

## Microscopic Polyangiitis

### Clinical Background

Microscopic polyangiitis (MPA) is a necrotizing vasculitis of the small vessels without granulomatous inflammation.

#### Epidemiology

- Incidence – 16-25/1,000,000 for systemic vasculitis as a group (includes Churg-Strauss, MPA, polyarteritis nodosa [PAN] and Wegener granulomatosis)
- Age – peak onset is 40 years
- Sex – M>F

#### Pathophysiology

- Necrotizing vasculitis of the microscopic vessels (small arteries, arterioles, capillaries or venules) in sites other than kidney
- Necrotizing cresenteric glomerulonephritis on renal biopsy

#### Clinical Presentation

- Constitutional – weight loss, fever, myalgias
- Otorhinolaryngological – oral ulcers, epistaxis, sinusitis
- Renal – glomerulonephritis, proteinuria
- Cardiovascular – pericarditis, endocarditis
- Neurological – mononeuropathy multiplex, sensorimotor polyneuropathy
- Dermatological – purpura, papules, ulcers
- Ophthalmological – scleritis, uveitis, episcleritis
- Pulmonary – alveolar hemorrhage, interstitial fibrosis

### Diagnosis

- Indications for testing – multiorgan system involvement in patient
- Laboratory testing
  - Nonspecific – blood urea nitrogen (BUN)/creatinine, CBC
  - Urinalysis (UA) – hematuria, proteinuria
  - Erythrocyte sedimentation rate (ESR) – elevated >50% of the time
  - ANCA – antineutrophil cytoplasmic antibodies – myeloperoxidase predominant
  - Antiglomerular basement membrane antibodies – negative; perform to rule out Goodpasture syndrome
- Histology
  - Small and mid-size artery, venule, and capillary necrotizing vasculitis (no granulomas visualized)
  - Renal biopsy – focal segmental necrotizing, glomerulonephritis with crescents

#### Differential Diagnosis

- Vasculitis
  - Churg-Strauss syndrome
  - Polyarteritis nodosa
  - Wegener granulomatosis
- Autoimmune disease
  - Systemic lupus erythematosus (SLE)
  - Mixed connective tissue disease (MCTD)

- Goodpasture syndrome
- Endocarditis

## Lab Tests

### Indications for Laboratory Testing

Tests generally appear in the order most useful for common clinical situations. For test-specific information, refer to the test number in the ARUP Laboratory Test Directory on the ARUP Web site at [www.aruplab.com](http://www.aruplab.com).

Test Name and Number	Recommended Use	Limitations	Follow Up
Urea Nitrogen, Serum or Plasma <b>0020023</b> Method: Spectrophotometry	Test for elevated renal function		
Creatinine, Serum or Plasma <b>0020025</b> Method: Spectrophotometry	Test for elevated renal function		
CBC with Platelet Count & Automated Differential <b>0040003</b> Method: Automated Cell Count with Flow Cell Differential	Rule out infectious process		
Urinalysis, Complete <b>0020350</b> Method: Reflective Photometry/Microscopic by Yellow IRIS	Useful in assessing renal involvement	Not specific for diagnosis of microscopic polyangiitis	
Sedimentation Rate, Westergren (ESR) <b>0040325</b> Method: Westergren	Use as initial screen in vasculitis	Not specific for diagnosis of microscopic polyangiitis	
Anti-Neutrophil Cytoplasmic Antibody with Reflex to Titer & MPO/PR-3 Antibodies <b>2002068</b> Method: Indirect Fluorescent Antibody/Multi-Analyte Fluorescent Detection	Most effective test to aid in diagnosis of microscopic polyangiitis Components include anti-neutrophil cytoplasmic antibody, IgG; myeloperoxidase antibody; and serine proteinase 3 antibody		

Renal Pathology Special Studies arup029 Method: Microscopic Exam	Confirm type of vessel involvement or confirm renal glomerulonephritis	May not demonstrate disease due to skip lesions	
Glomerular Basement Membrane Antibody Panel 0051001 Method: Multi-Analyte Fluorescent Detection/Indirect Fluorescent Antibody	Rule out Goodpasture syndrome		

**Additional Tests Available**

Test Name and Number	Comments
Anti-Neutrophil Cytoplasmic Antibody, IgG 0050811 Method: Indirect Fluorescent Antibody	
Myeloperoxidase Antibody 0050526 Method: Multi-Analyte Fluorescent Detection	

**General References**

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**Diagnostic Algorithm(s)**

PDF algorithm(s) available at [www.arupconsult.com](http://www.arupconsult.com).

Vasculitis in Adults Testing Algorithm

**Related Content**

Goodpasture Syndrome - Anti-GBM Disease

Polyarteritis Nodosa - PAN

Vasculitis - ANCA

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