

## 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 <b>arup029</b> 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 <b>0051001</b> 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 <b>0050811</b> Method: Indirect Fluorescent Antibody	
Myeloperoxidase Antibody <b>0050526</b> Method: Multi-Analyte Fluorescent Detection	

**General References**

Beauvillain C, Delneste Y, Renier G, Jeannin P, Subra JF, Chevaller A. Antineutrophil cytoplasmic autoantibodies: how should the biologist manage them?. Clin Rev Allergy Immunol. 2008; 35 (1-2) 47-58.

Collins CE, Quismorio FP Jr. Pulmonary involvement in microscopic polyangiitis. Curr Opin Pulm Med. 2005; 11 (5) 447-451.

Falk RJ, Nachman PH, Hogan SL, Jennette JC. ANCA glomerulonephritis and vasculitis: a Chapel Hill perspective. Semin Nephrol. 2000; 20 (3) 233-243.

Hughes LB, Bridges SL Jr. Polyarteritis nodosa and microscopic polyangiitis: etiologic and diagnostic considerations. Curr Rheumatol Rep. 2002; 4 (1) 75-82.

Jayne D. Challenges in the management of microscopic polyangiitis: past, present and future. Curr Opin Rheumatol. 2008; 20 (1) 3-9.

Kawakami T, Soma Y, Saito C, Ogawa H, Nagahuchi Y, Okazaki T, Ozaki S, Mizoguchi M. Cutaneous manifestations in patients with microscopic polyangiitis: two case reports and a minireview. Acta Derm Venereol. 2006; 86 (2) 144-147.

Merkel PA, Seo P, Aries P, Neogi T, Villa-Forte A, Boers M, Cuthbertson D, Felson DT, Hellmich B, Hoffman GS, Jayne DR, Kallenberg CG, Krischer J, Mahr A, Matteson EL, Specks U, Luqmani R, Stone J. Current status of outcome measures in vasculitis: focus on Wegener's granulomatosis and microscopic polyangiitis. Report from OMERACT 7. J Rheumatol. 2005; 32 (12) 2488-2495.

Mukhtyar C, Flossmann O, Hellmich B, Bacon P, Cid M, Cohen-Tervaert JW, Gross WL, Guillevin L, Jayne D, Mahr A, Merkel PA, Raspe H, Scott D, Witter J, Yazici H, Luqmani RA. Outcomes from studies of antineutrophil cytoplasm antibody associated vasculitis: a systematic review by the European League Against Rheumatism systemic vasculitis task force. Ann Rheum Dis. 2008; 67 (7) 1004-1010.

Savige J, Dimech W, Fritzler M, Goeken J, Hagen EC, Jennette JC, McEvoy R, Pusey C, Pollock W, Trevisin M, Wiik A, Wong R. Addendum to the International Consensus Statement on testing and reporting of antineutrophil cytoplasmic antibodies. Quality control guidelines, comments, and recommendations for testing in other autoimmune diseases. *Am J Clin Pathol*. 2003; 120 (3) 312-318.

Watts RA, Scott DG, Lane SE. Epidemiology of Wegener's granulomatosis, microscopic polyangiitis, and Churg-Strauss syndrome. *Cleve Clin J Med*. 2002; 69 Suppl 2 S1184-S1186.

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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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