

Lies, Damned Lies, Metrics & Semantics: Exploring definitions of the end of Leprosy (Hansen's Disease) & their implications

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Many a statistic is false on its face. It gets by only because the magic of numbers brings about a suspension of common sense.¹

Leprosy (also known as Hansen's Disease), is an infectious disease that is both potentially disabling and highly stigmatised.² Leprosy can cause blindness, disfigurement and deformity. This disease is named after G. H. Armauer Hansen, a physician who identified the causative agent, *Mycobacterium leprae*, in Norway in 1873.³ *Mycobacterium leprae* is a strongly acid-fast rod-shaped organism with parallel sides and rounded ends (see Figure One), rather similar to the tubercle bacillus.⁴

Figure One: *Mycobacterium Leprae*



It is interesting to note that *Mycobacterium leprae* was the first bacterium to be identified as causing disease in humans. Given this significant history and impact of this disease on human civilisation,⁵⁻⁶ and the length of time that has elapsed since its cause was identified, it is alarming that this disease still remains a scourge.

Although for many people the mention of leprosy may evoke biblical associations,⁷ or images of remote segregated and excluded communities of a bygone era,⁸⁻⁹ leprosy is an ongoing and significant problem. As can be seen from Table One, although there has been a gradual decline in the number of new cases reported in official statistics over the last decade globally, approximately 200,000 are still diagnosed annually.¹⁰

Table One: New Leprosy Cases (thousands) 2006 – 2015 ¹⁰

Year	2006	2007	2008	2009	2010	2011	2012	2013	2014	2015

No. of new cases	244.6	237.4	229.7	224.2	211.1	208.0	217.5	202.9	200.8	200.0
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The global incidence of leprosy has declined significantly since Multi-Drug Therapy (MDT) was introduced as the standard treatment in the early 1980s. The specific form of MDT given depends on the type of leprosy diagnosed, but routinely includes two or three of the following drugs: Rifampicin, Clofazimine & Dapsone.¹¹ The decline in the global impact of leprosy has been significantly aided by the involvement of major international charitable efforts from bodies such as The Nippon Foundation (TNF), formerly known as the Japan Shipbuilding Industry Foundation, the Sasakawa Memorial Health Foundation (SMHF),¹² and the pharmaceutical company Novartis and its philanthropic arm.¹³

However, the very success of MDT has served to undermine the future success of leprosy elimination programs, and this may help explain a recent upsurge in leprosy rates. As Thompson notes “*To prematurely consign leprosy to the history books guarantees unnecessary future morbidity*”.¹⁴ In 1991 the 44th World Health Assembly called for the elimination of leprosy as a public health problem globally by 2000.¹⁵ Although this aim is of course laudable, significant issues have emerged over the definition of elimination. The WHO unilaterally defined elimination as an incidence of less than one case per 10,000 people.¹⁶ Based on this criterion and available statistics, the WHO was therefore able to declare leprosy eliminated at a global level in the year 2000.¹⁰

However, there are a significant number of issues associated with the metric used by the WHO to define the elimination of leprosy.²

The first issue is that of a lack of a scientific basis to the rather arbitrary decision to settle on the figure of less than one new case per 10,000 people. The editor of the International Journal of Leprosy addressed this issue stating that:

Others are highly sceptical of the scientific basis for current elimination policies, and think that implementation of these policies is being unnecessarily and prematurely rushed to meet arbitrary bureaucratic goals, to the detriment of patient care.¹⁷

The second issue is the focus by the WHO on such a short-term target. The decision in 1991 to target elimination by the year 2000 is rather bizarre given the nature of leprosy infection and development. Although MDT treatment is 98% successful, a notable aspect of leprosy is that it can take 5 to 20 years to appear.¹⁰ Though the incubation period is typically 3 to 8 years. Therefore, any such short-term aim was unfeasible from its inception.

The third issue is the WHO combination of the term 'elimination' with a numerical metric, i.e. less than 1 new case per 10,000 population. Dowdle has outlined, with examples, the routine definition of terms such as Control, Elimination, Eradication and Extinction from an epidemiological perspective.¹⁸ Perhaps one of the best examples of WHO efforts around infectious disease can be seen in the Smallpox eradication program.¹⁹⁻²¹ However, using Dowdle's widely accepted criteria (see Box 1),¹⁸ this process involved mass population level vaccination programs that slowly achieved the 'elimination' of smallpox in defined geographical areas (usually countries), until the disease could be described as eradicated globally, only now existing in a small number of Government/Military research labs.¹⁹⁻²¹ The important issue here is that countries were defined as having achieved the elimination of smallpox when their incidence declined to zero, not some ratio of 1 per 10,000, or per 100,000 or per 1,000,000. It should be noted that smallpox is very different from leprosy in that it is significantly

more infectious, and has a much shorter incubation period. Smallpox can also result in the death of up to a third of those who contract it, unlike leprosy which results in disability. However, it is the use of terminology that is important, rather than the nature of the disease.

Box 1: Dowdle's Principles of Disease Control, Elimination, Eradication & Extinction ¹⁸

Control: The reduction of disease incidence, prevalence, morbidity or mortality to a locally acceptable level as a result of deliberate efforts; continued intervention measures are required to maintain the reduction. Example: diarrhoeal diseases.

Elimination of disease: Reduction to zero of the incidence of a specified disease in a defined geographical area as a result of deliberate efforts; continued intervention measures are required. Example: neonatal tetanus.

Elimination of infections: Reduction to zero of the incidence of infection caused by a specific agent in a defined geographical area as a result of deliberate efforts; continued measures to prevent re-establishment of transmission are required. Example: measles, poliomyelitis.

Eradication: Permanent reduction to zero of the worldwide incidence of infection caused by a specific agent as a result of deliberate efforts; intervention measures are no longer needed. Example: smallpox.

Extinction: The specific infectious agent no longer exists in nature or in the laboratory. Example: none.

Based on Dowdle's criteria ¹⁸ the WHO definition of elimination is blatantly inappropriate. Based on standard epidemiological terminology a target of less than 1 new case per 10,000 would perhaps be more appropriate as a control target, rather than a definition of elimination.

The term elimination in popular discourse does not imply simply reduced. The Oxford English Dictionary defines elimination as "*The complete removal or destruction of something*".²² Semantics are vitally important in this instance as it impacts the awareness, focus and commitment of individuals, groups and governments to combating HD.¹⁰ Lockwood discusses the unfortunate impact of the use of the term elimination in relation to leprosy:

The rhetoric on elimination has discouraged dermatologists from engaging with leprosy programmes, even though they may be diagnosing cases in the private sector, because they believe leprosy is eliminated. Academic work on leprosy has declined; it rarely figures in medical school curriculums even in endemic countries, and research has declined. Young researchers perceive that the disease is eliminated. ¹⁰

The fourth issue in relation to WHO declaration of leprosy having been eliminated relates to consistent under-reporting of the condition. Concerns over under-reporting of leprosy cases are not confined to any one country. For example, Pedrosa et al. conducted a cross-sectional study of 34,547 schoolchildren aged under 15 years in the Amazon region of Brazil.¹⁹ This was followed by an active case finding exercise among household contacts for children identified with leprosy. The researchers noted a prevalence rate of 11.58 per 10,000. Alarmingly, this rate was 17 times higher than the registered rate.¹⁹ Lockwood et al. observed similar issues in India noting the blatant switch to voluntary rather than mandatory reporting of cases combined with a cessation of active case seeking in order to meet the WHO target:

The difference between the reported and observed estimates suggests that up to half of India's leprosy cases are not being reported. India has been reporting about 130,000 new cases a year, which keeps it safely in the eliminated leprosy category. There is therefore no incentive to find new cases.¹⁰

In 2010 the Eighth WHO Expert Committee on Leprosy recommended the use of a new international indicator to monitor leprosy.²³ The metric proposed was a Case Detection Rate (CDR) of Grade 2 Disability (G2D) cases per 1 million inhabitants.²⁴⁻²⁶ Box 2 details the disability grading system used by the WHO. A target CDR-G2D of 1 per million has been proposed.^{6,23,24}

Box 2. WHO Disability Grading System ²⁴

Hands and Feet

Grade 0 No anaesthesia, no visible deformity or damage

Grade 1 Anaesthesia present, but no visible deformity or damage

Grade 2 Visible deformity or damage present

Eyes

Grade 0 No eye problem due to leprosy; no evidence of visual loss

Grade 1 Eye problems due to leprosy present, but vision not severely affected as a result (vision: 6/60 or better; can count fingers at 6 metres).

Grade 2 Severe visual impairment (vision worse than 6/60; inability to count fingers at 6 metres); also includes lagophthalmos, iridocyclitis and corneal opacities.

However, it has been suggested that this revised metric may be equally problematic:

However, the CDR-G2D is less precise than the CDR due to small numbers in the numerator and this difference will make it difficult to use for monitoring small areas, i.e., small countries and local control programmes. Moreover, the CDR-G2D is influenced by early and late diagnoses and by the total incidence of leprosy, as measured by the total CDR.⁶

Despite current strategies,²⁷ leprosy remains a significant global threat,²⁸ particularly in an era of growing microbial resistance.²⁹ Although MDT has significantly reduced the global incidence of leprosy, official figures still record approximately 200,000 new cases per year. Evidence suggests that this figure may be a gross underestimate of the actual incidence of the disease. The WHO decision to arbitrarily adopt Elimination as a term for control is a misnomer that has served to minimize an important and ongoing issue that will undoubtedly have tragic consequences. The more recent adoption of a new measure based on CDR-G2D by the WHO appears equally problematic.

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