Leprosy

Background

Leprosy is also known as Hansen's Disease, Bungarun Disease or Big Sickness

Think of leprosy in unexplained skin lesions or nerve problems

Epidemiology

In Australia, leprosy is rare. Most cases are in people born overseas or in Aboriginal people.

In Western Australia:

- Half of cases occur in remote or very remote places
- Notification rates in the Kimberley Aboriginal population is significantly higher than the overall WA rate
- New cases continue to be diagnosed in the Kimberley

Leprosy in the Kimberley

It is believed that leprosy was introduced into the Kimberley in the late 1800s through European settlement and pearling activity. Aboriginal families in the Kimberley were disproportionately affected by the number of infections, with large case numbers by the 1930s. From 1936 to 1986, a purpose-built leprosarium (known as 'Bungarun') operated outside of Derby. Mandatory isolation continued long after effective treatment became available. Leprosy caused significant social disruption among generations of Kimberley Aboriginal families.

It is important to recognise this history in raising conversations about diagnosis and screening for leprosy. Some people may remember and find solidarity with family history and connections; others may fear diagnosis or treatment. There may be a belief that leprosy disappeared when Bungarun closed, so

Refer to Kimberley Regional Physician Team for further assessment

diagnosis or requirements for screening may cause shock. All discussions need to be undertaken in a sensitive and confidential manner.

Leprosy (*Mycobacterium leprae)*

A chronic granulomatous disease affecting skin and peripheral nerves caused by infection with *Mycobacterium leprae*.

- Leprosy has a wide spectrum of disease expression.
- Classification: **paucibacillary** (2-5 lesions) or **multibacillary** (≥6 lesions)
- Transmission: not well understood, but is **not** highly contagious
- Incubation period: highly variable, but can be very long (2-30 years)
- Treatment: with three drugs for between 12 and 24 months
- Sequelae: can include deformity, disability, stigma and psychological distress
- Complications: include acute immunological reactions called lepra reactions

Diagnosis

Think of leprosy if there are any of the following:

- Non-healing skin lesions
- Neurological symptoms (sensory or motor)
- A personal or family history of leprosy

Common presenting symptoms and signs include:

- Skin lesions: pale (hypo-pigmented), coppery, or reddish (erythematous) patch
- Shiny thickened skin on the face (lion/leonine facies)
- 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 cardinal signs of leprosy are:

1. Skin lesions: hypopigmented (pale), coppery or red skin lesions with reduced or absent sensation

Early diagnosis and treatment of leprosy can prevent permanent disability

2. Nerve thickening: thickened peripheral nerves, with or without loss of sensation and muscle weakness
3. Demonstration of *M. leprae*: by slit skin smear (SSS), biopsy or PCR

Delays in diagnosis are common due to a lack of expertise and awareness of leprosy.

For image of characteristic skin lesions see page 18 of the Guidelines for the control of Leprosy in the Northern Territory (2018).

Assessment

When considering a diagnosis of leprosy:

Take a history, including asking;

- Have you noticed any skin lesions?
- Have you felt numb or tingly in your face, hands or feet?
- Have your hands or feet felt dry?
- Have you been weak in your hands or feet?
- Has it been hard to close your eyes?
- Have you had any burning or shooting pains?

Tread gently and explore the language preferred by the patient before asking about personal or family history. Some people might know of family who stayed at Bungarun, but not that the person was treated for leprosy.

Kimberley Aboriginal Health Planning Forum

- Have you been treated for leprosy/Hansen's disease/Bungarun disease before?
- Do you know if anyone in your family has had leprosy/Hansen's disease/Bungarun disease?

Carefully examine the skin and peripheral nerves;
Record all findings in the Template (Appendix 1)

- Examine whole body for skin lesions:
  Lesions can be single or multiple, well or poorly defined, macular or papular, nodular, infiltrative, hypopigmented, coppery or red, with reduced sensation, dry with loss of sweating/hair.
- Palpate nerves for thickening:
  Ulnar nerve at the elbow
  Median nerve at the wrist
  Facial nerve near the ear
  Common peroneal nerve at the knee
  Posterior tibial nerve at the ankle
  (See Box 1 for method for nerve palpation).
- Assess sensory and motor nerve function:
  The combination of sensory testing and voluntary muscle testing used for standardized leprosy assessments is called VMT-ST (see Appendix 2 for a guide).
- Eye examination:
  Inspect eye, eyebrows, eyelashes, blink, surrounding skin check for gap between lids when asked to close eyes. Perform visual acuity.

Exclude other causes for signs and symptoms;

- Perform skin scrapings of skin lesions suspicious for fungal infection and treat with 6 weeks of a topical antifungal
- Investigate for other causes of peripheral neuropathy

If skin signs or symptoms not explained by other cause or nerve or eye signs or symptoms present; Arrange investigations as advised by Kimberley Regional Physician Team (call on-call physician or KPHU for further advice);

- Slit skin smear for AFB microscopy
  - Usually taken from 3+ sites
  - The physician team can advise you on how to perform this procedure
- Skin biopsy (ideally a 1cm full thickness elliptical biopsy or a 4mm punch) with samples placed in:
  - Formalin: Fite-Faraco stain, histology, fungal stains
  - Fresh sample (nothing added): fungal MC+S
  - Saline: *M. leprae* PCR
- Other: nerve biopsy (by neurology/neurosurgery)

Principles of Management

Case Management Model

In the Kimberley, there is a case management model for the management of leprosy to optimise drug therapy, early detection of lepra reactions, prevention of disability and provision of social support.

All leprosy cases are managed by an interagency team including the

- Primary care team
- Kimberley Regional Physician Team,
- Kimberley Population Health Unit, and
- Anita Clayton Centre (Perth)

Drug Treatment

A three-drug antibiotic regimen is used, with duration determined by disease classification, often between 6 months – 2 years. Common drugs used include dapsone, clofazimine, rifampicin, fluoroquinolones and minocycline. Sometimes prednisolone is needed to suppress immune reactions, so leprosy patients can be more vulnerable to infections (immunocompromised). All treatment is managed by Infectious Diseases and Regional Physician Teams.

Drug treatment is free under the WA department of health.

Disease Notification

Leprosy is a notifiable disease and on diagnosis should be notified via the Notification Form and faxed to KPHU (9194 1631).

Prevention

Contact Tracing

Contact tracing identifies close contacts (which may include household, extended family and community contacts) to facilitate early diagnosis of secondary cases, surveillance of contacts and provision of chemoprophylaxis if indicated.

KPHU will facilitate contact tracing with local health professionals. Note that consent from the index case should be sought before proceeding with contact tracing. The identity of the index case must not be disclosed unless informed consent has been provided to do so.

If community wide screening is indicated consent to undertake contact tracing from the index case is not required. However, the identity of the case must not be disclosed under any circumstance.

Chemoprophylaxis

Chemoprophylaxis using single-dose rifampicin for contacts should be administered with guidance from KPHU under the direction of the Anita Clayton Centre. This should be offered to all household contacts of a newly diagnosed case of leprosy, following screening for signs of leprosy and exclusion of contraindications.

BCG Vaccination

BCG vaccination should not be offered routinely to Australian residents. However, it is indicated in particular settings including:

- Newborn children of parents with Leprosy or a family history of Leprosy, and
- Children <6 years old who have not previously been vaccinated and are household contacts of newly diagnosed leprosy.

Leprosy

To arrange BCG vaccination, refer babies of parents with Leprosy or a family history of Leprosy to KPHU.
Enquiries for BCG vaccination for children <6 years of age undertaking extended overseas travel to TB endemic countries can be directed to the Anita Clayton Centre via the Enquiry Form.

Follow Up

Case Monitoring

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.

The local case manager supported by KPHU is responsible for ensuring recommended follow up is conducted. Involvement of Aboriginal Health Workers / Aboriginal Health Practitioners can assist in providing culturally appropriate case management

Establish recalls for:

- Monthly doctor review: VMT-ST should be performed monthly.
- Three-monthly specialist (physician) review.
- Blood tests: FBP, UEC and LFT should be performed monthly for the first 3 months and if normal, then 3 monthly.

Contact Screening

See Box 2 for what is involved in screening of contacts.

- Contacts of multibacillary cases require annual review for at least 6 years
- Contacts of paucibacillary cases require a single review, with no follow up necessary in the absence of suspicious findings

Ophthalmology

Ophthalmology review should be arranged for anyone with multibacillary leprosy, abnormal findings on eye examination, or other eye concerns.

Allied Health

Refer to podiatry, occupational therapy, physiotherapy as required due to deformity or disability.

Refer / Discuss

Kimberley Population Health Unit

Ph: (08) 9194 1630
Fax: (08) 9194 1631
Email: WACHSKimberleyCommunicableDiseaseControl@health.wa.gov.au

Kimberley Regional Physician Team

Ph: via Broome Hospital switch (08) 9194 2222
Email: krpt@health.wa.gov.au

Anita Clayton Centre

Ph: (08) 9222 8500 (infectious disease specialist)

Resources / References

WA Health: Guidelines for the Diagnosis, Management and Prevention of Leprosy

Boxes

Box 1: Method for Nerve Palpation

- Position patient and examiner correctly, ideally seated comfortably
- Enquire as to whether any of the nerves are tender
- Locate the nerve
- Use the pulp of two or three fingers to roll the nerve gently against the bone (do not use the tips of the fingers)
- Observe the patient's face during examination to detect tenderness. Feel along the nerve as far as possible in both directions
- Compare features of nerve with opposite side
- Assess for: thickness, consistency, tenderness, irregularity, number of nerves involved, symmetry
- Document in template (Appendix 1). Enlargement is scored as *None (N)*, *Possible (P)*, *Definite (D)*.

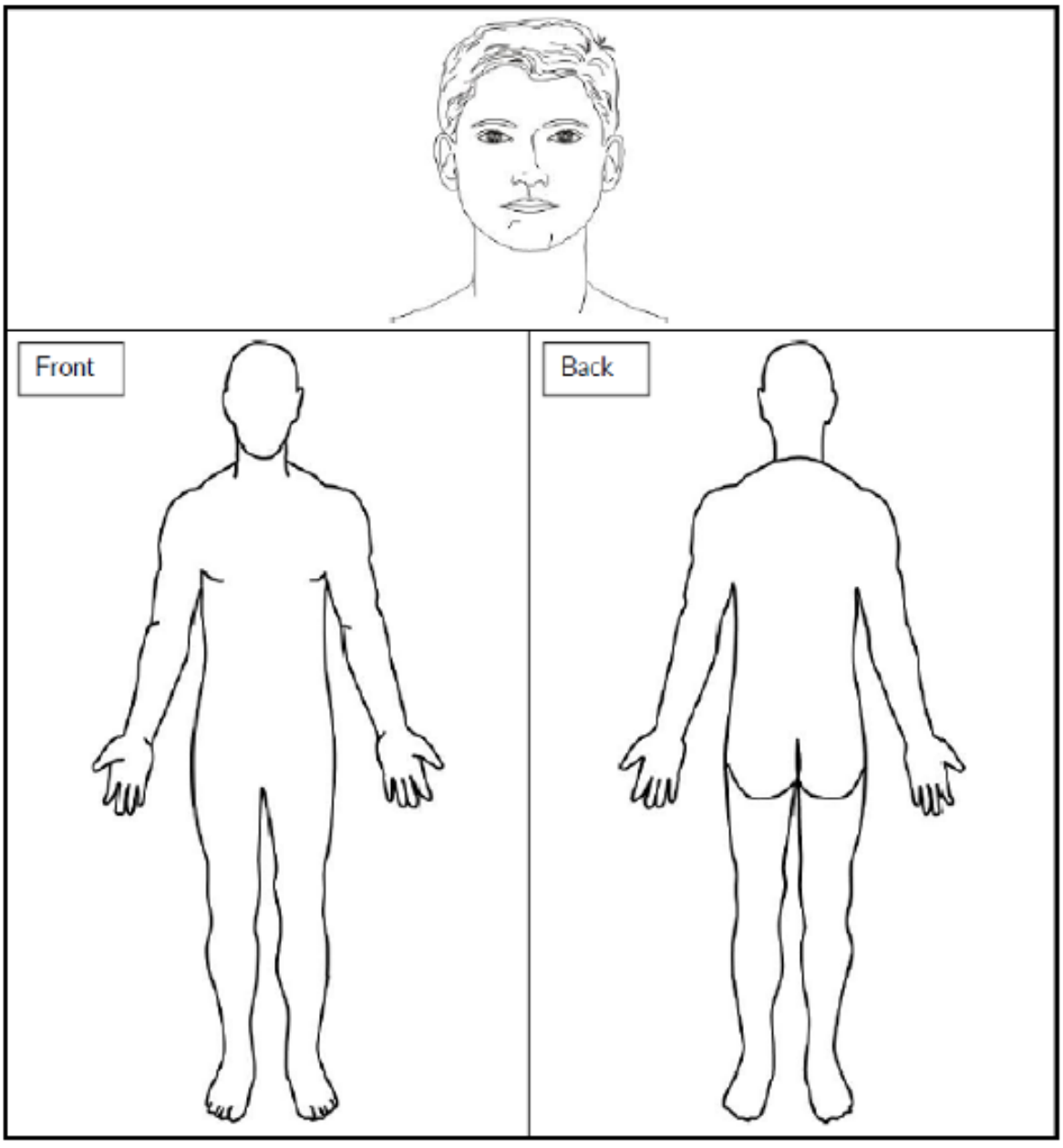
Box 2: Contact Screening

- Interview: symptoms of leprosy, history of BCG vaccination, previous TB exposure or treatment, co-existing medical conditions
- Examination: skin and peripheral nerves – complete VMT-ST (see Appendices 1 & 2)
- Education: signs and symptoms of leprosy and the importance of early presentation
- Refer: refer to physicians if clinical features of leprosy are present for investigations, and call KPHU to notify
- Vaccinate: refer to KPHU to arrange vaccination for those eligible
- Chemoprophylaxis: follow advice of KPHU to determine eligibility for single dose rifampicin (if no contraindications)
- Follow up: arrange annual review for contacts of multibacillary leprosy

Appendix 1 – Template for Leprosy 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.



Leprosy

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

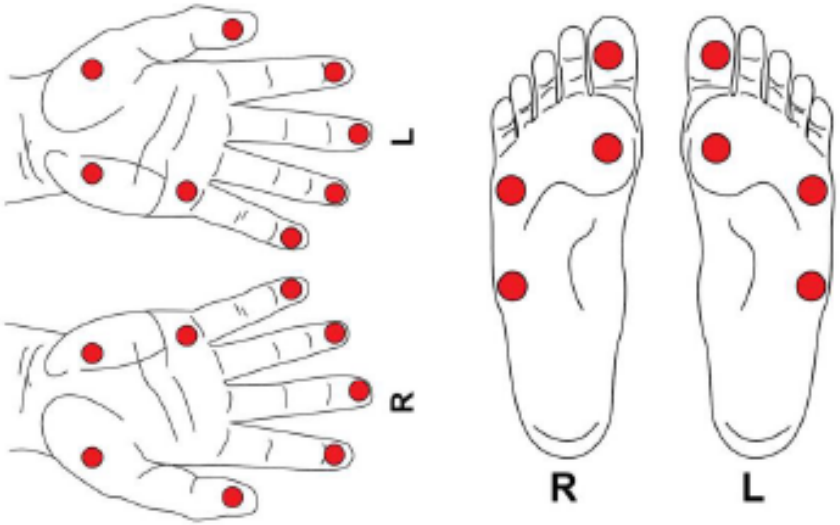
S=strong W=weak P=paralysed

	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

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From WA Health Guidelines for the Diagnosis, Management and Prevention of Leprosy

Appendix 2 – 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.

From WA Health Guidelines for the Diagnosis, Management and Prevention of Leprosy