ACR criteria for the classification of Giant Cell Arteritis

Criterion		Definition
1	Age at onset > 50 years	Development of symptoms or findings beginning aged 50 years or older.
2	New headache	New onset of, or new type of, localized pains in the head.
3	Temporal artery abnormality	Temporal artery tenderness to palpation or decreased pulsation, unrelated to atherosclerosis of cervical arteries
4	Increase ESR	ESR > 50mm/h by Westergren method
5	Abnormal artery biopsy	Biopsy specimen with artery showing Vasculitis characterized by a predominance of mononuclear infiltration or granulomatous inflammation.

Note for purposes of classification a patient shall be said to have giant cell arteritis if at least 3 of these 5 criteria are present. The presence of any 3 or more criteria yields a sensitivity of 93.5% and specificity of 91.2%.

From Hunder [15]

ACR Classification criteria for Takayasu's arteritis

Criterion		Definition
1	Age < 40 years old	Development of symptoms or signs related to Takayasu's arteritis at age < 40 years
2	Claudication of extremities	Development and worsening of fatigue and discomfort in muscles of one or more extremity while in use, especially the upper extremities
3	Decreased brachial arterial pulse	Decreased pulsation of one or both brachial arteries
4	BP difference > 10 mmHg	Difference of > 10mmHG in systolic blood pressure between arms
5	Bruit over subclavian arteries or aorta	Bruit audible on auscultation over one or both subclavian arteries or abdominal aorta
6	Arteriogram abnormality	Arteriographic narrowing or occlusion of the entire aorta, its proximal branches, or large arteries in the proximal upper or lower extremities, not due to atherosclerosis, fibromuscular dysplasia, or similar causes; changes usually focal or segmental.

Note for purposes of classification a patient shall be said to have Takayasu's arteritis if at least 3 of these 6 criteria are present. The presence of any 3 or or more criteria yields a sensitivity of 90.5% and specifcitty of 97.8%.

From Arend [16]

ACR Classification criteria for Henoch-Schönlein purpura

Criterion		Definition
1	Palpable purpura	Slightly elevated purpuric rash over one or more areas of the skin not related to thrombocytopaenia
2	Bowel angina	Diffuse abdominal pain worse after meals, or bowel ischaemia , usually bloody diarrhoea
3	Age at onset < 20 years	Development of first symptoms at age 20 years or less
4	Wall granulocytes on biopsy	Histological changes showing granulocytes in the walls of arteries or venules

Note for purposes of classification a patient shall be said to have Henoch Schonlein purpura if at least 2 of these 4 criteria are present. The presence of any 2 or more criteria yields a sensitivity of 87.1% and specificity of 87.7%

From Mills [17]

ACR Classification criteria for hypersensitivity vasculitis

Criterion		Definition
1	Age at disease onset > 16 years	Development of symptoms age > 16 years
2	Medication at disease onset `	Medication was taken at the onset of symptoms that may have been a precipitating factor
3	Palpable purpura	Slightly elevated purpuric rash over one or more areas of the skin, does not blanch with pressure and is not related to thrombocytopaenia
4	Maculopapular rash	Flat and raised lesions of various sizes over one or more areas of the skin
5	Biopsy including arteriole and venule	Histologic changes showing granuocytes in a perivascular or extravascular location

Note for purposes of classification a patient shall be said to have Hypersensitivity vasculitis if at least 3 of these 5 criteria are present. The presence of any 3 or more criteria yields a sensitivity of 71.0%and specificity of 83.9%

From Calabrese [18]

ACR classification criteria for the classification of polyarteritis nodosa

Criterion		Definition
1	Weight loss	Loss of 4kg or more of body weight since the illness began, not due to dieting or other factors
2	Livedo reticularis	Mottled reticular pattern over the skin of portions of the extremities or torso
3	Testicular pain or tenderness	Pain or tenderness of the testicles, not due to infection, trauma, or other causes
4	Myalgias, weakness, or leg tenderness	Diffuse maylgias (excluding shoulder or hip girdle) or weakness of muscles or tenderness of leg muscles
5	Mononeuropathy or polyneuropathy	Development of mononeuroapthy, multiple mononeuroapthies or polyneuropathy
6	Diastolic BP > 90mmHg	Development of hypertension with diastolic BP > 90mmHg
7	Elevated blood urea or creatinine	Elevated BUN > 40 mg/dl or creatinine 1.5 mg/dl, not due to dehydration or obstruction
8	Hepatitis B virus	Presence of hepatitis B surface antigen or antibody in serum
9	Arteriographic abnormality	Arteriogram showing aneurysms or occlusion of the visceral arteries, not due to arteriosclerosis, fibromuscular dysplasia or other non-inflammatory causes
10	Biopsy of small or medium sized artery containing PMN	Histologic changes showing the presence of granulocytes or granulocytes and monuclear leucocytes in the artery wall

Note for purposes of classification a patient shall be said to have polyarteritis nodosa if at least 3 of these 10 criteria are present. The presence of any 3 or more criteria yields a sensitivity of 82.2% and specificity of 86.6%

From Lightfoot [19].

ACR Criteria for classification of Churg-Strauss Syndrome

Criterion		Definition
1	Asthma	History of wheezing or diffuse high pitched rales on expiration
2	Eosinophilia	Eosinophilia >10% on white cell differential count
3	Mononeuropathy or polyneuropathy	Development of mononeuropathy, multiple mononeuropathies, or polyneuropathy (i.e.glove/stocking distribution) attributable to systemic vasculitis
4	Pulmonary infiltrates, non fixed	Migratory or transient pulmonary infiltrates on radiographs (not including fixed infiltrates), attributable to a systemic vasculitis
5	Paranasal sinus abnormality	History of acute or chronic paranasal sinus pain or tenderness or radiographic opacification of the paranasal sinuses
6	Extravascular eosinophils	Biopsy including artery, artiole, or venule showing accumulations of eosinophils in extravascular areas

For purposes of classification, a person shall be said to have Churg Strauss Syndrome if at least 4 of these 6 criteria are present. The presence of any 4or more criteria yields a sensitivity of 85.0% and specificity of 99.7%.
Masi et al [20].

ACR Criteria for classification of Wegener's granulomatosis

Criterion		Definition
1	Nasal or oral inflammation	Development of painful or painless oral ulcers
or		
		purulent or bloody nasal discharge
2	Abnormal Chest radiograph	Chest radiograph showing the presence of nodules, fixed infiltrates or cavities
3	Urinary Sediment	Microhaematuria (>5 red cells per high power field) or red cell casts in urinary sediment
4	Granulomatous inflammation on biopsy	Histological changes showing granulomatous inflammation within the wall of an artery or in the perivascular or extravascular area (artery or arteriole)

For purposes of classification, a person shall be said to have Wegener's granulomatosis if at least 2 of these 4 criteria are present. The presence of any 2 or more criteria yields a sensitivity of 88.2% and specificity of 92.0%.
Leavitt [21]