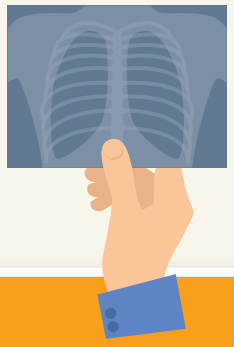
PULMONARY EOSINOPHILIC Syndromes

Diagnostic Approach

Characteristics

- Increased respiratory symptoms with increased eosinophils in peripheral blood, lung tissue, sputum or BAL fluid
- Pulmonary infiltrates
- Possible signs or symptoms of systemic disease



STEP 1 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

Tailor according to history and physical:

- **IMAGING:** CXR, chest CT scan, sinus CT scan, echocardiogram
- **LABORATORY:** CBC with differential, ESR, CRP, ANCA, vitamin B12, electrolytes, LFTs, total protein, urinalysis
  Serum tryptase level, serum Ig's (include IgE)
  Bone marrow biopsy with reticulin and tryptase stains

STEP 2 Exclude major secondary causes of pulmonary eosinophilia

DISORDERS TO EXCLUDE	EVALUATIONS AND TESTS TO CONSIDER
Drug or toxin reaction	• Drug and dietary supplement history • Discontinuation of potential causative agents
Parasitic/ helminthic infection	• Travel history • Stools for ova and parasites • Serologies for parasitic infection (based on travel history)
Nonparasitic infection (TB, Cocci)	• TB skin test, IGRA, M. tuberculosis cultures • Coccidioides serologies, fungal cultures
Leukemia/ lymphoma	• Bone marrow aspirate/biopsy with cytogenetics, immunophenotyping • RT-PCR analysis of PBMCs for BCR-ABL fusion gene • Abdomen/chest CT, gallium scan, PET scan
HIV	• HIV serology
Primary allergic disorders	• Allergen skin testing, Aspergillus precipitins, specific RASTs
Autoimmune disorders	• Auto-antibodies (ANA & ANCA), specialized clotting studies
Systemic mastocytosis	• c-KIT mutation analysis, serum tryptase level
Solid tumors	• CT/MR scan

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STEP 3 Exclude other diseases associated with eosinophilia

LUNG DISEASES

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

SYSTEMIC DISEASES

- Rheumatoid arthritis
- Sarcoidosis
- Sjögren syndrome

STEP 4 Distinguish between the primary pulmonary eosinophilic disorders

	ACUTE ESOINOPHILIC PNEUMONIA	CHRONIC EOSINOPHILIC PNEUMONIA	HYPEREOSINOPHILIC SYNDROME	EOSINOPHILIC GRANULOMATOSIS WITH POLYANGIITIS*
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**

*** 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; **PBMC**, peripheral blood mononuclear cell; **PET**, positron emission tomography; **RAST**, radioallergosorbent test.

Reference: Wechsler ME. Pulmonary eosinophilic syndromes. Immunol Allergy Clin N Am. 2007;27:477-492.