

Essential Lymphedema Information for All Health Care Providers: from a Physician with Secondary Lymphedema

I am a physician who acquired secondary lymphedema while being treated for breast cancer. Despite being a medical school educator, board certified in family medicine and involved in the care of many women with breast cancer, I had essentially no knowledge of lymphedema--there was one question on my boards, and no continuing education covered it. After I developed lymphedema, I acquired both a personal and professional education on the subject. I now care for oncology patients, and I see many patients who are compliant and are examined by many providers and yet their lymphedema is over-looked. Lymphedema is not addressed in medical school education, residency or continuing medical education. I'd like to share the information I've learned in an effort to provide a short primer on the subject and general advice in managing your patients who are at risk for lymphedema. This information is for all members of the health care team--if you touch, talk to, run tests or treat patients at risk for lymphedema, knowledge of the condition will enhance the care and health of your patients.

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“Therapeutic nihilism (i.e. no treatment at all) is deplorable, although common. The fact that the average clinician is ill prepared to recognize the early signs of lymphedema must be remedied, because the sooner the treatment is started, the less treatment is required to prevent further progression.”

Cancer, Principles and Practice of Oncology, 7th Edition, DeVita et al, p.1485

1) LEARN ABOUT IT: Education about Lymphedema and the lymphatic system is lacking: Dr. Stanley Rockson (Stanford) has shown that most physicians have received 15 minutes or less of education on the lymphatic system. Lymphedema is not a thing of the past: it is common and providers need to understand the disease and management. This is a chronic, progressive disease and early diagnosis and proper treatment is essential. Lymphedema is a disease of inflammation and derangement of the lymphatic and connective tissues of the affected area—it is NOT just a swollen arm. Dr. Rockson's lectures can be accessed for free on YouTube: <http://www.youtube.com/watch?v=XR-9duCfgF0> Or via Stanford: <http://healthlibrary.stanford.edu/videolibrary/index.html> - rockson09

Provider training can be obtained through the major lymphedema training

schools and the National Lymphedema Network. Here is a free review article in the journal Cancer: Lymphedema Primer: CA Cancer J Clin 2009; 59:8-24 <http://caonline.amcancersoc.org/cgi/content/full/59/1/8>

2) LYMPHEDEMA IS NOT A DISEASE OF THE PAST: The true incidence of lymphedema is unknown as there is no single validated diagnostic standard, but Jane Armer, PhD who has followed over 250 women for greater than 8 years has found the prevalence varies from 43-94% depending on the measurement method employed: this includes women with “just” sentinel node biopsies <http://www.ncbi.nlm.nih.gov/pubmed/21226414>
Lymphology. 2010 Sep;43(3):118-27.

3) LEARN TO RECOGNIZE AND DIAGNOSE LYMPHEDEMA: Do a physical exam to look for lymphedema at every follow up visit, or when there are concerns: Examine limbs, chest, breast and axilla for subtle changes and swelling. Symptoms often precede swelling—take pain, aching or tightness in the at risk area seriously. Look for pitting edema, skin texture changes—the swollen area will be either tight or boggy or on the breast it can be reddened thickened skin, skin color changes as lymph is light colored and obscures blood vessels, in hands check for swelling of the dorsum or palm and have the patient make a fist and check the MCP joints between the hands for obscuring, have the patient bend their elbow and observe the delineation of the olecranon and tendons between limbs. If patients have bilateral lymphedema or are at risk bilaterally, the exam becomes much more difficult, as there is no “control” side to use for comparison.

On this site there is an excellent series of photographs of a patient with controlled, but significant lymphedema—utilize the images and realize the presentation may be more subtle:
http://www.stepup-speakout.org/What_does_lymphedema_look_like.htm

There is no diagnostic standard for diagnosing lymphedema: common methods that are used are inter-arm circumference discrepancy: measure the arms at several points and compare the circumferences, every 4 cm is the standard method of calculating limb volume. While the 2 cm difference has been clinically adopted as “the gold standard” for diagnosis, it is a clinical standard that began with one clinical trial and became widely adopted. The sensitivity and specificity of the “2 cm rule” are unknown. So, while a 2 cm size discrepancy—at any one point on the two arms—strongly suggests lymphedema, due to fibrosis and “patchy” distribution of disease, patients can have lymphedema without a 2 cm inter-arm circumference discrepancy. A.W. Stanton, PhD has published that the clinician must be alert to early changes in the limb: obscuring of bony or

tendon landmarks, and that a 2 cm discrepancy is not required to diagnose lymphedema. Link to Stanton, et al, Diagnosing Breast Cancer-Related Lymphoedema, full article

[Http://britishjournaloflymphoedema.com/journal/0101_arm.pdf](http://britishjournaloflymphoedema.com/journal/0101_arm.pdf)

Early lymphedema, stage 1 or 0, is spontaneously reversible and the swelling is highly variable.

More sophisticated measures of lymphedema are perometry—a laser method of assessing volume, cylinder water volume displacement, and a less sensitive and specific method of serial bioimpedance values can be utilized to measure and with history and exam, confirm the diagnosis of lymphedema.

-- History is crucial in evaluating patients. Take a history of swelling, pain, achiness, and/or tightness in the entire quadrant “at risk.” Validated surveys have indicated high incidences of intermittent swelling in women who have been treated for breast cancer. Yet even if swelling resolves spontaneously, it is still Stage 0 lymphedema.

---Be aware of breast and truncal lymphedema: The incidence has increased with sentinel node biopsy: *Lymphology*. 2006 Mar;39(1):33-40 “Radiotherapy after breast conservation surgery leads to increased long-term changes in basal lymph circulation and smaller increases in lymph flow in the contralateral breast receiving 2-4 Gy and after surgery. If maximal lymph transport capacity is unchanged, edema may be more likely in this circumstance of reduced lymphatic transport reserve.” <http://www.ncbi.nlm.nih.gov/pubmed/16724508>.

This full length article is an excellent review of the subject: *Journal of Lymphoedema*, 2006: http://www.lymphoedema-uk.com/journal/0101_breasttrunk.pdf

4) DISEASE MANAGEMENT:

Know the sequela and co-morbidities of lymphedema:

High risk of severe cellulitis—patients should have prescriptions on hand of antibiotics that treat cellulitis so that they can start them at the first sign of redness, pain, swelling or fever—and then contact their physician. Please provide antibiotics for patients with lymphedema—this is not a case of inappropriate antibiotic use.

Cellulitis is both a risk factor for lymphedema and a poor prognostic indicator as cellulitis irreversibly destroys lymphatics.

From NE Journal Review of Cellulitis: “A distinctive form of cellulitis, sometimes recurrent, may occur weeks to months after breast surgery for cancer. Cellulitis in the ipsilateral arm has been well described after radical mastectomy where it occurs because of associated lymphedema; cellulitis in the ipsilateral breast is more common now, occurring after breast-conservation therapy. Local lymphedema from the combination of partial mastectomy, axillary lymph-node dissection, and breast irradiation is a predisposing factor.” <http://www.nejm.org/doi/full/10.1056/NEJMcp031807> This link is to an image of breast/arm cellulitis: <http://www.nejm.org/doi/full/10.1056/NEJMicm065836>

Pain: Lymphedema is often quite painful and that pain should be validated and managed. The pain is multi-factorial: neuropathic, mechanical, and/or inflammatory. The National Lymphedema Network issued a statement confirming that the condition can be painful, http://www.lymphnet.org/pdfDocs/NLN_Washington_Post_Reply.pdf

Emotional response: Patients find lymphedema emotionally distressing: patients feel abandoned, stigmatized and generally distressed. The disease is disfiguring, disabling and requires daily care. They often find that the condition is under-recognized—to the point of being denied, and that their providers are not educated enough about the disease to manage it or supervise their treatment by lymphedema therapists. Lymphedema reduces the quality of life for breast cancer survivors. Multiple studies have documented this distress. Please address this with your patients and support them.

<http://www.ncbi.nlm.nih.gov/pubmed/20797846> : 2010 Study: ”
CONCLUSIONS: In this cohort of women with breast cancer, women with lymphoedema after surgery for breast cancer had significantly worse overall emotional well-being and adjustment to life compared to women without lymphoedema.”

---Learn about lymphedema therapy and form therapeutic alliances with well- trained therapists:

Read the NLN position papers on therapy, therapist training, and utilize the therapist locator web page to find therapists. There is no standard for lymphedema therapy training, although the NLN recommends that a properly trained therapist receive 135 hours of training and supervision. Even with adequate training, therapists require supervision and consultation from referring physicians—therapists should not manage this chronic disease without consultation from health care providers. <http://www.lymphnet.org/lymphedemaFAQs/positionPapers.htm>

The standard physical therapy model of acute intervention and transition

to home management is not sufficient for this chronic disease: ideally patients should have regular maintenance evaluations and follow up and treatment as needed. This disease is chronic and progressive; patients require hands on periodic evaluation.

---Understand Lymphedema “Gear”: patients will require compression garments, which should be replaced every 6 months, and a sleeve should not be dispensed without hand protection—either a gauntlet or glove, <http://lymphedivas.com/lymphedema/gauntletandsleeve/> Treatment involves lymphedema compression wrapping, specialized nighttime garments, daytime compression garments and at times, pneumatic pumps. None of these treatments are fully covered by insurance and often they will require prescriptions from a treating physician. Again, review the National Lymphedema Network Position paper on treatment, and please provide prescriptions for your patients.

There are only a handful of rehabilitation physicians in the country who specialize in lymphedema. Your patients need you to know about the condition, to be informed to diagnose it, to treat the complications of it, to help manage the therapy and to follow them for this chronic condition.

5) WORK TO MINIMIZE LYMPHEDEMA IN YOUR PATIENTS:

Education, Advice, Anticipatory Guidance:

---Educate your patients about lymphedema: Most patients are not given accurate information, and most physicians do not consider lymphedema counseling or advice to be their responsibility. An article from Breast by Electra Paskett PhD revealed: “Overall, women knew little to nothing about lymphedema before they developed it . . . Most physicians reported that they did not routinely counsel women or provide written information on lymphedema prevention to their patients, and the extent to which women's daily living was affected by the condition was not always recognized.” <http://www.ncbi.nlm.nih.gov/pubmed/11348395>

--- Pre-operative lymphedema therapy evaluation and post-operative limb measurements for early detection of lymphedema should be standard practice: The National Lymphedema Network issued a position paper in 2011, "Screening and Measurement for Early Detection of Breast Cancer Related Lymphedema", calling for pre-treatment baseline measurements, consistent follow up measurements, inquiring about arm heaviness, achiness and/or swelling of the affected arm and trunk at all follow up visits: <http://www.lymphnet.org/pdfDocs/nlnBCLE.pdf>

---Close and careful surveillance of patients with early intervention

minimizes lymphedema:

<http://www.ncbi.nlm.nih.gov/pubmed/18428212>

Cancer. 2008 Jun 15;112(12):2809-19. In this study, women's arms were measured pre-op and at every post op visit and if a 3% increase occurred, they were placed in compression garments, with excellent results; "CONCLUSIONS: A short trial of compression garments effectively treated subclinical LE."

Currently Massachusetts General Hospital is replicating this study: <http://www.clinicaltrials.gov/ct2/show/NCT00959985>

---Radiation increases the risk of lymphedema: Even when women are receiving whole breast radiation without nodes intentionally included in the field, radiation oncologists should work to minimize radiation to the level one nodes in node negative women. Andrea Cheville, MD (Mayo) is pioneering a radiation technique where lymph nodes that drain the arm are blocked to minimize lymphedema. http://www.abstracts2view.com/sabcs10/view.php?nu=SABCS10L_1220

<http://www.ncbi.nlm.nih.gov/pubmed/20951451> (publication)

interview: <http://newsblog.mayoclinic.org/2010/12/06/lymphedema-study/> - 2

Cheville AL, Petersen IA, Brinkman DH, Ward SB, Mullan BP, Durski J, O'Connor MK, Schomberg PJ, Yan E, Garces YI, Laack NN. Mayo Clinic, Rochester, MN

"Lymphedema (LE) is a common complication of cancer treatment, and has been identified as the number one issue by breast cancer survivors with incidence as high as 70%. Given the long-term survivorship of these women, LE is a widely prevalent condition. Irradiation of axillary and supraclavicular lymph nodes increases a patient's risk of LE onset and progression by as much as 2-fold."

---Inform all of your patients of the National Lymphedema Network's risk reduction behaviors-and all office staff should be aware of them as well: <http://www.lymphnet.org/lymphedemaFAQs/positionPapers.htm>

The knowledge reduces the risk of developing lymphedema: <http://www.ncbi.nlm.nih.gov/pubmed/21186149>

"CONCLUSIONS: Exposing women to lymphedema risk information at the time of breast cancer diagnosis facilitates increased awareness and enactment of risk-minimization behaviors." Oncol Nurs Forum. 2011 Jan 1;38(1):E27-36.

---Learn which patients should wear compression garments while flying: all patients with lymphedema and those at high risk are advised to wear a sleeve and hand garment while flying. They will need a prescription from

you for garments. Read the position paper by the NLN on flying:

“It is the position of the National Lymphedema Network that: Individuals with a confirmed diagnosis of lymphedema should wear some form of compression therapy while traveling by air. Individuals at risk for developing lymphedema should understand the risk factors associated with air travel and should make a decision to wear compression based on their individual risk factors.”

<http://www.lymphnet.org/pdfDocs/nlnairtravel.pdf>

---There is a movement to “debunk” risk reduction behaviors as not evidence-based, yet the clinical experience to support these behaviors are based in broad experience and if a permanent disease occurs due to an avoidable, iatrogenic behavior, the patient’s incidence of lymphedema becomes 100%.

---For patients with bilateral lymphedema or bilateral risk, learn how to take leg blood pressures, foot blood draws and foot or neck IV’s if necessary. <http://www.medscape.com/viewarticle/471829>

While popliteal thigh pressures can be obtained, many patients find posterior tibial ankle blood pressures are more comfortable: http://www.ehow.com/how_5729015_check-blood-pressure-ankle.html

<http://www.ncbi.nlm.nih.gov/pubmed/8912031>: “Since the mean blood pressure readings obtained at the arm and at the ankle were statistically equivalent, we concluded that the ankle cuff placement provided a reliable alternative to the placement of the cuff on the arm.”

Concerns have been raised over the safety of intravenous lines in the lower extremity: they have been associated with a higher risk of infection, should be avoided when artificial joints are in the limb and this author could find no study that documented increased risk of deep vein thrombosis. The higher risk of infection needs to be weighed against the risk of exacerbation or triggering of lymphedema with the use of an IV in a limb with compromised lymphatic function.

---Learn about exercise and lymphedema: Exercise is a key component of lymphedema therapy and all patients are encouraged to exercise. Patients with lymphedema or at risk for it, should not exercise their upper extremities to the point of fatigue and need to monitor their arms for swelling or discomfort after exercise. There are far too many cases of anecdotal reports of women developing lymphedema in exercise programs designed for cancer survivors, using exercise trainers who are unaware of lymphedema precautions or even with standard physical therapy. Resistive exercise, despite recent studies, can pose a risk to women both with lymphedema and at risk for it. From the NLN position

paper on exercise-<http://www.lymphnet.org/pdfDocs/nlnexercise.pdf>:
"Studies: Resistive exercise has been formally studied in "at risk" individuals, and when begun at a low level and increased gradually, has not been found to trigger or worsen lymphedema. An isolated study in lymphedema patients suggests that resistive exercise, in the absence of compression, may lead to increased lymph accumulation and eventually worsen limb swelling. Potential Benefits: Resistive exercise may enhance lymph flow and prevent limb swelling from muscle overuse. Potential Risks: Strength training increases local blood flow and metabolic waste production. These effects increase the demand on the lymphatic system, potentially triggering or worsening limb swelling."

In carefully selected patients, who participated in a specific exercise protocol, (PAL protocol), utilizing specially trained trainers and frequent assessment by trained lymphedema therapists, in this highly controlled setting, weight-lifting was shown to do little or no harm to patients with lymphedema and at risk patients, Schmitz et al—but weight-lifting does not cure nor definitively prevent lymphedema, and needs to be done slowly and carefully. Read the articles, not the press releases.

The initial article on weight lifting with the PAL protocol for women with lymphedema is a free article on pubmed: <http://www.nejm.org/doi/full/10.1056/NEJMoa0810118>

The second article reviewed use of the protocol with women "at risk" for lymphedema:
<http://www.ncbi.nlm.nih.gov/pubmed/21148134>
http://www.abstracts2view.com/sabcs10/view.php?nu=SABCS10L_9027
"Slowly progressive weight-lifting had no significant impact on arm swelling among breast cancer survivors with lymphedema, and resulted in a reduced incidence of lymphedema flares and improvement in symptoms. In breast cancer survivors at risk for lymphedema, a program of slowly progressive weight-lifting, compared with no intervention, did not result in increased incidence of lymphedema. In secondary analyses, women with 5+ nodes removed who did weight-lifting were less likely to experience increases in arm swelling."

Unfortunately, on the official University of Pennsylvania PAL site: <http://www.penncancer.org/physical-activity-and-lymphedema> This statement will be found: "Several key findings from the study include: 50% reduction of the likelihood of lymphedema worsening among women with lymphedema 70% reduction of the likelihood of arm swelling increases among women who had 5 or more lymph nodes removed"

Please know the facts and limitations of the study. Weight lifting does not cure or prevent lymphedema, and can precipitate it in some women. Read the NLN guidelines and counsel your patients with the facts.

Peri-operative and Post-operative Management:

--Handle the tissues gently during surgery: Janice Cormier, M.D., NLN lecture 2010, Physicians' Intensive. Lecture slide: Critical Surgical Technique 1) Gentle handling of tissues, 2) Orientation of incisions (extremities) and 3) Hemostatic control at time of surgery

--Treat seromas promptly: from San Antonio Breast Conference 2010, "Seroma usually leads to prolonged wound healing, tissue inflammation and subsequent fibrosis and necrosis. It has been hypothesized that seroma formation is one of the main risk factors for post-breast cancer lymphedema." Seroma Formation Is Associated with Lymphedema-Related Symptoms.

http://www.abstracts2view.com/sabcs10/view.php?nu=SABCS10L_478

---Consider limiting stretch on the axillary area for 10-14 days post-operatively, as lymphatics have limited time to regenerate: NLN Conference Lecture, 2010, Jodi Winicour PT

From Foldi Textbook of Lymphology: Lymphatic regeneration occurs as the stumps of the afferent or efferent collectors of a removed node connect as the result of proliferation of the endothelium at the terminal portion of the damaged vessel. Regeneration of superficial vessels in dogs takes 4 days, and deep vessels in 8 days.

Have your patients limit their arm movement to shoulder height for the first 10-14 days post-op to allow the efferent and afferent vessels to connect during the limited time of lymphatic regeneration.

Systematic review of early vs. delayed exercise has shown delayed exercise decreases seroma formation: <http://www.ncbi.nlm.nih.gov/pubmed/15830140>

A study in 2008, published in Physiotherapy, showed higher risk of development of lymphedema in women who had axillary node dissection and performed exercise early vs. delayed exercise: <http://www.lymphoedemaleeds.co.uk/Pages/Research.aspx>

---Axillary Web Syndrome is a risk factor for lymphedema: In a study of early PT to minimize lymphedema, a subset of patients who developed axillary web syndrome at 3-4 weeks post-op had an almost universal development of lymphedema, despite intervention <http://www.ncbi.nlm.nih.gov/pubmed/20068255> BMJ. 2010; 340: b5396. Published online 2010 January 12. "We also found that 12 of the 18 women who developed

secondary lymphoedema had axillary web syndrome during the second and third week after surgery. The axillary web syndrome is a known but poorly studied complication of surgery. No study has shown any link between the axillary web syndrome and the onset of secondary lymphoedema. We and others suggest that the axillary web syndrome may be a sign of injury to the lymphatic system and it could produce a lymphatic overload as a result of failure of the lymphatic system. This overload, together with other factors, could be responsible for the onset of secondary lymphoedema”

Learn about axillary web syndrome: http://www.stepup-speakout.org/Cording_and_Axillary_Web_Syndrome.htm

---Lymphedema Myths and Falsehoods: Learn why they're incorrect

1) You can't get lymphedema from “just” a sentinel node biopsy: the risk of lymphedema after sentinel biopsy is unknown, but is at least 10%. Lymphedema is multilateral, with genetic predisposition, inherent lymphatic pump variations, and inflammation all playing important roles in its development.

2) None of MY patients has ever gotten lymphedema: statistically impossible, and it only indicates that you need to do more careful examinations and take more detailed histories. Lymphedema doesn't equate with poor medical care, and ignoring it will lead to progressive disease. There are behaviors, described above, that can lessen the chances of your patients developing it, but even with optimal medical care, some patients will develop lymphedema.

3) Risk reduction behaviors are myths: they have not been proven with randomized, double blind placebo controlled trials, due to ethical and funding issues, but clinical experience has validated that they are reasonable and should be taught to patients and staff and adhered to as medical practice. Patients remain at risk for life—studies of axillary node dissection patients show that the majority develop lymphedema in the first few years after surgery, and then at least 1%/year develop it, so that the incidence at 20 years of follow up is 50%. The triggering event that overwhelms lymphatic filtration capacity can be minimal and should be avoided.

4) Lymphedema is just cosmetic: it's a derangement of the immune system, causes chronic inflammation and connective tissue derangement. There is the risk of life threatening cellulitis with lymphedema, and the psychosocial issues are considerable. It is a “real” disease, and deserves prompt recognition and treatment by informed medical staff.

5) Why bother diagnosing it, there's nothing that can be done to treat it: Lymphedema therapy is effective and most effective when started early.

In summary: lymphedema is common—even with sentinel node biopsy, and its true prevalence is unknown as there is no diagnostic standard, it is under-recognized, under-treated, and health care providers are under-educated about the disease. Your patients need you to understand the reality of their disease and help them manage it. Lymphedema is a disease of abandonment and misunderstanding. It is NOT just swelling; it is derangement of the lymphatic system, derangement of connective tissue, progressive disability and impaired lymphatic functioning. It is a dreaded complication for very good reasons.

An excellent handout for quick reference: <http://www.livestrong.org/Get-Help/Learn-About-Cancer/Cancer-Support-Topics/Physical-Effects-of-Cancer/Lymphedema>

***The author of this page works in radiation oncology, although trained in primary care. In my practice, the risk of lymphedema is on the consent form for breast irradiation. The practice protocol is to measure all breast cancer patients' arms at every visit, and every week I diagnose lymphedema in compliant patients who see their breast surgeons, medical oncologists and primary care doctors—and I read the notes, which state “no edema”. All the patients are offered treatment and they overwhelmingly accept it. As one patient said, “I didn't know there was anything I could do.” If you don't look, you won't find it, and patients will suffer needlessly. And I don't just diagnose lymphedema in breast cancer patients: patients with sarcoma, endometrial cancer, melanoma, head and neck cancer, rectal cancer and prostate cancer have presented with it as well. Any time medical therapy disrupts the lymphatic system; patients are at risk for lymphedema.

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