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THE CLASSIFICATION OF LEPROSY WITH A PRIMARY DIVISION INTO "BENIGN" AND "MALIGN" CLASSES [1]

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The question of the classification of leprosy quite justifiably preoccupies a great many leprologists. Leprosy control is extending year by year throughout the world, and the number of qualified specialists—already relatively low—is becoming increasingly inadequate. For this reason, effective leprosy control in most regions in which the disease is endemic is possible only through the properly organized cooperation of the whole medical profession. The establishment of a leprosy classification has, therefore, become an urgent problem to which a solution must be found without delay.

The classifications used up to the present are all easily understood by leprologists, but their diversity generally makes classification of cases difficult and often impossible for the majority of nonspecialized physicians. These physicians now insistently demand the adoption of a uniform classification which will be simple and universally acceptable, and based principally on the clinical signs of the disease.

The use of a classification based on histopathological examinations cannot be generalized in the underdeveloped countries. Furthermore, the interpretation of histological findings, even by competent laboratories, is often far from uniform. On the other hand, the correct classification of cases is almost always possible on the basis of clinical symptoms with the aid of the bacteriological examination.

[1] This article was prepared as a working document for the WHO Expert Committee on Leprosy which met in Brazil in November 1952.

We have no intention of presenting a personal classification. We have merely endeavored to bring together the most useful data provided by the two best-thought-out classifications, i.e., those proposed by the Cairo and Havana congresses.

We have therefore divided leprosy into two types: benign leprosy and malign leprosy, the former including the tuberculoid and indeterminate forms of the disease. As will be seen from the scheme given hereunder, the cutaneous lesions of the tuberculoid and indeterminate forms have so many elements in common that we feel justified in grouping them together and indicating them as benign, in contrast to the malign lepromatous forms. In our experience, this division of leprosy into two types has great advantages from the didactic point of view. It also enables an inexperienced physician to classify his patients correctly in a simple and rapid manner.

Certain authors assert that the indeterminate form cannot be classified as benign since some of the cases may develop into the malign form. This argument, however, does not seem to us to be valid, since the tuberculoid form may also develop into the malign form. On the other hand, some indeterminate cases are stable, remaining as such indefinitely, while others change to the tuberculoid form. Moreover, if the existence of a malign type of leprosy is admitted, all the other forms which cannot be classified as malign should, logically, be included in the benign type.

In this scheme of classification we have endeavored to use only terms which have the same roots in most modern languages. The meaning of the symbols used in the abbreviated nomenclature can thus be easily understood in English, Spanish, Portuguese, Dutch, Italian, German and French.

The generally accepted use of the term "macule" to define certain cutaneous lesions in leprosy is not satisfactory. The word "macule" in dermatology refers particularly to marks which are discolored as compared with the general color of the skin but are not elevated above the surface and show no change in texture, whereas from the beginning the prominent cutaneous deformations of leprosy, particularly those of tuberculoid leprosy, have been called "macules" by the great majority of leprologists. However, we consider that the use of this term, which has been adopted for lack of a more precise word by eminent dermatologists such as Jeanselme and Klingmüller, should be retained in leprosy terminology.

The following is the classification scheme proposed by us:

I. PRIMARY CLASSIFICATION

BENIGN LEPROSY	MALIGN LEPROSY
In general, the initial form of leprosy.	In general, secondary to the benign form.
Characterized by cutaneous lesions called leprids (macules, papules, very rarely nodules); or by neural acroteric lesions (degeneration of peripheral sensibility, polyneuritis, trophic disorders, paralysis, mutilations); or by an association of cutaneous and neural lesions.	Characterized by cutaneous lesions called lepromas (macules, papules, nodules, infiltrations); which are nearly always associated with neural acroteric lesions, sometimes clinically little evident (degeneration of peripheral sensibility, polyneuritis, trophic disorders, paralysis, mutilations).
Cutaneous lesions usually clearly delimited. Marked tendency to peripheral extension, to coalescence, and to resolution from the center. Often asymmetrical arrangement.	Cutaneous lesions not clearly delimited, particularly when smooth or only slightly infiltrated. No tendency to peripheral extension or to resolution from the center. Often asymmetrical arrangement.
More or less pronounced disorders of the superficial sensibility always associated with the lesions.	Modification of superficial sensibility inconstant and sometimes not even associated with the lesions.
Negligible lesions of the mucous membrance, ocular system proper, and internal organs in slight and mild cases. Such lesions are rare and little evident even in advanced cases.	Lesions of mucous membrane, of the ocular system, and of the internal organs in mild cases fairly rare and often only slight, increasing in incidence and gravity with advancement of the disease.
Bacteriological examination of the nasal mucosa generally negative (91%).[2] Bacilli usually rare and not in "globi."	Bacteriological examination of the nasal mucosa very often positive (73%).[2] Nearly always positive in advanced cases (97%). Bacilli usually numerous and partly in "globi."
Skin lesions bacteriologically negative in more than half of the cases (57%). Bacilli usually rare, except during an active stage or a state of "reaction." With few exceptions, no bacilli in globi.	Skin lesions always bacteriologically positive (100%). Bacilli usually very numerous, partly in globi.

[2] The percentages shown refer to the results of our examination of 1,290 untreated patients.

I. PRIMARY CLASSIFICATION—Continued

BENIGN LEPROSY	MALIGN LEPROSY
Lepromin reaction frequently positive (80%). A negative reaction indicates, in untreated patients, that a malign development is to be feared.	Lepromin reaction always negative. In exceptional cases the reaction may become feebly positive following prolonged treatment.

II. SECONDARY CLASSIFICATION

BENIGN LEPROSY		MALIGN LEPROSY
TUBERCULOID LEPROSY (T)	INDETERMINATE LEPROSY (I)	LEPROMATOUS LEPROSY (L)
Cutaneous lesions (c) erythematous (pink to purplish-red), raised (macules, papules, rarely nodules), frequently soft in consistency.	Cutaneous lesions (c) smooth and flat (macules; hypopigmented, weakly erythematous or hyperpigmented). The appearance of slight elevation of the edge of the lesion indicates probable change to tuberculoid or lepromatous leprosy.	Cutaneous lesions (c) erythematous (salmon pink, copper red, purplish red in rare cases; little tendency to hypopigmentation), more or less infiltrated, frequently very prominent (macules, papules, nodules, infiltrations), often very firm in consistency.
Very frequent and usually very marked thickening of the superficial cutaneous nerves;	No thickening of the superficial cutaneous nerves;	Fairly frequently, very slight thickening of the superficial cutaneous nerves, particularly the cervical nerves which are palpable.
or neural lesions (n), acroteric (pronounced thickening, frequently moniliform, of the nerve trunks, polyneuritis). Tendency to abscess formation, caseation and sclerosis.	or neural lesions (n), acroteric (moderate and uniform thickening of the nerve trunks, polyneuritis). No abscess formation. Sclerosis not pronounced.	The pure acroteric neural form (n), without accompanying cutaneous changes, probably does not exist in initial lepromatous leprosy. It could not be diagnosed, however, except by histological examination.

II. SECONDARY CLASSIFICATION—Continued

BENIGN LEPROSY		MALIGN LEPROSY
TUBERCULOID LEPROSY (T)	INDETERMINATE LEPROSY (I)	LEPROMATOUS LEPROSY (L)
Differential clinical diagnosis often difficult. It may be based on the pronounced moniliform thickening of the nerve trunks and the intense reaction to lepromin.	Differential clinical diagnosis often difficult. It may be based on the moderate and uniform thickening of the nerve trunks and on the weak reaction to lepromin.	Since the pathogenic agent is spread by the blood stream, nerve lesions are almost always associated with skin lesions. However, in recent cases, polyneuritic disturbances are often absent or very slight (moderate thickening of the nerve trunks; slight degree of sclerosis). In cases of long standing, the cutaneous lesions may disappear while the polyneuritic symptoms become more severe (very marked thickening, often fusiform, of the nerve trunks; pronounced tendency to sclerosis). Such residual polyneuritic leprosy may be clinically diagnosed by means of the stigmata left by cutaneous lesions and the negative result of the lepromin test.
Precise classification can be achieved only by histological examination;	Precise classification can be achieved only by histological examination;	Histological examination may be necessary.
or association of cutaneous and neural lesions (cn).	or association of cutaneous and neural lesions (cn).	Practically always association of cutaneous and neural lesions (cn).

II. SECONDARY CLASSIFICATION—Continued

BENIGN LEPROSY		MALIGN LEPROSY
TUBERCULOID LEPROSY (T)	INDETERMINATE LEPROSY (I)	LEPROMATOUS LEPROSY (L)
Histological examination of the skin and nerve lesions show presence of tuberculoid granulomas (epithelioid cells, giant cells of the Langhans type).	Histological examination of the skin and nerve lesions show infiltrations of the mild chronic inflammation type (histio-lymphocytic cells).	Histological examination of the skin and nerve lesions show presence of granulomas consisting of histiocytes (macrophages filled with bacilli, or "lepra cells," Virchow's vacuolate cells containing globular clusters or lysed bacilli).
THREE VARIETIES	NO DEFINITE VARIETIES	NO DEFINITE VARIETIES
(1) Minor Tuberculoid (T′)	Indeterminate Anesthetic (Ia)	Diffuse Lepromatous (Ld)
Moderately raised cutaneous lesions (macules, papules) with an irregular surface which is micropapulate, often of sarcoid type. Pronounced tendency to thickening of the superficial cutaneous nerves.	Rare cases show, at the outset of the disease, an area of cutaneous anesthesia without any apparent change in the skin and without neuritis. These cases may provisionally be classified as indeterminate anesthetic leprosy. The histopathology and, above all, the subsequent course will permit more precise classification.	In rare cases the cutaneous lesions are extensive and diffuse, without localization in macules, nodules, or infiltrations. These cases may be designated as diffuse lepromatous leprosy.
Bacteriological examination rarely positive (nasal mucosa 8%, cutaneous lesions 26%). Bacilli rare except in an active period of the disease. No globi.	Bacteriological examination only infrequently positive (nasal mucosa 3%, cutaneous lesions 34%). Bacilli generally rare and, except in some prelepromatous cases, never in globi.	Bacteriological examination always positive (nasal mucosa 73%, cutaneous lesions 100%). Bacilli usually very numerous, especially in the skin lesions, and partly in globi.
Lepromin reaction very often positive (92%). Reaction generally very intense.	Lepromin reaction frequently positive (87%). Reaction usually weak.	Lepromin reaction always negative.

THREE VARIETIES	NO DEFINITE VARIETIES	NO DEFINITE VARIETIES
(1) Minor Tuberculoid (T')	Indeterminate Anesthetic (Ia)	Diffuse Lepromatous (Ld)
No state of reaction proper.	No reaction state.	Frequent reaction states, often grave (high fever, adynamic state, exacerbation of existing lesions and sudden formation of new ones, tendency to relapse). (In diffuse lepromatous leprosy, the reaction state may lead to the appearance of necrotic erythema (Lucio).)
Spontaneous regression possible (skin lesions flatten, become hypopigmented and then repigmented before disappearing). The residual skin and nerve lesions frequently show the "indeterminate" histology (round cell infiltration).	Spontaneous regression possible (normal pigmentation, then disappearance of the skin lesions.	Tendency to progressive aggravation of the disease.
Relatively stable variety. Nevertheless, may develop into the malign type.	Unstable form. Develops fairly frequently into the malign type (negative lepromin), but may remain indeterminate or sometimes change to tuberculoid (strongly positive lepromin).	Stable form. However, in long-standing cases the skin lesions may disappear while the nerve disturbances increase in intensity. The histology of the skin lesion is then frequently indeterminate (round-cell infiltrations) while the nerve lesions usually retain typical lepromatous aspect.

(2) Major Tuberculoid (T″)	(3) Borderline Tuberculoid (BT)
Very prominent cutaneous lesions (macules, papules, rarely nodules) with uniform surface. Pronounced tendency to thickening of the superficial cutaneous nerves.	Evolutive stage of major tuberculoid, frequently following a reaction state. Cutaneous lesions sometimes very prominent (macules, papules) and soft in consistency. Surface often smooth, shiny and dark red. Arrangement frequently asymmetrical. Slight tendency to thickening of the superficial cutaneous nerves.
Bacteriological examination fairly frequently positive (nasal mucosa 12%, skin lesions 59%). In the skin lesions, bacilli may be frequent during reaction states and in advanced cases. Presence of globi exceptional.	Bacteriological examination always positive (nasal mucosa 38%, skin lesions 100%). Bacilli usually numerous. Rare globi, especially in the skin lesions.
Lepromin reaction very often positive (94%), generally very intense.	Lepromin reaction generally negative (86%), or very feebly positive (14%). Histological examination often necessary for differential diagnosis of this variety (presence of granulomas of both tuberculoid and lepromatous appearance, sometimes in the same lesion).

(2) Major Tuberculoid (T″)	(3) Borderline Tuberculoid (BT)
Fairly frequent appearance of reaction states (fever, sometimes high, exacerbation of existing lesions and sudden formation of new ones. Nodules which may appear are paucibacillary. Subsequently, tendency of the skin lesions to regression). Reactions with the formation of bullous lesions ("lazarine") are rare.	No reaction state proper.
Spontaneous regression possible (process identical to that described under minor tuberculoid).	Regression to the major tuberculoid variety possible. Unstable variety, frequently developing into the lepromatous type.
Relatively stable variety. Nevertheless, may develop into the "borderline" variety and the malign type (lepromin negative).	

III. ABBREVIATED NOMENCLATURE

(1) *Classification of the different forms of leprosy.—*

BENIGN LEPROSY				MALIGN LEPROSY	
TUBERCULOID LEPROSY (T)		INDETERMINATE LEPROSY (I)		LEPROMATOUS LEPROSY (L)	
minor (T′) major (T″) "borderline" (BT)					
Cutaneous (c)		*Cutaneous* (c)		*Cutaneous* (c)	
macular	(m)	macular	(m)	macular	(m)
proliferating [3]	(p)	anesthetic	(a)	proliferating [3]	(p)
"lazarine" (bullous lesions)	(l)			diffuse	(d)
				"Lucio" type	(l)
Neural (n)		*Neural* (n)		*Neural* (n)	
cutaneo-neural	(cn)	cutaneo-neural	(cn)	cutaneo-neural	(cn)

[3] The definition "proliferating" (p) is suggested to designate cases of leprosy showing nodules or infiltrations, so as to avoid the double use of the letters "n" and "i."

(2) *State of the disease.*—Slight case (1); moderate case (2); advanced case (3).[4] (The numbers should be placed before the letters c or n.)

(3) *Degree of activity of the disease.*—Florid (f); quiescent (q); residual (r). (The letters f, q or r should be placed after T, I, or L.)

(4) *Lepra reaction.*—Moderate reaction: underline once or put in italics. Marked reaction: underline twice or put in underlined italics.

(5) *Results of bacteriological examinations.*—Use the signs + or –, firstly for the nasal mucosa and then for skin and possibly nerve lesions.

(6) *Examples.*—Simplified classification: L3++. Advanced lepromatous leprosy. Bacilli present in the nasal mucosa and the skin lesions.

T2–+. Moderate tuberculoid leprosy. Nasal mucosa, negative. Bacilli present in the skin lesions.

I1––. Slight indeterminate leprosy. Bacteriological examination negative.

More detailed classification: T″3cIn–+. Advanced major cutaneoneural tuberculoid leprosy, in a state of moderate reaction. Numerous skin lesions, slight neural lesions (peripheral anesthesia of limited extent without trophic disturbances, or slight atrophy or paresia). Bacteriological examination negative for the nasal mucosa, positive for the skin lesions.

Complete classification: *Lf3c(pm)3n*++. Advanced florid, cutaneoneural lepromatous leprosy in a pronounced state of reaction. Presence of numerous extensive and very marked skin lesions (nodules, infiltrations, and macules) and very considerable neural lesions (extensive peripheral anesthesia, trophic and motor disturbances: paralysis, atrophy, contractures, trophic ulcers and mutilation). Bacteriological examination positive for both the nasal mucosa and the skin lesions.

Lr2n––. Moderate residual neural lepromatous leprosy. No visible skin lesions. Moderate neural lesions: peripheral anesthesia, very extensive on one extremity, less extensive on two or more extremities and moderate trophic disturbances (atrophy, possibly paralysis with commencement of contractures). Bacteriological examination negative.

Iq2c––. Quiescent, moderate, cutaneous, indeterminate leprosy. Presence of fairly numerous macules (hypo- or hyperpigmented). No neural disturbances. Bacteriological examination negative.

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[4] The stage to the disease from the neural point of view is indicated schematically in the "examples," later.