

## Guideline of Diagnosis Classification for Vasculitis: March 2002

The Research Committee on Intractable Vasculitides, the Ministry of Health and Welfare of Japan

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### Proposed diagnostic criteria for microscopic polyangiitis (MPA)

(Research Committee on Intractable Vasculitis and Research Committee on Epidemiology of Intractable Diseases, The Ministry of Health and Welfare of Japan, 1998)

#### 1. Clinical manifestations

- 1) Rapidly progressive glomerulonephritis
- 2) Pulmonary hemorrhage or interstitial pneumonia
- 3) Organ involvement besides kidney and lung: purpura, subcutaneous bleeding  
GI bleeding, mononeuritis multiplex, etc.

#### 2. Histological findings

Necrotizing vasculitis in capillaries, venules, or arterioles, with perivascular inflammatory infiltrate.

#### 3. Laboratory findings

- 1) MPO-ANCA positive
- 2) Elevated level of CRP
- 3) Proteinuria and hematuria, with elevated levels of BUN and/or creatinine
- 4) Chest X-ray findings: infiltration (pulmonary hemorrhage), or interstitial pneumonitis

#### 4. Diagnosis

- 1) Definite
  - A) At least two clinical findings with the histological finding
  - B) At least two clinical findings including items 1) and 2) with a positive MPO-ANCA.
- 2) Probable
  - A) At least three clinical findings
  - B) One clinical finding with a positive MPO-ANCA

#### 6. Exclusion criteria

- 1) Polyarteritis nodosa
- 2) Wegener's granulomatosis
- 3) Allergic granulomatous angiitis (Churg-Strauss syndrome)
- 4) Goodpasture's syndrome

#### 7. Reference

	PN	MPA
<b>Histological feature</b>	Necrotizing	Necrotizing
Type of vasculitis	Medium, small	capillary, venules,
Size of vessels	-sized muscle	arterioles
	arteries,	
<b>Clinical feature</b>		
RPGN	rare	common
Hypertension	common	rare
Pulmonary bleeding	rare	common
Relapse	rare	common
MPO-ANCA	negative	positive
Angiography		
(aneurysm, stenosis)	Yes	No
<b>Diagnosis</b>	angiography or	biopsy
	biopsy	