CLASSIFICATION OF LEPROSY

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Since 1931, that is to say since we have specialised in leprosy, much ink has flowed on the problem of the classification of leprosy and without much success, for leprologists are not yet able or willing to agree on one classification that might at last be adopted by all.

I. Primary Classification

At the present moment there are four primary classifications in existence, which are more or less widely accepted. They are: the classifications of the 1st WHO Expert Committee on leprosy and of Madrid which only differ in a few details; the Indian classification; the classification worked out by the Japanese leprologists; and finally Cochrane's classification. Needless to say, we have no intention of proposing a fifth.

We are of the opinion, with ROSS INNES[5] that the WHO and Madrid classifications are acceptable, in spite of several imperfections. They seem to us to be markedly clearer and more practical than the others. What are the criticisms that are most frequently levelled at them?

First of all, the nomenclature used in the primary classification is not unanimously accepted. Thus although the expression "tuberculoid leprosy" is used by the vast majority of leprologists, DAVISON and his co-workers[3] have just recently proposed its suppression, on the ground that the histological structure characteristic of this form is transient. This objection does not appear to us to be valid. The exact classification of a patient ought to be made on his admission to antileprosy treatment and it is not permissible to modify this classification "a posteriori", solely because histological changes have intervened in regressive lesions.

It has been fully established that the histopathology of the cutaneous lesions of tuberculoid and lepromatous patients is gradually modified, and before the cure is complete it is possible to detect the picture of an ordinary non-specific chronic inflammation. It would be a grave error to try at this stage to classify these patients as indeterminate leprosy (as we saw certain leprosy services doing), making the claim that the histology of their lesions is analogous with that of the pathological changes that take place in indeterminate leprosy.

It is obvious that it is not possible to reclassify a tuberculoid or lepromatous patient as indeterminate if the only reason for doing so is that the histology of the regressive skin lesions shows the picture of ordinary chronic inflammation.

While the term "lepromatous" is universally accepted, the word "indeterminate" has been strongly criticised. We cannot understand why it should be considered useless in leprosy classification. Since the descriptions "tuberculoid leprosy" and "lepromatous leprosy" are terms based on histological data, the expression "indeterminate leprosy" seems to us to be correct and intelligible, for it is based just as much on histological observations. A case of indeterminate leprosy is a patient presenting the indisputable clinical signs of leprosy, but whose lesions show the histological picture of an ordinary chronic inflammation. This picture may be called "indeterminate" if one takes into account the more distinctive "determinate" histology of tuberculoid lesions and, more markedly, of lepromatous lesions. Furthermore the definition "indeterminate" implies that we are dealing with a frequently unstable form. *On the other hand we feel there is no profit in describing indeterminate leprosy as a "group" (Madrid classification). It is in fact a clinically defined initial "form" of the disease which may either remain unchanged or evolve in the end into one of the other two forms.*

It would be unfortunate to use, as the Indian leprologists wish to do, histological definitions for the tuberculoid and lepromatous forms and the clinical definition of maculo-anaesthetic leprosy for the indeterminate form. And the more so since certain skin lesions of tuberculoid leprosy, and sometimes even lepromatous ones, may equally well be described clinically as maculo-anaesthetic.

We feel that the terms "tuberculoid", "indeterminate", and "lepromatous" ought to be retained in the primary classification of leprosy. They are already known and accepted by the majority of leprologists and it seems unlikely to us that simple and easily understood clinical definitions could be found to replace successfully the present histologically based terms. *One might all the same wonder if we ought to reserve a place for borderline leprosy in the primary classification.* Personally we consider borderline leprosy to be an unstable variety of the tuberculoid form capable either of regressing to the major tuberculoid variety or of evolving into the lepromatous form. To us it seems hardly necessary to include it in the primary classification. RAMOS E SILVA[4] tries to resolve the difficulty by dividing borderline leprosy into two groups, one predominantly tuberculoid and the other predominantly lepromatous. It seems to us that it is rather difficult to make this distinction in a primary classification. We think it would be preferable to consider borderline leprosy, as long as it remains really "borderline", as an unstable variety of tuberculoid leprosy and so being logically placed in the secondary classification.

However the WHO and Madrid classifications and also those recommended by the Indian leprologists and by COCHRANE include borderline leprosy in the primary classification. We feel that this is

an illogical procedure but strictly speaking it is admissible, since it does not cause any confusion when classifying patients. *On the other hand we cannot allow borderline leprosy to be included in the different classifications under completely different names.* Thus both the Latin-American leprologists and Cochrane prefer the terms "dimorfo" or "dimorphous", while the Indian and Japanese leprologists use "intermediate" and "atypical". This would not matter much if all the terms had exactly the same meaning, but unfortunately this is not the case.

Borderline leprosy is described by WADE as an unstable intermediate stage between major tuberculoid leprosy and lepromatous leprosy, and capable of regressing towards major tuberculoid leprosy, from which it derives, or of evolving towards the lepromatous form. Now although the Madrid classification admits this definition and gives eaxctly the same meaning to the word "dimorfo", Cochrane groups under the heading "dimorphous" not only Wade's "borderline" cases, but also patients in the intermediate phase between the clinical beginning of leprosy, which is always, according to this author, potentially lepromatous (we certainly do not share this opinion) and tuberculoid leprosy In the Indian and Japanese classifications the borderline cases are put together with cases of indeterminate leprosy in a group called respectively "intermediate" and "atypical".

It is evident that a unique word is necessary for an international classification and the most appropriate term, one which avoids confusions during the classification is Wade's term "borderline" unless the word "dimorphous" be henceforth used only as a synonym of the word "borderline".

Certain authors describe borderline leprosy as "bipolar", basing their description on RABELLO who considers the tuberculoid and lepromatous forms as the "polar" types of the disease. But in geography the north pole never changes into the south pole, and equally in electricity the positive and negative poles are not interchangeable. Thus the description "polar types" which Rabello gives to the tuberculoid and lepromatous forms of leprosy seems to us to be very questionable since poles are immutable. Now it is no longer possible to claim that tuberculoid leprosy is an immutable form which never evolves towards lepromatous leprosy. The polar conception of leprosy and, therefore, the expression "bipolar", ought to be abandoned.* It would be more logical to substitute the word "extreme" for "polar", the tuberculoid and lepromatous forms of leprosy being thus defined as the two extreme types of the disease *But we do not approve of the inclusion, proposed by Wade and by the Indian leprologists, of a pure polyneuritic form in the primary classifi-*

* Certain Brazilian authors even use the adjective "infrapolar" to describe indeterminate leprosy.

cation. We would then have in the same group patients with tuberculoid or indeterminate leprosy, as well as lepromatous cases who only show polyneuritic lesions since their cutaneous lesions have disappeared. It is inconceivable this group should be given a place in the primary classification since this classification has the precise object of defining the principal forms of the disease with a view to an orderly scientific classification of patients. And for the rest, this procedure is hardly to be recommended from a clinical point of view since the prognosis and the necessary duration of treatment differ greatly for tuberculoid, indeterminate and lepromatous patients.

We know that it is sometimes difficult to classify correctly patients who have pure polyneuritic leprosy, but this is a rare occurrence. A positive Mitsuda reaction permits us to exclude the lepromatous form, and if it is strongly positive allows us to assert that we are dealing with a tuberculoid leprosy. A weakly positive lepromin reaction, however, indicates rather an indeterminate leprosy, especially in patients with a moderate and even hypertrophy of nerve trunks. As for subjects insensitive to lepromin, indeterminate leprosy is probably what exists, unless the cutaneous stigmata or alopecia of the eyelashes indicate that we have a lepromatous patient whose cutaneous lesions have disappeared. In fact pure nerve leprosy in lepromatous cases probably does not exist, or, if it does, remains purely neuritic only for a short time since the skin is rapidly invaded by *M. leprae* in this form of the disease.

In very rare cases which cannot be classified by a result of clinical methods and the result of the lepromin reaction, the classification is helped by the histological examination of a small biopsy taken of a swollen nerve. In this way we were able to establish a diagnosis of tuberculoid leprosy in three lepromin-positive patients who showed only a single unilateral facial paralysis with a mild hypertrophy of one or of several cervical nerves. These biopsies had absolutely no harmful consequences. In two of these patients treatment with diamino-diphenylsulphone brought only a slight improvement, but in the third the facial paralysis had practically disappeared after 11 months of treatment.

We think that patients with polyneuritic leprosy, whether pure or secondary, ought to be classified under one of the three forms of leprosy—tuberculoid, indeterminate, or lepromatous. In case of doubt the patient could be placed provisionally in the group that seems the most likely one, but with a question mark until the classification has been confirmed or rejected by a histological examination.

The adoption of *a binary classification* which covers the primary classification could be achieved by using, in their biological sense, the terms "benign" and "malignant". In our opinion this binary

classification has above all the advantage of avoiding the continual repetition of the words "tuberculoid", "indeterminate", and "lepromatous" in the literature. However, this division might not fit exactly in the case of borderline leprosy, though in fact this unstable variety cannot be called biologically malignant until it has definitely evolved towards the lepromatous form. The terms benign and malignant seem to us to be preferable to lepromatous and non-lepromatous, proposed by certain authors.

On the other hand we do not advocate the use of the terms "open" and "closed" for the classification of leprosy patients. These terms would be grammatically acceptable if they were used as follows: "open" to mean that the nasal mucosa is positive or that the cutaneous lesion is ulcerated, and "closed" to mean that the nasal mucosa is negative and the cutaneous lesion is non-ulcerated. However, at present we find in the "open" group patients with only very few bacilli in non-ulcerated cutaneous lesions and also patients with strongly bacilliferous nasal mucosa and cutaneous lesions, and this seems to us undesirable.

The majority of leprologists consider that patients with few bacilli and negative nasal mucosa can to all intents and purposes be described as non-contagious. All such cases would thus be classified, under the present system, as "open" and so are subject to the sometimes irritating administrative consequences of this designation.

One could perhaps use for the Administration *in place of the terms "open" and "closed" the following expressions*, which would be more easily understood by the non-medical: (in French, "contagieux") *contagious* (with positive nasal mucosa or highly bacilliferous cutaneous lesions, above all when ulcerated); (French, "présumé non-contagieux") *presumed non-contagious* (with negative nasal mucosa, few bacilli in non-ulcerated cutaneous lesions); (French, "non-contagieux") *non-contagious* (negative to bacterial examination).

II. Binary Classification

In order that it might be universally accepted the binary classification should be simple and based principally on clinical observation. The most elementary classification would thus be to subdivide each of the three forms of leprosy into "cutaneous", "neuritic", and "cutaneous-neuritic". But the usefulness of a more detailed classification is undeniable. And thus it is necessary to attempt to define the different varieties of the forms of leprosy.

But it should always be borne in mind that there are certain intermediate and transitory stages that exist between different forms and even between certain varieties of leprosy, and which can sometimes be detected only by histological examination. In our opinion these intermediate stages cannot be considered as varieties as we

describe them, and they ought not, except for borderline leprosy, to be taken into account in the binary classification. Similarly the reactional states, whether of long or short duration, which alter, for good or for ill, the normal course of the disease cannot be classified as different varieties. The use of the terms "pretuberculoid", "tuberculoid reaction", "tuberculoid reactional transformation", "prelepromatous", "lepromatous reaction" and "nodular erythema" will permit us to describe these transitory stages of the disease.

The distinction between the different varieties of leprosy is essentially grounded on the clinical aspect of the cutaneous lesions, except of course for the purely neuritic cases.

Tuberculoid Leprosy

According to the Madrid classification this form of leprosy is divided, from the cutaneous aspect, into the three varieties "macular", "minor" and "major". We would add to this list borderline leprosy.

One may wonder whether there is any profit in considering pure macular tuberculoid leprosy as a true variety. (We would mention that in this article we are using the terms "macule" and "macular" in their strict dermatological sense.) In fact it is rare for an undoubted case of tuberculoid leprosy to show only typical macular changes. A careful clinical examination generally allows us either to detect a very mild infiltration or to recognise previously-infiltrated tuberculoid lesions that are now regressing. Besides, the most of the strictly macular erythematous lesions which are included in this variety exceptionally prove to be purely tuberculoid. They are, more often than not, pretuberculoid or even prelepromatous indeterminate lesions, whose exact nature can often only be determined by bacteriological or histological methods.

As for the terminology, "macular" is a descriptive word, whereas "minor", "major", and "borderline" indicate different degrees of the infection. So if we wish to include this variety in the binary classification it would be preferable to replace the word "macular" by a more appropriate term. The adjective "atypical" might be suitable, since the infiltration, absent from the macule, is one of the principal clinical characteristics of the tuberculoid cutaneous lesions.

Finally we prefer the term "major tuberculoid" to "reactional tuberculoid", for the latter is often confused with the expression "tuberculoid reaction" by doctors unfamiliar with leprology.

We thus have the following list of varieties of tuberculoid leprosy:

Tuberculoid leprosy

atypical	(macular, well-defined) (?)
minor	(micropapular, well-defined)
major	(infiltrated, in a plaque or a ring, well-defined)

borderline (more or less infiltrated, in a plaque or a ring, ill-defined). One may however object to this method of classification which is based principally on the degree of the infection while the terminology at present used to describe the varieties of the lepromatous form is certainly clinically descriptive.

Indeterminate leprosy.

In this form of leprosy there are, from the cutaneous point of view, no varieties, since all the lesions are strictly macular. At most one might make distinctions on the grounds of colour. But these lesions are almost always hypopigmented.

As for erythematous macules, bacteriological and above all histological methods reveal that we are most often dealing with indeterminate pretuberculoid, or even prelepromatous lesions. Finally hyperpigmented macules are extremely rare.

Lepromatous leprosy.

In lepromatous leprosy there are in reality only two cutaneous varieties: lepromatous leprosy with figurate lesions and diffuse lepromatous leprosy.

However, we may find cases, of ordinary recent lepromatous leprosy, with nothing but figurate lesions of the same type. It will therefore be of use for the prognosis and for the assessment of therapeutic results to classify such patients in a more precise way. To do this we might subdivide the variety "figurate" into "papular", "macular", "nodular", and "infiltrated".

But such patients (that is showing skin lesions all of the same type), are relatively rare. Most lepromatous patients have, in varying proportions, skin lesions of widely differing types. And this subdivision could only be applied to them with difficulty. But one could then specify that a certain type of lesion is "predominating".

We may thus list the following binary classification for lepromatous leprosy:

Binary Classification
Lepromatous Leprosy

figurate		*diffuse*
papular macular nodular infiltrated	"pure" or "predominating"	

The essence of this study of leprosy classification is summarised in Table I which is appended. So as not to overload the scheme we have not mentioned the bacteriological, immunological, and histological features of the different varieties and forms of leprosy. Besides, these features are not now in question. Those varieties

which do not seem to be absolutely indispensable have been marked with a question mark.

Conclusion

An acceptable classification of leprosy could be rapidly decided on if leprologists would agree to remove from consideration certain regional or personal preferences, to which it is hard to attach any real importance. And this result could be achieved easily since no doctrinal differences exist in clinical, immunological, or histological aspects. It is high time that we attained such a result for it is hard to believe that only a few years from the 90th Anniversary of the discovery of the bacillus by ARMAUER HANSEN, leprologists are still searching for an acceptable classification of leprosy.

Acknowledgement

We are very grateful to Dr. J. Ross Innes for arranging for the translation into English of this article, which was submitted in French.

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TABLE I

CLASSIFICATION OF LEPROSY

TUBERCULOID	INDETERMINATE	LEPROMATOUS
CUTANEOUS	CUTANEOUS	CUTANEOUS
atypical (macular) (?) *minor* (micropapular) *major* (in a plaque or a ring, infiltrated, well-defined) *borderline* (in a plaque or a ring, more or less infiltrated, ill-defined)	*macular* (?) (hypopigmented, and rarely erythematous or hyperpigmented)	*figurate* papular, macular, nodular, infiltrated } "pure" or "predominating" *diffuse*
NEURITIC	NEURITIC	NEURITIC
pure *secondary*	*pure* *secondary*	*pure* (?) *secondary*
CUTANEOUS-NEURITIC	CUTANEOUS-NEURITIC	CUTANEOUS-NEURITIC