LYMPHOEDEMA

RACGP Accepted clinical resource

Guide for diagnosis and management in general practice

WHAT IS LYMPHOEDEMA?

A chronic swelling of one or more regions of the body caused by the accumulation of protein rich fluid in the tissue spaces. It occurs when the demand for lymphatic drainage exceeds the capacity of the lymphatic circulation. There are two types: primary arising from developmental compromise of the lymphatic system; and secondary due to damage to the lymphatic vessels and/or lymph nodes[1].

TYPES OF LYMPHOEDEMA

Primary

- caused by abnormal development of the lymphatic system
- swelling may be present from birth, or develop in adolescence or middle age
- some may be hereditary

In primary lymphoedema, often no triggering factor can be identified.

Consider primary lymphoedema as a differential diagnosis in patients with unexplained symptoms of limb swelling/morbidity especially if unilateral.

Secondary

- occurs when there is damage to the lymphatic system, lymphatic decompensation or failure
- acquired following surgery, radiotherapy, trauma or other damage to the lymphatic system

Secondary lymphoedema may develop many months or even years after damage to the lymphatic system.

Causes

- surgery:
  - cancer related
  - orthopaedic
  - venous stripping
  - skin grafting
- DVT / venous disease
- radiotherapy
- trauma/burns
- infection:
  - cellulitis
  - filariasis
- prolonged dependency oedema:
  - immobility
  - chronic neurological disorders
- obesity related
- can occur in combination with lipoedema

Lymphoedema can occur in any part of the body e.g. limbs, head and neck, breast and genital area.

A qualified lymphoedema practitioner can help with this.

RISK FACTORS FOR DEVELOPING LYMPHOEDEMA[2, 3, 4]

- any surgery (not just cancer surgery) where there is damage to the lymphatic system/nodes
- radiotherapy to the lymphatic system
- cellulitis or history of infection in the affected region
- injury or trauma to the lymphatic system
- immobility including post CVA/wheelchair bound
- obesity
- longterm chronic venous insufficiency
- family history

EARLY WARNING SIGNS AND SYMPTOMS OF LYMPHOEDEMA[2]

- transient swelling
- feelings of heaviness in the affected region
- pain or tension in the affected region
- tightness and a 'bursting' feeling in the affected region
- clothing or jewellery becoming tighter

Early warning signs can be present for three years or more prior to the development of swelling.

Intervention at this stage can have a significant impact on reducing the risk of developing lymphoedema and the severity of lymphoedema if it does develop.

For those at risk of lymphoedema, bioimpedance can detect changes early. Bioimpedance is available from some lymphoedema practitioners.

LIPOEDEMA

Lipoedema is a different condition to lymphoedema. It is caused by abnormal deposition of subcutaneous adipose tissue

- occurs almost exclusively in women
- usually bilateral and generally does not involve the feet
- swelling is mostly due to fat, not fluid
- adipose tissue can be painful and sensitive
- primarily occurs from the waist down, but can also affect the arms
- body shape is disproportionate, differentiating from obesity
- tendency to bruise
- patients can have a chronic oedema develop due to their lipoedema
  - oedema develops due to overloading of the functional capacity of the normal lymphatic system
  - frequently occurs with co-existing obesity and venous insufficiency

Lipoedema can be misdiagnosed as lymphoedema.

HealthPathways Melbourne
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An Australian Government Initiative
Lymphoedema Association Australia
Australasian Lymphology Association

PHYSICAL EVALUATION

- assess subcutaneous tissue
  - pitting/non-pitting oedema
  - tissue tone
- check for the presence of Stemmer's sign
  - thickened skin at the base of the 2nd toe indicates lymphoedema
- assess skin condition
  - dryness or cracking
  - infection
  - bruising
  - check for interdigital tinea
  - check nails for paronychia and in-grown toe nails
- assess presence and severity of swelling by measuring circumference of affected limb(s) compared with unaffected limbs or pre-operative measurements using a tape measure
- measurement forms can be downloaded from **www.lymphoedema.org.au**
- weight and height/BMI/waist to hip ratio, waist to height ratio
- cardiac and respiratory parameters
- examination for presence of masses - abdominal/pelvic, check for lymphadenopathy
- assess arterial circulation (e.g. ABPI)

INVESTIGATIONS

- duplex scan to exclude venous insufficiency/DVT
- pathology tests if clinically relevant
  - FBC
  - U&E&CR (EUC)
  - TFT
  - LFT
  - ESR/CRP
  - HbA1c if BMI > 35
  - Urinary protein/ ACR
- CT scan or Ultrasound to exclude masses/tumours
- chest x-ray (echocardiogram if appropriate) to exclude cardiac/respiratory causes of oedema
- bioimpedance, especially if they have had initial measure prior to surgery in context of cancer treatment
- lymphoscintigraphy if diagnosis is still unclear (often occurs in primary lymphoedema). Exclude all other causes first
- MR lymphangiography where available provides more info
- ICG lymphography or fluoroscopy (not available in all states)

GENERAL MANAGEMENT PRINCIPLES

Management principles for lymphoedema are primarily based on clinical consensus[1, 2]

Management essentials

- effective management can reduce symptom severity and improve quality of life
- minimising the risk of cellulitis is essential to reduce the risk of developing or exacerbating lymphoedema
- acknowledging patient concerns and challenges of living with lymphoedema is important and should include practical and emotional aspects
- regular and appropriate exercise depending on patient's general ability must be prescribed

Comprehensive Lymphoedema Management

Management is multimodal and may include the following:

- education on limb/body part care including skin care to maintain a protective barrier against infection
- physical exercises to improve lymphatic flow
- compression individually prescribed and fitted by a qualified lymphoedema practitioner e.g. compression bandage, garment, wrap, pump **(N.B. not every patient will require compression bandaging/garment)**
- Manual Lymphatic Drainage (MLD), or intermittent pneumatic compression pump, to improve lymphatic flow

Other treatment could include:

- photobiomodulation (formerly low level laser therapy)
- kinesiotaping
- negative pressure therapy

Precautions

- it is important to note that diuretics are ineffective in lymphoedema alone (however, they can still be important to use in the context of heart failure)
- some medications may exacerbate the condition (e.g. calcium channel blockers, steroids, pregabalin, anti-inflammatory agents)
- patients with existing lymphoedema who experience an unexplained exacerbation should be assessed for tumour recurrence or DVT and referred as appropriate

SPECIFIC MANAGEMENT PRINCIPLES

Cellulitis

- people with lymphoedema are more susceptible to recurrent episodes of cellulitis (usually Streptococcus)
- cellulitis risk can be reduced by good skin care and compression garment use
- urgent antibiotic treatment is essential to control the spread of infection (e.g. phenoxymethylpenicillin 500mg orally q6h or clindamycin 450 mg orally q8h for patients allergic to penicillin for minimum 14 days)[5]
- in cases of frequent recurrence, consider continuous prophylaxis (e.g. phenoxymethylpenicillin 250 mg orally bid for 6 months initially)[6]
- when Staphylococcus suspected (e.g. folliculitis/pus) use flucloxacillin 500mg q6h
- advise elevation of the affected limb/body part and the use of compression garment can continue as tolerated
- lower limb cellulitis is usually unilateral
- bilateral leg redness, consider lipodermatosclerosis or 'Red Leg Syndrome'[7]
- refer to Australasian Lymphology Association (ALA) Consensus Guideline: Management of Cellulitis in Lymphoedema: **www.lymphoedema.org.au/about-lymphoedema/consensus-guideline/**

Skin care

- daily skin care with non-perfumed/soap free washes and moisturisers is essential for skin to act as a barrier to infection
- avoid sunburn
- tight jewellery or clothing which constricts the affected limb or body part should be avoided

Foot care

- check for interdigital tinea or infections
- feet should be cleaned and dried daily
- treat any infection/injury promptly
- referral to podiatry may be required

Exercise and Movement

- exercise is a cornerstone of management
- combinations of flexibility, resistance and aerobic exercise may be beneficial in controlling lymphoedema[2]

Weight control

- weight management is important as excess body weight may impair lymphatic function

Travel

- consider providing a script for prophylactic antibiotics for patients when travelling
- encourage consultation with a Lymphoedema Practitioner for individualised advice regarding compression garments and travel advice

Heat

- patients should avoid outdoor activities in the hottest part of the day
- saunas and hot water may exacerbate swelling
- patients could consider financial support from Energy Cooling Rebate Schemes

Clinical procedures

- where possible, use non-affected arm/area of the body for injections, IV cannulas, BP readings and other clinical procedures
- avoid immunisations in the affected limb
- take care when excising skin lesions and using liquid nitrogen in the lymphoedematous area

Surgical Treatments

- surgical options are still emerging and may include, liposuction, lymphovenous anastomosis and lymph node transfer
- compliance with conservative measures is essential prior to surgical referral

ACCESSING TREATMENT

There are various ways to access lymphoedema treatment, depending on your state or territory.

Lymphoedema Practitioners

- best practice recommends people with lymphoedema are referred for treatment to a trained Lymphoedema Practitioner - a therapist that has completed an ALA endorsed training course
- Lymphoedema Practitioners can be accessed through private or public services depending on where you live
- visit the National Lymphoedema Practitioners Register **www.lymphoedema.org.au**
- private treatment can be subsidised via the Department of Health Chronic Disease Management Plan 5 sessions, if the Lymphoedema Practitioner is an approved allied health service provider

Financial Assistance for persons with lymphoedema

- Lymphoedema Compression Garment Subsidy Schemes are available in each state, and all vary in eligibility criteria
- visit this website to find out the correct information for your state: **www.lymphaustralia.org.au/resources/find-a-service/**
- financial assistance via Energy Cooling Rebate Schemes may be available for people with lymphoedema, as they require cooling to prevent the severe worsening of their condition.

Australian Lymphology Association (ALA)

- ALA is the peak professional organisation promoting best practice in lymphoedema management, research and education in Australasia.
- visit the National Lymphoedema Practitioners Register **www.lymphoedema.org.au**

Lymphoedema Association Australia (LAA)

- LAA aims to ensure people living with lymphoedema get the information, treatment and support they need to live well
- P: 1300-852-850 **www.lymphaustralia.org.au**

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References

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5 HealthPathways Melbourne: Lymphoedema. Document 177312. Jan 2023
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DIAGNOSIS AND ASSESSMENT OF CHRONIC OEDEMA

Presenting symptoms in limb/body part

- persistent swelling – may be permanent or transient

Additional symptoms:

- heaviness
- tightness
- fullness or pain

Take a medical history including:

- cancer
- trauma to the limb
- past cellulitis/infection/ulcers
- travel
- family history of limb swelling

Perform full physical examination including:

- skin condition
- pitting
- Stemmer's sign
- limb circumferences

HISTORY OF CANCER

Details of:

- surgery
- lymph node removal
- chemotherapy
- radiotherapy
- other surgery
- complications (e.g. post-operative infection)

Consider:

- tumour recurrence
- DVT
- cellulitis

ABSENT

Secondary Cancer Related Lymphoedema

Is there a recent exacerbation?

PRESENT

Investigate appropriately and refer back to patient's specialist

NO HISTORY OF CANCER

Exclude other causes of oedema:

- cardiac, renal or hepatic failure
- mass (pelvic, abdominal, lymphadenopathy)
- thyroid disease
- medication side-effects
- venous insufficiency (including past DVT, chronic ulcers)
- immobility and chronic neurological disorders
- obesity
- other surgery

Investigations:

- FBE
- EUC
- LFT
- TFT
- ESR
- CRP
- HbA1c if BMI >35
- Urinary protein/ACR

If lower limb affected:

- CXR +/- echocardiogram
- abdo U/S or CT scan
- venous duplex scan

Consider lymphoscintigraphy if diagnosis is still unclear.

Consider differential diagnosis: LYMPHOEDEMA

(Primary, Secondary non-cancer, Vaso-lymphoedema or mixed)

Refer to lymphoedema practitioner or clinic for comprehensive assessment and management

For your nearest specialist practitioner or clinic go to:
National Lymphoedema Practitioners Register **www.lymphoedema.org.au**
Support for your patient can be provided by the Lymphoedema Association Australia. www.lymphaustralia.org.au