What is lymphedema?

- Lymphedema is an abnormal swelling that results from a malfunction of the lymphatic system. The lymphatic system is composed of organs, lymph nodes and lymph vessels that function in the removal of excess fluids from tissues and transport of WBC and other cells involved in the body’s immune response. Lymph vessels collect excess fluid from tissues and lymph nodes filter the fluid before it is returned to the circulatory system.

- Damage to or malfunction of the lymphatic system can affect the overall lymph load and trigger lymphedema.
- Lymphedema is a diagnosis, edema is a symptom.
- Lymphedema is classified as a high protein edema.

High protein vs low protein

- Edemas can be classified as high protein or low protein depending on the concentration of plasma proteins in the fluid.
- High protein – concentration of plasma proteins in fluid is > blood
- Low protein – concentration of plasma proteins in fluid is < blood
- Differential diagnosis is imperative to determine nature of edema and appropriate treatment
High protein vs low protein

Compression and manual techniques may be useful in treating both high and low protein edemas, but may also be contraindicated or ineffective depending on differential diagnosis. Diuretics are helpful in low protein edemas. They remove fluid only, leaving protein in interstitial spaces, thereby increasing overall concentration, and are not effective in treating high protein edemas.

Differential diagnosis

- Conditions resulting in extracellular edema include:
  - Increased capillary hydrostatic pressure
  - Decreased plasma proteins
  - Increased capillary permeability
  - Blockage of the lymphatic system

Primary vs Secondary

- Primary lymphedema involves a malformation, developmental defect or inefficiency of the lymphatic system
- Can be seen alone or as part of a syndrome
- Can manifest at birth or any point in the lifespan

Primary

- Present at birth – congenital lymphedema
- Onset up to 35 YO (usually 11-17) – lymphedema praecox
- Onset after 35 YO – lymphedema tardum
- Most common in females at all ages of onset
- More common in lower extremities
- Can be triggered by minor trauma or no trauma
### Primary
- Defect may be hypoplasia, hyperplasia or aplasia of lymph structures
- Hereditary types
- Typically symmetrical
- Starts distally, progresses proximally
- 10% of all lymphedema cases worldwide

### Secondary
- Secondary lymphedema results from injury, damage or overload of the lymphatic system
- In US, most common cause is radiation or removal of lymph nodes in the treatment of cancer and from chronic venous insufficiency

### Non cancer secondary lymphedema
- Vessel obstruction (scar or tumor), damage/injury or surgery
- Rheumatoid arthritis
- Filiariasis
- Silica dust
- Self induced injury
- Orthopedic injuries or surgeries

### Non cancer secondary lymphedema
- Neurological conditions (eg muscle paresis, CVA, post polio, spastic paralysis)
- Integumentary injury – degloving, burns, long term steroid use from COPD or RA
- Infection
- Cosmetic surgery – fat removal, liposuction, skin flap removal
- Often cumulative of multiple causes — surgery, obesity, trauma, chronic inflammation, steroid use, vein harvesting, etc.
<table>
<thead>
<tr>
<th><strong>Secondary lymphedema</strong></th>
<th><strong>Lipedema/Lipolymphedema</strong></th>
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<tbody>
<tr>
<td>• Tends to progress through stages more quickly, particularly UE lymphedema following axillary dissection</td>
<td>• Lipedema is a metabolic disorder of adipose tissue with unknown etiology</td>
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<tr>
<td>• Typically asymmetrical</td>
<td>• Seen almost exclusively in women</td>
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<tr>
<td>• Starts proximally, progresses distally</td>
<td>• Symmetrical bilateral edema, generally lower extremities, extending hips to ankles, but can occur in upper extremities as well</td>
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<td>• Currently no evidence of specific genetic disorder but often see family history?????</td>
<td>• Often starts in puberty</td>
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<td>• Can develop lymphedema overlying lipedema when fatty tissues hold fluid (Lipolymphedema)</td>
<td>• Low tolerance for compression</td>
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<tr>
<td>• Tissue becomes nodular and tough in later stages. Palpation resembles small styrofoam ball, but surface will appear smooth in early stages</td>
<td>• Diurectics not helpful</td>
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<td></td>
<td>• Liposuction can lead to lymphedema</td>
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<td>• Pneumatic pump may be helpful</td>
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<td>• Often tender to palpation</td>
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Other edemas

• Malignant lymphedema – tumor obstructing or compressing lymph vessels
• Mixed edemas – phlebo lymphedema, phlebo-lipo-lymphedema

Triggers

• Any person who has had lymph nodes removed or radiated is considered to be “at risk” for development of lymphedema
• Lymphedema can be “triggered” in at-risk persons by a variety of things including infection, weight gain, morbid obesity, circulatory deficiencies, air pressure changes including air travel, constriction or crush injuries, repetitive or heavy lifting, temperature extremes, chronic inflammation, sunburn, changes in fluid volume including pregnancy

Symptoms

• Swelling
• Numbness/tingling
• Discomfort or pain (typically achiness)
• Pressure, skin tightness
• Heaviness in the limb
• Fatigue of limb
• Increased incidence of infection

Symptoms

• Fibrotic changes to the skin
• Decreased mobility
• Decreased wound healing
• Lymphatic cysts/fistulas
• Papillomas
• hyperkeratinosis
Precautions to decrease risk

- Avoid blood pressure readings on affected extremity (lymph vessels collapse at approx 60 mm Hg)
- Avoid needle sticks/IVs, cuts and insect bites on affected extremity. Clean any opening with alcohol and apply antibiotic ointment. Shave with electric razor.
- Avoid fungal infections of skin and nails
- Avoid temperature extremes and sunburns
- Wear compression during air travel
- Avoid tight, constricting clothing and jewelry
- Maintain healthy weight (higher BMI is associated with increased incidence of lymphedema)

Stages of progression

- Left untreated, lymphedema generally is a progressive disorder
- Stage 0 – latent stage
  - no clinical signs of swelling (not measureable)
  - may complain of heaviness or aching
  - sub clinical changes are occurring in the tissues

Stages

- Stage 1 – acute stage
  - swelling reverses spontaneously with elevation
  - May demonstrate pitting late in stage 1
  - Texture may be soft, puffy or taut
  - May have bursting pain if sudden onset
  - Symptoms include heaviness, aching or fatigue, mild erythema, measurable swelling, puffiness, warmth

Stages

- Stage 2 – chronic stage
  - Does not reverse fully with elevation
  - Texture becomes firm, rubbery, viscous (fibrosis forming)
  - May no longer pit at end of this stage
  - Continued complaints of heaviness, aching, fatigue
  - May see rubor and warmth
Stages

• Stage 3 – elephantiasis
• Firm, non pitting edema
• Trophic changes noted (thickening of the skin, papilloma form, hair and nail changes, permanent skin color change; more common in lower extremity lymphedema)
• Skin folds and lobuli form
• Weeping may occur
• Infections increase
• Increased risk for lymphangiosarcoma (malignancy)

Fibrosis

• High protein concentration in interstitial fluid causes increased connective tissue cell proliferation (Fibroblasts), increased collagen fiber production leads to formation of sclerosis and fibrosis.
• Many changes in the progression of lymphedema are due to this process.

Standard treatment

• No cure
• Most effective evidence based treatment currently is Complete Decongestive Therapy (CDT)
• Addresses physical and psychosocial aspects of lymphedema
• Developed in Europe in early 1900s

Treatment

• CDT consists of 5 components
• Compression bandaging and garments
• Remedial exercises
• Manual lymphatic drainage
• Skin care and hygiene to decrease infection risk
• Home program
Goals of treatment

Utilize remaining lymph pathways
Encourage alternate routes of drainage
Improve efficiency of available drainage
Encourage new vessel formation
Remodel skin to regain elasticity and support

Compression bandaging and garments

• Compression bandaging involves use of short stretch bandages in multi layer fashion to build a compression gradient to work against the effects of gravity
• Short stretch bandages have high working pressure, low resting pressure to increase effectiveness of the muscle pump and comfort
• Bandaging decreases ultrafiltration, prevents re-accumulation of fluid, breaks up fibrosis, provides support for lost elasticity
• Used to reduce edema prior to fitting of garments

Garments

• Can be ready made or custom
• Variable compression levels, classes 1-4 (20-60+ mmHg pressure)
• Variable styles, fabrics and options available
• Day and nighttime garments available
• Gradient compression is built into garments
• Daytime garments are used to contain edema once it has been reduced, they are not designed to reduce edema
• Covered by most insurance plans for breast cancer patients, but NOT by Medicare

Remedial Exercises

• Utilizes the muscle and joint pumps to help remove excess fluid
• Can assist in reducing excess weight, a common trigger of lymphedema
• Strength training is appropriate but should be progressed slowly
• Aerobic exercise, yoga, pilates also helpful
• Water exercise excellent option
• Compression garments should be worn (except during water exercise)
• Exercise classes for cancer survivors are available here at MECC
• Patients who have undergone mastectomy or lumpectomy with axillary lymph node at Methodist Hospital are educated in Phase I exercises before discharge and Phase II exercises are offered at 3-6 weeks post op to address ROM. Lymphedema education is also completed.
Manual Lymphatic Drainage

- Increases frequency of lymph vessel contraction
- Increases volume of lymph transported
- Can alter directional flow to “detour” around malfunctioning areas
- Very gentle and unique to each patient
- NOT massage
- Performed by therapist in clinic and taught to patient to complete at home
- Other manual techniques are often used to address scar tissue and restrictions to improve lymph flow and improve ROM

Skin care

- Decrease infection risk by preserving integrity of skin “barrier” to environmental harm
- Decrease risk of nail fungus by avoiding salon manicures and pedicures
- Cracks in dry skin or any opening in skin are open portal for bacteria

Home Program

- Goal is to educate patient to continue with home program of self care and edema management. Since there is no cure for lymphedema, management is a lifetime commitment
- Can include skin care and precautions, compression bandages and garments, exercise, MLD and monitoring, and will be unique to each person

Contraindications to treatment

- Untreated cellulitis (May begin treatment within 24-48 hrs of antibiotic initiation)
- Renal failure
- Uncontrolled or acute exacerbation of CHF
- Untreated cancer
- DVT
- Active TB
Precautions to treatment

- Skin reactions or allergic reactions
- Active cancer
- Uncontrolled pain
- Uncontrolled hypertension

Typical patient populations

- CVI
- CVA
- Cancer patients
- Post surgical patients
- Crush injuries
- Burn patients
- Post vein stripping
- Obese patients

Appropriate referrals

- Patients who have been evaluated to determine cause of edema and rule out contraindications for treatment are appropriate.
- Therapist may modify treatment plan dependent upon patient’s relative precautions and concurrent medical treatments. Patients can receive modified treatment during radiation therapy and chemotherapy.

Truths vs misconceptions

- Can occur with radiation and/or lymph node dissection in any type of cancer
- Ace bandages are NOT used in CDT
- Diuretics are not helpful in true lymphedema, but may be helpful in other types of edema
- No weight lifting limit; appropriate exercise, including strength training, is helpful
- Risk lasts a lifetime
Truths vs misconceptions

- Compression garments do not reduce edema, they only contain it once it has been reduced
- Lymphedema is not curable
- Although new lymph vessels are formed and body adapts over time and most lymphedema is diagnosed within 3 years of cancer treatment, patients are considered at risk for the remainder of their lives, thus precautions should continue to be observed.
- Sentinel node biopsy alone carries a 3-10% risk of lymphedema development

What can you do?

- Listen to patient’s questions and concerns. Descriptors “heavy, full, tight, achy” are indicators patient may have lymphedema developing.
- Patients often need repeat therapy and garments need to be replaced every 6-12 months. Don’t assume they don’t need additional therapy if just because they’ve been treated before.
- Follow precautions within reason and educate patients that risk lasts a lifetime, but emphasize it need not interfere with daily life.

What can you do?

- Refer to lymphedema therapy. Services are available at Methodist Hospital both inpatient and outpatient (2N and MECC clinics).
- Encourage patients to seek treatment with therapist who is a Certified Lymphedema Therapist who has completed 135 hour certification course.

QUESTIONS?