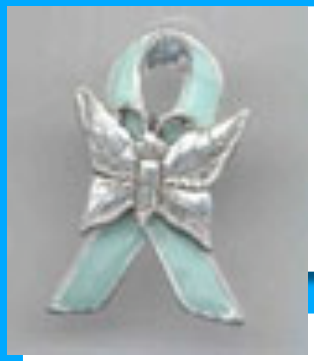


When Is It Lymphedema And What Can Be Done For It?

Colin Kanar, MD

Ellen Poage-Hooper, ARNP, CLT-LANA

Overview

- Definition
- Common Causes
- Problems with diagnosis
  - How?
  - When?
  - What should we do?

Secondary LE

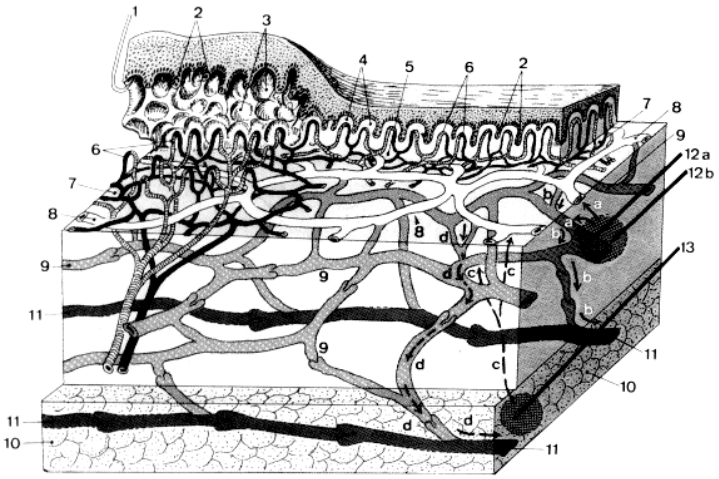
- LE results from the disruption or obstruction of the lymphatic pathways
- Reduced lymphatic flow with increased pressure in remaining lymphatic channels
- Accumulation of protein-rich fluid in the interstitium which causes chronic inflammation and reactive fibrosis of the affected tissues - mechanisms are not fully understood
- ~ 90% cases of LE in U.S. are due to tumors or as a consequence of treatment

Lymphatic System
Anatomy

Components:

- Lymph vessels
- Lymph nodes
- Spleen
- Thymus gland
- Tonsils
- Peyer's patches
- Lymphocytes

Lymphatic System
Anatomy

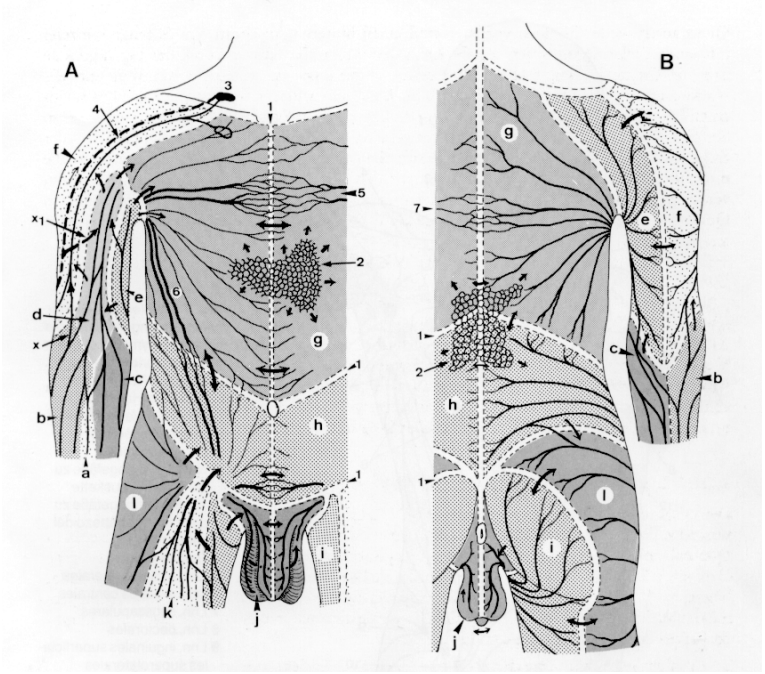


Lymph vessels:

- Initial Lymphatics
  - Capillaries
  - Pre-collectors
- Transport Vessels
  - Collectors
  - Trunks

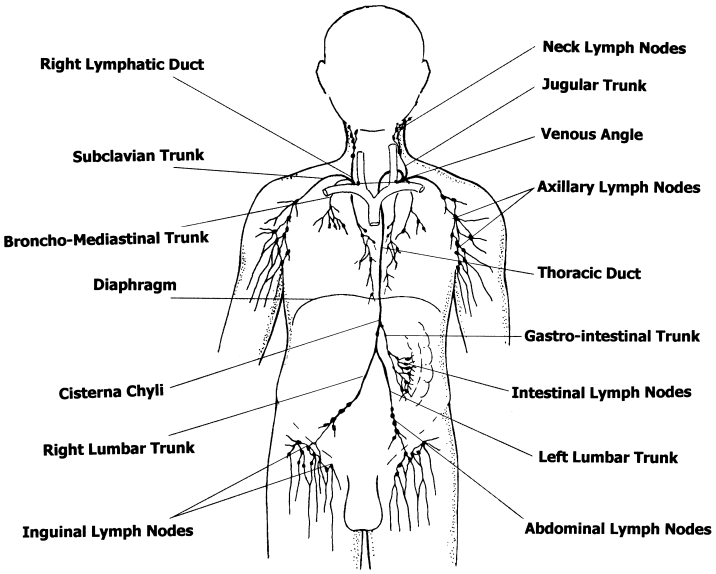
Lymphatic System

Anatomy –Superficial Lymph Collectors

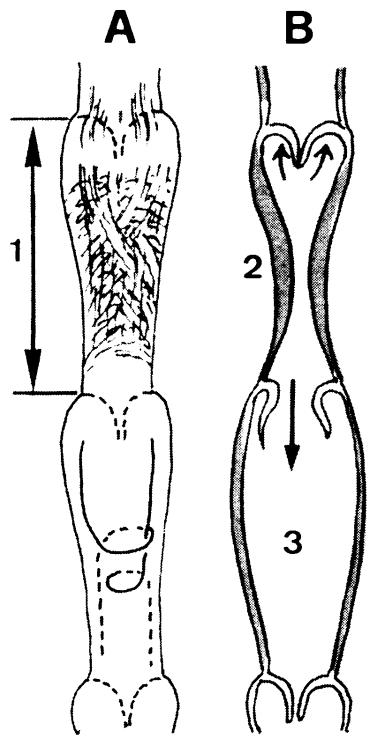


Lymphatic System

Anatomy - Lymph trunks



Lymphatic System
Physiology



Lymph vessels:

- Lymphangion

  (muscle tissue, valves)

- Intrinsic contractions (6-10 x/ min.)

- 2 - 2.5 l/day

Lymphatic System
Physiology

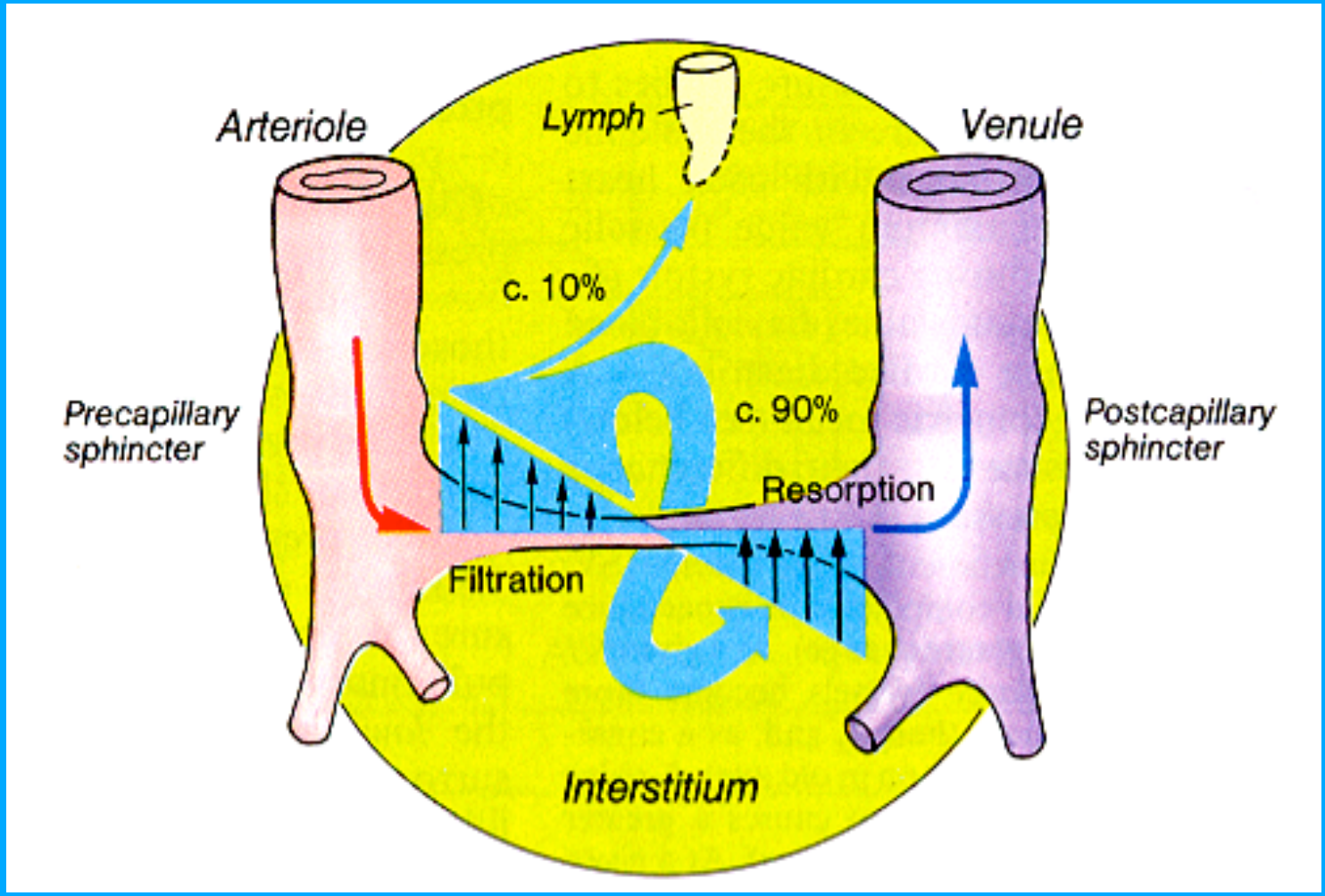
Lymph vessels/nodes:

- Return protein to circulatory system
- Remove excess fluid from interstitium
- Intestinal lymph vessels transport fat (Chyle)
- Immunological functions

Lymph sufficiency:

Transport capacity (TC) > Lymph (obligatory) load (LL)

Lymphatic System
Physiology - Microcirculation



Lymphatic System
Pathophysiology

Factors in edema formation:

+ Increased capillary hydrostatic pressure
+ Decreased plasma proteins (Hypoproteinemia)
+ Increased capillary permeability
+ Blockage of lymphatic return (Lymphedema)

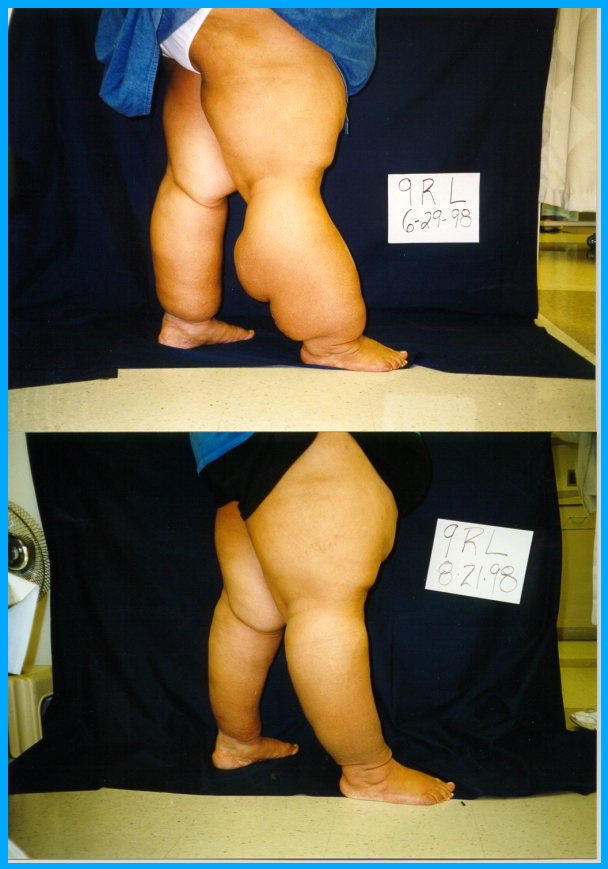
Lymph Insufficiency

+ Transport capacity (TC)< lymph (obligatory) load

Lymphedema
Characteristics

- Slow onset, progressive
- Rapid onset
- Pitting (early stages)
- Puffiness
- When it starts distally
  - Squaring of toes, Stemmer's sign positive
  - Dorsum of foot or hand
  - Loss of ankle contour
  - Asymmetric (if bilateral)
- Cellulitis is common

Lymphedema
Characteristics cont.



- Rarely painful
- Discomfort is common
  - Heaviness, achiness, fullness
- Skin changes
  - Hyperkeratosis, Papillomas, Peau d'orange
- Ulcerations unusual
- In contrast with venous disease the skin maintains hydration and elasticity for longer in the disease process

Stages of Lymphedema

Stage 0: *Latency* Transport capacity reduced	Stage I: *Spontaneously Reversible*	Stage II: *Spontaneously Irreversible*	Stage III: *Lymphostatic Elephantiasis*
Tissue: • No visible/ palpable edema	• Skin may pit or hold indentation after activity or pressure. • Reversible	• Spongy • Non-pitting • Early signs of hardening.	• Irreversible swelling • Hard, fibrotic
Limb size: ▪ Subjective complaints possible -	• Normal or close to normal.	• Increased in size	• Very large limb

Lymphedema
Anatomical Locations

- Extemities
  - Arm, Leg (unilateral, bilateral)
  - Trunk proximal to the limb
- Head and Neck
- Intestinal
- Genital

- Affects people at any age!

Lymphedema
Diagnostic Tests

- Physical Exam and History
- Lymphoscintigraphy

Lymphoscintigraphy

Lymph nodes missing in second photo on right

Lymphedema
Differential Diagnosis

- Lymphedema
- Lipedema
- Chronic Venous Insufficiency
- Acute Deep Venous Thrombophlebitis
- Cardiac Edema/Congestive Heart Failure
- Malignancy
- Myxedema
- CRPS (RSD)

Common causes

- Malignant tumors
- Trauma
- Surgery
- Radiation

Malignant Tumors

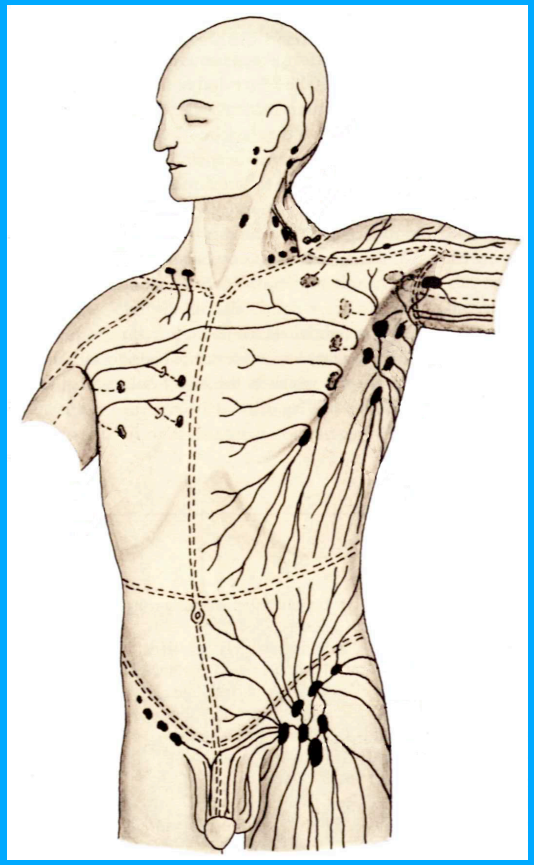
- Tumors that develop within a limb causing obstruction of lymphatic flow
- Sarcomas

Trauma

- Direct injury causing disruption of the lymphatic channels

Surgery

- Surgical disruption of lymphatic system
  - Vascular surgery
  - Orthopedic procedures
- Tumors that spread to lymph nodes
  - Breast cancer
  - Melanoma
  - Genitourinary (GU)
  - Gynecologic (GYN)
  - Head and Neck cancers

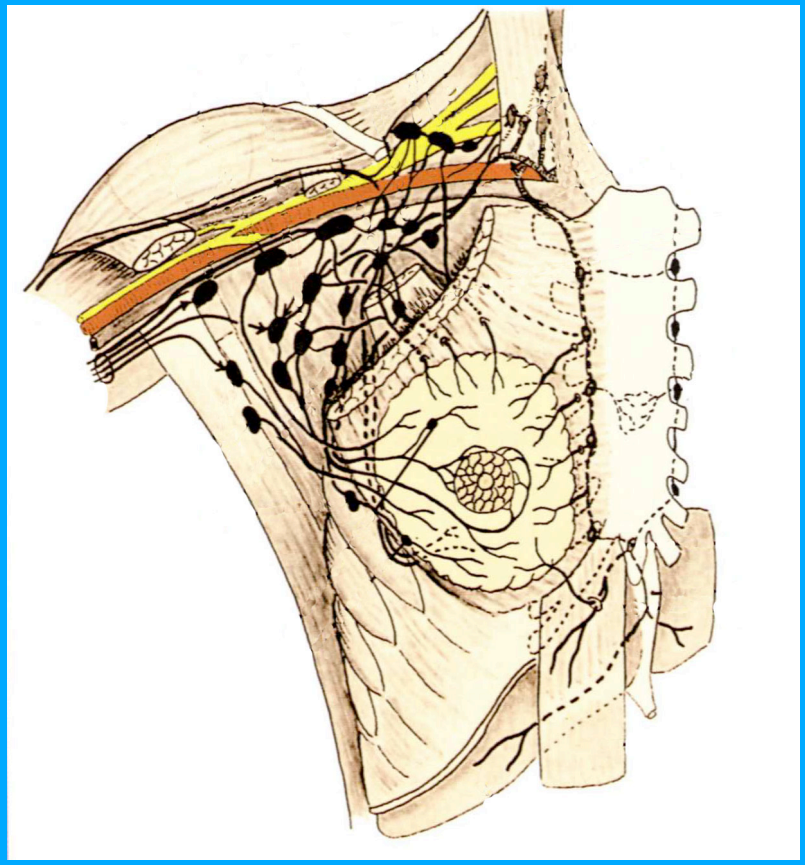


Upper Extremity

Axillary metastasis

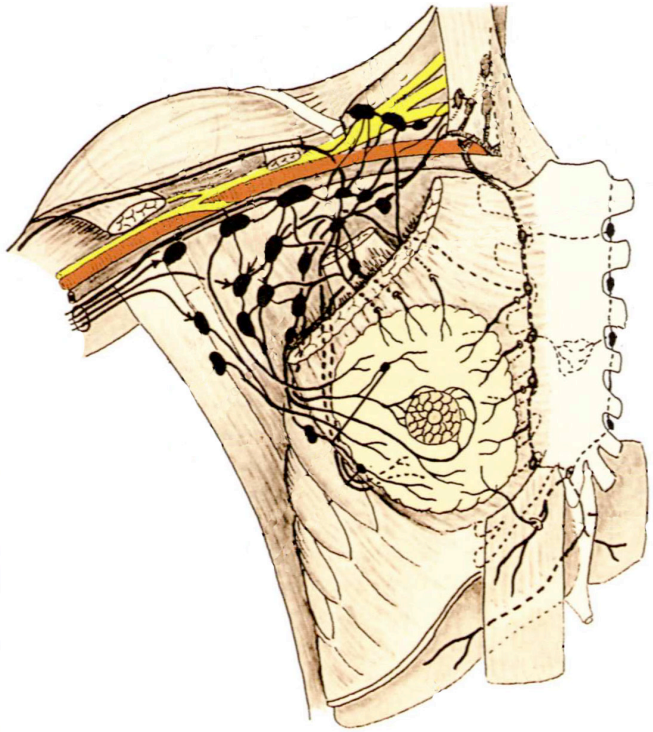
+ Breast cancer

+ Melanoma



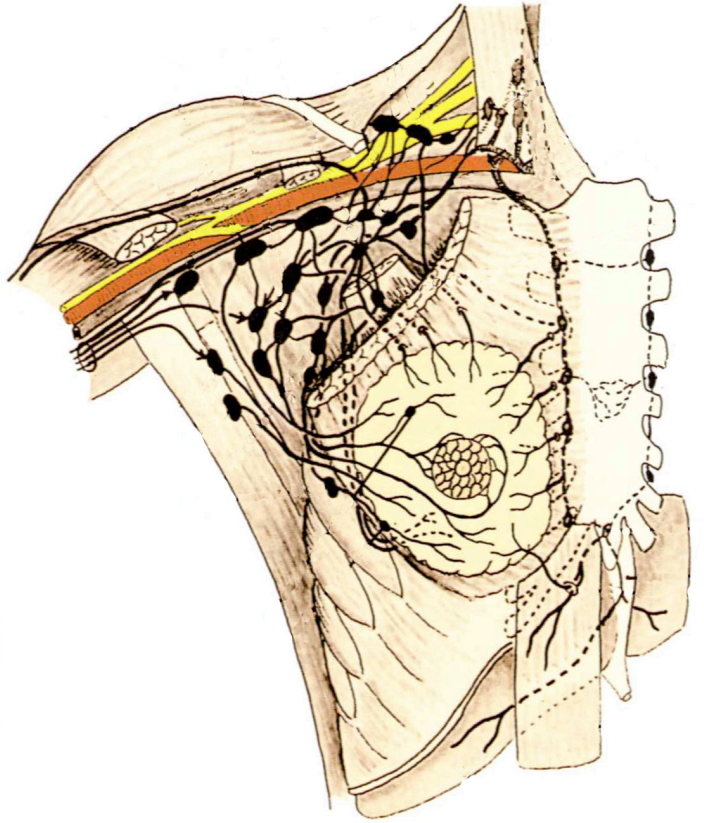
Breast Cancer

- Removal of level I / II lymph nodes
- Incidence LE ~20-30% in arms
  - Most by 3 years
  - 10% by two years
- Influencing factors:
  - Socio-demographic Variables (Ages, Income, etc) Did not influence
  - Body Mass Index
    - Correlates with everything measured
    - Influences development (>30 BMI= 4 times more likely), arm size and symptoms

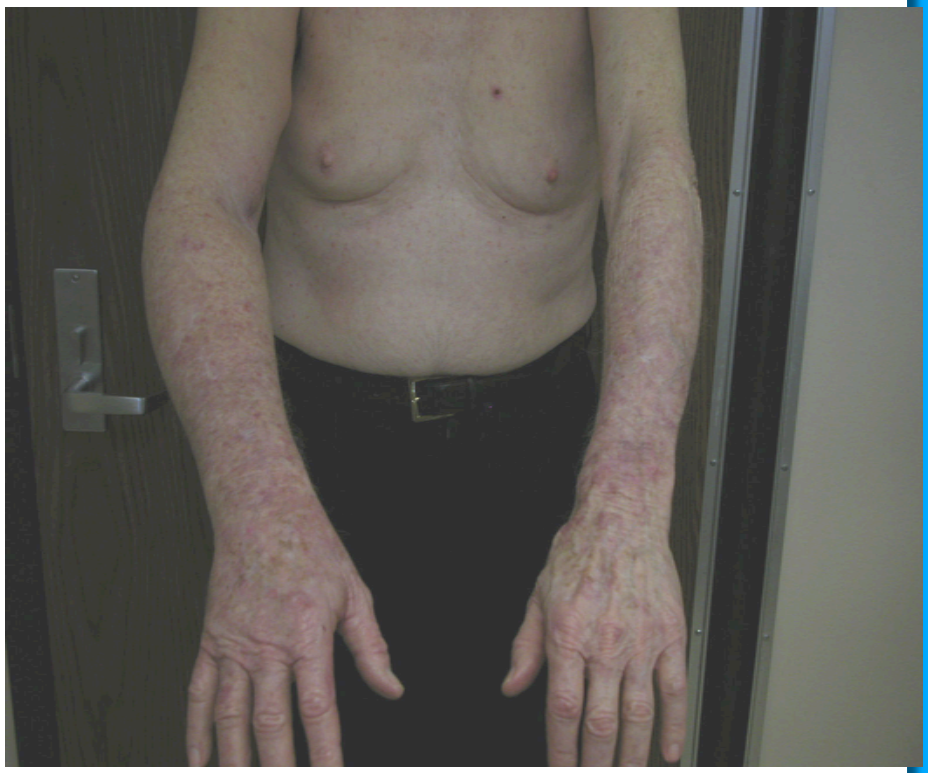


Melanoma

- Primary sites located on extremities
- More extensive in lymph node dissections



Melanoma



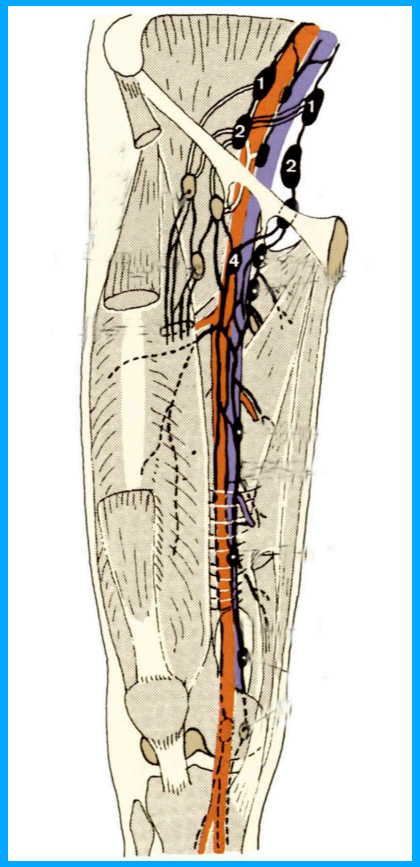
Lower Extremity LE

- Melanoma (40 - 75%)
- Gynecological cancers (~ 50%)
- Genitourinary cancers
  - Prostate cancer (1 – 66%)
  - Penile cancer (14 – 57%)
  - Bladder cancer (10 – 23%)
- Soft tissue
  - Sarcoma (12 – 20%)

Lower Extremity

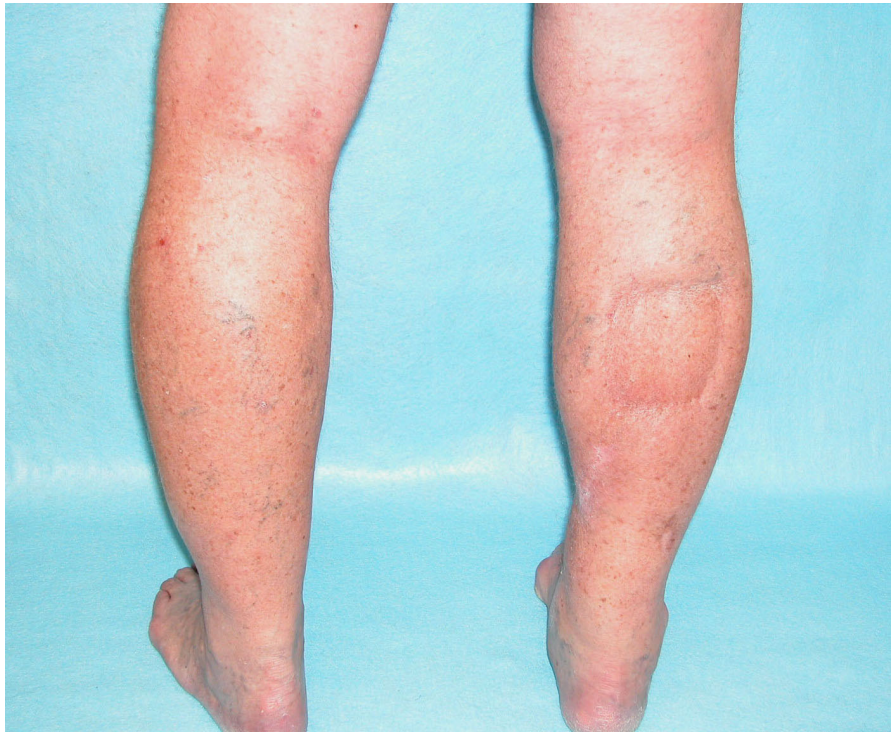
Groin Metastasis

- Melanoma
- GU malignancies
- Gyn malignancies
- GI malignancies



Melanoma

Lower Extremity



Head and Neck

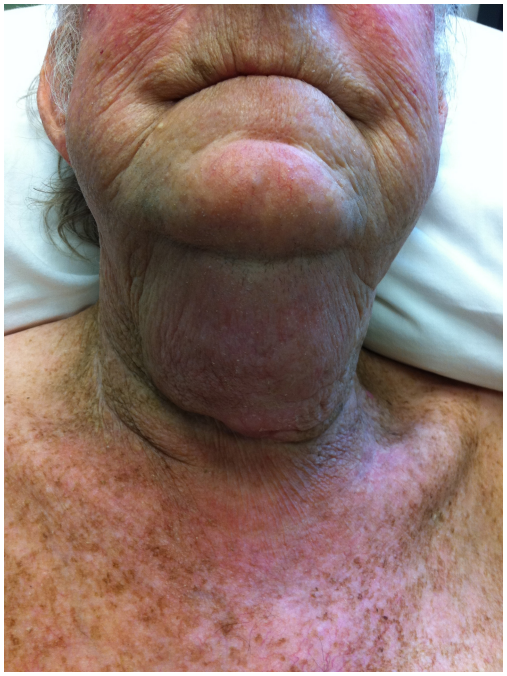
Number of Head and neck cancer survivors is Increasing due to HPV virus causing a high Percentage of new cases and combined chemo-Radiation is extending life.

Influencing factors Radiation:
Risk of Lymphedema is dose Dependent.
Getting radiation and chemo simultaneously influences risk.
Intensity of treatment Not location influences the development of LE with head and neck cancers.

Head and neck LE

Neck reconstruction

Waddle



Radiation therapy

- Goal is to destroy malignant cells but can also damage normal cells
- Causes fibrosis of lymphatics interfering with passage of lymphatic fluid

Combination Therapies: Surgery & Radiation Treatment

- Bentzen & Dische (2000) meta-analysis of combination therapy and the interaction with other modalities

  Results:

  **Showed marked increase in the incidence of lymphedema after combination treatment.*

  **Must find optimal balance between surgery and radiation- Sentinel Lymph node biopsy*

- Doe et.al. (2004) conducted a retrospective analysis of 300 women.

  Results:

  42.4 % of patients who underwent combination therapy reported lymphedema vs. 13.4% of patients treated with surgery only.

Lymphedema
Incidence

Worldwide:

- Parasites 90 million
- Breast Cancer 20 million
- Primary 2-3 million

(WHO Technical Report, 1984)

Breast Carcinoma-Related:

- 6% - 30% Breast Carcinoma Survivors

(Jeanne A. Petrek et al., 1998)

Review of Current Literature

- An estimated 212,920 persons will be diagnosed with breast cancer in 2006 (ACS, 2005).
- 77% of women were not told prior to treatment about the risk of developing lymphedema and physicians failed to recognize the symptoms (Paskett & Stark, 2000).
- The estimated incidence of lymphedema is reported in 10% - 60% of breast cancer patients.

Problems with Diagnosis

- Estimated that ~ 24,000 people diagnosed with LE annually in U.S.

- True incidence unknown
  - Lack of awareness
  - Lack of convenient, accurate, objective measurement methods
  - Variations in criteria used to define LE
  - Inadequate follow-up reporting

Measurement of limb/symptoms

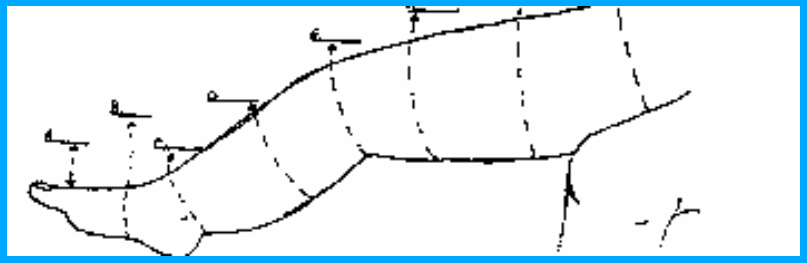
Tape measure

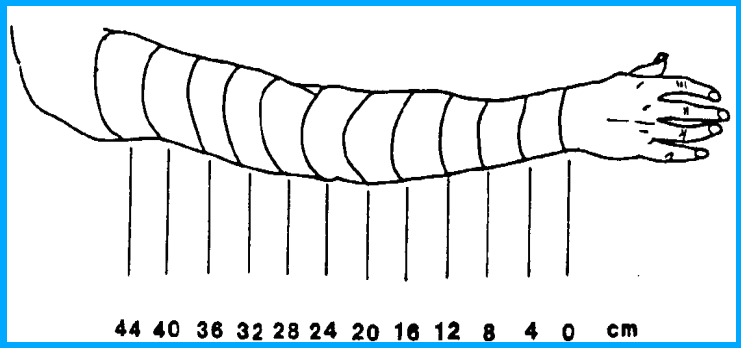
Self report of symptoms:

- tightness
- heaviness

Comparative limb assessment

History, history, history…





Pediatric Lymphedema

No standard definition

Circumference

- > 2 cm or more difference between limbs

Clinical grades

- Grade I - Asymptomatic
- Grade II - Mild
- Grade III - Moderate
- Grade IV - Severe

Volume

- > 200 mL difference, >10% volume difference

Few studies demonstrate correlation between

volume and function or QOL

Severity often associated with duration of symptoms

Co-Morbidities

Age:

- Conflicting research
- Tengrup et al. (2000) showed no correlation with age being a risk factor.
- Gellar et. al. (2003) increased risk in patients

  < 50 years of age.

Weight:

- *Memorial Sloan Kettering Cancer Center* reported BMI > 29.2kg/m2 significant predictive factor. Incidence rose to 36% compared to 12%with lower BMI.
- Increase weight =Increased radiation dose (Gellar et.al, 2003).

Co-Morbidities:

Infection:

- Development of fibrotic tissue.
- Lymphedematous tissue susceptible to infection.
- Burns, abrasions or puncture wounds may increase lymphatic destruction and blockage.
- Diabetic patients and frequent finger sticks = increased risk.

Hypertension:

- Encouraged to monitor BP
- The link is not yet understood.
- Lowering pressure, less fluid leaks out through capillaries
- Patients being treated for HTN had decreased incidence (Gellar et al., 2003).

Coexisting Medical Problems

- Orthopedic:
  - Neck and shoulder (important to get hx pre-operative)
  - Arthritis
  - Fibromyalgia
- Diabetes
- Carpal tunnel

External Factors:

Air Flight:

- Women report increased lymphedema with air flight.
- Casley-Smith (1996) suggests low cabin pressure may trigger lymphedema.

Work Activity:

< 50 years of age

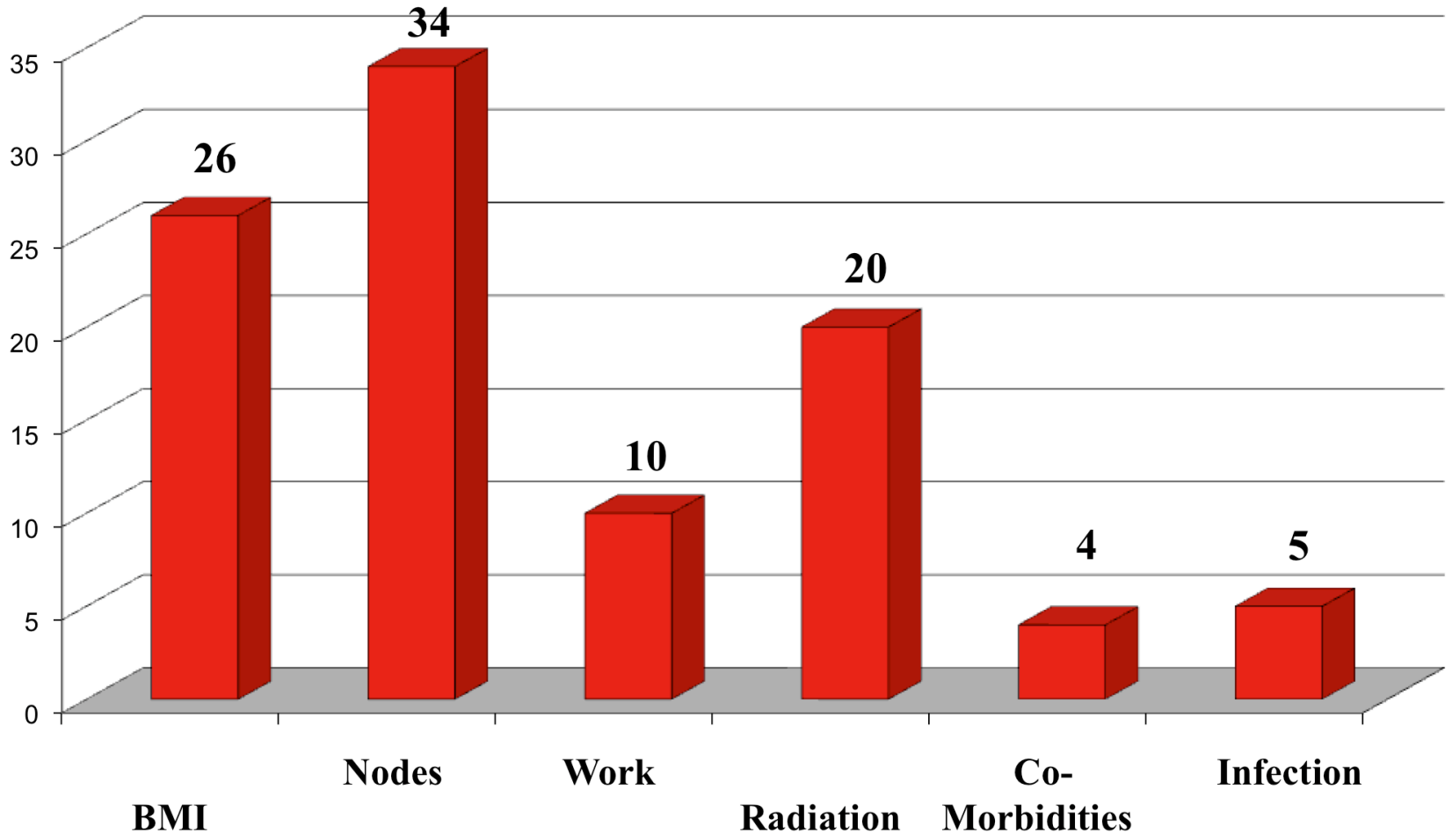
Repetitive movement or excessive use of limb

(Gellar et.al., 2003)

Heat:

- With ineffective perspiration, body's response is vasodilation (Guyton & Hall, 2000)
- Increased interstitial fluid in the skin can increase lymphatic fluid load and overwhelm limb (Szuba & Rockson, 1997).

Lymphedema Risk Criteria



Lymphedema Risk Assessment Tool

Risk Factors	*Score*	0	1	3
BMI		(<25kg/m2)	25-29	>29
Nodal Status/ Surgery		SNB	AND	Axillary Recurrence
Work Activity			Repetitive Movement or Moderate lifting	Heavy Lifting
Radiation		None	Breast Only	Breast & Axilla
Co-Morbidities •Diabetes •Hypertension		No	Yes	
Prior Cellulitis/ Infection		No	Yes	
Total				

Scale 0-10. Low Risk <3 Moderate Risk 4-6 High Risk > or equal to7
SND=sentinel node biopsy, AND= Axillary Node Dissection

Category of Risk

Low Risk:

+ 0-3

Medium Risk:

+ 4-6

High Risk:

+ > or equal to 7

Prevention / Management

- Early detection
  - Post-operative surveillance
  - Prompt reporting of symptoms
  - Risk Reduction Practices
- Early referral to LE treatment center

Evidence Based Reduction/Prevention of Lymphedema

**Avoid trauma or puncture of the skin to affected arm.*

1. Restrict venipuncture or injections to affected limb [Ex].
2. Monitor cuts, abrasions or hang nails to in affected extremity[Ex].
3. Meticulous skin care after injury [Ex].
4. Wear gloves when doing dishes, gardening, or similar activities [Ex].
5. Patient should be knowledgeable of the signs and symptoms of infection. (E.g. Elevated temperature, chills, hot to touch or redness to skin) [Ex].
6. Avoid blood pressure monitoring to affected extremity [Ex].
7. Avoid heavy lifting/strenuous exercise to affected extremity (0-15 pounds) [Ex].
8. Avoid inactivity or overprotection of affected extremity [Ex].
9. Avoid extreme heat or humidity [Ex].

Evidence-based Treatment Options for Lymphedema

1. Elevation and Compression Garments (1)
2. Pneumatic Compression Pumps (3)
3. Compression Bandaging (1)
4. Manual Lymphatic Drainage (2)
5. Debulking Procedures (3)
6. Low level Laser therapy (3)

Findings: Source of Self-Care Education other than Therapists

- 39% had never received self-care information from anyone other than their lymphedema therapist.
- Physicians (22%) followed by a variety of other sources, e.g., "someone on the internet" "someone else with lymphedema".
- NO ONE CITED A NURSE.

Lymphedema
Treatment

- Pneumatic Compression Pump
- Compression Garments
- Surgery
- Medications
- Complete Decongestive Therapy (CDT)

Lymphedema
Pneumatic Compression Pump

Problems:

- Does not remove protein
- Disregards ipsilateral trunk involvement
- ↑ Incidence of genital edema
- Does not reduce fibrosis
- Requires many hours/day
- May traumatize residual functioning lymph vessels

Lymphedema
Compression Garments

- Essential in "Self Care Phase"
  - prevents re-accumulation of edema fluid
- Custom Made/ Ready to Wear Garments
  - various compression grades and styles
  - flat knit, circular knit, cut & sew
- Prophylactic benefit

Compression garments

Lymphedema
Medications

- Penicillin/ Antibiotics
  - Tx. Infections (e.g. Erysipelas,, Cellulitis, Lymphangitis)
- Diuretics
  - Not useful, may make lymphedema worse

Complete Decongestive Therapy
Two Phase Treatment

Intensive Phase

- Daily Treatments
  - MLD
  - Compression bandaging
  - Remedial exercises
  - Skin & nail care
  - Instructions in self-care
- 2-4 Weeks
  - depends on severity

Self Care Phase

- Garments (Daytime)
- Bandaging (Night)
- Exercises (Daily)
- Skin & nail care
- MLD as needed
- Follow-up visits

Complications

Fungal infection

Cellulitis

Complications

Folliculitis

Inflammatory Breast CA

Complications

Skin reactions

Chemo reactions

Complications

Axillary web syndrome

+ Axillary web syndrome

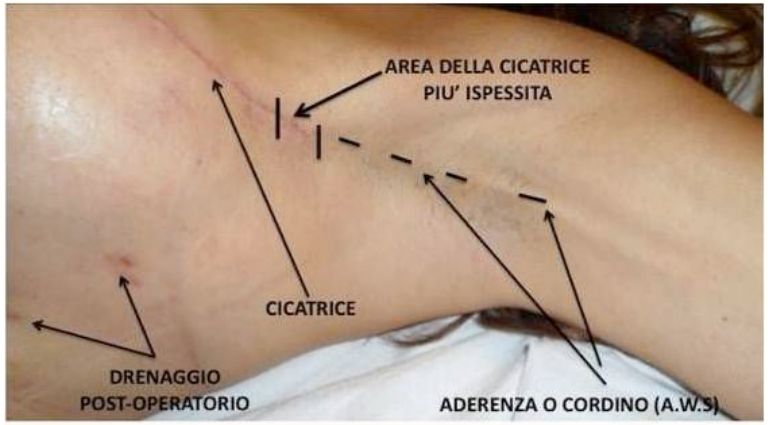


Fig. 2: Cordón (AWS) visible a nivel axilar, que prosigue incluso en el brazo y antebrazo (foto Glowinski, 2011)

Deep Vein Thrombosis

+ Deep vein thrombosis

Complications

Inflammation

Inflammation, xerosis

Exercise

Kinesiotaping

Self Care

- About 50% so some form of self care
- Barriers:
  - Expense
  - Time
  - Lack of outcomes
  - Disrupts life
- Benefits
  - Decrease in cost of acute care
  - Decrease in infections
  - Increase in perception of control

Complete Decongestive Therapy
UE Lymphedema - Before and After CDT

Complete Decongestive Therapy
UE Lymphedema - Before and After CDT

Complete Decongestive Therapy
UE Lymphedema - Before and After CDT

Complete Decongestive Therapy

LE Lymphedema - Before and After CDT

Summary

- Secondary LE is a common condition associated with the surgical treatment of many solid tumors.
- Underreported in the literature
- Education, surveillance and early management are critical