

TECHNICAL REPORTS

We here reprint (from the International Journal of Leprosy, Vol. 21, No. 4, Oct. Dec. 1953) the reports of the Technical Committees of the Madrid Congress, as adopted by the plenary session of the Congress.

CLASSIFICATION¹

The criteria which bear on the classification of leprosy cases are: (1) clinical, (2) bacteriological, (3) immunological, and (4) histopathological. Existing systems of classification differ with respect to the priority given certain of these criteria.

The Committee agrees unanimously that the basic criteria of primary classification should be clinical, comprising the morphology of the skin lesions and the neurological manifestations. Indispensable in connection with the clinical examination is the bacteriological criterion, involving examination of smears from skin lesions and the nasal mucosa.

In the study of cases full use should be made of the immunological criterion (the lepromin test) and of the histopathology of the lesions. These factors are of value in the determination of types, and may be essential in the determination of sub-groups.

The histological examination, though important in the diagnosis of the form of leprosy and consequently in connection with prognosis, should not govern the primary classification, except when, as may happen, it definitely indicates the clinical classification of the case to have been in error. In such instances—if the lepromin reaction is in agreement with the histologic findings—the case should be reclassified.

Cases should be classified according to findings at the time of the examination. They may or may not present evidence, by history or by objective stigmata, of a previous form or phase of evolution, and sometimes these features are significant with respect to present classification.² The evidence obtainable may indicate a likelihood of change to another form or phase in the future evolution of the disease, but that factor does not affect the determination of form (as to type or group) until such a change actually occurs.

The Committee considers that this system of classification offers every possibility for further progress.

PRIMARY CLASSIFICATION

The Committee recommends that two distinct *types* of leprosy,

¹ The Committee on Classification was composed as follows: Dr. H. W. Wade, *chairman*, Dr. José Gay Prieto, *vice-chairman*, Dr. Martín Vegas, *secretary*, and Drs. G. Basombrío, R. G. Cochrane, V. R. Khanolkar, Kané-hito Kitamura, Francisco Lapatí, and Francisco E. Rabello, *members*; also, Dr. Harry L. Arnold, Jr., *co-opted member*.

² For example, it may be known, or be evident from existing stigmata, that a case presenting only simple macules was previously tuberculoid. Such a case should be classified as "residual tuberculoid" and not as "indeterminate."

lepromatous and tuberculoid, be recognized, thus maintaining the concept of polarity. It also recommends that two *groups* be recognized, indeterminate and borderline (dimorphous).

The following definitions are offered:

Type connotes clinically and biologically stereotyped features, characterized by marked stability and mutual incompatibility.

Group connotes less distinctive or positive characteristics, less stability, and less certainty with respect to evolution.

Variety connotes a subdivision of a type or group.

PROPOSED TYPES, GROUPS AND VARIETIES

Lepromatous type (L)

Macular
Diffuse
Infiltrated
Nodular
Neuritic, pure (?) ³

Tuberculoid type (T)

Macular
Minor tuberculoid (micropapuloid, etc.)
Major tuberculoid (plaques, annular lesions, etc.)
Neuritic, pure.

Indeterminate group (I)

Macular
Neuritic, pure

Borderline (dimorphous) (B)

Infiltrated
(Others?) ⁴

LEPROMATOUS TYPE (L)

A malign type, especially stable;⁵ strongly positive on bacteriological examination; presenting more or less infiltrated skin lesions; negative to lepromin. The peripheral nerve trunks become manifestly involved as the disease progresses, habitually in symmetrical fashion and often with neural sequelae in advanced stages.

TUBERCULOID TYPE (T)

Usually benign; stable; generally negative on bacteriological examination; presenting in most cases erythematous skin lesions which are elevated marginally or more extensively; positive to lepromin.

Sequelae of peripheral nerve trunk involvement may develop in a certain proportion of cases, and this may give rise to serious and disabling deformity. This frequently appears to occur as a result of extension from or through cutaneous nerve branches, rather than of systemic dissemination, and consequently it is often asymmetric and unilateral.

³ Cases of this variety have been observed by some workers, but have not as yet been reported in the literature.

⁴ See addendum by Drs. Khanolkar and Cochrane.

⁵ The word "stable" implies stability as regards the type, not as regards the severity of the disease.

Tuberculoid leprosy should be subdivided as follows:

Macular tuberculoid (Tm). These cases present macules with clear-cut and definite margins, the surface generally smooth and dry, invariably with some loss of cutaneous sensibility; almost always negative on bacteriological examination, or with at most only a few bacilli.

Minor tuberculoid (micropapuloid, etc.) (Tt). Skin lesions are only slightly to moderately elevated, often only at the margin or even a part of the margin, usually with irregularity of the surface. The condition tends to be relatively superficial, and palpable enlargement of cutaneous nerves associated with the lesions is infrequent.

Major tuberculoid (plaques, annular lesions, etc.) (TT). Skin lesions, often smooth of surface, are more markedly elevated and thickened than those of the minor variety, the affected zone usually broader; the more recent lesions may show only partial central recession or no recession. Because of the degree of the condition in the deeper levels of the skin, manifest extension in the associated cutaneous nerves is relatively frequent and marked.

INDETERMINATE GROUP (I)

A benign form, relatively unstable; seldom bacteriologically positive; presenting flat skin lesions which may be hypopigmented or erythematous; the reaction to lepromin negative or positive. Neuritic manifestations, more or less extensive, may develop in cases which have persisted as of this group for long periods. The indeterminate group consists essentially of the "simple macular" cases. These cases may evolve towards the lepromatous type or the tuberculoid type, or may remain unchanged indefinitely.

BORDERLINE (DIMORPHOUS) GROUP (B)

A malign form, very unstable; almost always strongly positive on bacteriological examination; the lepromin reaction generally negative. Such cases may arise from the tuberculoid type as a result of repeated reactions, and sometimes they evolve to the lepromatous type. The nasal mucosa often remains bacteriologically negative, even when the skin lesions are strongly positive.

The skin lesions are usually seen as plaques, bands, nodules, etc., with a regional distribution similar to that of lepromatous leprosy, except for conspicuous asymmetry. The ear lobes are likely to present the appearance of lepromatous infiltration. The lesions frequently have a soft or succulent appearance, and peripherally they slope away from the centre and do not present the clear-cut, well-defined margins seen in the tuberculoid type; they are therefore liable to be mistaken for lepromas. The surface of

the lesions is generally smooth, with a shiny appearance and a violaceous hue, sometimes (in light skins) with a brownish (sepia) background.

REACTIONAL PHASES

All forms of leprosy may go through phases of reactivation or reaction. We would particularly draw attention to three main reactional phases of leprosy, as follows:

Reactional lepromatous leprosy.—Two forms must be distinguished:

(1) Lepra reaction (of which there may be two or more varieties) consists essentially of the aggravation of pre-existing skin lesions, usually with fever and extension of the lepromatous process.

(2) Erythema nodosum leprosum is characterized by the appearance of erythematous nodular skin lesions, accompanied at times by fever, and has as a rule a favourable prognosis.

We would further draw attention to the special condition known as the "Lucio phenomenon" or "erythema necroticans," occurring only in diffuse lepromatous leprosy and more particularly in Central America.

Reactional tuberculoid leprosy.—Infiltrated lesions of active, succulent appearance, without central retrogression, develop abruptly from major tuberculoid lesions or from lesions of lesser degree (minor tuberculoid or even indeterminate), or on sites not previously involved. In some cases more or less numerous and widely scattered small metastatic nodules may appear. The lesions of the peripheral trunk nerves may become marked, and necrosis and even abscess formation may occur. On bacteriological examination, although the cutaneous lesions are found to be positive, sometimes strongly so, the nasal mucosa frequently remains negative. During the reaction the response to lepromin may decrease in intensity. Fever and constitutional symptoms do not ordinarily occur.

Reactional borderline (dimorphous) leprosy.—In reactional borderline cases the lesions show extreme oedema, erythema and desquamation. The reaction frequently extends to nerves, and marked nerve pain and dysfunction develop. The skin lesions, during this phase, may ulcerate superficially, or sometimes widely and deeply; and the skin is acutely tender. Bacteriologically the lesions are strongly positive. The lepromin reaction is usually negative.

ADDITIONAL NOTE

1. The classifying factor is mainly clinical, but it is advisable for workers to give consideration to the immunological and histo-

pathological criteria. These factors may decisively influence the placing of a case in a particular type or group.

2. Cases of the lepromatous or tuberculoid types with only recessive or residual lesions remain in their respective types. Such cases may be described as recessive or residual, and these terms should be added to the description of the main types, e.g., L(res), T(res), etc.

ADDENDA

DR. WADE registered a dissenting opinion regarding the recognition of a "macular" variety of the tuberculoid type, on the following grounds:

For classification to be understandable to all serious workers in leprosy, and not merely to the expert specialist, the line of demarcation between the tuberculoid type and the group or groups which present "simple" flat macules should be as distinct as possible. The distinction must necessarily be based on clinical aspects, primarily the morphology of the lesions, elevation and usually certain other features being characteristic of the lesser tuberculoid cases. In his opinion the inclusion, in this type, of a variety of simple macular cases, commonly known in the past as "maculo-anaesthetic," would cause much confusion. This same proposal was made by the classification committee of the Havana Congress, in that part of the report which was rejected by the Congress in plenary session.

Regarding the argument that a great majority of the lesions which it is proposed to call "macular tuberculoid" will, if active, show histologically some degree of tuberculoid change if it is sought with sufficient care, it is to be said that that change is not outwardly evident because of the relatively low degree of tissue reactivity. In keeping with that circumstance, cases with such lesions are as a rule less responsive to treatment than are frank tuberculoid cases.

Incidentally, the creation of a "macular" tuberculoid variety would increase confusion in terminology. All of the skin lesions of tuberculoid leprosy are commonly referred to by many leprologists as "macules," and the Japanese leprologists use the term "lepra maculosa" for the tuberculoid form as a whole.

For these reasons he adheres primarily to the definitions of the tuberculoid type and the indeterminate form adopted by the Expert Committee on Leprosy of the WHO, the pertinent parts of which are as follows:

(1) Cases of the indeterminate group present "flat skin lesions." The group "consists essentially of the 'simple macular' cases and comprises those cases previously known as 'maculo-anaesthetic'."

(2) Cases of the tuberculoid group present "erythematous lesions which are elevated, marginally or more extensively. . . ." and in the minor variety the "skin lesions are only slightly to moderately elevated, often at the margin or even a part of the margin, usually with irregularity of the surface."

Agreeing fully that those cases which have become established in the "maculo-anaesthetic" form should not be retained in the "indeterminate" group, he holds that they should be recognized as a distinct "group," a view which is in accord with the conclusions of a special classification committee recently set up by the Indian Association of Leprologists.

DRS. KHANOLKAR and COCHRANE hold that there exist macular dimorphous lesions which have clinical, bacteriological, immunological and histological features which justify their inclusion in the borderline (dimorphous) group. They further are of the opinion that, if a careful study is made, a pure neuritic form of the borderline (dimorphous) group could be established.

The following is a description of what is considered a dimorphous macule:

These macules show, clinically, characteristics of both the tuberculoid and lepromatous types. Their distribution is that of lepromatous leprosy; the margin of the lesion is less definite than that of the tuberculoid macular lesion, but not so vague as that of the lepromatous macule; the surface

tends to be dry and may show a wrinkled or creased appearance. On careful examination some loss of cutaneous sensibility can be elicited.

NOTE: In compiling this classification, the Committee is indebted to the report of the WHO Expert Committee on Leprosy, kindly supplied by Dr. Mario Giaquinto.