REPORT OF THE COMMITTEE ON CLASSIFICATION SECOND PAN-AMERICAN LEPROSY CONFERENCE [1]

Rio de Janeiro, October 1946

The Committee on Classification of the Second Pan-American Leprosy Conference recommends the adoption, for the American countries, of the so-called "South American Classification" with the modifications here proposed.

Holding as a basis of classification the separation of a tuberculoid type distinct from the neural one, this Committee follows in a definite way the recommendations made by the Cairo congress that "future research be in the direction indicated by questions raised by the minority . . . ." which was composed of South American members of the Subcommittee on Classification. Investigations carried on since then on this continent [have been published, discussed in certain meetings, investigated in several countries, and amply confirmed].

The classification proposed by us holds as the point of departure the different structural types of leprosy, attempting to integrate them, by recognition of the clinical and immunological elements, into a coherent doctrine capable of orienting prophylaxis, epidemiology, prognosis and therapy.

In these premises, this Committee recognizes three fundamental types: lepromatous (L), incharacteristic (I) and tuberculoid (T), the incharacteristic type representing the dynamic element between the two polar types. Each of these three fundamental types presents its own particular clinical forms, which are defined later.

These clinical varieties, or forms, are distinguished on the basis of objective and evolutive characteristics. Thus the *lepromatous* type includes simple macular forms, because these frequently mark the marginal zone (*fronteira*) between this type and the incharacteristic one. These are to be distinguished from the infiltrative and nodular forms, which characterize the stable phase of the lepromatous type. There being cases

[1] A new translation of the Portuguese text as published in Vol. III of the official transactions, *Arq. Serv. Nac. Lepra* **5** (1947) 176-186, and also in *Rev. brasileira Leprol.* **14** (1946) 334-344, checked against the over-free and incomplete translation in the *Internat. J. Leprosy* **15** (1947) 100-107 and by Dr. Ernani Agricola, of Rio de Janeiro, and Dr. Lauro de Souza Lima, of São Paulo. The originals have not been followed entirely with respect to type-face and paragraphing, which were not consistent, and certain portions not contributory to the delineation of the classification scheme have been dealt with summarily.

of exclusively nerve involvement in which the diagnosis of the lepromatous type can be arrived at, the present classification includes a neural form of this type. Although it is admitted in principle that there is a certain degree of general dissemination in all forms of the lepromatous type, there is distinguished here a generalized form, to comprise the advanced cases with extensive cutaneous, neural and visceral involvement.

In the *incharacteristic* type there are distinguished macular, neuro-macular and pure neural forms, the definitions of which are self-evident. Although we recognize that there are almost always changes of the nerve branches of the dermis, we distinguish simple macular forms for clinical reasons, these being the lesions which are most evident in the examination of cases of this type. For the same reasons there are recognized as neuro-macular forms those which present, besides the characteristic macular eruption, lesions of the nerve trunks which are manifested by neuritic symptoms.

The *tuberculoid* type comprises macular, papuloid-circinate, neural and reactional forms.[2] As in the lepromatous type, there are distinguished simple macular forms, which frequently are marginal (*fronteiriças*) between the tuberculoid type and the incharacteristic one, from which they arise. By the term neural is meant in this type those cases with thickening of the nerve trunks, manifested by clinical signs which are characteristic of this localization of the disease. There are cases, particularly typical of this [neural] form, which present signs of caseation.

DEFINITIONS OF THE TYPES

Lepromatous type (L).—The serious cases of the disease, of high degree of stability, always negative to lepromin, which present lepromatous lesions of the skin, mucous membranes, nerves and other organs. Bacteriologically, the lesions are always positive. Histologically, the aspect is that of a specific

[2] Marginal note of the rapporteurs, Drs. F. R. Tiant and Nelson Souza Campos: Due to an omission by the Editorial Committee of the report on classification there were not included, among the tuberculoid forms, the following clinical eventualities which, because of their frequency, should be taken into account: (1) The macular and papuloid-circinate cases presenting neurotrophic symptoms, which may be designated "neuro-macular" and "neuro-circinate," respectively. (2) Similarly, the reactional form includes borderline (*limitantes*) lesions. This note is made in the margin of the text to be approved for the purpose of calling attention to these terms.

granuloma. This is the outstandingly contagious type of the disease.

Incharacteristic or undifferentiated type (I).[3]—In general, benign cases of the disease, relatively unstable, with positive or negative lepromin reactions, and presenting distinctive (*definidos*) cutaneous and neural lesions. Bacteriologically, the findings are generally negative, or only rare bacilli are found. Histologically [the lesions] present a chronic inflammatory condition without distinctive (*definidos*) characteristics. Such cases may eventually become contagious.

Tuberculoid type (T).—Benign cases of the disease, of great stability, almost always reacting positively to lepromin, which present characteristic (*peculiares*) lesions of the skin and nerves. Bacteriologically, the lesions are negative as a rule, and when positive the bacilli are few. The histology is that of a tuberculoid granuloma. These cases are held to be practically noncontagious.

Besides this type [as described], which we may call quiescent (*tórpido*), there are acute or reactional tuberculoid cases which differ from the foregoing description in that positivity to lepromin is less frequent, the lesions have a special clinical aspect, and bacteriologically the findings are frequently positive in the lesions and occasionally in the mucosa, although temporarily. The histology is the only thing that unites them [with the nonreactional kind], although it presents the characteristics of the acute phase (edema, dissociation and loosening of the foci, vacuolization, etc.). These cases are to be considered contagious during the stage of bacteriological positivity, and eventually they may transform into the lepromatous type.

Type	*Variety or clinical form*
Lepromatous (L)	Macular Infiltrative (in plaques or diffuse) Nodular Neural Generalized
Incharacteristic (I) or undifferentiated	Macular Neural Neuro-macular

[3] The designation given this type is provisional, for lack of a more precise term to express the peculiar clinical aspects, the anatomical substratum of which is that of an inflammatory lesion without definite characteristics.

Type	*Variety or clinical form*
Tuberculoid (T)	Macular Papulo-circinate Neural Reactional [4]

A. Lepromatous Type

The lesions corresponding to this type may be:

(*a*) *Lepromatous macules.*—Erythematous macules, rose-colored, reddish or reddish-purplish (violaceous). Other alterations of color [such as] yellowish (tawny), brownish, coppery or rust-colored may exist from the outset in the pigmented macules, or these may be associated with the congestive element and explain the initiation of the process as mixed or erythemato-pigmented macules. In any one case, macules of one of the above varieties may predominate, while in others there may exist at the same time macules of different varieties. The macules may have a uniform aspect, and may have irregular and diffuse contours; but the central part of the lesion may appear normal or hypochromic, contrasting clearly with the erythematous or pigmented part which surrounds it.

(*b*) *Infiltrations.*—When the lesions assume this character they usually have the same color characteristics as the various macules, and they present more or less marked elevation, or at times .frankly salient and prominent. The infiltrative lesions may be circumscribed—lepromatous plaques—of variable dimensions, or [they may be] diffuse lepromatous infiltrations. Not infrequently their coloration is made distinctive by a peculiar greasy aspect. As for size and shape, both the circumscribed and diffuse infiltrations may present the same peculiarities as the lepromatous macules.

(*c*) *Tubercles.*—Circumscribed elements originating in the dermis, which may or may not be salient, firm to the touch, whose color may be reddish, violaceous, brownish or bronzy, of various sizes, miliary, pisiform, lenticular, or at times even larger. They may be isolated, confluent or compounded by coalescence to form masses of more or less large size, at times of mammilate aspect. Tubercles, as well as the other infiltrative lesions, may involve, besides the skin, the semi-mucosas and the visible mucosas, presenting in these places the same characteristics as those in the skin.

(*d*) *Nodules.*—Subcutaneous elements, firm to the touch, which may or may not cause external elevation, of spheroid form, variable in size, smaller than a pea in some instances, much larger in others. In those cases in which, in their evolution, the nodules become adherent to the dermis, they usually impart to the skin a rosy or violaceous color, and the so-called orange-peel appearance.

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[4] Under this term are included both the tuberculoid lepra reaction (Wade) and cases in which the acute episode assumes a different reactional modality (reactional tuberculoid leprosy, Souza Campos). The former is a reactivation of preexistent lesions of the tuberculoid type, generally undergoing involution to the original form; the latter develops ordinarily from the incharacteristic type and may, in its progressive evolution, transform to the lepromatous type, or, by involution, it may return to the original type.

(*e*) *Ulcerations.*—These lesions are frequently seen when there has been a loss of substance due to the softening of nodules, tubercles or infiltrations, whether in the skin or in the mucous membrane.

(*f*) *Cicatricial lesions.*—These may be seen as sequelae of the infiltrative or ulcerative lesions, endowed with specific characteristics.

(*g*) *Lepromatous reaction.*—A very frequent intercurrent episode in the course of lepromatous leprosy is the appearance of reactional outbreaks (*surtos*) commonly called leprotic reaction ("lepra reaction"). Its clinical picture is characterized by cutaneous manifestations of the erythema multiforme or erythema nodosum type which may or may not be associated with neural, ocular or visceral reactions. The process evolves with fever, general weakness and algias. It occurs in acute, subacute or chronic forms.

B. Incharacteristic or Undifferentiated Type

There are three elementary forms of the incharacteristic lesions: erythematous, erythemato-dyschromic, and achromic. These three varieties of lesions are, in general, devoid of infiltration. In rare cases, however, they may show distinct elevation. In all of them there are, constantly, disturbances of sensitivity, although of varying degrees.

Usually the lesions of the incharacteristic type are of small size [5] and few in number. At times they may be roseoliform and disseminated. The outlines of these lesions are either relatively distinct, or indistinct and diffuse, and it is a peculiar trait that both characteristics may coexist in the same lesion. With regard to color it is to be emphasized that, besides the rosy tone of erythema, there may be seen various degrees of hypochromia, due to disturbances of the normal melanogenesis. These three forms of incharacteristic lesions are far from exclusive, for there may be combinations of them.

The *achromic* lesions appear in some cases as relatively well-circumscribed areas, at other times in a more diffuse form, and there are not infrequent cases in which one sector of the lesion is diffuse while others are well-defined. Frequently these lesions are few, two or three on the entire skin surface, or even solitary.

The *erythemato-dyschromic* macule almost always appears in the diffuse form, with ill-defined outlines. Such macules present a mixture of erythema and hypochromia, there being cases of intermediate tone over all of the surface and others with a peripheral rosy halo with or without margination.

The *erythematous* macule has the aspect of a congestive lesion, pale-rose to reddish, or variable sizes, and at times with relatively distinct borders but at other times diffuse.

C. Tuberculoid Type

The tuberculoid type may be manifested by simple erythematous or erythemato-pigmented macules similar to those seen in the incharacteristic type, or by brownish macules, of clear-cut outline, and showing a fine pityriasis-like desquamation.

The element which characterizes the tuberculoid type, however, is the tubercle, which in this instance has a distinctive aspect represented by

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[5] In the original, *tamanho numular.* Variously translated, "size of a coin, a shilling or half-dollar," and "3 to 5 cm."—EDITOR.

miliary papuloid lesions, pin-head in size or smaller, of reddish-purple or reddish-bronzy color, very slightly elevated, at times showing slight desquamation, isolated or more often confluent and even conglomerate. These elements may be dispersed, but more commonly they are united to form plaques of variable size and configuration and with a finely granular surface.

In other cases there may be seen fusion of these elements to form plaques of circular or elliptical configuration, well-defined as to the outline, of succulent aspect and irregular surface, finely scaling. More often, however, the miliary tubercles are clustered in the periphery of the plaque to form a more or less broad border, well-defined at the outer edge but ill-defined on the inside, and formed of an aggregation of numerous papuloid elements; they may be isolated (*distintos*) or fused to form a granular-surfaced zone. These plaques, which constitute the most characteristic feature of the type, are annular, irregularly oval, or of capricious, geographic configuration, the border either continuous or interrupted, the center usually hypochromic and covered with fine scales.

The foregoing description refers to the quiescent, torpid lesions of the tuberculoid type. The reactional form presents eruptive lesions which are much more polymorphous, the predominant elements being tuberiform, congestive, salient, generally of purplish color, and variable as to size. The eruption appears abruptly, then following a subacute course.

In childhood there may be observed a variant of the reactional condition, manifested by one or a few tuberiform lesions which leave characteristic depressed scars.

NEUROLOGICAL SYMPTOMS USEFUL FOR THE CLASSIFICATION OF CASES

Contrary to the situation with respect to the cutaneous symptomatology, the neurological syndrome, which is an expression of a preferential localization of the leprosy infection, does not have characteristics distinctive of each of the three principal types. For this reason it becomes necessary to emphasize the few features which may permit, or at least help in, the diagnosis of the type. These are in this case, with certain reservations, areas of anesthesia, thickening of the nerves, muscular atrophy, and certain trophic phenomena such as perforating ulcer, mutilation, etc.

Areas of anesthesia.—Leprosy cases in which the disease is manifested only by [local] areas of anesthesia, with or without anhydrosis, changes of the pilo-sebaceous follicles, etc., may in practice be classified as of a pure neural form of the incharacteristic type, unless there exists in connection with the anesthetic area a thickened subcutaneous nerve branch. In that event one may, almost without error, classify the case as of the pure neural form of the tuberculoid type.

Thickening of nerves.—Leprosy cases which present only thickening of the nerves, with complete absence of an eruption or any other manifestations in the skin or other organs, are relatively frequent. It seems practically impossible [in such cases] to make a purely clinical diagnosis of the type, without the aid of the lepromin test and of puncture or biopsy of the nerve. If the lepromin reaction is negative, doubtful or weakly positive, and the nerve puncture is negative for acid-fast bacilli,

the case should be classified as of the pure neural form of the incharacteristic type, if one cannot make a biopsy which would confirm or disprove that type diagnosis. If the lepromin reaction is negative and the nerve puncture positive for *Mycobacterium leprae,* the biopsy will determine whether the type is lepromatous or incharacteristic. It must be said, however, that these cases are extremely rare. [Referring to lepromatous?]

Relatively less rare are the cases with pure thickening of a nerve or nerves, the lepromin reaction clearly positive and nerve puncture negative. Especially [significant] is the finding of tumefactions in the course of the nerve, with or without fluctuation, or even fistulas and retracted scars adherent to the nerve. All of these cases should in practice be classified as of the pure neural form of the tuberculoid type.

Trophic phenomena.—Leprosy cases presenting partial or total atrophy of the muscles of the extremities, mutilations, trophic ulcers, etc., with regional anesthesia (exceptionally without anesthesia), are relatively difficult to classify satisfactorily. In these cases the problem of type diagnosis is resolved in the same manner as already specified for cases of nerve thickening.

BACTERIOSCOPY

[This section deals with the technique of the bacteriological examination, giving instructions for the collection of material and for staining, with instructions for grading smears as follows:]

Results.—Negative: When no bacilli can be found in at least 100 microscope fields. Positive: (+), rare, one or less than one bacillus per field; (++), many, when bacilli are seen in all fields; (+++), abundant, when a large number of bacilli or globi are seen in all fields.

HISTOLOGY

[Because of the significance given the histopathology of the lesions in this classification, the essential part of this section is included here.]

Definitions.—(*a*) The lepromatous lesion is a specific granuloma characterized by the presence of the vacuolated cells of Virchow. The reactional lepromatous lesion is a perifocal exudative one characterized by edema, hyperemia and polymorphonuclear infiltration associated with the specific granuloma.

(*b*) The tuberculoid lesion is a granuloma in which are encountered epithelioid cells with a tendency to follicular disposition, with or without giant cells, usually surrounded by a lymphocytic halo. The reactional tuberculoid lesion is the tuberculoid granuloma accompanied by exudative phenomena, hyperemia and edema which modify its characteristic aspect (vacuolization by edema).

(*c*) An incharacteristic lesion is one which is represented by slight perivascular, perineural, periglandular and perifollicular lymphocytic infiltration; it is devoid of Virchow and epithelioid cells.[6]

Interpretation.—The histological examination has an absolute diagnostic value in the lesions of the lepromatous type, and relative value in

[6] This definition is from the previous translation, the original version being obviously incomplete.—EDITOR.

the tuberculoid and incharacteristic types; in the last two [it is so] in correlation with the clinical diagnosis.

IMMUNOLOGY

[This section deals with the Fernandez (early) and Mitsuda (late) reactions to lepromin, dosage of antigen (0.1 to 0.2 cc.), the preferred sites of the test, and the antigen, and investigations which should be carried out. The antigen recommended is the Dharmendra bacillary one "because it can be standardized" and "because in practice no categorical difference" between it and the classical one has been proved. The schedules of readings, somewhat condensed, are as follows:]

Fernandez reaction.—Reading: At 48 hours. Interpretation: negative (—), absence of halo, or halo less than 5 mm. Doubtful (±), halo greater than 5 mm. but less than 10 mm. Positive (+), reaction with a well-defined infiltrated erythematous halo not less than 10 mm. and up to 15 mm.; (++), the same type of reaction but more than 15 mm., up to 20 mm.; (+++), the same type of reaction greater than 20 mm.

Mitsuda reaction.—Reading: Between 20 and 30 days, taking into account the diameter, color, infiltration and evolution. Interpretation: Negative (—), absence of any visible or palable element. Doubtful (±), perceptible element without the characteristics of positivity. Positive (+), elevated and infiltrated element, of rose to purplish color, progressive and persistent, from 3 to 5 mm. in diameter; (++), the same, greater than 5 mm; (+++), when there is ulceration.