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Lymphedema Management

Introduction

Lymphedema is a chronic condition that involves an excess of swelling in one or more extremities due to a blocked or otherwise impaired lymphatic system. With up to 10 million Americans living with lymphedema and other forms of lymphatic dysfunction, healthcare providers must have an understanding of this condition and how it comes about. Occupational therapists are just one type of healthcare professional who can assist individuals in rehabilitating from lymphedema and similar disorders. Lymphedema has the potential to impact a person's strength, range of motion, endurance, and other client factors. Individuals with lymphedema may also experience impaired ability to participate in occupational areas such as work or school, leisure, sleep, ADL function, IADL function, and more. The physical effects of lymphedema can cause someone to experience concerns related to body image, self-esteem, hygiene, and quality of life. For these reasons and more, lymphedema is an important area of specialization within the field of OT since it requires unique treatment and expertise on behalf of the therapist.

Section 1: Background

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In order to understand how lymphedema is treated and the impact it can have on a person, it's important for all occupational therapists to know the anatomical structures and functions that are affected by this condition. As you might have guessed from the name, lymphedema primarily impacts the lymphatic system. While the lymphatic system has several duties, it is most often likened to a drainage system that operates one way, since its chief job is to transport fluid from bodily tissues to the bloodstream. This colorless, water-like fluid is also known as lymph. Lymph is primarily made of cell waste that will later be eliminated from the body, including damaged cells, cancerous cells, and foreign bodies such as viruses

and bacteria. Lymph also includes minerals, nutrients, white blood cells, fats, and proteins. Most often, traces of fats and proteins come from the digestive system and are called chyle. The other substances often end up a part of lymph after having leaked from blood vessels into tissues.

The lymphatic system has three main functions, including regulating the levels of fluids throughout the body; helping the body absorb, store, and move fat from the digestive tract to the bloodstream; and boosting the immune system. The lymphatic system aids a person's immune function by creating, storing, and transporting white blood cells. Since white blood cells play an important part in someone's ability to fight off diseases, infections, and other types of illness, lymphatic organs are considered an integral part of the immune system. It may seem like there is a lot of overlap between the lymphatic system and the immune system, and this is because the lymphatic system is a big part of the immune system. Organs that are part of the lymphatic system include the spleen, bone marrow, thymus, lymphatic vessels, and lymph nodes. The immune system encompasses these same organs along with the tonsils, parts of the bowel, skin, and mucous membranes found in the genitalia, throat, nose, and bladder.

The spleen is the largest of all the organs in the lymphatic system. This organ helps the body fight infection and regulate the amount of blood cells (red, white, and platelets) in the body. Bone marrow, which is the pulpy center of bone, is what creates stem cells that later become red blood cells, white blood cells, and platelets. The thymus modifies stem cells (called T-cells) to become white blood cells. Interestingly enough, the greater majority of this process takes place before a child is even born and the rest of a person's T-cells are specialized by the end of puberty. So, as important as the thymus is for someone's immune system, it does not serve much purpose for adults.

Lymphatic capillaries are thin tubes that carry lymph throughout the body. They have closed ends and are slightly wider than blood capillaries, which are the smallest class of blood vessels that connect veins to arteries. Within lymphatic

capillaries, fluid can only flow forward through a series of valves and is not able to leave through the cell wells. Lymph exits lymphatic capillaries by flowing into larger lymphatic vessels that eventually empty into ducts. Lymph nodes are small clumps of tissue that contain white blood cells. There are hundreds of lymph nodes found throughout the body – namely in the chest, underarm, neck, abdomen, and groin – and they are considered one of the body's first lines of defense against infection. Lymph nodes act as filters that sift through substances in the lymphatic fluid. This filtering process also helps prevent fluid from building up in tissues. Each group of lymph nodes throughout the body is assigned to filter through fluid from lymphatic vessels in a certain region. It is common for lymph nodes to swell periodically when the body is fighting an infection, but it could point toward cancer if they are swollen for a long period of time.

Lymphedema vs. Edema

Lymphedema is a chronic condition characterized by significant swelling (or accumulated lymph fluid) that results from damaged or blocked lymph nodes. Lymphedema results when a person's lymphatic system is too overloaded to correct the fluid imbalance on their own or the damage to the lymphatic system is unable to be reversed. Edema is also the result of accumulated lymph fluid in a certain part of the body. However, it is not usually severe or significant and, in many cases, it's considered part of the body's natural defenses. Most people experience edema as a normal bodily response to an infection, injury, or surgery. When the body senses one of these concerns, it sends a rush of white blood cells and fluid to the area to assist with healing. This is what causes visible swelling. Someone may also develop edema more often as they age, due to static postures such as sitting or standing for too long, as a result of dietary habits such as eating too much salt, or due to medical conditions such as obesity or pregnancy. Edema (also known as inflammation) is typically acute or short-lived and usually resolves on its own. It's also common for edema and lymphedema to present or feel similarly in the early stages. Patients with either edema or lymphedema may

notice increased fatigue, a new heaviness in certain body parts, or abnormal sensations in their limbs.

It is possible for edema to be a symptom of circulatory disorders such as congestive heart failure, deep vein thrombosis, and chronic venous insufficiency. When this occurs, edema usually appears in the lower limbs because of the impact gravity has on fluid movement. In these cases, the edema will need to be addressed and the best way to do this is by treating the condition that is causing it. Chronic edema is characterized as lasting longer than three months and occurs when someone is unable to independently rebalance the fluid levels in their body.

Some differential diagnoses for lymphedema include cellulitis, CLOVES syndrome, erysipelas, lymphangioma, lipedema, and thrombophlebitis. Each of these conditions can potentially have symptoms similar to lymphedema. Cellulitis is a non-contagious bacterial infection that causes severe swelling, redness, and tenderness. Cellulitis most often impacts the legs and arms. If cellulitis is not diagnosed and treated soon enough, it has the potential to spread to the lymph nodes. This causes the condition to become much more severe and further mimic the presentation of lymphedema. Lipedema is characterized by abnormal fat build-up in the legs and, in some cases, the arms along with pain and bruising in these areas. This condition results in fat developing unevenly under the skin of the inner and outer legs and buttocks. People with lipedema often appear to have an excess of swelling, which is why typical treatments for edema management are not effective. The feet are not affected by lipedema, which is a tell-tale sign that differentiates this condition from lymphedema.

Erysipelas is an infection of the inner layer of the skin (called the dermis), which may also affect the superficial structures of the lymphatic system within the skin. This infection causes patches of redness on the skin that present like a rash, also called demarcated erythema. Erysipelas most often impacts the legs, which is a common body part affected by lymphedema, but it may also present on the face. Lymphangiomas are benign cysts that typically develop during childhood.

Lymphangiomas are filled with fluid and often form on the head and neck. These fluid-filled cysts are created from a backup of lymphatic fluid but, unlike with lymphedema, they do not typically require treatment because they do not grow large enough to cause functional concerns. Doctors can surgically remove them if they become painful or overly swollen.

Congenital lipomatous overgrowth, vascular malformations, epidermal nevus, and spinal and skeletal scoliosis anomalies syndrome, also known as CLOVES syndrome, is a congenital condition that may initially be mistaken for lymphedema. Since this condition is very rare, it makes sense that physicians may assume CLOVES symptoms such as vascular and skin abnormalities are attributed to a somewhat more common condition such as lymphedema.

For these reasons, it is important to seek help from a lymphedema specialist for proper diagnosis and treatment. This will lead to the best prognosis regardless of the severity of the condition.

Section 1 Personal Reflection

Based on the basic differences between lymphedema and edema, would conservative management strategies for edema help a person with lymphedema?

Section 1 Key Words

Differential diagnosis - The act of healthcare providers (usually physicians) completing tests to compare the symptoms and signs associated with at least two conditions to make a diagnosis

Section 2: Lymphedema Staging, Etiology, & Risk Factors

9,10,11,12,13,14,15,16,17,18,19,20,21,22,23,24,25,26,27,28,29

There are four stages of lymphedema. The process of determining what stage a person is experiencing is called staging. Individuals with Stage 0 lymphedema begin to experience abnormal flow in the lymphatic system, but they do not demonstrate any visible symptoms. For this reason, Stage 0 is also called the latent or subclinical phase. Some people with Stage 0 lymphedema may report feelings of skin tightness, heaviness, or fullness in the affected limb or area. Other sensation changes, including burning or itching of the skin, may result during this stage. Those with Stage 0 lymphedema have typical results when their limb or body measurements are taken. Clinically, these individuals are considered normally functioning, but providers can see the abnormalities in their lymphatic system through a test called lymphoscintigraphy. While people may remain in this stage for up to a few years before progressing to the next, they are considered at high risk due to the prognosis of this condition.

Stage 1 lymphedema (also known as the mild stage) is characterized by slight fluid accumulation and swelling in the affected limb or area. This swelling occurs occasionally and may cause slight stiffness. If the affected area is an arm or leg, the swelling can be managed by elevating the limb. It is common for individuals in this stage to experience pitting edema, which means the affected area is soft and will be temporarily left with a small dimple or pit when someone presses a finger on it. During this stage, individuals with lymphedema often experience an increase in existing sensation changes or new ones altogether. This may include itching, burning, and feelings of numbness and tingling (also known as pins and needles). During this phase of lymphedema, individuals have between 1 and 3 cm of swelling in the affected body parts.

As part of Stage 2 lymphedema (also known as the moderate stage), individuals will experience greater levels of swelling that do not respond to elevation. If the skin was not already tighter in Stage 1, it will become this way during Stage 2. Pitting edema will become much more obvious at this point. However, in late Stage 2, pitting may not occur due to the development of hardened skin, tissue, and subcutaneous fat. This is called fibrosis or fibroadipose deposition, and occurs

as a result of excess, unorganized collagen. At this point, an individual's lymphedema has become irreversible and will not improve without treatment. During Stage 2, individuals may experience bursting or shooting pain in the affected area. During this phase of lymphedema, individuals have between 3 and 5 cm of swelling in the affected body parts.

Stage 3 lymphedema is considered severe due to the presence of extensive swelling. Pitting no longer takes place, since the skin has become very thickened. Individuals may also notice changes to their skin's color and texture. These can include a brownish tint called hyperpigmentation, a leathery feel to the skin, fat deposits, significant dryness, and added skin folds along with the formation of blisters or papillomas, which are similar to warts. Papillomas may leak or ooze fluid, which places someone at a high risk of infection and deep, persistent wounds. It is possible for individuals to experience relief from symptoms and drop down to a less severe stage if they receive treatment. If someone's limbs are affected by Stage 3 lymphedema, they will likely have a lot of difficulty moving their arm or leg on their own and may also experience discoloration of the nails on their hand(s) or feet. Providers identify a person's lymphedema stage based on the aforementioned symptoms, but also by the severity of the condition. If someone's extremity volume increases by less than or equal to 20%, they have mild lymphedema. When this number enters the 20% to 40% range, their condition is considered moderate. If their extremity volume increases by 40% or more, their condition is labeled severe. In order to meet the criteria for severe lymphedema, individuals must have 5 or more centimeters of swelling in the affected body part.

The majority of lymphedema cases involve the arms or legs, with most people experiencing swelling and other symptoms in one of these extremities (e.g. one arm or one leg). It is more uncommon for someone with lymphedema to experience symptoms in both the arms and the legs. Lymphedema can also impact the chest wall, neck, face, trunk, abdomen, and genitalia. If an individual has a diagnosis of lymphedema that causes abdominal swelling, this is most often

due to lymphedema of the intestines. This type of lymphedema may also cause digestive symptoms such as diarrhea and an inability to digest fatty foods.

Studies show that, in general, lymphedema symptoms often worsen in response to menstruation, warm or humid weather, or a body area being in a dependent position. This is a natural position for most people, but it can cause an increase in swelling for some individuals with lymphedema. In particular, this is not a recommended position for those with lymphedema of the arms or legs, since it involves the arm hanging down by the side of the body while someone sits or stands and the leg in a fixed position with the foot flat on the ground. Lymphedema-related swelling also gets worse when someone exerts that body part or limb too much, does not change positions often enough, and is inactive for too long. Swelling typically increases throughout the day and eases overnight.

Additional symptoms of lymphedema include hair loss and difficulty sleeping. Individuals with early-stage lymphedema may notice a periodic increase in flu-like symptoms such as fever, body chills, and generalized weakness. This is common since the fluid buildup can trigger an immune response as the body tries to compensate for a weak lymphatic system. If someone with lymphedema experiences a fever in addition to prolonged warmth, tenderness, or redness around their swelling, this may point toward an infection. Infections are a complication of lymphedema and must be identified and treated immediately for the best results.

Signs of lymphedema that someone without a clinical background may notice include:

- Difficulty putting on clothing, bras, and jewelry due to a more snug fit
- Inability to feel or see the veins or tendons in the hands or feet (in lymphedema of the limbs)
- Less pronounced knuckles (in lymphedema of the arms)

- Less pronounced markings on the knee (in lymphedema of the legs)
- Gradual weight gain as the swelling progresses
- Smoothing of natural skin wrinkles in the early stages
- Presence of skin folds in the later stages

Causes of Lymphedema

The causes of lymphedema differ based on the type of lymphedema someone develops. There are two main types of lymphedema: primary and secondary.

Primary Lymphedema

Cases of primary lymphedema are rare and considered to be genetic. It is estimated that 1 in 6,000 to 10,000 people are living with primary lymphedema, which is characterized by a partially-absent or abnormally developed lymphatic system. Aplasia is the term used to describe an organ, limb, or other bodily structure that never develops. This phrasing may be attributed to primary lymphedema that involves the absence of lymph nodes or other lymphatic structures in certain parts of the body. Hypoplasia and hyperplasia are the two other reasons for primary lymphedema. Hypoplasia is when portions of a bodily system (in this case, the lymphatic system) do not fully develop and hyperplasia describes bodily systems that are larger than normal. Hyperplasia causes oversized lymphatic structures, which prevents them from working properly. Primary lymphedema may also develop as a result of scarred, hardened lymph nodes, also known as fibrosis.

Primary lymphedema may arise at various stages in a person's life, which determine the type of diagnosis someone carries. Some people develop primary lymphedema during infancy, at which point it appears as a condition called Milroy's disease. This form of primary lymphedema is also called congenital lymphedema, as it may even be present at birth. Milroy's disease accounts for 10-25% of all primary lymphedema cases. When someone develops primary

lymphedema between the ages of 2 and 35, this is considered lymphedema praecox. Lymphedema praecox is four times more common in women than in men, and develops once women reach puberty or become pregnant. 65 to 80% of individuals with primary lymphedema carry a diagnosis of lymphedema praecox. Research shows that about 70% of individuals who develop lymphedema praecox experience lymphedema unilaterally in the lower extremity. Most people affected by lymphedema praecox possess less lymphatic ducts than individuals without the condition. Meige's disease is the autosomal dominant form of lymphedema praecox. This means that Meige's disease can be passed down from parent to child through a mutated gene that causes the condition. Individuals who develop Meige's disease have about a 50% chance of inheriting that gene from their parents. Individuals who develop primary lymphedema after the age of 35 are considered to have late-onset lymphedema, also known as lymphedema tarda or tardum. Less than 10% of individuals with primary lymphedema have this form of the condition, which exclusively causes leg swelling. While most physicians still diagnose lymphedema tardum through a process of elimination, recent technology has allowed them to identify genetic mutations that are the root cause of this lymphedema subtype.

Secondary Lymphedema

It is estimated that 1 in 1,000 individuals have secondary lymphedema, sometimes known as acquired lymphedema. Secondary lymphedema primarily impacts adults, and occurs in less than 1% of children. This is the more common of the two lymphedema types, which is caused by one or more lymphatic system injuries, traumas, or blockages. Some of the most well-known causes of secondary lymphedema include:

- Malignant cancer and cancerous tumors, since they block lymphatic vasculature
- Surgery of any kind, since this causes scarring that can impact lymphatic structures

    - Surgeries involving veins, blood vessels, and lymphatic or other vasculature – such as excisions of burns or scars, axillary node dissections, vein stripping, pelvic floor surgery, lipectomies, sentinel lymph node biopsies, and surgery of the peripheral vascular system – place someone at any especially high risk of lymphedema.
    - It's a myth that the only cancer surgery someone can experience lymphedema after is an axillary node dissection and not a sentinel lymph node biopsy. While someone's risk of lymphedema is higher (at around 19-25%) if they undergo an axillary node dissection, they still have a 2-8% chance of developing lymphedema after a sentinel lymph node biopsy.
    - Someone's lymphedema risk is not dependent on how many lymph nodes they had surgically removed. Individuals who had one or two lymph nodes removed for any reason do not necessarily have a lower risk of developing lymphedema compared to someone who had multiple lymph nodes removed. The extent of scarring that surgery causes in and around the lymphatic system is a bigger indicator of their lymphedema risk.
- Cancer treatment, especially radiation treatment and surgical removal of cancer
    - It's most common for lymphedema to develop after treatment for breast cancer; skin cancer; cancer of the head and neck; cancer of the cervical, vulvar, or other gynecological areas; cancer of the prostate, penis, or other genitourinary areas; and lymphoma, since each of these areas either house or are in close proximity to clusters of lymph nodes.
- Traumatic injuries, such as car accidents or sports injuries, especially those that cause deep cuts, bruises, or other soft tissue injuries

- Deep vein thrombosis (DVT)
- Varicose veins
    - Varicose veins and DVT both cause damaged veins, which results in overflow from that vasculature penetrating nearby tissues. This overwhelms the lymphatic system and causes disruptions.
- Severe cases of chronic venous insufficiency
    - This results in a condition called phlebolymphedema, which is a mix of lymphatic insufficiency and severe venous insufficiency.
    - In combination, this causes significant backups in the veins, which then leads to blockages in lymphatic drainage.
- Rheumatoid arthritis and psoriatic arthritis
    - Research shows that inflammatory markers in the synovium (which are present in both forms of arthritis) also travel to the lymphatic system, which causes blockages.
- Atopic eczema
    - This condition causes redness and swelling of soft tissue, which can cause permanent damage to the lymphatic system in severe cases.
- Severe obesity
    - While the mechanisms behind lymphedema in individuals with severe obesity are still unclear, it is likely that the excess fatty tissue lowers the flow of fluid through lymphatic vessels.
- Infections and parasites, such as cellulitis and Wuchereria bancrofti
    - Lymphatic filariasis, also known as elephantiasis, is a very rare parasitic disease that impacts the lymph nodes. Elephantiasis is

spread by mosquitoes infected with the Wuchereria bancrofti parasite. In most cases, an individual will be asymptomatic, but long-term complications can include extreme swelling of the arms, legs, and/or genitalia. This condition also places someone at a greater risk of bacterial infections that lead to thickened and hardened skin.

- Prolonged immobility
    - This can lead to extended periods of blockages and stagnant fluid in the lymphatic system, and is most likely to develop in individuals who are also immunocompromised.

In the United States, the most common cause of secondary lymphedema is malignant cancer or treatment related to malignant cancer (specifically radiation therapy). Yet, the prevalence of secondary lymphedema differs worldwide, as its main cause across the globe is a parasitic infection called Wuchereria bancrofti, also known as lymphatic filariasis.

Research shows that 20% of individuals who receive breast cancer treatment will develop lymphedema. While lymphedema can develop after treatment for any type of cancer, this condition occurs most often as a result of breast cancer treatment due to the high concentration of lymph nodes near the breast, underarm, and axilla. Individuals who undergo surgical or chemical interventions to treat breast cancer can experience secondary lymphedema months or even years after their last round of treatment. However, the condition most often develops between 12 and 18 months after the lymphatic vessels are operated on or receive radiation. Around 75% of individuals are impacted by lymphedema within 3 years of the lymphatic vessel damage. For the individuals who do not develop lymphedema in that time frame, their risk increases by 1% each year after that.

There is extensive research to support the connection between lymphedema and other cancers, as well. Over 90% of people who recover from head and neck

cancer will experience soft tissue dysfunction and/or lymphatic complications (combined, external, or internal lymphedema), which occur most often in the first 18 months after treatment. External lymphedema affects neck and head soft tissue, while internal lymphedema impacts the larynx and pharynx (also known as the upper digestive tract). More than 50% of these patients also develop fibrosis. Additionally, studies show that up to 37% of women who receive treatment for any type of gynecological cancer develop symptoms of lymphedema within 12 months after their treatment concludes.

Newer research shows that many people who develop secondary lymphedema after an injury to the lymphatic system have a genetic variation (called single nucleotide polymorphisms, or SNPs) that contributed to their acquired lymphedema. This same mutation is present in babies born with primary lymphedema. In the end, this shows that genetics also play a part in whether or not someone develops secondary lymphedema after a lymphatic injury.

Risk Factors

Understandably, many of the conditions that cause lymphedema place someone at risk for developing the condition. Additional risk factors for lymphedema include:

- Dermatologic signs of cardiac disease
    - These are visible manifestations of edema and skin changes that people with cardiac conditions may have. These signs include clubbing (enlarged tips of the toes or fingers), Osler's nodes (raised, red lesions on the hands and feet), Janeway lesions (painless bumps on the skin of the hands and feet), splinter hemorrhages (the bursting of capillaries under the nails), and purpura (the leakage of capillaries under the skin). Each of these concerns place someone at risk for lymphedema due to their involvement of lymphatic and circulatory structures.

- Dermatologic signs of renal disease
    - Purpura may also result in individuals with renal disease, specifically in the lower legs. Individuals with renal disease, particularly those who rely on hemodialysis, may also experience nodules just under the skin and ulcers called calciphylaxis. Each of these concerns place someone at risk for lymphedema due to their involvement of lymphatic and circulatory structures.
- Thrombophlebitis
    - This is an infection that leads to the formation of blood clots in one or more of a person's veins. Thrombophlebitis typically takes place in the legs, but it can be superficial or deeply embedded in muscle. In the latter instance, this is called deep vein thrombosis, or DVT. As with other circulatory conditions, either type of thrombophlebitis can cause blockages in lymphatic vessels.

Due to the difference in etiology of both lymphedema types, primary and secondary lymphedema each come along with their own sets of risk factors. Since primary lymphedema itself is a congenital condition, individuals with other congenital conditions are at a greater risk of developing this disorder. The following conditions are considered risk factors for primary lymphedema:

- Noonan Syndrome
    - A genetic disorder that causes skeletal dysfunction, heart defects, and bleeding problems. It's somewhat rare for individuals with Noonan Syndrome to develop lymphedema but, when it does occur, it consistently results in swelling of the bilateral legs, genitals, and intestines.
- Prader-Willi Syndrome

    - This genetic disorder results in symptoms such as muscle weakness and developmental delays. Newborns and infants with Prader-Willi Syndrome will often have difficulty latching to breastfeed or take a bottle. This later develops into a heightened appetite in childhood. There is not much research on the link between these two conditions, but it is thought that chromosomal abnormalities that cause Prader-Willi Syndrome may also cause malformations in the lymphatic system.

- Yellow Nail Syndrome

    - Yellow Nail Syndrome is a rare condition that can affect multiple bodily systems. Individuals with this condition must possess two of the following criteria to be diagnosed: peripheral lymphedema, stunted nail growth with hardened and yellow nails, or diseases of the respiratory tract. While the cause of Yellow Nail Syndrome is still unknown, it is thought that abnormalities in the lymph vessels under the nails lead to its development.

- Turner Syndrome

    - This genetic condition affects only females and is characterized by an underdeveloped reproductive system. Many infants with Turner Syndrome have lymphedema present in the hands and feet at birth, but this sometimes resolves on its own. Research shows that 2 out of 3 women with Turner Syndrome will develop lymphedema at some point in their lives.

- Neurofibromatosis Type 1

    - Neurofibromatosis Type 1 causes someone to experience skin color changes along with tumors on the brain, skin, nerves, and other areas. Since symptoms vary greatly, it's possible for individuals with

Neurofibromatosis Type 1 to experience lymphedema as a result of lymphatic blockages from tumors throughout the body.

- Hemangioma
    - These are non-cancerous birthmarks that consist of excess blood vessels within the skin. Their exact cause is still unknown. Hemangiomas can occur anywhere in the body, and lymphedema can develop if these clusters form over areas with a lot of lymph nodes or vessels. This would lead someone to experience additional, more noticeable swelling around the area of the hemangioma.
- Lymphedema Distichiasis Syndrome
    - It's a bit more clear why this condition predisposes someone to lymphedema, as Lymphedema Distichiasis Syndrome develops due to an underdeveloped lymphatic system. Individuals who have this condition often experience bilateral leg swelling that begins around adolescence. This condition is also associated with varicose veins, which also place someone at risk of developing lymphedema.
- Eruptive Xanthomatosis
    - This skin condition causes someone to develop visible yellow or orange bumps. The bumps, also called xanthoma, are formed as a result of lymphatic system abnormalities that cause the leakage of lipoproteins. This leakage eventually accumulates until someone develops symptoms of lymphedema.
- Anonychia congenita
    - This condition results in the congenital absence of nails. It's quite rare to be missing nails from birth and some people diagnosed with this condition may be born with partial nails on many digits. Either way, this condition can cause lymphedema similarly to how Yellow Nail

Syndrome can: due to abnormalities in the development of lymphatic vessels in the nail beds.

- Down Syndrome
    - Down Syndrome is a genetic condition that can cause many bodily changes, including chylous ascites. Chylous ascites involve the leakage of lymphatic fluid (rich in triglycerides and other fats) into the area between the abdomen and the pelvic wall. This leads to intestinal swelling, which is common with lymphedema.
- Klippel-Trenaunay Syndrome (KTS)
    - This is another rare congenital condition that prevents the proper development and functioning of many aspects of the musculoskeletal system, including soft tissues, blood vessels, and bones. This often also extends to the lymphatic system, which places someone at a higher risk of lymphedema.

Any other disorders that cause lymphatic malformations (such as Klinefelter Syndrome and many other genetic conditions) also place someone at risk of developing primary lymphedema. It's also of note that many lymphatic formations able to be detected in utero typically resolve and are not present at birth. However, this is not usually the case for most genetic conditions, even those that can be identified through prenatal surveillance and testing.

Risk factors for secondary, or acquired lymphedema include:

- Aging
- Diabetes
- Family history of lymphedema or arthritis
- A lack of physical activity

- Obesity
    - Individuals who are overweight or obese and also experienced damage or injury to the lymphatic system are at risk for developing lymphedema.
- Receiving a combination of chemotherapy and radiation for the treatment of cancer
- Having edema before receiving treatment for cancer
- Smoking
- Having low breast density
    - Research shows that low breast density can cause someone to have poor adipose homeostasis, which prevents the body from regulating, transporting, and storing fats. Low breast density may also lead to structural changes that impact the lymphatic vasculature.
- Being a current or prior user of intravenous drugs
    - This places an excess of stress on the lymph nodes and can cause permanent damage.

There are several myths about activities and practices that can cause lymphedema or raise someone's risk of developing the condition. For example, being in overly hot temperatures cannot cause lymphedema nor can flying in an airplane. Despite this, it is recommended that people who have other risk factors for lymphedema avoid excess sources of heat such as saunas because any extreme temperatures (either hot or cold) place added stress on the lymphatic system, which can lead to slowed movement of fluid through lymphatic vessels. There is no evidence to suggest that flying in airplanes or traveling to places with higher altitudes will cause or worsen lymphedema. But many people who already have lymphedema

often feel more comfortable wearing compression garments during flights to offer themselves added support.

It's also untrue that vigorous exercise will cause lymphedema in those who are at risk for the condition. In fact, individuals with lymphatic system damage or a diagnosis of lymphedema should regularly engage in controlled, gentle exercises and stretches to best manage their symptoms.

Another myth about lymphedema is regarding the use of the affected side of the body. Some people mistakenly believe that those who had lymph nodes removed near one of their arms should avoid lifting heavy items with that arm or getting blood pressure readings and blood draws from that extremity. These practices will not cause lymphedema, but they can lead to discomfort and complications in someone who already has lymphedema. If a healthcare professional draws a patient's blood improperly, this places someone at risk of infection at the site where the needle was inserted. In most people, this is a mild concern with a simple resolution. However, someone with lymphedema can develop cellulitis in their affected arm as a result of a minor infection. For this reason, many people who have lymphedema are advised to avoid getting blood drawn in the affected limb(s). There are no risks associated with getting blood pressure taken from a limb that has lymphedema-related swelling, but people with this condition are often more comfortable getting their vitals checked on a different extremity.

Similarly, it's untrue that individuals with lymphedema in one or both upper extremities cannot lift anything with their affected arm(s). As long as someone can lift items without exerting too much force or straining themselves, they can continue their daily activities as they normally would.

Section 2 Personal Reflection

How can an occupational therapist increase awareness of lymphedema risk factors? How would the target audience and preventive interventions differ for primary and secondary lymphedema?

Section 2 Key Words

Hyperkeratosis - A symptom that involves the thickening of the outer layer of skin and the appearance of scaliness

Lymphangioma - A condition that causes children to experience small, non-cancerous blisters and bumps on the skin; they are most common on the head and neck and don't usually require treatment unless they are severe

Lymphorrhea - The leakage of lymphatic fluid from the skin; lymphorrhea is a symptom of lymphedema and can lead to complications if it is not properly managed

Lymphoscintigraphy - A diagnostic test that involves the use of a radioactive substance to visually view the flow of fluid through a person's lymphatic system; early stages of lymphedema can be diagnosed through this test even if symptoms are not apparent yet

Papillomas - Skin changes similar to warts that are common in those who have lymphedema; they can lead to complications if not managed, since they ooze and may become infected

Phlebolymphedema - A hybrid type of lymphedema that results from both lymphatic dysfunction and an impaired circulatory system; this commonly causes lymphedema in the legs

Pitting edema - A type of inflammation that is apparent when someone presses a finger into the skin, which temporarily leaves a dimple; the normal response is for the skin to bounce back immediately

Tissue fibrosis - Scarring and excess extracellular growth that leads to hardening of soft tissues; this often results due to extra collagen deposits

Section 3: Lymphedema Diagnosis, Prognosis, & Complications

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Individuals who present to their doctor with potential symptoms of lymphedema will first be asked about their medical and family history. This will help determine if patients have any risk factors that increase their chances of developing lymphedema. Doctors will visually inspect the skin to look for any infections or injuries that may cause symptoms similar to those of lymphedema. They will also check the entire body for swelling, since this may reveal other swelling with a different source. Palpation is another part of the physical examination. When palpating, doctors will use their hands to manually feel the tissues in certain areas of the body. It is common for them to check areas where there are many lymph nodes, including the underarms and behind the ears. They will be looking to see if these areas are hard or soft and if any lymph nodes are tender, sensitive, or are larger than they should be. While completing the physical exam, doctors will also complete the Stemmer sign. This is a physical test where a doctor or therapist will pinch the skin on the back of the foot or hand. If the examiner can do this, the test is negative and the patient likely does not have lymphedema-related swelling. If the examiner cannot do this, the test is positive and the patient is more likely to have swelling associated with lymphedema. If patients have a positive Stemmer sign or other signs of lymphedema and their doctor suspects this condition is a possibility, they may run one or more tests to get a closer look at their lymphatic system.

Lymphoscintigraphy is one test that can help with the diagnosis of lymphedema. This is most often used to help diagnose individuals with primary lymphedema, since such patients may not demonstrate visible signs of the condition. Lymphoscintigraphy involves taking video imaging of the lymphatic system using a scanner or probe after injecting someone with radioactive dye. This dye travels through someone's system and lights up structures such as lymphatic ducts,

vessels, and drains. Doctors can then use this video footage to point out any blockages. This test can take up to a few hours to complete, since it's important that the dye flow through much of someone's lymphatic system. While lymphoscintigraphy may be used to confirm lymphedema in certain less obvious cases, it's possible to make a diagnosis of lymphedema without completing this test. This test is also a good way for providers to measure the extent of lymphatic damage a person is experiencing so they can best inform interventions. Lymphoscintigraphy has replaced lymphangiography as the gold standard test for diagnosing lymphedema. Lymphangiography uses a similar radioactive dye along with X-ray imaging to take photos of potential areas of blockage within the lymphatic system.

Doctors may also use magnetic resonance imaging (MRI), computed tomography (CT) scans, and ultrasound to view potential lymphatic blockages. Ultrasound, specifically Doppler ultrasound, can help doctors rule out differential diagnoses such as deep vein thrombosis and venous insufficiency. Doppler ultrasounds and optical scattering methods can also be used to measure the diameter of blood vessels, veins, and arteries along with the flow inside of them. Since ultrasound provides a view of soft tissue, this diagnostic test can also point out masses, tumors, or other tissue changes that may be contributing to lymphatic compression. CT scans and MRIs are accurate ways to explore sources of soft tissue edema. However, these tests are expensive, so they are not often used for lymphedema diagnosis unless other similar conditions are suspected.

After a diagnosis is made, it's common for doctors and/or therapists to take measurements of the affected and unaffected limbs so they can compare the two sides, monitor the affected side over time, and determine if treatments are working. The gold standard method for limb measurement is volumetry, also known as water displacement. Volumetry involves placing the affected limb in a two-sided water tank and measuring how much water flows from one side to the other to determine the volume of the limb. Providers can also use a tape measure to calculate the circumference of the limb at certain anatomical landmarks. When

measuring the arm, providers should be mindful of the olecranon process on the elbow and the acromion process on the shoulder. When measuring the leg, providers should do so around the popliteal fossa and the malleoli of the ankle.

Perometry is another diagnostic method that utilizes infrared light to outline an affected limb and calculate its volume. Additionally, doctors may use bioimpedance testing, which involves placing electrodes on the body via tiny metal discs that measure electrical charges. Any changes in the strength of the current between these discs notifies the provider that the tissue beneath that area has fluid buildup.

Prognosis & Complications

As of now, there is no cure for lymphedema. The best way to keep this condition under control is to get regular treatment to manage its symptoms and prevent complications from arising. Someone's prognosis is usually based on the severity of their condition and how consistent they are with receiving treatment. Without treatment, it is more likely that patients will experience complications. One of the most common complications of lymphedema is cellulitis, since bacteria can enter the body through even a small break in the skin and the infection can spread. Individuals with untreated or severe lymphedema may also experience other complications, including:

- Lymphangitis, which is an inflammatory infection of lymphatic structures that may come about through viruses, fungi, bacterial or other pathogens
- Acute dermato-lymphangio-adenitis (ADLA)
    - This condition often presents similar to cellulitis or erysipelas because it develops when bacteria enters damaged lymphatic structures as a result of parasitic filarial infections. Individuals with Stage 0 lymphedema who do not have any symptoms may develop bouts of ADLA that are difficult to differentiate between.

- Deep vein thrombosis
- Severe functional impairment
- Psychosocial dysfunction
- Cosmetic embarrassment
- Sepsis
- Superficial bacterial and fungal infections
- Skin leakage
- Amputation
- Lymphangiosarcoma
    - This is a highly aggressive tumor that begins in the fibrous tissue, muscle, fat, blood vessels, or other soft tissues and spreads elsewhere. Lymphangiosarcomas must lead to amputation of the affected extremity for the best results. Even so, lymphangiosarcoma has a poor prognosis, with its five-year survival rate being less than 10%.
    - People with primary lymphedema, secondary lymphedema, or other lymphatic system conditions for 10 years have a 10% risk of developing lymphangiosarcoma.

People who have surgery to manage lymphedema may also experience different complications, which are quite common and include hematomas, seromas, skin necrosis, and partial wound separation.

Section 3 Personal Reflection

How might lymphedema complications impact the occupational therapy treatment process?

Section 3 Key Words

Hematoma - A pool of semi-clotted blood that may form within tissues or organs anywhere in or on the body; this usually results from a broken blood vessel and appears as a large, dark purple or brown bruise on the surface of the skin

Seroma - An abnormal cluster of clear or light yellow fluid (called serous fluid) that accumulates in dead spaces within the body; this is a common occurrence after surgery

Skin necrosis - Tissue death that may occur after trauma; causes of skin necrosis include surgery, injury, radiation treatment, and exposure to chemicals

Wound separation - When the edges of a wound fully or partially separate despite stitches or other wound closures; this happens most often around 5 to 7 days after surgery and is also called dehiscence

Section 4: Lymphedema Assessment

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A well-rounded evaluation process is the best start to any occupational therapy plan of care. This also applies to treatment plans for individuals with lymphedema. Occupational therapists can get a good idea of a patient's priorities, needs, personal goals, social support, and more by putting together an occupational profile. Important aspects of a complete evaluation for individuals with lymphedema include:

- Prior level of function (PLOF)
- Family and caregiver support
- Professional/employment-related responsibilities, roles, and expectations
- Leisure activities and hobbies

- Social and family roles
- Past and current lymphedema treatments and results
- Past compliance with home recommendations and exercise programs
- Occurrence of lymphedema complications
- Comorbidities, especially those that increase the risk of complications for someone with lymphedema
- Current medications
    - Analgesics, narcotics, chemotherapy agents, hormonal treatments, and neuromuscular medications can all have an impact on fluid balance in the body, which can impact medical and therapeutic treatment.
- Physical examination of the skin, including texture, appearance, and wounds
    - For any wounds, the therapist should note the size, color, drainage, location, dressing, and sutures, since these will be monitored closely during the therapy process.
- Strength
- Range of motion and overall joint mobility
    - It is common for patients with upper extremity involvement to have associated shoulder pathologies, so basic testing should be done to determine if there is any shoulder impingement, acromioclavicular osteoarthritis, rotator cuff tears, or labral tears. If any of these conditions are suspected, a therapist should refer the patient back to their doctor for confirmation via diagnostic testing.
- Edema measurement

- Pain
- Postural symmetry of the upper and lower body
- Sensation
- Cognition
- Coordination and motor control
- Endurance

Functional aspects of an evaluation should address any tasks someone frequently engages in. Specifically, therapists should discuss and observe a patient's ability to engage in tasks that require upper or lower extremity weight-bearing, excessive reaching, lifting, or carrying loads with both upper extremities. Therapists should pay particular attention to the impact that lymphedema symptoms have on a patient's ADL and IADL function along with any mental health concerns that may be present.

There are a range of standardized assessments that are suitable for the evaluation of individuals with lymphedema. Many of them focus on functional performance, but there are also a lot of assessments that take a look at patients' quality of life related to lymphedema. These assessments are particularly important since mental health is often affected in individuals who have lymphedema. Some standardized assessments are general and can be used with individuals who have a range of diagnoses, while others were designed specifically for those with lymphedema. Some examples of both include:

- Medical Outcomes Study 36-Short-Form
- Nottingham Health Profile (NHP)
- European Organization for Research and Treatment (EORTC) QLQ-C30

- The Functional Living Index-Cancer (Manitoba Functional Living Cancer Questionnaire)
- Chronic Venous Insufficiency Quality of Life Questionnaire (CIVIQ)
- Upper Limb Lymphedema-27 (ULL-27)
    - This tool assesses social, psychological, and physical health in those with upper extremity lymphedema.
- Functional Analysis of Cancer Therapy - Breast (FACT-B)
    - The FACT-B covers emotional, social, and physical health along with functional performance. This is a subtest for breast cancer patients, so it is more sensitive to those who developed lymphedema as a result of a cancer diagnosis.
- Freiburg Life Quality Assessment for Lymphoedema (FLQA-L)
    - This variation of the Freiburg Life Quality Assessment for Veins covers treatment, general satisfaction, emotional health, household management, professional engagement, social life, physical health, and daily activities.
- Gynecologic Cancer Lymphedema Questionnaire (GCLQ)
- Lower Extremity Function Scale (LEFS)
- Lymphedema Life Impact Scale (LLIS)
- Lymphoedema Quality of Life (LYMQOL) - ARM
- Lymphoedema Quality of Life (LYMQOL) - LEG
- The Lymphedema Symptom Intensity and Distress Survey (LSIDS-L) - Lower Limb
- The Lymphedema Symptom Intensity and Distress Survey (LSIDS-A) - Arm

- The Lymphedema Symptom Intensity and Distress Survey - Trunk, Head, & Neck
- Lymphoedema Functioning, Disability, and Health Questionnaire for Lower Limb Lymphedema (Lymph-ICF-LL)
- Lower Extremity Lymphedema Screening Questionnaire (LELSQ)

Once the evaluation process is complete, occupational therapists must determine whether or not the patient is a good candidate for occupational therapy. Some indications that a patient with lymphedema needs occupational therapy include the inability to functionally use a limb due to loss of motion, increased weight, or greater size; the presence of scar tissue that limits someone's range of motion and functional performance; and lymphedema-related swelling greater than 2 centimeters in the affected limb.

After a therapist determines a patient would benefit from occupational therapy treatment to manage lymphedema, they should develop appropriate goals based on the deficits identified. Based on the occupational therapy scope of practice, goals for a patient with lymphedema may include:

- To comprehend and follow lymphedema precautions to reduce the risk of infection
- To tolerate wearing multiple layers of short-stretch bandages to reduce edema
- To demonstrate independence in wearing multiple layers of short-stretch bandages to reduce edema and/or prevent fluid from reaccumulating
- To demonstrate lower levels of pitting edema to improve tissue health
- To comply with a home exercise program with minimal assistance to improve lymphatic flow

- To increase range of motion to experience more success with functional transfers, functional mobility, and/or functional tasks
- To practice manual lymphatic drainage according to the outlined protocol with minimal assistance to lower edema and improve range of motion
- To independently don and doff compression garments for regular daily use according to therapist wearing schedule
- To independently don and doff nighttime compression garments or use nighttime compressive devices to prevent fluid from reaccumulating
- To achieve maximum edema reduction to allow the patient to return to their prior level of function, improve their balance, reduce their fall risk, and wear standard clothing and shoes

These goals will pertain directly to the treatments an occupational therapist provides to help a patient better manage lymphedema symptoms. These treatments will be discussed in greater depth in the next section.

Section 4 Personal Reflection

How might the goals of an occupational therapist treating a patient with lymphedema differ from the goals of a physical therapist treating a patient with lymphedema?

Section 4 Key Words

Comorbidities - A medical condition that exists alongside one or more other medical concerns

Section 5: Lymphedema Treatment

41,42,43,44,45,46,47,48,49,50,51,52

As we mentioned before, there is no cure for lymphedema. Therefore, all therapeutic and medical treatments for lymphedema are intended to help manage symptoms, slow the progression of the condition, and improve a person's quality-of-life.

One of the most evidence-based treatments for lymphedema is complete decongestive therapy (CDT), sometimes also known as complex decongestive therapy. Research supports the use of CDT as an effective way to manage Stage 1 or Stage 2 lymphedema symptoms. There are two phases of CDT: Phase 1, which is the active phase; and Phase 2, also known as the maintenance phase. During Phase 1, the goal of CDT is to get excess lymph fluid out of the body, lower overall swelling, and manage other uncomfortable symptoms such as decreased motion. The length of Phase 1 CDT depends on how severe a person's symptoms are. This phase may last from 2-4 weeks up until 8 weeks. Since this phase is intended for people whose symptoms can be slowed, its schedule is typically quite rigorous. Patients are expected to attend 1-hour sessions 4 to 5 days per week. Treatments in all phases of CDT include bandaging/compression garments, exercises, skin care, and a technique called manual lymphatic drainage. However, certain aspects of these treatments – such as the wearing schedules for bandages – differ between Phase 1 and Phase 2. Patients receiving Phase 1 CDT are expected to wear bandages for 23 hours each day and only remove them to bathe.

Since Phase 2 CDT involves the maintenance aspect of lymphedema, treatments focus on maintaining any positive outcomes yielded from Phase 1 CDT and allowing someone to gain long-term control of their lymphedema symptoms. As we discussed earlier, treatments for Phase 2 CDT are nearly identical to treatments in Phase 1 CDT, and include manual lymphatic drainage, bandaging/ compression garments, exercises, and skin care. The wearing schedule and type of bandages worn are part of what set Phase 2 treatment apart from Phase 1 treatment. Patients in Phase 2 CDT wear very thin, elastic compression garments during the day to maintain their sizing and switch to bandages with foam padding at night, which are intended to decrease any swelling that accumulated during the

day. During this phase, patients are also instructed to wear compression garments when exercising and complete self-manual lymphatic drainage for around 20 minutes each day. It is not uncommon for lymphedema patients to remain in Phase 2 for years or even the rest of their lives.

There are some general contraindications therapists should be aware of when implementing CDT. Therapists must not use any form of heat on the affected extremity since this is known to increase inflammation. It is also contraindicated to lead patients through exercises if they have an active infection or excessive pain. Therapists should also avoid using therapeutic ultrasound on any part of the body that has lymphedema-related swelling if a patient also has a history of cancer.

Manual Lymphatic Drainage

While there is sufficient evidence to support the utility of each treatment within CDT, these methods are considered most effective when implemented together. CDT is considered highly effective at managing lymphedema symptoms, which is why therapists providing any of these treatments must follow specific protocols. Manual lymphatic drainage (MLD) involves providing light touch and light stretching of the skin to stimulate the lymphatic system and encourage fluid drainage. The MLD technique will differ slightly depending on the part of the body where someone has swelling. However, it's considered best practice to engage in deep breathing sequences both before and after MLD for the best results. Patients should receive MLD while laying down. The therapist begins by performing MLD slowly and rhythmically on any unaffected body regions or lymph nodes.

The therapist carries out four main strokes – the scoop technique, stationary circles, the rotary technique, and the pump technique – while moving proximally to distally to encourage drainage. This is referred to as the Vodder approach, but there are several other variations used. The Foldi approach also includes encircling strokes along with a thrust and relaxation phase. The Casley-Smith technique requires the therapist to use the side of their hand rather than the fingertips.

Lastly, the Leduc approach uses “call up” and “reabsorption” movements to help the lymphatic fluid enter the initial lymphatic structures and then travel to the larger structures to be redistributed.

Research shows that MLD itself is most effective for people with mild lymphedema and may also help people who are recovering from breast cancer surgery. Other studies show that an MLD variation called the Godoy method led patients to display a significantly lower limb volume in the arm. MLD is considered one of the more impactful and efficacious aspects of CDT for individuals with lymphedema of the head and neck, partly due to difficulty and discomfort when wearing compression garments in these areas.

Contraindications for MLD include cellulitis, malignant tumors that are in the early stages of treatment or have not yet been treated, and impaired arterial perfusion, which means arteries are not sending enough blood to the extremities. If someone receives MLD when they have cellulitis or a malignant tumor, the drainage process could worsen those conditions and cause them to spread. Additionally, lack of blood flow from impaired arterial perfusion can cause circulatory complications.

Compression Therapy

Compression therapy, including compression bandages and compression garments, are another crucial aspect of CDT. Compression works by giving the musculature counter force or working pressure, which prevents fluid from reaccumulating after therapies such as MLD. Research shows that people who receive MLD without compression afterwards usually do not experience a decrease in swelling. If patients receive compression therapy without MLD, they are at risk of experiencing proximal edema and fibrosis. These two modalities work together to prevent fluid from accumulating proximal to the affected body part. This is why patients wearing compression bandages are advised to keep them on until their next MLD session.

During Phase 1 of CDT, patients with lymphedema wear multiple layers of short-stretch (or low-stretch) bandages. These bandages have some extensibility, meaning they stretch between 30% and 60% farther than their resting length. In Phase 2 of CDT, patients progress to flat-knit, elastic compression garments and night compression systems. Therapists will usually layer bandages with foam underneath for added comfort and to maintain the patient's skin integrity.

Depending on the part of the body affected by lymphedema, compression garments and bandages may be worn around the entire arm or leg like a sleeve. This style of dressing offers resting pressure, which means there is more compression being given when the limb is at rest. The elastic moves along with the limb, which in turn causes the pressure to decrease. For this reason, sleeves are sufficient for people with mild lymphedema, but bandages are more suitable for those with moderate to severe swelling.

Individuals with lymphedema of the chest may use support bras, while those with swelling across the torso may need a vest. If swelling is concentrated in the hands, therapists may give patients fingerless gloves that offer some functionality. An alternative to this is a hand gauntlet, which is more like a mitten with a sleeve that extends up the forearm. Regardless of the style of dressing, wearable compression is made of flexible fabric and may consist of materials such as latex or wool. Most compression garments are graduated, meaning they are tighter at the farthest point from the heart (often the bottom of the garment) and slightly looser as the garment gets closer to the heart. The compression gradually gets less between the top and the bottom of the garment. This keeps the fluid moving out of the limb and toward the heart, which offers additional pumping to keep it moving.

There are some myths surrounding compression therapy and one is that they cause pain or are too tight to be comfortable. When they are sized properly for the individual wearing them, compression bandages and garments alike should not cause any pain or discomfort. If you ever find multi-layer bandages are causing you pain, remove one layer of compression and see if that helps. If the pain

persists or you notice changes in the color of your limb, numbness, and/or tingling, remove the bandages altogether and notify your provider. Another myth about compression therapy is that some compression is better than none. Individuals who have significant swelling and wear garments that do not offer enough compression are likely to notice that the garment will fall down or bind, which can worsen swelling. It's also a myth that compression socks and other compression garments will lower swelling. Compression garments are actually intended to maintain a person's current limb size and not further reduce swelling. This is why it's important for individuals with lymphedema to get compression socks, and other garments fitted to their body *after* they begin CDT and their swelling has decreased to a manageable degree. Bandages, on the other hand, are considered reductive therapy that makes the limb smaller. This is why they are used in the early stages of CDT.

When patients participate in prescribed exercises or functional activities while wearing bandages, this triggers an internal pump mechanism that moves fluid out of the tissues and into lymphatic vessels. While this initial movement is important, it is only effective if the fluid does not reaccumulate. This is why bandage wearing schedules are typically extensive, since they must prevent fluid from flowing back into the limb. Bandages also help soften thick lumps of tissue under the skin that develop due to lymphatic buildup. Compression bandages and compression garments are contraindicated for individuals with active skin ulcers, arterial disease, or signs of an infected wound.

Compression pumps are another wearable treatment option that may be used along with other aspects of compression therapy. Flexitouch is one of the most well-known devices for pneumatic compression. While it's recommended to use all types of CDT together for the best results, there is not much evidence to support the effectiveness of pumps when used in isolation. However, they may be recommended for patients with lymphedema who are non-ambulatory and cannot engage in prescribed exercises or other functional activities to encourage more lymphatic circulation. Compression pumps are contraindicated on

extremities that do not properly fit in the pump, meaning they may not be suitable for patients with advanced lymphedema. It's best practice for therapists placing compression pumps on patients to soften the tissues prior to use by washing and/or moisturizing the area.

Many insurance companies, including Medicare and Medicaid, offer varying degrees of reimbursement for CDT modalities such as compression therapy. In fact, some insurance companies consider it "out of network," meaning they have not established financial negotiations with providers for that service. However, federal legislation called the Lymphedema Treatment Act aims to change this and mandates that insurance companies view compression supplies as medically necessary for patients with manageable lymphedema. This act was passed in December 2022 and is expected to take effect on January 1, 2024, which means patients with lymphedema can benefit from much-needed coverage for an evidence-based treatment that can manage their condition.

Exercises

Supervised and independent exercises are both crucial for CDT. Individuals with arm lymphedema will be led through a series of exercises intended to improve their active motion while increasing the flow of lymphatic fluid out of the limb and into the lymphatic system. These exercises include ball squeezes; shoulder shrugs; elbow flexion while seated without weights; elbow extension, horizontal abduction/adduction of the shoulder in supine; and shoulder flexion and shoulder abduction in standing. Pole walking is another great exercise for lymphedema, which can also help improve coordination. A patient starts by standing upright and taking one step forward with their right foot while swinging their left arm forward from their waist. The left pole will then hit the ground behind the person's right foot, at which point they will let their right arm straighten behind them. While alternating feet and poles, patients should roll their left foot from the heel to the toe and push off with their toe, then repeat these steps for their right foot.

Patients with leg lymphedema would benefit from exercises such as hip and knee bends in sitting, standing, and supine. Other exercises include ankle circles, marching, gluteal squeezes, bridges, and clamshells. Those with lymphedema of the head and/or neck should practice head tilts, chin tucks down toward the chest, and trunk rotation. Contraindications related to exercise include any repetitive or vigorous motions such as scrubbing. Once getting clearance from their doctor and therapist, patients are encouraged to participate in exercises such as swimming, pilates, walking, tai chi, and yoga on their own.

Self-Massage

In addition to MLD provided by therapists, patients with lymphedema are instructed on the use of self massage to further improve fluid redistribution. Depending on what part of the body lymphedema has affected, patients may perform self massage to the stomach, thighs, calves, and/or feet. When massaging to decrease lymphedema near the stomach, patients will keep their hands on their pelvis and make short gentle strokes up toward the stomach. Self-massage to assist with lymphedema of the thighs should include massaging up the thighs to the pelvis. When massaging the lower legs, patients should massage up from the feet to the thighs.

Risk Reduction

An important part of an OT's role for all patients is providing education to help patients avoid injuries or complications. For lymphedema, this includes practices that fall under the heading of risk reduction, which includes some of the following:

- Maintaining a healthy weight with diet and exercise
    - A healthy diet for someone with lymphedema should be low in sodium

- Avoiding injuries of any kind to the affected limb, including physical trauma, pin pricks, sunburn, and cuts or abrasions
- Performing appropriate skin care
    - Cleaning and thoroughly (but gently) drying the affected limb daily
    - Lotioning the affected limb daily
    - Cutting nails regularly
    - Checking skin with a mirror to identify and monitor any scrapes or minor cuts
    - When cuts do occur, cleaning them with soap and water, then adding ointment and sterile dressings to prevent infection
- Elevating the limb whenever possible
- Using electric shavers when removing hair from the affected limb
- Not wearing clothing with elastic bands, since this can impair circulation
- Avoiding extreme temperatures on the affected limb
- Carrying heavy items with the unaffected limb
- Using gloves during certain tasks such as when soaking laundry, washing dishes, gardening, or sewing

Physical Agent Modalities (PAMs)

Since extreme temperatures are contraindicated for lymphedema, patients with this condition cannot receive some PAMs, including moist heat, cryotherapy, and fluidotherapy. Patients who have lymphedema-related wounds that need debriding may benefit from whirlpools to assist with preparing the skin and relieving pain. Some therapists may also choose to use PAMs such as iontophoresis, which are intended to assist with edema management on patients

with mild lymphedema. Low-level laser therapy is another option that can be incorporated into CDT. This modality targets underlying tissues to modify cells that are excessively accumulating fluid. Low-level laser therapy can lower swelling, alleviate skin tightness, and lead patients to experience greater range of motion.

In terms of functional treatments for lymphedema, therapists should make adaptations to any ADLs or IADLs that patients are having difficulty with. This may or may not include recommendations for environmental modifications that allow patients to more safely and efficiently ambulate and function within their chosen contexts.

Therapists should be aware that rapid exacerbations of lymphedema may indicate new malignancy or deep vein thrombosis. Similarly, any new redness in the affected limb may point toward an infection. For this reason, therapists should do skin checks during each session and closely monitor a patient's symptoms and tolerance for treatment throughout the plan of care.

While any occupational therapist can treat related conditions that a patient with lymphedema is experiencing, therapists must have credentials to perform CDT. These credentials are that of a certified lymphedema therapist (CLT). Both occupational and physical therapists with relevant experience may become a CLT after completing 135 hours of training (both home study and live lectures with hands-on workshops), logging past experience working with this population, and passing a certification exam. This allows general practitioners to gain the experience and skills they need to effectively provide specialized CDT services for those with lymphedema.

Section 5 Personal Reflection

What indications should a therapist look for to determine how a patient with lymphedema is tolerating PAMs?

Section 5 Key Words

Debriding - The process of cleaning and removing dead tissue from an open wound; this is usually done using small instruments and is often performed after softening the skin

Section 6: Case Study #1

A 45-year-old woman with mild lymphedema presents to an occupational therapist with reports of pain, numbness, and tingling in her right arm. She was diagnosed with lymphedema 3 weeks ago and recommended to begin CDT. She reports being able to complete her morning self-care routine independently, though it takes her a little longer than usual. She is beginning to have difficulty with dressing and bathing later in the evening, which is usually when she experiences most of her symptoms along with excessive fatigue. The patient has not reported any mental health concerns, but she is becoming worried she will not be able to care for her two young children.

1. What CDT stage is this patient most appropriate for?
2. How can the therapist best address this patient's self-care concerns?
3. Should the therapist address mental health during this patient's sessions? If so, using what treatments?

Section 7: Case Study #1 Review

This section will review the case studies that were previously presented. Responses will guide the clinician through a discussion of potential answers as well as encourage reflection.

1. What CDT stage is this patient most appropriate for?

Since this patient was just diagnosed with mild lymphedema and has not yet had lymphedema treatment, this patient will be in phase 1 CDT and receive treatments accordingly.

2. How can the therapist best address this patient's self-care concerns?

   The therapist should educate the patient that it's normal to have more symptoms in the evening, since the effects of gravity over the course of the day can lead fluid to accumulate more. The OT should instruct the patient to elevate her arm whenever possible and alternate arms during functional tasks to give herself a break while also encouraging movement of fluid in the affected arm. Another helpful recommendation would be offering energy conservation strategies to maximize her stamina during the day. Accordingly, the OT can suggest that the patient shower and wash her hair in the morning instead of the evening so that her symptoms are less impactful.

   Even though lymphedema treatment can greatly minimize the negative effects of the condition, the therapist may want to recommend some adaptive equipment to assist with her fatigue and allow her to more safely complete her ADLs. This will help maintain her independence in the event the patient cannot always adjust her schedule to shower during the morning hours. A shower chair with a back can prevent falls and help manage her fatigue. A handheld shower head, soap on a rope, and a long-handled scrubber can also help maximize the use of her affected arm. The therapist can also recommend adaptive equipment to help with dressing, such as a reacher, a long-handled shoe horn, a button hook, and a sock aid, depending on the patient's specific needs.

3. Should the therapist address mental health during this patient's sessions? If so, using what treatments?

The patient did report some fears over her long-term function and prognosis, so it's fitting that the OT provides some education about these areas to give the patient a realistic view of her condition. The OT can also recommend journaling (using a dictation feature on the computer if typing is not possible), deep breathing, and a support group to help this patient better cope with her condition and manage any anxiety that she may carry.

It's also a good idea to administer a lymphedema-related quality-of-life assessment to help with the early identification of any other emotions this patient may be experiencing. This will alert the therapist to other emotions that may impact the treatment process, and help them make a referral to a mental health provider, if necessary.

Section 8: Case Study #2

A 9-month-old child is diagnosed with primary lymphedema after demonstrating persistent swelling in both arms since birth. He is currently experiencing gross and fine motor delays as a result of moderate swelling. His parents are concerned, since he also appears to avoid interacting with objects due to his motor difficulties. They are also concerned that he is having difficulty settling down at night and sleeping through the night.

1. Is a lymphedema assessment or another assessment appropriate for this child?
2. What interventions will be most effective for this child?

Section 9: Case Study #2 Review

This section will review the case studies that were previously presented. Responses will guide the clinician through a discussion of potential answers as well as encourage reflection.

1. Is a lymphedema assessment or another assessment appropriate for this child?

   Since this patient is so young, it's more appropriate to use a pediatric assessment that evaluates motor skills. Some examples include the WeeFIM, Bayley Scales of Infant and Toddler Development Motor Scale, and Peabody Developmental Motor Scales. While it is a bit early to use the self-care portions of these assessments, they will give the therapist an idea of how the child is developing in terms of mobility, social interaction, motor planning, balance, coordination, strength, and bilateral integration.

2. What interventions will be most effective for this child?

   This child is a good candidate for participating in the active phase of CDT, since they can benefit from interventions such as compression therapy, MLD, infant massage provided by the parents, and PAMs, if any are indicated. The therapist can educate parents about home recommendations such as stretching and limb elevation, though some of these may be difficult to implement on a child of this age. Based on the results of the functional assessment, the therapist can determine what specific needs the child has and write goals accordingly. In addition to this, the therapist should provide the parents with some sensory strategies that can help with the child's alertness, regulation, and sleep.

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