Lymphedema 101

Part 1: Understanding the pathology and diagnosis

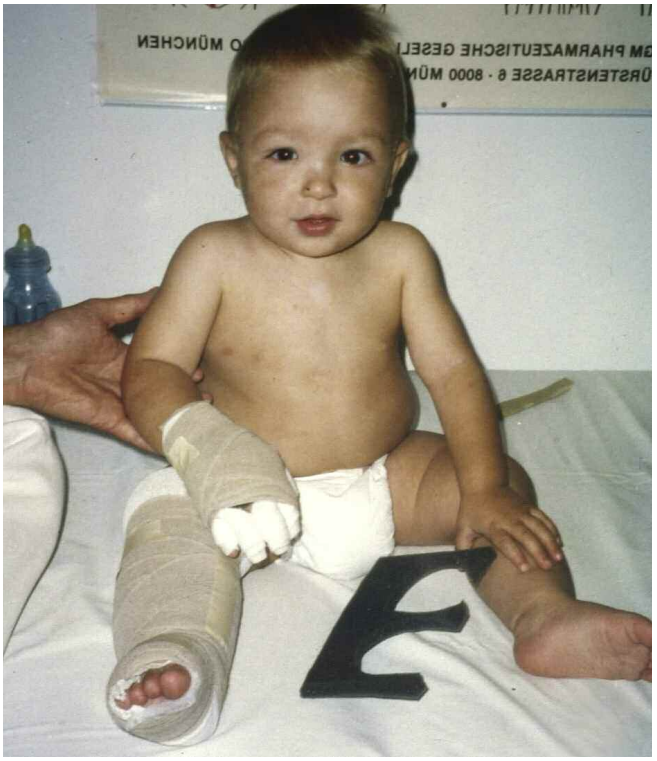
By Steve Norton, CDT, CLT-LANA

Lymphedema is characterized by regional immune dysfunction, distorted limb contours, and such skin changes as papillomas, hyperkeratosis, and increased girth. The condition may involve the limbs, face, neck, trunk, and external genitals; its effects may include psychological distress. For optimal patient management, clinicians must understand what causes lymphedema and how it's diagnosed and treated.

This two-part series provides an overview of lymphedema. Part 1 covers etiology, pathology, and diagnosis. Part 2, which will appear in the November-December issue, will focus on treatment.

Causes of lymphedema

Lymphedema occurs when protein-rich fluid accumulates in the interstitium due to impaired lymphatic function. Proteins, other macromolecular wastes, and water constitute lymphatic loads. These wastes rely on specially structured absorptive and transport structures in peripheral regions for their return to central circulation. When lymph stasis prevails, inflammatory processes and lymphostatic fibrosis trigger tissue-density changes, further entrapping superficial vessels and accelerating mechanical insufficiency. (See *Physiologic changes caused by lymphatic disruption*.)



Primary lymphedema in a 2-year-old child

Classifying lymphedema

Lymphedema can be primary or secondary. *Primary* lymphedema either is congenital (present at birth) or arises around puberty. In the vast majority of cases, it is associated with structural changes in the lymphatic system and isn't associated with another disease or condition. Most structural changes (87%) manifest before age 35 and cause hypoplasia of vessels and nodes. Syndromes involving hyperplasia, node fibrosis, or aplasia also may occur, although they're much less common. Dysplasia (either hypoplasia, hyperplasia, or aplasia) predisposes drainage regions to inadequate lymph collection, resulting in edema and secondary tissue changes, such as chronic inflammation and reactive fibrosis. Genetic variability in lymphatic constitution may explain why seemingly similar patients receiving the same surgical protocol have different lymphedema risks over time.

Secondary lymphedema stems from a significant insult to lymphatic tissues, as from lymphadenectomy, radiation therapy, trauma, infection, or cancer. It commonly results from direct trauma to regional

nodes or vessel structures. Slow degradation of lymphatic function also occurs when adjacent tissues (such as superficial and deep veins) become diseased, when cellulitis occurs, or when accumulations of adipose or radiation fibrosis mechanically disrupt drainage of skin lymphatics.

Lymphedema stages

Lymphedema progresses in stages, which involve secondary connective-tissue disease combined with disturbed fluid update and transport. These conditions cause a universal and classic clinical picture.

- *Stage 0* (latency stage) is marked by reduced transport capacity and functional reserve. The patient has no visible or palpable edema, but has such subjective complaints as heaviness, tightness, and waterlogged sensations.
- In *Stage 1* edema (reversible lymphedema), edema decreases with elevation. Pitting edema is present, but fibrosis is absent.
- During *Stage 2* (spontaneously irreversible lymphedema), lymphedema doesn't resolve entirely, although it may fluctuate. Pitting is more pronounced and fibrosis is present.
- *Stage 3* (lymphostatic elephantiasis) is marked by dermal hardening, nonpitting edema, papillomas, hyperkeratosis, and in some cases, extreme girth.

Assessment and diagnosis

Diagnosing lymphedema can be challenging because edema may be associated with other diseases and disorders. For a summary of signs and symptoms, see *Clinical findings in lymphedema*.

Discomfort and skin appearance

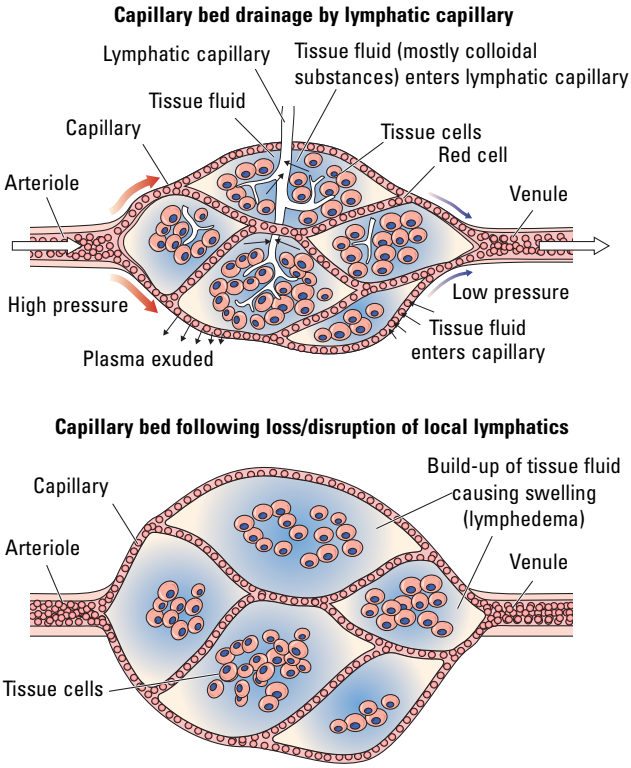
Lymphedema rarely causes pain because the skin accommodates gradual, insidious fluid accumulation. However, secondary orthopedic discomfort may result from increased weight of the affected limb due to deconditioning or decreased range of motion.

Because lymphedema usually progresses slowly, gravity and centrifugal forces pull fluids toward distal limb areas, causing an entrenched, stubborn pitting edema. Later, further valvular incompetence contributes to worsening distal edema in the fingers, toes, and dorsal regions of the hand and foot. Prominent lower-extremity structures, such as the malleolus, patella, tibia, anterior tibialis tendon, and Achilles tendon, become progressively less distinct. This creates a columnar limb appearance; the swollen limb has the same girth from distal to proximal aspects, unlike the natural cone shape of a normal limb.

Lymphatic failure doesn't tax the venous system, so skin color remains normal. Blood supply remains patent, helping to prevent secondary ulcers.

Physiologic changes caused by lymphatic disruption

These illustrations compare normal capillary bed drainage with disruption or loss of local lymphatics.



Clinical findings in lymphedema

Although signs and symptoms vary with lymphedema stage, they generally include the following:

- Slow, gradual progression
- Pitting during early stages
- Distal-to-proximal advancement (may spare the hand or foot)
- Loss of bony contours, causing a columnar appearance
- Dorsal "buffalo hump" if the hand or foot is involved
- Normal skin color (except during Stage 3)
- History of infection

Although many patients with lymphedema complain of discomfort, pain and ulcerations (pure lymphedema) are rare.

Severity

Lymphedema severity correlates directly with such factors as onset of the condition and extent of cancer therapy, if given (number of nodes resected, number of positive nodes, and use of radiotherapy). Lymphedema may worsen with a greater number of infection episodes, weight gain, injury, diuretics, limb disuse, pneumatic compression therapy (when used for pure lymphedema), and ill-fitting compression garments. The single most important contributor to increasing lymphedema severity is lack of patient education, which can result in improper treatment or none at all.

Opportunistic infections

Lymphedema causes regional immune suppression and leads to an increase in opportunistic infections such as cellulitis. As skin integrity suffers, scaling and dryness allow resident skin pathogens (such as streptococci and staphylococci) to gain access through the defective skin barrier into protein-rich interstitial fluid, creating a medium favorable to bacterial colonization. Lymphocyte migration decreases, and dissected or irradiated nodal sites are slow to detect invaders. Furthermore, stagnant lymph promotes further delays in the immune response. Patients with opportunistic infections may exhibit high fever, local erythema, regional hypersensitivity or acute pain, flulike symptoms, and rapidly advancing "map-like" borders in the skin.

Differential diagnosis

Several methods can aid differential diagnosis.

Clinical findings. Lymphedema can be diagnosed from patient history, physical examination, palpation, and inspection. Trauma to lymph nodes (each of which governs a distinct body region) decreases the transport capacity of lymph formed in that region, in turn causing local swelling (lymphedema). Trauma to the axillary or inguinal lymph nodes, which exist on both the left and right of the body and in both the upper and lower regions, predisposes these quadrants to swelling. Therefore, if lymph nodes on only one side are damaged, lymphedema occurs only on that side of the body. Using the universal characteristics cited above as a guide, while ruling out cancer recurrence, acute deep vein thrombosis, or plasma protein abnormalities, yields sufficient data to form a diagnosis.

Imaging. Lymphography involves subcutaneous injection of a lymph vessel–specific dye (Patent Blue V), followed by X-ray. Although it provides high-resolution images of lymphatic structures, this technique is invasive, painful, damaging to lymphatics, and potentially lethal—and therefore is no longer recommended.

Lymphangioscintigraphy (LAS) uses interdigital subcutaneous injection of protein-labeled radioisotopes, followed by imaging at specific intervals to gather information about uptake and transport time. Images are hazy and false-negatives are common, so well-trained radiotherapists familiar with lymphology and lymphedema should administer and interpret the test.

Also, experts don't agree on standard criteria for LAS administration, so measures may not be similarly conclusive.

Limb-measuring instruments and methods. Serial measurement of affected limb circumference using a standard garment tape measure is the most widely accessible approach. Intra-rater reliability is comparable to that of currently used tools; however, these methods can't be used for early detection, for screening, or when various raters are used to assess the same patient. Circumferences are measured at four points and are considered positive if a distance of 2 cm or more separates the involved from uninvolved extremity in comparison. Water displacement techniques for limb-volume calculation, although accurate, are impractical in most clinical settings and rarely used.

Most often used in research settings, the Perometer® uses infrared beams to scan the girth of a limb.
Courtesy Julius Zorn, Inc.

Impedimed XCA® uses bioelectrical impedance to obtain measurements.
Courtesy Impedimed, Inc.

Various devices have been used to obtain measurements. For instance, the Perometer® uses optoelectronic volumetry. By scanning the limb with infrared beams circumferentially, the device accurately records girth at 4-mm intervals along the limb length and transmits these measurements to a computer. The Perometer is used mainly in the research setting. Preoperative and postoperative measurements at intervals can detect lymphedema early.

Impedimed XCA® uses bioelectrical impedance to calculate ratios of intracellular to extracellular fluid. A weak electrical current is passed through affected and unaffected limbs, allowing comparison of results. Impedance is lower in edematous tissue, supporting an accurate diagnosis.

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- May be able to be cut to accommodate tubes

Disadvantages

- Could be expensive if exudate requires daily dressing change
- Wound bed may desiccate if there is no exudate from the wound
- May require secondary dressing
- Can lead to maceration of the peri-wound if it becomes saturated
- Contraindicated for use with third-degree burns, dry eschar, and sinus tracts

Tips

- Dressing should be 1" to 2" (2.5 to 5 cm) larger than the wound.
- Change the dressing every 3 to 7 days or as necessary.
- When using nonadhesive foam, add a secondary wound dressing for securement.
- You may facilitate dressing removal by stretching the adhesive border laterally.

Examples/Coding

- Mitraflex, Flexan, Hydrasorb, Lyofoam, Allevyn, PolyMem
- Healthcare Common Procedure Coding System (HCPCS) Code A6209 – A6215 ■

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Next step: Treatment

Once a diagnosis is made, the next step is treatment. Part 2 of this series covers lymphedema treatment. ■

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