THE AMERICAN COLLEGE OF RHEUMATOLOGY 1990 CRITERIA FOR THE CLASSIFICATION OF CHURG-STRAUSS SYNDROME (ALLERGIC GRANULOMATOSIS AND ANGIITIS)

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Criteria for the classification of Churg-Strauss syndrome (CSS) were developed by comparing 20 patients who had this diagnosis with 787 control patients with other forms of vasculitis. For the *traditional format classification*, 6 criteria were selected: asthma, eosinophilia >10% on differential white blood cell count, mononeuropathy (including multiplex) or polyneuropathy, non-fixed pulmonary infiltrates on roentgenography, paranasal sinus abnormality, and biopsy containing a blood vessel with extravascular eosinophils. The presence of 4 or more of these 6 criteria yielded a sensitivity of 85% and a specificity of 99.7%. A *classification tree* was also constructed with 3 selected criteria: asthma, eosinophilia >10% on differential white blood cell count, and history of documented allergy other than asthma or drug sensitivity. If a subject has eosinophilia and a documented history of either asthma or allergy, then that subject is classified as having CSS. For the tree classification, the sensitivity was 95% and the specificity was 99.2%. Advantages of the traditional format compared with the classification tree format, when applied to patients with systemic vasculitis, and their comparison with earlier work on CSS are discussed.

In 1951, Churg and Strauss (1) reported 13 cases of severe asthma with a "strikingly uniform clinical picture," including fever, hypereosinophilia, and evidence of vascular abnormality in various organ systems. Pathologic examination of these patients revealed granulomatous extravascular lesions as well as inflammatory, necrotizing, and granulomatous vascular changes. All but 2 patients died of the illness; these 11 cases were detected through autopsy files, having been originally categorized as having periarteritis nodosa (1). The presence of the characteristic generalized granulomatous lesions suggested that these cases constituted an entity apart from periarteritis nodosa (as it was then called) and apart from Wegener's granulomatosis (1,2). They reviewed 15 cases of periarteritis nodosa without asthma. None showed the granulomatous lesions. Histologically, these "allergic granulomas" were composed of necrotic eosinophilic exudates, severe "fibrinoid" collagen changes, and granulomatous proliferation of epithelioid and giant cells. It was suggested that other allergic syndromes may represent more benign forms of "allergic granulomatosis," while angiitis is its most malignant expression. Hence arose the concept of "allergic granulomatosis and angiitis" or Churg-Strauss syndrome (CSS).

Over the 25-year interval of 1950 to 1974, 30

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cases of allergic granulomatosis and angiitis (Churg-Strauss syndrome) were diagnosed in patients at the Mayo Clinic, including 6 identified at autopsy (3). All patients had bronchial asthma, peripheral eosinophilia (at some time), and systemic vasculitis that was deemed different from polyarteritis nodosa (PAN). Although no specific comparison with PAN was made, renal disease was not prominent in CSS. Only 1 patient with CSS had renal failure. Pathologically, all patients had necrotizing vasculitis of small arteries and veins, with prominent eosinophilia of vessels and perivascular tissues, accompanying lymphocytes, plasma cells, and some histiocytes. Necrotizing extravascular granulomatosis was reported in 22 (73.3%) of the series, and fibrinoid necrosis of vessel walls in 12 (40%).

As in the series described by Churg and Strauss (1), the 15 patients in the Mayo Clinic series (3) who died had had asthma for a mean interval of only 3 years prior to the onset of vasculitis. However, among the remaining 15 survivors, the mean duration of asthma prior to the onset of CSS was 13 years. In 1981, Churg and Strauss (4) broadened their concept of allergic granulomatosis and angiitis to include eosinophilic pneumonitis, eosinophilic non-necrotizing angiitis, bronchocentric granuloma, and allergic granuloma, as well as necrotizing angiitis.

In 1984, Lanham and associates (5) emphasized, as previously indicated by Chumbley and associates (3), that not all cases have the 3 major histologic features originally described (i.e., tissue infiltration by eosinophils, necrotizing vasculitis, and extravascular granulomas) (1). Conceptually, CSS was portrayed as a point of overlap among: hypereosinophilic disease (e.g., Löffler's syndrome), systemic vasculitis (e.g., PAN and hypersensitivity vasculitis), and granulomatous disorders (e.g., Wegener's granulomatosis).

A clinical approach to this syndrome was emphasized (5), and no histologic criteria for a diagnosis of systemic vasculitis were defined. They indicated, moreover, that the histologic "Churg-Strauss granuloma" (6) is not pathognomonic of CSS. Sixteen patients seen at their institution (5) between 1976 and 1982, and a further 138 patients culled from the literature, met their proposed criteria: asthma, peak peripheral blood eosinophil count in excess of 1.5×10^9/liter (1,500/mm^3), and systemic vasculitis involving 2 or more extrapulmonary organs. The mean peak white blood cell (WBC) count and eosinophil count were 18.3×10^9/liter and 8.4×10^9/liter, respectively (18,300/mm^3 and 8,400/mm^3, respectively, or 46% eosinophils, on the average).

A phasic pattern of CSS was described by Lanham et al (5), that is, an initial allergic disease, usually allergic rhinitis, evolving into asthma, followed by peripheral blood eosinophilia, and eosinophilic tissue infiltrates, and, finally, a vasculitic phase. CSS proved fatal in only 1 of 16 patients reported in their personal series. All but 1 patient had a rapid initial response to oral prednisone or prednisolone (30–60 mg daily). In contrast, among the 138 cases culled from the English language literature (5), the majority of whom were seen prior to the advent of corticosteroids, most had died of CSS. In that earlier literature series, granulomas were seen in 40% of subjects at autopsy and in 38% of positive tissue biopsies, compared with only 1 of 14 positive tissue specimens from Lanham's own series (between 1976 and 1982). No important difference in clinical expression of disease was found in patients with granulomas versus those without.

The classification of vasculitis and a reappraisal of allergic granulomatosis and angiitis (Churg-Strauss syndrome) was reviewed recently (7). The "Churg and Strauss granuloma" (6) may occur as a localized, isolated, or limited entity, and its diverse associations in various systemic diseases further complicates the nosology of CSS. The necrotizing vasculitis in Churg-Strauss syndrome may be indistinguishable from that in PAN and that in hypersensitivity angiitis (7). Nevertheless, a review of all 54 cases previously described by investigators at the Mayo Clinic, patients who fulfilled the diagnostic criteria for CSS proposed by Lanham et al (5), seemed to yield a group with features that constituted a distinctive morphologic pattern, albeit a syndrome that overlaps with PAN and Wegener's granulomatosis (7).

For current concepts of the stereotypical pathologic features of CSS, see the article by Lie et al (8), which appears elsewhere in this issue of *Arthritis and Rheumatism*. For a description of the methods of patient selection and evaluation, see the papers by Bloch et al (9) and Hunder et al (10), which also appear elsewhere in this issue.

RESULTS

Patient population. The 20 submitted CSS patients whose data we studied had a mean age at disease onset of 50 years (±13.2 SD), with a range of 16–74 years, were predominantly males (65%), and included 1 non-Caucasian. The mean age at onset of vasculitis in both the personal series and the literature series described by Lanham et al (5) was 38 years, and most patients were males.

Selected variables found to discriminate the 20 CSS patients from the 787 control patients with other

Table 1. Comparison of the sensitivity and specificity of potential criteria variables for Churg-Strauss syndrome*

Criterion	Cases studied (n = 20)	Controls studied (n = 787)	Sensitivity (%)	Specificity (%)
Clinical				
1. Allergy, seasonal	18	753	39	94
2. Allergy, other (besides drug)	17	744	35	95
3. Allergy, seasonal or other	17	739	59	89
4. Asthma	19	782	100	96
5. Chronic paranasal sinus pain or tenderness over sinuses	20	778	40	95
6. Eosinophilia >5%	20	708	95	87
7. Eosinophilia >7.5%	20	708	95	95
8. Eosinophilia >10%	20	708	95	97
9. Mononeuropathy or mononeuritis multiplex	20	785	65	86
10. Polyneuropathy	20	780	45	89
11. Mononeuropathy or polyneuropathy	20	781	75	80
Radiologic				
12. Pulmonary infiltrates, fixed	20	741	25	92
13. Pulmonary infiltrates, migratory	20	731	15	98
14. Pulmonary infiltrates, transitory	20	736	30	94
15. Pericardial effusion	20	740	25	98
16. Paranasal sinus opacification	12	354	83	82
Biopsy†				
17. Periarterial eosinophils	11	517	27	95
18. Wall arterial eosinophils	11	520	36	93
19. Extraarterial eosinophils	11	509	64	95
20. Periarteriolar eosinophils	18	473	39	93
21. Wall arteriolar eosinophils	19	474	37	95
22. Extraarteriolar eosinophils	18	474	39	94
23. Perivenular eosinophils	15	454	47	94
24. Wall venular eosinophils	15	455	33	95
25. Extravenular eosinophils	15	454	40	94

* Entries are the number of cases or controls with the variable described or tested. The sensitivity is the proportion of positive cases among Churg-Strauss syndrome cases defined on that variable. The specificity is the proportion of negative controls among other vasculitis syndrome controls defined on that variable.
† Biopsy showing eosinophils in specified locations: arterial, arteriolar, or venular.

vasculitis syndromes are shown in Table 1. These items were selected on the basis of univariate analysis of all coded data from the protocol forms of cases and controls, as described in the methodology paper by Bloch et al (9). All 19 patients for whom data on the history of asthma were available were positive for this item, compared with only 4% among the controls. Interestingly, only 4% of the patients with PAN also had a history of asthma. All but 1 CSS patient had eosinophilia of >10% on WBC differential count, compared with 3.4% among the controls. About one-third of the CSS patients gave a positive history of seasonal allergy, other allergy (besides drugs), or chronic paranasal pain/tenderness, versus 5–6% among the controls. Mononeuritis or mononeuritis multiplex and polyneuropathy were each found in approximately 50% of the patients versus 14% and 11%, respectively, of the controls. Chest roentgenograms showed modest frequencies of lung infiltrates of various kinds, as well as pericardial effusion, and in each instance, the frequency was significantly greater than that among controls. Paranasal sinus opacification was found in 10 (83%) of 12 patients who underwent this radiographic examination, versus only 18% among the controls. Biopsied vessels of all sizes and types showed eosinophils in moderate frequency: 27–64% in perimural, intramural, and extramural locations (versus 5–7% in the controls).

Traditional format classification. Using multivariate analytic techniques, as reviewed in Bloch et al's methodology paper (9), and combining selected individual variables listed in Table 1, we selected 6 criteria that most effectively discriminated Churg-Strauss syndrome patients from other vasculitis patients, when 4 or more of the criteria are positive (Table 2). This particular set of 6 criteria was chosen over 33 other potential criteria sets that included from

Table 2. 1990 criteria for the classification of Churg-Strauss syndrome (traditional format), their sensitivity and specificity versus other defined vasculitis syndromes*

Criterion	No. of CSS patients (n = 20)	Sensitivity (%)	No. of control patients (n = 787)	Specificity (%)
Asthma	19	100	782	96.3
Eosinophilia >10%	20	95	708	96.6
Neuropathy, mono or poly	20	75	781	79.8
Pulmonary infiltrates, non-fixed	20	40	736	92.4
Paranasal sinus abnormality	14	85.7	366	79.3
Extravascular eosinophils	16	81.3	385	84.4

* For classification purposes, a patient shall be said to have Churg-Strauss syndrome (CSS) if at least 4 of these 6 criteria are positive. The presence of any 4 or more of the 6 criteria yields a sensitivity of 85% and a specificity of 99.7%. (See Table 3 for criteria definitions.)

1 (i.e., asthma alone) to 8 criteria and several different combinations of histologic abnormalities. The criteria set selected provided good sensitivity (i.e., percentage of correctly classified cases) and excellent specificity (i.e., percentage of correctly classified controls).

Considering the data available in the protocols, 17 (85%) of the 20 CSS patients submitted were positive for 4 or more of the 6 criteria, versus only 2 of the 787 controls. The sensitivity of this criteria set is 85%, and the specificity is 99.7%. Thus, only 5 (0.6%) of the 807 vasculitis patients were misclassified according to these criteria, i.e., 3 (15%) of the 20 cases and 2 (0.3%) of the 787 control patients.

Tree classification. The classification tree process, as reviewed by Bloch et al (9), yielded criteria for CSS, as shown in Figure 1. Asthma was the most discriminative in segregating the cases and controls. All 19 CSS patients with data on history of asthma were positive for this variable, as were 29 of the 787 controls. In patients with a history of asthma, eosinophilia >10% on a WBC differential smear was found in 18 cases and 2 controls. Thus, this combination of 2 criteria correctly classified 90% of the CSS cases (i.e., 18 of 20 cases) and misclassified only 2 of the controls (i.e., 785 of 787 controls; specificity 99.7%) (Figure 1, subset 5). Five study subjects who did not have a positive history of asthma, satisfied the tree structure criteria of eosinophilia >10% on WBC differential count plus a history of seasonal or other documented, non-drug allergy (Figure 1, subset 3), and 4 of these 5 were misclassified as CSS cases. Thus, this subset correctly classified 1 additional case (5% of cases) and misclassified 4 controls (0.5% of controls).

The variable, history of seasonal *or* other allergy, besides drug, has a sensitivity of 58.8% among 17 CSS cases and a specificity of 89.3% among 739 control vasculitis patients for whom such information was available (Table 1). The overall sensitivity of the classification tree is 95% (i.e., 19 of 20 cases are correctly classified), and the specificity is 99.2% (i.e., 6 of 787 controls are misclassified).

Criteria definitions used for the classification of Churg-Strauss syndrome, either for the traditional format (Table 2) or for the classification tree (Figure 1), are specified in Table 3. The selective variables for the 2 case and 3 control subsets of the classification tree, represented by the boxes in Figure 1, and their efficacy are shown in Table 4.

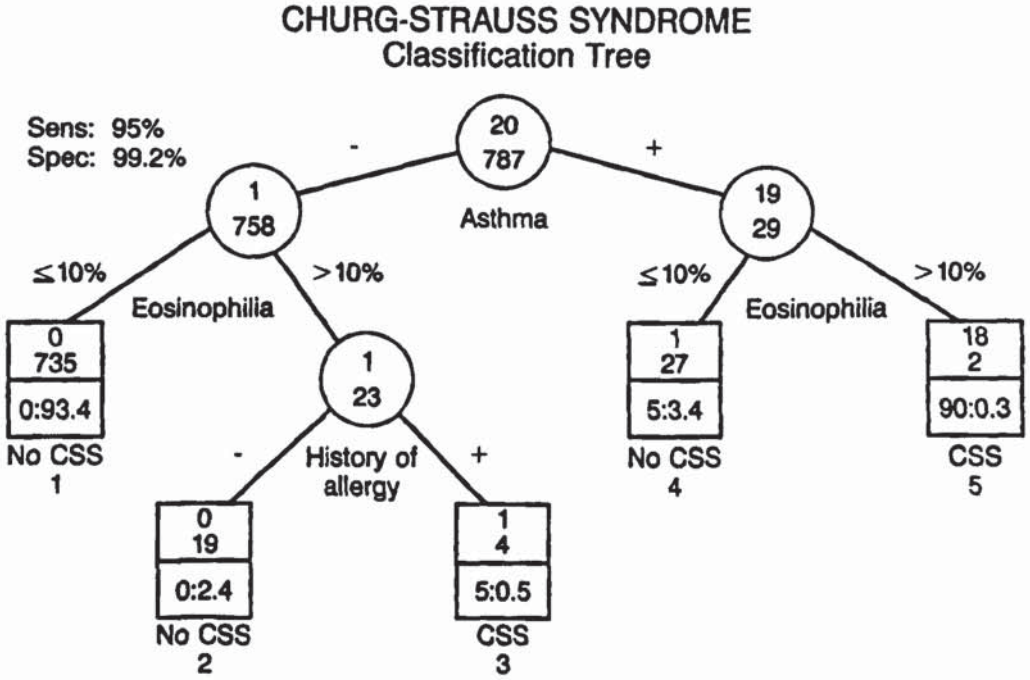

Figure 1. Classification tree for Churg-Strauss syndrome (CSS). The circles and boxes contain the number of patients with CSS (top number) and the number of control patients with other forms of vasculitis (bottom number). The bottom half of the boxes shows the percentage of patients with CSS (out of all CSS cases) (left number) and the percentage of controls (out of all controls) (right number). Boxes include subsets of patients either classified as having CSS or not having CSS (No CSS); the numbers under these specifications are the subset numbers (see Table 3 for definitions of criteria and Table 4 for explanations of subsets).

Table 3. Criteria and definitions used for the classification of Churg-Strauss syndrome

Criterion	Definition
Asthma	History of wheezing or diffuse high-pitched rales on expiration
Eosinophilia	Eosinophilia >10% on white blood cell differential count
History of allergy*	History of seasonal allergy (e.g., allergic rhinitis) or other documented allergies, including food, contactants, and others, *except* for drug allergy
Mononeuropathy or polyneuropathy	Development of mononeuropathy, multiple mononeuropathies, or polyneuropathy (i.e., glove/stocking distribution) attributable to a systemic vasculitis
Pulmonary infiltrates, non-fixed	Migratory or transitory pulmonary infiltrates on radiographs (not including fixed infiltrates), attributable to a systemic vasculitis
Paranasal sinus abnormality	History of acute or chronic paranasal sinus pain or tenderness or radiographic opacification of the paranasal sinuses
Extravascular eosinophils	Biopsy including artery, arteriole, or venule, showing accumulations of eosinophils in extravascular areas

* History of allergy, other than asthma or drug-related, is included only in the tree classification criteria set and not in the traditional format criteria set, which requires 4 or more of the 6 other items listed here (and in Table 2).

The composite variable, eosinophils located in the wall of an artery, arteriole, or venule (individual items in Table 1), was not selected as a criterion. It was found in 60% of cases and 17% of controls, which is not as sensitive or specific as the criterion of eosinophils in an *extravascular* location (Tables 2 and 3), which has a sensitivity of 81% and specificity of 16%. Use of the combination of *either* biopsy variable did not give better discrimination than the item of extravascular eosinophils alone. In order for a biopsy to be relevant to these criteria, it must include an artery, arteriole, or venule (8), as specified in Table 3.

DISCUSSION

The criteria listed in Table 3 provide a combination of simple features from the patient's medical history, physical examination, and laboratory, radiographic, and histologic findings that can be applied concurrently or retrospectively. In patients with well-documented systemic vasculitis, as in this study, the classification tree criteria might be advantageous because of their simplicity. The combination of only 2 criteria, asthma *and* eosinophilia, or the alternative criteria set, for patients without asthma, of eosinophilia *and* other documented allergy (including allergic rhinitis), discriminated CSS cases effectively. The combination of asthma and eosinophilia yielded a sensitivity of 90% and a specificity of 99.7% in this study population.

Alternatively, the traditional format criteria rule can be applied. This requires the presence of 4 or more of the 6 items, as shown in Table 2. All CSS patients, except for a doubtful case (case 301, see below), had eosinophilia on peripheral WBC differential counts that was >10% (range 11–77%, in either past or present hospital admissions, with a mean value of 40%). All but 2 CSS patients (case 301 being 1 of them) had absolute eosinophil counts of $>1.5 \times 10^9$/liter (1,500/mm^3), a diagnostic criterion proposed by Lanham et al (5). In contrast, only 1 of 29 control vasculitis patients with a history of asthma had this degree of eosinophilia.

Certain skin manifestations are common in CSS, for example, palpable purpura (45% in this series), maculopapular rash (40%), and even subcutaneous nodules (20%), but these clinical variables were not discriminating for CSS compared with the other

Table 4. 1990 classification tree criteria for Churg-Strauss syndrome (CSS)*

CSS subsets	No. of patients CSS/non-CSS	% correctly classified within subset	% of total CSS patients in subset	Non-CSS subsets	No. of patients CSS/non-CSS	% correctly classified within subset	% of total non-CSS patients in subset
5. Asthma and eosinophilia >10%	18/2	90	90	1. No asthma or eosinophilia >10%	0/735	100	93.4
3. No asthma, eosinophilia >10%, history of allergy	1/4	20	5	2. No asthma, but eosinophilia >10%, no history of allergy	0/19	100	2.4
				4. Asthma without eosinophilia >10%	1/27	96.4	3.4

* The subset numbers also appear below the subset boxes in Figure 1. See Table 3 for definitions of criteria.

vasculitis syndromes. Nevertheless, skin involvement is an important feature of CSS (1,5) and is often the site of a positive biopsy (8). In this series, 6 CSS cases had a skin biopsy; 5 of them showed eosinophilic infiltration in extravascular locations. Other positive biopsies included 2 of 3 lung samples, 2 of 3 muscle samples, and 1 each of pericardium and temporal artery (case 301) specimens. None of the 3 nerve biopsies was reported as showing changes positive for the histologic criterion.

In the era before corticosteroids, CSS patients were usually younger and had a more severe, rapidly progressing course (1,5); these features contrast with those found in the present series. These 20 patients had an older mean age at onset of CSS (50 years), and none died of the CSS. Nevertheless, impressive similarities between the original series (1) and this series are seen in the frequency of positive criteria variables among cases with defined values: asthma (100% in both), eosinophilia (100% versus 95%), peripheral neuropathy (69% versus 75%), migratory or transitory pulmonary infiltrates of the Löffler's pneumonia type (38% versus 40%), and paranasal sinus abnormalities (77% versus 86%), respectively.

In regard to the histopathologic findings, granuloma formation was infrequent in this criteria series, unlike that in the original report (1). Granulomas were found in relation to arteries, arterioles, or venules (i.e., in peri-, intra-, or extramural locations) in 10–20% of CSS cases, and none were noted in relation to veins. A low occurrence of granulomas was also found in the series reported by Lanham et al (5) and may have been partly due to limited pathologic material from the biopsy sampling. Also, earlier, less severe disease and frequent use of glucocorticoids may be other reasons for the low frequency of granulomas in recent series. Although, the "allergic granuloma" lesion described by Churg and Strauss (1) is not included among the proposed classification criteria, it may nevertheless raise suspicion of Churg-Strauss syndrome, especially if no other defined clinicopathologic process can be inferred (6–8).

In the report by Churg and Strauss (1), granulomatous nodules were found most commonly in the epicardium, and 5 of their 13 patients had heart failure. In this criteria study, the primary indicator of heart involvement was the radiographic finding of pericardial effusion in 5 (25%) of the 20 CSS cases submitted versus only 2% in the controls ($P < 0.001$). However, this variable was not selected for inclusion in the criteria set because of its relatively low sensitivity. Unlike PAN or Wegener's granulomatosis, renal involvement is mild in CSS, at least in more recent experience (3,5), and it rarely progresses to renal failure.

Both classification methods provided excellent discrimination in this study, the classification tree being the more sensitive and the traditional format the more specific. In either set, the numbers of true positives identified by these criteria far outnumbered the false positives: 19:6 with the classification tree and 17:2 with the 6-criteria set.

Utilizing the traditional format rule, 2 CSS patients satisfied only 2, and 1 patient only 3, of the 6 proposed criteria. One of these patients (case 301) satisfied 2 criteria and was the only "case" misclassified by the tree format. This patient, a 71-year-old white man, appears to have little evidence to support the diagnosis of CSS. His illness started at age 63 and included fever, asthma attacks, and bilateral pulmonary infiltrates and pulmonary nodules. Lung biopsy showed necrotizing granulomas and perivascular inflammation without eosinophils. Seven cultures grew atypical mycobacteria. The patient responded to isoniazid and ethambutol therapy. The asthma remitted without corticosteroid therapy. At age 71, the patient developed spermatic cord tenderness and swelling, a temperature of 38.5°C, and hepatomegaly. A WBC count showed leukocytosis and 3% eosinophils. Temporal artery biopsy showed granulomatous inflammation and eosinophils in vessel wall and extravascular locations. Followup of this patient revealed no further findings of vasculitis or eosinophilia. If this patient is excluded as a case of CSS, then he would be a control, and only 2 of the CSS patients submitted (10.5%) would be misclassified by the traditional format rule and none by the classification tree format.

The other patient submitted as a case of CSS who satisfied only the 2 criteria of asthma and eosinophilia was a 54-year-old white man with a history of nasal polyps and asthma for at least 5 years. He developed recurrent arthritis, rash, worsening of the asthma, bilateral "fluffy" pulmonary infiltrates (coded as "fixed"), and eosinophilia (17% of 9,100 WBC/mm^3). Over a 9-month period, with tapering doses of prednisone, the patient had several bouts of recurrent symptoms. Lung biopsy showed eosinophils cuffing the venules, but no eosinophils were recorded in extravascular locations. Paranasal sinus radiographs were not obtained. The traditional format rule may not have been satisfied for reasons of a difference in definition of his pulmonary infiltrates, lack of paranasal sinus radiographs, and limited biopsy findings of eosinophils only cuffing venules, after prednisone therapy.

The remaining CSS patient misclassified by the

traditional format rule was a 57-year-old white man who satisfied only 3 criteria: asthma, eosinophilia (56% of 8,800 WBC/mm^3), and extravascular eosinophils on pleural biopsy. He had a history of nasal polyps, but paranasal sinus radiographs were not obtained. Fixed pulmonary infiltrates were recorded.

Utilizing the traditional format rule, only 2 of the control vasculitis patients submitted were misclassified, and both of them had Wegener's granulomatosis. Interestingly, neither had asthma. Both had sinus abnormalities, eosinophilia >10% on peripheral smear, and extravascular eosinophils on biopsy. One patient satisfied the criteria for pulmonary infiltrates and the other for neurologic abnormalities proposed for CSS. One of these 2 control patients had allergies other than asthma and, thus, was also misclassified by the tree format (subgroup 3, Figure 1).

Utilizing the classification tree, 6 control patients were misclassified (Figure 1 and Table 4). Two misclassifications occurred in subgroup 5 (Figure 1), characterized by patients having asthma and eosinophilia, and 4 were in subgroup 3 (Figure 1), characterized by patients having eosinophilia and other allergy. In the former subgroup, 1 misclassified patient had vasculitis of unspecified type, and the other had Henoch-Schönlein purpura. In the latter subgroup, 2 patients had polyarteritis nodosa, 1 had Wegener's granulomatosis (described above as also misclassified by the traditional format rule), and 1 had vasculitis of unspecified type.

This multicenter criteria study was not designed to test the nosologic issue of whether CSS is a "separate entity" (1) versus an "overlap syndrome" among various other systemic vasculitis conditions (3–5,7). Neither are the criteria intended for clinical diagnosis of the individual patient with systemic vasculitis (10) or hypereosinophilic syndromes (4,13). These criteria define classification boundaries for CSS derived from current perspectives of this systemic vasculitis syndrome, as contributed by knowledgeable rheumatologists at multiple rheumatology centers. Although classification is imperfect in the absence of knowledge of etiology and pathogenesis, Lie (7,11) considered Churg-Strauss vasculitis to be a valid addition to the 5 varieties of pulmonary angiitis and granulomatosis that were previously defined by Liebow (12): classic Wegener's granulomatosis, limited forms of Wegener's granulomatosis, lymphomatoid granulomatosis, necrotizing sarcoid angiitis, and bronchocentric granulomatosis.

One may expect that criteria for CSS will change with future research addressed toward its etiology and pathogenesis. Such was not the purpose of this study. However, it is interesting that all of the 5 IgE values obtained in CSS cases in this study were elevated, with a range of 184–2,200 units/ml. The possibility that hyperergic mechanisms contribute to this syndrome and to elevated IgE levels in the active, vasculitic phase has been suggested previously (5).

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