occurs	Commonly Occurs	Rarely Occurs	Frequently Occurs	
Smoking history	Commonly Occurs	Rarely Occurs	Rarely Occurs	Rarely Occurs	
Vasculitis	Rarely Occurs	Rarely Occurs	Rarely Occurs	Frequently Occurs	
ANCA	Rarely Occurs	Rarely Occurs	Rarely Occurs	Commonly Occurs	
Cardiac involvement	Rarely Occurs	Occasionally Occurs	Frequently Occurs	Commonly Occurs	
Neurologic involvement	Rarely Occurs	Occasionally Occurs	Frequently Occurs	Frequently Occurs	
Requirement for therapies other than corticosteroids	Rarely Occurs	Rarely Occurs	Frequently Occurs	Commonly Occurs	

STEP 5

Comparison of ANCA-associated vasculitides

Small to medium vessel necrotizing vasculitides with differentiating features

GPA	EGPA
Almost Always ANCA positive	Sometir
C-ANCA and PR3 specificity	p-ANCA
Unlikely to exhibit asthma	Asthma
Unlikely to exhibit blood eosinophilia	Always
Glomerulonenhritis common	Glomer

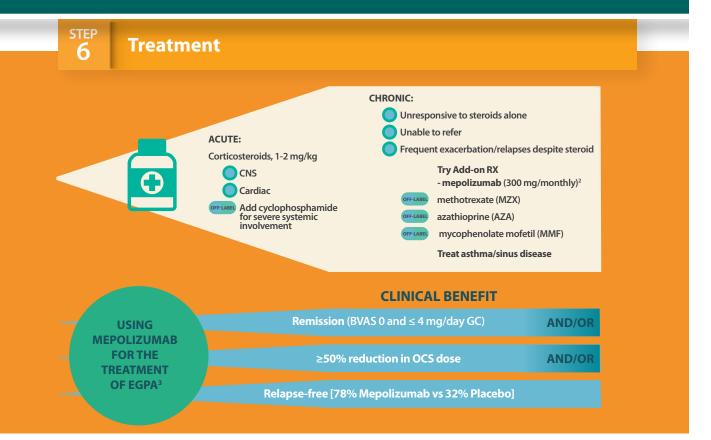
Sometimes ANCA positive p-ANCA and MPO specificity Asthma common Always exhibits blood eosinop

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Almost always ANCA positive p-ANCA and MPO specificity

MPA

- Unlikely to exhibit blood eosinophilia
- Glomerulonephritis common
- Cioncratonepintas common



• Eosinophilic granulomatosis with polyangiitis (EGPA) was previously called Churg-Strauss syndrome.

BAL, transbronchial or open-lung biopsy may be necessary, depending on clinical and radiographic findings.
Additional testing may be needed to evaluate for extrapulmonary disease.

Abbreviations:

BAL, bronchoalveolar lavage; ESR, erythrocyte sedimentation rate; CRP, C-reactive protein; ANCA, antineutrophil cytoplasmic antibody; Ig, immunoglobulin; LFT, liver function test; TB, tuberculosis; IGRA, interferon gamma release assay; RT-PCR, reverse transcriptase-polymerase chain reaction;

References: 1 Wechsler ME. Pulmonary eosinophilic syndromes. Immunol Allergy Clin N Am. 2007;27:477-492.3 2 Wechsler, ME, et al. Mepolizumab or Placebo for Eosinophilic Granulomatosis with Polyangiitis. N Engl J Med. 2017 May 18;376(20):1921-1932 3 Steinfeld J, et al. Evaluation of clinical benefit from treatment with mepolizumab for patients with eosinophilic granulomatosis with polyangiitis. J Allergy Clin Immunol 2019 Jun;143(6):2170-2177