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A QUANTITATIVE APPRAISAL OF SYSTEMIC LUPUS ERYTHEMATOSIS: DIAGNOSIS BY POINT SYSTEM

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As experience with systemic lupus erythematosis accumulates the need for a more objective definition of this protean disease becomes more essential. Unfortunately the histopathology of SLE is not clear-cut enough to serve as a referee to clinical diagnosis and the study of the natural history of the disease. This lack of definition of the disease has greatly hindered establishment of the range of sensitivity and specificity of the LE phenomenon and the newer tests for anti-nuclear anti-body. Indeed, how can a confirmatory test be assigned a range of sensitivity or specificity for a disease lacking concrete definition?

The value of the Jones Criterion for acute rheumatic fever may be cited in retrospect as an example of the value of an empirical, generally accepted, and widely used definition of a disease in the further development of an understanding of its natural history. Likewise, the definition of rheumatoid arthritis by the American Rheumatism Association has served the purpose of allowing Rheumatologists to communicate with clarity and has crowded out the "fellow-travelers" which clouded understanding of the natural history of rheumatoid arthritis and evaluation of various modes of therapy.

The fact that there is a core of characteristic clinical features and some confirmatory laboratory tests would seem to qualify SLE as a single disease entity. If this is so, the question of a concrete definition of SLE is principally one of finding the "outer boundaries." The literature bears eloquent testimony to the wide variation between individual cases and thus has defined the "within variance." The problem remaining is the definition of "between variance," or the objective effort to separate all cases of SLE from any case of another disease.

Method

The clinical and laboratory manifestations of SLE in 143 patients were collected and divided into groups depending on the frequency of that finding in other diseases. The less frequent a sign, symptom or laboratory finding occurred in other diseases,

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the more weight it was assigned insofar as its value in predicting the presence of SLE. A numerical value was then assigned to each group and a summation of these values for each of the 143 patients was tabulated. It was found that a total of 20 or more points was needed to make the diagnosis of SLE. A provisional or probable diagnosis could be made with 14 or more points. The validity of these values were then checked by determining the total points in a group of 15 patients with an autopsy diagnosis of SLE but who had consistently negative LE preparations during life. A further validation was the application of the point system to the cases described in Harvey's classical paper on SLE. Finally, the points were summarized for patients who have shown a positive LE preparation, but who by autopsy or other concrete evidence were shown to have a disease other than SLE.

10 points:	Positive LE preparation (5 or more cells in 500 counted).
	"Butterfly" rash.
	"Wire loop" or other definitive finding on renal biopsy.*
8 points:	Renal biopsy showing glomerulitis and/or basement membrane thickening.
	Arthritis plus sun-sensitivity or allergic diathesis.
5 points:	Arthralgia.
	Fibromyositis, severe.
	History of acute migratory polyarthritis.
3 points:	Myocarditis and/or pericarditis.
	Pleurisy and/or pneumonitis.
	Raynaud's phenomenon.
	Hemolytic anemia.
	Thrombocytopenic purpura.
	Psychosis and/or convulsions.
	False positive serology.
	Positive LE cell test (1-4 cells in 500 counted).
	Skin eruption compatible with LE (angioneurotic edema, erythema nodosum, erythema multiforme, vesicular dermatitis).
1 point:	Leucopenia.
	Abnormal urine sediment or albuminuria.
	Positive liver "flocculation" tests and/or hyperglobulinemia and/or cryoglobulinemia.
	Jaundice.
	Edema.
	Sun sensitivity.

*Do not use other renal manifestations.

½ point:	Elevated sedimentation rate.
	Anemia.
	Lymphadenopathy, hepatosplenomegaly.
	Thrombophlebitis.
	Conjunctivitis, iritis.
	Diarrhea, abdominal crisis.
— 5 points:	Patient with "certain" rheumatoid arthritis by ARA standards.
	Age 10 and under.

EXCEPTION: Lymph node or bone marrow biopsy revealing another disease known to give false positive LE phenomenon.

The point system has now been in clinical use at this hospital since 1960 and has proved useful in evaluating the patient with a false positive LE preparation; in patients with nonclassical manifestations of lupus erythematosus; and patients with 1 or 2 classical manifestations but lacking sufficient other proof of diagnosis; and in evaluating the patient with a suspected false positive LE cell phenomena. It is felt that it may have future value in determining prognosis, or in grading activity particularly in evaluation of efficacy of therapy. In this regard, although the empirically chosen point values are based on the diagnostic significance of the manifestation, a rough parallel between the total sum of points and prognosis has been noted.