# THE AMERICAN COLLEGE OF RHEUMATOLOGY 1990 CRITERIA FOR THE CLASSIFICATION OF POLYARTERITIS NODOSA

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**Criteria for the classification of polyarteritis nodosa were developed by comparing 118 patients who had this disease with 689 control patients who had other forms of vasculitis. For the *traditional format classification*, 10 criteria were selected: weight loss ≥4 kg, livedo reticularis, testicular pain or tenderness, myalgias, mononeuropathy or polyneuropathy, diastolic blood pressure >90 mm Hg, elevated blood urea nitrogen or serum creatinine levels, presence of hepatitis B reactants in serum, arteriographic abnormality, and presence of granulocyte or mixed leukocyte infiltrate in an arterial wall on biopsy. The presence of 3 or more of these 10 criteria was associated with a sensitivity of 82.2% and specificity of 86.6%. A *classification tree* was also constructed, with 6 criteria being selected. Three of these, angiographic abnormality, biopsy-proven granulocyte or mixed leukocyte infiltrate in arterial wall, and neuropathy, were criteria used in the traditional format. The other 3 criteria used in the tree format included the patient's sex, weight loss >6.5 kg, and elevated serum aspartate aminotransferase or alanine aminotransferase levels above the range of normal. The classification tree yielded a sensitivity of 87.3% and a specificity of 89.3%.**

Kussmaul and Maier described the syndrome of polyarteritis nodosa (PAN) in 1866 (1). In that and subsequent descriptions, PAN has been depicted as a necrotizing arteritis of small and medium-sized muscular arteries, affecting multiple organ systems throughout the body. The incidence and prevalence of PAN in the population are unknown, possibly because of difficulties in diagnosing and classifying the vasculitis syndromes, but it is most commonly reported in middle-aged adults, with a male predominance. A study of mortality in the city of New York from 1951 through 1959, disclosed that polyarteritis nodosa caused one-third the number of deaths as systemic lupus erythematosus during that same decade (2). Leavitt and Fauci described vasculitis overlap syndromes in several patients who fulfilled the then-extant, and largely anecdotal, diagnostic criteria for both PAN and another vasculitis syndrome (3).

The etiology of PAN remains unknown, but the histopathologic resemblance to chronic serum sickness has suggested an immune complex pathogenesis. There is substantial evidence that at least one subset of PAN patients experiences systemic vasculitis as a result of chronic hepatitis B antigen–associated immune complex disease (4). Other reports suggesting an immune complex etiology have included the description of classic PAN following shortly after the occurrence of serous otitis media in adults (5), and the

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finding of histopathologic lesions indistinguishable from PAN in coronary blood vessels of children recovering from the childhood febrile exanthem, Kawasaki disease. The finding that vasculitic lesions are usually in various stages of development in a patient with PAN suggests chronic, cyclic insults, rather than a single moment of onset.

The clinical findings in PAN are nonspecific, and they include systemic complaints, such as fever or weight loss, and focal symptoms resulting from vascular impairment in specific organ systems (e.g., mononeuritis multiplex, Raynaud's phenomenon, gastrointestinal infarction).

There are no specific serologic tests for PAN. In general, laboratory abnormalities simply reflect the degree of inflammation (i.e., elevated acute-phase reactants, leukocytosis, thrombocytosis) and the relative vascular impairment of specific organs. While angiographic studies may be extremely helpful in establishing a diagnosis, routine radiographs offer little more than documentation of organ system involvement, and may only be helpful in directing biopsy studies.

In the present study, patients with vasculitis occurring in the presence of a connective tissue disorder, mainly, systemic lupus erythematosus and rheumatoid arthritis, were excluded from analysis. Of the patients remaining, those whose physicians considered their disorder to be PAN were compared with all those diagnosed as having other vasculitis conditions.

For review of the selection criteria, see the introductory report by Hunder et al, which appears elsewhere in this issue of *Arthritis and Rheumatism* (6). For a discussion of the methods, see the article by Bloch et al, which also appears in this issue (7).

## RESULTS

**Patient population.** One hundred eighteen patients were considered by their physicians to have a diagnosis of PAN. Sixty-two percent of these patients were male, and their average age was 48.4 years (±1.7 SEM). The criteria for classifying PAN, as described below, were developed by comparing these 118 patients with the 689 other patients with vasculitis in this study. This number excluded patients with vasculitis in the presence of a known connective tissue disorder, such as systemic lupus erythematosus or rheumatoid disease.

**Data analysis.** The several hundred individual items in these patients' reports were subjected to univariate analysis, and from these, the subcommittee derived several combined variables that were also analyzed. Table 1 shows the data on a number of the individual and combined variables examined. Included are some variables that the committee intuitively believed were candidate criteria, which on analysis, proved not to be so. Table 1 also indicates the number of cases and controls in whom a particular variable was reported. Sensitivity indicates the percentage of PAN patients in whom the variable was present or abnormal. Specificity indicates the proportion of the 689 control patients in whom the variable was absent or normal.

**Traditional format classification.** A "short list" of 14 variables that would have the greatest potential for separating cases of polyarteritis nodosa from the other cases of vasculitis was selected (see Table 1). Members of the Subcommittee on Classification of Vasculitis were asked to submit for analysis as many combinations of the 14 "short-list" variables as they intuitively considered likely to yield good discriminatory criteria in the traditional "n choose k" format, as described by Bloch et al (7).

Of the 35 potential criteria sets submitted for analysis, the 10 criteria listed in Table 2 gave the best discrimination. The presence of 3 or more of the 10 criteria was associated with a sensitivity of 82.2% and a specificity of 86.6%. Addition of abdominal angina or bowel perforation as an eleventh criterion would have increased the sensitivity to 83.9%, but the specificity would have fallen slightly, to 84.9%.

The levels of sensitivity and specificity achieved with the "3 or more of 10 criteria" rule indicate 17.8% false-negative and 13.4% false-positive classifications, respectively. Ninety-two of the 689 controls were found to have been misclassified as having PAN. The 6 most common false-positive misclassifications were: Wegener's granulomatosis in 24 patients, vasculitis of unspecified type in 14 patients, Churg-Strauss syndrome in 10 patients, giant cell (temporal) arteritis in 9 patients, hypersensitivity vasculitis in 8 patients, and Takayasu arteritis in 8 patients. Sixty-three of the 92 misclassified controls had not had visceral angiography, and 24 had not had vessel biopsy. In fact, the 14 patients with vasculitis of unspecified type may have had PAN, but the cases were not sufficiently investigated. It is also likely that the misclassified control patients with Wegener's granulomatosis, Churg-Strauss syndrome, and hypersensitivity vasculitis were diagnosed on the basis of extravascular manifestations (e.g., eosinophilia, history of allergy, sinus involvement, granulomatous inflam-

**Table 1.** Comparison of the sensitivity and specificity of potential criteria variables for the classification of polyarteritis nodosa*

| | Criterion | No. of cases (n = 118) | No. of controls (n = 689) | Sensitivity (%) | Specificity (%) |
|---|---|---|---|---|---|
| 1. | Male sex†‡ | 118 | 686 | 61.9 | 59.9 |
| 2. | Weight loss ≥4 kg†‡§ | 112 | 645 | 67.0 | 67.8 |
| 3. | Testicular pain | 90 | 510 | 21.1 | 97.8 |
| 4. | Testicular tenderness | 90 | 510 | 24.4 | 97.8 |
| 5. | Testicular pain or tenderness†‡§ | 90 | 509 | 28.9 | 97.4 |
| 6. | Abdominal angina | 118 | 683 | 20.3 | 92.7 |
| 7. | Ischemic bowel | 116 | 684 | 11.2 | 95.5 |
| 8. | Bowel perforation | 117 | 684 | 5.1 | 99.0 |
| 9. | Abdominal angina or ischemic perforation† | 118 | 683 | 23.7 | 88.7 |
| 10. | Muscle tenderness, legs | 117 | 681 | 38.5 | 86.6 |
| 11. | Muscle weakness, general | 117 | 684 | 35.0 | 87.0 |
| 12. | Myalgias (excluding shoulder and hip girdle) | 113 | 680 | 36.3 | 86.8 |
| 13. | Myalgias, general muscle weakness, or leg muscle tenderness†§ | 114 | 680 | 69.3 | 69.1 |
| 14. | Polyneuropathy | 117 | 683 | 35.9 | 92.2 |
| 15. | Mononeuritis | 118 | 687 | 25.4 | 95.9 |
| 16. | Mononeuritis multiplex | 118 | 687 | 42.4 | 94.3 |
| 17. | Mononeuropathy or polyneuropathy†‡§ | 118 | 683 | 65.3 | 85.9 |
| 18. | Diastolic BP >90 mm Hg†‡§ | 84 | 456 | 36.9 | 84.9 |
| 19. | Ecchymoses, petechiae, or splinter hemorrhages | 115 | 685 | 24.3 | 79.6 |
| 20. | Dermal or periungual infarction or peripheral gangrene | 116 | 679 | 20.7 | 87.5 |
| 21. | Cutaneous ulcers | 117 | 683 | 16.2 | 88.7 |
| 22. | Cutaneous ulcers or pitted scars | 117 | 681 | 18.8 | 88.1 |
| 23. | Cutaneous ulcers, dermal or periungual infarction, or peripheral gangrene | 116 | 679 | 26.7 | 82.8 |
| 24. | Livedo reticularis†‡§ | 117 | 681 | 24.8 | 94.0 |
| 25. | BUN >40 mg/dl | 110 | 488 | 22.7 | 87.5 |
| 26. | Creatinine >1.5 mg/dl | 117 | 655 | 33.3 | 84.4 |
| 27. | BUN >40 mg/dl or creatinine >1.5 mg/dl†§ | 110 | 493 | 40.0 | 77.3 |
| 28. | Elevated serum AST (SGOT) | 101 | 563 | 54.5 | 76.9 |
| 29. | Elevated serum ALT (SGPT) | 55 | 207 | 67.3 | 68.8 |
| 30. | Elevated serum AST or ALT†‡ | 74 | 269 | 83.8 | 43.5 |
| 31. | Serum hepatitis B surface antigen | 105 | 322 | 21.0 | 96.0 |
| 32. | Serum hepatitis B surface antibody | 70 | 214 | 7.1 | 91.6 |
| 33. | Serum hepatitis B surface antigen or antibody†§ | 76 | 211 | 31.6 | 87.7 |
| 34. | Visceral arteriographic aneurysms | 67 | 156 | 67.2 | 94.9 |
| 35. | Visceral arteriographic occlusion | 66 | 160 | 36.4 | 93.8 |
| 36. | Visceral arteriographic aneurysms or occlusions†‡§ | 68 | 157 | 73.5 | 89.2 |
| 37. | Artery wall granulocytes | 87 | 445 | 32.2 | 96.4 |
| 38. | Artery wall granulocytes and mononuclear cells | 87 | 444 | 17.2 | 93.9 |
| 39. | Biopsy of small or medium-sized artery showing granulocytes, with or without mononuclear cells†‡§ | 87 | 444 | 48.3 | 90.3 |
| 40. | Visceral arteriographic abnormality or biopsy with granulocytes, with or without mononuclear cells† | 92 | 123 | 92.4 | 53.7 |

* Values are the number of cases or controls with the variable described or tested. The sensitivity is the proportion of cases positive for the variable tested or described. The specificity is the proportion of controls negative for the variable tested or described. BP = blood pressure; BUN = blood urea nitrogen; AST = aspartate aminotransferase; SGOT = serum glutamic oxaloacetic transaminase; ALT = alanine aminotransferase; SGPT = serum glutamic pyruvic transaminase.
† Criterion is one of the final "short list" of variables (n = 14) (see text).
‡ Criterion is used for the tree classification (except that weight loss is >6.5 kg).
§ Criterion is used for the traditional format classification.

Table 2. 1990 criteria for the classification of polyarteritis nodosa (traditional format)*

| Criterion | Definition |
|---|---|
| 1. Weight loss ≥4 kg | Loss of 4 kg or more of body weight since 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 myalgias (excluding shoulder and hip girdle) or weakness of muscles or tenderness of leg muscles |
| 5. Mononeuropathy or polyneuropathy | Development of mononeuropathy, multiple mononeuropathies, or polyneuropathy |
| 6. Diastolic BP >90 mm Hg | Development of hypertension with the diastolic BP higher than 90 mm Hg |
| 7. Elevated BUN or creatinine | Elevation of 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 occlusions of the visceral arteries, not due to arteriosclerosis, fibromuscular dysplasia, or other noninflammatory causes |
| 10. Biopsy of small or medium-sized artery containing PMN | Histologic changes showing the presence of granulocytes or granulocytes and mononuclear leukocytes in the artery wall |

* For classification purposes, 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 a specificity of 86.6%. BP = blood pressure; BUN = blood urea nitrogen; PMN = polymorphonuclear neutrophils.

mation on biopsy, or recent challenge with a hypersensitizing agent).

Twenty-one of the 118 patients deemed by their physicians to have PAN were misclassified as not having PAN by these traditional format criteria. Of these 21 patients, 13 had not had visceral angiography, 6 had not had a biopsy, 10 had not been tested for hepatitis B reactants, and 11 had no diastolic blood pressure value reported. Thus, most misclassifications of either type in this data set may well have resulted from incomplete exploration for the 10 criteria in question or from too prompt labeling of alternative vasculitides according to the presence of manifestations typical for them.

**Tree classification.** Figure 1 represents the best of several tree classifications derived using the CART computer program as described by Bloch et al (7). All 14 short-list variables in Table 1 were included as potential discriminators. When data concerning a given criterion are missing, then surrogate criteria that best approximate the split of the main criterion are used. The classification tree shown incorporated misclassification costs of 6:1 (see ref. 7). In other words, the correct classification of a PAN patient was deemed to be 6 times as important as the correct classification of a control, since there was one-sixth as many cases as controls.

Table 3 describes the classifying subsets of the tree. Seven of the criteria or surrogates are defined in Table 2 (except that the cut-point for weight loss is 6.5 kg in the tree structure). Of the other 2, one is the sex of the patient, and the other indicates elevation of the level of either aspartate aminotransferase or alanine aminotransferase above the normal range. The tree classification yielded a sensitivity of 87.3% and a specificity of 89.3%. Of the 7 classifying subsets, 3 (numbers 1, 2, and 5, Figure 1) represent groups classified as control patients (i.e., those where optimally all control patients should appear) while 4 subsets (numbers 3, 4, 6, and 7) represent groups classified as cases.

Fifteen PAN patients were misclassified as controls, 8 in subset 1 and 7 in subset 2; there was no misclassification in subset 5. Three of the 8 PAN patients of subset 1 had no visceral angiographic data, and 3 lacked biopsy information. Of the 7 PAN patients misclassified in subset 2, 6 were women (could not fulfill the criterion for testicular abnormalities), 4 had no visceral arteriography data, and 1 had no vessel biopsy data.

Of the 689 control patients, 74 (10.7%) were misclassified as having PAN (subset numbers 3, 4, 6, and 7, Figure 1). Several factors may explain this misclassification. First, 14 of the misclassified patients were thought to have "vasculitis of unspecified type" and may well have had PAN but were not completely examined. In addition, Churg-Strauss syndrome, giant cell (temporal) arteritis, Wegener's granulomatosis, and hypersensitivity vasculitis, respectively, accounted for 9, 10, 13, and 5 of the 74 misclassified controls. For reasons stated in the above paragraphs, these diagnoses may have been based on typical manifestations of those illnesses, which do not appear as negative criteria in the PAN classification. Second, a significant number of the 74 misclassified controls lacked important data for the classification tree shown in Figure 1. Fifty-five had no angiographic study report, and 15 had no biopsy report.

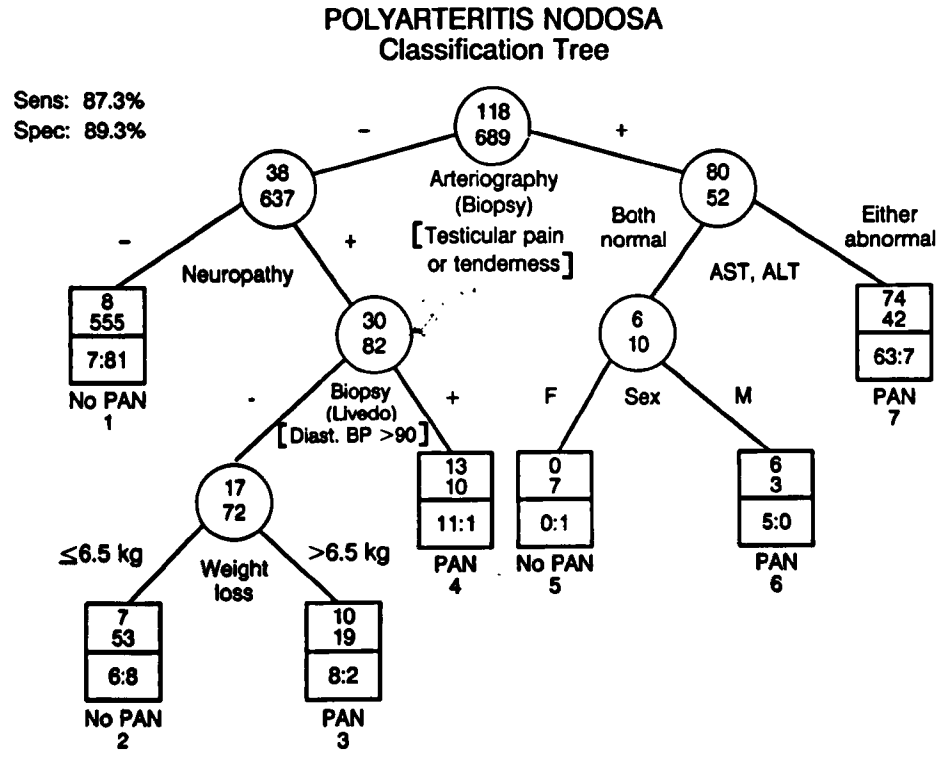


**Figure 1.** Classification tree for polyarteritis nodosa (PAN). The circles and boxes contain the number of patients with PAN (top number) and the number of control patients with other forms of vasculitis (bottom number). The bottom half of the boxes shows the percentage of patients with PAN (out of all PAN cases) (left number) and the percentage of controls (out of all controls) (right number). Boxes specify whether subjects are classified as having PAN or not having PAN (No PAN); the numbers under these specifications are the subset numbers (see Table 2 for definitions of criteria and Table 3 for explanations of subsets). Parentheses and brackets indicate the first and second surrogate criteria, respectively, that are to be used when the dividing criterion is unavailable. AST = aspartate aminotransferase; ALT = alanine aminotransferase; Diast. BP = diastolic blood pressure (in mm Hg).

**Table 3.** 1990 classification tree criteria for polyarteritis nodosa (PAN)*

| PAN subsets | No. of patients PAN/ non-PAN | % correctly classified | % PAN patients in subset | Non-PAN subsets | No. of patients PAN/ non-PAN | % correctly classified | % non-PAN patients in subset |
|---|---|---|---|---|---|---|---|
| 3. Neuropathy and weight loss >6.5 kg; negative arteriogram and artery biopsy | 10/19 | 34 | 8 | 1. Negative arteriogram and no neuropathy | 8/555 | 99 | 81 |
| 4. Positive artery biopsy and neuropathy; negative arteriogram | 13/10 | 57 | 11 | 2. Neuropathy; negative arteriogram and artery biopsy; weight loss ≤6.5 kg | 7/53 | 88 | 8 |
| 6. Positive arteriogram, male sex; normal AST and ALT | 6/3 | 67 | 5 | 5. Positive arteriogram; normal AST and ALT; female sex | 0/7 | 100 | 1 |
| 7. Positive arteriogram and abnormal AST or ALT | 74/42 | 64 | 63 | | | | |

* The subset numbers also appear below the subset boxes in Figure 1. Missing data rules: If angiogram not available, substitute vessel biopsy (first surrogate); if this is not defined, substitute testicular pain/tenderness (second surrogate); analogous rule is to be applied for the other dividing variable whose surrogates are provided (see Figure 1). The classification tree yields a sensitivity of 87.3% and a specificity of 89.3%. AST = aspartate aminotransferase; ALT = alanine aminotransferase. See Table 2 for definitions of criteria.

## DISCUSSION

The sensitivity and specificity of using either the traditional or the classification tree formats indicate that PAN remains one of the most difficult of the vasculitides to classify. Among the reasons suggested by the current analysis is the likelihood that, in many cases, complete ascertainment of potentially important criteria was not deemed necessary by the treating physician. It is conceivable that severe necrotizing arteritis of unknown type is thought by some to be a sufficient reason to warrant high-dose steroid treatment in the absence of additional studies to confirm the diagnosis. This may explain the frequency of missing data in the 74 misclassified controls. Many of the control patients misclassified as having PAN had syndromes such as giant cell (temporal) arteritis, Wegener's granulomatosis, and Churg-Strauss syndrome, which were probably diagnosed by the clinicians submitting these cases based on the classic findings of necrotizing upper airways disease, long history of atopy, and eosinophilia or granulomatous histopathology, which mitigated the need for further studies.

In a recently reported hypothetical analysis of strategies for diagnosing PAN, Albert et al (8) used sensitivity and specificity numbers derived from the literature and found that a conservative strategy consisting of one biopsy procedure plus visceral angiography was as cost-effective as an aggressive strategy of repeated tests until a positive finding was encountered or until available tests were exhausted. Many of the controls misclassified in the current analysis simply lacked angiographic or biopsy data and, presumably, had an alternative diagnosis made by the clinician based on classic extravascular features. It would appear that many patients with documented necrotizing arteritis are started on therapy for the diagnosis of "vasculitis of unspecified type" when initiation of treatment is deemed more critical than absolute documentation of the specific form of vasculitis present.

Rheumatoid arthritis and systemic lupus erythematosus patients with vasculitis were excluded from the control population in the current analysis, and it seems reasonable in practice to exclude these illnesses prior to submitting a patient to the tests required by the PAN criteria proposed here. In addition, because Wegener's granulomatosis responds dramatically to alkylating agents without necessarily requiring steroids to abate the acute inflammatory process, and because the above data suggest that many patients with Wegener's granulomatosis are diagnosed as such on the basis of manifestations other than the 10 listed criteria for the traditional format classification of PAN, it is reasonable to attempt to exclude Wegener's granulomatosis by the appropriate criteria (described elsewhere in this issue by Leavitt et al [9]) before subjecting a patient to the PAN classification studies. Having excluded these more easily diagnosed illnesses, both Albert and coworkers' data (8) and our experience with the missing data for biopsy and angiographic studies suggest that no patient with a potential diagnosis of PAN should be considered not to have PAN until angiograms or relevant vessel biopsies are performed.

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