The Central African
Journal of Medicine

Volume 2 May, 1956 Number 5

Leprosy and Childhood *

BY

J. A. KINNEAR BROWN, B.SC., M.D., D.T.M.
Specialist Leprologist, Uganda.

Disabilities in young people stir the emotions easily. There is an immediate response to children who cannot see or who have been crippled by accident or disease. Expressions such as "stricken," "handicapped for life" and "unable to join in the games of other children" are a measure of the sympathy that is awakened —a feeling, however, that does not necessarily follow them as easily as they get older, when the consequences of their limitations may be greater. It is, therefore, not difficult to regard disease in children as essentially different from that in adults. Certain conditions one expects to see predominantly in children just as others appear only in the more elderly. When considering those diseases that are common to all ages we ought to be certain that our emotional regard for the child is not causing us to put into separate houses what really belong to the same room.

The valuable adoption scheme of the British Empire Leprosy Relief Association, in which the Royal family has taken part for so many years, has focussed attention on those who are under sixteen. Control measures are frequently based on the assumption that, if the children are not infected, the disease will gradually die out. Some authorities have referred to leprosy as a disease of children. It will not be out of place, therefore, to try to see if there is anything significant about the age of onset, and in what respects leprosy in early life is different from the disease in older people.

The age frequency of the disease has sometimes been assessed by analysing the ages of patients in institutions, even when limited only to children. Such returns are notoriously misleading. In many cases the proportion of children in leprosaria is little different from that in the ordinary population. When the writer was in Southern Nigeria one-third of the patients in settlements were under fifteen. At the present time in all the leprosaria in Uganda 29 per cent. are children. This figure could be easily misinterpreted, for the proportion of children in the whole country is 40 per cent. In one of the larger leprosaria at a recent examination 225 (36 per cent.) of 631 resident patients had been admitted during childhood, but in this particular district children form about half the population. When seen against the general background the returns of leprosaria in Uganda do not support the conclusion that leprosy has any predilection for children.

In a large series of surveys in which practically every person was examined, of 882 patients with leprosy 19 per cent. were under 15, 32 per cent. between 15 and 30, 31 per cent. between 30 and 45, and 18 per cent. over 45. The number of those under age 20 was 242, little more than the 233 who were over age 40. Ross Innes had previously examined large numbers of patients in two widely separated districts of Uganda, and an analysis of his figures leads to the same inference that in this part of the world childhood is not the period of greatest incidence. It is not suggested that it is never greater among children. In any community where, from their earliest days, children are in intimate relationship with large numbers of people, the incidence of any and every infectious disease may be higher among them; but a high frequency of leprosy among children does not necessarily indicate a characteristic feature of the disease; it may only reflect the conditions under which the people live.

It is, naturally, very sound policy to do everything possible to prevent children from coming in contact with the disease, but the error should not be made of thinking that susceptibility diminishes with age; that if there is no exposure in childhood there will never be any risk, or that leprosy will automatically die out. Saving the children is a legitimate form of propaganda, but if concentration on the children leads to forgetfulness about the adults there will be little alteration in the prevalence of leprosy. It may be distressing to know that 27 per cent. of the

* This paper is published by kind permission of the Director of Medical Services, Uganda.

leper population in East Africa is under the age of 20, but it is important to remember that the other 73 per cent. got through their childhood and adolescence safely, only to become infected after reaching maturity.

It should also be remembered that not all children who are in contact with leprosy become infected. This is no reason for relaxing precautions. It is simply a well recognised fact, which is most easily explained on the hypothesis that some are susceptible and some are not. The more susceptible are usually those, one of whose parents had leprosy. Where both parents have the disease the susceptibility is presumably greater still. A parent with leprosy also provides a continuous and intimate source of contact, and it is the interplay of susceptibility and contact that determines whether infection will take place at all and at which age the first signs will appear.

The incubation period is an unknown quantity, but is generally accepted as from three to five years. In one group of 53 children, whose ages were known exactly and one or more of whose parents had leprosy, the ages of observed onset were:—

Age in Years	*Number of Children*
1— 2	1
2— 3	8
3— 4	9
4— 5	10
5— 6	8
6— 7	7
7— 8	5
8— 9	3
9—10	2
	53

Some of these children had been brought up during infancy segregated in a special creche, which may explain why infection was, apparently, so long delayed. Nine of the 53, however, showed the first signs before the age of three. It is possible that, in the very susceptible children of leper parents the incubation period is less, and that in those who are not totally without resistance the infection is contained until some event reduces that resistance to the point when the appearance of clinical signs indicates that dissemination has begun.

There are two distinct types of leprosy: (a) lepromatous; and (b) tuberculoid (Fig. 1). They are called types because they possess well-recognised clinical and biological features and tend to be stable. There are two groups, one "indeterminate" and the other "borderline" or "dimorphous." They are less stable and often represent a state of transition from one type to the other.

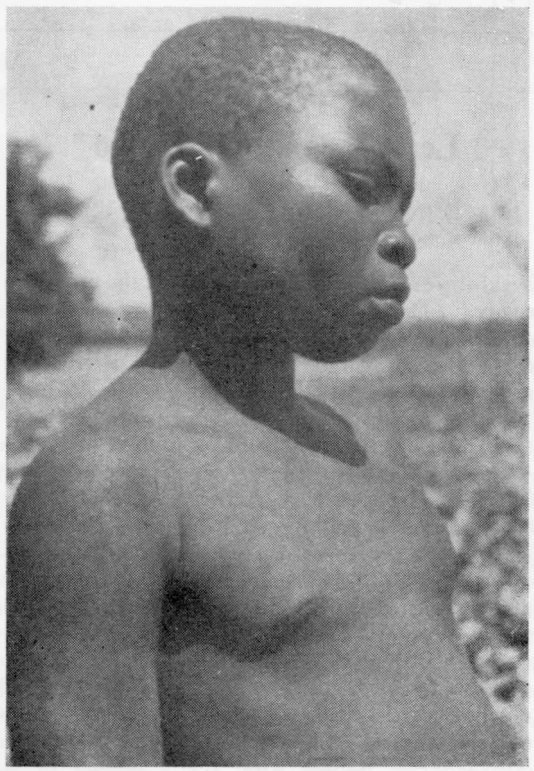

Fig. 1—Macular tuberculoid leprosy of the face treated with Native caustic.

Lepromatous leprosy can be sub-divided into macular, diffuse, infiltrated, nodular and neuritic forms; tuberculoid leprosy into macular, minor tuberculoid, major tuberculoid and neuritic. The "indeterminate" group includes macular and neuritic forms and the "borderline," infiltrated and non-infiltrated. It is not necessary for the general physician to be familiar with all the details of these categories. They are listed for the sake of completeness, but some of them are of particular importance in children.

Lepromatous lesions are strongly positive bacteriologically and the lepromin reaction is negative. The distribution is symmetrical and, whilst they are found on all parts of the body, the classical manner in which they affect the face and ears produces the typical leonine facies. Nerve involvement follows at a later stage and is also symmetrical. Tuberculoid lesions are usually negative bacteriologically and the lepromin test is positive. The skin changes consist

of erythema with hypopigmentation which is reversible, and the loss of pigment is never complete. Many of the lesions are raised to a greater or lesser degree, particularly at their margins, and the edge is quite distinct from the surrounding skin. Nerve involvement is frequent and is usually asymmetrical. In the minor tuberculoid form the elevation is slight and limited to the margin; the major tuberculoid variety consists of plaques and annular lesions, and the elevation and thickening are greater.

"Borderline" cases are usually recognisable clinically. They are generally positive bacteriologically and the lepromin reaction is weakly positive or negative. They may begin in this group or they may arise from tuberculoid lesions (Figs. 2 and 3). They may revert to the tuberculoid type, but more frequently they proceed to the lepromatous. The lesions include plaques, bands and nodules, generally asymmetrical and with a soft succulent appearance. They slope away from the centre to the periphery without presenting the well-defined margins seen in the tuberculoid type. They may be mistaken for lepromatous leprosy.

There is no difference between the incidence in childhood and adult life of either lepromatous or tuberculoid leprosy, but, whereas the majority of adult patients begin with lesions characteristic of one or other type, there is a tendency in children for both types to develop from a simple macule. Typical nodular lepromatous leprosy is often preceded by multiple, symmetrical, hypopigmented, erythematous macules, very slightly raised in the centre and distributed over the whole body. These lesions are not always easy to distinguish because the alteration in pigment is not marked; they appear to merge into the ordinary skin and there is no alteration in lustre due to anhydrosis. They are a typical form of lepromatous leprosy, and there is no proof that they ever become tuberculoid. Frequently in children both tuberculoid and lepromatous leprosy arise from another form of macule called tuberculoid by some and indeterminate by others. Such macules are flat, hypopigmented with definite edges, but dry and lacking the lustre of the ordinary skin due to some interference with sweating. They occur

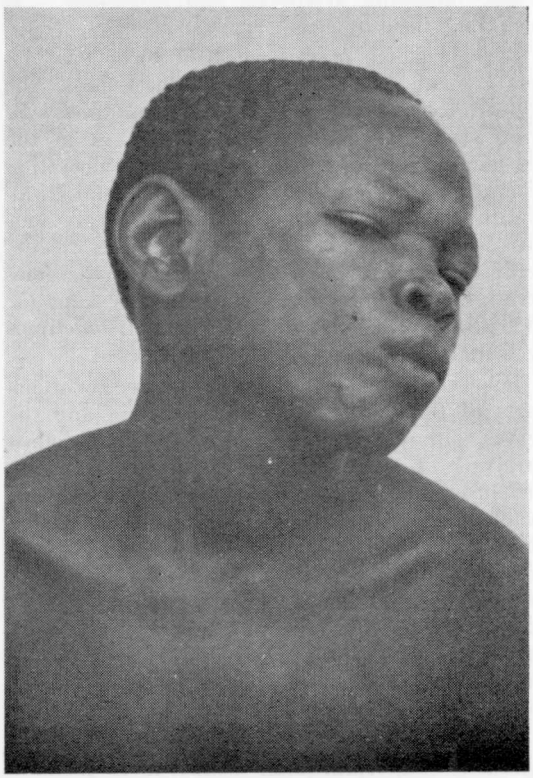

Fig. 2—Borderline leprosy becoming tuberculoid.

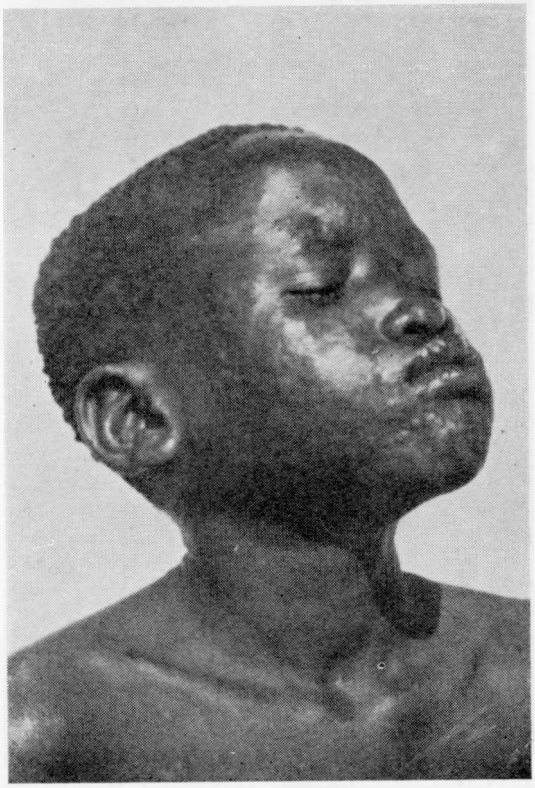

Fig. 3—Borderline leprosy becoming lepromatous.

most commonly on the face, arms, legs and buttocks.

If when a biopsy is taken the only histological changes are those of non-specific inflammation, indeterminate is a legitimate classification. If the pathology is more organised it may be more satisfactory to designate it accordingly. The essentials to remember are that, like most other diseases in which there are fluctuations one way or the other, the lesions do not remain in the same pigeon-hole all the time, and that when a dry, lustreless area of hypopigmentation is seen in a child it may become tuberculoid, borderline or lepromatous. The possibilities are more important than the name.

The onset of the disease is frequently accompanied by a period of vague ill-health, during which the appetite is reduced and the child fails to put on weight. Areas of numbness may develop before there is any change in pigmentation, and neuralgia and paraesthesia in the arms and legs in the course of the peripheral nerves are not uncommon. The appearance of nodules, areas of infiltration and hypopigmented patches

Fig. 4—Extensive deformity in a girl of 18 years suggesting more rapid progress in children.

is gradual and progressive over a matter of years, with frequent periods of quiescence. Ultimately peripheral neuritis with fibrosis in the nerve trunks may lead to claw hand and drop foot, and interference with sensation to trophic ulceration. Burns and other forms of trauma produce whitlows of varying severity, and bone absorption, even in the absence of sepsis, can result in complete disappearance of the digits until the finger nails appear to be poised grotesquely on what are virtually fingerless hands (Fig. 4).

In major tuberculoid leprosy the patches may become swollen, erythematous and oedematous, but not necessarily all at the same time. This reactional phase may last for weeks or months and is often unaccompanied by any change in temperature, although lesions which were bacteriologically negative may become temporarily positive. Frequently the patient is much better after the phase is over. In lepromatous leprosy the picture is rather different and, although some patients may appear eventually to have benefited, in the majority the outlook in lepra reaction is less encouraging. It may be precipitated by intercurrent infection or the giving of certain drugs, including arsenicals and sulphones. It is characterised by:—

Pyrexia which may reach 104° F. or 105° F., enlargement and swelling of old lesions, the appearance of new lesions, *erythema nodosum leprosus*, pain, tenderness and swelling in the peripheral nerves, inflammation and ulceration of mucosa of nose, throat and larynx, and iridocyclitis, mainly where there has been previous eye involvement. The fever is often hectic, being subnormal in the morning. If the temperatures and pulse rates are regularly kept for all lepromatous cases, the pyrexia can sometimes be anticipated and the reaction aborted.

Tuberculoid leprosy does not directly attack parts of the body other than the skin and the nerves, although damage to the eye may follow paralysis of the trigeminal and facial nerves. In lepromatous leprosy ulceration of the mucous membrane of the nose, throat and larynx obstructs the breathing and produces hoarseness or loss of voice, whilst the eyes may be seriously involved as a result of blood-borne infection or extension from the nose through the lachrymal duct. The testicle is the only other organ to be attacked, and fibrosis of it leads to gynaecomastia.

The differential diagnosis in children is extremely important and, whilst it is possible to miss the earliest lesions, the commoner and

often graver error is to label as leprosy something which is innocuous. Where leprosy is endemic and there is a general fear of contracting it, it is not difficult to see a wood when there is only a solitary shrub, and the incidence of the disease can be proportional to the zeal of those who are looking for it.

I have been to schools in which there has been an alleged incidence of leprosy of between 30 and 40 per cent. This is mentioned only as a measure of the confusion that may arise. Such an incidence would be uncommon in any typical section of the community, and in a unit as select as a body of school children it would be phenomenal. In an area with a general incidence of 2 per cent., in which one-fifth of those affected were children, a school incidence greater than 0.4 per cent. would not occur normally. One would not expect the incidence in a school to be anything like as great as the average, and a proportion as high as 30 per cent. would imply that one was dealing with a school restricted to the children of leper parents. There is no doubt about the difficulty in children, but whenever the number of "cases" in a school approaches or exceeds the average, one should begin to look for alternative diagnoses.

A frequent source of error is the small, isolated lesion on the face. Some of the children brought for examination have had indefinite patches less than one centimetre in diameter. Sometimes they have been paler than the ordinary skin; at other times they have shown no alteration in colour, but have been raised or thickened or have had an irregularly elevated margin. The fact that most people of all colours have some small area of skin in which there is an alteration in texture or pigmentation should suggest extreme caution in diagnosing or treating as leprosy any single lesion which is not absolutely typical, and no child should ever be admitted to a leprosarium unless the diagnosis has been established beyond all doubt.

The commoner conditions which can mislead are urticaria, herpes zoster, birthmarks lighter or darker than the ordinary skin, scars or disfigurement following smallpox, burns or other forms of trauma, keloids, lichen pilaris, flat warts, hypopigmentation associated with or resulting temporarily from impetigo or other skin infection on the forehead and cheeks, scaly hypopigmentation near the mouth often the result of nutritional deficiency, seborrhoea on the scalp margin and eczema behind the ears. The differential diagnosis has frequently to be made from tinea circinata, tinea versicolour and fungus infections which may arise in the margin of the scalp, the flexures, on the dorsum of the hands and feet or elsewhere on the body (Figs. 5, 6 and 7). Comedones and acne vulgaris can sometimes suggest lepromatous leprosy, and lupus vulgaris may have to be excluded. Hypopigmentation due to yaws or as an intermediate stage in the production of leucoderma is occasionally confusing.

A history limited to a few weeks or going back to earliest infancy without any neural change should create a doubt. The presence of other cases within the family, a long history, alterations in sensation and thickening of nerve trunks suggest leprosy. Even here, however, great care is needed. Tinea often occurs in more than one member of a family, and people who do not wear shoes, who walk regularly on the roughest roads and who easily handle objects which to others are far too hot are not always able to appreciate the more subtle changes in sensation or temperature. It should be remembered that a negative bacteriological result does not exclude leprosy. It only means, as in

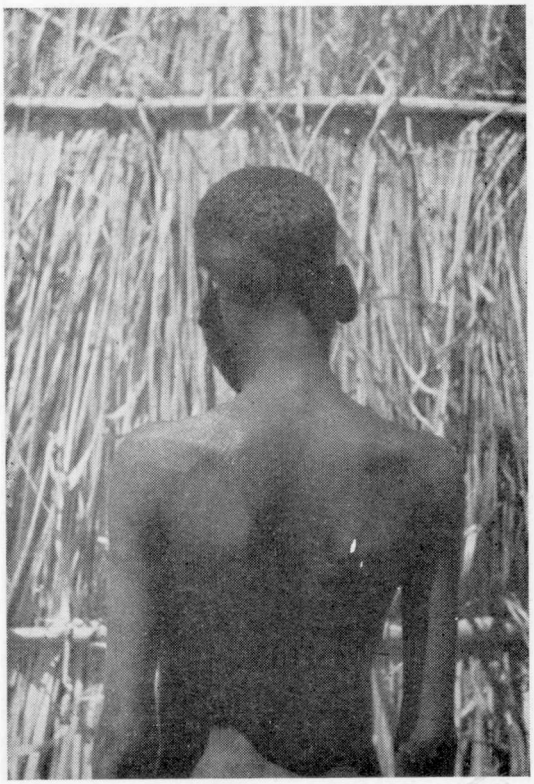

Fig. 5—Tinea on the shoulder resembling leprosy. Two members of the family had similar lesions.

tuberculosis, that bacteria have not been discovered. A positive finding in the skin is conclusive. Smears from the nose must be Gram stained to exclude other pathogenic organisms. The lepromin reaction is of value only in prognosis or in helping to determine the type. It is of no value whatever in the diagnosis.

Although, with the exception of the points which have been discussed, the broad picture of leprosy in children is very similar to that in adults, the progress of the disease may be more rapid because this is the period of growth, and children generally are less able to withstand the assault of most major illnesses. When the parents have had leprosy my impression has been that the downward course of events has been quicker. There is a greater tendency for those with lepromatous leprosy to go into repeated reactions which leave the patient more derelict than before. In tuberculoid cases, bone absorption in the hands and feet is of greater consequence, lack of adult caution tends to more frequent injuries and, as these are so often painless, a childish disregard of what may follow results in sepsis and ulceration. Contractures and deformities also occur at an earlier stage. in general, the prognosis in children in all forms of the disease is worse, and it is for this reason that attempts should be made to prevent them from contracting the disease or to get them at the earliest moment into an institution.

As in so many diseases, prevention is better than cure. What cannot be treated could often have been avoided. There has been much speculation about the reason for the disappearance of leprosy from some of the European countries. The usual explanation is that it has been due to better living conditions, but there are areas in some of the less advanced countries where the incidence is not greatly different from that where the standard of living is much higher.

The significantly higher incidences in Uganda are in the smaller tribal groups where differences of language and custom have restricted the field of marriage; the lower incidences are among those people who have responded to the opening up of their country by marrying in a larger circle. The inference is that leprosy establishes itself where the in-breeding of

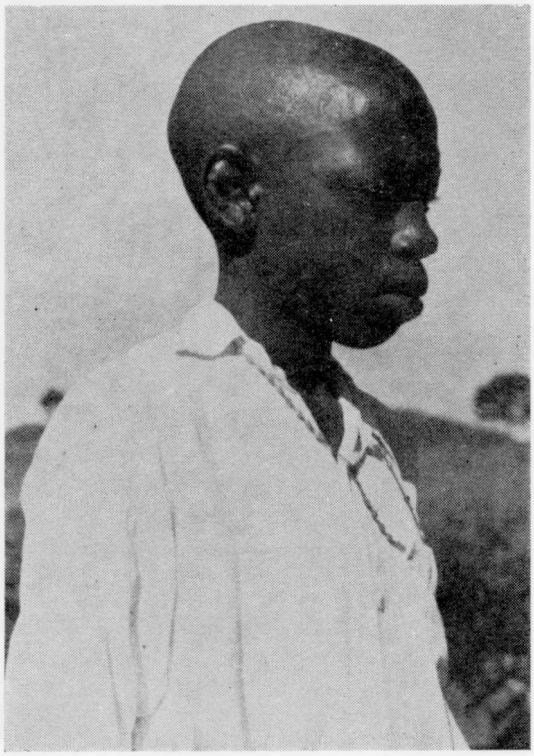

Fig. 6—Tinea on the scalp margin in a patient with lepromatous leprosy of the ears.

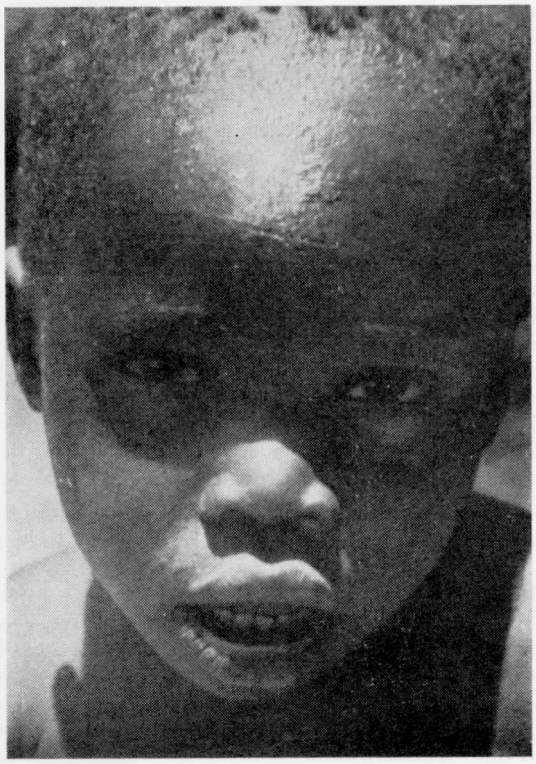

Fig. 7—Vague lesion on the forehead probably due to trauma—the type frequently produced as leprosy.

susceptible children is a consequence of the local marriage customs, and it disappears gradually as people travel and marry in a less restricted sphere. This could be the explanation of the disappearance of leprosy from England. The breaking down of barriers of language. customs and caste may, by broadening the basis of society, lead to the disappearance of the disease from those parts of the world where it is still highly endemic. Such a social adjustment will evolve naturally as education becomes more universal and broader in scope. As an immediate measure it is worth while trying to persuade patients with leprosy not to have families until they have been adequately treated. This alone would reduce the number of susceptible children, and it would reduce the risk of infection within the household. This is a counsel of perfection and may be somewhat of a cry in the wilderness, but it is no reason to withhold the advice because in many cases it will not be taken.

As a more active measure, B.C.G. vaccination has been suggested as a means of prevention because it converts a lepromin negative reaction in children to positive. It is not certain that the lepromin reaction is specific, especially in those in whom there is no evidence of leprosy, and there are other aspects which are controversial. Investigations are being pursued in most parts of the world, and it is hoped they will provide a valuable method of reducing the prevalence of the disease. There will, however, be certain practical limitations. It might be possible to vaccinate all the children of leper parentage, but in an undeveloped country, with a scattered population, there is always the difficulty of finding them when they are needed, of obtaining the parents' co-operation, and of making frequent follow-up examinations. Moreover, the children of leper parents form only one section of those who are susceptible. If, as Hale and others believe, it is the Mantoux negative section of the community that is liable to infection, the widespread use of B.C.G. to produce Mantoux rather than lepromin conversion should simultaneously protect against leprosy and tuberculosis.

The use of diaminodiphenylsulphone has completely revolutionised the treatment of leprosy, although it should not be concluded that all that now needs to be done is to distribute tablets to large numbers of out-patients whenever they present themselves. The first essential is accurate diagnosis—no specific treatment should be given until that has been established. The second is caution to avoid creating sulphone sensitivity, and the third continuity to avoid any possibility of drug resistance. Children normally tolerate sulphones well. The commonly accepted dosage for adults is 800 milligrammes weekly in divided doses issued either twice weekly or every day; that for children should be calculated in the usual way. The routine in Uganda is to try to reach the maximum over a period of three months. The following procedure is adopted for adults:—

Weeks 1— 4	100 mgs.	twice weekly
„ 5— 8	200 „	„ „
„ 9—12	300 „	„ „
Thereafter	400 „	„ „

Injections of sulphetrone are recommended by some workers, but the drug is more rapidly eliminated than the parent sulphone. Twice weekly injections are not likely to appeal to children, and their administration requires a technique more complicated than the issuing of tablets. Where, for any reason, sulphetrone is preferred, a 50 per cent. solution should be prepared by adding sufficient distilled water to 500 grammes of the solid to make one litre. The solution is sterilised by boiling for 30 minutes (or in an autoclave) and making the volume up to one litre when cold by adding sterile water; 1.4 grammes sodium bicarbonate per litre makes the injection less painful.

The solution is normally injected twice weekly. The following plan is for adults:—

First two weeks	0.5 c.c. twice weekly
Second two weeks	1.0 c.c. „ „
Third two weeks	1.5 c.c. „ „
Fourth two weeks	2.0 c.c. „ „
Fifth two weeks	2.5 c.c. „ „
Thereafter	3.0 c.c.

Tablets of sulphetrone (0.5 grm.) are chiefly reserved for cases of sulphone sensitivity. The drug is poorly absorbed and its routine use has long been given up. Sensitivity is better avoided by exhibiting the sulphone drug slowly, and should it ever develop it is better to change altogether to daily doses of thiosemicarbazone.

Questions frequently arise about how long treatment should be continued, and whether sulphones have any prophylactic value. No patient should be considered for discharge until successive smears from different parts of the body are negative, and in any case not until there has been continual treatment for a period of two years. One is doubtful about the preventive or suppressive use of D.D.S. I have seen the disease appear in children despite their

having had small doses of D.D.S. for one or two years whilst their parents had been having active treatment. The dosage may have been too small or the disease may have become resistant. It is difficult to prove without a culture that drug resistance does develop, but from time to time one sees patients who on normal doses improve considerably at first, and then suddenly become stationary even when the drug is increased to the margin of toxicity. The best form of prophylaxis is always the avoidance of risk.

It goes without saying that no form of therapy should be initiated in the presence of severe anaemia, malaria, active yaws and helminth infestation or other concurrent infection. These should be adequately treated at the very beginning, and whenever the patient's progress is retarded or complications threaten or signs of reaction appear, a full investigation should be made. I have always placed the greatest reliance on a morning and evening pulse and temperature chart; it often gives warnings that despite appearances all is not well, and so enables avoiding action to be taken in time. When complications or lepra reaction develop, treatment should be conservative and symptomatic, specific therapy being withheld in the presence of reaction, iritis, iridocyclitis, acute neuritis or signs of sensitivity. The treatment of late sequelae such as contractures is preventive and surgical. Early treatment, massage and exercises may help to reduce their incidence and severity, but when fully developed, plastic surgery and tendon transplantation form the only remedy.

REFERENCES

Brown, J. A. K. (1955). *Trans. R. Soc. trop. Med. Hyg.*, 49, 3.

Hale, J. H., Molesworth, B. D., Grove-White, R. J., Sambamurthi, C. M. & Russell, D. A. (1955). *Int. J. Leprosy*, 23, 2.

Innes, J. R. (1948). *E. Afr. med. J.*, 25, 379.

——— (1950). *Ibid.*, 26, 32.

——— (1950). *Int. J. Leprosy*, 18, 507.