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Looking into secondary lymphedema

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LYMPHEDEMA is a congenital (primary) or mechanical (secondary) abnormality in the lymphatic system that results in an abnormal accumulation of interstitial fluid and fibrous tissue in a specific area in a person's body.[1] This article focuses on secondary lymphedema, usually caused by lymph node dissection or lymphadenectomy for cancer staging in conjunction with tumor removal. Recent advances in microvascular surgery are discussed. Primary lymphedema, generally due to congenital or inherited disorders, is beyond the scope of this article.

This article also emphasizes patient teaching. Nurses can help postsurgical patients learn how to prevent lymphedema and recognize the early signs and symptoms of lymphedema or infection. Lymphedema can't be cured once diagnosed and can be challenging to treat in its later stages.[1] Early treatment for lymphedema can minimize complications such as fibrosis, hyperpigmentation, and fissures. Teaching patients how to prevent or manage lymphedema improves their self-esteem and quality of life.[2-4]

Pathophysiology

The edema associated with lymphedema differs from the generalized edema resulting from chronic venous insufficiency or fluid overload. Although both general edema and lymphedema cause edema in the extremity, lymphedema is caused by protein-rich lymph fluid and general edema is caused by tissue-related fluid.[5,6] Lymphedema can occur when the lymphatic load exceeds the lymph system's transport capacity, which causes filtered fluid to build up in the interstitium. Secondary lymphedema results from a disorder or treatment, such as cancer or treatment for cancer such as axillary node dissection, infection, inflammatory disease, obesity, trauma, burns, and chronic venous insufficiency.[1,6]

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The most common cause of secondary lymphedema is surgical intervention for a malignancy when lymph node dissection is performed.[1,6] Radiation therapy for cancer is another common cause.

Most common sites for lymphedema

Most cancer surgeries require lymph node removal or, at the least, lymph node sampling, also called lymph node biopsy. In larger areas such as the abdomen and the chest, other parts of the lymphatic system such as the thoracic or lymphatic duct can help with collateral drainage, and generally more room for edema is found in the abdomen.[7] The areas of the body that most commonly demonstrate lymphedema are the upper extremities, related to the axillary lymph nodes; the lower extremities, related to the inguinal lymph nodes; and the neck, related to cervical and supraclavicular lymph nodes.[1,6] (See *Normal anatomy and physiology of the lymphatic system*.)

Incidence

Lymphedema is seen more frequently in females than males.[1] The incidence of upper extremity lymphedema for breast cancer survivors is 17%, with the greatest incidence within the first 2 years after breast cancer diagnosis. The National Cancer Institute reports that women who've undergone treatment for breast cancer may experience lymphedema within days of treatment or up to 30 years posttreatment.[8] (See *Lymphatic drainage of the breast*.)

Lower extremity lymphedema occurs most often in patients who've undergone treatment for uterine or prostate cancer, lymphoma, or melanoma.[6]

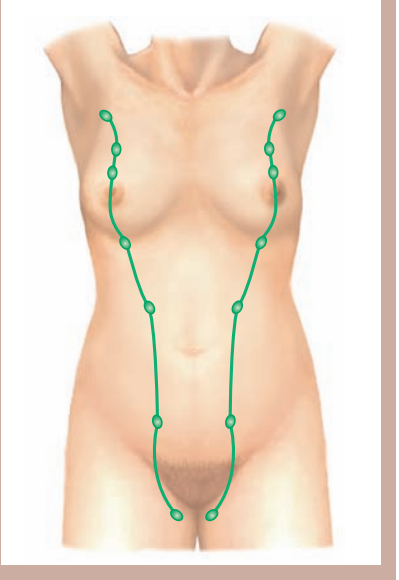
Risk factors

The main risk factor for secondary lymphedema is a lymph node dissection as part of surgical cancer treatment, most commonly after a modified radical mastectomy or a lumpectomy.[6,8] Because clinicians have been using more breast-conserving techniques in the past decade, many patients are having a lumpectomy and a sentinel lymph node biopsy of just the first few nodes that drain the tumor being removed. Statistics show that these patients can still develop lymphedema, but at a lower frequency—5% for sentinel lymph node dissection versus 19% for axillary lymph node dissection. Patients having less radical surgeries must still follow lymphedema precautions.[9]

A surgical resection of an upper body melanoma sometimes requires an axillary node dissection. Lower extremity lymphedema can be caused by inguinal lymph node dissections after a resection for a melanoma of the lower extremity or extensive gynecologic procedures.[6,8,10,11]

Normal anatomy and physiology of the lymphatic system

The lymphatic system is an extensive vascular network that drains lymph fluid from body tissues and returns it to the venous circulation. The system starts peripherally as blind lymphatic capillaries, continues centrally as thin vascular channels, then as collecting ducts, and empties into the major veins at the neck. Lymph fluid transported through these channels is filtered through lymph nodes interposed along the way.



Lymph nodes are round, oval, or bean-shaped structures that vary in size according to their location. Some lymph nodes, such as the preauricular nodes, if palpable at all, are typically very small. The inguinal nodes, by contrast, are relatively larger—often 1 cm in diameter and occasionally even 2 cm in an adult.

In addition to its vascular functions, the lymphatic system plays an important role in the body's immune system. Cells within the lymph nodes engulf cellular debris and bacteria and produce antibodies.

Only the superficial lymph nodes are accessible to physical assessment. These include the cervical nodes, the axillary nodes, and nodes in the arms and legs.

Recall that the axillary lymph nodes drain most of the arm. Lymphatics from the ulnar surface of the forearm and hand, the little and ring fingers, and the adjacent surface of the middle finger, however, drain first into the epitrochlear nodes. These are located on the medial surface of the arm approximately 3 cm above the elbow. Lymphatics from the rest of the arm drain mostly into the axillary nodes. A few may go directly to the infraclavicular nodes. The lymphatics of the lower limb, following the venous supply, consist of both deep and superficial systems.

The superficial inguinal nodes include two groups. The horizontal group lies in a chain high in the anterior thigh below the inguinal ligament. It drains the superficial portions of the lower abdomen and buttock, the external genitalia (but not the testes), the anal canal and perianal area, and the lower vagina.

Source: Bickley LS. *Bates' Guide to Physical Examination and History Taking*. 11th ed. Philadelphia, PA: Wolters Kluwer Health/Lippincott Williams & Wilkins; 2013.

Lymph node dissections are often performed during head and neck surgeries. Neck lymphedema can be very discouraging, especially after an already-disfiguring procedure.[12] (See *Head and neck lymph nodes.*)

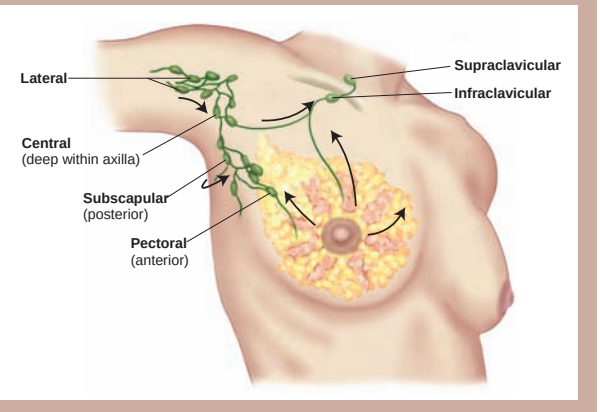
Other risk factors for lymphedema include tumor compression of the lymphatic system in the axilla, the neck, the groin, the abdomen, or the chest. The risk of lymphedema is increased when the patient has large tumors, which may compress the lymphatic system, and in later stages of disease, when the patient has positive lymph nodes because larger tumors tend to be associated with more positive lymph nodes.[1,8,13]

External beam radiation can cause fibrosis and muscle wasting, leaving the lymphatic system functioning improperly. Obesity is another risk factor because parts of the lymphatic system can be blocked by excessive adipose tissue.

Cellulitis is a localized skin infection that can lead to lymphedema, and long-standing repeated exacerbations of lymphedema can lead to recurrent incidents of cellulitis.[14]

Lymphatic drainage of the breast

Lymphatics of most of the breast drain toward the axilla. Of the axillary lymph nodes, the *central nodes* are most likely to be palpable. They lie along the chest wall, usually high in the axilla and midway between the anterior and posterior axillary folds.



Into them drain channels from three other groups of lymph nodes, which are seldom palpable:

- *Pectoral nodes—anterior*, located along the lower border of the pectoralis major inside the anterior axillary fold. These nodes drain the anterior chest wall and much of the breast.
- *Subscapular nodes—posterior*, located along the lateral border of the scapula; palpated deep in the posterior axillary fold. They drain the posterior chest wall and a portion of the arm.
- *Lateral nodes*—located *along the upper humerus*. They drain most of the arm. Lymph drains from the central axillary nodes to the *infraclavicular* and *supraclavicular* nodes.

Not all the lymphatics of the breast drain into the axilla. Malignant cells from a breast cancer may spread directly to the infraclavicular nodes or into the internal mammary chain of lymph nodes within the chest

Source: Bickley LS. *Bates' Guide to Physical Examination and History Taking.* 11th ed. Philadelphia, PA: Wolters Kluwer Health/Lippincott Williams & Wilkins; 2013.

Signs and symptoms

Clinical manifestations of lymphedema related to axillary node dissection include complaints of jewelry or clothing being too tight; a thick, heavy, or full feeling in the affected arm; pain; edema; decreased flexibility in the elbow or shoulder area; and feeling that it's an effort to lift things or write using the affected limb.[1,3,8]

At first, peripheral lymphedema may present as pitting edema or cellulitis. (See *Picturing lymphedema.*) It could be related to an injury or an infection. Lymphedema can involve patients' fingers, making it very hard to put on rings or even form a fist.[3]

If not diagnosed and treated properly, lymphedema evolves into nonpitting edema and fibrosis. The patient feels that the arm is heavier, and it's firm to palpation.[1,5]

The skin of an area with lymphedema that's hyperpigmented and thickened is very fibrotic. If the lymphedema is left untreated, the involved area can be at high risk for breakdown and fissures.[1]

With lower extremity lymphedema, patients may say that their clothes, the elastic on their underwear, or shoes feel too tight. Some patients have difficulty ambulating.[15]

Picturing lymphedema

Lymphedema is soft in the early stages, then becomes indurated and nonpitting. Skin is markedly thickened; ulceration is rare.

Source: Bickley LS. *Bates' Guide to Physical Examination and History Taking.* 11th ed. Philadelphia, PA: Wolters Kluwer Health/Lippincott Williams & Wilkins; 2013.

Assessment

Because the onset of lymphedema is subtle, clinicians should perform ongoing assessments of high-risk patients at every clinical evaluation. This includes comparing the involved extremity with the contralateral extremity. Measuring limb

circumference and tracking those measurements is a general method used by clinicians that's also taught to patients.

When comparing two extremities, the same areas should be measured every time. The joints of the fingers and toes should be measured if they're edematous. The wrist and/or the ankle should be measured as well as 10 cm above and below the elbow and/or 10 cm above or below the patella. Many patients are followed by physical therapists (PTs) or occupational therapists (OTs) at a facility for these ongoing measurements.[1] (See *Staging lymphedema.*)

Staging lymphedema

Lymphedema can be categorized by using the National Cancer Institute Common Terminology Criteria for Adverse Events. The stage is based on assessment findings and the degree of functional impairment, which are generally the clinical stages listed here.

- Grade 1–Trace thickening or faint discoloration
- Grade 2–Marked discoloration, leathery skin texture, papillary formation, limiting instrumental activities of daily living
- Grade 3–Severe signs and symptoms limiting self-care and activities of daily living.

Source: Mohler ER III, Mehrara B. Clinical staging and conservative management of peripheral lymphedema. Up To Date; 2017. www.uptodate.com.

Diagnosis

Early diagnosis is critical so that treatment will be as effective as possible. Clinicians should monitor high-risk patients by obtaining an accurate health history and performing a comprehensive physical assessment including circumference measurements of the affected limb and the contralateral limb.[1]

Lymphedema is often diagnosed using radiologic imaging.[6] Lymphedema may not be noticeable, and it can even be microscopic.[2,10]

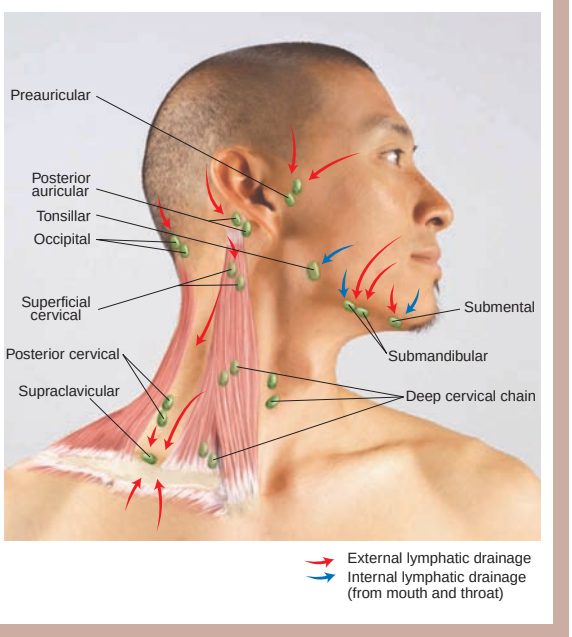
Ultrasound can assist in differentiating lymphedema from other vascular disorders. Computerized tomography or magnetic resonance imaging can help to determine if fluid is in the peripheral soft tissues.[1] Lymphoscintigraphy images the flow of fluid from the skin to the lymph nodes, especially in the extremities.

Bioimpedance analysis uses resistance to electrical current in comparing the composition of body fluid compartments to help detect lymphedema.[6]

A newer method of diagnosis involves an off-label use of indocyanine green (ICG) for lymphangiography. ICG is injected between the fingers, and infrared light near the skin demonstrates the lymph flow. Patients with lymphedema don't have good linear flow or have a dim-looking flow.[16] This exciting new testing is dynamic, allowing lymphedema to be diagnosed in real time.[1]

Head and neck lymph nodes

The lymph nodes of the head and neck are classified in a variety of ways. One classification is shown here, together with the direction of lymphatic drainage. The deep cervical chain is largely obscured by the overlying sternomastoid muscle, but at its two extremes, the tonsillar node and supraclavicular nodes, may be palpable. The submandibular nodes lie superficial to the submandibular gland, and should be differentiated. Nodes are normally round or ovoid, smooth, and smaller than this gland. The gland is larger and has a lobulated, slightly irregular surface. Note that the tonsillar, submandibular, and submental nodes drain portions of the mouth and throat as well as the face.



Source: Bickley LS. *Bates' Guide to Physical Examination and History Taking.* 11th ed. Philadelphia, PA: Wolters Kluwer Health/Lippincott Williams & Wilkins; 2013.

Noninvasive therapies

Patients with peripheral lymphedema should avoid using an affected limb to lift or carry heavy objects. Patients with lower extremity lymphedema shouldn't cross their legs for extended periods. Because signs and symptoms are more common in those with an increased body mass index, weight management is important.[6]

Compression, the main conservative treatment for lymphedema, can be obtained with compression garments or compression bandages that wrap around the extremity. Intermittent pneumatic compression is used to achieve an optimal

amount of pressure. One benefit is that patients can be taught to use this therapy themselves at home.[2] Although compression is usually provided by PTs or OTs who've been specially educated in lymphedema management, nurses can also be educated.[2,17]

Sleeve and leg garments can have layers of compression material. Patients are measured and garments are sized for their original limb size plus the amount of lymphedema. Smaller garments may be used as the lymphedema is reduced. Although compression garments and bandages may feel tight and constricting at first, they're the first line of treatment for this disorder.[6,17-19]

During the first level of compression therapy, patients are fitted with layers of bandaging or garments; the greatest amount of pressure that patients can tolerate is best for decreasing the edema. In the second level of therapy, patients use garments fitted to their present or maintenance level of edema to maintain that level during regular high-risk activities such as a plane flight or rides in amusement parks that put the limb under a high degree of pressure and can exacerbate lymphedema.[2,5,20] In general, edema can be reduced by approximately 31% with compression bandages and 46% with garments.[5]

Daily moisturizing is imperative under garments or bandages and when the patient has edema. Using moisturizers, preferably water-based, maintains the skin's integrity, prevents fissures, and avoids infection.[3,5]

Some patients may have to use additional dressings to collect drainage or protect the skin. Once the lymphedema lessens, patients may wear the garments occasionally to keep the lymphedema under control or to participate in high-risk activities as mentioned above.[2,20]

Another method of treating lymphedema is manual lymphatic drainage (MLD) therapy, which is also often performed by specially trained PTs or OTs, nurses, and massage therapists. MLD uses mostly massage and light pressure to assist in the manual drainage of lymphatics. The fluid is moved from the distal or most peripheral area proximally into other areas where the lymphatics are functioning properly. The patient's compression garments should be placed after MLD for the most effective treatment.[2] Together the two techniques have produced as much as a 60% decrease in edema.[5,17]

Lymphedema can't be cured once diagnosed and can be challenging to treat in its later stages.

Complete decongestive therapy is similar to MLD with very thorough manual lymphatic drainage and the use of compression bandages and garments to follow. Excellent skin care is stressed for the prevention of infection and irritation from the garments. As with MLD, patients are taught to continue to use their compression garments and practice optimal skin care at home.[2]

No pharmacologic agent has shown any positive results in treating lymphedema although research on anti-inflammatory and antifibrotic agents continues.[21,22]

Surgical options

Indications for surgery include exhaustion of conservative measures such as compression and complete decongestive therapy. Candidates for surgical management include patients with recurrent cellulitis, pain, disfigurement, and localized primary lesions.[23] Surgical management involves physiologic or reductive techniques.

Physiologic techniques reroute lymphatic drainage by creating new conduits for the fluid so that it can empty into different lymphatic drainage areas or the venous circulation.[23] Lymphovenous bypass procedures, performed by a surgeon trained in microvascular surgery, is the most commonly used physiologic approach to surgical management of lymphedema. The approach to bypass surgery can be either lymphatic-lymphatic, lymphovenous, or lymphaticovenular.[23]

- Lymphatic-lymphatic bypass transfers healthy lymph tissue that's microsurgically anastomosed to the affected tissue.[23]
- The lymphovenous bypass uses a vein graft to connect distal lymph vessels with vessels proximal to the obstruction.[23]
- Lymphaticovenular anastomosis connects distal subdermal lymphatic vessels to adjacent venules; the pressure in the subdermal venules is lower than in deeper veins, creating less venous backflow to help improve lymphedema.[23]

Reductive techniques remove the fibro-fatty tissue generated by prolonged lymphatic fluid pooling and are considered palliative, not

curative, for secondary lymphedema. The goal is to alleviate signs and symptoms.[23] Reductive surgery includes direct excision and liposuction.

Direct excision is most often used for genital and extremity lymphedema, with excised areas often covered with a skin graft or tissue flap, which functions more effectively for patients who will retain excessive skin following the procedure or following liposuction. Liposuction with specialized suction cannulas, which can be used to remove some of the subcutaneous fatty tissue, may improve the cosmetic appearance of the limb.[5]

Postoperative care and complications

Following lymphovenous bypass procedures, keep the affected limb elevated during patients' entire hospital stay. Frequently monitor circulation, sensation, and movement of the affected limb. Elastic support bandages will be in place for the first 24 to 48 hours postoperatively. Garments aren't usually used on the limb immediately postoperatively because of the risk of skin shearing; however, once the surgical site is stabilized, compression garments are used until clinicians and patients determine that the surgery was successful enough to discontinue their use.[23,24]

After a direct excision procedure, most patients have a drain in place. Teach patients drain care, including how to empty the drain and record the amount and characteristics of drainage; perform dressing changes; and care for the skin around the drain. Many patients have long hospital stays and poor wound healing after the direct excision of skin and fat.[23] Teach patients the signs and symptoms of infection and tell them to notify their healthcare provider immediately in case of any problems, such as fever or purulent drainage from the incision site or drain.

Early treatment for lymphedema can minimize complications such as fibrosis and fissures.

After liposuction procedures, patients are at high risk for bleeding due to the large volume of fat and adipose tissue that's been removed, but the use of tourniquets and epinephrine during the procedure reduces the risk.[23] Monitor the patient for signs and symptoms of bleeding postoperatively. Compression garments will help to prevent bleeding. The assistance of the lymphedema specialist (usually the PT or OT) will most likely be required because the garments will probably need to be resized. Patients will need to continue to wear compression garments or bandages for the long term.

Postoperative management includes using compression garments immediately after the liposuction procedure and up to 12 months afterward. Prophylactic antibiotics may be prescribed for up to 1 year after surgery and excellent skin care, including moisturizing, will be needed because of the high risk of infection.[23]

Cellulitis is a common issue for patients with lymphedema. Patterns of recurrent and increasingly worse cellulitis can occur in 23% to 35% of patients with lymphedema.[3,16] Teach patients the vital importance of skin hygiene and how to monitor for early signs and symptoms of infection.

Lymphangiosarcoma is a rare but serious complication of chronic lymphedema. Seen most often in patients who've had a mastectomy, this secondary malignancy most often affects patients with massive and protracted lymphedema.[25]

Patient education and quality of life

The overall goals for patients vulnerable to lymphedema are prevention and early detection and treatment.[26] Patient education is crucial in achieving these goals. Patients need to understand how to avoid engaging in high-risk behaviors and advocate for themselves by warning healthcare workers to avoid risky practices whenever possible. These include BP measurements and venipunctures in the affected limb. Patients should also be taught to recognize the signs and symptoms of lymphedema and to seek care from their oncologist promptly if they appear.

Lifestyle modifications and self-care are essential to prevent lymphedema or manage symptoms. A 2016 study found that 85% of patients with head and neck lymphedema practiced self-care activities.[27] Patients in this study reported on the importance of in-person interaction, lymphedema education before treatment, follow-up evaluation, and visualization of self-care. Teach

patients with lymphedema or at risk for it that they'll need to continue these self-care measures for their entire lifetime.

To optimize their health and meet treatment goals, patients should strive to balance physical activity with rest. Encourage patients to be active, as tolerated, and to avoid a sedentary lifestyle.

An infection in a person with lymphedema or at risk for it can potentiate or worsen the lymphedema. Teach patients about proper precautions, as discussed below, such as good skin care and hygiene to help prevent infection. Meticulous skin care is vital in the postoperative period; skin should be well moisturized and cleaned daily. Patients should use gentle, nonalcoholic, water-based creams or emollients to prevent cracked or dry skin. Encourage patients with lymphedema to use an electric razor; avoid insect bites; cut nails straight across or see a podiatrist for nail care; avoid cuticle cutting; and be aware of the early signs and symptoms of infection such as new onset of edema, erythema, or warmth.[3-5,14]

Patients should avoid anything that applies pressure to the affected limb, including tourniquets used for venipuncture. Everyday precautions include avoiding constricting jewelry, such as tight bracelets or watches, and gloves. Patients with lower extremity lymphedema should avoid clothing such as socks and pants with elastic bands. Women with upper extremity lymphedema should wear a loose-fitting bra to avoid pressure to the axillary region.[1,3,4]

Air travel can trigger or worsen lymphedema. For example, lower air pressure during flights may allow pooling in the lymphatic system.[2,14] Wearing a compression garment during the flight may help offset this risk. However, a recent study suggests that air travel for patients with upper extremity lymphedema isn't

Patients with lymphedema or at risk of it should continue self-care measures for the rest of their lives.

associated with increased extremity volume.[20] Sitting still for prolonged periods is another risk; patients should be sure to walk and stretch frequently during their flight. Deep abdominal breathing may also help facilitate lymph flow. Lifting and carrying luggage can also trigger or worsen lymphedema; patients should seek assistance with their luggage.

Patients should monitor for signs and symptoms of lymphedema, including limb heaviness, weakness, or aching; skin tightness, pain, or numbness; and an impaired range of motion. Any of these warrants prompt evaluation by a healthcare provider.

Most of the quality of life research on lymphedema has studied breast cancer survivors, but even that research is limited. Lymphedema is a chronic disease. After patients have completed disease treatment and are feeling better about themselves, lymphedema can occur, affecting their quality of life.[28,29]

Wrapping up

For patients who develop lymphedema, the primary goal is early diagnosis and treatment to get them back to baseline as soon as possible. Patient education is essential to help them resume their usual activities and improve their overall quality of life. Nurses' knowledge and care can make a difference in these patients' lives. ■

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Lippincott Professional Development is also an approved provider of continuing nursing education by the District of Columbia, Georgia, and Florida CE Broker #50-1223. This activity is also provider approved by the California Board of Registered Nursing, Provider Number CEP 11749 for 1.5 contact hours.