LYMPHOEDEMA

Guide for diagnosis and management in general practice

WHAT IS LYMPHOEDEMA?

Chronic swelling of a body part (usually limbs) caused by the accumulation of fluid and protein in the tissue spaces arising from congenital malformation of the lymphatic system, or damage to lymphatic vessels and/or lymph nodes¹.

TYPES OF LYMPHOEDEMA

Primary

- caused by abnormal development of the lymphatic system
- swelling may be present from birth ('congenital'), or develop in adolescence ('lymphoedema praecox') or middle age ('lymphoedema tarda')
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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

- acquired following surgery, radiotherapy, trauma or other damage to the lymphatic system
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Secondary lymphoedema can develop many months or even years after surgery or radiotherapy

Mixed

Mixed lymphoedema describes lymphatic decompensation or failure associated with:

- venous disease
- lipoedema
- obesity
- immobility
- chronic neurological disorders
- skin grafting
- vein stripping or harvesting
- arthroscopy

Information is adapted with permission from Cancer Australia.

We would like to acknowledge the support of Dr Yvonne Zwar and Dr Susan Hodson for their assistance with this document.

RISK FACTORS FOR DEVELOPING LYMPHOEDEMA^{2,3,4}

- any surgery (not just cancer surgery) where there is damage to the lymphatic system
- radiotherapy to the lymphatic system
- history of infection in the affected limb/body part
- injury or trauma to the lymphatic system
- immobility
- obesity
- filariasis
- genetic predisposition

EARLY WARNING SIGNS OF LYMPHOEDEMA²

- transient swelling following exercise or physical activity
- feelings of heaviness in the affected limb or body part
- pain or tension in the affected limb or body part
- tightness and fullness (a 'bursting' feeling) in the limb or body part
- 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

LIPOEDEMA

- lipoedema, which is caused by abnormal deposition of subcutaneous adipose tissue, can be misdiagnosed as lymphoedema
- with lipoedema:
 - → swelling is bilateral and generally does not involve the feet which distinguishes it from lymphoedema
 - → there is pain on indent pressure and a tendency to bruise
 - → it is almost exclusively restricted to women
 - → patients can have mixed lipoedema/lymphoedema oedema develops due to overloading of the functional capacity of the normal lymphatic system







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PHYSICAL EVALUATION

- 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
- assess skin condition
 - → dry
 - → cracked
 - → infection
 - → bruising
 - → check for interdigital tinea
 - → check nails for paronychia and in-grown toe nails
- 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
- weight and height/BMI
- cardiac and respiratory parameters
- examination for presence of masses abdominal/pelvic, check for lymphadenopathy
- assess arterial circulation ABPI

INITIAL INVESTIGATIONS

- pathology tests if clinically relevant
 - → FBC
 - → U&E&CR
 - → TFT
 - → LFT
 - → ESR
 - → GTT if BMI > 35
- chest X-ray
 - → to exclude cardiac/respiratory causes of oedema
- CT scan or Ultrasound scan
 - → to exclude masses/tumours
- Duplex Scan
 - → to exclude venous insufficiency/DVT

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

Complex Lymphoedema Therapy (CLT)

- Complex Lymphoedema Therapy (CLT) is the most effective management option^{1,2}, and may include one or more of the following:
 - → education on care of the limb/body part including skin care to maintain a protective barrier against infection
 - → physical exercises designed to improve lymphatic flow
 - → Manual Lymphatic Drainage (MLD) (a specific type of massage) to improve lymphatic flow
 - → compression bandaging/garment individually fitted by a lymphoedema practitioner (N.B. not every patient will require compression bandaging/garment)

Cautions

- it is important to note that diuretics are ineffective in lympheodema, while other medications may exacerbate the condition (e.g. anti-hypertensive agents, steroids, HRT, anti-inflammatory agents)
- patients with existing lymphoedema who experience an exacerbation should be assessed for tumour recurrence or DVT and referred as appropriate

SPECIFIC MANAGEMENT PRINCIPLES

Cellulitis

- people with lymphoedema are prone to recurrent episodes of cellulitis
- advise bed rest and elevation of the affected limb/body part during cellulitis episode
- use of compression garment can continue if comfortable and tolerated
- urgent antibiotic treatment is essential to control the spread of infection (e.g. dicloxacillin/flucloxacillin 500mg orally q6h for 7-10 days or clindamycin 450 mg orally q8h for patients allergic to penicillin)
- in cases of frequent recurrence, consider continuous prophylaxis (e.g. phenoxymethylpenicillin 250 mg orally bid for 6 months initially)⁵
- cellulitis risk can be reduced by good skin care and compression garment use to minimise swelling
- refer to Australasian Lymphology Association (ALA)
 Consensus Guideline: Management of Cellulitis in
 Lymphoedema: www.lymphoedema.org.au/index.cfm//about-lymphoedema/consensus-guideline/

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Skin care

- good skin care is essential as healthy skin acts as a barrier to infection
- tight jewellery or clothing which constricts the affected limb or body part should be avoided

Foot care

- feet should be cleaned and dried daily
- treat any infection/injury promptly
- podiatry may be required

Clinical procedures

- use non-affected arm/area of the body for injections, IV cannulas, BP readings and other clinical procedures when possible. Avoid immunisations in the affected limb.
- take care when excising skin lesions and using liquid nitrogen in the lymphoedematous area
- exercise therapy 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 slow lymphatic flow

Overheating

- advise patients that hot baths, spas, and saunas may exacerbate swelling
- patients should avoid strenuous activities (e.g. sport, gardening) in the hottest part of the day

Travel

 if patients are planning air travel or a long-haul road or train trip (e.g. longer than 4 hours), they should seek advice from a lymphoedema practitioner before travelling

Lymphoedema Compression Garment Program (LCGP)

- subsidised garments are provided if receiving a Centrelink pension or assessed as being a low or medium income earner in Victoria
- LCGP is funded by the Department of Health and Human Services (DHHS) and administered by the State-wide Equipment Program (SWEP)
- application forms are available from https://swep.bhs.org.au/lymphoedema-compressiongarment-program.php
- the initial LCGP eligibility form must be certified by a medical practitioner.
- further assessment and garment prescription is undertaken by the lymphoedema practitioner
- P: 1300-747-937 / (03) 5333-8100 Fax: (03) 5333-8111
 E: swep@bhs.org.au

Australasian Lymphology Association (ALA)

- ALA is the peak professional organisation promoting best practice in lymphoedema management, research and education in Australasia.
- P: 1300-935-332 (Toll free in Australia)W: www.lymphoedema.org.au

Lymphoedema Association of Victoria (LAV)

- LAV provides information, education and support for people with lymphoedema
- contact LAV for lists of lymphoedema practitioners/clinics in your local area P: 1300-852-850 W: www.lav.org.au



References

- International Society of Lymphology *The diagnosis and treatment of peripheral lymphedema*. 2016 revision of the Consensus document of the International Society of Lymphology. Lymphology, 49(2016):170-184.
- ² Lymphoedema Framework (2006). Best Practice for the Management of Lymphoedema. International consensus. London: MEP Ltd.
- Queensland Health Lymphoedema Clinical Practice Guideline 2014 (found on Queensland Health site)
- 4 NSW Agency for Clinical Innovation: Key principles of care for people living with, or at risk of lymphoedema Amended August 2017
- ⁵ Therapeutic Guidelines Ltd: Antibiotic (eTG July 2018 edition) Skin and Soft tissue infections: bacterial.

DIAGNOSIS AND ASSESSMENT OF CHRONIC OEDEMA

Presenting symptoms in limb/body part (one or more of the following, AND of chronic duration, i.e. > 3 months) swelling heaviness pain or tension tightness and fullness HISTORY OF CANCER NO HISTORY OF CANCER **Details of: Exclude other causes of oedema:** surgery mass (pelvic, abdominal, lymphadenopathy) lymph node removal cardiac, renal or hepatic failure radiotherapy thyroid disease other surgery medication side-effects complications (e.g. post-operative infection, etc) venous insufficiency (including past DVT, chronic ulcers) chronic neurological disorders obesity immobility other surgery Consider: tumour recurrence In medical history, exclude the above and ask cellulitis about: trauma to limb past cellulitis, infection, ulcers travel history **ABSENT PRESENT** family history of limb swelling Secondary Perform full physical examination Investigate appropriately Lymphoedema and refer back to patient's Also assess degree of pitting, skin condition, presence of specialist Stemmer's sign and record limb circumferences Investigations Is there a recent FBE, U&E&CR, TFT, LFT, CXR, ESR, GTT if BMI >35 exacerbation? Legs: ultrasound or CT scan (abdomen, pelvis), venous duplex scan

(either primary, secondary or mixed)

Consider lymphoedema as differential diagnosis

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**N.B. patients should be advised that some practitioners and clinics have waiting lists