THE AMERICAN COLLEGE OF RHEUMATOLOGY 1990 CRITERIA FOR THE CLASSIFICATION OF HENOCH-SCHÖNLEIN PURPURA

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Criteria for identifying Henoch-Schönlein Purpura (HSP) and distinguishing HSP from other forms of systemic arteritis were developed by comparing the manifestations in 85 patients who had HSP with those of 722 control patients with other forms of vasculitis. By the *traditional format* of choosing different combinations of candidate criteria and comparing the combinations for their ability to separate HSP cases from controls, 4 criteria were identified: age ≤20 years at disease onset, palpable purpura, acute abdominal pain, and biopsy showing granulocytes in the walls of small arterioles or venules. The presence of any 2 or more of these criteria distinguish HSP from other forms of vasculitis with a sensitivity of 87.1% and a specificity of 87.7%. The criteria selected by a *classification tree* method were similar: palpable purpura, age ≤20 years at disease onset, biopsy showing granulocytes around arterioles or venules, and gastrointestinal bleeding. These were able to distinguish HSP from other forms of vasculitis with a sensitivity of 89.4% and a specificity of 88.1%.

The syndrome of acute purpura and arthritis in children was first described by Schönlein in 1837 (1). The manifestations of colicky abdominal pain and of nephritis were added by Henoch in 1874 (2). Although the term "Schönlein-Henoch purpura" is more appropriate historically, "Henoch-Schönlein purpura" (HSP) is used commonly by rheumatologists in the United States and will be used in this report. The disease occurs predominantly in children between the ages of 2 and 10; however, there are many well-documented cases of what appears to be the same syndrome in adults (3,4).

In the early 1950s, Zeek published several papers describing an acute form of systemic arteritis in adults, characterized by skin purpura, arthritis, acute abdominal signs, and glomerulonephritis (5). Many of those cases occurred in close temporal relation to sulfonamide therapy or the administration of heterologous antisera. Although it was Zeek's purpose to distinguish what she called "hypersensitivity angiitis" from microscopic periarteritis nodosa, it is apparent that hypersensitivity angiitis bears a close resemblance to HSP. In fact, there is no published evidence of a consistent attempt to distinguish between the two.

Henoch-Schönlein purpura in children has a seasonal incidence that peaks in winter. Although it has been linked in numerous reports to a preceding

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infection, particularly streptoccal pharyngitis (6), there is no strong evidence to support such an association, and the etiology of HSP remains unknown (7). In contrast to hypersensitivity vasculitis, drug reactions do not appear to be involved. The incidence of HSP in the 2–14-year-old population is about 14 cases per 100,000 per year (8).

Fever and palpable purpura, predominantly on the extremities and buttocks, are usually the first signs of HSP. Early skin lesions may appear urticarial (9). Arthralgia and abdominal pain usually accompany the rash, but these manifestations may precede or follow the rash. Melena is common, and signs of peritonitis are often present. Although rare, complications that require surgery, such as intussusception, demand close observation of all patients (9,10). The arthritis is characteristically oligoarticular, affects large joints, and is transient, with pain out of proportion to objective evidence of synovitis. Proteinuria and hematuria of variable severity are found in about 40% of cases. The renal pathologic findings present a spectrum, from mild focal glomerulitis to necrotizing or proliferative glomerulonephritis (11). The renal disease is usually milder in children and almost always heals. In 10–20% of cases in older children or adults, the nephritis can progress despite the resolution of other disease manifestations (6,12).

The pathology of HSP is that of an acute vasculitis of arterioles and venules in the superficial dermis and the bowel (13). Immunofluorescence staining of tissues usually reveals the presence of IgA in the walls of the arterioles and in the renal glomeruli (14). The serum IgA level is frequently higher than normal. In some adult patients, IgG-containing immune complexes are present, and their deposition in the renal glomeruli may be an important factor in the prognosis of the nephritis (15). However, the more widespread deposition of IgA and properdin in tissues, together with normal levels of the second and fourth components of serum complement, suggest that activation of the alternative pathway of complement activation is the predominant pathogenic mechanism. The renal pathology of HSP closely resembles that of Berger's nephritis, and some investigators have proposed that the two diseases are related (16).

A full recovery is typical for HSP patients, at least in children. Several relapses of purpura, abdominal pain, and arthritis may occur over 3–6 weeks before there is a complete resolution of the disease.

Clinical data for 85 patients who were diagnosed as having HSP by the physicians who submitted cases for the vasculitis criteria study were analyzed. The clinical and laboratory features that best identified the 85 cases of HSP were compared with those of 722 patients diagnosed as having other forms of vasculitis. From that analysis, classification criteria that best separated HSP from the other vasculitis syndromes were identified.

The data on all of the 85 patients who were entered into the study with a diagnosis of HSP were reviewed before being analyzed by a subcommittee to ensure that the diagnosis was based on sufficient data. This subcommittee included physicians who regularly practice pediatric rheumatology.

For a description of the methods of patient selection and evaluation, see the articles by Hunder et al (17) and Bloch et al (18), which appear elsewhere in this issue of *Arthritis and Rheumatism*.

RESULTS

Data completion for the 85 cases of HSP entered into the study was excellent for all clinical features, but was variable for some of the laboratory tests that are not done routinely.

The HSP patients included 46 males and 39 females. The mean age of the patients was 17.4 years; 71% of the patients were younger than age 20, and 63.5% were younger than age 16.

The variables that were chosen as potential discriminators for HSP, by means of univariate analyses of all items for which data were collected, are shown in Table 1. The number of HSP cases or controls (the number of subjects for whom the particular variable was recorded), the sensitivity (the proportion of HSP cases in which the variable was positive), and the specificity (the proportion of controls in which the variable was negative) are also shown in Table 1.

The presence of palpable purpura, defined as a slightly elevated purpuric rash, especially on the extremities and buttocks, had the highest sensitivity. Although the specificity for palpable purpura was slightly less than that of several other variables, such as abdominal angina or hematochezia, the latter variables were less sensitive.

Abdominal manifestations included diffuse abdominal pains, often worse after meals, which was referred to as "bowel angina," and "bowel ischemia," which was defined by the presence of bloody diarrhea. Abdominal pain was recorded in more than a third of the cases, but gross gastrointestinal bleeding was

Table 1. Comparison of the sensitivity and specificity of potential criteria variables for Henoch-Schönlein purpura*

Criterion	No. of patients (n = 85)	No. of controls (n = 722)	Sensitivity (%)	Specificity (%)
History				
1. Age ≤20 at disease onset†‡§	85	722	70.6	90.7
2. Abdominal angina	83	718	37.3	94.0
3. Bowel ischemia	84	716	16.7	95.8
4. Bowel angina (variables 2 or 3)†‡	83	717	51.8	91.6
Physical				
5. Palpable purpura†‡§	85	718	88.2	79.9
6. Monarticular synovitis	85	716	11.8	94.8
7. Oligoarticular synovitis	84	716	39.3	86.7
8. Synovitis (variables 6 or 7)†	84	717	50.0	82.6
Laboratory				
9. Gross hematuria	85	720	17.6	95.3
10. Microhematuria ≥5 RBC/HPF	81	696	38.3	79.2
11. Hematuria (gross hematuria or microhematuria ≥1 RBC/HPF)†	82	695	54.9	59.7
12. Melena	85	718	15.3	94.4
13. Hematochezia	85	716	23.5	94.8
14. Positive stool guaiac	54	473	53.7	82.9
15. GI bleeding (variables 12 or 13 or 14)†	61	477	67.2	78.6
16. Proteinuria	80	681	43.8	70.5
17. Decreased C3 level	46	298	10.9	83.2
Biopsy				
18. Granulocytes in arteriole wall	33	460	36.4	85.4
19. Granulocytes in venule wall	37	431	54.1	83.3
20. Wall granulocytes (variables 18 or 19)†‡	38	428	63.2	75.0
21. Periarteriolar granulocytes	32	459	40.6	87.6
22. Perivenular granulocytes	36	432	58.3	85.2
23. Extraarteriolar granulocytes	33	456	27.3	92.5
24. Extravenular granulocytes	37	432	35.1	89.8
25. Extravascular/perivascular granulocytes (variables 21 or 22 or 23 or 24)†‡	37	430	73.0	74.9

* 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. RBC = red blood cells; HPF = high power field; GI = gastrointestinal.
† Criterion is one of the final "short list" of variables (n = 8) (see text).
‡ Criterion is used for the traditional format classification.
§ Criterion is used for the tree classification.

infrequently reported. However, both of these findings were highly specific for the diagnosis of HSP.

Oligoarticular synovitis, defined as involvement of 4 or fewer joints, was present in 39% of the patients with HSP and in only 13% of the patients with other kinds of vasculitis. Polyarthritis or synovitis that involved more than 4 joints occurred in only 24% of the HSP patients and was less disease-specific.

The presence of nephritis in HSP was documented by proteinuria or microhematuria. Proteinuria alone was not sufficiently specific, since that finding was present in almost one-third of the control patients with all other forms of vasculitis. Hematuria, gross or microscopic, although more sensitive (54.9%) than proteinuria, was not very specific at a minimum level of only 1 red blood cell per high power field (59.7%). Red blood cell casts were reported in only 7% of the patients with HSP.

Table 2. 1990 criteria for the classification of Henoch-Schönlein purpura (traditional format)*

Criterion	Definition
1. Palpable purpura	Slightly raised "palpable" hemorrhagic skin lesions, not related to thrombocytopenia
2. Age ≤20 at disease onset	Patient 20 years or younger at onset of first symptoms
3. Bowel angina	Diffuse abdominal pain, worse after meals, or the diagnosis of bowel ischemia, usually including bloody diarrhea
4. Wall granulocytes on biopsy	Histologic changes showing granulocytes in the walls of arterioles or venules

* For purposes of classification, a patient shall be said to have Henoch-Schönlein 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 a specificity of 87.7%.

The biopsy findings were combined to form several variables that included the histologic changes in both arterioles and venules. One of these was defined as the presence of granulocytes in the walls of vessels; the other was defined as the distribution of granulocytes in a perivascular or extravascular location. In both cases, venules were involved more commonly than arterioles (35–50% versus 27–41%). The presence of granulocytes in the vessel wall or in a perivascular location were retained as separate variables. A combined "either/or" variable improved sensitivity slightly, but reduced specificity. Other cell types, mononuclear cells or eosinophils, were prominent in less than 14% of HSP patients. No larger vessel involvement was reported in this series of patients with HSP.

Diastolic hypertension and depressed total serum complement levels were found in only 17% and 19% of the cases, respectively. The serum C3 level was measured in only 46 HSP patients, and it was lower than normal in 5 of them. Circulating immune complexes were sought in 12 patients and were found in 3. Serum IgA was measured in 36 patients, but the levels were found to be above the age-related range of normal in only 5.6%. The erythrocyte sedimentation rate exceeded 50 mm/hour in only 16 patients; 5 had a serum creatinine level over 1.5 mg/dl.

Traditional format classification. Eight of the variables shown in Table 1 were selected for a "short list," based on their potential to provide the best discrimination of HSP from other kinds of vasculitis. To increase their discriminatory value, some closely related criteria were combined, as indicated in Table 1.

Twenty-four combinations of these 8 criteria were tested to determine which of them best separated cases of HSP from controls, using a traditional format rule (17). The final criteria selected by that method are shown in Table 1 and are defined in Table 2. The presence of any 2 or more of these 4 criteria correctly classified 74 of the 85 cases of HSP while correctly excluding all but 89 of the 722 control patients. The sensitivity and specificity of 2 or more criteria are 87.1% and 87.7%, respectively. Only 4 patients with HSP had 4 of these selected criteria, and only 1 patient with HSP had all 8.

Of the 89 control patients with other forms of vasculitis who were misclassified as having HSP because of the presence of at least 2 of these criteria, the entry diagnoses were hypersensitivity vasculitis in 40, polyarteritis nodosa in 12, Wegener's granulomatosis in 9, and unspecified vasculitis in 10. Eighty-four percent of the misclassified control patients had palpable purpura, and 84% met the histologic criteria. Of the misclassified controls, only 18% were younger than age 20.

Tree classification. Figure 1 shows the distribution of HSP patients and the control patients with other vasculitis when the criteria are applied in a tree format. The criteria used in the tree classification are identified in Table 1 and are defined in Table 3 (classifying subsets are numbered for reference). When information for a given variable was not available, a "surrogate variable" was used to classify subjects (see ref. 18).

The best first split is the presence or absence of palpable purpura, which identified 75 of the 85 HSP patients, but includes 144 of the 722 control patients with other forms of vasculitis (Figure 1 and Table 4). Applying the age criterion (age ≤20 years at disease onset) to the subset with palpable purpura correctly classified 51 cases in this subset and misclassified 10 controls. Three of these misclassified control patients were originally diagnosed as having hypersensitivity vasculitis and 3 as having Wegener's granulomatosis. Twenty-four of the HSP patients with palpable purpura were over age 20 at disease onset. Eighteen of these patients were correctly classified as having HSP according to the histologic criterion or the criterion of gastrointestinal bleeding if the histologic findings were not specified. Of the 65 control patients who were also in this subset (subset 6), 32 had hypersensitivity vasculitis, 8 had polyarteritis nodosa, and 7 had Weg-

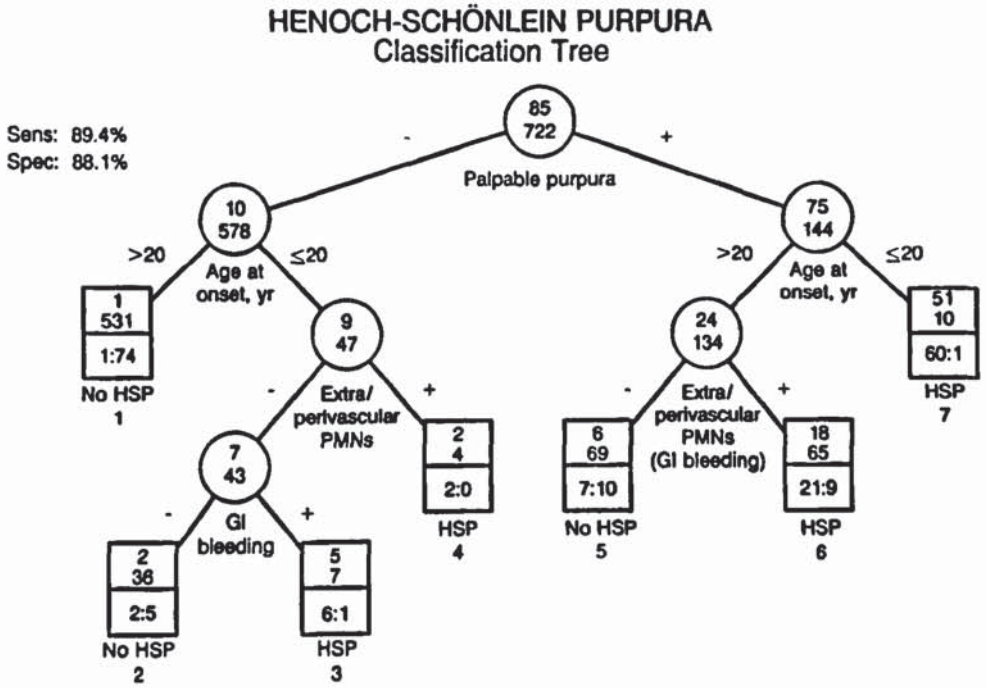

Figure 1. Classification tree for Henoch-Schönlein purpura (HSP). The circles and boxes contain the number of subjects with HSP (top number) and the number of control patients with other forms of vasculitis (bottom number). The bottom half of the boxes shows the percentage of subjects with HSP (out of all HSP cases) (left number) and the percentage of controls (out of all controls) (right number). Boxes specify whether subjects are classified as having HSP or not having HSP (No HSP); the numbers under these specifications are the subset numbers (see Table 3 for definitions of criteria and Table 4 for explanations of subsets). Parentheses indicate the surrogate variable "gastrointestinal (GI) bleeding" to be used when "extravascular or perivascular polymorphonuclear neutrophils (PMNs)" is not defined by biopsy.

ener's granulomatosis. It should be noted that the histologic criterion selected by the tree analysis (granulocytes in an extravascular and perivascular location) is different from that selected by the traditional format rule (Tables 2 and 3). Also, gastrointestinal bleeding emerged in the tree classification method, whereas bowel angina was used in the traditional format, possibly because of the greater sensitivity of gastrointestinal bleeding.

Of the 10 cases submitted as HSP without purpura (Figure 1, left side), 9 were correctly classified according to the criterion of age at disease onset, but 47 controls were also included. That subset was further subdivided into 3 subsets (numbers 2, 3, and 4) by the successive application of the histologic criterion and the gastrointestinal bleeding criterion, resulting in the correct classification of 36 of the controls. The only HSP patient who did not have purpura and was over age 20 at disease onset had gastrointestinal bleeding and hematuria and would have been correctly classified according to the histologic criteria. A detailed description of each subset is given in Table 4.

Table 3. Criteria and definitions used for the classification of Henoch-Schönlein purpura (tree format)*

Criterion	Definition
1. Palpable purpura	Slightly raised, "palpable" hemorrhagic skin lesions; not related to thrombocytopenia
2. Age ≤20 at disease onset	Patient 20 years or younger at onset of first symptoms
3. Gastrointestinal bleeding	The passage of melena, grossly bloody stool, or a positive result for occult blood in stool (usually by the guaiac method)
4. Extravascular or perivascular granulocytes on biopsy	Histologic changes showing granulocytes in a perivascular cuff around arterioles or venules, or in an extravascular location

Table 4. 1990 classification tree for Henoch-Schönlein purpura (HSP)*

HSP subsets	No. of patients HSP/non-HSP	% correctly classified	% HSP patients in subset	Non-HSP subsets	No. of patients HSP/non-HSP	% correctly classified	% non-HSP patients in subset
7. Palpable purpura; age at disease onset ≤20	51/10	84	60	1. No palpable purpura; age >20 at disease onset	1/531	100	74
6. Palpable purpura; age at disease onset >20; biopsy showing granulocytes around small blood vessels	18/65	22	21	2. No palpable purpura; age ≤20 at disease onset; biopsy negative for granulocytes around small blood vessels; no GI bleeding	2/36	95	5
4. No palpable purpura; age ≤20 at disease onset; biopsy showing granulocytes around small blood vessels	2/4	33	2	5. Palpable purpura; age >20 at disease onset; negative biopsy	6/69	92	10
3. No palpable purpura; age ≤20 at disease onset; negative biopsy, but GI bleeding	5/7	42	6				

* The subset numbers also appear below the subset boxes in Figure 1. The classification tree yields a sensitivity of 89.4% and a specificity of 88.1%. See Table 3 for definitions of criteria. GI = gastrointestinal.

By the successive application of these 4 criteria, the tree correctly classified 89.4% of the 85 cases of HSP while excluding all but 11.9% of the non-HSP patients (86 patients). The tree classification is slightly more sensitive than the traditional format selection method, and it excludes an additional 3 non-HSP cases.

DISCUSSION

Although the bedside diagnosis of HSP is easily made in most cases, no diagnostic criteria have previously been established by comparing the clinical manifestations of HSP with those of other forms of vasculitis. The classification criteria that were identified by 2 methods in this study identify HSP and separate it from other vasculitis syndromes with a high degree of accuracy.

The disease manifestation that combined the best sensitivity and specificity was palpable purpura, which emerged as the primary criterion in the classification tree and as 1 of the criteria in all of the best criteria combinations identified by the traditional format rule. Only 10 of the 85 patients diagnosed as having HSP did not have palpable purpura. Although many of the purpuric lesions may not be palpable in some patients who have HSP, that feature helps to distinguish vasculitic purpura from simple purpura, which is much less disease-specific.

It should be pointed out that these criteria may not distinguish HSP from such disorders as infectious purpuras or allergic reactions, which were not included in the control population. For example, the presence of only 2 criteria, such as palpable purpura and the vascular histology, could occur frequently in a number of different diseases.

Age at disease onset was identified as the second most important criterion. HSP is predominantly a disease of children, but what seems to be the same disorder clearly occurs in adults as well. A number of age splits were tested for their ability to separate HSP from other forms of vasculitis. Onset at age 20 or less emerged as the most accurate. Even so, 25 of the HSP cases, or almost 30%, were over age 20 at disease onset. All but 1 of these patients had palpable purpura, and 18 of them met the histologic criteria. Although a biopsy is rarely necessary to diagnose HSP in children, it may be helpful in adults. In fact, the biopsy findings were able to exclude more than 50% of the non-HSP patients with palpable purpura who were over age 20 at onset.

Of the patients with other vasculitis syndromes who might be diagnosed as having HSP by meeting the biopsy criteria (subset 6, Figure 1), almost 50% were

patients who had hypersensitivity vasculitis (HSV). The clinical similarity of HSP and HSV is reflected in the fact that 3 of the 8 candidate criteria were the same for both diseases (see ref. 19). Apart from age at disease onset, only the gastrointestinal manifestations, gastrointestinal bleeding or bowel angina, in patients with HSP and the taking of medication at disease onset in patients with HSV distinguished the 2 diseases. The age criterion is problematic, since it is not a disease manifestation, and it is possible that some patients with palpable purpura were originally submitted as cases of HSP or HSV simply on the basis of age at onset.

The 4 final criteria selected by the traditional format method identified only 1 criterion common to HSP and HSV: palpable purpura. Different histologic criteria were chosen, though at first glance, they appear to be similar. Extravascular and perivascular granulocytes, chosen as a criterion for HSV was less sensitive and specific for HSV than was the presence of granulocytes in vessel walls. In fact, the former is a better criterion for HSP, as demonstrated by its selection by the tree classification method.

It must be reemphasized with respect to the previous discussion that the criteria identified for HSP and HSV were developed by comparing HSP and HSV individually with all other forms of vasculitis, rather than specifically comparing the two. That objective explains what, at first, might seem to be arbitrary differences between some HSP and HSV criteria.

It can be argued that HSP is predominantly a pediatric disease and that criteria should be developed by analyzing only childhood vasculitis. That contention does not acknowledge the incidence of HSP in adults (3). Other forms of vasculitis are much less common in the pediatric age group, and gathering a statistically comparable group of children with other vasculitis syndromes would be difficult.

Two features of the original descriptions of the disease in the literature, arthritis and nephritis, failed to be included in either classification scheme, although both were in the final list of candidate criteria that were considered. Hematuria, as a sign of nephritis, lacked both specificity and sensitivity. The presence of synovitis, although more specific than palpable purpura, lacked sensitivity. Arthritis defined only as joint pain was very nonspecific.

In summary, by each of 2 selection methods, 4 criteria were indentified that distinguish HSP from other forms of vasculitis. By a classification tree method, palpable purpura and age $\leq$20 years at disease onset appear to be the best discriminators. The 2 additional criteria, which, by the tree format, identify HSP in patients over age 20 or in the few who do not have purpura, are the presence of granulocytes around small blood vessels and gastrointestinal bleeding.

The distinction between HSP and HSV in clinical and pathophysiologic terms is unclear. The resolution of that problem requires a more complete understanding of the pathogenesis of systemic vasculitis.

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