

Peripheral neuropathy in leprosy and its consequences

W. H. VAN BRAKEL

TLM India, New Delhi, India

Summary Leprosy causes a 'mononeuritis multiplex' of immunological origin that results in autonomic, sensory and motor neuropathy. When detected and treated early, *primary impairments* may be reversible. However, 11–51% of patients do not recover. In addition, 33–56% of newly registered patients already have clinically detectable impairments, often no longer amenable to drug treatment. Among new patients, 6–27% present with *secondary impairments*, such as wounds, contractures and shortening of digits. All patients with impairments should be taught methods to prevent further impairment and subsequent disability (POID). As the result of impairments, many people experience *limitation of activities* of daily living, which can be partially overcome with the help of assistive devices, training, and surgery. As a result of these limitations, because of visible impairments, or simply because of the diagnosis 'leprosy', many people are *restricted in their participation* in society. Many overcome activity limitations and participation restrictions without assistance, despite residual impairments. However, some require intervention, such as physical or occupational therapy, reconstructive surgery or temporary socioeconomic assistance. Information on these issues is not collected routinely, and the few tools that exist to measure the severity or extent of impairment have not been widely used, nor have they been used to generate cohort-based statistics. There are no agreed indicators for monitoring POID activities or rehabilitation interventions. Work in the general field of rehabilitation has resulted in the ICIDH-2, which provides a conceptual framework for rehabilitation and the entire area of 'consequences of health conditions'. Although experience to date is very limited, the conceptual framework appears appropriate to leprosy. Data on the prevalence and incidence of primary and secondary impairments have been reported from several countries, the link between impairments and activity limitations has been investigated, and a few studies of the magnitude of the need for rehabilitation have been reported. Research priorities include studies of methods to improve detection of autonomic, sensory and motor neuropathy; trials of alternative drugs or regimens for treating neuropathy; studies of the use of various POID-monitoring systems that may be derived from these; studies of the design and use of instruments to assess limitations of activities and restrictions on participation; assessments of needs for rehabilitation and the development of methods to do these; studies of the efficacy of various types of rehabilitation interventions for particular conditions; and studies of the cost-effectiveness of such interventions.

Introduction

Leprosy is characterized by the peripheral neuropathy it causes. A 'mononeuritis multiplex'

of immunological origin results in autonomic, sensory and motor neuropathy. These neural impairments lead to well-known *primary impairments*: dryness of the skin,¹ impaired vasomotor reflexes,^{2,3} impairment of various sensory modalities, such as pain, touch and temperature sensation,^{4-7,8,9} and motor impairment causing muscle weakness or paralysis.⁸⁻¹⁰ If not treated in time, these primary impairments often give rise to *secondary impairments*, such skin cracks, wounds, clawing of digits, contractures and shortening of digits and even hands or feet, and blindness.¹⁰⁻¹³ Impairments may lead to *limitations of activity* (formerly termed 'disabilities').¹³⁻¹⁶ Activity limitations, impairments, or even a current or past history of leprosy may lead to *restrictions in (social) participation* (formerly termed handicap).^{14,17,18}

Issues

When detected and treated in time with corticosteroids, primary impairments may be reversible. However, a substantial proportion of patients, 11-51%, does not recover, or the situation worsens.¹⁹⁻²² In addition, depending upon the country and the programme, 16-56% of newly registered patients have clinically detectable impairments, often no longer amenable to drug-treatment, at the time they are first seen. Table 1 summarizes recent data on the prevalence of primary impairments (WHO grade 1) and secondary (or visible) impairments (WHO grade 2), based upon studies of more than 100 subjects. The prevalence of nerve-function impairment (NFI) per 100 person-years at risk (PYAR) has been reported as 1.3-3.5 for PB and 7.5-24 for MB leprosy during and after MDT.^{9,23,24} Early detection of leprosy along with early detection and treatment of neuropathy are the means to prevent permanent primary impairments.

Among newly-detected patients, 6-27% present with *secondary impairments*, such as wounds, contractures and shortening of digits, usually preventable consequences of autonomic, sensory or motor neuropathy. Among MB patients, who form the majority of new cases in some countries, the frequency of secondary impairments is even greater, 33-56%, as shown in Table 1. All of the patients with primary or secondary impairments require careful and repeated teaching with respect to methods of preventing further impairment and subsequent disability (POID).

Although little is known about the number of people requiring such interventions, estimates may be based on two recent studies. Among 417 PB patients in Thailand, using changes in voluntary muscle testing (VMT), sensory testing (ST), wound count and bone loss as measures of severity of the impairment, 2.2% improved between diagnosis and release from treatment (RFT), and 2.9% developed new or additional impairments during this time. Among the MB patients, these figures were 19 and 16%, respectively. At RFT, 4.6% of PB patients had WHO grade 1 impairments and 6.5% grade 2.²⁵ Among the MB patients, these figures were 24 and 10%. Among a cohort of 706 MB patients in western Nepal, 28% had improved and 16% had become worse, using WHO 0-2 grading as a measure of the severity of the impairment.²⁶ After follow-up of the same cohort for 2 years, 15% had grade 1 and 29% grade 2 impairments.¹⁵ These figures indicate that one-third or more of those who have had MB disease, and as many as 10% of those who have had PB leprosy, may be in need of life-long POID activities (mostly self-care).

As a result of their impairments, many people experience *limitations of activities* (of daily living), formerly termed 'disability', which can be partially overcome with the help of

Table 1. Prevalence of impairment in leprosy patients newly diagnosed in the field (data from studies of more than 100 patients in MDT programmes)

Country & authors	Patients	WHO grade*	Prevalence					
			PB Number	%	MB Number	%	All patients	
							Number	%
West Nepal; van Brakel (unpublished data)	704 PB	0	6	0.9	41	2.1	47	1.7
	1999 MB	1	30	4.3	369	19	399	15
		2	22	3.2	566	29	588	22
		1 + 2	52	7.5	935	48	987	37
West Nepal; Reed <i>et al.</i> ²⁶	1082 MB	0		0				
		1		280	26			
		2		324	30			
		1 + 2		604	56			
East Nepal; Schipper <i>et al.</i> ¹²	106 PB	0					18	6.9
	154 MB	1					42	16
		2					52	20
		1 + 2					94	36
Thailand; Schreuder ²⁵	420 PB	0	0		0		0	
	220 MB	1	19	4.0	52	24	71	11
		2	26	6.2	21	9.5	47	7.3
		1 + 2	45	11	73	34	118	18
Malawi; Pönnighaus & Boerrigter ³⁹	305 MB	0**			0			
		1			37	12		
		2			67	22		
		1 + 2			104	34		
Ethiopia; deRijk <i>et al.</i> ³⁵	128 PB	0	0		0		0	
	158 MB	1	32	25	51	32	83	29
		2	38	30	38	24	76	27
		1 + 2	70	55	89	56	159	56
Bangladesh; Croft <i>et al.</i> ⁴⁰	2220 PB	0	0		0		0	
	444 MB	1	134	6.0	122	28	256	9.6
		2	78	3.5	81	18	159	6.0
		1 + 2	212	9.5	209	47	415	16

* Graded according to the 1988 WHO 0–2 scale.

** Re-graded from 1960 WHO 0–5 scale.

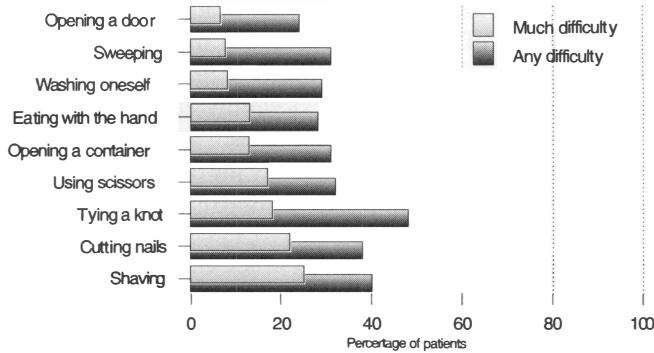


Figure 1. A survey of difficulty experienced with activities of daily living among 269 people affected by leprosy in west Nepal: selected results of indoor activities.

assistive devices, training, and surgery. A survey among people affected by leprosy attending field-clinics revealed that, depending on the type of activity, as many as 25% had ‘much difficulty’ with indoor activities, and as many as 34% with outdoor activities.²⁷ The results for some selected, common activities are shown in Figures 1 and 2. Based on this work, an activity assessment scale, the Green Pastures Activity Scale (GPAS),²⁸ was developed.

As a result of the activity limitations, because of visible impairments, or simply because of the diagnosis ‘leprosy’, many people are *restricted in their (social) participation*, formerly termed ‘handicapped’.^{14,18,29,30} A large, but unknown proportion of people succeed in overcoming these activity limitations and participation restrictions by themselves, and do not need assistance, in spite of residual impairments. However, a certain proportion need rehabilitation interventions, such as physical or occupational therapy, reconstructive surgery or temporary socioeconomic assistance. A study among 53,000 people affected by leprosy, conducted by Gopal and his coworkers,³¹ found that 34% were in need of some kind of social or economic rehabilitation assistance. A study in Ethiopia found that, of people affected by leprosy, only 20% identified themselves as needing rehabilitation (cited in³²). Kopparty found that 57% of 150 people with leprosy-related deformities faced social or economic problems.³³

There are no routine information systems in place that collect information on these issues.

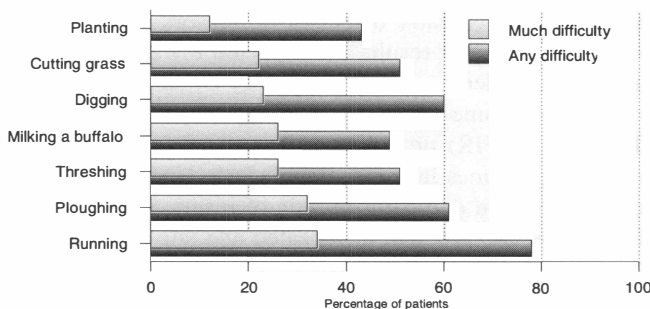


Figure 2. A survey of difficulty experienced with activities of daily living among 269 people affected by leprosy in west Nepal: selected results of outdoor activities.

WHO 'disability' grading according to the 0–2 scale, which was introduced in 1988, is used very widely to grade severity of impairment in newly diagnosed patients. However, this information is used almost exclusively to calculate a proxy indicator for delay in case-detection—the 'proportion with WHO grade 2 disability among all newly detected cases'. The WHO grading system is based on six sub-scales, each grading one eye or one limb on the 0–2 scale; the 'proportion grade 2' indicator uses only the maximum of these six grades as its value. Other scores have been proposed, which are based on the same system, but make better use of the available information; these are the HF (hands, feet) and the EHF (eyes, hands, feet) scores. However, because the grading is very crude and not sensitive to change, none of these is suitable for monitoring impairment status in individuals.^{15,36} The Impairment Summary Form (ISF, formerly termed the 'District Disability Summary Form') represents a more elaborate system, much more suited to monitoring POID activities. Although it has been introduced in a number of programmes, this system has not been used widely and consistently. As yet, no agreed indicators for monitoring POID activities have been derived from any of these measurement tools.

The situation with regard to monitoring and evaluation of rehabilitation-interventions and programmes is even worse. By and large, reports only make mention of process-indicators, e.g. the number of people who have received surgery or vocational training. Anandaraj introduced a scale to measure 'dehabilitation' in people affected by leprosy.³⁸ However, this scale does not appear to have been used in programme management or further research. A scale for measuring activity limitations was developed recently in Pokhara, Nepal.²⁸

Current and planned work

The design of a 'participation scale' for monitoring and evaluating socioeconomic rehabilitation of people affected by leprosy is underway in the RELEASE Project in Pokhara, Nepal (Dr Engelbrektsson, personal communication). A proposal for pilot studies of the usefulness among people affected by leprosy of the World Health Organization (WHO) Disability Assessment Schedule (WHODAS II), a generic instrument for measuring impairment, activity limitation and participation restriction, based on the ICIDH-2, has been submitted to the WHO. An international workshop is planned for November 2000, to address the entire area of measuring disablement in leprosy.

An international research programme, named the TRIPOD trials, is investigating the benefits of prophylactic steroids, early detection of sensory impairment and treatment of long-standing neuropathy. The first results are expected towards the end of this year. Two years ago, the International Federation of Anti-Leprosy Organisations (ILEP) commissioned another large research programme on reactions and neuropathy. The ILEP Nerve Function Impairment and Reaction (INFIR) programme consists of four separate research projects, involving centres and programmes in India, Bangladesh, Nepal, Ethiopia and Malawi, that investigate prediction, detection and pathogenesis of reactions and neuropathy; alternative drug treatment of reactions; the problem of repeated and late reactions; and the problem of delayed presentation for treatment.

The type I reaction study is a multicentre, double-blind, randomized, controlled trial of different steroid regimens for the treatment of type 1 reactions in leprosy, conducted at several centres in India. Results may be expected later this year or early 2001.

Future research priorities

1. POID

1.1. Prevention of primary impairment

1.1.1. Prevention of reactions and nerve function impairment (NFI)

Double-blind controlled trial of clofazimine in high dosage in the prevention of reactions

1.1.2. Early detection of NFI

1.1.2.1. Detection of autonomic nerve damage: laser and sound Doppler; other techniques?

1.1.2.2. Improvement of monofilament sensory testing: prospective study of the optimal number and combination of test sites on hands and feet; reliability of testing by multipurpose health workers

1.1.2.3. Design and testing of an optimal series of monofilaments (including number, weights and diagnostic thresholds) in a format suitable for field use

1.1.2.4. Study of the feasibility of affected people testing themselves at home with a monofilament

1.2. Treatment of primary impairment

1.2.1. Study of short-term high-dose steroids in the treatment of severe reactions with NFI by means of a controlled trial against the current standard regimen

1.2.2. A controlled trial of nerve decompression plus steroids versus steroids alone in patients with recent NFI, with independent outcome assessment

1.2.3. Treatment of paraesthesia

1.2.4. A trial of the standard steroid regimen *versus* variable steroid treatment based on clinical decision-making

1.3. Monitoring of POID

1.3.1. Studies of the reproducibility and responsiveness to change over time of the WHO maximum grade, the eyes hands feet (EHF) score, and the Impairment Summary Form (ISF) for grading severity of impairment

1.3.2. Studies of cohort reporting using indicators derived from these scoring methods

1.3.3. Studies of the cost-effectiveness of different components and approaches in POID

1.3.4. Mapping of body image in patients with NFI

2. Rehabilitation

2.1. Activity limitation and disability

2.1.1. Survey of difficulty experienced in activities of daily life among persons with various leprosy related impairments, using the GPAS and WHODAS II

2.1.2. Effects of different surgical procedures (particularly hand surgery) on activity limitations

2.1.3. Effects of occupational therapy interventions in different types of activity limitation

2.2. Participation restriction

2.2.1. Development or adaptation of a scale to measure participation among people affected by leprosy

- 2.2.2. Survey of participation restriction among people affected by leprosy
- 2.2.3. Study of the correlation among impairments, activity limitation (disability), and the extent of participation restriction
- 2.2.4. Evaluation of socioeconomic rehabilitation interventions using a participation scale
- 2.3. Community-based rehabilitation (CBR)
 - 2.3.1. Surveys to assess to what extent people affected by leprosy are included in existing CBR programmes
 - 2.3.2. Development of methods to estimate the number of people in need of CBR in a given area
 - 2.3.3. Studies of different approaches in CBR and of the inclusion of POID activities within such programmes

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