# CLASSIFICATION OF LEPROSY CASES.

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The importance of a detailed and reliable method of classification and case-taking cannot be too much emphasised. Without this the progress of the disease cannot be correctly traced, and a true, continuous picture of its course arrived at. Also, now that more effective forms of treatment are becoming available, it is even more necessary to be able to estimate with precision the signs of gradual recovery.

It is equally important to be able to compare cases of diverse races and those living in different countries, or under dissimilar climatic, dietetic or social conditions. Without a precise and universally recognised system of typing and case-taking an accurate comparison is impossible.

There are six chief criteria by means of which cases of leprosy may be classified: Clinical, topographical (skin, nerves etc.), histopathological or structural, bacteriological, immunological (lepromin test), reactional.

## PRIMARY CLASSIFICATION.

A clinical classification was originally used and, as far as it goes, this is the simplest and most convenient. But clinical aspects are so numerous, and often so confusing, that we require a more reliable basis of grouping, a final court of appeal when the clinical, topographical and even bacteriological findings leave us in doubt. The microscopic structure of the lesions gives us a standard which, though not always entirely satisfactory, is at least the best available.

What is known as the Panamerican Classification* divides leprosy primarily according to the histological picture into three types. In two of these types, the lepromatous and tuberculoid, the picture is characteristic and distinctive, but in the third it is uncharacteristic and indistinguishable from other chronic inflammatory conditions.

The microscopic structure thus provides a simple and reliable basis of primary classification. But it is only in a well-equipped laboratory and in the hands of a trained pathologist that this criterion can be applied, and these facilities are not available in the majority of cases.

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*Adopted at the Panamerican Conferenec held in Rio de Janeiro in October, 1946.

It is generally necessary therefore to use a combination of the other more easily available criteria, especially the clinical signs, bacteriological findings and, where possible, the lepromin test to find out the structural type to which the case belongs.

It is important, therefore, to discuss in turn the usual clinical, bacteriological and lepromin findings associated with each of the three structural types: lepromatous, tuberculoid and uncharacteristic.

(1) The structural picture of *leproma*, with its Virchow or foamy lepra cells surrounded by small round cell infiltration, is not confined to the skin or subcutaneous tissue; the same picture is found in the mucosa, nerves and internal organs.

The lepromatous case has in its advanced stages certain clinical appearances which mark it out from the other two types, namely diffuse thickening of the skin (especially of the face, arms and legs), nodulation, and loss of supercilliary hair.

In early cases the clinical appearance is less distinctive, but the nature of macules, their shape, size, number and distribution, and especially their central thickening and absence of a raised or incisive margin, are at least an indication of the lepromatous type, Nerve thickening is not nearly as marked as in the tuberculoid.

The bacteriological smears are most important. Globi are characteristic of this type, and a case with many bacilli is almost certainly lepromatous unless it be a reacting tuberculoid, when the clinical peculiarities and the lepromin test will, as a rule, give a clear distinction. The lepromin test is invariably negative.

Acute reaction is generally much more severe in the lepromatous than in the tuberculoid type, and this is natural considering the infinitely greater number of bacilli present.

(2) The *tuberculoid* structure, with its denser cellular formation in the shape of follicles or cords with clear-cut edges, and with its epithelioid and giant cells surrounded by round cell infiltration, is found in the skin and peripheral nerves alike.

Clinically, the most typical skin lesions (leprides) show a tessilated appearance, due to small tubercles one or two milimetres in size, which stand out from the skin surface especially at the advancing margin, but coalesce and often flatten out in the older and more central part of the lesion. The tubercles correspond with the cellular cords which press on and cause projection of the epithelium.

The peripheral nerves are most affected in the tuberculoid type and may be markedly thickened and tender; they sometimes even caseate and form abscesses. There is in consequence a greater

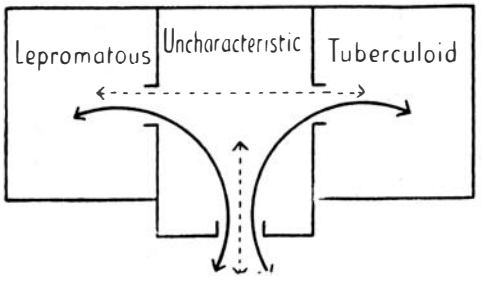
degree of anaesthesia, and the distal parts of the limbs suffer more from trophic and sensory changes.

When lesions are in acute reaction a few or more bacilli may be easily found, but they often disappear as the reaction subsides. Otherwise bacilli are few and hard to find.

The lepromin test is almost always positive, generally giving a fairly strong reaction.

(3) Unlike the first two types which are sometimes described as " polar " because of their strongly divergent signs, the third type does not show a characteristic histological picture. There are neither foamy Virchow cells, nor yet epithelioid and giant cells, only the small cell infiltration common to the other two types and to most chronic inflammations. It has therefore been called " uncharacteristic."

Uncharacteristic cases may be *static*, remaining in this type throughout the course of the disease and never passing on into another type, till perhaps in the end they recover. Or they may be in one out of three forms of transition: (a) *initial*, in the course of passing on into the tuberculoid or the lepromatous type; (b) *intermediate*, passing from one of the characteristic types into the other, or (c) *vestigial*, passing out of one of the characteristic types on the way towards recovery. In initial cases, it may be compared to the entrance hall of a house (see diagram) with doors opening to the right (tuberculoid) or to the left (lepromatous). This is the hall of anergy. If effective allergic response comes in time before the bacilli are too many, the case enters to the right (tuberculoid); failing this it may gradually drift to the left (lepromatous) as there is no effective check to bacillary multiplication.




But in the actual classification of a case at any one time we are not concerned with change and transition. We have to dot his present position, though a series of such dots made at subsequent examinations may indicate the curve he has followed, or chart his course in the house of classification.

The chief indication of an uncharacteristic case is one of exclusion, its failure to belong to either of the other two types.

Clinically the lesions are large or small, reddish or whitish (or a combination of the two) in comparision with the surrounding skin, flat and thin when picked up between the finger and thumb, and without the thickened leproma or nodule of the first type or the outstanding and thickened tubercle-formation of the second.

Initial lesions are generally limited in size and require a good light to recognise them. They would often be missed were it not that knowledge of leprous relatives or other contacts has led to careful examination. Changes in sensation are slight as a rule and require careful testing. Such lesions may disappear, to be followed later by a more serious form of the disease.

Residual lesions on the other hand are often more extensive and more easily recognised. Anaesthesia is more marked and there may be definite polyneuritic signs in the extremities and the face, and thickening and tenderness of the nerves.

Bacteria are nil or few in number, and the lepromin test varies from negative to moderately positive.

Unlike the other two types, the uncharacteristic one seldom, if ever, shows a reactionary phase; when reaction does occur it is generally a sign that the case has passed on into one of the two polar types.

Regarding transition from the *one polar type to the other*, there is a difference of opinion, Some hold that the tuberculoid is never transformed into the lepromatous, others that it is not an infrequent occurrence. This is a matter which requires further investigation. There seems little doubt that leprosy varies considerably in different countries, or even in different parts of one country, and this may, at least in part, account for the divergence of view.

The writer's opinion is that this phenomenon does undoubtedly occur, in fact there are many cases in which this has been checked up histologically, clinically, and by the lepromin test. It is not uncommon to find an advanced lepromatous case with one or more circular limited patches which mark the site of former definite tuberculoid lesions, and which still maintain their resistance to invasion from the surrounding lepromatous skin.

Passage from lepromatous to tuberculoid is more doubtful, as would be expected. Definite lepromatous cases not infrequently become arrested in the sense of becoming and remaining bacteriologically negative as far as careful routine examination is concerned; but it is a less common occurrence for such a case to devolop a strong or even moderately positive lepromin test, such as would be expected if it were transformed into a tuberculoid. Whether this

(transformation into tuberculoid type with positive lepromin test) will occur in future under treatment with sulphones remains to be seen. Certainly such a transformation in the course of recovery would prognostically be of great value, as showing that the natural resistance of the body to leprosy had increased.

### SECONDARY CLASSIFICATION.

It is important after making the primary or basal structural classification with the help of the other criteria, to pass on to secondary classifications, so as to describe the case in greater detail.

*Clinical and Topographical.* In sub-classification we consider first the various organs affected: skin and subcutaneous tissue, mucosa of nose, mouth and throat, the nerves and polyneuritic changes, the internal organs, etc.

The following is a brief list of points to be noted in making a clinical examination:—

(1) *Skin lesions* are as a rule the most important, whatever the primary type. Macules and leprides should be described as to their number, size and distribution; their colour, thickness, centre and margin; the presence of tubercles, keratosis and changed superficial markings; loss of hair and sweat function.

Diffuse lesions should be delineated, giving their extent and the changes from the normal as above; and the presence, size and number of nodules should be specified. Lesions of the subcutaneous tissue should be mentioned.

(2) *Mucous membranes* affected, of nose, mouth, throat, air passages, should be described.

(3) *Nerves.* An account should be given of tenderness, thickening, nodulation, caseation or abscess formation of peripheral nerves, and of secondary, trophic and sensory changes in the hands, feet and face.

(4) *Internal Organs* are often affected, but clinical signs such as enlargement of the liver and spleen are seldom apparent. Atrophy of the testicles, followed by gynecomastia, is a not uncommon occurrence in advanced lepromatous cases.

(5) Of the *special sense organs,* apart from those of the skin, the eye is the most seriously affected. Careful examinations, if necessary with the use of atropine, should be made for early signs of conjunctivitis, keratitis and irido-cyclitis. In tuberculoid and uncharacteristic cases,

lagophthalmia and consequent changes in the eye-ball and adnexa should be kept in mind.

In all clinical examinations the presence and degree of reactive signs, whether chronic, subacute or acute, should be noted.

*Bacteriological Examination.* It is necessary to note not only whether a case is bacteriologically positive or not, but also the degree of positivity; whether there are globi, single bacilli or peculiarities in staining. The distribution of areas found positive should be noted, in the nose, in clinically apparent skin lesions or in seemingly normal skin. Such details are of increasing importance, not only because they distinguish lepromatous from reacting tuberculoid cases, but because in the new and more effective forms of treatment the chief criterion of improvement is bacteriological rather than clinical.

*Lepromin Test.* In one sense this test is the most important of the criteria available for the practical classification of leprosy. More than anything else it indicates the power of the tissues to react to and destroy the lepra bacillus.

The results of the test vary in three respects:

(a) Variation according to the type of case and the individual.

(b) From time to time the degree of positivity varies in the same case. It should therefore be repeated frequently, if possible every three months.

(c) There may be a distinct difference in the degree of reaction inside and outside of a skin lesion at any one time.

For the sake of both primary and secondary classification it is important that the lepromin test be done accurately. For this it is necessary to have a standard antigen, and a standard method of reading results. These standards should be fixed by an accepted authority, such as an international congress.

A standardised test would make it possible to compare cases all over the world, and might also be of great value in confirming recovery if lepromatous cases under new forms of treatment were found to develop a positive result.

## Case-Taking.

In taking a case a great many details may be gathered under the descriptions suggested above, and especially under the clinical definitions.

For practical purposes, however, I propose the use of three tabular forms, the first being of a general nature, the second describing circular skin lesions, and the third dealing with nodules,

FORM 1.

| | *Skin and Mucosa.* | | | | *Nerves and Polyneuritic.* | | | | *Bact.* | | | | *Lepromin.* | | | | *Reaction.* | | | |
|---|---|---|---|---|---|---|---|---|---|---|---|---|---|---|---|---|---|---|---|---|
| | - | + | ++ | +++ | - | + | ++ | +++ | - | + | ++ | +++ | - | + | ++ | +++ | - | + | ++ | +++ |
| Lep | | | | | | | | | | | | | | | | | | | | |
| Un | | | | | | | | | | | | | | | | | | | | |
| Tub | | | | | | | | | | | | | | | | | | | | |

ulcers and the nose and eyes. These include the main elements in the classification, but they can, if so desired, be elaborated in further detail elsewhere in the patient's chart.

In Form 1 the three primary types are given in longitudinal columns. The first two vertical columns are clinical, giving the degree of affection of skin and mucosa, and nerves and polyneuritic lesions, while the other three deal with bacteriological examination, lepromin test and reaction. Each vertical column is subdivided into four for negative, and one, two and three plus.

In the first column, negative would mean that there were no lesions of the skin and mucous membranes of nose, mouth, etc. One plus = lesions covering in aggregate not more than 150 sq. cm. (25 sq. inches), two plus covering up to one quarter of the body surface, and three plus more than that area.

In the second column, one plus indicates thickening and tenderness of one or two nerves without trophic changes in hands, feet or face; two plus means involvement of more than two nerves and/or trophic changes of the face or one hand or one foot; three plus indicates more extensive neural or trophic involvement.

In the bacteriological column one plus indicates bacilli present, but no globi and not more than 10 bacilli in any one field of a smear taken by biopsy from nose or skin by the ordinary routine method. Two plus indicates one or more globi and/or more than 10 bacilli in any one field. Three plus indicates more than ten globi in the whole of any ordinary skin or nasal smear.

In the lepromin column, one plus indicates in the immediate reading a flare up to 5 mm. diameter and/or in the delayed reading a nodule up to the same diameter; two plus up to 7 mm. in early flare and/or delayed nodule, but without tissue distruction at the centre; three plus more than 7 mm. and/or tissue destruction at the centre.

In the reaction column, minus indicates no reddening or local swelling or thickening of the skin or mucosa, and no thickening or tenderness of a nerve. One plus would indicate one or more erythematous macules or other lesions without anything of the nature of a flare up, and/or thickening and tenderness of minor degree in nerves. Two plus indicates lepra reaction of moderate degree in either lepromatous or tuberculoid type. Three plus means a severe reaction with considerable swelling of lesions, fever and/or liquifaction of lesions and, in tuberculoid cases, severe and painful swelling of leprides and/or nerves.

## FORM II.

*Circular Skin Lesions.*

1. Number −, 1 to 10 or 10+.
2. Area index in sq. cm.
3. Area central flattening in sq. cm.
4. Margin breadth in cm.
5. Margin serration − to +++.
6. Hypopigmentation + or −.
7. Anaesthesia − to +++.
8. Analgesia + or −.

Form II deals with more or less circular, circumscribed lesions such as macules and leprides which need more detailed description than is given in Form I. The following eight points should be noted.

1. Number of discrete lesions, viz. 1, 2, 3, 4, 5, 6, 7, 8, 9, 10, and 10+ when more than 10.
2. Area index. This is arrived at by measuring in centimetres ($2\frac{1}{2}$ to 1 inch) the greatest diameter of the largest discrete circular lesion, and multiplying it by the length of the diameter at right angles. If no discrete skin lesions, mark '−'.
3. Area of central flattening. Here again take the largest discrete lesion, and, if there is a flattened area in the centre, measure its size in the same way, otherwise mark '−'.
4. Margin breadth. When there is a flattened centre measure the breadth of the margin in cm.; otherwise mark '−'.
5. Margin serration. Serration of the edge is one of the clearest signs of activity and spread. Mark the degree of serration negative or one to three plus.
6. Colour changes. Hypopigmentation is due to loss of pigment, to be marked + or −. Reddening is due to reaction and is marked in Table I.
7. Anaesthesia to light touch, as tested with a feather. Ask the blind-folded patient to place his finger on the points touched. This can be graded from − to +++ by using three grades of touching instrument.
8. Analgesia. This can be tested by the two pin method. A healthy and a suspected area being pricked simultaneously, and the patient asked each time which he feels most.

## FORM III.

Nodules −, 1 to 10 or 10 +.

Ulcers. Nodular −, 1 to 10 or 10 +.
Trophic −, 1 to 10 or 10 +.

| | | |
|---|---|---|
| Nose condition: | | R. −, +, + +, + + +. |
| | | L. −, +, + +, + + +. |
| Eye. | Lepromatous. | R. −, +, + +, + + +. |
| | | L. −, +, + +, + + +. |
| Eye. | Trophic. | R. −, +, + +, + + +. |
| | | L. −, +, + +, + + +. |

In Form III other details are given:— the number of nodules, and of lepromatous or trophic ulcers, the condition of the nose, and lepromatous or trophic eye changes.

In the nose condition + indicates infection without nodulation, + + nodulation, partial blocking, but no ulcers, + + + ulceration and blocking.

In lepromatous eyes + means conjunctival affection with punctate keratitis, or slight ground-glass appearance spreading from the limbus, and/or pupil irregular but free; + + = nodulation of the cornea and/or fixed pupil; + + + = vision destroyed.

In trophic eye conditions + = lagophthalmia, + + = ulceration of cornea or scars of former ulceration, + + + = vision destroyed.

## SUMMARY.

1. A classification of leprosy, following that adopted by the Panamerican Conference in Brazil, is explained and expanded.

2. Primarily there are three types: lepromatous, tuberculoid and uncharacteristic, based on the histological picture, but usually indicated by clinical, bacterioscopic and lepromin findings.

3. Subclassification gives further clinical, topical, bacteriological and lepromin details.

4. A simple method of case-taking in three tabular forms is suggested.