Government of **Western Australia**
North Metropolitan Health Service
Mental Health, Public Health and Dental Services

Guidelines for the Diagnosis, Management and Prevention of Leprosy

Western Australian Tuberculosis Control Program

Version 1.1

April 2023

Version control

1. As new information becomes available relevant to leprosy in WA, this guideline will require updating.
2. This guideline is approved by the North Metropolitan Health Service, Mental Health, Public Health and Dental Services, Director of Public Health.
3. Amendments shall be detailed in the revision history table below following endorsement.
4. This document has been compiled and informed by the documents, and other resources, listed in the References at the end of each chapter.

Revision History		
Version	**Date**	**Changes/Comments**
1.0	February 2019	Initial Endorsement
1.1	April 2023	• General formatting, grammar and references • Update Chapter 2 - Epidemiology • Update Chapter 3 - Global Considerations • Update Table 5 - Interpretation of sensory testing with monofilaments • Update to Chapter 5 – 5.2 Laboratory diagnosis, 5.4 Disease Classification and Assessment • Update to Chapter 7 – Treatment of ENL • Update Chapter 10 – 10.3 Chemoprophylaxis • Update Appendix 2 section 6 formatting • Addition of Appendix 8 K6 Scale

Contents

Abbreviations

AFB	Acid Fast Bacilli
BB	Borderline-borderline
BCG	Bacillus Calmette-Guerin
BI	Bacillary index
BL	Borderline-lepromatous
BT	Borderline-tuberculoid
CDCD	Communicable Disease Control Directorate
CDNA	Communicable Diseases Network Australia
CMI	Cell mediated immunity
DN4	Douleur Neuropathique en 4
DNA	Deoxyribonucleic Acid
DOT	Directly observed therapy
EHF	Eye hand foot score
ENL	Erythema nodosum leprosum
FD-MDT	Fixed duration multi drug therapy
G2D	Grade 2 Disability
GLP	Global Leprosy Program
Hpf	High power field
HRUS	High resolution ultrasonography
ISF	Impairment severity form
LL	Lepromatous-leprosy
MDT	Multi drug therapy
MI	Morphological index
MRC	Medical Research Council
MRL	Mycobacterium Reference Laboratory
NCS	Nerve conduction studies
NFI	Nerve function injury
NNDSS	National Notifiable Diseases Surveillance System
PCR	Polymerase Chain Reaction
PEP	Post exposure prophylaxis
POD	Prevention of Disability
QEII	Queen Elizabeth II (Sir Charles Gairdner Hospital)
ROM	Rifampicin Ofloxacin Minocycline
SDR	Single dose rifampicin
SSS	Slit skin smear
ST	Sensory testing
T1R	Type 1 reaction
TB	Tuberculosis
TT	Tuberculoid
VMT-ST	Voluntary muscle testing-sensory testing

WA	Western Australia
WANIDD	Western Australian Notifiable Infectious Disease Database
WGS	Whole Genome Sequencing
WHO	World Health Organization

Chapter 1 Introduction

Leprosy (also known as hansen's disease) is chronic granulomatous disease caused by infection with *Mycobacterium leprae*. The disease is characterised by skin lesions and nerve damage, with involvement of the respiratory tract, eyes, and other organs occurring less commonly.

The diagnosis of leprosy is based primarily on the clinical findings of peripheral nerve enlargement, characteristic skin lesions (hypoanaesthetic hypopigmented or reddish patch), and compatible microbiology (detection of acid-fast bacilli in skin smears or biopsies) or histopathology. The disease is classified broadly into two categories based on the number of skin lesions; paucibacillary leprosy (2-5 lesions) and multibacillary leprosy (6 or more lesions). A three-drug regimen is used for treatment, with duration of therapy determined by disease classification.

Case management is an important component of treatment, ensuring optimisation of drug therapy, early detection of lepra reactions and provision of social support. Identification and surveillance of contacts plays an important role in the prevention of leprosy, facilitating early diagnosis of secondary cases, and provision of chemoprophylaxis if indicated.

The clinical manifestations of leprosy are determined largely by the host immune response to *M.leprae*, with the spectrum of disease ranging from localised cutaneous disease, to disseminated disease with widespread skin lesions and nerve involvement.

While leprosy is not considered to be a highly contagious disease, the dynamics of transmission are not well understood. An individual's susceptibility to disease is thought to be influenced by genetic determinants, with the majority of people unlikely to develop infection if exposed. The incubation period is highly variable and potentially very long, ranging from 2-30 years. The sequelae of disease including stigma, psychological ramifications and deformity contribute to a significant burden, which affects the patient well beyond the completion of drug therapy. A long-term sustainable approach is therefore required to address the individual and public health consequences of leprosy.

In areas of low incidence, such as Australia, waning expertise and awareness of leprosy can lead to a delay in diagnosis. Clinical examination of the patient is of great importance for both diagnosis and monitoring of leprosy (reflected in further detail in Chapter 5).

Leprosy is frequently complicated by acute immunological reactions, called lepra reactions, which are the main cause of nerve damage and subsequent deformity and disability. Early diagnosis and treatment of leprosy and associated immunological reactions are required to prevent permanent disability.

Leprosy in Western Australia (WA) is managed primarily through a central clinic at the Anita Clayton Centre (Western Australia Tuberculosis Control Program), in collaboration with regional population/public health units, community health centres, regional Physician teams and district medical officers throughout the state.

While guidelines for leprosy control have been published in other Australian States and Territories, a guideline is also needed to address the unique cultural and geographic features of leprosy in WA. The vast land area of WA poses challenges for service delivery, with more than half of the leprosy cases occurring in remote and very remote parts of the state.

The guidelines are based on thorough literature review, expert opinion, and the recently published World Health Organization (WHO) Guidelines for the Diagnosis, Treatment and Prevention of Leprosy[1]. The WHO *Global Leprosy Strategy 2016-2020 "Accelerating towards a leprosy-free world"* [2] and the *WHO Operational Manual*[3] and *Monitoring and Evaluation Guide*[4] have been used to guide recommendations on contact tracing and other operational aspects of management, with the aim of promoting early detection and prompt initiation of treatment to prevent disability and reduce transmission. Recommendations made in these guidelines have been adapted to the local setting, taking into consideration an assessment of quality of evidence, harm-benefit balance, acceptability (for patients and health care workers), resource provision, feasibility, and effectiveness.

The most important changes included in these guidelines are;

1) The recommendation of a three-drug regimen for all leprosy patients, regardless of classification (see Chapter 6). This recommendation is in line with the WHO guidelines, the rationale being to simplify therapy, overcome problems associated with misclassification of leprosy, and the potential for beneficial anti-inflammatory effects of clofazimine in paucibacillary patients with neuritis.

2) Formal recommendation for the use of single dose rifampicin as chemoprophylaxis for contacts (>2 years) of newly diagnosed leprosy patients (see Chapter 10).

The guidelines aim to raise awareness of leprosy in WA, and to equip health care workers with the information necessary to suspect, diagnose and manage leprosy. In addition, there is a focus on prevention and operational aspects of leprosy control specific to WA, including provision of a framework for high quality data collection to facilitate quality assessment and optimisation of service delivery.

Through ongoing collaboration with involved stakeholders and sustained political commitment, the policy aims to ensure delivery of high-quality clinical care and reduce the burden of disease for all patients with leprosy in WA, with particular focus on early diagnosis and prompt initiation of treatment, contact surveillance, prevention of disability, and disease prevention through BCG vaccination and chemoprophylaxis.

References

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2. Regional Office for South-East Asia, World Health Organization. (2016). Global Leprosy Strategy 2016-2020: Accelerating towards a leprosy-free world. WHO Regional Office for South-East Asia. http://www.who.int/iris/handle/10665/208824

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Chapter 2 Epidemiology

2.1 Global epidemiology

Leprosy is an ancient disease that has historically been documented in nearly all parts of the world. Following the introduction of multidrug therapy in the 1980s, there has been a dramatic decline in the global prevalence of leprosy, from over 4 million, to 202, 256 at the end of 2019[1]. The global registered prevalence of leprosy at the end of 2020 was 129 192, however this is likely an under representation due to less detection and reporting during the COVID-19 pandemic[1]. Halving of the recommended treatment duration for multibacillary disease in 1998, in addition to changes in case definitions, has also had a significant impact on reported prevalence.

The elimination of leprosy as a public health problem is defined by the World Health Organisation (WHO) as a prevalence of less than one case per 10 000 population. This target was achieved at a global level in 2000, and in most countries by 2005, however elimination at a subnational level remains an ongoing challenge. The WHO Global Leprosy Strategy 2021 – 2030 has reset the target for leprosy elimination, now defined as no new autochthonous cases as a result of interruption of transmission.[2]

Despite these prevalence figures appearing promising, leprosy disease burden is measured more accurately by assessing the new case detection rate, number of new cases detected, and rates of grade 2 disability (G2D) in new cases.

Following a fall in new case detection between 2002 and 2005, there has been a stabilisation, with a gradual decline of approximately 2% per year, with a small increase from 2015 to 2016.

In 2019, 202 256 new cases of leprosy were detected, with 79% of cases in Indonesia, Brazil and India. Data from 2020 noted that multibacillary disease accounted for 67.3% of new cases, and 6.8% of all new cases occurred in children, indicating ongoing transmission. Whilst a number of regions have noted a reduction in the number of new cases with G2D at the time of diagnosis, there were still 13, 043 new cases with G2D at the time of diagnosis globally in 2020. Of note, 91.4% of new cases in global priority countries had G2D, indicating delay in diagnosis.

Until recently, the WHO has classified countries by utilising the absolute number of new cases as a measure of disease burden. In order to provide a more accurate measure of leprosy burden at a national and subnational level, the most recent Global Leprosy Strategy includes the following indicators to calculate a composite score to guide classification: prevalence, case detection, case detection rate, percentage of children among newly diagnosed cases (marker of ongoing transmission), percentage of G2D among newly diagnosed cases (marker of delayed case finding and therefore ongoing transmission), and G2D rate per million cases.

Based on this scoring system, 22 high burden countries have been identified, allowing prioritisation for resource allocation and control activities.

Table 1: Global list of 23 high burden countries for leprosy

Angola	**Madagascar**
Bangladesh	**Micronesia**
Brazil	**Mozambique**
Comoros	**Myanmar**
Cote d'Ivoire	**Nepal**
Democratic Republic of the Congo	**Nigeria**
Egypt	**Philippines**
Ethiopia	**South Sudan**
Micronesia	**Sri Lanka**
India	**Sudan**
Indonesia	**United Republic of Tanzania**
Kiribati	

2.2 National epidemiology

Leprosy is an uncommon disease in Australia, with a low notification rate of <0.1 per 100,000 population, equating to an average of 10 cases reported annually since 1996[3] (Figure 1). In 2016 there were 21 new notifications of leprosy, which is the highest number of notifications since 1991. The disease affects two vulnerable populations, migrants from leprosy endemic countries, and Aboriginal people. The majority of leprosy cases have been acquired overseas, with only a small proportion of cases acquired in Australia, exclusively in the Aboriginal population. Since 1925, Western Australia (WA) has reported the highest number of leprosy cases, followed by the Northern Territory and Queensland.

Between 2017 and August 2022 a total of 47 cases of leprosy have been notified to the NNDSS in Australia, with the majority of cases occurring in Western Australia, NSW and Queensland (Figure 2). It is important to note that case detection is highly dependent on a number of operational factors including robustness of contact tracing, active case finding and clinical expertise. The reported notifications in Australia are likely to be an underestimate of true prevalence due to limited allocated resources and health service factors related to the COVID-19 pandemic.

Figure 1: Notifications of leprosy, Australia, 1996-2016, by year and Indigenous status. Source: National Notifiable Diseases Surveillance System[15]

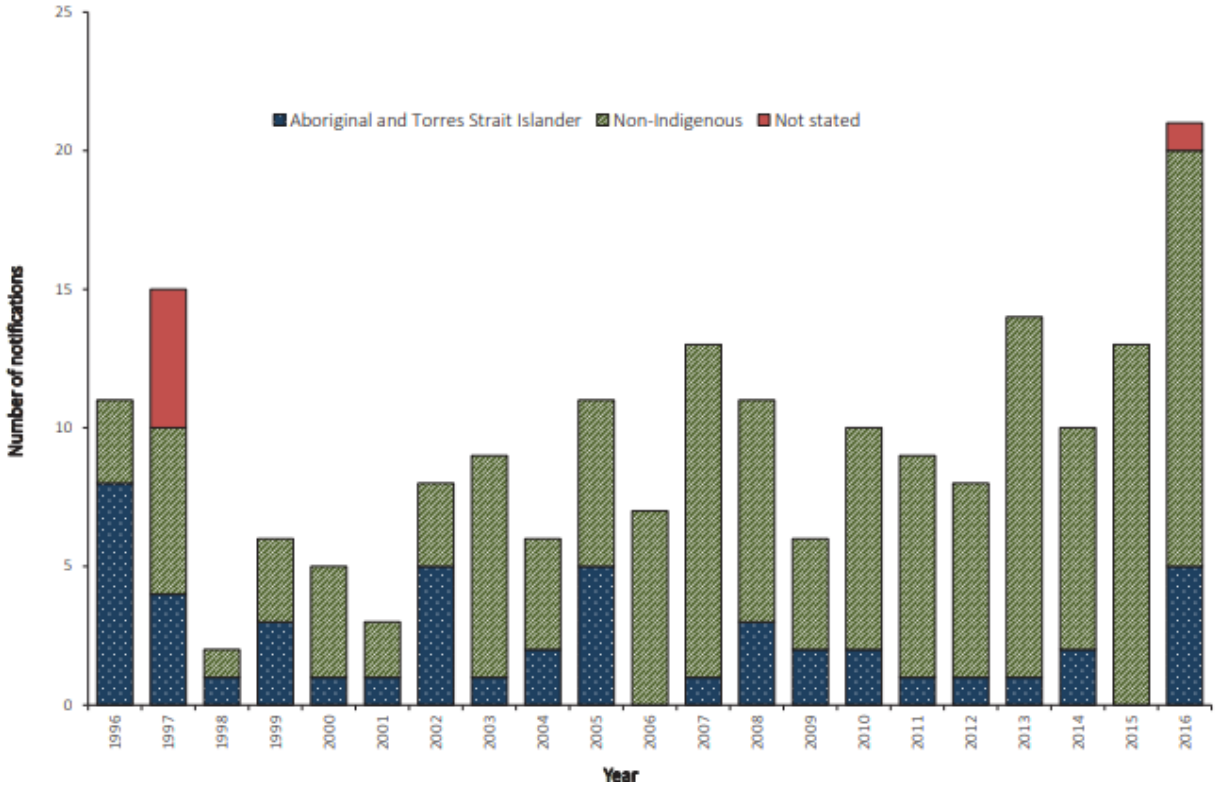
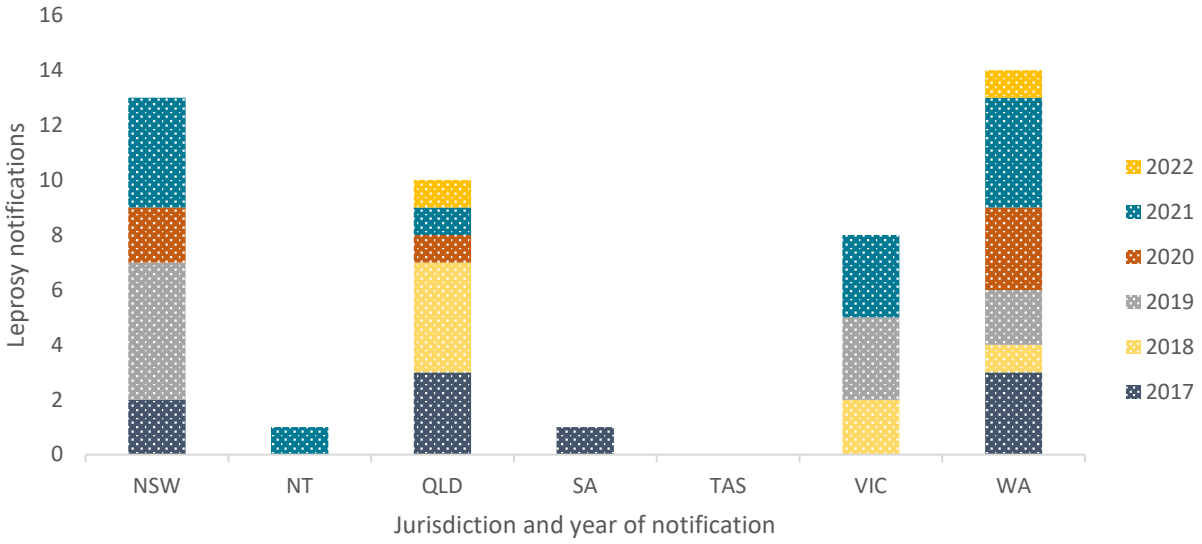


Figure 2: Leprosy notifications, by jurisdiction and year, 2016-2022 (5 August)



NB: Data extracted from the NNDSS on 5 August, by diagnosis date. Due to the dynamic nature of the NNDSS, data in this extract are subject to retrospective revision and may vary from data reported in published NNDSS reports and reports of notification data by states and territories.

2.3 Western Australia epidemiology

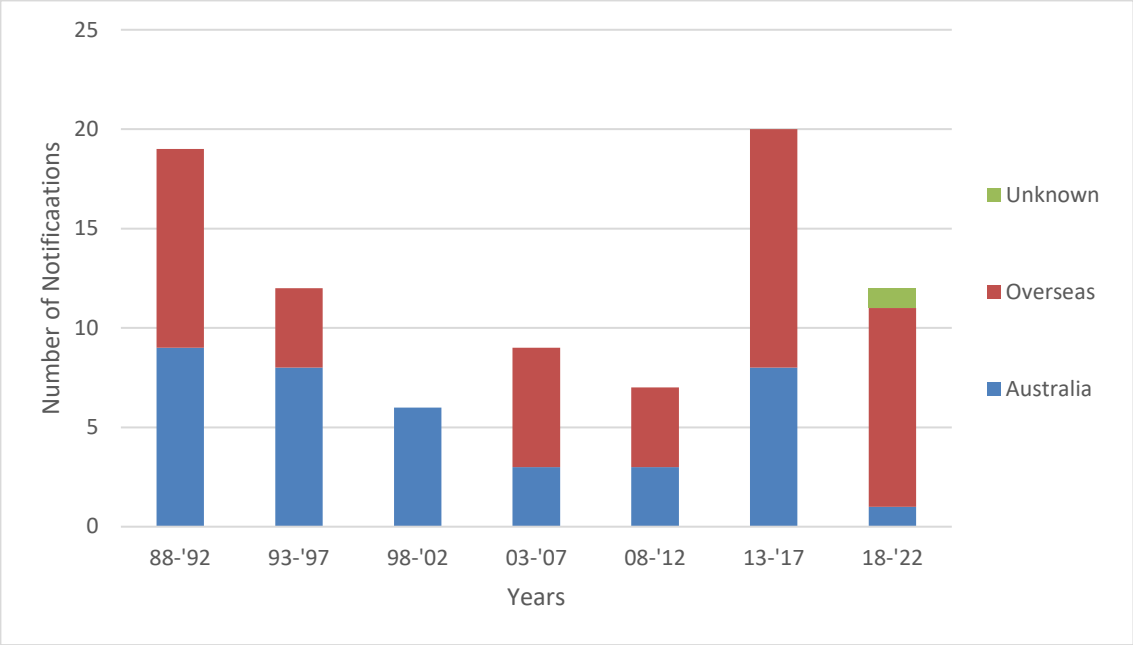
While the WA leprosy notification rate is extremely low at 0.1 per 100,000 population, pockets of higher endemicity exist within the state which are not reflected in state or national figures. The Kimberley Aboriginal population has a leprosy notification rate significantly higher than WA overall, highlighting the need for careful data collection, analysis, reporting and allocation of resources. The issue of addressing hotspots of leprosy at a subnational level in the push for elimination is addressed in the most recent Global Leprosy Strategy and operational Manual.

Leprosy is thought to have been introduced to WA, with the first recorded case in a Chinese migrant in Roebourne in 1889, and the first Aboriginal case occurring shortly after near Broome in 1893. There is no evidence that Aboriginal Australians were affected by mycobacterial disease prior to settlement, however leprosy infection spread rapidly throughout the Kimberley, with approximately 10% of the population affected by the 1950s. A leprosarium was established near Derby in 1935, with approximately 1200 Aboriginal patients being treated there before its closure in 1986.

Since 1988, there have been 85 cases of leprosy notified in WA, with 44% of cases being Aboriginal and the majority of these residing in remote or very remote parts of the state. Of cases acquired overseas, 48% were from the South East Asian WHO region, and 43% from the Western Pacific region. The male to female ratio was 1.3:1, with an age range of 11-77. Multibacillary disease accounted for 62% of cases and there is clear evidence of ongoing transmission of leprosy in WA, with secondary cases occurring in contacts.

Figure 3 shows leprosy notifications in WA between 1988 and 2022 by place of acquisition. Of cases acquired in Australia during this period all except one were in Aboriginal Australians. While there has been a trend toward a predominance of overseas acquisition, an increase in notifications in cases acquired in Australia occurred between 2013 and 2017 due to enhanced surveillance activity in the Kimberley region of WA.

Figure 3: Notifications of leprosy, Western Australia, 1988-2022 by place of acquisition



Leprosy enhanced surveillance commenced at the WA TB Control Program in 2013. Since this time 32 cases of leprosy have been notified, of which 35% had G2D at the time of diagnosis, 16% were retreatment cases, and 65% suffered lepra reactions either before, during or after completion of therapy.

These figures highlight the need for ongoing surveillance of both contacts and previously treated patients, as well as education to facilitate opportunistic diagnosis by health care workers. In addition, there is a need for strengthened referral pathways to ensure optimal care, particularly for those suffering lepra reactions and those residing in remote locations, in order to prevent disability. The guidelines aim to address these issues to optimise outcomes, particularly for patients residing in remote parts of the state.

References

1. Global leprosy update, 2019: time to step-up prevention initiatives. Weekly Epidemiological Record. 2020; 95(36):417- 440.

2. Regional Office for South-East Asia, World Health Organization. (2021). Global Leprosy Strategy 2021-2030: Towards Zero Leprosy. WHO Regional Office for South-East Asia. Towards zero leprosy. Global leprosy (Hansen's Disease) strategy 2021–2030 (who.int)

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Chapter 3 Global considerations

The epidemiology and burden of leprosy are highly variable at a national and subnational level. Therefore, adaptation and implementation of guidelines at a local level is essential to achieve optimal outcomes.

In December 2020, the WHO published 'Towards Zero Leprosy – Global Leprosy Strategy 2021 – 2030' , highlighting the shift in leprosy focus to interrupting leprosy transmission and achieving zero autochthonous cases with the eventual elimination of leprosy[1].

Targets of the strategy for the end of 2030 include:[1]

- To have 120 countries with zero new autochthonous cases
- 70% reduction in the annual number of new cases detected
- 90% reduction in rate per million population of new cases with G2D
- 90% reduction in rate per million children of new child cases with leprosy

The Global Leprosy Strategy aims to employ several strategic pillars to achieve these targets, including[1];

1. Implementing integrated, country-owned zero leprosy roadmaps in all endemic countries
2. Scaling up leprosy prevention alongside integrated active case detection
3. Managing leprosy and its complications and preventing new disability
4. Combating stigma and ensuring human rights are respected.

The *WHO Guidelines for the Diagnosis, Treatment and Prevention of Leprosy* published in 2018, is also accompanied by an *Operational Manual*[2] and *Monitoring and Evaluation Guide,*[3] assisting leprosy coordinators with program implementation and evaluation.

Global leprosy data collection by the WHO has been simplified with the introduction of an electronic open-source software tool, encouraging improved reporting and more accurate measures of disease burden on which to base program recommendations.

The WHO Guidelines for the Diagnosis, Treatment and Prevention of Leprosy[4] were developed through a formal and robust guideline development process based on the GRADE (Grading of Recommendations Assessment, Development and Evaluation) approach. These guidelines provide comprehensive clinical recommendations with a global focus, but particularly relevant for low and middle-income countries with high leprosy burden. The Western Australian Leprosy Guidelines provide recommendations adapted to local epidemiology and resources.

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Chapter 4 Case definition

The Australian Government Department of Health released an updated leprosy case definition in 2013[1]. Leprosy cases meeting this definition should be notified to the National Notifiable Diseases Surveillance System (NNDSS) (see Chapter 12).

Australian leprosy case definition:

A confirmed case requires:

1. Laboratory definitive evidence OR laboratory suggestive evidence;

   AND

2. Clinical evidence:

Laboratory definitive evidence

- Detection of *Mycobacterium leprae* by nucleic acid testing from the ear lobe or other relevant specimens

Laboratory suggestive evidence

- Demonstration of characteristic acid-fast bacilli in slit skin smears and biopsies prepared from the ear lobe or other relevant sites

  OR

- Histopathological report from skin or nerve biopsy compatible with leprosy examined by an anatomical pathologist or specialist microbiologist experienced in leprosy diagnosis.

Clinical evidence

- Compatible nerve conduction studies

  OR

- Peripheral nerve enlargement

  OR

- Loss of neurological function not attributable to trauma or other disease process

  OR

- Hypopigmented or reddish skin lesions with definite loss of sensation

World Health Organization (WHO) definition:

Presence of at least one of three cardinal signs of leprosy:

1. Definite loss of sensation in pale (hypopigmented) or reddish skin patch.

2. Thickened or enlarged peripheral nerve with loss of sensation and/or weakness of the muscles supplied by that nerve.

3. Presence of acid-fast bacilli in a slit skin smear.

References

1. Australian Government Department of Health. Leprosy case definition. http://www.health.gov.au/internet/main/publishing.nsf/content/cda-surveil-nndss-casedefs-cd_leprosy.htm Accessed 9 May 2018.

Chapter 5 Diagnosis of leprosy

5.1 Clinical diagnosis

5.1.1 Presentation

The presenting features of leprosy are highly variable, and dependent upon the host immune response to the pathogen, as well as duration of infection.

Common presenting symptoms and signs include:

- skin lesions - pale (hypo-pigmented), coppery, or reddish (erythematous) patch
- shiny thickened skin on the face (lion/leonine faces)
- swelling or nodules in the face and earlobes
- loss of sensation with or without skin lesions
- loss of sweating in a skin lesion
- numbness or tingling of hands or feet
- weakness of hands, eyelids and feet
- injury secondary to nerve injury e.g. burn, ulceration
- visible deformity of the hands, feet and eyes
- nerve pain, acute nerve palsy, inflamed skin lesions, eye pain or fever in association with lepra reaction.

The diagnosis of leprosy is heavily reliant on clinical symptoms and signs, especially in settings where access to slit skin smears and laboratory tests are limited. At least one of the three cardinal signs of leprosy must be present to make a diagnosis, with laboratory evidence required to meet the Australian National Notifiable Diseases Surveillance System definition of a confirmed case (see Chapter 4).

Cardinal signs of leprosy

1. **Skin lesions**: Hypo-pigmented, coppery or erythematous skin lesions with reduced or absent sensation

2. **Nerve thickening**: Involvement of peripheral nerves in sites of predilection, as demonstrated by thickening or enlargement, with or without loss of sensation and weakness of the muscles supplied by that nerve

3. **Demonstration of *M.leprae***: either by slit skin smear (SSS), biopsy or polymerase chain reaction (PCR).

Standard questions asked as part of the history taking:

- Have you been treated for leprosy before?
- Has anyone in your family or community had leprosy?
- Have you noticed any skin lesions?
- Have you noticed any loss of sensation in your face, hands or feet?
- Have you noticed any dryness of the palms of your hands or soles of your feet?
- Have you noticed any weakness in your hands or feet?
- Have you noticed any weakness in your face or difficultly closing your eyes?
- Have you noticed any new sensations of pins and needles or tingling in your face, hands or feet?
- Have you noticed any new pain sensations such as burning or shooting pain?

If the patient answers yes to any of the questions, enquire further as to location of the symptom and how long it has been present for.

> When a diagnosis of leprosy is suspected or proven, careful examination of the skin and peripheral nerves should be done, in addition to testing sensory and motor function of commonly affected nerves. Findings should be documented in detail in the templates located in Appendices 1, 2 or 3, with a guide for examination in Appendix 4.

5.1.2 Skin examination

The entire body should be examined, preferably in natural sunlight. Lesions can appear anywhere on the body but tend to spare the axillae and groin.

Findings of the skin examination should be recorded on the template in Appendix 1.

Skin lesions in leprosy can be:

- single or multiple
- well defined or poorly defined
- variable in size
- macules (flat lesions)
- papules (raised lesions)
- nodular
- infiltrative
- hypo-pigmented (lighter in colour compared to surrounding skin)
- erythematous, reddish or coppery
- hypo-anaesthetic (reduced sensation for temperature, touch and/or pain)
- dry with loss of sweating and/or loss of hair.

The following features are not suggestive of leprosy:

- White (depigmented), dark red or black lesions.
- Scaly lesions.
- Lesions present since birth.
- Lesions which appear or disappear suddenly (exception with lepra reaction).

- Pain or itchiness (exception with pain associated with lepra reactions).

Testing sensation in a skin lesion

If a skin lesion is present, sensation should be tested using light touch and temperature. Light touch sensation is tested by using a cotton wool ball rolled to a point to touch the skin at a 90-degree angle. It is important not to stroke the skin. Demonstrate the process on an area of normal skin with the patient's eyes open, asking them to point to the area where the touch is felt. Light touch in the skin lesion is then tested with the patient's eyes closed. Temperature sensation can be tested using glass test tubes or cups filled with cold water (25 degrees Celsius) and warm water (45 degrees Celsius).

It is important to note that skin lesions on the face and in lepromatous leprosy may have normal sensation. Testing light touch in areas with callus or thickened skin is not reliable, and interpretation of findings in children can be difficult.

Digital photography of skin lesions at the time of diagnosis and at the completion of therapy with patient consent is recommended. Photography can also be a useful tool to monitor suspicious lesions during the diagnostic process.

5.1.3 Nerve palpation

Nerve palpation is an important part of the clinical assessment, with nerve enlargement being one of the cardinal signs of leprosy. Detailed information and photographs to guide palpation of specific peripheral nerves can be found in Appendix 4.

M.leprae grows best at temperatures lower than that of the body's core temperature, and therefore affects nerves at their coolest and most superficial location. This means that affected nerves are generally easily felt.

The peripheral nerves most commonly affected are:

- Ulnar nerve at the elbow
- Common peroneal (lateral popliteal) nerve at the knee
- Posterior tibial nerve at the ankle
- Median nerve at the wrist
- Facial nerve near the ear.

Other nerves that may be affected include, but are not limited to, the greater auricular, sural and radial nerves.

The ulnar nerve at the elbow is the easiest to palpate, with a section of nerve (up to 10cm) able to be felt above the elbow. This allows for detailed assessment of features of thickening, such as nodularity or rope like consistency. A normal nerve feels compressible, and generally flat. Table 2 outlines the method for palpating peripheral nerves.

Table 2: Method for nerve palpation

Method for nerve palpation:

1. position patient and examiner correctly, ideally seated comfortably
2. enquire as to whether any of the nerves are tender
3. locate the nerve
4. use the pulp of two or three fingers to roll the nerve gently against the bone (do not use the tips of the fingers)
5. observe the patient's face during examination to detect tenderness feel along the nerve as far as possible in both directions
6. compare features of nerve with the opposite side

Assess for:

- Thickness
- Consistency (may be rope like in advanced disease)
- Tenderness
- Irregularity (nodular, beaded)
- Number of nerves involved
- Symmetry

Nerve enlargement is scored as follows1:

- None (N)
- Possible (P)
- Definite (D)

5.1.4 Assessment of nerve function

Nerve involvement with leprosy can cause abnormalities in autonomic, sensory and motor function, with motor loss generally occurring later in the course of the disease. Both cutaneous nerves and peripheral nerve trunks can be affected. While nerve damage occurs slowly, patients may present with acute deterioration in the setting of lepra reactions.

Delayed detection of leprosy is strongly associated with an increased risk of nerve function impairment at the time of diagnosis,[2,3] which in turn is a strong predictor of the risk of further lepra reactions and progressive neuropathy.[4,5]

Careful standardised assessment of nerve function is an important part of the diagnostic work up of a patient, facilitating prompt treatment of reactional states when detected, and a means of allowing comparison in function over time to assist in early detection of deterioration.

Assessment of peripheral nerve function should be made at the time of initial presentation, as well as monthly while on treatment and following completion of therapy as outlined in Figure 5 in Chapter 6.5.5.

The presence of autonomic dysfunction, sensory loss, weakness and secondary impairments are diagnostic of nerve damage and should be carefully assessed in the patient suspected of having leprosy.

Table 3: Primary and Secondary impairments associated with peripheral nerve damage in leprosy

Peripheral nerve	Primary impairment		Secondary impairment
	Sensory	Motor	
Ulnar nerve *	LF and ulnar half of RF	Weakness of LF abduction	Ulnar claw Contractures
Radial nerve *	Dorsum hand (thumb, IF MF, half RF)	Weakness of wrist and finger extension	Wrist drop
Median nerve *	Palmar aspect (thumb, IF, MF, half RF)	Weakness of thumb abduction	Wasting of interossei and clawing Contractures
Cranial nerve VII (zygomatic branch)		Weakness of orbicularis oris: abnormal eye closure	Lagophthalmos Corneal ulceration / inflammation Blindness
Cranial nerve V	Corneal anaesthesia: impaired blink reflex		Corneal ulceration / inflammation Blindness
Common peroneal nerve	Lateral leg and dorsum of foot	Weakness of dorsiflexion of the foot	Foot drop
Posterior tibial nerve *	Plantar aspect of foot		Claw toes

IF = index finger, LF=little finger, RF=ring finger, MF=middle finger

*In addition, damage to autonomic fibres results in loss of sweating and loss of hair with subsequent predisposition to callus formation and fissuring of the skin.

5.1.4.1 Sensory testing:

The assessment of sensory function is important for the diagnosis of leprosy, as well as for monitoring the patient while on therapy to ensure early detection of neuritis and lepra reactions.

The pattern of sensory neuropathy in leprosy often varies according to the type of leprosy.

Table 4: Patterns of sensory neuropathy in leprosy

Leprosy classification	Pattern of sensory neuropathy	Mechanism of neuropathy
Indeterminate	Localised cutaneous sensory loss, no nerve trunk involvement	Injury to cutaneous nerve endings
Tuberculoid	Mononeuropathy, localised sensory loss in distribution of one peripheral nerve trunk	Involvement of single peripheral nerve
Borderline states, early Lepromatous leprosy	Multiple mononeuropathy	Multiple peripheral nerves affected
Lepromatous leprosy	Polyneuropathy, often in a glove and stocking distribution	Diffuse nerve involvement

Sensory testing is performed on the hands and feet in the areas supplied by the peripheral nerves commonly affected in leprosy. Testing is done preferably using standardised, graded nylon monofilaments[6] as outlined in Appendix 2 Referral Centre Template. In the field situation, or where monofilaments are unavailable, the tip of a ballpoint pen can be used, and is equivalent to a 10g (orange) monofilament[7] as outline in Appendix 3 Peripheral Centre Template. When using a pen to test sensation, touch the skin lightly with the pen from a perpendicular angle. It is important just to use the light weight of the pen, and to avoid pressing and indenting the skin. For further guidance see Appendix 4.

Six colour coded monofilaments are used to test sensation in defined points, providing a means of monitoring sensory function over time. Table 5 outlines the features, force and interpretation of sensory testing with monofilaments.

Table 5: Interpretation of sensory testing with monofilaments

Colour	Force	Hands & dorsal foot	Sole of the foot
Green	0.07g	Normal	Normal
Blue	0.4g	Diminished light touch	Normal
Purple	2.0g	Diminished protective sensation ↑risk of injury and ulceration	Normal - reduced sensation
Dark Red	4.0g	Loss of protective sensation	Diminished protective sensation ↑risk of injury and ulceration
Orange	10.0g	Loss of protective sensation	Loss of protective sensation
Bright red	300.0g	Deep pressure sensation only	Deep pressure sensation only

When testing the hands, all monofilaments should be used, starting with the finest filament. For testing sensation on the soles of the feet, omit 0.05g (Green) and 0.2g (Blue) monofilaments. he finest/lightest filament felt is recorded.

Practice notes for using monofilaments:

- Ensure the monofilament is in good condition.
- Apply to the points indicated on the assessment form.
- Apply the monofilament at a 90-degree angle to the skin surface.
- Pressure should be slow and gentle to form a “C” shape in the monofilament.

5.1.4.2 Assessing motor function:

- Motor function is assessed using the Voluntary Muscle Test (VMT) and is scored using the modified Medical Research Council (MRC) Scale or alternative simplified system (Table 6).

Table 6: Modified MRC grading score for VMT[8]

Alternative scoring	VMT score	Description
Strong (S)	Grade 5	Full ROM against gravity and maximum resistance
Weak (W)	Grade 4	Full ROM against gravity and moderate resistance
	Grade 3	Full ROM against gravity, not added resistance
	Grade 2	Partial range of movement, no resistance, gravity eliminated
	Grade 1	Visible or palpable contraction with no movement
Paralysed (P)	Grade 0	Complete paralysis

ROM=range of movement

Considerations for performing a VMT:

- Ensure correct movement to test the selected muscle (patients may compensate for weakness in one muscle by using others).
- Test muscle strength using appropriate resistance (for example, the examiner should use their own little finger to assess strength of little finger abduction in the patient).

The combination of sensory testing and voluntary muscle testing is called VMT-ST. Templates for recording findings can be found in Appendices 2 and 3**,** with a guide to performing VMT-ST in Appendix 4**.** The referral centre template (Appendix 2) is more detailed and requires greater clinical expertise. The peripheral centre template (Appendix 3) is designed for simple assessment at the community level.

5.1.5 Pain assessment

Neuropathic pain is a common feature of leprosy, occurring due to nerve inflammation, nerve entrapment and nerve damage, with or without other symptoms of lepra reaction.

Neuropathic symptoms can include:

- paresthesia
- pain: burning, stabbing, squeezing, stinging, aching, shooting
- stimulus induced pain.

While the pain is often acute, chronic pain can develop in patients who have completed therapy, with significant impact on their quality of life.

Pain is most easily assessed using a visual analog scale, which is incorporated into the VMT-ST template. Douleur Neuropathique en 4 questions (DN4) is a screening tool for

neuropathic pain with a higher sensitivity and specificity and can be used if clarification is required (Appendix 5).

5.1.6 Eye Examination

Leprosy remains one of the leading causes of preventable blindness and visual disability, with studies reporting rates of potentially sight-threatening leprosy related eye pathology varying between 11% and 48%[9,10,11] and prevalence of blindness between 2.8% and 3.2%.[10,12]

Like all leprosy related disability, eye complications can develop before, during and after the completion of drug therapy.

It is therefore important to perform careful examination of the eyes in a suspected case of leprosy, during therapy and following completion of treatment in patients with risk factors for progression.

Patients with MB leprosy and any patient with eye abnormality should be seen by an ophthalmologist at the time of diagnosis, annually during treatment and for a minimum of five years following completion of treatment.

Eye involvement in leprosy can occur in three ways:

1. Direct infiltration of the eye and surrounding tissues.
2. Inflammation as part of lepra reactions.
3. Involvement of cranial nerves V and VII.

Risk factors for eye involvement in leprosy:

- Multibacillary disease
- Presence of other deformities
- Facial skin lesion
- Lepra reaction involving the face
- Increasing age

Method for examining the eyes:

Inspect for:

- loss of eyebrows (particularly outer 1/3) (Madarosis)
- loss of eyelashes
- inability to close the eyes (Lagophthalmos)
- infrequent or abnormal blink (reduced corneal sensation)
- dryness or scarring of the cornea (exposure keratitis)
- inflammation / infection of the lacrimal sac (between eye and nose)
- redness of the eye (iridocyclitis)
- sagging / turned out lower lid (ectropion)
- lid turned in towards eyeball (entropion)
- turned in lashes (trichiasis), often associated with corneal scarring

- lens opacity (cataract)
- abnormal pupil shape
- facial skin lesions.

Examine:

- Ask the patient to close their eyes (as if asleep).
    - Look for a gap between the lids.
    - If present, estimate the gap in millimetres.

- If there is no gap visible, ask the patient to close their eyes tightly.
    - Use your index finger and thumb to try to open the lids.
    - If you are easily able to open the eye, the muscles are weak.

- Abnormal lid closure is caused by involvement of cranial nerve VII.

- Take note of the number of times the patient blinks. If they blink infrequently i.e. less than twice per minute, it implies anaesthesia due to involvement of cranial nerve V the trigeminal nerve. For safety reasons, testing corneal sensation with cotton wool should not be performed.

- Measure visual acuity in both eyes with a Snellen chart.

Potentially sight-threatening ocular complications associated with leprosy:

- Cataract
- Glaucoma
- Lagophthalmos
- Reduced corneal sensation
- Corneal ulceration
- Iridocyclitis / uveitis
- Scleritis.

Occur more commonly in Borderline Tuberculoid and Tuberculoid forms of leprosy.

5.1.7 Leprosy in children

Leprosy in children is a marker of ongoing transmission within a community, and the *WHO Global Leprosy Strategy 2021 – 2030* emphasizes the importance of early diagnosis and reduction of disability in children.[13]

Leprosy in a child is often difficult to diagnose due to subtle skin findings and difficulties encountered performing a neurological examination.

Children are more likely to present with indeterminate or early leprosy with the following skin lesion features:

- Small to medium size (1-3 cm)
- Hypopigmented
- Ill-defined edges (particularly smaller lesions)
- Normal hair growth
- Sensation usually normal
- Often located on the face & extensor aspect of limbs

The diagnosis should be suspected in a child from a leprosy endemic region, or with a known family history of leprosy. A biopsy is required to confirm the diagnosis of indeterminate leprosy however this is often not acceptable from a cosmetic perspective for a facial lesion.

As approximately 75% of indeterminate lesions heal spontaneously, a reasonable approach is to apply topical antifungal for 4 weeks and observe for 6-12 months.

Changes in the lesion including development of well-defined margins, more marked hypopigmentation, and development of hypoanaesthesia are suggestive of progression into definite leprosy and requires further investigation.

5.2 Laboratory diagnosis

5.2.1 Introduction

The diagnosis of leprosy is hindered by the inability to culture *M.leprae* in the laboratory. While there is no true gold standard, full-thickness skin biopsy examined by an experienced pathologist is the most reliable and informative test. In many resource-poor settings, the diagnosis is purely clinical, sometimes supplemented with slit skin smear (SSS) results.

Polymerase Chain Reaction (PCR) offers enhanced sensitivity compared with biopsy and SSS. PathWest Laboratory Medicine has developed in-house PCR assays with confirmation by sequencing to increase diagnostic certainty. Collection of nasal swabs for PCR is now recommended for all cases in view of high sensitivity and ease of collection.

5.2.2 Slit skin smears

SSS are performed to detect *M.leprae* bacilli in tissue fluid of the skin. The test has low sensitivity (10-50% depending on expertise) but high specificity (100%). SSS have low diagnostic utility and are useful only in patients with borderline lepromatous and lepromatous leprosy, with smears often negative in many patients with other forms of leprosy. Despite this, SSS are relatively non-invasive, cheap and simple to perform, and help to identify and monitor patients who are most infectious and at highest risk of relapse.

Indications:

- For diagnosis in patients with suspected or diagnosed leprosy before commencing treatment.
- Monitoring response to therapy of multibacillary cases.
- Detecting relapse.

It is recommended that smears be taken from a minimum of three sites, avoiding the face for cosmetic reasons.

Suggested sites include:

- One ear lobe and two active lesions
    - Smears should be taken from the active edge if the lesion is distinct, and from the centre if the lesion is indistinct
- Extensor surface of elbow (s)
- Extensor surface of knee (s)
- Dorsal surface of fingers.

When utilising SSS for disease monitoring and detecting relapse, smears should be taken from previously positive sites.

Method for taking a slit skin smear:

1. Consider applying topical anaesthetic cream if sensation preserved in selected site.
2. Clean site with alcohol swab and allow to dry.
3. Roll site between index finger and thumb until it becomes bloodless.
4. Using a size 15 scalpel blade, make an incision 5mm long and 3mm deep.
5. Maintaining pressure with the fingers, turn the blade at a right angle to the incision and scrape the slanted edge of the blade several times in one direction to obtain tissue fluid.
6. Smear the fluid gently on the slide in a circle approximately 8-10mm in diameter.
7. Samples with visible blood content are unlikely to be useful. A second sample can be obtained after wiping the incision site with cotton wool while maintaining finger pressure.
8. Place a small dressing over the incision.
9. Fix the slide by passing the underside for 2 seconds over a naked flame.
10. Label the slide with patient details and site, and place in slide container.
11. Send to PathWest requesting acid-fast bacilli (AFB) microscopy for leprosy.

Nasal mucous membrane smears for microscopy can be obtained using a sterile cotton bud wiped firmly in the nasal passage. Prepare the smear as outlined from points 5-8 above. Nasal smears are often positive in patients with untreated lepromatous leprosy.

Laboratory reporting of skin smears / nasal smears

Samples are reviewed at the Mycobacterium Reference Laboratory (MRL) at the PathWest Laboratory Medicine Department at the Queen Elizabeth II (QEII) site.

Smears with AFB seen are reported with the number of bacilli quantified using a semi logarithmic score called the Bacillary Index (BI). The BI ranges from 1+ to 6+, depending on the number of bacilli seen in an average microscopic field using an oil immersion lens or high power field (hpf). Untreated lepromatous leprosy patients generally have a BI of 5+ or 6+, falling by approximately 0.75-1.0+ BI units per year with treatment.

Table 6: Bacillary index definitions for skin smears

Bacillary Index	Description
1+	1-10 bacilli in 100 hpf
2+	1-10 bacilli in 10 hpf
3+	1-10 bacilli in 1 hpf
4+	10-100 bacilli in 1 hpf
5+	100-1000 bacilli in 1 hpf

In addition to the BI, the laboratory will report a morphological index (MI) for specimens where AFB are seen. The MI is the percentage of all bacilli that appear viable. These bacilli stain solidly and are of standard shape and size.

Fragmented and granular bacilli are not considered viable and are often seen after several months of effective therapy. Occasionally globi are reported, describing clumps of bacilli, and are generally found in patients with lepromatous leprosy and high bacillary load.

The MI in an untreated multibacillary patient generally ranges from 25% to 75% and should fall to 0% after 4-6 months of effective treatment. It is important to note that the MI is a more useful measure of response to therapy and potential for transmission, with many patients, particularly those with high BI, remaining smear positive at the completion of therapy.

5.2.3 Skin biopsy

A full thickness skin biopsy is the preferred test when leprosy is suspected on clinical grounds, having higher sensitivity than SSS, particularly when examined by an experienced pathologist. While biopsy is more invasive than SSS, it has a number of advantages.

Advantages of skin biopsy over SSS

- Facilitates diagnosis of paucibacillary cases and those with low bacillary load not detectable by SSS.
- Allows classification according to Ridley-Jopling criteria
    - More accurate classification
    - Ensures appropriate therapy
    - More accurate prediction of prognosis
- Can identify lepra reactions.
- Facilitates diagnosis of conditions which mimic leprosy.
- Optimal specimen for molecular testing
    - Enhance sensitivity
    - Allow detection of drug resistant determining regions in certain cases.

Method for skin biopsy

1. Clean the site and inject local anaesthetic deep into the subcutaneous tissue around the biopsy site (do not inject intradermally as it ruins the biopsy).

2. Full thickness elliptical (1cm long) or punch (4mm) biopsy from the advancing margin of an active lesion or centre of a subtle hypopigmented lesion with indistinct edge (two biopsies are preferred if the punch technique is used to allow for optimal mycobacteriological processing).
3. Place a small part of the elliptical biopsy or one punch biopsy into sterile saline for microscopy and PCR.
4. Place the remainder of the biopsy, or second specimen (punch biopsies) in buffered formal saline (10%) formalin and request histopathology with Fite-Faraco stain for AFB and fungal stains.
5. Label the biopsies and send to PathWest Perth.

Multiple biopsies should be obtained if there are numerous skin lesions with varying morphology.

5.2.4 Nerve biopsy and fine needle aspiration.

Pure neuritic leprosy, sometimes called pure neural, primary neural, primary neuritic, or poly-neuritic leprosy, is a form of leprosy characterised by nerve involvement without skin lesions. It is not included in the WHO or Ridley-Jopling classification systems, and debate continues as to optimal treatment.

Pure neuritic leprosy is most frequently seen in patients of Indian or Nepalese ethnicity.[14] The ulnar, lateral popliteal, posterior tibial and sural nerves are most commonly affected, with patients presenting either with a mononeuritis or mononeuritis multiplex when several nerves are involved. While classic skin patches are absent, skin along the distribution of the involved nerve is often hypoanaesthetic and changes such as anhidrosis, xerosis, fissuring and trauma can be present.

Pure neuritic Leprosy

When to suspect

- Patient from leprosy endemic area, particularly India.
- Mononeuritis or mononeuritis multiplex.

Clinical features

- Thickened, tender peripheral nerves.
- Sensory, motor or autonomic impairment.
- Skin lesions absent.

Diagnosis

- Nerve biopsy is gold standard.
- Fine needle aspiration cytology from nerve.
- Nerve conduction studies often non-specific.
- High resolution ultrasonography.
- SSS negative.

Nerve biopsy is the gold standard for diagnosis, but is prone to sampling error, with low sensitivity and the risk of permanent nerve damage. Biopsies are most commonly taken from a branch of a purely sensory nerve such as the superficial sensory radial nerve branch at the wrist, ulnar cutaneous nerve near the hand, and sural nerve near the ankle. The procedure is generally done by an experienced neurologist or neurosurgeon. Specimens should be sent to anatomical pathology for haematoxylin and eosin stain and Fite-Faraco stain.

Fine needle aspiration cytology is a less invasive, simpler method that can be used prior to nerve biopsy if pure neuritic leprosy is suspected.[15] The sensitivity is low but improved with use of *M.leprae* PCR in addition to cytology and Fite-Faraco stain for AFB.[16]

5.2.5 Molecular testing

***M.leprae* PCR**

PCR is a rapid, reliable, sensitive and specific tool which can be used to detect *M.leprae* deoxyribonucleic acid (DNA) in a variety of tissue sources including skin biopsy samples, oral or nasal swabs, SSS fluid,[17] and whole blood. Multi-copy targets are increasingly used to improve sensitivity and current assays are reported to have a lower limit of detection than microscopy. PathWest is unable to validate its in-house Leprosy PCR to NPAAC standards due to the infrequency of cases. All PCR detections are confirmed by sequencing.

While skin biopsy in saline is the preferred sample for optimal DNA extraction[18] it is strongly recommended that nasal swabs are sent for PCR in all cases where leprosy is suspected.

Situations where PCR may be useful include:

- paucibacillary cases with atypical clinical findings and non-diagnostic histological findings
- pure neuritic leprosy.

5.2.6 Susceptibility testing

Surveillance for antimicrobial resistance has been identified as an important component of the Global Leprosy Strategy 2021 – 2030. In 2008, the Global Leprosy Programme (GLP) established a surveillance network to collect data on antimicrobial resistance in *M.leprae*.

Global data from sentinel sites of 19 countries collected by the WHO between 2009 and 2015 reveals rates of rifampicin resistance of 5.1% in 1143 cases of relapsed infection (proxy for secondary resistance), and 2.0% in 789 new cases (proxy for primary resistance). The global rate of rifampicin resistance was 3.8%, with higher rates in Brazil (9%) and India (5%). The global rate of dapsone resistance was 5.3% with rates in Brazil and India again higher, at 12.7% and 6.4%, respectively.[19] These figures highlight the need to expand the surveillance network and implement susceptibility testing for all retreatment cases.

The inability to grow *M.leprae* in vitro, and the expense and time-consuming nature of the mouse footpad technique, has prompted a recommendation from the WHO for susceptibility testing to be performed using molecular techniques to detect the mutations known to be associated with rifampicin (*rpoB*), ofloxacin (*gyrA*) and dapsone (*folP1*)[20] resistance. Susceptibility testing has not been standardized, with varying techniques utilised. Amplicon-based sequencing is available at PathWest Laboratory Medicine.

Susceptibility testing should be requested for:

- all retreatment cases
    - with the exception of transferred cases who have documentation of uninterrupted treatment.
- all new multibacillary cases with a BI of >2+.

Required specimens include either two slit skin smears or one skin biopsy, with collection performed by an experienced clinician following discussion with the Infectious Diseases Specialist at the WA TB Control Program. Skin biopsy is the preferred specimen.

Method for sample collection:

Slit skin smear samples:

- Two samples to be obtained using method outlined in 5.2.2.
    - Either 2 from most prominent lesion with BI >2+, or one from most prominent lesion and one from the earlobe.
- Extra caution should be taken to prevent contamination.
- Following collection of tissue fluid from the incision site, the blade should be rinsed into a 1.8 mL centrifuge tube with screw cap pre-filled with 1mL of 70% ethanol (molecular biology grade absolute ethanol at 70% v/v + sterile deionized water from MilliQ or human injection quality 30% v/v).
- Transport to the MRL at QEII at room temperature.

Skin biopsy:

- New cases: 4mm punch biopsy of the most prominent lesion with BI >2+.
- Retreatment cases: 6mm elliptical biopsy of most prominent lesion is preferred for cases with BI <3+ to improve yield.
- Place the biopsy in a 1.8mL centrifuge sterile tube filled with 70% ethanol as described above.
- If the ethanol mixture is unavailable, place the biopsy in an empty 1.8mL sterile centrifuge tube with a screw cap.
- Transport to the MRL at QEII at room temperature.

WA TB Control will report the following annually to the WHO:

- Number of new MB cases tested, and the number and proportion of resistant cases (to one or more drug).
- Number of retreatment MB cases tested, and the number and proportion of resistant cases.
- The Infectious Diseases Specialist responsible for leprosy at WA TB Control will complete the forms for data collection with assistance from the Mycobacterial Reference Laboratory (Appendix 6).

5.2.7 Quality control

All PathWest laboratories are accredited by National Association of Testing Authorities and undergo regular audits. Proficiency is assured through participation with national (Royal College of Pathologists Australasia) and the Special Interest Group for Mycobacteria within the Australian Society for Microbiology. These programs cover all aspects of tertiary mycobacteriology. PathWest laboratories offering AFB microscopy also undertake quality

assurance to ensure competency. The MRL offers training and quality control materials as required.

5.2.8 Laboratory notification of results

New smear positive, PCR positive and results of susceptibility testing are communicated by the scientist to the requesting doctor or laboratory. Hard copy and electronic reports are managed via a laboratory information system. The MRL will notify all new smear positive and PCR positive results to the Medical Director, Western Australia TB Control Program by fax followed by a hard copy of the report.

5.3 Other diagnostic tools

5.3.1 Nerve conduction studies

Nerve conduction studies (NCS) are rarely indicated in the diagnosis of leprosy, and should be reserved for difficult cases, in consultation with the neurology department. NCS are invasive, uncomfortable and time consuming. Furthermore, the sensitivity of sensory testing with monofilaments in conjunction with VMT and nerve palpation for detecting nerve damage in leprosy is similar to that of NCS.[21]

5.3.2 Imaging

High resolution ultrasonography (HRUS):

HRUS may be indicated in the following situations:

- In cases where the diagnosis of leprosy is unclear
    - Used with colour doppler to demonstrate nerve enlargement and inflammation.
- Early detection of neuritis (with use of colour doppler).
- To localise nerve pathology to guide aspiration or biopsy.
- When nerve abscesses are suspected.

While some ultrasonographic features including extensive nerve enlargement, increased endoneural and epineural blood flow and thickening of the epineurium are suggestive of leprosy, HRUS cannot distinguish leprosy from other causes of nerve pathology,[22,23] Hence the findings must be used in conjunction with clinical findings and microbiological results.

5.4 Disease classification and assessment

5.4.1 Introduction

Leprosy is a chronic disease which presents with a wide spectrum of clinical features. Disease expression is determined by the patient's cell mediated immune response, with the potential for variation over time.

Classification systems have been designed to simplify categorisation for treatment purposes, as well as determining infectivity, risk of leprosy reaction and nerve damage. It is important to note that the systems are designed to be used as a guide, with some patients presenting with atypical features, and recognising that clinical features can change during the course of disease.

The most common classification systems currently in use are the WHO classification 1998,[24] the National Leprosy Eradication Programme (NLEP) of Government of India classification,[25] and the Ridley-Jopling classification.[26] The choice of classification system depends upon availability of resources and operational factors.

In WA, the Ridley-Jopling classification is used when skin biopsy is available, in view of greater utility in terms of predicting prognosis. In situations where only SSS are available, the NLEP classification is preferred due to inclusion of the number of nerves involved, providing greater sensitivity and allowing for incorporation of pure neuritic leprosy into the diagnostic spectrum.

The WHO classification is used in situations where neither SSS nor skin biopsy are available, and for reporting to the WHO. Treatment is guided by correlation of Ridley-Jopling and NLEP classification with WHO classification as outlined in Table 8.

5.4.2 Ridley-Jopling classification

The Ridley–Jopling classification system identifies five forms of leprosy on the basis of clinical, histological, bacteriological and immunological features:

- TT: Tuberculoid leprosy
- BT: Borderline Tuberculoid leprosy
- BB: Borderline Borderline leprosy
- BL: Borderline Lepromatous leprosy
- LL: Lepromatous leprosy

This classification represents a spectrum from localised disease with high cell mediated immunity (CMI), robust granulomatous response and few to no AFB in TT leprosy, through three immunologically unstable borderline forms, to the polar lepromatous (LL) form which is characterised by disseminated disease with abundant *M. leprae* and poor CMI. It is important to note that in a small proportion of cases clinical features do not correlate with the histopathological classification. This may represent immunological instability with a lag in clinical manifestations. Indeterminate and pure neural leprosy are not included in the Ridley-Jopling classification.

Indeterminate leprosy is believed to represent an early form of leprosy, which occurs in a proportion of patients following exposure to *M.leprae*. It is characterised by a hypopigmented patch with ill-defined edges and in most cases preservation of hair and nerve function. Diagnosis requires demonstration of typical perineural inflammation or more rarely, the presence of scanty bacilli. The majority of cases will self-heal without therapy, with a small proportion progressing to TT leprosy.

The features of pure neural leprosy are outlined in section 5.2.4.

5.4.3 WHO Classification

The WHO classification was initially proposed in 1982 to guide fixed duration MDT (FD-MDT). It was revised in 1998 to simplify diagnosis in the field and remove the need for slit skin smears. The primary clinical criterion for classification is the number of skin lesions, with paucibacillary disease having (1-5 lesions) and multibacillary disease (6 or more lesions).

5.4.4 NLEP Classification

The NLEP classification proposed in 2009 is based on the WHO classification, with the addition of assessment of the number of nerves involved. A MB case is defined as the presence of six or more skin lesions, involvement of more than one nerve (irrespective of the number of skin lesions) and skin smears positive at any site. As stated previously, inclusion of the number of nerves involved improves the sensitivity of the classification and ensures inclusion and accurate treatment facilitates of pure neural leprosy.

Table 8: Classification of Leprosy

<table>
<tr><th>Classification</th><th>Paucibacillary (PB)</th><th>Multibacillary (MB)</th></tr>
<tr><td>WHO</td><td>1-5 skin lesions</td><td>More than 6 skin lesions
Smear + at any site</td></tr>
<tr><td>NLEP</td><td>1-5 skin lesions
and / or
no nerve or only 1 nerve involved
Smear negative at all sites</td><td>More than 6 skin lesions <u>or</u> > 1 nerve involved
Smear + at any site</td></tr>
<tr><td rowspan="3">RIDLEY-JOPLING</td><td>TT
1-3 skin lesions: asymmetrical, anaesthetic
1-2 nerves involved with regional nerve function impairment (NFI)</td><td>BB
Many skin lesions: more symmetrical, hypoanaesthetic, inflamed
>1 nerve involved with more widespread NFI
BI 1-3+
High risk of reactions</td></tr>
<tr><td rowspan="2">Some BT
3-5 skin lesions: asymmetrical, anaesthetic / hypoanaesthetic
Few nerves involved
BI 0-1+</td><td>BL
Many skin lesions: symmetrical, hypoanaesthetic, some skin infiltration
Multiple nerves involved, enlargement less pronounced, NFI variable
BI 3-5+
High risk of reactions</td></tr>
<tr><td>LL
Widespread numerous skin lesions, often confluent with skin infiltration and nodularity
Multiple nerves, late enlargement, NFI variable, often glove and stocking neuropathy
BI 5-6+</td></tr>
</table>

*NB: If the skin smear is positive, irrespective of number of skin lesions or nerve involvement, the case is classified as MB leprosy. In situations where the classification is unclear, the patient should be treated as having MB leprosy.

5.4.5 Disability grading

Leprosy remains an important cause of disability worldwide, with prevention and management of disability being one of the components of the current WHO Global Leprosy Strategy. The rate of Grade 2 disability (G2D) in WA is 50%, well above accepted targets.

The proportion of G2D cases amongst newly diagnosed cases and the G2D rate in a population are markers for delayed leprosy detection. Factors influencing the rate of G2D include community awareness of the early signs of leprosy, access to leprosy services, skills of healthcare workers, and stigma.

Grading disability in leprosy is important for assessing program efficiency, as well as assessing changes in individual patients over time.

Three grading systems are currently in use: the WHO disability grade, the Eye Hand Foot (EHF) score and the Impairment Summary Form (ISF).

WHO disability grading:

The grading system currently in use was proposed by the WHO in 1997 (Figure 4). Grading should be performed at the time of diagnosis, primarily for reporting. Detailed operational definitions for the grading scale have been proposed to enhance reliability of data, facilitate comparison between programs or groups of patients, and assist with interpretation of the EHF score.[27] The WHO disability grading should be used primarily to assess program efficacy in terms of early case detection.

Figure 4: WHO disability grading 1998[28] with proposed operational and expanded grading.

Hands and Feet

Grade 0: no anaesthesia, no visible deformity or damage.

Grade 1: anaesthesia present, but no visible deformity or damage.

Includes scars of healed ulcers when sensation is impaired, muscle weakness without clawing or contracture.

Grade 2: visible deformity or damage present.

Includes ulcers, severe cracks, severe atrophy, contractures, digital absorption, clawing

Eyes

Grade 0: no eye problem due to leprosy, no evidence of visual loss.

Grade 1: eye problems due to leprosy, but vision not severely affected (VA 6/60 or better, can count fingers at 6m).

Includes absence of regular blink.

Grade 2: severe visual impairment (VA worse than 6/60, inability to count fingers at 6m, lagophthalmos, iridocyclitis, corneal opacities.

NB:

1. Facial deformity in association with lepromatous leprosy such as loss of eyebrows, enlarged earlobes and collapse of the nose are not included but should be recorded
2. Impairments which are not due to leprosy should not be included. Record separately and annotated with "non-leprosy"
3. The highest score for either the eye or hands / feet is reported

EHF impairment score:

The EHF impairment score is more complex than the WHO grading, providing more detailed information on the site and degree of impairment.

Individual impairment grades for eyes, hands and feet are added together to give the final score (Figure 5). The maximum score for each hand, foot and eye is two, with a maximum final score of 12. The EHF score provides an indication of extent of nerve impairment, with a score of 5 or more reflecting involvement of 3 or more nerves. While the EHF can provide some information on changes in impairment over time, it is not a highly sensitive or reliable indicator at an individual level, with the potential for deterioration in one area to be masked by improvement in another.[29,30] For this reason, it is recommended that the EHF, like the WHO grading system,[31] be used primarily to assess program effectiveness, rather than to assess change in impairment in individual patients.

Figure 5: WHO grading and EHF score[31]:

Site	R eye	L eye	R hand	L hand	R foot	L foot	Total
Grade	(0-2)	(0-2)	(0-2)	(0-2)	(0-2)	(0-2)	(_ /12)

Impairment Summary Form:

The ISF has a total of 24 variables including 3 for each eye, 5 for each hand and 4 for each foot. It should be performed at least every 6 months while on therapy, and after completion of therapy for patients at high risk of further impairment. ISF scoring provides more detail than that provided by the EHF and WHO grading, with greater sensitivity for assessing deterioration at an individual level.[29] It is useful for monitoring both secondary impairments, as well as detecting development or deterioration in primary nerve function.

The ISF is detailed and time consuming, and therefore most suitable for assessments at the referral centre level. An ISF Scoring table can be found in Appendix 7.

5.4.6 Case notification

When a new case of leprosy is confirmed, it is a requirement that a notification form is completed as soon as possible. It is important that cases not meeting the National Notifiable Diseases Surveillance definition, but fulfilling criteria required to meet the WHO leprosy case definition, are notified to the Director of the WA TB Control Program (for definitions, see Chapter 4).

In addition, the enhanced surveillance database should be completed by the treating physician and case manager. Assistance with completing the database and notification forms can be provided by WA TB Control Program staff.

5.4.7 Functional assessment

Leprosy is a chronic infectious disease which has the potential to significantly impact the patient's Health-related Quality of Life (HRQoL).[32,33,34]

Factors contributing to impaired quality of life include:

- stigma associated with disease
- physical consequences of the disease
- chronic pain
- lepra reactions and their consequences
- physical deformity and disability
- medication side effects.

Measurement of the impact of disease on a patient's overall wellbeing provides valuable information that can guide care planning and interventions to optimise patient outcomes.

HRQoL can be measured using a number of tools. The World Health Organisation Quality of Life Questionnaire (WHOQOL-BREF) has been validated for assessing quality of life in patients with leprosy. The scale has four domains; physical health, psychological health, social relationship and environmental health. The questionnaire is very comprehensive with a complex scoring system which may not be suited to the clinical setting. Initial screening with the Kessler Psychological Distress Scale (K6) is recommended for all patients at the time of diagnosis and annually while on treatment. For patients with acute neuritis, lepra reactions or nerve damage, the survey should be completed six-monthly during treatment and follow up (Appendix 8).

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Chapter 6 Medical treatment of leprosy

6.1 Introduction

Multidrug therapy for leprosy

In 1982, prompted by the emergence of dapsone resistance and in an attempt to improve compliance and efficacy, the WHO recommended the use of multi drug therapy (MDT) for the treatment of leprosy[1]. Since this time, the WHO has made a number of changes to both the classification of disease and the duration of therapy.

Significant controversy exists regarding duration of treatment and removal of the need for skin smears to determine classification. Uniform MDT (U-MDT), involving 6 months of treatment for all patients, regardless of classification, has not been widely adopted. Reasons for this include minimal long term follow up data and concerns regarding the risk of relapse, particularly in multibacillary patients with a high bacillary index (BI ≥4+).

Similarly, there are concerns that Accompanied MDT (A-MDT), where the patient is provided with the entire supply of MDT drugs at the time of diagnosis, with the supervision of a nominated person, may compromise adherence, resulting in relapses which may not be detected in periods of short term follow up.

Several studies where long term follow up has been performed have noted higher rates of relapse in cases with a high BI (≥4+) managed with shorter duration of MDT[3]. Review of the literature using the GRADE (Grading of Recommendations Assessment, Development and Evaluation) approach has revealed that evidence for clinical improvement and relapse rate with MDT for MB and paucibacillary (PB) leprosy is low (relapse rate MB leprosy) to very low (clinical improvement MB and PB, relapse rate PB leprosy)[4].

For these reasons, Western Australia guidelines recommend at least 24 months of therapy for patients with BI ≥4+.

WA guidelines align with the United States National Hansen's Disease Program (NHDP) guidelines[5] for treatment duration of patients with BI>4+ but differ with regards to dosing of rifampicin. WA has adopted the WHO recommendation for monthly rifampicin, rather than daily dosing as suggested by NHDP, based on results from a multicentre RCT demonstrating similar clinical improvement but increased adverse events with daily rifampicin[6]. Uncertainty still exists regarding optimal dosing of rifampicin, with some suggestion that patients with high bacillary index (>4+) may benefit from daily rifampicin in the first month of therapy.

The recently published WHO guidelines for diagnosis, treatment and prevention of leprosy[7] recommend using a three-drug regimen of rifampicin, dapsone and clofazimine for all patients, regardless of classification. The advantages of this approach include simplification of prescribing, reduction in impact of misclassification of MB leprosy as PB leprosy, and possible anti-inflammatory benefits for PB patients with neuritis, who otherwise would not receive clofazimine. In view of this, the WA guidelines have followed this recommendation.

6.2 Principles of drug therapy for leprosy

- Accurate classification of disease.
- Individual case management to optimise compliance and outcomes.
- Patient education
    - Symptoms and signs of lepra reactions
    - Adverse drug reactions
    - Importance of compliance.
- Careful clinical monitoring of patients to
    - detect adverse drug reactions.
    - ensure early detection and treatment of lepra reactions.

6.3 Pre-treatment investigations and documentation

- FBC, EUC, LFT, Calcium, Vitamin D, HBA1C.
- HIV, Hepatitis B & C serology.
- Screen for Glucose 6 phosphate deficiency (G6PD)
    - Can predispose to dapsone related haemolysis.
- Exclude co-existent latent or active TB: TST or Quantiferon (QIFN) TB-Gold Plus assay and Chest X-Ray.
- Strongyloides serology (if prednisolone required).
- Complete notification and inform WATBCP.
- Photography of skin lesions with informed consent.
- Documentation of clinical findings and laboratory results.

6.4 Treatment planning

- Assign case manager at WATBCP.
- If outside metropolitan Perth a local case worker is also required.
- Ophthalmology review for MB patients or if any concerns about eye abnormality (see 5.1.6).
- Podiatry, occupational therapy, physiotherapy referral if required.
- Weekly review & provision of medication by case manager /worker for the first month of therapy.
- Once compliance established, monthly clinical review by case manager and medical practitioner.
- Specialist review at least 3 monthly (for non-metropolitan patients this can be coordinated between WATBCP specialists and regional physicians.)
- In presence of lepra reaction, clinical review is required more frequently.

*NB: For male patients with ENL, screen for hypogonadism with serum total testosterone (ideally taken 8-10am), Luteinizing hormone (LH) and Follicle stimulating hormone (FSH).

6.5 Treatment regimens

Tables 9 and 10 outline treatment regimens recommended for leprosy in WA. Information on the drugs used to treat leprosy can be found in Appendix 10.

Table 9: WA first line regimen for leprosy treatment

	Regimen	Duration
Paucibacillary	Monthly supervised *	6 months
≤5 skin lesions	Rifampicin	
0-1 nerves involved	Clofazimine	
Skin smears negative at all sites.		
Compatible histopathology	Daily (or intermittent see Table 10)	
	Clofazimine	
	Dapsone	
Multibacillary	Monthly supervised *	12 months
6 or more skin lesions	Rifampicin	24 months ** for BI ≥4+
more than one nerve involved (irrespective of number of skin lesions)	Clofazimine	
Skin smears positive at any site.	Daily (or intermittent see Table 10)	
Compatible histopathology	Clofazimine	
	Dapsone	

*Monthly doses of rifampicin and clofazimine should be taken in the presence of the case manager or health care worker. This is referred to as being “Directly Observed Therapy” (DOT)
**For patients with BI ≥4+ specialist advice should be sought regarding use of daily rifampicin therapy for the first month of therapy.

Table 10: Medication doses for multi drug therapy

	Adult >15 years **WHO MB Adult blister pack**	10-14 years and >40kg **WHO MB Child blister pack**	<10 years or 20-40kgs*	<20kgs
Monthly				
Rifampicin	600mg	450mg	300mg	10mg/kg
Clofazimine	300mg	150mg	150mg	6mg/kg
Daily or intermittent dosing				
Clofazimine	50mg daily	50mg daily	50mg twice weekly	Not given
Dapsone	100mg daily	150mg second daily	25mg daily	2mg/kg

*MB child blister pack can be adapted for use: omit 150mg Rifampicin, give clofazimine twice weekly instead of alternate days, halve dapsone tablet

It is advisable that treatment of children is initiated following specialist consultation, and with ongoing close supervision to avoid dosing errors and adverse drug reaction.

6.5.1 Patient monitoring while receiving leprosy treatment

All patients receiving treatment for leprosy require close clinical monitoring to detect lepra reactions, assess response to therapy, identify adverse drug effects, and provide psychosocial support.

It is recommended that all patients are seen by a doctor at least monthly, with specialist review at least every three months. More frequent reviews are required for patients with lepra reactions and treatment complications.

Suggested monitoring while on treatment (see Clinical Template Appendix 9):

Clinical item	Frequency
Dispensing medication	Weekly for first month, then monthly
Clinical review including skin examination, VMT-ST	Monthly More frequently in presence of lepra reaction (usually 1-2 weekly depending on severity of reaction)
Blood tests: FBC, EUC, LFT	Monthly for first 3 months, then if normal 3 monthly Additional investigations required for patients receiving prednisolone (see 7.2.2.4)
Skin smears	For patients with positive smears at time of diagnosis, repeat smears 12 monthly until completion of therapy. Smears should be taken from previously positive sites. More frequent or further investigation may be required in cases where treatment failure is suspected.
Disability grading	WHO grading and EHF score at time of diagnosis and completion of treatment Impairment summary form (ISF) 6 monthly (see 5.4.5 and Appendix 7)

6.5.2 Treatment completion

Treatment completion is defined as:

> **PB leprosy**: 6 months of doses (or 6 x 4 weekly cycles) within 9 months.
>
> **MB leprosy**: 12 months of doses (or 12 x 4 weekly cycles) within 18 months or for BI ≥4+ 24 months of doses (or 24 x 4 weekly cycles) within 36 months.

Treatment completion is not necessarily synonymous with improvement in clinical signs and symptoms, or with microbiological cure. For this reason, while fixed duration therapy is recommended by the WHO, therapy often needs to be individualised where resources permit. In the absence of a true test of cure, the relapse rate is the main method of

assessing the efficacy of treatment. In multibacillary cases, SSS should be taken at previously positive sites at the completion of therapy to allow assessment of treatment response and facilitate ongoing monitoring to detect relapse.

In patients with high BI at the time of diagnosis (>4+) the BI will remain positive at completion of 24 months fixed duration therapy however the MI should reveal dead bacilli which stain irregularly and are reported as being fragmented or granular. The BI should fall by 0.75-1+ each year while on treatment and following completion of therapy (See 5.2.2).

6.5.3 Follow up after completion of therapy

The aims of reviewing patients following completion of therapy are:

To detect relapse (see 6.10.1):

- The suggested duration of follow-up for MB patients with high BI is based on clinical data suggesting that the average interval from completion of therapy to relapse ranges from 3 years to 26 years, with a mean of 10.5 to 14 years.[8,9,10.]
- Relapse rate is an important indicator of effectiveness of drug therapy and program quality.
- Estimations of relapse rate following completion of MDT vary widely due to variations in definitions, treatment regimens and durations, patient characteristics including smear positivity, and duration of follow up.
- The WHO estimates that relapse rates following 12-24 months MDT are extremely low, with average rates of 0.1% per year for PB leprosy and 0.06% per year for MB leprosy.
- The rate of relapse is higher when regimens without rifampicin (dapsone monotherapy, clofazimine, injectable thiambutosine (CIBA-1906 / DPT) are used to treat multibacillary leprosy. Rates of relapse are reported to range from 2.5%[11] to 8.6%[12] with one study reporting a rate of 25%[13], driven primarily by sulphone resistance. The majority of cases diagnosed in WA prior to 1986 did not receive rifampicin as part of their therapy.
- Patients with a high bacillary index at the time of diagnosis (4-6+) have higher rates of relapse compared with those with lower bacillary index, hence more intensive follow up is recommended in this group of patients.[14,15,16]
- Early detection of relapse is important to prevent transmission and avoid further complications of leprosy including nerve function injury.

To prevent disability and deformity

- Evidence suggests that the majority of initial nerve function injury (NFI) occurs within 2 years of diagnosis.[17]
- Any NFI at diagnosis is predictive of further impairment of nerve function[18]
- Disability progression and burden of deformity are significant problems following completion of therapy
    - In a prospective cohort study, 40% of patients showed disability progression in the 10 years after treatment completion, with the majority presenting after 5 years.[19]
- Suggested duration of follow up reflect these findings and aim to deliver a high standard of care.
- Early detection and treatment of lepra reactions is facilitated by close follow up following completion of MDT.

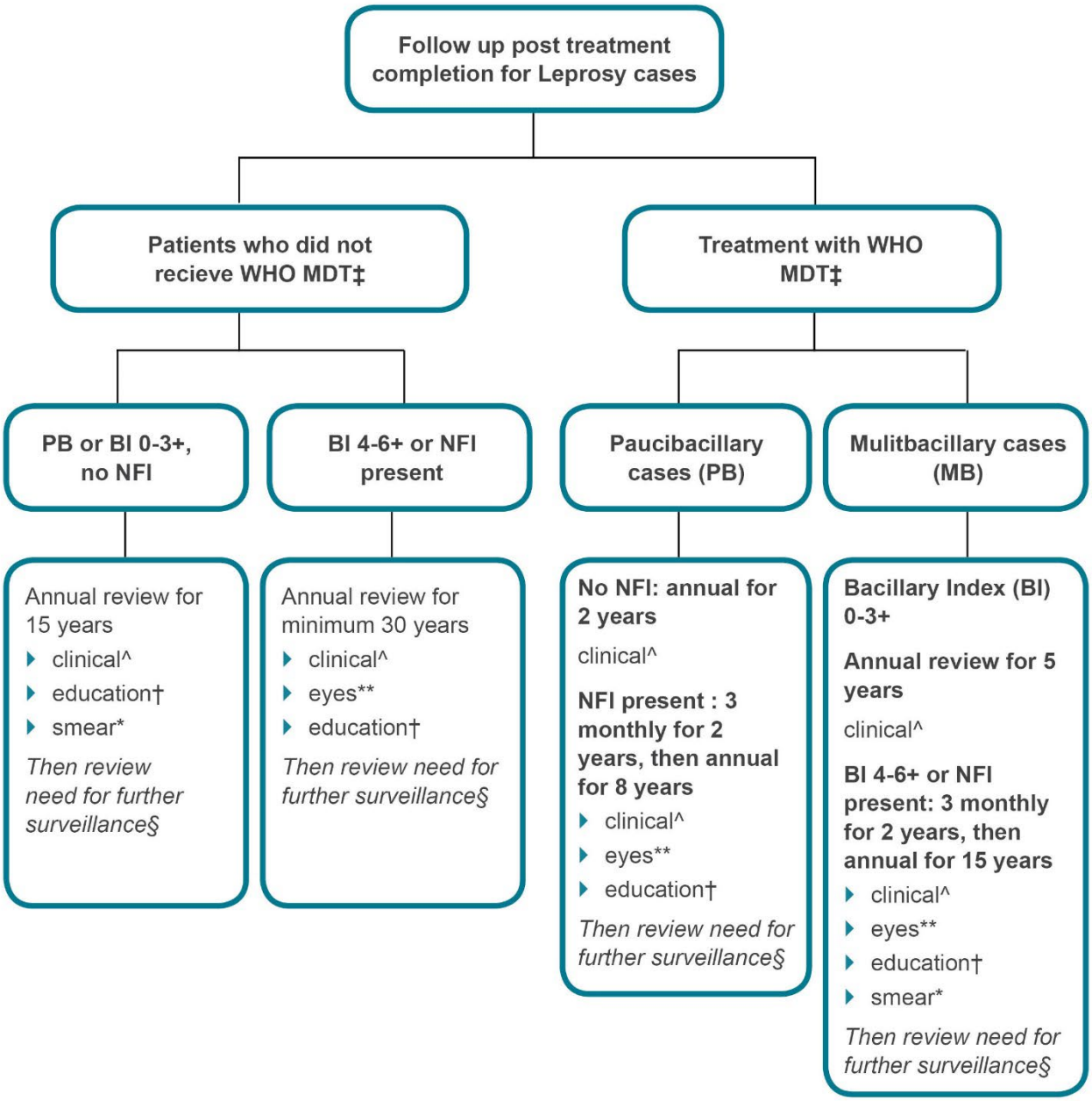
- Education for self-care of hands, feet and eyes is an essential component of prevention of disability.
- Coordination of the multidisciplinary team involved in the care of patients with nerve function injury is facilitated by the follow up suggested.

Case detection

- Education about signs and symptoms of leprosy.
- Enquiring about contacts and reviewing completeness of contact tracing.

Figure 6 outlines the suggested follow up of leprosy patients in WA once they have completed recommended therapy.

Figure 6. Follow up after completion of leprosy treatment



NFI= Nerve Function Injury

BI = Bacillary Index

‡ World Health Organisation Multi Drug Therapy where rifampicin + dapsone ± clofazimine given for at least 6 months (PB) or 12-24 months (MB) regardless of whether rifampicin given daily or monthly

^ Skin examination, nerve palpation, voluntary motor and sensory testing (Appendices 1-4)

* Skin smears should be taken from previously positive sites annually until negative. Thereafter, smears should be taken if clinically indicated (new skin lesions, suspicion of relapse; see 6.10.2)

** Visual acuity and ophthalmology review to detect potentially sight threatening ocular manifestations (see 5.1.6)

† Aimed at prevention of disability through self-care, early presentation, signs of relapse, opportunity to enquire about contacts needing review

§ Indications for ongoing surveillance include: worsening or severe NFI, need for ongoing supervision of self-care and coordination prevention of disability activities

6.6 Alternative regimens

Unable or refusal to take rifampicin (adult).

In case of allergy to rifampicin, seek advice from Immunology regarding safety of rechallenge with a reintroduction schedule, and feasibility of substituting with rifamycin analogues such as rifapentine.

Where rifampicin cannot be given (in case of allergy or drug resistance):

- 300mg clofazimine monthly AND
- 50mg clofazimine daily
    - *In addition to at least two second line agents:*
        - 400mg moxifloxacin / or ofloxacin daily
        - 100mg minocycline daily
        - 500mg clarithromycin daily.

To be given daily for 6 months, followed by clofazimine + one second line drug for an additional 18 months.

In cases where ofloxacin resistance is also present, a fluoroquinolone should not be used as part of second line treatment. The recommended regimen is clarithromycin, minocycline and clofazimine for 6 months, followed by clarithromycin or minocycline plus clofazimine for an additional 18 months (conditional recommendation, based on expert opinion).

Unable or refusal to take dapsone (adult).

Dapsone can be replaced by

- 400mg moxifloxacin daily.

Unable or refusal to take clofazimine (adult).

Clofazimine can be replaced by:

- moxifloxacin 400mg daily OR
- minocycline 100mg daily.

In cases where alternative or second line regimens include use of a fluoroquinolone, care should be taken to exclude active tuberculosis infection. For cases where second line therapy includes the use of fluoroquinolones, clarithromycin or minocycline, electrocardiographic monitoring should be performed due to the risk of QT-prolongation and associated cardiac arrhythmias.

6.7 Regimens for drug-resistant leprosy

In the situation where single or multiple drug resistance is identified, expert opinion from an Infectious Diseases Physician at the WATB Control Program should be sought.

Alternative regimens are outlined above in section 6.6.

6.8 Adverse drug effects

Dapsone

Dapsone Hypersensitivity Syndrome is an uncommon but serious reaction that appears to occur at a higher incidence amongst Aboriginal people. It occurs most commonly between the 2nd and 8th weeks of dapsone therapy and requires immediate cessation. Rechallenge is also contraindicated.

Adverse effect	Features	Treatment
Haemolytic anaemia	↓ Hb ↓ haptoglobins ↑ reticulocytes ↑ LDH ↑ unconjugated bilirubin	Trial 5mg folic acid If severe and not improving, cease dapsone
Dapsone Syndrome / Dapsone hypersensitivity syndrome	Criteria proposed by Richardus and Smith[20]: 1. Presence of at least 2 of the following: fever, skin eruption, lymphadenopathy, liver abnormalities 2. Appearance of adverse events between the 2nd and 8th week after starting dapsone, which disappear on cessation of dapsone 3. Symptoms not attributed to any simultaneously used drugs or to lepra reaction 4. Symptoms not attributed to any other diseases	**Immediate cessation of dapsone: high mortality rate** Prednisolone commencing at 1mg/kg daily, then weaning regimen for 4-6 weeks Supportive therapy

Rifampicin

Serious adverse effects are infrequently encountered with monthly administration, however respiratory distress with dyspnoea, fever, cough, and flu like symptoms, gastrointestinal upset, thrombocytopaenia, shock and acute renal failure can occur. Consultation with an Immunology service is strongly recommended.

Clofazimine

Adverse effect	Features	Treatment
Skin discoloration	Brownish-red discoloration of skin	No treatment required, drug cessation if stigmatising Usually takes 6-12 months to resolve following cessation
Ichthyosis	Violaceous brown lesions on extensor surfaces	10% urea cream and dermeze - apply topically to affected areas on alternate days
Enteropathy (occurs with doses of >200mg for > 3 months	Abdominal pain, subacute bowel obstruction	Reduce dose of clofazimine

6.9 Special considerations

6.9.1 Pregnancy

It is widely accepted that pregnancy can have an adverse effect on women with leprosy, however there is limited good quality data to support this, and there is little data from the MDT era.

The changes in cell mediated and humoral immunity that occur during pregnancy and in the post-partum period may have the following effects on the course of leprosy for women either on therapy or following completion of therapy:

- Increased risk of ENL in LL / BL patients.
- Increased risk of progression of infection towards the lepromatous end of the spectrum (new skin lesions, increase in BI, progressive NFI).
- Increased risk of neuritis.
- Increased risk of relapse.
- Increased risk of Type 1 reaction (post-partum).

In addition, there appears to be an increased risk of presenting with leprosy during pregnancy or post-partum period because of the altered immunological state. It is important to note that pregnancy is safe for women who have completed effective MDT, however those affected by lepra reaction during treatment should be monitored with voluntary motor and sensory testing as well as nerve and skin examination three monthly during the antenatal period and at one month following delivery (see Appendix 1-4).

The WA BCG policy recommends that babies of parents with leprosy or a family history of leprosy be offered BCG. This is best facilitated through ensuring documentation in the antenatal record for current and future pregnancies.

Drug therapy of Leprosy during pregnancy

Standard MDT should be continued throughout pregnancy and breast-feeding and is safe to be commenced if a new diagnosis of leprosy is made.

Lepra reactions should be treated as per standard recommendations, however thalidomide is absolutely contraindicated.

Effects of MDT on pregnancy:

- Dapsone: anecdotal reports of haemolytic anaemia and hyperbilirubinemia in breast fed infants.
- Rifampicin: association with haemorrhagic disease of the newborn, prophylactic vitamin K for newborn.
- Clofazimine: temporary discoloration of foetal skin.

Leprosy drugs contraindicated in pregnancy:

- Quinolones (ofloxacin and moxifloxacin)
- Minocycline
- Clarithromycin
- Thalidomide.

6.9.2 Co-existent active tuberculosis

Patients with leprosy and active tuberculosis (TB) require full treatment for both conditions. Screening for tuberculosis should be done in all patients with newly diagnosed TB and vice versa.

6.9.3 Co-existent latent tuberculosis

All patients with leprosy should be screened for tuberculosis with a tuberculin skin test (TST) or Quantiferon Gold and chest X-Ray. If there is evidence of latent tuberculosis infection (LTBI) and active disease has been excluded, latent infection can be opportunistically treated with daily rifampicin for four months, in addition to standard MDT. Alternatively, leprosy and LTBI can be treated sequentially.

6.9.4 Co-existent Human Immunodeficiency Virus infection

Despite the host immune response playing an important role in all aspects of leprosy infection, there is little evidence to suggest that Human Immunodeficiency Virus (HIV) has a significant impact on the course of disease. Similarly, it does not appear that leprosy has a significant impact on the course of HIV infection, unlike tuberculosis[21].

HIV co-infection may be related to an increase in the frequency of lepra reactions, particularly in unstable borderline states, however the evidence from a small number of studies is conflicting[22,23]. A number of case reports have described an association between leprosy and Immune Reconstitution Inflammatory Syndrome occurring following initiation of highly active antiretroviral therapy (HAART), with two main forms identified[24]:

1. Unmasking of previously untreated and unrecognised infection with *M.leprae.*

2. Type 1 reaction occurring in a patient with pre-existing leprosy.

In light of the lack of evidence to support significant interaction between *M.leprae* and HIV, the following is recommended:

- Standard MDT should be given with no change in duration of therapy.
- Patients should be closely monitored for development of lepra reactions (particularly type 1 reaction) following institution of HAART.
- Reactional states should be treated without modification from routine guidelines, however prolonged courses of prednisolone may be required, even after completion of MDT.
- Leprosy should be suspected in a patient from a leprosy endemic area presenting with skin lesions or neurological symptoms shortly after HAART initiation.

6.9.5 Treatment interruption and default

Poor adherence to leprosy therapy remains a significant problem in many endemic regions, with implications for both the individual, as well as for the success of disease control.

Contributing factors:

- Socioeconomic disadvantage
- Cultural
- Geographic isolation
- Psychosocial
- Drug related
- Disease related
- Health service related
- Stigma

WHO definitions for treatment default:[25]

- Failure to complete 6 months of prescribed therapy within 9 months
- Failure to complete 12 months of prescribed therapy within 18 months.

To incorporate the 24 month treatment duration recommended by WA TB Control Program for patients with BI ≥4+, this definition also includes:

- Failure to complete 24 months of prescribed therapy within 36 months

Patients who fail to attend for a monthly dose of directly observed therapy (DOT) should be located immediately to facilitate early intervention and address factors that may be contributing to incomplete adherence.

The management of treatment defaulters is often extremely challenging and resource intensive. The long duration of therapy for leprosy is a significant barrier for a number of patients, with little evidence to support the use of shorter or monthly regimens.

Suggestions for managing poor adherence:

- Patients who have poor adherence related to a single drug in the MDT regimen can be changed to an alternative regimen as outlined in 6.6.
- Monthly supervised rifampicin, ofloxacin and minocycline (ROM) can be considered for persistent defaulters with PB (minimum 12 months) or in rare situations with MB leprosy (minimum 24 months), however every attempt should be made to re-establish the patient on daily therapy.
- Patients receiving ROM require careful long term follow up to detect relapse.[26,27]

Methods to assess and improve adherence are discussed further in Chapter 8.

6.10 Retreatment

6.10.1 Definitions

Retreatment case: a patient diagnosed with leprosy who has already received treatment for the disease in the past.

Retreatment cases are further classified into the following groups[28]:

(a) **Retreatment after loss to follow-up**: a patient diagnosed with leprosy who has abandoned treatment before its completion and returns to the health facility to complete treatment.

(b) **Relapse**: a patient who has completed full treatment course for leprosy in the past and who returns with signs and symptoms of the disease that are not deemed due to a reaction according to the clinician.

(c) **Transferred in**: a patient who has started treatment in one facility and reports to another facility to continue treatment.

(d) **Other retreatments**: any leprosy case that does not fall in any of the above categories and requires treatment.

6.10.2 Differentiating relapse from reactional state

Differentiating relapse from reactional state is often challenging. Features that can help to ensure the correct diagnosis is made are shown in Table 11.

Factors predisposing an individual to relapse:

- Inadequate or irregular therapy.
- Dapsone monotherapy (pre MDT era).
- BI >4 + (higher risk of having persisting organisms: “persisters”).

Table 11: Features of relapse and reactional state

Clinical feature	ENL	Relapse
Relationship to treatment	Before, during, or after treatment	After completion of therapy
Onset	Sudden	Gradual
Constitutional symptoms	Often present	Absent
Clinical signs	Tender, warm, erythematous superficial nodules, appear in crops	Full thickness lesions, not warm or tender
SSS	Fragmented AFB	BI>2+, solid staining AFB
Clinical feature	**Type 1 reaction**	**Relapse**
Relationship to treatment	Before, during, or after treatment	Usually > 1 year after completion of treatment
Clinical signs	↑ erythema, swelling, tenderness of skin lesions	Appearance of new skin lesions, non-tender, often satellite lesions
	Ulceration may occur in severe reactions	Ulceration does not occur
	Oedema of hands and feet	Rarely seen
	Few new lesions may appear	Many new lesions
	Acute painful neuritis which may be associated with motor or sensory loss	Neuritis may be present, but generally not acute
SSS	BI continues to fall	BI may increase
	Bacilli granulated in reactions	
Response to prednisolone	Prompt improvement in skin lesions and neuritis	No or minimal response

6.10.3 Management of retreatment cases

- Retreatment after loss to follow up and transferred-in cases should be re-evaluated and treated according to guidelines.
- Drug resistance testing should be requested for all MB retreatment cases with BI>2+ (excluding transferred-in cases unless thought to be at risk due to irregular treatment)
    - Procedure described in Chapter 5.2.6.
- Where the diagnosis of relapse is unclear and type 1 reaction suspected, a trial of prednisolone is warranted
    - Improvement in clinical signs within 2 weeks of commencing prednisolone is suggestive of lepra reaction.
- If drug resistance is detected, specialist advice should be obtained.
- Drug sensitive relapse and relapse where sensitivity testing is not possible (BI <2) or available should be reclassified and treated according to guidelines.
- Patients who have previously received either dapsone or clofazimine monotherapy should be retreated with MDT as per WA guidelines.

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Chapter 7 Diagnosis and management of neuritis and reactional states

7.1 Diagnosis of neuritis and reactional states

7.1.1 Introduction

Leprosy or lepra reactions occur as a result of immune mediated inflammation and can involve the skin, nerves, eyes, and body organs.

Lepra reactions and acute neuritis can occur:

- before diagnosis or at the time of diagnosis (prompting presentation).
- during treatment for leprosy.
- after completion of treatment.

Reactions and neuritis are a major cause of nerve damage and subsequent disability in leprosy.

Principles of management:

- Early diagnosis.
- Prompt treatment.
- Referral for specialist advice.
- Close clinical follow up.

Urgent referral for assessment is required for a patient presenting with any of the following:

- Red, painful nodules with or without ulceration.
- Pain or tenderness in one or more nerves, with or without impairment of function.
- Any new nerve function impairment.
- Red, swollen skin patch on the face or overlying a major nerve trunk
- High fever.
- Marked oedema of hands, feet or face.
- Pain and / or redness of the eye(s) with or without impaired vision.
- Painful swelling of joints or testes with fever.
- Mild reactions that have not responded to two weeks of anti-inflammatory treatment.

7.1.2 Neuritis

Neuritis is the inflammation of nerves. In leprosy it can manifest in the following ways:

1. Subacute, low-grade neuritis associated with disease.
2. Acute, more severe neuritis associated with reactions.
3. Silent neuritis (nerve function impairment without symptoms).
4. Neuropathic pain.

Clinical features of neuritis:

- Nerve thickening.
- Nerve tenderness.
- Nerve pain
    - includes paraesthesia, dysaesthesia, hyperaesthesia, allodynia.
- Nerve function impairment (NFI)
    - sensory
    - motor
    - autonomic.
- Secondary complications of NFI
    - deformity
    - trauma.

Diagnosis of neuritis:

- Sensory impairment detected using graded monofilament testing (Appendix 2)
    - Monofilament threshold sensation increased by 1 or more levels at any one site.
- Motor impairment detected with voluntary muscle testing (VMT) using modified Medical Research Council (MRC) scale (Appendix 2)
    - Motor impairment present if VMT score for any muscle or group of muscles ≤ 4 on the modified MRC scale.
    - Reduction in MRC score for any muscle group.
- Eye signs
    - Reduced corneal sensation.
    - Weakness of orbicularis oris (lid closure).

7.1.3 Type 1 Reaction

7.1.3.1 Clinical features

Type 1 (reversal or upgrading) reactions (T1R) are Th1 mediated delayed hypersensitivity responses to *M.leprae* antigens, which cause a sudden increase in cell mediated immunity and a shift towards the polar tuberculoid end of the Ridley-Jopling leprosy spectrum.

Type 1 reactions occur primarily in borderline leprosy (BT, BB and BL), affecting approximately 30% of cases[1]. Inflammation involves the skin, nerves (neuritis), or in some cases both. Neuritis causes loss of sensory and muscle function, generally associated with

nerve pain, but in some cases without pain or nerve tenderness (silent neuritis). Skin inflammation can be disfiguring and painful but is less serious than neuritis.

Type 1 reactions occur most commonly after starting treatment but can occur at any time in the course of infection.

Risk factors for Type 1 Reaction

- Borderline group
- Previous lepra reaction
- Pregnancy and the post-partum period
- Starting treatment with MDT
- Extensive disease (≥ 3 body areas involved)
- Skin lesions overlying nerve trunks
    - Facial lesions are high risk for ocular NFI.

Features of Type 1 Reaction

Skin (changes in existing and new leprosy lesions)

- Pain
- Swelling
- Redness
- Warmth
- Necrosis and ulceration in severe cases.

Nerves

- New sensory loss
- New muscle weakness
- Nerve pain or tenderness.

General

- Oedema of hands, feet or face.

Type 1 reactions are usually diagnosed on the basis of clinical findings. There are no laboratory tests useful for the diagnosis of type 1 reactions in clinical practice. The C-Reactive Protein (CRP) may be elevated but is a non-specific marker. Skin biopsies can be useful if the diagnosis is uncertain, with skin specimens typically showing lymphocytic infiltration, extracellular oedema in and around epithelioid granulomas, Schwann cell destruction and nerve fibre ischemia.

Table 12: Criteria for the clinical diagnosis of type 1 reaction [2]

One major or at least two minor criteria (without signs of ENL*) required for the diagnosis of T1R

Major

1. Pre-existing and/or new skin lesions become inflamed, red and swollen.

Minor

1. One or more nerves become tender and may be swollen
2. Crops of new, painless lesions appear
3. Sudden oedema of face and extremities
4. Recent loss of sensation in hands and feet, or signs of recent nerve damage (loss of sweating, sensation, muscle strength) in an area supplied by a particular nerve.

* ENL = Erythema Nodosum Leprosum

7.1.3.2 Severity Index

A severity score for type 1 reactions was developed and validated in 2008.[3] While detailed and requiring skills in the assessment of VMT and use of monofilament testing, the scale is useful in the referral centre setting, and for research purposes. The severity scale is designed for assessment of NFI present for less than 6 months (see Appendix 11).

Table 13: Grading scale for severity of type 1 reaction

Mild

- Small number of inflamed skin lesions (excluding facial lesions).
- No nerve pain or loss of nerve function.

Severe

- Nerve pain or sensory change.
- Loss of nerve function.
- Fever.
- Peripheral or facial oedema.
- Inflammation of skin lesion on the face.
- Ulceration or necrosis of skin lesions.

7.1.4 Erythema Nodosum Leprosum

7.1.4.1 Clinical features

ENL (Type 2 reaction) is a systemic inflammatory condition, which occurs in up to 50% of patients with lepromatous leprosy, and 5-10% of patients with Borderline Lepromatous Leprosy (BL).[4,5]

ENL can occur prior to, during, or following the completion of curative therapy for leprosy. Patients may experience a single episode of ENL or have recurrent episodes occurring over several years, with some developing chronic ENL.[6]

Classification of ENL:

Acute: single episode lasting less than 24 weeks.

Recurrent: occurrence of a second or subsequent episode of ENL 28 days or more after stopping treatment for initial episode.

Chronic: episode occurring for 24 weeks or more during which a patient has required ENL treatment either continuously or where any treatment free period has been 27 days or less.

While the exact cause of ENL is unclear, it is associated with a complex array of immune activation resulting in inflammation at multiple body sites. Immune complex formation and deposition are thought to play a role, with IgG, IgM, complement and mycobacterial antigens identified in ENL skin lesions. Pro-inflammatory cytokines including tumour necrosis factor α, interferon gamma, interleukin-6, interleukin-17 and interleukin-1 may also be important in the pathogenesis of ENL.[7]

The main risk factors for ENL are:[8]

- lepromatous leprosy with skin infiltration
- bacterial index >4+
- age less than 40 years.

The diagnosis of ENL is based primarily on clinical findings. Skin biopsies may be useful in cases where the diagnosis is unclear.

Clinical features of ENL:

- Sudden onset
- Crops of tender subcutaneous nodules
- Pre-existing leprosy skin lesions usually unchanged
- Fever
- Peripheral oedema
- Lymphadenopathy
- Neuritis (generally not as severe as in type 1 reactions)
- Pain, often bony in nature
- Joint swelling
- Dactylitis (swelling of fingers or toes)
- Eye inflammation (iridocyclitis)

The clinical presentation can often mimic an acute infection, with high fever, leucocytosis and elevated inflammatory markers. Renal dysfunction and liver dysfunction occur in some cases, generally indicating a more severe reaction.

ENL is often a severe illness that has a significant impact on the patient and their families. It is associated with increased mortality,[9] economic hardship[10] and a negative impact on health-related quality of life.[11]

7.1.4.2 Severity Index

Assessment of ENL severity is useful to guide appropriate treatment, predict prognosis and monitor progress.

A 10-item severity score for ENL has recently been validated.[12] In clinical practice, the score is helpful to determine the severity of the reaction, monitor response to therapy, and in the research setting, to compare the effectiveness of various treatments. The severity scale and user guide are included in Appendix 12.

When using the ENL ENLIST Severity Scale, a score of 8 or less indicates mild ENL, and when comparing scores, the minimal important difference is 5.

In situations where it is not practical to use the severity scale, a general classification into mild and severe ENL can be made according to the features outlined in Table 14.

Table 14: Features of mild and severe ENL

Clinical feature	Mild ENL	Severe ENL
Skin lesions	Few, limited in distribution	Multiple, red, painful nodules, widespread
Ulceration / necrosis of skin lesions	Absent	May be present
Fever, arthralgia	Absent	Present
Neuritis	Absent	Present
Peripheral oedema	Mild	Often marked, including face
Eye involvement	Absent	May be present
Lymphadenopathy	Absent	Often present
Systemic inflammation (nephritis, epididymo-orchitis)	Absent	May be present

7.1.5 Lucio's Phenomenon

Lucio's Phenomenon is an uncommon immunological reaction that occurs in diffuse non-nodular form of lepromatous leprosy, called Lucio's Leprosy. This form of leprosy has been associated with *Mycobacterium lepromatosis* and is only seen in certain populations, including Mexicans and Brazilians. It primarily occurs in patients who have not received treatment.

Features include:

- appearance of irregular painful plaques which become haemorrhagic and subsequently evolve into necrotic ulcers.
- absence of fever (unlike severe ENL).

ENL and Lucio's phenomenon are sometimes difficult to differentiate clinically and have been reported to occur simultaneously.[13]

7.2 Treatment of neuritis and lepra reactions

7.2.1 Introduction

Early identification and prompt treatment of reactions is one of the most important aspects of leprosy management.

Lepra reactions can result in permanent deformity and disability, including blindness.

If a lepra reaction or new NFI is suspected, urgent referral should be made through the WA TB Control Program for specialist review, and advice should be sought regarding immediate management.

Early diagnosis can be facilitated by:

- educating patients, their families and health care workers about the early signs and symptoms of reactions, and the need for urgent medical review.
- monthly clinical review, including neurological examination, of patients while on therapy, and following completion of therapy for those at higher risk of lepra reaction.
- ensuring systems designed to maintain good communication between regional / remote and referral centres.

Principles of management of lepra reactions:

- early diagnosis and prompt treatment
- multi drug therapy (MDT) should be continued or initiated (in those who first present with a T1R)
- anti-inflammatory therapy should be commenced at a dose to reduce inflammation rapidly, then gradually tapered down
- adequate analgesia
- physical therapy including rest and splinting where required
- close monitoring of neurological function.

7.2.2 Type 1 Reaction and acute neuritis

7.2.2.1 Treatment of mild type 1 reaction

Mild T1R is characterized by erythema, pain and tenderness in a few of the existing skin lesions. Nerve symptoms are absent.

- Analgesia (aspirin or paracetamol) for 2-3 weeks.
- Fortnightly review until resolved.
- Escalation of treatment if reaction becomes severe or symptoms / signs of nerve involvement appear.

7.2.2.2 Treatment of severe T1R and acute neuritis (<6 months duration NFI)

Severe T1R is characterized by one or more of the following:

- inflammation of multiple existing skin lesions and appearance of new lesions
- nerve pain, tenderness, paraesthesia, loss of nerve function
- fever, arthralgia
- ulceration of skin lesions
- oedema of hands and/or feet.

Patients may present with symptoms of type 1 reaction, acute neuritis or recent (<6 months old) NFI at the time of diagnosis. These patients should be commenced on MDT and treated according to the guideline below.

Despite a paucity of high quality evidence to guide treatment for type 1 reaction and acute neuritis, it is generally accepted that prednisolone should be the cornerstone of therapy[14]. Prednisolone reduces inflammation of skin and nerves and helps to improve pain and nerve function.

Optimal dosing and duration have not yet been established, with guidelines varying between institutions. An individualised approach based on careful and frequent assessment of neurological function using VMT-ST is ideal.

The recommendation is for patients with severe type 1 reaction to be reviewed fortnightly until symptoms of acute neuritis have improved. VMT-ST should be performed and recorded at each visit.

7.2.2.3 Assessment, therapy and monitoring of patients prescribed long term prednisolone

Baseline	• Weight, height, BMI • Blood pressure • TST/QFN and CXR to exclude LTBI or active TB • Strongyloides serology and stool specimen • Treat active infection (osteomyelitis, cellulitis, scabies) • Random blood glucose level • HIV and Hepatitis B serology • Melioid serology if clinically indicated • Lipid profile • FBC, Calcium, Vitamin D • BMD assessment • Serum testosterone, LH and FSH in men with LL and clinical suspicion of hypogonadism (recurrent ENL) • Visual acuity, obtain history for personal or family history of open angle glaucoma • Commence H2 blocker or proton pump inhibitor for gastric protection • Osteoporosis prevention with calcium and vitamin D supplementation if required
Subsequent monitoring	• Measure BP and weight at each visit • Assess lipids one month after initiation of prednisolone then every 6-12 months • Measure blood glucose level 2 weeks after commencing prednisolone, then 3-6 monthly for first year and annually thereafter. Refer to Endocrinology if abnormal blood glucose levels. • Ophthalmology referral for those with symptoms of cataract, diabetes mellitus, personal or family history of glaucoma, • BMD one year post initiation of prednisolone, then frequency as guided by results. Consider referral to Endocrinology if BMD is decreasing

BMI = Body mass index; TST = tuberculin skin test; QFN = Quantiferon test; CXR = Chest X-Ray; LTBI = latent tuberculosis infection; TB = tuberculosis; HIV = Human immunodeficiency virus; FBC = full blood count; BMD = Bone mineral density; LH = luteinising hormone; FSH = follicle stimulating hormone; LL = lepromatous leprosy; ENL = erythema nodosum leprosum; BP = blood pressure

7.2.2.4 Prednisolone regimen

Table 15: Suggested regimen for treatment of severe type 1 reaction in a patient weighing 50kg

Week number	Prednisolone dose (daily)	Alternate regimen to reduce pill burden
1-2	50mg	
3	45mg	1.5 x 25mg tablet + 1 x 5mg tablet = 42.5mg
4	40mg	1.5 x 25mg tablet = 37.5mg
5	35mg	1.5 x 25 mg tablet = 37.5mg
6	30mg	
7-8	25mg	
9-14	20mg	
15-16	15mg	
17-18	10mg	
19-20	5mg	
21-22	2.5mg	

When utilising this suggested regimen, it is important to note the following:

- Patients must be assessed prior to commencing prednisolone and monitored closely for adverse events.
- Prednisolone should be commenced at 1mg/kg. If symptoms or NFI fail to improve after 2 weeks the dose should be increased.
- For less severe reactions, a starting dose of 40mg may be adequate.
- Once symptoms are controlled and NFI improves, begin reducing the dose of prednisolone by approximately 5mg every 1 – 2 weeks until 20mg is reached.
- If symptoms recur or NFI worsens, the dose should be increased to the level at which symptoms were controlled, and maintained at this level for a minimum of 2 weeks before reattempting dose reduction.
- Once the dose of 20mg is reached, this dose should be continued for at least 4 weeks or until symptoms have resolved and NFI is stable or optimally improved.
- Further tapering should then occur with dose reduction of 5mg every 2 weeks.
- A proportion of patients may not respond to prednisolone therapy.
- Improvement in nerve function with prednisolone is more likely to occur when NFI has been present for < 6 months.[15]
- Evidence from a small number of randomised studies suggests that a 5 month course of prednisolone is as effective as an 8 month course and associated with better outcomes compared with a 3 months course[16]. Despite this, some patients will require prolonged courses of therapy, particularly patients with BL or Borderline Borderline (BB) leprosy.

- While there does not appear to be a benefit of giving higher doses of prednisolone[17], in some circumstances, higher doses may be required to control inflammation, particularly in patients with a higher body weight.
- Recurrence of type 1 reaction or acute neuritis following completion of prednisolone treatment should be retreated as per the suggested guideline.
- Supplementary doses of prednisolone or steroid may be required in the event of illness or surgery, either during or for up to 12 months after completion of a prolonged course of prednisolone. The patient should be made aware of this and an alert placed on their medical record.
- Live vaccines should be avoided whilst on high dose prednisolone (40mg / day) and for 3 months after cessation.
- Prednisolone should be given in the morning to minimise suppression of the adrenal axis.

7.2.2.5 Alternative agents

A proportion of patients, particularly those with severe Type 1 reaction or BL leprosy, will fail to respond to prednisolone, or require prolonged periods at high doses. A number of studies have been conducted to examine the role of alternative immunosuppressive agents including azathioprine[18], cyclosporine A[19]and methotrexate[20] in the treatment of type 1 reaction.

While there is no evidence to support the first line use of these agents, they may be considered in certain cases where the patient is failing to respond to prednisolone, or as a steroid sparing agent in patients with significant adverse effects. Specialist advice should be sought from the ACC in these circumstances.

7.2.2.6 Non-pharmacological therapy

In addition to prednisolone, patients with severe type 1 reaction may require analgesia and splinting of the affected limb(s).

7.2.2.7 Surgical therapy

Surgical therapy for Type 1 reactions includes decompression of affected nerves by incision of the epineurium and / or opening of the fibro-osseous tunnel through which the affected nerve travels. While the available evidence is limited and of poor quality, there does not appear to be any enhanced benefit of surgery over prednisolone for patients with Type 1 reaction[21].

Surgical therapy should therefore be reserved for patients with nerve abscesses, or in very select and rare cases where the patient suffers intractable severe pain despite maximal immunosuppressive therapy. Such cases should be discussed with the specialists at WA TB Control Program prior to surgical referral.

7.2.3 Treatment of Erythema Nodosum Leprosum

Treatment for ENL aims to resolve skin lesions, relieve pain, as well as reduce and prevent complications by reducing systemic and neural inflammation.

Corticosteroids, clofazimine, and thalidomide, either alone or in combination, are the drugs most commonly used to treat ENL. Recent data from small case series and a single centre

pilot study, as well as local experience support the use of apremilast as an alternative to thalidomide in both chronic and recurrent ENL[22, 23].

Due to the tendency for ENL to become recurrent or chronic, treatment is often prolonged, requiring many months to years of therapy. It is preferable to initiate steroid sparing agents early to induce faster remission, avoid the side effects of prolonged steroid therapy and reduce chance of recurrence. It is important to note that at present there is not a therapeutic regimen demonstrated to be reliably effective, and extended use of currently recommended agents is often complicated by serious side-effects.

There is limited high quality clinical data to guide drug choice, dosing and duration, with recommendations based on a small number of randomised studies and expert opinion.

Two open-label randomised studies comparing treatment options for ENL provide evidence to support the use of thalidomide for both acute and chronic/recurrent forms of ENL. An open-label randomised study performed in India showed that significantly greater clinical improvement was achieved in patients with acute ENL with a 20-week reducing schedule of thalidomide compared with a 20-week reducing schedule of prednisolone, and in patients with chronic / recurrent ENL, with a combination of prednisolone and thalidomide compared with a combination of prednisolone and clofazimine. In addition, thalidomide appears to act as a steroid sparing agent, with reduced total prednisolone requirements[24].

An earlier open-label randomised study, also performed in India, compared the efficacy of thalidomide with prednisolone for moderate to severe ENL[25]. Results of the study showed that clinical response was significantly faster with thalidomide, with cutaneous lesions resolving in a mean of 5.54 days compared with 13.25 days in the prednisolone group, and constitutional symptoms resolving in a mean of 2.04 days vs 4.37 days. The period of remission was significantly longer, and number of relapses significantly lower in the thalidomide group. Importantly, adverse events were more common in the prednisolone group, with somnolence being the most common side effect of thalidomide.

There is a need for high quality randomised studies using validated severity scoring to determine optimal treatment for ENL, and to assess in more detail adverse effects associated with thalidomide. Research aimed at identifying other agents for ENL is essential, to provide treatment options for patients in regions where thalidomide is unaffordable, intolerable, unavailable or highly restricted.

Suspicion of ENL should prompt immediate referral for specialist review.

Features of ENL impacting on therapy:

- Neuritis and NFI occur less commonly in ENL compared with type 1 reaction.
- ENL has a greater tendency to recur or become chronic compared with type 1 reaction, placing the patient at risk of steroid dependency and high cumulative steroid doses.

7.2.3.1 Treatment of mild ENL, not associated with neuritis or NFI:

- Bed rest.
- Aspirin
- Prednisolone should be commenced if there is no improvement within two weeks.

7.2.3.2 Treatment of moderate - severe acute ENL associated with neuritis or NFI (<6 months):

- If acute neuritis or NFI of < 6 months duration present, commence prednisolone at 1mg/kg and reduce as per recommendation outlined for type 1 and acute neuritis outlined above.
- If no symptoms of neuritis, commence prednisolone at 1mg/kg and reduce by 5-10mg every 1-2 weeks
    - Shorter courses of prednisolone are preferred for ENL to prolonged courses and steroid dependency in light of the tendency for the condition to recur or become chronic.
- If symptoms of ENL (excluding neuritis) fail to resolve within 2-4 weeks, seek specialist advice and consider adding thalidomide (see below for guidelines and precautions).
- Thalidomide can be considered as first line therapy for severe ENL.
- If eye symptoms are present, urgent ophthalmology review must be arranged.
- If ENL becomes chronic, consider adding extra clofazimine and seek specialist advice (see below for guidelines and precautions).

7.2.3.3 Thalidomide:

Mechanism of action and indications:

- Exact mode of action unclear, however reduction in TNFα appears to be important in treating ENL.
- Very effective in controlling skin lesions and fever associated with acute ENL and preventing recurrences.
- Rapid onset of action, within 48-72 hours.
- Indicated for acute treatment of moderate to severe ENL, maintenance therapy and suppression of ENL recurrences.
- If neuritis is present, thalidomide needs to be used in combination with prednisolone.

Prescribing requirements:

- Available through a restricted distribution program similar to the United States FDA System for Thalidomide Education and Prescribing Safety (STEPS programme) https://www.iaccesscelgene.com/Prescriber/Register.
- Treatment must be initiated and monitored under the supervision of a specialist in the management of leprosy experienced in the treatment of ENL.
- Informed patient consent must be obtained prior to prescribing.

Adverse effects:

- Common: drowsiness, somnolence and sedation (care with driving and operating heavy machinery).
- Peripheral neuropathy (baseline and 6 monthly nerve conduction studies recommended for use > 6 months).
- Development of peripheral neuropathy requires drug cessation except in rare cases of severe ENL where alternative options are not appropriate.

- Thrombogenicity (thromboprophylaxis recommended for patients with additional thrombotic risk factors).

Contraindications:

- Hypersensitivity to thalidomide or its components.
- Age < 12 years.
- Pregnancy or breastfeeding.
- Patients unable or unwilling to comply with adequate contraceptive measures to prevent pregnancy.

Precautions and contraceptive requirements:

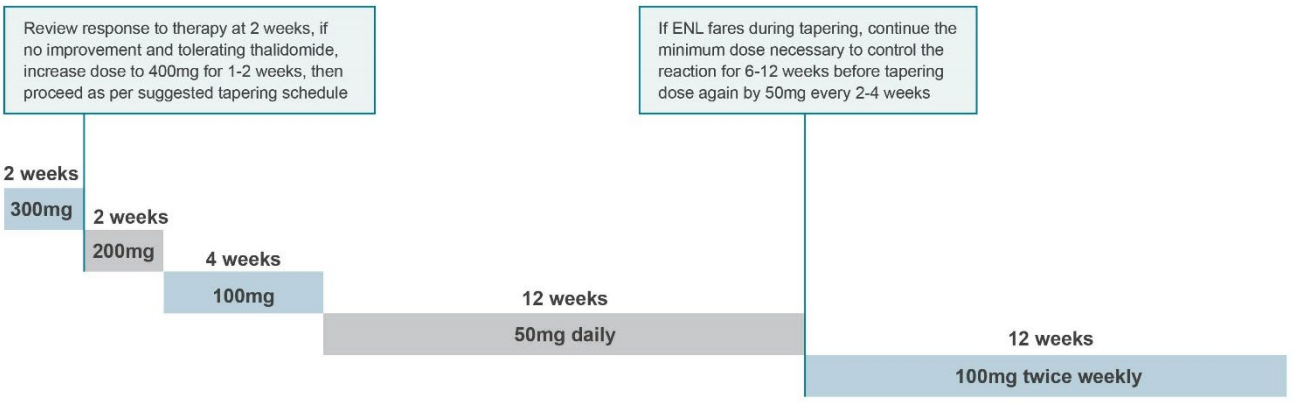
- Risk of teratogenicity requires extreme caution with prescribing, particularly for women of childbearing potential.
- Contraception must be continued for one month after stopping thalidomide.
- Women of childbearing potential must take two forms of contraception, receive detailed counselling and have monthly medically supervised pregnancy test

Dosing:

- Optimal dosing schedule for thalidomide has not been established.
- Suggested regimen for severe ENL is outlined in Figure 7.
- Lower starting doses (eg.100mg nocte) can be used for less severe ENL.

Figure 7. Suggested regimen for thalidomide dosing in erythema nodosum leprosum

Adapted from: VV Pai. Bombay Leprosy Project, Sion-Chunabhatti, Mumbai, Maharashtra, India. Thalidomide in ENL/Type II Reactions in Leprosy – A Decadal Experience. Indian Journal of Leprosy. 2017;89(4): Oct-Dec.



- If neuritis is present, prednisolone should be used in combination with thalidomide
- Prescribe as per guidelines outlined www.guildlink.com.au/gc/ws/celgene/pi.cfm?product=cjpthalo
- Adverse effects include: teratogenicity, venous thromboembolism (VTE) (assess risk), peripheral neuropathy (examine monthly), drowsiness & somnolence, dizziness & orthostatic hypotension, neutropaenia, thrombocytopaenia, bradycardia, SJS/TEN, seizures
- Drug interactions: care with CNS depressants, Oral contraceptive pill (may increase risk of VTE), drugs with potential for peripheral neuropathy
- No dose adjustment required for renal impairment

7.2.3.4 Clofazimine for chronic ENL:

- Clofazimine has anti-inflammatory activity at higher doses.
- May help to facilitate dose reduction of corticosteroids in chronic ENL.
- Increase clofazimine to 300mg daily for one month (maximum of 3 months).
- Allow 4-6 weeks for any beneficial effect to be apparent.
- Reduce to 200mg daily for 3-6 months, then 100mg daily until completion of MDT.
- High doses of clofazimine are associated with gastrointestinal side effects, skin discoloration and ichthyosis.
- Abdominal symptoms in a patient on high dose clofazimine should be investigated promptly.

7.2.3.5 Apremilast for recurrent or chronic ENL:

- Selective phosphodiesterase (PDE)-4 inhibitor with anti-inflammatory and immunomodulatory effects.
- Titration of dosing over 5 days to 30mg bd maintenance dose as follows: d1 10mg mane ; d2 10mg bd ; d3 10mg mane 20mg nocte ; d4 20mg bd ; d5 20mg mane 30mg nocte ; d6 30mg bd.
- Oral agent.
- Adverse effects mild and usually transient, including diarrhoea, nausea, headache, depression.

7.2.3.6 Alternative agents for ENL:

In patients in whom prednisolone, clofazimine or thalidomide are either ineffective or contraindicated, specialist advice should be sought. Agents including azathioprine, methotrexate, TNFα inhibitors (including infliximab and etanercept), pentoxifylline, cyclosporine A, mycophenolate mofetil and oral zinc can be considered.

7.2.4 Treatment of Lucio's Phenomenon

Patients with Lucio's Phenomenon usually respond well to the initiation of MDT. In the uncommon situation where it occurs after commencing MDT, prednisolone can be commenced and tapered as per the schedule outlined for Type 1 reaction. (see Table 15). Thalidomide is not effective.

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Chapter 8 Prevention and management of disability

8.1 Introduction

Despite widespread use of multi-drug therapy (MDT) and a fall in the prevalence of leprosy, global rates of Grade 2 disability (G2D) among new cases of leprosy have remained largely static. In some regions, including South East Asia, rates of G2D among new diagnoses have increased.[1] In Western Australia, the current rate of G2D amongst new cases of leprosy is greater than 50%, well above the WHO target of less than 5%. Historical data is not available to allow assessment of trends.

The current WHO global strategy identifies reducing leprosy related disability as one of its key focus areas, with global targets for a 90% reduction in rate per million population of new cases with G2D and a 90% reduction in rate per million children of new child cases with leprosy by 2030.[2]

The G2D rate among new cases is an important indicator of the efficiency of early detection of leprosy, as well as an indirect marker of community awareness of the early signs of leprosy and expertise of healthcare workers in leprosy diagnosis.

Leprosy related disability is caused primarily by nerve damage, which can occur before, during or after completion of curative MDT. Nerve damage resulting in sensory, motor or autonomic deficits predisposes the patient to injury with development of secondary impairments including ulcers, shortening of digits, contractures, bone destruction, and visual loss. Disability results when these impairments make it difficult for a person to carry out their activities of daily living.

Disability can lead to stigma and discrimination and has a negative impact on quality of life.

Prevention of disability (POD) can be defined as "a concept comprising all activities at an individual, community and program level, aimed at preventing impairments, activity limitations and participation restrictions".[3]

The success of POD is reliant on interplay between personal attitudes and circumstances, and provision of medical care in the form of pharmacological and physical therapies.[4]

The goals of prevention of disability are to:

1. prevent the development of any disability or deformity not present at the time of diagnosis.
2. prevent the progression of disability or deformity.

The components required to achieve these goals include:

1. early detection and treatment of leprosy before nerve damage has occurred
2. accurate assessment of disability at the time of diagnosis and at subsequent assessment
3. early detection and treatment of leprosy reactions
4. education and support to encourage self-care of hands, feet and eyes
5. access to a multidisciplinary team including podiatry, occupational therapy, physiotherapy, and surgery
6. ongoing surveillance of patients at high risk of progression of disability following completion of MDT.

8.2 Early detection and treatment

Clinical and laboratory diagnosis of early leprosy is often extremely challenging. A high degree of suspicion is required, as well as awareness of the early symptoms and signs amongst communities and health care workers.

In areas of low endemicity with disease affecting primarily vulnerable populations such as in Western Australia, the following activities are recommended[5]:

1. Targeted education of health care workers involved in the care of affected populations
    a. Focus on early symptoms and signs which may raise suspicion
    b. Information regarding referral pathways.
2. Education of index cases and affected communities
    a. Simple educational material
    b. Community awareness campaigns.
3. Integration of screening for leprosy (skin check) into school health checks for Aboriginal children.
4. Comprehensive contact tracing of household and community members of diagnosed cases.
5. Optimise treatment adherence through
    a. Patient centred approach
    b. Case holding: supervision of treatment until compliance is established, then monthly directly observed therapy
    c. Ensuring uninterrupted drug supply.
6. Maintaining up to date knowledge and support of developing innovative methods of early diagnosis of leprosy.

8.3 Accurate assessment of disability at the time of diagnosis and at subsequent assessment

Grading disability in leprosy is important for assessing program efficiency, as well as for assessing changes in impairment in an individual patient over time.

Three grading systems are currently in use, the WHO disability grade, the Eye-Hand-Foot (EHF) score (See 5.4.5) and the Impairment Summary Form (ISF)[6] (Appendix 7).

See Chapter 5.4.5 for further detailed information on these scoring systems.

It is important to identify patients with Grade 1 disability (G1D), mainly sensory impairment, and institute measures for POD including education on self-care, provision of adequate footwear and referral for ongoing multidisciplinary care coordinated by the case manager.

Patients with G1D and G2D require lifelong care to prevent progression of their disability.

8.4 Early detection and treatment of leprosy reactions and acute neuritis

Leprosy reactions and acute neuritis are the most important case of nerve damage and subsequent disability and deformity. Approximately 10-30% of all leprosy patients will experience a lepra reaction either before, during, or after completion of MDT. Prednisolone is effective in 50-70% of cases where nerve damage has been present for less than 6 months however optimal dose and duration have not been clearly defined. Early diagnosis and prompt treatment of reactional states reduces the risk of permanent nerve damage[7].

The following measures are suggested to optimise early detection and treatment among cases of Leprosy:

1. **Patient education:** At the time of diagnosis and treatment completion, educate the patient regarding the signs and symptoms of reactions, and the need to present promptly for review.

2. **Regular nerve function assessment** using VMT-ST (ideally monthly), particularly for high risk patients including
    a. patients with existing nerve function impairment
    b. post-partum women
    c. patients with multibacillary leprosy.

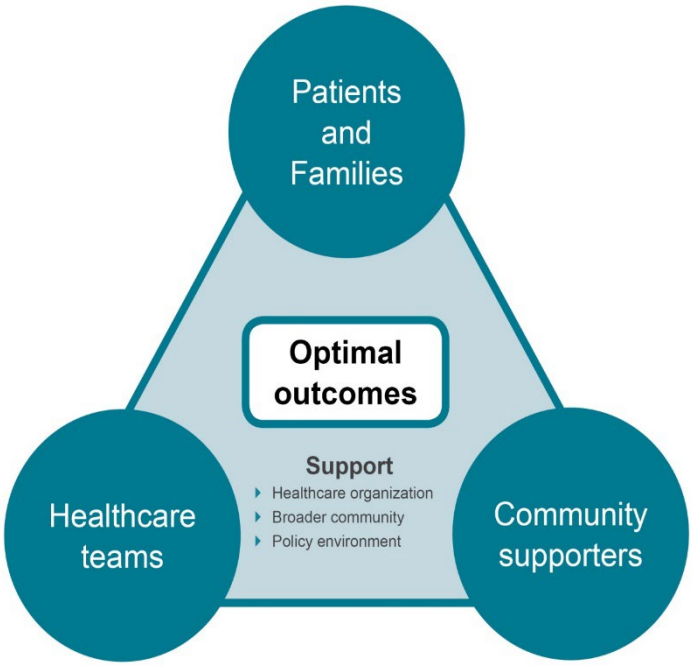
3. **Robust referral systems** with **urgent referral** required in situations where lepra reaction is suspected.

8.5 Self-care

In many cases, leprosy becomes a chronic condition, with peripheral neuropathies predisposing the patient to secondary impairments and subsequent disability and deformity. Self-care is central to the management of chronic conditions, as outlined in the WHO Innovative Care for Chronic Conditions (ICCC) framework[8].

The framework is based on achieving optimal outcomes through a healthcare triad, illustrated in Figure 8.

Figure 8. Healthcare triad (ICCC framework)



The concept of patient-centred care is increasingly being recognised as a core principle of management of chronic disease. The emphasis is on the central role of the patient in the management of their condition, assuming responsibility for their own health outcomes, rather than being a passive recipient.

Self-care is important for all patients with leprosy, particularly those with any degree of nerve damage, and those at risk of lepra reaction.

Patients and their families should be taught self-care practices to prevent disability, with a focus on developing problem solving ability, and ongoing partnership between healthcare teams and the community.

The principles of hand and foot care include[9]:

- **Inspection**
  - Daily inspection of both hands and feet
    - Looking for wounds and cracks
    - Areas of redness or swelling.
  - Daily inspection of footwear

        - Foreign bodies
        - Damage to footwear.
- **Soaking, scraping and moisturizing**
    - To prevent development of callus, cracks and subsequent ulceration.
    - Soak daily in clean water for 30 minutes.
    - After soaking, scrape dry skin away.
    - Follow with an oil based moisturisers or Vaseline.
- **Exercises** (active and passive) to maintain joint mobility and strengthen weak muscles
    - In consultation with physiotherapy/hand therapist.
    - Audio-visual aids can be used to teach patients.
- **Protection from injury**
    - Education regarding appropriate footwear
        - Sports shoes
        - Sandals with heel strap, preferably velcro
        - Firm undersole and soft insole
        - Custom made footwear.
    - Aids to avoid injury
        - In consultation with occupational therapy.
        - Devices to assist with cooking.
        - Splints.

<u>The principles of eye care include:</u>

- **Lagophthalmos**
    - Acute lagophthalmos should be treated as a lepra reaction.
    - Wear glasses and a hat.
    - Keep the eye clean and moist
        - Daily inspection and washing with a clean cloth
        - Lubricating eye drops.
    - Check vision daily by looking at the same fixed object 6 metres away.
    - Daily exercises
        - Try to close the eyes with force daily.
        - Passive exercise required for complete paralysis of orbicularis oris.
    - Surgical assessment is required for chronic lagophthalmos.

- **Corneal anaesthesia**
    - “Think blink”
        - Encourage the patient to force themselves to blink frequently, can be prompted by seeing a common object, such as a certain tree or household appliance.
    - Daily lubricating eye drops.
    - Wear glasses and a hat.

The booklet “I can do it myself!” produced by the WHO is a useful resource to help educate and empower patients affected by leprosy to embrace self-care.

http://english.aifo.it/leprosy/documents/selfcarebooklet-who07.pdf

8.6 Access to a multidisciplinary team

Leprosy patients, especially those who suffer lepra reactions and those with permanent NFI, frequently require a multidisciplinary approach to their care. Services including podiatry, occupational therapy, physiotherapy, ophthalmology and surgery can provide preventative and therapeutic management.

- All patients with any degree of nerve damage involving the feet should be taught self-care of their feet and referred to either a local podiatrist or to the Podiatry service at Royal Perth Hospital (RPH). Telehealth is available for remote patients who cannot attend the RPH clinic.

- Patients with a facial patch or any degree of eye involvement should be referred to an ophthalmologist for initial assessment and ongoing review.

Referral should be facilitated through the WA TB Control Program physician and case manager. Where consultation at RPH is not feasible, alternative arrangements for review by visiting podiatrists and specialists should be made. Communication between involved parties is extremely important, and can be facilitated by email, telehealth and multidisciplinary meetings.

8.6.1 Reconstructive surgery

The aim of reconstructive surgery is to restore form and function.

Indications for surgical referral include:

- Deformity (secondary impairment) present for > 12 months
    - Claw hand
    - Chronic foot ulceration
    - Lagophthalmos
    - Foot drop.
- Patient not currently receiving prednisolone.
- No active neuritis or lepra reaction.
- Patient motivated and committed to pre and post-operative physiotherapy.

8.7 Ongoing surveillance

Patients at high risk of progression of disability require ongoing surveillance and care following completion of MDT.

The following patients are at high risk of progression of disability following completion of MDT.[10,11]

- Patients with existing nerve function injury.
- Patients with MB disease.
- Women of childbearing age (post-partum risk).
- Patients who have had lepra reactions or neuritis.

These patients require a patient-centred chronic disease approach to their management, with ongoing self-care, multidisciplinary team involvement and surveillance.

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Chapter 9 Case Management

9.1 Introduction

Case management is a form of healthcare delivery in which a management plan is individualised for the patient according to their personal circumstances and developed by a multidisciplinary team to achieve a specific and measurable outcome.

Case management is central to the success of the WA TB Control Program's management of leprosy.[1] This is the nurse-led, individual patient-based activity that ensures that treatment is adhered to and completed satisfactorily, and that contacts are identified and screened. In the treatment of leprosy, it is never enough to prescribe the correct drug therapy alone. The prescription must always be accompanied by case management.

Rationale

Cure of leprosy requires medication to be taken for an extended period of time. Adherence with prescribed drug regimens can be further hampered by the perceived stigma associated with the diagnosis, language and cultural barriers, social stressors, competing work demands, and potential for side effects related to the medication. Patients left to take treatment unsupervised are at risk of having difficulty adhering to the treatment regime.

Complications from leprosy, as well as the medication used to treat the disease, can occur at any time throughout the treatment process. Patients require considerable support and care during the treatment period, provided by the case manager in collaboration with the treating physician. Case management ensures early detection of lepra reactions and acute neuritis, facilitating early treatment with the aim of minimising permanent deformity and disability.

Contact tracing, a secondary component of case management, aims to interrupt leprosy transmission by early identification of secondary cases and provision of chemoprophylaxis for eligible contacts. Regular screening of contacts and institution of therapy for newly diagnosed active cases aims to reduce the overall burden of leprosy and leprosy related disability at diagnosis.

All patients undergoing treatment for leprosy in Western Australia are assigned a case manager from the WA TB Control Program based at the Anita Clayton Centre (ACC), Perth. This includes patients diagnosed and treated for leprosy by both private and public physicians outside the WA TB Control Program.

The case manager is allocated according to the index case's home address and pre-defined regions assigned by the program. This allocation can be altered at the case manager's discretion and by agreement.

For patients located outside metropolitan Perth, a local case worker will be assigned with coordination and care delivered as outlined in the Memorandum of Agreement between WA Country Health Service (WACHS) and the WA TB Control Program.[1] The case manager works closely with medical staff and other professionals involved with the care of the patient to support the patient to completion of his or her leprosy treatment.

9.2 Components of case management

Case management in leprosy involves a range of activities commencing from the identification of the leprosy patient, through to contact tracing, assessment and education of patients early after the diagnosis, support and care coordination during the treatment period and case review once treatment is completed.

The components of case management include.[2]

1. Case detection including contact tracing investigation.
2. Assessment.
3. Care planning.
4. Care coordination
    a. Medication management including Directly Observed Therapy (DOT)
    b. Self-care support
    c. Advocacy and negotiation
    d. Psychosocial support
    e. Monitoring and review.
5. Case review and ongoing follow up at completion of treatment.

9.3 Case detection

Case detection is the early identification of patients with leprosy, to ensure that treatment and control activities can be initiated as soon as possible.

This may involve:

- liaison, networking and communication with both hospital-based and private. practitioners to ensure the early identification of patients suspected of having leprosy
- coordination of contact tracing (See Chapter 11).

9.4 Assessment

The assessment phase involves the gathering of information about the patient's disease and social circumstances to assist with the planning of leprosy treatment. Information may be gathered from a variety of sources including the patient, other health care providers, community-based agencies, and other government departments e.g. housing and schools. Assessment should be initiated as early as possible after diagnosis. It may take place at the first clinic appointment or at the first home visit by the case manager. Prior to the first home visit a risk assessment must be undertaken and a mutually suitable time to visit the patient at their home confirmed.

During the assessment phase the case manager should aim to:

- complete Nursing Assessment: identifying demographic and medical history on relevant database.
- collect and record surveillance data as a requirement for statutory medical notifications.
- obtain or review previous medical history.
- identify household and close contacts to guide contact tracing activity..
- evaluate the patient's knowledge and beliefs about leprosy
- assess any leprosy medication that has been prescribed.

- identify any barriers to treatment adherence e.g. difficulty swallowing tablets, transportation problems to attend appointments, issues that may require directly observed therapy
- develop an understanding of the patient's social circumstances that may impact on completion of treatment i.e. living arrangements, housing issues, employment, education, residency status, travel plans, welfare issues, cultural background and presence of any drug or alcohol misuse.

Patient assessment should continue throughout the treatment period in order to detect any changes in the patient's circumstances that might affect treatment compliance.

9.5 Care planning

The patient's care plan is pivotal to case management and should be developed with consideration to the individual's personal circumstances, their health needs and service provision. The plan should be developed in consultation with the patient and their medical team. The care plan will change according to the patient's individual situation and clinical progress. Care planning should be an ongoing process throughout the treatment period and should include the commencement of the contact tracing investigation, monitoring of drug therapy, regular medical review and assessment of adherence.

9.5.1 Case management meeting

A case management meeting is held at least monthly, to plan and review the care of active leprosy cases. The purpose is to document new cases, discuss issues that may be a barrier to successful treatment, discuss the extent of contact tracing, address problems which arise, provide a forum for peer review, and ensure that critical outcomes are achieved. For patients being cared for outside metropolitan Perth, case meetings are held 4-6 weekly via videoconference.

Case histories are collated by the assigned case managers on a shared spreadsheet, with new cases added at the time of diagnosis. All new notifications in the interval since the last meeting are discussed, in addition to any case with adherence issues, lost to follow up, transferred out of WA or completing treatment.

A summary of case discussion and any proposed changes to management should be documented in the patient's record and communicated with their care providers. This is particularly important for patients managed outside metropolitan Perth and for those cared for by Physicians outside the WA TB Control Program.

Attendance at the meeting is open, but it is primarily for the WA TB Control Program physicians and case managers. The meeting is chaired by the Clinical Nurse Manager (or Medical Director of TB in her / his absence) but is an open forum format with decision making by discussion and consensus. Videoconference case meetings with WACHS organisations are attended by a WA TB Control Program Physician, the WA TB Control Program WACHS case manager, relevant local case workers, a member of the relevant Regional Physician team, a representative from KPHU and the regional leprosy nurse specialist. Other involved health care providers attend where required.

9.6 Care coordination

The case manager should act as the central point of contact for the patient for the duration of their treatment. The case manager is the coordinator of care and primary source of support for the patient and should be in continuous communication with the patient throughout the treatment duration, via telephone, home visits or clinic attendances.

Care coordination for leprosy can be structured as follows:

- Diagnostic assessment
  - Initial medical assessment and completion of relevant investigations
- Start of Treatment: Clinic visit
  - Seen by physician and case manager
  - Begin discussions on contact tracing
  - Supply one month's medication
- One week: Home visit
  - Assess environment
  - Complete management plan
  - Assess medication related issues
- Two weeks: Clinic visit
  - Physician and case manager
  - Clinical review
  - Assess medication adherence
- Three weeks: Home visit
  - Assess medication adherence
- Four weeks: Clinic visit
  - Physician and case manager
  - Clinical review
  - Supply one month's medication
  - Assess medication adherence
- Monthly review: Clinic visit
  - Physician and case manager
  - Clinical review
  - Supply one month's medication
  - Assess medication adherence
- Fortnightly or more frequent review may be required in the following circumstances:
  - Issues with adherence
  - Management of lepra reaction or acute neuritis
  - In the setting of adverse drug reaction or coexisting illness

9.6.1 Medication management

An essential part of successful leprosy treatment is the completion of prolonged antibiotic therapy. In addition, patients who experience lepra reactions require close supervision of prednisolone and other medication.

Important roles of the case manager include monitoring patient adherence to the prescribed medication regimen, supporting the patient through medication side-effects, recognising lepra reactions, and identifying and addressing promptly any barriers to medication compliance. The treatment should be directly observed on the first day of the monthly prescription, and either self-administered or directly observed at various intervals as required for the remainder of the monthly schedule. The case manager should work closely with the medical team to ensure that the medication is being taken according to the prescription, and to ensure optimal adherence.

Case managers are responsible for ensuring that patients have an adequate supply of medication. This includes patients living in rural and remote areas. In rare instances where patients with leprosy are managed by Physicians without consultation with the WA TB Control Program, medication and case management can still be provided through the WATBCP. These patients must have a medication chart at the WA TB Control Program.

9.6.1.2 Non-adherence and directly observed therapy (DOT)

Case managers must encourage adherence thorough education outlining the indication for and importance of treatment, management of adverse effects, the use of dosette boxes or Webster packing as required and ensuring a regular supply of medication. Adherence should be checked regularly by direct questioning of patients, pill counts, and rapid contact whenever a patient does not attend planned appointments. If there is evidence of non-adherence it should be discussed with the treating physician as soon as possible, and a plan made between the case manager and physician for enhanced monitoring, formulation of timelines for tolerance of incomplete adherence and measures to be taken if this threshold is reached.

Direct observation of the patient taking the first dose of monthly therapy is a requirement of leprosy treatment in WA. DOT may be recommended more frequently in the following circumstances:

- Demonstrated poor adherence that jeopardises successful leprosy treatment
- Relapsed leprosy where non-adherence is suspected
- All hospital inpatients (while in hospital)
  All patients within correctional services
- Any other patient where the case manager considers there to be a high risk of non-adherence.

DOT involves observing the patient swallowing every dose of treatment.

The process should be explained to patients at the start of leprosy treatment, and the value of DOT should be reinforced by the treating physician and the case manager.

DOT is established and managed by the case managers at the WA TB Control Program.

DOT is most commonly provided by the case manager, but with consent from the patient, can be provided by a community nurse or service, local doctor, local pharmacist, correctional staff or hospital staff. The external DOT providers should be given information and instruction on DOT and a Directly Observed Therapy Log sheet.

The DOT Log sheet can be found at https://healthpoint.hdwa.health.wa.gov.au/policies/Policies/NMAHS/Public%20Health/WATBCP.Directly%20Observed%20Therapy%20Log.pdf and is reproduced in Appendix 13. The log should be returned to the service on a monthly basis. It is not recommended that family members observe therapy as they are not typically neutral or objective about the patient's health.

DOT can be arranged for any location convenient and safe to the patient and the provider. Community based DOT can be provided more efficiently by establishing partnerships with services based in the community.

When a patient refuses treatment and cannot be managed by routine case management or DOT, there is provision in the Public Health Act 2016 Part 9: Notifiable Infectious Diseases and Related Conditions;[3] to enforce a test order or public health order. The provisions for doing this are extremely limited, and the situation should be discussed with the Medical Director of the TB Control Program. All socially and culturally appropriate avenues to assist with adherence should be exhausted before considering further action.

Incentives and enablers may assist with adherence. Incentives are small, non-monetary rewards given to patients to encourage them to take their medications or attend appointments. Enablers can assist clients to take treatment and attend appointments by overcoming barriers such as transportation issues. Incentives and enablers can be provided after consultation with the Clinical Nurse Manager.

9.6.2 Self-care support

The level of support offered to leprosy patients by their case manager will vary according to the complexity of the disease and needs of the individual. While patients are supported to manage their own condition, the case manager may:

- ensure the patient has a good understanding of his or her condition and provide continuous education regarding leprosy and its treatment .
- provide and/or make referrals for general health education and advice e.g., diet, exercise, smoking cessation.
- provide and/or make referrals for advice on health conditions occurring as a complication of leprosy or specific to the patient's circumstances e.g., podiatry, ophthalmology, physiotherapy, ensuring general practitioner involvement for management of diabetes.
- provide education on navigating the healthcare system and services to contact regarding non-urgent issue.

9.6.3 Advocacy and negotiation

A key role in case-management is advocating for and negotiating on behalf of the patient for access to services and needs identified in the care plan. This may involve liaising with

other government departments e.g., housing, social security, liaising with employers on behalf of the patient; ensuring the patient is aware of and attends appointments for referrals made to other providers; providing education for the patient's family and friends regarding leprosy and its treatment.

The case manager plays an important role in keeping the treating Physician informed of the patient's condition and treatment. The case manager should ensure that the Physician is aware of any difficulties or compliance issues and assist with explaining the Physician's recommendations to the patient.

For patients treated in regional areas, the WA TB Control Program case manager has a responsibility to communicate weekly with the local case manager regarding patient progress and medication adherence. Documentation of clinical information should be made both at the WA TB Control Program and in the local record.

9.6.4 Psychosocial support

The leprosy case manager has the most contact with the patient and should remain with the patient from time of diagnosis until discharge from the program. The case manager should provide continuity of care throughout the treatment duration, either directly or through the local case worker for non-metropolitan patients. This regular contact ensures support for the patient and promotes completion of therapy. Being diagnosed with leprosy and the social stigma associated with the diagnosis can be a source of great distress for the patient, and the case manager has an important role to help them through this difficult time. The aim of case management beyond the successful treatment of leprosy is to rehabilitate the patient to full pre-morbid health and function.

9.6.5 Clinical handover

It may be necessary for case managers to handover the care of their patients when they go on leave. The patient should be made aware of the handover and given contact details for the relieving case manager. The handover should be documented in the patient record at the WATBCP and in the local record for patients treated outside Perth.

If a client is transferred to regional Western Australia, interstate or outside of Australia, a nursing summary with a management plan should be provided to the receiving health service. The document can be found at https://healthpoint.hdwa.health.wa.gov.au/policies/Policies/NMAHS/Public%20Health/ACC%20Clinical%20Handover%20Procedure%20Guidelines.pdf . This document can be sent with the medical summary. Acknowledgement that the document has been received by the health service should be documented in the patient record.

9.6.6 Monitoring and review

A case-manager's role is to determine if a patient is receiving and complying with appropriate leprosy treatment and revising the care plan as necessary. The frequency of monitoring is dependent on the patient's circumstances and level of need. It may vary during the treatment program i.e. more frequently at the beginning of treatment or increase in times of personal crisis or treatment issues e.g. medication side effects.

Monitoring may take place daily, weekly or monthly and may occur in a variety of forms i.e. direct contact through clinic appointments, home visits or telephone contact. Email can be

used, but only for simple information e.g. confirming appointments, and not for collecting personal medical details or conveying medical advice. All contacts with patients and other care providers should be recorded in the patient's record.

Home visiting can be used to evaluate the patient's home environment and social situation and is a tool to monitor patients and provide support to promote adherence. Not all patients require regular home visits, but it is recommended that the case manager meets with patients taking treatment for leprosy at least monthly. This can be done at home or in clinic. Prior to any new site or home visit a risk assessment is to be completed and reviewed by the Clinical Nurse Manager or Area Manager. The Risk Assessment tool can be found on the patient database. If it is not possible to carry out a risk assessment, two people are required to attend the site or home.

9.7 Case closure

The process for discharge of patients from the case management program or 'case closure' should be clear and defined in time. The aim for case management for leprosy should be the successful completion of medical treatment. The decision to discharge a patient should be determined by the case manager and treating physician.

The timing of case closure after leprosy treatment varies significantly, and must be assessed on an individual basis, considering disease classification, bacillary index at time of diagnosis, presence of lepra reactions, presence of nerve function injury and the need for ongoing prevention of disability.

The decision on follow up and case closure is made by the medical physician and case manager (see Chapter 6.5.3 Follow up after completion of leprosy treatment).

In a number of cases, case closure may not occur for several years after completion of drug therapy. For this reason, the case manager should update the Western Australian Notifiable Infectious Disease Database (WANIDD) (see the WA TB control policy Chapter 12.0 *Notification of Leprosy and enhanced surveillance data)* and ensure all contacts have been assessed, screened and managed (see WA TB Control program Policy Chapter 11 *Contact tracing*) when the patient completes drug therapy, and add additional information as required at the time of case closure.

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Chapter 10 Prevention of Leprosy

10.1 Introduction

The introduction and widespread implementation of multi drug therapy (MDT) for leprosy in the 1980s has been successful in achieving a dramatic fall in global prevalence, from over 5.2 million people, to 202, 256 at the end of 2019. The impact on new case detection rate and disease transmission has been less impressive, with stagnation in figures over the past decade. Several countries continue to have a high burden of disease, both at a national and subnational level with pockets of higher endemnicity,[1]

It is therefore clear that measures other than MDT are required to interrupt transmission and prevent leprosy on a global scale. This has been recognised and addressed by the WHO and the most recent Global Leprosy Strategy,[2] with a focus on strengthening political commitment to ensure adequate resources for clinical care and research, promoting early case detection, and promoting interventions for the prevention of infection and disease.

Interventions aimed at interrupting disease transmission and preventing leprosy include:

- MDT: optimisation of drug supply, compliance and addressing drug resistance
- Early case detection through contact tracing and active case finding
    - Enhanced through development of tests to facilitate early diagnosis and subclinical disease
- Immunoprophylaxis
- Chemoprophylaxis (post exposure prophylaxis)
- Addressing social factors such as overcrowding.

This chapter will focus on vaccination and post exposure prophylaxis, with other interventions discussed in other sections of the guideline.

10.2 BCG vaccination

Bacillus Calmette-Guerin (BCG) vaccine was developed in the early 20th century with the aim of preventing tuberculosis. While protection against pulmonary tuberculosis is relatively poor, BCG vaccine provides good protection against miliary tuberculosis and tuberculous meningitis[3].

BCG protection against leprosy varies between 20% and 90%, with study design (BCG efficacy in experimental studies = 41% versus observational studies 60% and features of the target population (efficacy higher in contacts of leprosy compared with the general population; 68% versus 53%) likely to be the most significant factors influencing observed vaccine efficacy[4]. The impact of vaccine strain, exposure to environmental non-tuberculous mycobacteria, number of doses of BCG and age at vaccination are less clear.

BCG is categorised as being a beneficial intervention for leprosy, with evidence graded as being of low quality, according to the most recent British Medical Journal Clinical Evidence publication[5].

The protection against leprosy offered by BCG alone is inadequate to interrupt transmission of leprosy. However, its importance as a component of preventative strategies remains significant. Results of a recent modelling analysis comparing the predicted impact of future

intervention strategies on leprosy case detection rates indicate that infant BCG vaccination, contact tracing, early diagnosis and treatment of leprosy, and chemoprophylaxis have the greatest impact[6].

Alternative leprosy vaccines under investigation include ICRC vaccine, *Mycobacterium indicum pranii* (*Mw*) vaccine[7] and killed *M.leprae*, either alone or in combination with BCG, and as both immunoprophylaxis and immunotherapy in combination with MDT.

10.2.1 Western Australia BCG Policy

BCG vaccination should not be offered routinely to Australian residents. However, it is indicated in the following people:

1. Children less than 6 years old who are going to live in another country with a high incidence of tuberculosis (defined as an annual incidence of > 40 / 100 000 population) for more than 3 months (once off or cumulatively). BCG should be given 2 – 3 months prior to departure.

   For country specific incidence rates see the World Health Organisation TB country profile website http://www.who.int/tb/country/data/profiles/en/index.html

2. Newborn children of migrants who have arrived from countries with a high incidence of tuberculosis (see definition above) in the last 5 years, or newborn children who have household contact with people who have arrived from a high incidence country in the last 5 years.

3. Newborn children of parents with Leprosy or a family history of leprosy.

4. Children less than 6 years old who have not previously been vaccinated with BCG and are household contacts of newly diagnosed leprosy.

5. Infant household contacts of TB after empiric prophylaxis if TST remains negative.

BCG vaccination is not recommended for general use in the Australian population. BCG can be considered for persons not included in these indications. However, care should be taken to adequately inform all persons of the potential risks and low efficacy of the vaccine, especially in adults.

It is recommended that the decision to give BCG outside of the above indications should be discussed with the Medical Director or Clinical Nurse Manager of the WA Tuberculosis Control Program.

Information on BCG contraindications, general considerations, and dose and administration can be found in the WA TB Control Program TB policy at https://ww2.health.wa.gov.au/Articles/A_E/About-the-Western-Australia-Tuberculosis-Control-Program.

10.3 Chemoprophylaxis

10.3.1 Review of evidence

Chemoprophylaxis for leprosy has been of interest since the 1960s. Early studies with dapsone and acedapsone given to contacts showed an overall reduction in leprosy of between 40% and 60%.[8,9] The long duration of treatment and side effects associated with these drugs prompted studies and programs utilising other regimens, including rifampicin, and the combination of rifampicin, ofloxacin and minocycline (ROM).[10]

Chemoprophylaxis with single dose rifampicin (SDR) has been shown to have a protective effect in leprosy contacts when used in high incidence settings. The COLEP study in Bangladesh, a randomised placebo controlled trial conducted in a highly endemic region in northwest Bangladesh showed an overall reduction in incidence of leprosy of 57% (CI 33-72%) at 2 years for contacts who received SDR[12]. No further risk reduction was seen at 4 and 6 years.[13] The overall number needed to treat to prevent one case of leprosy was 265 after 2 years, and the intervention was found to be cost effective.[14]

Interestingly, SDR seemed to be most effective in contacts determined as having a lower risk of developing leprosy:

- Those not closely related or in close physical contact
- Contacts of index cases with paucibacillary disease
- Social contacts.

Results of this study are consistent with those of an earlier unblinded intervention study conducted on five Indonesian islands using two doses of rifampicin given approximately 3.5 months apart[15]. The study noted that the cumulative new case detection rate in the control islands was 39/10 000 population while in islands using the blanket approach the new case detection rate was found to be approximately three times lower. These findings support the observation that protection from chemoprophylaxis appears to be greatest for those at lower risk, either through lower exposure or lower susceptibility.

While rifampicin chemoprophylaxis appears to have short term protective efficacy in subgroups of people in highly endemic regions, uncertainty remains regarding the applicability of results to areas of lower endemicity or with pockets of higher endemicity, optimal drug choice (rifampicin vs rifapentine), dosing frequency, adverse effects of rifampicin in the medium and longer term, and the suitability of blanket versus targeted chemoprophylaxis in different settings.

The results of the leprosy post-exposure prophylaxis (LPEP) programme, an international, multicentre feasibility study implemented within leprosy control programmes of Brazil, India, Indonesia, Myanmar, Nepal, Sri Lanka and Tanzania was published in October 2020. The programme assessed the feasibility of combining three key interventions: systematically tracing contacts of individuals newly diagnosed with leprosy, screening the traced contacts for leprosy, and administering SDR to eligible contacts. The overall findings were that post-exposure prophylaxis with SDR was safe, can be integrated into control programmes with minimal additional efforts once contact tracing has been established, and is generally well accepted by index patients, their contacts and health care workers.

The effectiveness of BCG vaccine alone versus BCG in combination with SDR in the prevention of leprosy among contacts of newly diagnosed leprosy cases was assessed in a

single centre, cluster randomized trial conducted in Bangladesh (MALTALEP trial).[18] Whilst the trial noted a 42% reduction in the incidence of paucibacillary leprosy amongst contacts who received SDR after BCG vaccination, the result was not statistically significant.

Post exposure prophylaxis (PEP) with single dose rifampicin is currently in use as part of national policy in Cuba, Morocco and Samoa, and as part of subnational policy in Australia (Northern Territory), India, Indonesia and Nepal. In addition to the LPEP study, other collaborations are investigating the feasibility and efficacy of one month of multibacillary multidrug therapy for household contacts in the Democratic Republic of Congo, and in Cambodia, SDR PEP is being administered contacts of retrospectively identified index cases in an area of low case detection.[19]

10.3.2 WHO Recommendations

In 2018, the WHO formally recommended chemoprophylaxis with SDR for contacts of leprosy cases (adults and children 2 years and above), after excluding leprosy and TB disease, and in the absence of any other contraindications. They specify that this intervention should only be implemented by programmes that can ensure: (a) adequate management of contacts, and (b) consent of the index case to disclose his/her disease. The recommendation has been made utilising the GRADE approach, with the strength of recommendation classified as “Conditional”, and the quality of evidence “Moderate”.[20]

10.3.3 Western Australian recommendations

Chemoprophylaxis with single dose rifampicin should be offered to all household contacts of a newly diagnosed case of leprosy, following screening for signs of leprosy and exclusion of contraindications as outlined below. Consent of the index case for disclosure of the diagnosis to the contacts is required.

In remote settings where there is frequent close contact outside the household, or where the household is extended (for example Aboriginal communities), chemoprophylaxis should be offered to each member of the community with individual medical assessment prior to prescription. In this situation, disclosure and consent of the index patient is not required. The community should instead be informed that leprosy cases have been found in the region over the past years, and that community screening is planned to find hidden cases and prevent further cases. Community screening must be accompanied by an education campaign aimed at raising awareness about the clinical signs of disease, and to address problems which may arise as a result of fear and stigma.[21]

Chemoprophylaxis should be administered as part of contact tracing activities. The program will be conducted through an operational research study aimed at assessing the feasibility, acceptability, adoption, cost, coverage and sustainability of the intervention in WA.

10.3.4 Process for Chemoprophylaxis

Following identification of a new active case of leprosy (Index Case):

- A decision on the extent of contact tracing will be made by the Physician at the WA TB Control Program in consultation with regional Public Health Unit for non-metropolitan patients.
- Definitions for household and community contacts can be found in Chapter 11.
- Contact tracing should occur between 1 and 6 months after the index case has commenced MDT.

- Where household screening is to be undertaken:
    - The rationale for contact tracing should be explained to the Index case, and their verbal consent for disclosure of diagnosis obtained
    - If consent obtained, the assigned case manager is responsible for creating a list of identified contacts
    - For contacts residing outside metropolitan Perth, collaboration with regional public health units should occur in the planning of tracing and screening activities.
- Where community screening is to be undertaken:
    - The assigned case manager and treating physician should liaise with relevant public health units, stakeholders and community health centres to carefully define the size of the targeted community
    - Consideration should be given to the local disease epidemiology, available resources, feasibility and acceptability screening.
- When conducting contact screening activities, the *Leprosy Contact Screening Form for Consideration of Chemoprophylaxis* should be used. (see Appendix 14)

- In addition, documentation of the following indicators related to performing contact screening is required:
    - Number listed (for household and neighbour screen)
    - Number estimated (for community screen)
    - Number traced
    - Numbers screened
    - Number confirmed or suspected leprosy
    - Number received SDR
    - Number refusing SDR

- Data obtained will be recorded in a secure central database at the WA TB Control Program, with information used for clinical follow up as required, and to facilitate reporting.

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Chapter 11 Contact Tracing

11.1 Rationale

Unlike many other infectious diseases, the pathogenesis and exact mode of transmission of leprosy are not well understood. This is mainly due to the inability to grow *M.leprae* in the laboratory, and lack of a suitable animal model. Formulating clear guidelines for leprosy contact tracing is therefore somewhat challenging, with recommendations based on limited evidence and expert opinion.

11.1.1 Transmission

Leprosy is thought to be transmitted primarily person to person via droplets from nasal secretions of infected patients, and to a lesser extent through direct contact with skin lesions. Patients with untreated multibacillary disease are believed to be the primary source of infection, however all cases of active leprosy should be considered potentially infectious. It is believed that subclinical cases may also contribute to transmission, with identification of the infective source not possible in a significant number of cases. Infection associated with inoculation from environmental sources has been documented, as has zoonotic transmission following contact with armadillos.[1]

Leprosy is not considered to be a highly infectious disease, generally requiring prolonged periods of close contact. It is believed that the majority of people have natural immunity to infection with *M.leprae*, with genetic factors being of primary importance in influencing susceptibility.[2] The likelihood of transmission is to a lesser extent influenced by various factors including extremes of age, hygiene, overcrowding, pregnancy and nutrition. It is estimated that only 3-5% of conjugal partners of patients with untreated lepromatous disease develop infection, with a higher risk observed in other family members, particularly children[3]. Evidence suggests that in areas of lower endemicity such as Australia, the majority of new cases will be found amongst contacts of known active cases, emphasizing the importance of contact tracing.[4]

11.1.2 Incubation period

The incubation period for leprosy is highly variable and potentially long, requiring extended periods of annual surveillance for effective contact tracing. The average incubation period is thought to be between 5 and 7 years, however disease may not manifest for 20 years or more.

The widespread use of drug therapy has not been successful in eradicating infection, with stabilization of the new case detection rate evidence of ongoing transmission of infection in a number of areas worldwide. Other interventions, including preventative strategies and contact tracing are therefore required.

Contact tracing is a key intervention in the World Health Organization Global Leprosy Strategy[5], with a target to screen all household contacts of active cases.

The objectives of contact tracing are to:

- promote early case detection
    - Reduce transmission
    - Minimise leprosy associated deformity.
- provide an opportunity for prevention
    - Use of post exposure prophylaxis in some settings.
- provide an opportunity for education
    Promote self-presentation.

11.2 Governance

Contact tracing will generally be undertaken by the Western Australia Tuberculosis Control Program. Primary oversight is provided by the Case Manager for the index case, who in turn reports to and seeks advice from the WA TB Control Program Physician responsible for Leprosy care and the Medical Director for TB Control.

In situations where contact tracing is undertaken by other agencies, there should be consultation with the WA TB Control Program.

In rural and remote areas, contact tracing can be done by the relevant WA regional population / public health unit in consultation with the WA TB Control Program.

There is no specific legislation governing contact tracing, other than the requirement that information on contacts must be collected and kept in compliance with privacy laws.

11.3 Definitions

11.3.1 Index case

The person diagnosed with leprosy prompting contact tracing. If the contact tracing identifies a new case of leprosy, this then becomes a new case that requires independent, although often overlapping, contact tracing.

11.3.2 Household (close) contacts

Household contacts are people who share a bedroom, kitchen, bathroom or sitting room with the index case. Such contacts may include immediate and extended family members, as well as people who cohabit in dormitories or other residential accommodation. A broad definition should be used to maximise the impact of contact tracing, with consideration of cultural and local factors.

11.3.3 Community contacts

Community contacts are those who have less direct exposure to the index case. The exposure is generally outside his or her primary place of residence and may include more distant family members, school or work contacts. The definition of community contacts will depend on local conditions and characteristics of the index case.

11.4. Extent of contact tracing

11.4.1 Timeframe

Contact tracing should begin as soon as possible after diagnosis of an index case with Leprosy. Determining the exact period of infectivity is often challenging. In general, contacts should be determined from the time of onset of symptoms, which may be neurological, skin changes or both. Given that the average incubation period is 5-7 years, contacts of the index case during this period may be included, particularly in situations where the aim of contact tracing is to find a source (for example in contact tracing of paucibacillary cases).

11.4.2 Who to contact trace

- All household or close contacts of the index case.
- In remote settings where there is close contact within communities, extended family and community members should be included.
- For contacts of paucibacillary cases, contact tracing is focused primarily on identifying a possible source of infection i.e. a multibacillary case.
- Retrospective contact tracing is recommended in regions where there is ongoing transmission and surveillance has been incomplete
    - Includes Kimberley and Pilbara regions for cases diagnosed after 1996 when local surveillance programs ceased.

11.5 Procedure for contact tracing

The procedure for contact tracing is outlined in Appendix 15.

11.5.1 Review of Index case

The index case should be carefully interviewed in a culturally appropriate setting to determine lists of contacts that require screening. Confirmation of case classification as multibacillary or paucibacillary, including bacillary index where available, is important to determine duration of contact follow-up.

Contacts of MB cases require annual review for a minimum of six years.
Contacts of PB cases require a single review with no further follow up necessary in the absence of any suspicious findings.

The index case should be educated regarding the risk of possible transmission of infection to their contacts, and potential benefits which may result from contact tracing and administration of chemoprophylaxis where appropriate. Consent from the index case for disclosure of the diagnosis must be obtained prior to proceeding with contact tracing. In situations where community wide screening is planned, consent from the index case is not required, however it is paramount that the index case is not identified during the screening process.

11.5.2 Stratification of Contact List

Contacts should be stratified into groups to reflect the intensity of exposure i.e. household contacts, family contacts, community contacts and casual contacts.

11.5.3 Contact screening

Once the contact list has been created, contacts should undergo the following:

- Interview
  - Questioning for symptoms of Leprosy including skin lesions, changes in sensation, new weakness or deformity
  - History of BCG vaccination
  - Previous TB exposure or treatment
  - Co-existing medical conditions
- Clinical examination
  - Skin, peripheral nerves, eyes as outlined in Chapter 5.1
  - VMT-ST should be performed if examiner is skilled in the procedure (see Appendices 2 and 3)
- Education
  - Educate the contact regarding symptoms and signs of leprosy, to identify early in themselves and others
  - Importance of early presentation
- Investigations
  - If clinical features of leprosy are present the patient should have further investigation including either skin smear or skin biopsy
  - In situations where investigations are not possible, referral should be made to a specialist
- BCG
  - Contacts who have not received BCG should be vaccinated according to WA BCG policy which can be found under policy links at https://ww2.health.wa.gov.au/Articles/A_E/About-the-Western-Australia-Tuberculosis-Control-Program
- Referral
  - Contacts with clinical features suggestive of leprosy should be referred to the Infectious Diseases Physician at the WA TB Control Program for review
- Assessment for eligibility for chemoprophylaxis with single dose rifampicin
  - See Appendix 14
- Arrange subsequent review
  - Contacts of PB cases without any suspicious findings do not require further review
  - Contacts of MB cases should be reviewed annually for a minimum of 6 years
- Contact tracing report
  - A list of all contacts should be maintained by the local case worker and WA TB Control Program case manager
  - The completeness and results of contact tracing should be recorded and communicated in an annual report

11.6 Other considerations

11.6.1 Maintaining confidentiality of the index case

The name of the index case should never be disclosed to contacts without the consent of the index case. Health professionals (including public health authorities) have a duty to maintain the confidentiality of all information that comes to them in the course of providing medical treatment and care to patients. Inadvertent disclosure of a patient's diagnosis of Leprosy to a third party could have adverse consequences for the patient, both at home and in the workplace.

Further information regarding patient confidentiality is provided in the Government of Western Australia, Department of Health, Patient Confidentiality Policy.[6]

11.6.2 Contacts declining screening

Contacts who refuse to be screened for Leprosy should be informed of the symptoms and signs of infection and advised to seek medical attention with any concerns. They should be informed that symptoms might take many years to appear.

11.6.3 Media attention

Media attention may develop when leprosy involves schools, hospitals, detention facilities or other public settings. This is most likely to arise through contacts speaking to the media. Contact tracing procedures and priorities should not alter in this situation. Attention should be paid to clear and prompt communication with contacts to alleviate anxiety and concerns that may prompt erroneous media reporting. Any media enquiry should be addressed as soon as possible, so to ensure accurate reporting. Pre-emptive statements in the setting of contact tracing are not recommended. All media enquiries should be referred to the Medical Director of the WA TB Control Program.

References

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2. Mira MT, Alcais A, Van Thuc N, Moraes MO, Di Fulmeri C, Thai VH, Phuong MC, Huong NT, Ba NN, Khoa PX, Sarno EN, Alter A, Montpetit A, Moraes ME, Moraes JR, Dore C, Gallant CJ, Lepage P, Verner A, van de Vosse E, Hudson TJ, Abel L, Schurr E. Susceptibility to Leprosy is associated with PARK2 and PACRG. Nature 2004;427:636-640.

3. Joyce MP. Historic aspects of human susceptibility to leprosy and the risk of conjugal transmission. Mem Inst Oswaldo Cruz, Rio de Janeiro. 2012;107(Suppl.1):17-21.

4. Richardus JH, Meima A, van Marrewijk CJ, Croft RP, Smith TC. Close contacts with leprosy in newly diagnosed leprosy patients in a high and low endemic area: comparison between Bangladesh and Thailand. In J Lepr Other Mycobact Dis. 2005;73:249-257.

5. Regional Office for South-East Asia, World Health Organization. (2021). Global Leprosy Strategy 2021-2030: Towards Zero Leprosy. WHO Regional Office for South-East Asia. Towards zero leprosy. Global leprosy (Hansen's Disease) strategy 2021–2030 (who.int)

6. Government of Western Australia, Department of Health. Patient Confidentiality Policy MP0010/16. 01 July 2016. http://www.health.wa.gov.au/circularsnew/pdfs/13317.pdf accessed 20/2/2018.

Chapter 12 Notification of leprosy and enhanced surveillance

12.1 Introduction

Leprosy is an infectious disease for which there is a legal requirement for clinicians and laboratories to report the diagnosis to the Western Australia Department of Health. All cases of active leprosy, in both children and adults, are notifiable.

12.2 Statutory medical notifications

Under the *Public Health Act 2016*[1] (the Act) if a medical practitioner, nurse practitioner or responsible pathologist at a pathology laboratory forms the opinion that a patient of the practitioner, or from whom a sample was taken, has or may have, a notifiable infectious disease or notifiable infectious disease-related condition, they have a legal requirement to notify the Chief Health Officer. Part 9 Division 2 of the Act outlines detail relating to notification.

Notification should be made using the approved notification form, by post, fax or telephone, depending on urgency. The Act states that notification should be made as soon as is practicable, or within 24 hours for urgently notifiable infectious disease, and within 72 hours for any other notifiable infectious disease or notifiable infectious disease-related condition. In situations where two or more practitioners may be involved in a patient's management and it is not clear if the case has already been notified, the case should still be reported. This ensures optimal ascertainment of all cases.

In addition to an obligation to notify the Chief Health Officer about notifiable infectious diseases or notifiable infectious disease-related conditions, medical practitioners, nursing practitioners and responsible pathologist have a duty to provide the patient with information about the disease or condition, including; the patient's obligations under section 88(2) – (4); the patient's rights under section 88(5); and preventing the transmission of the disease to any other person; information about the obligation to notify the disease; and any information prescribed by the regulations in subsection (2)(d).

12.3 Case definition for leprosy

The Communicable Diseases Network Australia (CDNA) definition for leprosy is outlined in Chapter 4.

All confirmed cases should be notified to the Department of Health. Cases not meeting the CDNA definition but where a high degree of suspicion of leprosy exists and criteria for the WHO definition of leprosy are met should also be notified. Reporting of cases meeting the WHO case definition ensures accurate reporting by WA TB Control Program to the WHO annually.

12.4 Notification and surveillance process

12.4.1 Western Australia

12.4.1.1 Notification

The responsible practitioner is required to complete a Department of Health Notification form, which is reproduced in Appendix 16 and available from the WA TB Control Program website at http://www.health.wa.gov.au/acc/tb/hp.cfm . Reply paid envelopes are provided with notification forms, and a fax number is provided on the form.

Notifications are sent to the Director of the Communicable Disease Control Directorate (CDCD) for cases diagnosed in the Perth metropolitan area, and to the appropriate Public Health Unit for cases diagnosed in country areas. Pathology laboratories generally provide notifications by automated electronic downloads or relevant information directly to the CDCD.

Core notifiable disease data from both paper-based clinician notifications and electronic or paper-based laboratory notifications are stored in the Western Australian Notifiable Infectious Disease Database (WANIDD), which is accessible to a limited number of authorised users, including designated staff of the WA TB Control Program. Notifications are entered into WANIDD within 24 hours of receipt at CDCD. The database provides real-time surveillance capacity on a state wide basis.

The responsibility for entry of notifications into WANIDD lies with designated staff in CDCD. Notifications are often generated by or sent directly to the WA TB Control Program (Anita Clayton Centre). These are immediately faxed to CDCD for data entry. Conversely, notifications received by CDCD, once entered into WANIDD are faxed to the TB Control Program to ensure the program is aware of the newly identified case. The receiving officer of the fax at the Anita Clayton Centre (usually the Senior Administrative Officer) will then inform the case manager (if already assigned) of the notification, or alternatively the Clinical Nurse Manager or Medical Officer if the case is not already known to the centre. The Mycobacterial Reference laboratory, in addition to sending electronic notifications to CDCD, also notifies the TB Control Program of all new positive *M.leprae* microscopy and PCR by fax.

Cases of leprosy treated in WA but diagnosed in another state or country are not notifiable. The WA TB Control Program is responsible for providing surveillance data back to the notifying state or country as required and where possible.

12.4.1.2 Enhanced surveillance

The WA TB Control Program collects additional information for notified leprosy cases using a specific enhanced surveillance form REDcap database. The form is completed by medical and case management staff at the WA TB Control Program.

12.4.1.3 Review of notification and surveillance data

Data cleaning of the initial notification for leprosy (and tuberculosis) is performed monthly by designated staff at CDCD. Requests to complete missing data fields from the form for notification of infectious diseases in WA for all patients diagnosed with leprosy are emailed to the Clinical Nurse Manager, Medical Director and case managers of the WA TB Control Program monthly.

In May each year, leprosy notifications from the previous 12 months are reviewed at the WA TB Control Program, to ensure completeness and accuracy, prior to submission of the surveillance data to the Commonwealth and WHO.

12.4.2 National Notifiable Diseases Surveillance System (NNDSS)

The National Notifiable Diseases Surveillance System (NNDSS) was established in 1990 and is maintained by the Australian Government Department of Health (DoH). Under this scheme, de-identified core information on cases of infectious diseases that are notified to State or Territory health authorities, are forwarded electronically to DoH on a daily basis for incorporation into the NNDSS database. CDNA, which comprises representatives from DoH and State/Territory Departments of Health, coordinates national surveillance of the agreed list of communicable diseases that are maintained in the NNDSS. Sharing of notifiable disease data across jurisdictions is auspiced under the terms of the *National Health Security Act 2007*.

In addition to the electronic transmission of core surveillance data on notified leprosy cases from WANIDD to the NNDSS, CDCD provides the Department of Health on an annual basis with the additional data collected from the enhanced surveillance scheme.

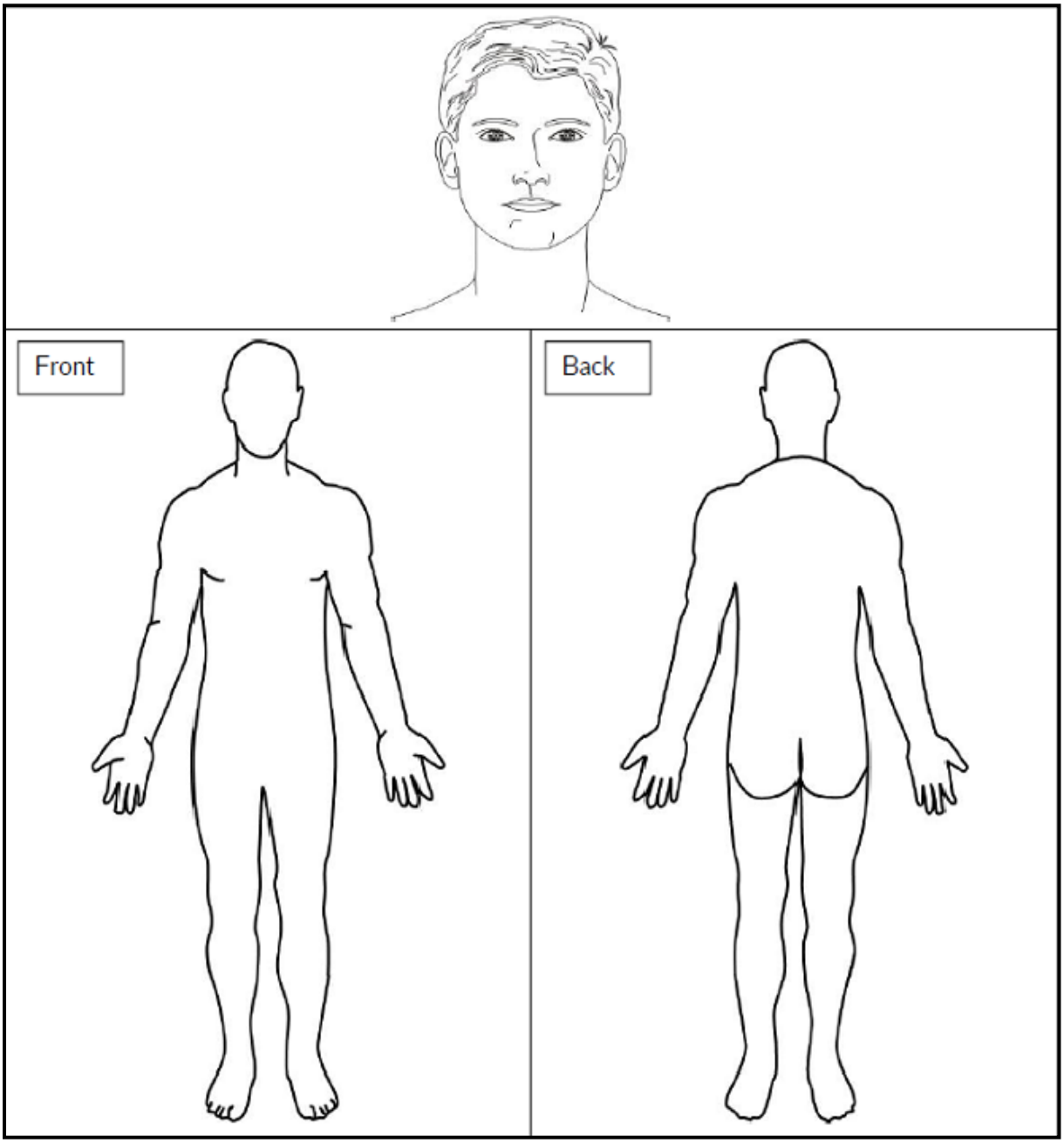
References

1. *Western Australia Public Health Act 2016* (WA). https://www.legislation.wa.gov.au/legislation/prod/filestore.nsf/FileURL/mrdoc_41553.pdf/$FILE/Public%20Health%20Act%202016%20-%20%5B00-g0-02%5D.pdf?OpenElement. [accessed 2022 Jun 6]

Appendix 1 Template for skin examination

DATE: SURNAME: NAME: UMRN:

Please draw skin lesions and include a description including colour, pigmentation, shape, sensation to light touch, edge, presence of satellite lesions, features of the border and presence of hair.



Appendix 2 Guide to performing a VMT-ST: Referral centre (See Appendix 4 for guide to performing VMT-ST)

Date: .....

Surname: .....

Name: .....

UMRN: .....

Section 1: Reactional state:

□ Type 1

□ Type 2

□ None

Section 2: Neuritis check

Sensory or strength change in past 6 months: Y □ N □

Nerve pain or tenderness: Y □ N □

If yes, give details: .....

.....

.....

Section 3: Nerve enlargement		
N= none P= possible D= definite. If nerve tender, mark (t) in relevant box		
	R	L
Supraorbital		
Greater auricular		
Radial		
Median		
Ulnar		
Radial cutaneous		
Common peroneal		
Posterior tibial		
Other(specify)		

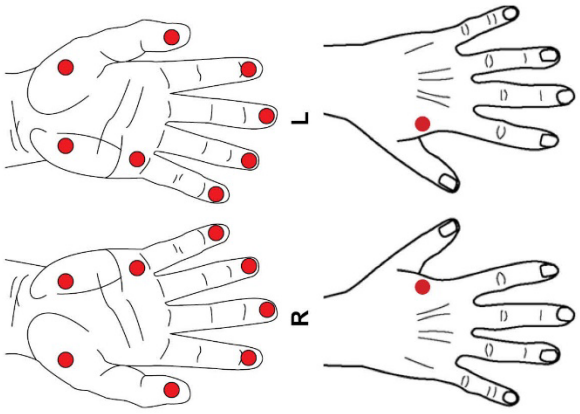
Section 4: Eyes	Right		left	
Blink problems	Yes	No	Yes	No
Power upper lid muscles (try to open on firm closure) (MRC grade 0-5)				
Lid gap light closure		mm		mm
Lid gap strong closure		mm		mm
VA corrected	6/_		6/_	
VA uncorrected	6/_		6/_	
Conjunctival abnormalities: Y/N. If yes, please circle:				
Redness, congestion, watering, other:				

Section 5: sensory testing

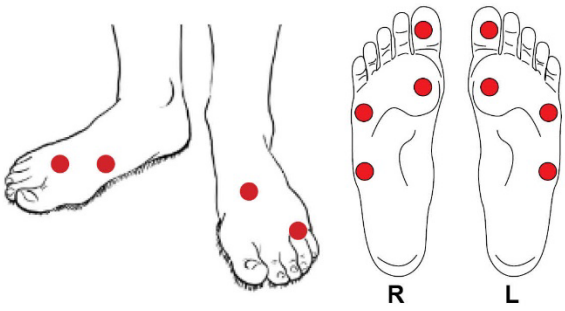
Record highest grade (1-4) felt at each site on diagrams. If no monofilament felt, mark "X"

If impairment of sensation is detected at any of the points illustrated, testing of sensation at more points in the relevant peripheral nerve distribution is recommended.

Document Shortening (=), Clawing (C), Ulceration / Wound (#), Dryness (D), Callus (O), Wasting (W)



Filament	Force	Interpretation	Grade
Blue	0.4g	Normal sensation	3
Purple	2g	↓ Protective sensation	2
Orange	10g	Loss of protective sensation	1



Filament	Force	Interpretation	Grade
Purple	2g	Normal sensation	4
Dark red	4g	↓ Protective sensation	3
Orange	10g	Loss of protective sensation	2
Bright red	300g	Loss of deep pressure sensation	1

Section 6: Muscle testing	Strong (5), Weak (4-2), Paralysed (1-0) R	L
MRC grade		
Ulnar nerve		
Little finger abduction		
Index finger abduction		
Median nerve		
Thumb abduction		
Thumb to little finger		
Radial nerve		
Wrist extension		
Finger extension		
Common peroneal nerve		
Foot dorsiflexion		
Great toe dorsiflexion		
Posterior tibial nerve		
Spreading of toes		

MRC grading: 5=full ROM against gravity & maximal resistance; 4=full ROM against gravity & moderate resistance; 3=full ROM against gravity, no added resistance; 2=partial ROM, no resistance, gravity eliminated; 1=visible or palpable muscle contraction only, no movement; 0=complete paralysis

Section 7: Disability grading

WHO grading (See Figure 3 page 44):

Disability grade hands ___

Disability grade feet ___

Disability grade eyes ___

Max disability grade =

ISF score (See Appendix 7): ___

Assessment of disability progression:

WHO grade	ISF score	Date

Appendix 3 Guide to performing a VMT-ST: Peripheral centre

(See Appendix 4 for guide to performing VMT-ST)

Date: .....

Surname: .....

Name: .....

UMRN: .....

Nerve enlargement

N=None P=Possible
D=Definite

	R	L
Supraorbital		
Greater auricular		
Median		
Ulnar		
Common peroneal		
Posterior tibial		

Eyes

	R	L
Blink problems (Y/N)		
Lid gap(mm)		
Can count fingers at 6m (Y/N)		
Redness (Y/N)		
Pain (Y/N)		

Muscle strength

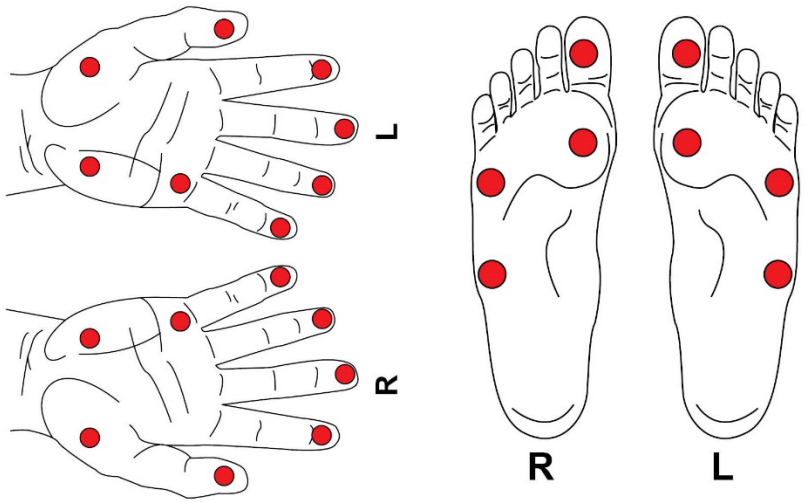
S=strong W=weak P=paralysed

	R	L
Little finger out		
Thumb up & across		
Wrist extension		
Foot up		

Sensory testing

Use ballpoint pen or 10g monofilament to assess sensation

Tick when felt within 3cm Cross when not felt (x) Shortening level (=)
Clawing (c) Draw wound or fissure Wasting (w)



Neuritis check

Sensory or strength change in past 6 months: Y □ N □

Nerve pain or tenderness: Y □ N □

If yes, give details: .....

.....

.....

.....

.....

Comments

.....

.....

.....

.....

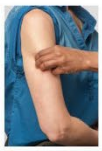
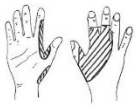
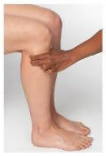
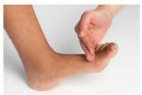
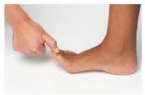
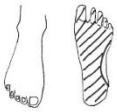
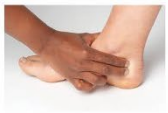
.....

.....

.....

.....

Appendix 4 Guide to performing a VMT-ST and palpating peripheral nerves

Peripheral nerve	Sensory distribution	Palpation	Motor testing
Supraorbital	Forehead, upper eyelid, anterior scalp	Run pulp of thumb just above eyebrow 1cm from inner edge	
Facial		Palpate below and in front of the ear lobe	Ask the patient to close the eyes lightly, then firmly. Measure any lid gap in mm
Trigeminal	Cornea		Examine blink frequency, if patient blinks less than twice per minute, suggestive of impaired corneal sensation
Greater auricular	Lower part of ear and surrounding skin		
Radial (above elbow)	Posterior forearm		Elbow extension
Ulnar			 Index finger abduction  Little finger abduction
Median			 Thumb opposition (up and across)  Thumb abduction
Radial (superficial cutaneous branch)			 Finger extension  Wrist extension
Common peroneal			 Foot dorsiflexion (Foot up)  Great toe dorsiflexion
Posterior tibial			 Fanning of toes

*NB the Sural nerve is composed of branches from the common peroneal and tibial nerves, and supplies sensation to the posterolateral aspect of the distal third of the leg and lateral border of the foot, heel and ankle. It has no motor supply.

Palpation of peripheral nerves.

Greater auricular nerve: With patient's head turned to one side, feel along upper third of opposite SCM muscle, the nerve is often visible if enlarged, running parallel to the external jugular vein

Radial nerve (above elbow): With elbow partly flexed and shoulder internally rotated, press pulp of fingers deep into radial groove at insertion of the deltoid muscle

Ulnar nerve: With patient's arm flexed to 90 degrees, feel in the olecranon groove just behind the medial epicondyle, palpate 10cm along the nerve toward the axilla if enlarged

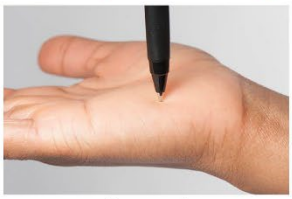
Median nerve: With palm facing upwards and hand slightly flexed, palpate at the wrist crease in line with the ring finger, medial to the palmaris longus tendon

Radial nerve (superficial cutaneous branch): With the hand held in slight flexion and forearm semi-prone, roll the nerve with 2-3 fingers over the lateral side of the radius near the wrist, just above the anatomical snuff box

Common peroneal nerve: With the patient sitting, palpate the nerve 2cm down and 1cm behind the fibular head

Posterior tibial nerve: With the patient sitting, palpate the nerve 2cm down and 2 cm behind the medial malleolus

Technique for using a pen to test sensation



Correct

Incorrect

Appendix 5 DN4 – Questionnaire

To estimate the probability of neuropathic pain, please answer yes or no for each item of the following four questions.

Interview of the patient	Yes	No
Question 1: Does the pain have one or more of the following characteristics?		
Burning		
Painful cold		
Electric shocks		
Question 2: Is the pain associated with one or more of the following symptoms in the same area?		
Tingling		
Pins and needles		
Numbness		
Itching		

Examination of the patient	Yes	No
Question 3: Is the pain located in an area where the physical examination may reveal one or more of the following characteristics?		
Hypoesthesia to touch		
Hypoesthesia to pinprick		
Question 4: In the painful area, can the pain be caused or increased by:		
Brushing		

Yes - 1 point
No - 0 points

Patient's score:___/ 10

Appendix 6 Laboratory reporting for *Mycobacterium leprae* drug resistance testing

Form 1 *Mycobacterium leprae* drug resistance testing: Clinical report form

1. **Reporting details**
    - Date of specimen collection(dd/mm/yy):
    - Date of report (dd/mm/yy):
    - Place of referral (region / state / country):
2. **Demographics and medical history of the case**
    - Date of birth (dd/mm/yy): Sex: M/F
    - Country of birth:
    - Chemoprophylaxis: Yes/No
        - If yes, which drug was given:
    - Previous TB treatment: Yes/No
    - Previous treatment with ofloxacin or other fluoroquinolones for longer than one month: Yes/No
3. **Case classification**
    - New
    - Retreatment
        - Retreatment after loss to follow up
        - Transferred in
        - Relapse
        - Other retreatment
4. **Treatment history (for retreatment cases only)**
    - MDT, year:
    - Rifampicin, Dapsone, Clofazimine (not in MDT pack), year:
    - Other, please specify with year:
5. **Clinical presentation at time of referral for resistance testing**
    - Clinical features:
    - Number of skin lesions
    - Skin smear results from specific sites
        - Site: BI: Date of smear:__/__/__
        - Site: BI: Date of smear:__/__/__
        - Site: BI: Date of smear:__/__/__
6. **Disease classification**
    - WHO: MB / PB
    - Ridley-Jopling: BT/BB/BL/LL
7. **Type of sample**
    - Biopsy (number of samples): Site of collection:
    - Skin smear (number of samples): Site of collection:

Form 2 Testing laboratory report

1. **Name of testing laboratory:**
2. **Case identification**
    - Case initials:
    - DOB (dd/mm/yy):
    - Sex: M/F
    - Place of referral (region / state / country or laboratory):
3. **Type of case**
   New □
   Retreatment □
4. **Specimen**
    - Date received (dd/mm/yy):
    - Type of specimen
        - Slit skin smear □ Biopsy: skin □ nerve □ Other:
5. **Sequencing results**
    - ***rpoB* gene**
        - Negative PCR □ (other PCR tested:)
        - No mutation (do not report silent mutation) □
        - Presence of a mutation known to confer drug resistance
            - Nucleotide mutation (X to Y at nucleotide Z*)□
            - Amino acid substitution (A to B at position C*) □
        - Presence of another mutation (A to B at position C*) □
    - ***folP1 gene***
        - Negative PCR □ (other PCR tested:)
        - Presence of a mutation known to confer drug resistance
            - Nucleotide mutation (X to Y at nucleotide Z*)□
            - Amino acid substitution (A to B at position C*) □
        - Presence of another mutation (X to Y at position Z*) □
    - ***gyrA gene***
        - Negative PCR □ (other PCR tested:)
        - Presence of a mutation known to confer drug resistance
            - Nucleotide mutation (X to Y at nucleotide Z*)□
            - Amino acid substitution (A to B at position C*) □
        - Presence of another mutation (X to Y at position Z*) □

*numbering system of *M.leprae* genome strain TN (NCBI accession number AL4550380.1)

Name of Scientist reporting: Date:

Signature:

Note:

The presence or absence of mutations known to confer drug resistance should be reported as:

- “no mutation known to confer resistance” if these mutations are not found and the sequence brings up wild-type codons;

- “presence of mutations known to confer resistance” if at least one of these mutations is observed. The substitution (e.g. Ser456Leu) should be stated on the basis of the numbering system of the *M.leprae* genome from the TN strain (see Table 1 and Figure 4 in *A guide for surveillance of antimicrobial resistance in leprosy: 2017 update)*

- “another missense mutation”: this could be a new mutation or a previously described mutation but not known to confer resistance. The mutation should be written with regard to the substitution (e.g. Lys411Asn), with the nucleotide change in brackets (e.g. aaa to aac) on the basis of the numbering system of the *M.leprae* genome from the TN strain. Corresponding DNA sequences and data concerning the description of new mutations will be also sent to a common database located at Ecole Polytechnique Federale de Lausanne;

- do not report silent mutations; ie. Mutations that do not change the amino acid.

Form 3 Reporting form for treatment outcomes of resistant cases

Reporting details

- UMRN:
- Initials / sex: M/F / age (in numbers)
- Type of case: New □ Retreatment □ ; Type of retreatment:
- Date of report (dd/mm/yy): __/__/__
- Date of treatment completion (dd/mm/yy): __/__/__

Type of resistance

- Rifampicin alone
- Dapsone alone
- Ofloxacin alone
- Rifampicin + dapsone
- Rifampicin + dapsone + ofloxacin
- Rifampicin + ofloxacin
- Dapsone + ofloxacin

Treatment prescribed by:

Dr: Name of facility:

Treatment regimen prescribed

- □ Option A: 400 mg ofloxacin + 100 mg minocycline + 50 mg clofazimine, daily for 6 months followed by 400 mg ofloxacin + 50 mg clofazimine for 18 months daily
- □ Option B: 400 mg ofloxacin + 100mg minocycline + 50 mg clofazimine, daily for 6 months followed by 100 mg of minocycline + 50 mg clofazimine daily for 18 months
- □ Other treatment (specify):

Treatment outcome

□ Treatment completed
□ Unsatisfactory response to treatment
□ Lost to follow up
□ Transferred out
□ Died

Appendix 7 ISF Score Summary Sheet

Variables		Outcome	Score	
			Right	Left
Eye	Blink	Yes	0	0
		No	1	1
	Vision	Normal >6/18	0	0
		Impaired <6/18	1	1
		Blind <3/60	2	2
	VMT	Lid gap in mm	__(mm)	__(mm)
Hand		Strong	0	0
	Little finger abduction	Resistance reduced	1	1
		Movement reduced	2	2
		Paralysed	3	3
	Thumb up & across	Strong	0	0
		Resistance reduced	1	1
		Movement reduced	2	2
		Paralysed	3	3
	Sensation	Number of points with sensory loss	0-10 __	0-10 __
	Wound count	Count number of wounds	__ wounds	__ wounds
	Bone resorption / loss	Number of sensory testing points lost due to digital resorption	0-10 __	0-10 __
UMRN:	**Name:**	**Date:**	**R=**	**L=**
			Total score (R + L):	

Appendix 8 K6 Scale

WA TUBERCULOSIS CONTROL PROGRAM KESSLER PSYCHOLOGICAL DISTRESS SCALE (K6)	AFFIX PATIENT IDENTIFICATION LABEL HERE UR NO: FAMILY NAME GIVEN NAME DOB SEX M F

The Kessler Psychological Distress Scale (K6)[1] involves 6 questions about a person's emotional state. Each question is scored from 1 (None of the time) to 5 (All of the time). Scores of the 6 questions are then summed, yielding a minimum score of 6 and a maximum score of 30. Low scores indicate low levels of psychological distress and high scores indicate high levels of psychological distress.

The following questions are about how you have been feeling during the past 30 days.	ALL	MOST	SOME	A LITTLE	NONE
1. About how often during the past 30 days did you feel **nervous** - would you say **all** of the time, **most** of the time, **some** of the time, **a little** of the time, or **none** of the time?	○	○	○	○	○
2. During the past 30 days, about how often did you feel **hopeless** - **all** of the time, **most** of the time, **some** of the time, **a little** of the time, or **none** of the time?	○	○	○	○	○
3. During the past 30 days, about how often did you feel **restless or fidgety**? *(If necessary prompt: **all, most, some, a little**, or **none** of the time?)*	○	○	○	○	○
4. How often did you feel **so depressed that nothing could cheer you up**? *(If necessary prompt: **all, most, some, a little**, or **none** of the time?)*	○	○	○	○	○
5. During the past 30 days, about how often did you feel that **everything was an effort**? *(If necessary prompt: **all, most, some, a little**, or **none** of the time?)*	○	○	○	○	○
6. During the past 30 days, about how often did you feel **worthless**? *(If necessary prompt: **all, most, some, a little**, or **none** of the time?)*	○	○	○	○	○

Total Score: _____ / **30**

K6 Dichotomous score groupings and categorisation[2]

Australian K6 Total Score Levels	Level of psychological distress
6-18	No probable serious mental illness
19-30	Probable serious mental illness

[1] Furukawa, T.A., Kessler, R.C., Slade, T. & Andrews, G. (2003) 'The performance of the K6 and K10 screening scales for psychological distress in the Australian National Survey of Mental Health and Well-Being', Psychological Medicine, 33, 357-362..

[2] Kessler, R.C., Green, J.G., Gruber, M.J., Sampson, N.A., Bromet, E., Cuitan, M., Furukawa, T.A., Gureje, O., Hinkov, H., Hu, C.-Y, Lara, C., Lee, S., Mneimneh, Z., Myer, L., Oakley-Browne, M., Posada-Villa, J., Sagar, R., Viana, M.C. & Zaslavsky, A.M. (2010) 'Screening for Serious Mental Illness in the General Population with the K6 screening scale: results from the WHO World Mental Health (WMH) survey initiative', International Journal of Methods in Psychiatric Research, Vol 19: 4-22.

Appendix 9(a) Leprosy clinical care template PB leprosy

	Month 1	Month 2	Month 3	Month 4	Month 5	Month 6
Bloods*						
DOT supervision of monthly Rifampicin						
VMT-ST**						
Skin examination						
Nerve palpation						
Ophthalmology review##						
WHO disability grading and EHF score (5.4.5)						
ISF 6 (Appendix 7)						
Specialist reviewϕ						

* Routine bloods: FBC, EUC, LFT. Additional investigations may be required in some circumstances.

** More frequent review required in setting of lepra reaction, generally 1-2 weekly depending on severity.

Formal ophthalmology review should be performed at time of diagnosis for patients with facial skin lesions or abnormal findings on eye examination (see 5.1.6)

ϕ Patients should be reviewed by a specialist at least three monthly. More frequent review required in the presence of lepra reaction or other complications.

Appendix 9(b) Leprosy clinical care template MB leprosy BI 1-3+

	Month 1	Month 2	Month 3	Month 4	Month 5	Month 6	Month 7	Month 8	Month 9	Month 10	Month 11	Month 12
Bloods*												
DOT supervision of monthly Rifampicin												
VMT-ST**												
Skin examination												
Nerve palpation												
Slit skin smears#												
Ophthalmology review##												
WHO disability grading and EHF score (5.4.5)												
ISF 6 (Appendix 7)												
Specialist reviewϕ												

* Routine bloods: FBC, EUC, LFT. Additional investigations may be required in some circumstances. If bloods remain within normal limits at six months, frequency can be reduced to three monthly.

** More frequent review required in setting of lepra reaction, generally 1-2 weekly depending on severity.

For patients with positive smears at time of diagnosis, repeat smears 12 monthly until completion of therapy. Smears should be taken from previously positive sites. More frequent or further investigation may be required in cases where treatment failure is suspected.

Formal ophthalmology review should be performed at time of diagnosis and annually while on treatment for all patients with MB leprosy

¥ 6 monthly for patients with acute neuritis, lepra reactions or nerve damage. Complete yearly for all other patients.

ϕ Patients should be reviewed by a specialist at least three monthly. More frequent review required in the presence of lepra reaction or other complications.

Appendix 9(c) Leprosy clinical care template MB leprosy BI 4-6+

	1	2	3	4	5	6	7	8	9	10	11	12	13	14	15	16	17	18	19	20	21	22	23	24
Bloods*																								
DOT supervision of monthly Rifampicin																								
VMT-ST**																								
Skin examination																								
Nerve palpation																								
Slit skin smears#																								
Ophthalmology review##																								
WHO disability grading and EHF score (5.4.5)																								
ISF 6 (Appendix 7)																								
Specialist review$^{\phi}$																								

* Routine bloods: FBC, EUC, LFT. Additional investigations may be required in some circumstances. If bloods remain within normal limits at six months, frequency can be reduced to three monthly.

** More frequent review required in setting of lepra reaction, generally 1-2 weekly depending on severity.

For patients with positive smears at time of diagnosis, repeat smears 12 monthly until completion of therapy. Smears should be taken from previously positive sites. More frequent or further investigation may be required in cases where treatment failure is suspected.

Formal ophthalmology review should be performed at time of diagnosis and annually while on treatment for all patients with MB leprosy and those with eye abnormalities

¥ 6 monthly for patients with acute neuritis, lepra reactions or nerve damage. Complete yearly for all other patients.

ϕ Patients should be reviewed by a specialist at least three monthly. More frequent review required in the presence of lepra reaction or other complications.

Appendix 10 Drugs used to treat leprosy

(Adapted with permission from Northern Territory Leprosy Guidelines)

Drug	Action	Presentation	Dose	Precaution	Adverse effects
Rifampicin	Potent bactericidal action Selective inhibition of DNA-dependent RNA polymerase	-150 mg and 300mg tablet -100mg/5ml liquid	10mg/ kg monthly Adult 600mg monthly	-Monitor LFTs -Drug interactions[1]	-Red discolouration of urine and tears -hepatitis -thrombocytopaenia -fever -flu like syndrome -rash -GI upset
Dapsone	Bacteriostatic & Weakly bactericidal Competitive inhibition of dihyro folate synthetase	50mg and 100mg tablet	1-2 mg/kg/ day Adult 100mg daily	-avoid if sulphur allergy, severe anaemia, G6PD deficiency, porphyria	-dapsone hypersensitivity syndrome (see 6.9) -haemolysis -fixed drug eruption -exfoliative dermatitis -GI irritation
Clofazimine	Mild bactericidal action Anti - inflammatory	50mg and 100 mg tablet	Children 1mg/kg/day 6mg/kg monthly Usual adult dose 50mg daily (up to 300mg daily for ENL[2])	Long half-life of 70 days Hepatic or renal impairment	-red/brown discoloration of skin (reversible) -dry skin, ichthyosis, pruritis -dry eyes -clofazimine induced enteropathy (doses of 300mg/d > 3 months): GI symptoms, sub-acute bowel obstruction -crystal deposition in liver and spleen
Ofloxacin	Bactericidal	200mg tablet	400mg monthly or daily (depending on regimen)	Avoid in pregnancy, children <18yrs Reduce dose in hepatic and renal impairment	-nausea, diarrhoea, dyspepsia -headache -rash, pruritis -dizziness -seizures (in epilepsy or in combination with NSAIDS)

[1] Increased doses of oral corticosteroids, oral contraceptives, oral anticoagulants and oral hypoglycaemics may be required, important drug interactions

[2] Doses of >100mg should not be continued for more than 3 months

Drug	Action	Presentation	Dose	Precaution	Adverse effects
Moxifloxacin	Bactericidal	400mg tablet	400mg monthly or daily (depending on regimen)	Avoid in pregnancy and children Severe hepatic impairment Patients with QTc prolongation, uncorrected hypokalaemia or patients receiving class IA or III anti-arrhythmic	-headache -dizziness -QTc prolongation -GI upset -raised transaminases -tendon inflammation and rupture -photosensitivity
Minocycline	Bacteriostatic	50mg tablet 100mg capsule	100mg monthly or daily (depending on regimen)	Avoid in tetracycline allergy, severe renal impairment, pregnancy, early childhood Monitor LFT	-photosensitivity -oesophagitis -abnormal bone development -tooth staining, enamel hypoplasia -morbilliform rash -urticaria, fixed drug eruption -cheilosis -glossitis
Clarithromycin	Bactericidal	250mg and 500mg tablets	500mg daily (adult)	Avoid in children <12 years, pregnancy & lactation QT prolongation	-pseudomembranous colitis -GI upset -headache, dizziness -hepatic dysfunction -photophobia

Appendix 11 Type 1 reaction severity scale

Walker SL, Nicholls PG, Butlin CR, Nery JAC, Roy HK, Rangel E, Sales AM, Lockwood DNJ. Development and validation of a severity scale for leprosy type 1 reactions. PLoS Negl Trop Dis. 2008;2(12):e351

	Criteria	0	1	2	3	Score
A1	Degree of inflammation of skin lesions	**None**	**Erythema**	**Erythema and raised**	**Ulceration**	
A2	Number of raised and/or inflamed lesions	**0**	**1-5**	**6-10**	**>10**	
A3	***Peripheral oedema due to reaction***	**None**	**Minimal**	**Visible, but not affecting function**	**Oedema affecting function**	
A SCORE						

	HANDS	**Purple 2g Monofilament scores**				**Orange 10g Monofilament scores**			**Score**
	Nerves	**0**	**0.5**	**1**	**1.5**	**2**	**2.5**	**3**	
B1	RIGHT Trigeminal	**Felt**						**Not felt**	
B2	LEFT Trigeminal	**Felt**						**Not felt**	
B3	***RIGHT ulnar***	**All sites felt**	**1 site not felt**	**2 sites not felt**	**3 sites not felt**	**1 site not felt**	**2 sites not felt**	**3 sites not felt**	
B4	***LEFT ulnar***	**All sites felt**	**1 site not felt**	**2 sites not felt**	**3 sites not felt**	**1 site not felt**	**2 sites not felt**	**3 sites not felt**	
B5	***RIGHT median***	**All sites felt**	**1 site not felt**	**2 sites not felt**	**3 sites not felt**	**1 site not felt**	**2 sites not felt**	**3 sites not felt**	
B6	***LEFT median***	**All sites felt**	**1 site not felt**	**2 sites not felt**	**3 sites not felt**	**1 site not felt**	**2 sites not felt**	**3 sites not felt**	
	FEET	**Orange 10g Monofilament scores**				**Pink 300g Monofilament scores**			**Score**
	Nerves	**0**	**0.5**	**1**	**1.5**	**2**	**2.5**	**3**	
B7	***RIGHT posterior tibial***	**All sites felt**	**1 site not felt**	**2 sites not felt**	**3 sites not felt**	**1 site not felt**	**2 sites not felt**	**3 sites not felt**	
B8	***LEFT posterior tibial***	**All sites felt**	**1 site not felt**	**2 sites not felt**	**3 sites not felt**	**1 site not felt**	**2 sites not felt**	**3 sites not felt**	
B SCORE									

	NERVE	0	1	2	3	Score
C1	RIGHT Facial	**MRC =5**	**MRC=4**	**MRC=3**	**MRC<3**	
C2	LEFT Facial	**MRC =5**	**MRC=4**	**MRC=3**	**MRC<3**	
C3	RIGHT Ulnar	**MRC =5**	**MRC=4**	**MRC=3**	**MRC<3**	
C4	LEFT Ulnar	**MRC =5**	**MRC=4**	**MRC=3**	**MRC<3**	
C5	RIGHT Median	**MRC =5**	**MRC=4**	**MRC=3**	**MRC<3**	
C6	LEFT Median	**MRC =5**	**MRC=4**	**MRC=3**	**MRC<3**	
C7	RIGHT Radial	**MRC =5**	**MRC=4**	**MRC=3**	**MRC<3**	
C8	LEFT Radial	**MRC =5**	**MRC=4**	**MRC=3**	**MRC<3**	
C9	RIGHT Lateral Popliteal	**MRC =5**	**MRC=4**	**MRC=3**	**MRC<3**	
C10	LEFT Lateral Popliteal	**MRC =5**	**MRC=4**	**MRC=3**	**MRC<3**	
C SCORE						

Total score	**Scores of A+B+C**	

Appendix 12 Enlist ENL Severity Scale

Walker SL, Sales AM, Butlin CR, et al, on behalf of the Erythema Nodosum Leprosum International Study Group. A leprosy clinical severity scale for erythema nodosum leprosum: An international, multicentre validation study of the ENLIST ENL Severity Scale. PLoS Negl Trop Dis.2017;11(7):e0005716. https://doi.org/10.1371/journal.pntd.0005716

Pain Rating - Visual Analogue Scale

(Ensure the line is 100 mm long)

How severe is your pain today? Mark the line below with an X to indicate how bad you feel your pain is today.

No pain _____ Worst possible pain

Item		Scores				Patient score
		0	1	2	3	
1	VAS - Pain (mm)	0	1-39	40-69	70-100	
2	Fever (in °C)	None (37.5 or less)	No fever now but history of fever in the last 7 days	37.6-38.5	38.6 or higher	
3	Number of ENL skin lesions	None	1-10	11-20	21 or more	
4	Inflammation of ENL skin lesions	Non-tender	Redness	Painful	Complex	
5	Extent of ENL skin lesions	0 regions	1-2 regions	3-4 regions	5-7 regions	
6	Peripheral oedema (hands or feet or face)	None	1 site	2 sites	All three sites	
7	Bone pain	None	Present on examination but does not limit activity	Sleep or activity disturbed	Incapacitating	
8	Inflammation of joints and/or digits due to ENL	None	Present on examination but does not limit activity	Sleep or activity disturbed	Incapacitating	
9	Lymphadenopathy	None	Enlarged	Pain or tenderness in 1 group	Pain or tenderness in 2 or more groups	
10	Nerve tenderness due to ENL	None	Absent if attention distracted	Present even if attention distracted	Patient withdraws limb on examination	

User Guide for the ENLIST ENL Severity Scale – final version which supersedes previously published version.

The score for each item should be **added** together to obtain the ENLIST ENL Severity Scale score.

Mild ENL is categorised as an ENLIST ENL Severity Scale score of **8 or less**.

The Minimal Important Difference of the ENLIST ENL Severity Scale is 5.

Scale Item	Notes
1. VAS Pain	Instruct the patient to point to the position on the line to indicate how much pain they are *currently* feeling. The far left end indicates 'No pain' and the far right end indicates 'Worst possible pain'. Take the measurement (in **mm**) using a ruler from the **LEFT** end of the line to the centre of the cross. **Ensure that the line when reproduced from this document is 100 mm long**.
2. Fever	Take temperature (in°C) using a thermometer. If the temperature is GREATER than 37.5 °C the patient has a fever. If it is less than or equal to 37.5 °C the patient scores 0 for this item UNLESS they give a history of having had a fever in the last 7 days in which case they score 1. The cause of the fever does not need to be established.
3. Number of ENL skin lesions	***Note: only skin lesions due to ENL are to be considered by this item***.
4. Inflammation of ENL skin lesions	***Note: only skin lesions due to ENL are to be considered by this item***. The term complex refers to the following type of skin lesions: vesicular, bullous, pustular, erythema multiforme-like, panniculitis, necrotic, ulcerated. ***If the participant fulfils criteria for more than one score then the highest scoring criteria should be used***. For example if there are red ENL skin lesions and some are ulcertated or vesicular or pustular then the patient scores 3 because "complex" lesions are present.
5. Extent to ENL skin lesions	***Note: only skin lesions due to ENL are to be considered by this item.*** *The separate regions are:* *a) Head and neck* *b) Left upper limb* *c) Right upper limb* *d) Torso – front (including genitals)* *e) Torso back (including buttocks)* *f) Left lower limb* *g) Right lower limb*
6. Peripheral oedema due to ENL	The three sites to be considered are the face, hands and feet. *Both feet count as one site. Both hands count as one site*. Oedema thought to be due to treatment such as corticosteroids or thalidomide should not be counted.

Appendix 13 DOT Log Sheet

Western Australian Tuberculosis Control Program

Directly Observed Therapy Log

Adverse drug reaction label

Chart No ……... of ……

DOT Month:	Case Manager:	Case Manager Phone:	REGION:
Patient details UMRN First Name Given Name DOB Sex: M / F	**DOT Start Date:**	**DOT Expected Completion Date:**	
	DOT Site: □ Home □ Work □ Clinic □ Pharmacy □ Other: _____ If Work/ Pharmacy/ Other Details: Name: Contact Details:		
Patient Contact Details Home: Work: Mobile:	**PPE Required?** □ Yes □ No Details: Location		

Day of Month	Time DOT observed	Dot observed signature / self tick		Comments (Please note any side effects) E.g. Abdominal discomfort rash/itch, visual disturbance, changes to appetite, fatigue.	Case Manager notified of adverse reaction?
1			□ Self		□Yes □No
2			□ Self		□Yes □No
3			□ Self		□Yes □No
4			□ Self		□Yes □No
5			□ Self		□Yes □No
6			□ Self		□Yes □No
7			□ Self		□Yes □No
8			□ Self		□Yes □No
9			□ Self		□Yes □No
10			□ Self		□Yes □No
11			□ Self		□Yes □No
12			□ Self		□Yes □No
13			□ Self		□Yes □No
14			□ Self		□Yes □No
15			□ Self		□Yes □No
16			□ Self		□Yes □No
17			□ Self		□Yes □No
18			□ Self		□Yes □No

Day of Month	Time DOT observed	Dot observed signature / self tick		Comments (Please note any side effects) E.g. Abdominal discomfort rash/itch, visual disturbance, changes to appetite, fatigue.	Case Manager notified of adverse reaction?
19			□ Self		□Yes □No
20			□ Self		□Yes □No
21			□ Self		□Yes □No
22			□ Self		□Yes □No
23			□ Self		□Yes □No
24			□ Self		□Yes □No
25			□ Self		□Yes □No
26			□ Self		□Yes □No
27			□ Self		□Yes □No
28			□ Self		□Yes □No
29			□ Self		□Yes □No
30			□ Self		□Yes □No
31			□ Self		□Yes □No

Appendix 14 Leprosy contact screening form for consideration of chemoprophylaxis

1) Index patient identification number: Date of diagnosis:

Consent for disclosure and household contact screening obtained: Y/N/NA

- *Not applicable when whole community screening undertaken where identity of index case will not be disclosed*

2) Contact information:

First name: Surname:

DOB: UMRN:

Gender: M/F/other BCG vaccination: Y / N / unsure

Relationship to index case:

Type of contact: Household / Community / Other

3) Refused screening: Y / N

4) Suspected leprosy: Y / N

If **Yes**, the contact is **ineligible** for chemoprophylaxis.

- Please refer to the Infectious Diseases Physician at WA Tuberculosis Control Program for review
  (Ph: 08 9222 8500)

5) Exclusion criteria for SDR: if any response answered "Y": contact is not eligible for SDR.
   - Age <2 years: Y / N
   - Pregnancy*: Y / N
   - Rifampicin use within past 2 years**: Y / N
   - Suspected TB***: Y / N
   - Liver or renal disease: Y / N
   - Refused SDR: Y / N
   - Other: please specify

6) If eligible for SDR: obtain informed consent and provide patient with information on possible adverse effects

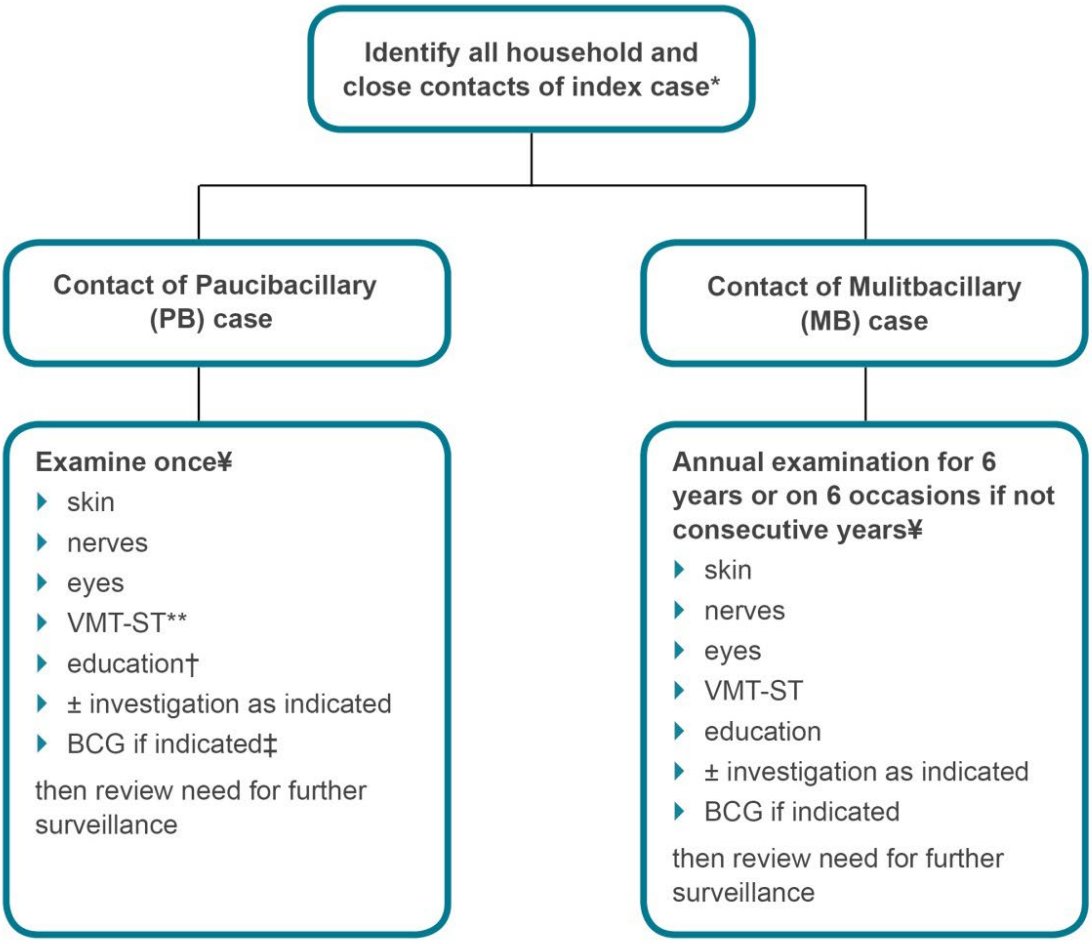
7) SDR dose:

Patient weight range	Rifampicin dose	Dose given
<20 kg	10-15mg/kg	
20-35 kg	450mg	
>35 kg	600mg	

Date SDR given: **Name:** **Signature:**

*SDR can be given after delivery
**e.g. for tuberculosis, leprosy or previous SDR for leprosy chemoprophylaxis
*** patients with any of the following symptoms require referral to Physician for exclusion of TB: cough present > 2weeks, night sweats, unexplained fever, unintentional weight loss.

Appendix 15 Guideline for leprosy contact tracing in Western Australia



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* Family, household contact, close social contacts (highest risk for children who are household contacts), includes index cases diagnosed post 1996 in regions where contact tracing has been incomplete. Community screen may be appropriate in some settings as guided by the Infectious Diseases Physician at WA TB Control Program

¥ At time of initial screen, assess eligibility for chemoprophylaxis (Appendix 12)

** Voluntary Motor and Sensory Testing

† Symptoms and signs of leprosy to look for in self and others, encourage early presentation

‡ Refer to WA BCG Policy

ww2.health.wa.gov.au/Articles/A_E/About-the-Western-Australia-Tuberculosis-Control-Program

Appendix 16 Notification form

Department of Health
GOVERNMENT OF WESTERN AUSTRALIA

Date received by DOH: [ ][ ]20

Notification ID: 20[ ][ ] – [ ][ ][ ][ ][ ]

Infectious and Related Diseases Notification Form

You may notify by post, telephone or fax
To: Communicable Disease Control Directorate,
PO Box 8172, Perth Business Centre WA 6849
Phone: (08) 9222 0255 or Fax: (08) 9222 0254
For urgent ☎ diseases after hours: Phone (08) 9328 0553

PATIENT DETAILS

Family name _____

Given name _____

Street address _____

Suburb/Town _____ **Postcode** _____

Tel. Home _____ **Mobile** _____

Date of birth ____/____/____ (dd mm yyyy)

Sex ☐ Male ☐ Female ☐ Transgender

Country of birth ☐ Australia ☐ Other, specify _____

Language spoken at home ☐ English ☐ Other, specify _____

Occupation or name of school/childcare centre attended:

_____

Is the patient of Aboriginal and/or Torres Strait Islander origin?

☐ No ☐ Yes, Aboriginal ☐ Yes, Torres Strait Islander

(For persons of both Aboriginal and Torres Strait Islander origin, tick both 'yes' boxes.)

DISEASE DETAILS

How was the infection identified?

☐ Clinical presentation ☐ Contact tracing ☐ Screening

Date of onset ____/____/____ (dd mm yyyy) **Date of death** (if applicable) ____/____/____ (dd mm yyyy)

Place infection acquired ☐ WA ☐ Interstate ☐ Overseas ☐ Unknown

If acquired interstate/overseas, specify _____

Was the patient hospitalised? ☐ No ☐ Yes

How was diagnosis made?

☐ Lab ☐ Result pending ☐ Linked to lab-confirmed case ☐ Clinical only

↳ Method: _____ Result: _____

FOLLOW-UP (tick one or more)

☐ Patient/carer aware of diagnosis and that it is a notifiable disease.
☐ Risk to contacts discussed with patient.
☐ Patient/carer aware Public Health Unit may contact them for information.
☐ Other _____

CLINICAL COMMENTS (presentation, treatment)

NOTIFIER DETAILS

Name ..... **Phone** .....

Clinic/Hospital .....

Address .....

..... **Postcode** .....

Signature ..... **Date** ...../...../..... (dd mm yyyy)

NOTIFIABLE DISEASES (tick box below) ☑

Pursuant to the WA ***Public Health Act 2016*** please notify diseases marked with a ☎ by telephone within 24 hours of diagnosis. Otherwise fax or post notification within 72 hours of diagnosis.

Multi-resistant organisms (MRSA, CRE, VRE) are notified by laboratories. Notification by doctors or nurse practitioners is not necessary.

- ☐ Acute post-streptococcal glomerulonephritis (APSGN)
- ☐ Adverse event following immunisation – **use separate form**
- ☎ ☐ Amoebic meningoencephalitis
- ☎ ☐ Anthrax
- ☐ Barmah Forest virus infection
- ☎ ☐ Botulism
- ☐ Brucellosis
- ☐ *Campylobacter* infection Species: _____
- ☐ Chancroid
- ☐ Chikungunya virus infection
- ☐ Chlamydia ☐ Lymphogranuloma venereum (serovar L1-3 detected)
- ☎ ☐ Cholera
- ☐ Creutzfeldt-Jakob disease (classical or variant)
- ☐ Cryptosporidiosis
- ☐ Dengue virus infection
- ☎ ☐ Diphtheria
- ☐ Donovanosis
- ☎ ☐ Flavivirus infection ☐ JE ☐ MVE ☐ West Nile/Kunjin ☐ Yellow fever ☐ Zika ☐ Other
- ☎ ☐ Food or water-borne gastroenteritis (≥2 linked cases)
- ☐ Gonococcal infection
- ☐ Haemolytic uraemic syndrome (HUS)
- ☎ ☐ *Haemophilus influenzae* type b (Hib) infection (invasive)
- ☎ ☐ Hendra virus infection
- ☎ ☐ Hepatitis A
- ☐ Hepatitis B ☐ newly acquired (<2 yrs) ☐ Chronic/unspecified
- ☐ Hepatitis C ☐ newly acquired (<2 yrs) ☐ Chronic/unspecified
- ☐ Hepatitis (other) ☐ D ☐ E
- ☐ HIV infection – **use separate form**
- ☐ Influenza
- ☎ ☐ Legionellosis ☐ Longbeachae ☐ Pneumophila ☐ Other
- ☐ Leprosy
- ☐ Leptospirosis
- ☎ ☐ Listeriosis
- ☎ ☐ Lyssavirus infection ☐ Rabies ☐ ABL ☐ Other _____
- ☐ Malaria Species: _____
- ☎ ☐ Measles
- ☐ Melioidosis
- ☎ ☐ Meningococcal infection ☐ Meningitis ☐ Septicaemia ☐ Other
- ☎ ☐ Middle East Respiratory Syndrome coronavirus (MERS-CoV)
- ☐ Mumps
- ☎ ☐ Paratyphoid fever
- ☐ Pertussis
- ☎ ☐ Plague
- ☐ Pneumococcal infection (invasive)
- ☎ ☐ Poliovirus infection
- ☐ Psittacosis (ornithosis)
- ☐ Q Fever
- ☐ Rheumatic fever/heart disease – **use separate form**
- ☐ Rickettsial infection Species: _____
- ☐ Ross River virus infection
- ☐ Rotavirus infection
- ☎ ☐ Rubella ☐ Non-congenital ☐ Congenital
- ☐ *Salmonella* infection Species: _____
- ☎ ☐ Severe Acute Respiratory Syndrome (SARS)
- ☎ ☐ Shiga toxin-producing *E.coli* (STEC) infection
- ☐ Shigellosis Species: _____
- ☎ ☐ Smallpox
- ☐ Syphilis ☐ 1° ☐ 2° ☐ Early latent (<2yrs) ☐ Late latent ☐ 3° ☐ Congenital
- ☎ ☐ Tetanus
- ☐ Tuberculosis
- ☎ ☐ Tularaemia
- ☎ ☐ Typhoid fever
- ☐ Varicella-zoster virus ☐ Chickenpox ☐ Shingles ☐ Unspecified
- ☐ *Vibrio parahaemolyticus* infection
- ☎ ☐ Viral haemorrhagic fever (Crimean-Congo, Ebola, Lassa, Marburg)
- ☐ *Yersinia* infection

CDC-002430 SEP17 Metro

☐ Tick this box if you require more forms and pre-paid envelopes (or print from ww2.health.wa.gov.au/Articles/N_R/Notification-of-infectious-diseases-and-related-conditions). Sept 2017

Policy Sponsor	Dr Justin Waring, Medical Director WA Tuberculosis Control Program				
Policy Contact	Dr Alison Keed, Consultant Infectious Diseases				
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Approved by:	WA Tuberculosis and Leprosy Advisory Council PH Document Review Committee			Date:	20/04/2023
Endorsed by:	Director Public Health			Date:	20/04/2023
NSQHS Standards Applicable:	x Std 1: Clinical Governance ☐ Std 2: Partnering with Consumers ☐ Std 3: Preventing and Controlling Healthcare Associated Infection x Std 4: Medication Safety		X Std 5: Comprehensive Care x Std 6: Communicating for Safety ☐ Std 7: Blood Management ☐ Std 8: Recognising and Responding to Acute Deterioration		
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The health impact upon Aboriginal people have been considered, and where relevant incorporated and appropriately addressed in the development of this health initiative (2206).

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