Current Practice

MEDICINE IN THE TROPICS

Leprosy

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Every doctor already working abroad or thinking of working abroad should familiarize himself with leprosy—a disease frequently overlooked or misdiagnosed, often neglected, and generally feared. The busy medical missionary or general duty medical officer, beset with milling crowds of outpatients with readily diagnosable infections and parasitic diseases and facing a long operating list comprising gross and remediable pathological conditions, may be understandably reluctant to spend time on a nondescript hypopigmented skin macule or an ulcerating foot.

Leprosy is a disease of insidious onset and slow and undramatic progress. It is, moreover, largely hidden, and the extent of its ravages in the community is usually underestimated—sometimes seriously. Yet, given a few basic principles, leprosy is generally not difficult to recognize despite its bewilderingly variegated pattern; nor is it in the main difficult to treat, though dealing with some of its manifestations may tax the resource and knowledge of the expert. Once word gets around that the doctor is doing something for those suffering from leprosy, patients begin to flock from nowhere. If, in addition to his hospital practice, the doctor in the tropics is concerned—as he should be—with preventive medicine, nutrition, and public health, and can integrate the treatment and control of leprosy into rural and urban schemes through trained auxiliary personnel, then he faces an opportunity as rewarding as it is challenging.

Diagnosis

Leprosy is frequently missed because not only does the doctor not think of it when dealing with a skin or nerve condition, but also because he may not be sufficiently acquainted with its salient and pathognomonic features. He should consider leprosy whenever confronted by a chronic and symptomless skin rash that does not correspond with the common dermatoses with which he is familiar, or which does not respond to the standard treatments he usually prescribes for similar lesions. Leprosy should also be considered in all cases of transient, recurrent, or persistent numbness or paraesthesiae, specially when this is localized to a more or less well-defined area of skin.¹

The history may or may not help. When the patient has been in close contact at any time previously with a person known to have leprosy—especially three to five years previously, and particularly if the index case has open leprosy—suspicions should be aroused. Frequently, however, such a history of contact may be denied through ignorance, fear, or shame.

The earliest symptoms, as indicated above, are usually neurological. They may precede by months—rarely by years—

visible changes in the skin or demonstrable impairment of function. Until such changes do occur it is not justifiable to diagnose leprosy or to treat the patient for leprosy. All such patients should be re-examined every three months.

The presenting signs in the skin may be varied and misleading, but most of them fit into well-defined patterns. The practising doctor would do well to build up a composite mental picture of the range of skin pigment among his patients and the appearances of the commoner dermatoses. He should get to know the common causes of local or general variations in depth of pigmentation, and the effect of sun and wind. As a rule, the skin lesions of leprosy are more easily recognizable in pigmented races than in Caucasians. The lighter the skin the more noticeably pink or violaceous are the typical leprosy lesions.

Indeterminate Leprosy

The commonest early skin lesion in leprosy is the one least often seen, except by those who do regular whole population surveys in highly endemic areas. Doctors may come across it in patients complaining of something else. It is a single small, slightly hypopigmented macule, with either vague or well-defined limits, set anywhere on the trunk or limbs. There is no demonstrable impairment of function in skin adnexa or nerves in the macule; notably, there is no anaesthesia to light touch. Another common mode of onset is an area of vague hyperaemic mottling. On ordinary methods of examination *Mycobacterium leprae* cannot be demonstrated in these lesions, though they can be found after prolonged examination of serial sections. These lesions are indeed leprosy, and often disappear without treatment. When the diagnosis is made, however, the patients should be treated, since it is impossible for non-specialists to predict which lesions will not resolve spontaneously. These early indeterminate lesions may persist as such, or may become more numerous and develop features of the more determined kinds of leprosy.

Tuberculoid Leprosy

If the patient's innate resistance to *Myco. leprae* is high, the edges of the lesion become well defined and papular, the depigmentation more marked, and indubitable signs of local nerve damage appear—for example, diminution in sensitivity to touch, to heat, and then to pain. The hair is lost, sweating is impaired, and the skin surface becomes dry and rough. Anaesthesia to light touch in a hypopigmented area of skin virtually means leprosy. Sometimes as the finger nail is passed around such a lesion it encounters a nerve fibril, greatly enlarged and tender. The lesion may slowly extend peripherally, healing and repigmenting in the centre, or successive crops of similar lesions may appear, producing an extremely varied clinical picture. Some or all of these skin lesions may pass through

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phases of acute inflammation during which they become red and succulent, erysipeloid, and perhaps desquamating and ulcerating around the raised margins. Only at this stage are *Myco. leprae* demonstrable in the lesions by standard methods.

Lepromatous Leprosy

Lepromatous leprosy usually presents as a rash composed of numerous small vague hypopigmented macules scattered diffusely over the trunk, face, and limbs. These macules are visible only in a very good light, preferably sunlight falling obliquely on the tensed skin. Slight differences in texture, such as shininess and pigmentation, should be looked for. Ordinarily there is no loss of tactile sensation in these lesions, but the bacteriological examination is almost always highly positive.

The signs of late lepromatous leprosy are so well known that they should present no difficulty in diagnosis: in nodular lepromatous leprosy, discrete or confluent masses of granulation tissue appear on the ears, face, trunk, and limbs; in diffuse lepromatous leprosy, a generalized infiltration leads eventually to the thickened corrugated skin of the leonine facies.

Intermediate Forms

The various intermediate forms of leprosy (borderline or dimorphous) present skin lesions in great variety—particularly plaques showing greater elevation in the centre than at the periphery, and wide raised succulent rings surrounding flat "immune areas." Slight impairment of tactile sensation in the lesions is the rule.

Acute Exacerbation

Patients who have had symptomless leprosy for years may present themselves when—because of some intercurrent illness, the ingestion of some drug, smallpox vaccination, mental shock, or parturition—they suffer a sudden attack of erythema nodosum. The rash affects the forearms, arms, thighs, face, and trunk, in that order, and may be superficial or deep, and come in a single attack or in recurring crops. Perhaps an acute iridocyclitis or painful polyneuritis makes the patient seek medical aid. He may be ill, with raised temperature and severe malaise, polyadenitis, arthralgia (perhaps with effusion), sudden anaemia, orchitis, and painful gynaecomastia.

Peripheral Nerves

In addition to the gross enlargement of unnamed small superficial nerves in direct relation to a leprosy lesion, the peripheral nerve trunks are enlarged, hard, and tender at certain sites of predilection: that is, the ulnar for some inches above the elbow, the posterior tibial midway between the internal malleolus and the point of the heel, the lateral popliteal as it winds round the neck of the fibula. The great auricular, the median, the musculo-spiral, the facial, the brachial plexus, etc., may also be involved. These abnormalities occur early and are widespread in borderline leprosy, localized and slight in tuberculoid leprosy, and generalized and late in lepromatous leprosy. Very rarely cases are seen where the nerves show the typical changes but no leprosy lesions are present in the skin.

Patients sometimes delay presenting themselves for treatment until they show the results of longstanding peripheral nerve damage, such as plantar ulceration (perhaps associated with dropped or everted foot), clawing of the hand from intrinsic paralysis, Bell's palsy with lagophthalmos, etc. Damage to anaesthetic fingers and toes may be more marked than muscular wasting.

Special Investigations

Skin Smears.—The demonstration of *Myco. leprae* in skin smears confirms the clinical diagnosis of multibacillary leprosy, but the absence of bacilli does not of course exclude those kinds of leprosy in which bacilli are either extremely scanty or no longer present.

The skin over the edge of the most active-looking skin lesion, and of the earlobe, is cleansed with spirit, compressed firmly between the thumb and forefinger, and incised for $\frac{1}{2}$ cm. with a sharp scalpel—the incision just penetrating the dermis. The tip of the blade, turned through a right angle, is drawn once or twice firmly along the slightly gaping edges of the incision. The tissue fluid on the tip of the scalpel is spread on a new clean microscope slide, fixed by heat, and stained by Ziehl-Neelsen's method. The presence, concentration, and morphology of *Myco. leprae* are shown by examination under oil immersion.

Biopsy.—When confirmation of the type of leprosy is required, a small portion of skin (deep enough to include some fat) is removed, fixed in Zenker's or Ridley's solution, and processed to show both the cellular response to infection and *Myco. leprae*.

In indeterminate leprosy, the histological picture is unhelpfully non-characteristic; in tuberculoid leprosy it shows a focalized tuberculoid granuloma involving nerve fibrils and adnexa, and in lepromatous leprosy a cellular granuloma replete with *Myco. leprae* in reticulo-endothelial cells.

Lepromin Reaction.—Since the lepromin reaction does not indicate past or present leprosy infection, it is only very rarely of use in the differential diagnosis of a suspected lesion. It is, however, of value in confirming or correcting the clinical classification, and in indicating the prognosis.

Differential Diagnosis

Leprosy is the great simulator, and the conditions that have at times been confused with leprosy are legion.² Most of these are readily excluded by greater knowledge of the typical presentations of these conditions, a closer acquaintance with leprosy, and the grounds for its positive diagnosis. This knowledge, a wisp of cotton wool (for testing tactile sensation), and a scalpel (for making skin smears) should enable the non-specialist practitioner to diagnose the vast majority of cases of leprosy.

If the lesions conform to the clinical pattern described—if either tactile sensation in the skin lesions is impaired, or the skin smears show numerous acid-fast bacilli, and if peripheral nerve trunks are enlarged, hard, and tender—the diagnosis of leprosy is certain. All these features may not be present in early or atypical leprosy, and less stringent criteria for a positive diagnosis are acceptable in practice.

Macular Lesions.—By far the commonest cause of confusion is some form of mycosis of the skin—frequently irritating and desquamating, sometimes with irregularly papulate margins, and even showing impairment of tactile sensation. If in doubt after careful clinical examination, scrape the surface with a sharp scalpel and put the material in 20% caustic potash solution on a microscope slide; examine it at once under reduced light, and again (having ringed the cover-slip with petroleum jelly) after 24 hours, for mycelial elements. A therapeutic test with an antifungal preparation may also help.

Other common causes of error are congenital or acquired hypopigmented areas of skin, varying from persistent Mongolian spots to bizarre patterns of vitiligo and naevi; from framboesial depigmentation to onchocercal achromia. Here again, better acquaintance with these sources of error will serve to exclude them. Of greater difficulty is nutritional hypochromia of the skin, specially when associated with some form of neuropathy. The hallmarks of this confusing condition are ill-defined, slightly scaly, and slightly hypopigmented macules on the cheeks, trunk, or limbs, tending to be symmetrical and occurring at certain seasons among the ill-nourished. The

powdery macules of seborrhoeic dermatitis, vague and hypopigmented on the dark facial skin, and occurring frequently among the educated (and hypochondriac), are also common sources of error.

Nodular Lesions.—When occurring in other diseases these rarely cause confusion with the nodules and papules of lepromatous leprosy; Kaposi's sarcomatosis, cutaneous leishmaniasis, onchocercosis, and framboesial juxta-articular nodules are all excluded by their own typical presentation and by the absence of the positive criteria for the diagnosis of leprosy.

Peripheral Nerve Lesions.—Traumatic or toxi-infective peripheral nerve lesions are almost never (in tropical practice) characterized by enlargement, hardness, and tenderness of the nerve trunks at the sites of predilection. The combination of sensory and motor damage excludes many neuropathies. In leprosy the peripheral neuropathy is generally associated with skin lesions of characteristic aspect, active or healed.

Management

Once the diagnosis has been made, classification determined, and the patient placed in either the "open" or the "closed" administrative category, the questions of general management and treatment must be faced.

No special advice is required regarding diet. Any intercurrent diseases and parasitic infections should be treated, and anaemia corrected.

All patients with active leprosy (except those presenting with acute polyneuritis, or in an acute reactive phase, in which cases treatment should be deferred) should be treated, and the great majority can be treated with perfect safety to themselves and their families as outpatients in general hospitals. Admission to hospital is reserved for patients showing drug sensitivity, or needing stabilization of drug dosage or transfer to another drug, and for patients in acute exacerbation (especially with acute polyneuritis or iridocyclitis).

Where hospitalization is necessary on other grounds, patients suffering from open leprosy may be admitted to general wards and nursed with a modified barrier nursing technique. No special precautions are needed in respect of patients suffering from closed leprosy, as are the majority needing reconstructive surgery.

Medical attendants need not wear gloves or take prophylactic drugs; cleansing the hands with soap and water is adequate, and a gown should be worn. Patients with open forms of leprosy who have been under treatment for a year may as a rule be regarded as no longer contagious. (Fragmented and non-viable bacilli may persist for years and may even provoke new "leprosy" lesions.)

Household contacts should be examined every three months, and child contacts given B.C.G. vaccination. Pregnant and parturient patients should continue treatment and be more closely supervised medically, since leprosy may become worse at these times.

Drugs.—The standard and best drug is dapsone (diaminodiphenyl sulphone). The dose regimen advised for an otherwise healthy adult weighing 50 kg. (110 lb.), and with a haemoglobin level of at least 9 g.%, is as follows:

1st month	...	25 mg. once weekly
2nd month	...	50 mg. once weekly
thereafter	...	100 mg. once weekly

Doses for children (who take the drug well) are in proportion. Increments should be made cautiously in borderline disease. Once the dose has been stabilized, most patients need be seen only every month, or even less frequently.

Duration of Treatment.—In closed leprosy (indeterminate or tuberculoid) treatment should be given for at least two years, or at least one year after all clinical signs of activity have disappeared.

In open leprosy (lepromatous and borderline), treatment is needed for at least four years, or at least two years after all clinical signs of activity have ceased, and acid-fast bacillary material is no longer present in skin smears. ("Clinical activity" in this sense means any extension of existing lesions, or the appearance of new lesions; persistent redness or elevation of the lesions; or an increase in the signs of peripheral nerve damage.)

Intelligent and co-operative patients who have had open leprosy should take half the therapeutic dose for life after clinical arrest of the disease and discharge from full treatment.

In cases of sensitivity or resistance to dapsone, when dapsone even in minute doses apparently precipitates acute exacerbation, in dapsone "psychosis" (very rare), and in acute peripheral neuritis, another drug (such as thiambutosine, thiacetazone, a long-acting sulphonamide, or a phenazine derivative, B 663, Geigy) may be substituted. Details of dosage, etc., should be sought in standard works.²

Acute exacerbation of leprosy, particularly lepromatous leprosy, calls for cessation of antileprosy treatment, bed rest, local rest to inflamed tissues (such as peripheral nerves, the uveal tract, etc.), and analgesics. If after two weeks the patient is still ill, antimonials may act dramatically. If after a further two weeks the condition is not controlled, a fortnight's course of chloroquine (400 mg. base every day) is advised. If the patient is still unrelieved, a short course of a corticosteroid preparation such as prednisolone (beginning with 30–60 mg. daily in divided doses, and rapidly diminishing) may be effective, but the dangers of such therapy must be faced. Sometimes a small (10 mg.) daily dose of prednisolone may be required to suppress the distressing signs of exacerbation, and dapsone medication may be cautiously resumed under such cover.

Resumption of treatment after the interruption made necessary by the acute exacerbation must be slow and tentative: dapsone should be given as at the beginning of treatment, with less frequent and smaller increments.

Surgical Treatment.—There is little call for surgical skill in the management of uncomplicated leprosy. Acute footdrop will usually recover if treated expectantly, and sudden facial paralysis is better treated with corticosteroids than by operation. Severe tenderness of a swollen peripheral nerve trunk, persisting for more than a few weeks and getting worse, is relieved by injection into the fibrous sheath of either 5 ml. of 1% procaine or the same solution with added hyaluronidase. If this procedure is unsuccessful, longitudinal incision of the sheath under local anaesthesia will relieve pain and may prevent neural damage.

Most busy doctors tackling the varied medical and surgical problems of a hospital practice in the tropics will acknowledge that reconstructive surgery for leprosy patients demands time they cannot afford and expertise they do not possess. However, such relatively simple and non-time-consuming procedures as tibialis posticus transfer for drop foot, triple arthrodesis stabilization for the flail foot, and tarsorrhaphy for lagophthalmos give better results than a complicated hand tendon transfer when the latter is undertaken infrequently.³ Walking-plaster-casts will allow the healing of neuropathic plantar ulceration, and simple protective footwear should keep the ulcer healed. Cosmetic and plastic operations for conditions such as severe lagophthalmos, sunken nose, madarosis, redundant earlobes, sagging face, embarrassing gynecomastia, obviously enlarged nerves, wasted thumb-web, etc., may be undertaken where time and opportunity permit.

The importance of team-work—nurses, physiotherapists, shoemakers, and social workers—needs no emphasis. Nor should the need for education be overlooked—education of the patient in the protection of anaesthetic extremities, of the family and community in revising their attitude towards leprosy, and of public health authorities to ensure that this slightly contagious

and severely crippling disease is accorded the attention that its importance merits.

If leprosy is diagnosed early enough and is treated properly, nerve damage can generally be prevented, social dislocation avoided, and the cycle of transmission interrupted. Every doctor working in the tropics should strive towards this ideal.

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Seamless Shoes in Rheumatoid Arthritis—Preliminary Trial

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In each foot there are approximately 30 joints or tendon sheaths which may be affected by rheumatoid arthritis, and patients often complain of pain on walking. In early disease pain is usually reduced by prescribing commercially obtainable shoes large enough to contain both the swelling of the foot and special insoles which redistribute the body weight from the painful metatarsophalangeal joints to relatively painless parts of the longitudinal arch.

As disease progresses numerous deformities develop. In typical late rheumatoid arthritis they include hallux valgus with broadening of the forefoot, dorsal subluxation or dislocation of the second to fifth toes at the metatarsophalangeal joints, valgus deformity at the subtalar joint with collapse of the longitudinal arch, and, less often, involvement of the ankle joint itself with painful limitation of flexion and extension. Under the metatarsophalangeal joints the fibrofatty "cushion" moves forward, leaving the bones with little protection between them and the plantar skin. Painful callosities form beneath the metatarsal heads (and may break down to form sinuses) while shoe pressures cause similar callosities on the dorsum of the cocked-up proximal interphalangeal joints. Contractures of the hips and knees also affect the way in which the foot is used and hence the ability to obtain comfortable shoes.

In Great Britain the National Health Service provides surgical shoes, free of charge, on the prescription of a hospital consultant. This is a service of great value. Unfortunately, we noted that a proportion of shoes prescribed for patients with rheumatoid arthritis could not be worn. Delays of up to six months occurred between prescription and delivery, and shoes too often did not fit when delivered. Further alterations might take more months.

Retrospective Study

A retrospective pilot study looked at what happened to conventional surgical shoes prescribed at St. Mary Abbots and St. Stephen's Hospitals, London, from 1 to 30 January 1965 for 116 patients (all diagnoses). Of these, 29 were not traceable (inadequate records), 87 were written to, and of 70 who replied 30 had no complaints about the shoes and 40 had one or more complaints—that is, shoes uncomfortable (26), returned for adjustment because of misfit (14), too tight (10), too heavy (16),

too clumsy (14), some other reason (8). Thirty-one patients had difficulties in "breaking-in" the shoes.

Most of these shoes were prescribed for patients with rheumatoid arthritis. Those with the more severe rheumatoid foot deformities showed a higher proportion of misfits and of shoes sent back for adjustment.

This study provided evidence of waste in the provision of surgical shoes and that many months might elapse between prescription and delivery. One possible reason for misfits was that the patient's foot might change further during the waiting period. A long period sometimes necessary to break-in the shoes was also notable. There was no evidence that poor quality material or workmanship was used—rather the reverse. However, the all-leather, weather-proof construction made for a firm and relatively unyielding shoe. In rheumatoid arthritis with thin atrophic skin covering bony prominences, the breaking-in period could be an agonizing experience.

Trial of Seamless Shoes

Nineteen patients with difficult shoe-fitting problems were each provided with one pair of conventional surgical shoes and one pair of seamless shoes.¹ A systematic comparison was then made. Of the 19 patients, 18 had rheumatoid arthritis and one had psoriatic arthropathy.

Photograph of seamless shoe

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¹ The brand known as Imprints made by Messrs. Jack C. Rich, of New York, was used; shoes and cast-making kits are available through Braywick Distributors Ltd., Maidenhead, Berkshire.