

**2022 AMERICAN COLLEGE OF RHEUMATOLOGY / EUROPEAN ALLIANCE OF ASSOCIATIONS FOR RHEUMATOLOGY**  
**CLASSIFICATION CRITERIA FOR GRANULOMATOSIS WITH POLYANGIITIS**

**CONSIDERATIONS WHEN APPLYING THESE CRITERIA**

- These classification criteria should be applied to classify a patient as having 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**

Nasal involvement: bloody discharge, ulcers, crusting, congestion, blockage, or septal defect / perforation	<b>+3</b>
Cartilaginous involvement (inflammation of ear or nose cartilage, hoarse voice or stridor, endobronchial involvement, or saddle nose deformity)	<b>+2</b>
Conductive or sensorineural hearing loss	<b>+1</b>

**LABORATORY, IMAGING, AND BIOPSY CRITERIA**

Positive test for cytoplasmic antineutrophil cytoplasmic antibodies (cANCA) or antiproteinase 3 antibodies (anti-PR3)	<b>+5</b>
Pulmonary nodules, mass, or cavitation on chest imaging	<b>+2</b>
Granuloma, extravascular granulomatous inflammation, or giant cells on biopsy	<b>+2</b>
Inflammation, consolidation, or effusion of the nasal/paranasal sinuses, or mastoiditis on imaging	<b>+1</b>
Pauci-immune glomerulonephritis on biopsy	<b>+1</b>
Positive test for perinuclear antineutrophil cytoplasmic antibodies (pANCA) or antimyeloperoxidase (anti-MPO) antibodies	<b>-1</b>
Blood eosinophil count $\geq 1 \times 10^9/\text{liter}$	<b>-4</b>

**Sum the scores for 10 items, if present. A score of  $\geq 5$  is needed for classification of GRANULOMATOSIS WITH POLYANGIITIS.**

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Arthritis Rheumatol. 2022  
 Ann Rheum Dis. 2022