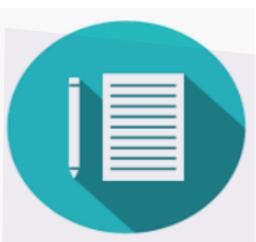
## Diagnosis and Treatment Selection for Eosinophilic Granulomatosis with Polyangiitis: Person-Centered Care in EGPA



## 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





Sinusitis Nasal Polyps

**Asthma** Pulmonary Infiltrates Alveolar Hemorrhage

Glomerulonephritis Renal insufficiency Renal infarct

Glomerulonephritis Renal insufficiency Renal infarct



Neurosensory Hearing Loss Motor neuropathy Sensory neuropathy

> Cardiomyopathy Myocarditis Heart Failure Arrhythmia

Ischemic Bowel Pancreatitis Cholecystitis

Palpable purpura Skin nodules Urticaria Livedo Reticularis



## Differential Diagnosis

	Acute Eosinophilic Pneumonia	Chronic Eosinophilic Pneumonia	Hypereosinophilic Syndrome	Eosinophilic Granulomatosis with Polyangiitis
Key Features 1	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
Frequently Occurs	Fulminant     respiratory failure	Chronic sinusitis	<ul> <li>Cardiac involvement</li> <li>Neurologic involvement</li> </ul>	<ul><li>Asthma/allergy history</li><li>Vasculitis</li><li>Neurologic involvement</li></ul>
Commonly Occurs	Smoking history	Asthma/allergy history	Requirement for therapies other than corticosteroids	<ul> <li>ANCA</li> <li>Cardiac involvement</li> <li>Requirement for therapies other than corticosteroids</li> </ul>

Treatment



Acute

Corticosteroids, I-2 mg/kg

Add cyclophosphamide for severe systemic involvement

- CNS

- Cardiac

OFF-**LABEL** 

Chronic

- Unresponsive to steroids alone

- Unable to taper

- Frequent exacerbation/relapses despite steroid

Try Add-on RX

- Mepolizumab (300 mg/monthly)<sup>2</sup>

- Methotrexate (MZX)

- Azathioprine (AZA)

- Mycophenolate mofetil (MMF)

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.

