

## I. THE CLASSIFICATION OF LEPROSY

### REPORT OF THE SUBCOMMITTEE ON CLASSIFICATION\*†

The problems of classification of cases of leprosy should be viewed broadly, bearing in mind both (a) the requirements and circumstances of work of the practical field worker to whom classification is necessary for purposes of prognosis, treatment or control, but who cannot apply elaborate or time-consuming methods of differentiating types of the disease, and (b) the refinements of such differentiation that are possible to the specialist who employs special methods of investigation. The great majority of persons who deal with leprosy, work under circumstances that require that the basic or primary classification be as simple as possible.

Progress in knowledge of the forms of leprosy and of the nature of the leprous processes has been made since the classification that is now most generally used was adopted by the Leonard Wood Memorial Conference on Leprosy in 1931, and it is now possible to modify some of the terms of that classification to eliminate certain causes of misunderstanding. However, our knowledge of the matter has not yet progressed to a point where it is possible to attain unanimity of opinion on certain essential features.

It is recommended that for the present the basic division of leprosy into two types, along the lines laid down in the Memorial Conference classification, be continued until such time as further study of the matter permits attainment of unanimity. It is further recommended that future research be in the direction indicated by the questions raised by the minority of this committee, the main question being whether or not the neural type of the Memorial Conference classification should be divided into two distinct main types, "simple neural" and "tuberculoid." For the present it is the predominant opinion that such divisions should be considered as subtypes or varieties.

\* This committee consisted of Dr. H. W. Wade (*Chairman*), Dr. R. C. Germond (*Secretary*), and Drs. P. L. Baliña, A. Dubois, J. M. M. Fernandez, V. Klingmüller, J. Lowe and Rabello, Jr.

† The French translation of this report, prepared in accordance with action of the Resolutions Committee by Prof. E. Marchoux, with the collaboration of Drs. R. C. Germond and A. Dubois, is here published for the first time. It follows the report of the subcommittee on cultivation of the leprosy bacillus.

Objections have repeatedly been raised to both of the current names of the two types (i.e., "neural" and "cutaneous") because of confusion arising from the special sense in which they are employed in leprosy classification, because of difficulties of translating them into other languages, and for other reasons. However, no other words have been proposed which are free from similar objections. It is the opinion of the committee: (a) That for the time being, at least, the word "neural" should be retained for the type to which it is now applied. (b) That because "cutaneous" has proved particularly confusing its use should be discontinued, and replaced by the term "lepromatous," the symbol of which is "L."

It is proposed that the definition of the two types of the Memorial Conference classification be amended as follows:

#### PRIMARY CLASSIFICATION

*Neural (N) type.*—All cases of the "benign" form of leprosy, with disturbances of polyneuritic nature (i.e., alterations of peripheral sensation, trophic disturbances, atrophies and paralyses, and their sequelae), or macules of nonlepromatous nature (i.e., leprides, usually with localized sensory disturbances), or both. These cases give evidence of relative resistance to the infection, are of relatively good prognosis as regards life, although mutilation may take place, and usually react positively to leprolin. Bacteriologically the skin lesions are typically but not invariably found negative by standard methods of examination, though the nasal mucosa may be found positive. Many of these lesions are histologically of tuberculoid nature.

*Lepromatous (L) type.*—All cases of the "malignant" form of leprosy, relatively nonresistant and of poor prognosis, usually negative to leprolin, exhibiting lepromatous lesions of the skin and of other organs, especially the nerve trunks. Bacteriological examination usually reveals abundant bacilli. Disturbances of polyneuritic nature may or may not be present; they are usually absent in the earlier stages and present in the later stages of primarily lepromatous cases, and are often present in cases arising secondarily from the neural form.

#### SUBCLASSIFICATION

Subdivision of the types of leprosy may be made from two points of view: (a) with respect to the degrees of advancement of the disease, and (b) with respect to the forms or

varieties of cases within a type (i.e., subtypes), based on the nature of the lesions. The former method of subdivision is that of the Memorial Conference classification and it has proved useful in the hands of many workers, especially in dealing broadly with large numbers of cases. The latter method of subdivision is generally employed in dealing more precisely with individual cases. Both methods have their uses and should be understood, but a generally applicable, practical formula for combining the two has not been arrived at. The two methods are dealt with independently here.

1. GENERAL SUBCLASSIFICATION  
(BY DEGREES OF ADVANCEMENT)

The following specifications indicate in a general way the basis of the division into three degrees of advancement of the disease in each type.

*Neural 1 (N1).*—Slight neural: (a) Cases with from one to several small macules, or a proportionally smaller number of larger macules, whether flat or elevated, without indications of polyneuritic changes; or (b) cases presenting only polyneuritic changes of slight degree: disturbances of peripheral sensation affecting one or two extremities, not of marked extent, with only minor trophic disturbances, muscular atrophy or paresis, if any; or (c) cases showing combinations of macular and polyneuritic manifestations in equivalent degree of total affection.

*Neural 2 (N2).*—Moderately advanced neural: (a) Cases with fairly numerous or large macules, of wide distribution, without evidence of polyneuritic changes, or with such manifestations of fairly slight degree; or (b) cases presenting only polyneuritic changes of moderate degree: peripheral anesthesia of considerable extent if affecting only one extremity, of less extent if affecting more than one; and moderate trophic changes, atrophy and paralyses, including beginning contractures if of limited extent; or (c) cases showing combinations of equivalent total degree.

*Neural 3 (N3).*—Advanced neural: (a) Cases with very numerous or very extensive macular lesions of the more marked kinds, with polyneuritic changes; or (b) cases presenting only advanced polyneuritic changes: extensive peripheral anesthesia and more or less marked motor and trophic disturbances: paralyses, atrophies, contractures, trophic ulcers and mutilations; or (c) cases showing combinations of equivalent total degree.

*Lepromatous 1 (L1).*—Slight lepromatous: Cases with lepro-

matous skin lesions consisting of one or a few macular areas, or a few small infiltrated patches or small nodules, or diffuse lepromatous changes of slight degree; lesions of the nasal mucous membrane are usually absent.

*Lepromatous 2 (L2).*—Moderately advanced lepromatous: Cases with numerous macular areas or fairly numerous small or fewer large infiltrations or nodules, or diffuse lepromatous changes of moderate degree; lesions of the nasal mucous membrane are frequently present.

*Lepromatous 3 (L3).*—Advanced lepromatous: Cases with numerous and extensive or very marked lepromatous lesions, which may vary in their stage of development or retrogression; lesions of the nasal mucous membrane are almost always present.

*"Mixed" cases.*—Recognition should not be given to "mixed leprosy" as a *type*. However, cases of the lepromatous type usually exhibit, sooner or later, varying degrees of polyneuritic involvement, and for precision such "mixed" or "complete" cases may be designated LN. The symbol L should be given precedence, regardless of the original nature of the case or the relative severity of the two elements, because of the predominant importance of the lepromatous element. In grading the degree of advancement of these cases the appropriate figure is placed after each symbol; e.g., L2-N1, or L1-N3.

*Secondary neural cases.*—Cases that have previously been of the lepromatous type with polyneuritic features (mixed cases), but in which the lepromatous lesions have resolved leaving only the polyneuritic manifestations, are called "secondary neural."

## 2. SPECIAL SUBCLASSIFICATION

(ACCORDING TO THE NATURE OF THE LESIONS PRESENTED)

1. *Lepromatous type.*—No varieties of the lepromatous type of leprosy have been established that are sufficiently distinct, frequent and general in occurrence to require recognition in formal classification. In some places (e.g., India) where many cases show, at least for a time, extensive "diffuse" involvement of the skin, not localized in macules or infiltrations, there might be an advantage in distinguishing such cases (which might be indicated by the symbol Ld), but it is not certain that this division would be generally useful.

2. *Neural type.*—The neural type of leprosy may be divided into two main subtypes, namely, "anesthetic" and "macular." For

some purposes such subclassification may be sufficient. However, for more exact work the macular variety should be divided into "simple" and "tuberculoid," and the latter may be further divided into "minor" and "major" forms. For such work, therefore, the following is proposed:

Neural (type)

Anesthetic (nonmacular, polyneuritic) (Na)

Simple macular (with flat macules) (Ns)

Tuberculoid macular (minor and major) (Nt)

*Anesthetic.*—This variety of neural leprosy presents evidence of involvement of nerve trunks only (polyneuritic changes and sequelae) without macular skin lesions.

*Simple macular.*—The simple macular cases, which comprise a considerable proportion of those encountered, present skin lesions (leprides) that have no or only very slight elevation or palpable infiltration. When elevation is present, it is often difficult to detect it in diffuse light, and the surface is smooth, not granular or pebbled in appearance; such elevation is usually limited to a narrow marginal zone. "Residual" lesions, which are without elevation and therefore are "simple" under this definition (though they may be affected by scarring) should not be considered as a separate variety.

*Tuberculoid macular.*—This subtype, as stated, may be divided into two groups, which are here described separately.

(a) *Minor tuberculoid:* The lesions so designated are the less marked ones of the kind that has become generally recognized to be histologically tuberculoid and that is clinically recognizable as such with certainty. These lesions show definite elevation of characteristic appearance, though there are considerable variations. They are usually marked by irregularity of the surface, due to the essentially focal nature and superficial location of the tuberculoid process. That condition may produce elevated bands or areas which may be continuous, or discontinuous even to the point of producing isolated papulations. Occasionally the process is relatively deep in the dermis, in which case the surface may be relatively smooth, and the appearance may therefore approach that of some of the major tuberculoid lesions, but the degree of the condition is less than in that form.

(b) *Major tuberculoid:* The lesions so designated are the more striking, grossly elevated ones to which recognition as tuberculoid has been largely confined in the past. They are "major"

both in degree and nature of the pathological process. Typically the process invades the deeper layers of the skin to a marked degree, and also the subcutaneous tissue, and, by further extension in the cutaneous nerves related to the macules, it may produce gross involvement of them. Macules of this variety are those most liable to be mistaken for lepromata, especially when they are (a) small but thick, morphologically nodular, (b) in a "reaction" condition, reddish, turgid and smooth, or (c) bacteriologically positive. One feature that helps to differentiate them is their typically sharp demarkation and frequently asymmetrical distribution. Another distinguishing feature is the occasional tendency to the development of marked enlargement of the local cutaneous nerves, which condition sometimes extends to the main trunks of an affected extremity, thus introducing a secondary polyneuritic element. A point of importance is the frequency with which these lesions start abruptly, as a "reaction" condition, and the relative rapidity—and, sometimes, the completeness—with which they may subside.

#### DEFINITIONS

For the purpose of amplifying certain features of these proposals, and of facilitating the attainment of uniformity in applying them, the following definitions are adopted.

*Leproma*.—The lepromatous condition, which is the distinguishing feature of the type of leprosy so named, is a granulomatous one in which the invaded tissues show maximal tolerance of the bacilli. The essential histological feature is an accumulation of "lepra cells," which may show little differentiation from their original form (the macrophage), or may contain globi, or may undergo multiple vacuolation to produce the so-called Virchow cells, which are often multinucleate. The lepra cells contain bacilli in considerable and often great numbers, though bacilli may also occur in cells of other types. Lepromatous lesions in the skin may be so slight as to be imperceptible, ranging up to marked, extensive infiltrations or conspicuous nodular masses. As a rule they are more ill-defined and diffusely outlined than the leprides, and they do not exhibit the same tendency to radial extension or the same changes of color or sensation.

*Lepride*.—This term is applied to the discrete macular lesions that are characteristic of neural leprosy when the skin is involved. The leprides vary greatly in appearance, size and as regards elevation; they may be flat or markedly thickened; they may be



smooth-surfaced or very irregular ("granular," "pebbled," or micropapulate); they tend to enlarge radially and to merge with adjacent ones, and to undergo central resolution. Disturbance of sensory perception, slight or marked, is a typical feature, though its development may be delayed. The definitely elevated leprides, at least, are granulomatous, the essential feature being the non-specific "tuberculoid" change, together with which there usually is banal chronic inflammatory infiltration of variable degree. Associated cutaneous nerves may be similarly affected and may undergo necrosis or even cold-abscess formation. Though these lesions result from the reaction of the tissue to the presence of the leprosy bacillus, ordinarily bacilli are not found in smears and only in very small numbers in sections. In occasional cases, however, especially during or after a reaction condition, bacilli can be found in smears, and they may be numerous.

*Leprotic and leprous.*—These terms should be used only in their general sense, signifying pertaining to or affected with leprosy.

*Lepromatous.*—This term signifies of the nature or possessing the qualities of the leproma. In classification, as here proposed, it applies to cases with this form of lesion.

*Macule.*—This term is specifically applied to the leprides (neural type leprosy) though it is sometimes used to designate lepromatous patches. It signifies a circumscribed area of skin of abnormal color—varying widely in this character in different races but usually hypopigmented, occasionally hyperpigmented, and often erythematous—and commonly with other surface abnormalities, such changes being evident in the whole or only in parts of the area. In the terminology of leprosy it is used without regard to the presence or absence of infiltration or elevation.

*Infiltration.*—This name is often applied clinically in a special sense to a diffuse thickening of lepromatous nature.

*Plaque.*—Ordinarily this term is applied only to large leprides in which central resolution is delayed or absent, and not to areas of diffuse infiltration.

*Papule.*<sup>1</sup>—A papule is a small, more or less solid, circumscribed, superficial elevation of the skin, usually but not necessarily circular, conventionally described as varying in size from that of a pinhead or less to five millimeters in diameter (split-pea size). Papules occur in both forms of leprosy and differ correspondingly in structure and often in appearance.

<sup>1</sup>The definitions of papules and nodules here given are those generally

*Nodule.*<sup>1</sup>—A nodule (synonymous with but preferable to "tubercle") is a solid elevation of the skin, often similar to a papule except that it is larger; in practice the application of this term is not limited as regards maximum size. Ordinarily it is applied only to lepromata. Nodules are usually more deep-seated than papules, and often arise from localized subcutaneous masses.

*Polyn neuritis.*—This term has been employed to designate involvement of the peripheral nerve trunks which results in sensory changes of the extremities that tend to spread centripetally ("acroteric" anesthesia), and in trophic changes of various kinds, and paralyses and atrophies which may also involve the face. Polyn neuritic manifestations do not include the sensory changes in the leprides, or lesions of superficial cutaneous nerves that develop by extension from leprides.

*Trophic changes.*—Under this head are included those changes that are ordinarily ascribed to disturbances of the vaso-motor system and of nutrition: anidrosis, glossy skin, ichthyosis, pigmentary changes, loss of hair, perforating ulcers, atrophy and necrosis of bones with consequent mutilations and neuropathic joint lesions. Strictly speaking, atrophy and paralysis of muscles, and contractures consequent on such changes, are not included, but in practice the distinction is often not made.

#### SPECIAL SYMBOLS

*Indication of the original phase of the disease.*—If it is desired to indicate symbolically in a mixed case the form that occurred first, this can be done readily by placing the prime accent mark (´=primary) after the appropriate letter, as L´N or LN´. This would not interfere with the use of the customary figures to indicate the degree of advancement.

*Indication of secondary neural cases.*—If it should be desired to indicate a secondary neural case, that can be done as N<sup>°</sup> (°=secondary).

accepted by British and American dermatologists. It is to be recognized that these terms are employed in different senses in other countries. The members of the committee from South America submitted the following comment:

"Quelques auteurs, parmi lesquels se rangent les léprologues de l'Amérique du Sud, n'admettent pas le terme 'papule' dans la lèpre, parce que en dermatologie la papule est une efflorescence spontanément résolutive, et que dans la lèpre les lésions cutanées, auxquelles les auteurs anglais et américains appliquent le nom de papule, ne s'effacent qu'en laissant après elles une lésion cicatricielle, au moins histologiquement. A la rigueur on pourrait employer pour les désigner le terme 'papuloïde.' Quand à ce qu'est à propos des lésions granulaires de la lèpre tuberculoïde, pour les mêmes raisons les dermatologistes de l'Amérique du Sud préfèrent adopter la désignation de lésions 'micropapuloïdes.'"



*Indication of bacteriological status.*—If, for epidemiological or other considerations, it is desired to indicate in a case symbol the bacteriological status of the case, that can be done by adding + or – (or, as suggested by Lie, B+ or B–) to the case symbol.

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Professor Baliña and Dr. Rabello, Jr. who took part in the deliberations on the subject of the classification of the clinical forms of leprosy, wish to point out that they have already published their personal views on this matter, either during or before the Congress. The definitions and descriptions here adopted are based on those in articles by H. W. Wade in *Internat. Jour. Lep.* **4** (1936) 409-430 and *American Jour. Trop. Med.* **17** (1937) 773-801.