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Complex Decongestive Therapy-based Management of Primary Lymphedema

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Primary lymphedema is related to a developmental defects (malformation, dysplasia) of the lymphatic vessels or lymph nodes. Many studies have reported an association between several genes and primary lymphedema. Although no curative surgical or medical intervention is available for lymphedema, current standard for treatment of either primary or secondary lymphedema is two-phase complex decongestive therapy (CDT). Many studies have revealed the short-term and long-term benefits of CDT such as decreasing edema volume and improving quality of life (QOL) in patient with secondary lymphedema on limb or even head/neck area. The ultimate goals of treatment for patients with primary lymphedema are to improve not only physical impairments of affected limb or area but also QOL and functional disabilities. Especially, the treatment of children with primary lymphedema should be performed with a long-term plan, utilizing CDT. This article will explore the two-phase CDT approach according to involved area especially lower limb and discuss clinical experience on primary lymphedema treatment, comparing it with characteristics of secondary lymphedema.

Key Words: Lymphedema · Primary congenital · Therapy · Rehabilitation

INTRODUCTION

Historically, primary lymphedema was classified into just three categories depending on the age of onset of swelling; congenital, praecox and tarda. Developments in clinical phenotyping and identification of the genetic causes of some of these conditions have demonstrated that primary lymphedema is highly heterogenous. Although a new classification and diagnostic pathway for primary lymphedema have been introduced [1], the mainstay of treatment is still limited.

CDT is now well established as the standard treatment for lymphedema regardless of the etiology (primary or secondary), its site (limb, trunk, or face), or its clinical stage [2]. The best treatment protocol among whole CDT or single therapy (MLD, pneumatic pump, short-stretch bandages, compression garments, and therapeutic exercises) still could not be identified because of heterogeneity in the subject populations, measured outcomes, and follow-up durations in the current studies. Many studies have revealed the short-term and long-term benefits of CDT such as decreasing edema volume and improving quality of life (QOL) in patient with secondary lymphedema on limb [3,4] or even head/neck area [5].

However, surprisingly, there is no study that reports the effect of CDT in patient with primary lymphedema only. Few studies have reported that patients with primary lymphedema experienced distressful symptoms, functional impairments, and frequent infection [6,7].

Primary lymphedema can occur in many sites such as leg, arm, face, or genital area. Also, it is common to see patients with bilateral limb or multi-segment, more than one segment affected by lymphedema, e.g., face and arms or arms and leg [8,9]. The ultimate goals of treatment for patients with primary lymphedema are to improve not only physical impairments of affected limb or area but also QOL and functional disabilities. Especially, the treatment of children with primary lymphedema should be performed with a long-term plan, utilizing CDT.

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MAIN

1. CDT on limb

Elements of CDT include manual lymph drainage (MLD), compression therapy, decongestive exercises, basic skin care and education for self- care and risk reduction. It is performed in two phases. In phase 1, known as the decongestive phase, the patient is treated with all components of CDT on a daily basis until the limb is decongested. The patient progresses into phase 2 of CDT, self-management phase [10,11].

The current standard of care for lymphedema is CDT. However, the optimal treatment protocol remains controversial [12]. Recent study reported no significant differences in limb volume between compression garment only and CDT with daily MLD and short-stretch bandaging for arm lymphedema [13].

1) Phase 1: Decongestive phase

In this phase, the patient has intensive therapy daily, and also is informed about all components of CDT. The end of this first phase is determined by the time that the degree of lymphedema limb volume reduction reaches a plateau. The duration of this phase varies with the severity of the lymphedema, and usually averages 2 to 4 weeks for patients with one limb. In extreme patients, this phase may be extended up to 6 to 8 weeks and can be repeated several times.

MLD is a gentle manual treatment technique, based on basic Vodder strokes - stationary circle, pump, scoop and rotary designed to reroute the accumulated lymphatic fluid from involved limb/body area to healthy lymphatic and/or venous system. The most common effects of MLD are (1) increase in lymphangiomotoricity, (2) reverse of lymph flow, (3) increase in venous return, (4) soothing, and (5) analgesic effects. MLD is different from other massage techniques in that it has light directional pressure on the superficial lymphatic territories. Another unique characteristic is that the proximal part of affected limb and even contralateral limb is decongested first to treat the more distal part. High pressure techniques like Swedish muscle massages could damage lymphatic structures including anchoring filaments especially in patient with the early stage of lymphedema. However, for patient with lymphostatic fibrosis, other fibrosis techniques are applied in the target area directly with more intensity and prolonged time than the basic strokes. In this phase, MLD is applied over 30 minutes at least once a day, 5 times a week.

The effects of compression therapy are achieved with compression bandages and garments. In decongestive phase, compression therapy is applied using short-stretch bandages (up to 60% elasticity) in combination with proper pads following MLD for all day and night. Short-stretch bandages exert a very low resting pressure to avoid a tourniquet effect and also a high working pressure generated during muscle contraction. To achieve a compression gradient, it is necessary to apply bandages in layers. Bandage techniques that are commonly used are the spiral or figure of eight wrapping. The purposes to use of special pads with foam materials are not only protecting bony prominences and making the limb to a cylindrical shape and also increasing the pressure over the focal fibrotic area. In addition, it is important to educate patient or a family member to make sense of the self-bandaging techniques and skin care [14,15].

Lymphedematous skin also tends to be dry and may become thickened and scaly. Lymphedema patients are susceptible to infections of the skin and soft tissues. The process of inflammation/infection may not only worsen the swelling and discomfort, but can also develop into a serious medical crisis. Therefore meticulous care of skin and nail is essential.

Exercises consist of active, non-resistive and repetitive protocols, which should be customized by the physician and/or therapist to meet the patient's goals. The stage and type of lymphedema, as well as additional co-morbidities need to be considered for specific restrictions and limitations of muscle/joint activity. Exercises varies interstitial tissue pressure, affects lymph propulsion and clearance, transports fluid and inflammatory proteins from the edematous area [16]. Ideally decongestive exercises are performed 2-3 times daily for 10-15 minutes. Compression should be worn during exercise. High intensity and repetitive activities such as tennis, running, and mountain biking etc. may not be beneficial for patient during and immediately after phase 1 therapy.

2) Phase 2: Self-management phase

To maintain and more improve the results achieved in phase 1, the patient progresses into phase 2 of CDT. The components in this phase are similar to those in phase 1, but the point should be emphasized is self-management. The patient assumes a duty like a therapist.

Patients should continue the meticulous skin care, perform simple self-MLD, compression therapy using

elastic stocking/garment and exercises. But it is not easy for patients to self-MLD properly implemented.

Main compression in this phase is applied by compression garments during the daytime hours. Patients with limb lymphedema will usually require a compression pressure range about 30 mmHg for arm or 40-60 mmHg garments for leg, respectively. If a patient's limb is deformed or need the high degree of compression garment over 50 mmHg, custom-made garments are a better choice. Compression garments should be checked regularly and replaced every 6 months. According to the severity and chronicity of lymphedema, many patients still require short-stretch multilayered bandaging during the night or weekends in this self-management phase. The multilayered short-stretch bandaging is very effective, however, many patients complain about a self-care burden and cosmetic issue with its bulkiness. Less bulky velcro-band devices such as CircAid® and AutoFit® are recently developed or improved, but still not widely used than bandages in practice. These are applied more easily and allow better movement, and also, a few reports show good results [8].

Exercise is beneficial not only for general health but also for lymphedema itself. Gradual progression is imperative when prescribing exercises. Under water leg exercises proved to significantly enhance the effect of CDT [17].

Good patient compliance is indispensable because this phase lasts lifetime. Therefore the physician and or the lymphedema therapists educate and checkup patient regularly to maximize their compliance.

2. CDT on genital/lower abdominal area or face

In the majority of cases, genital lymphedema is combined with leg lymphedema. Because lymph cysts or lymphangioma circumscriptum commonly occur, daily meticulous skin care with disinfecting agents should be necessary for cellulitis prevention. Parts of MLD including abdominal and genital treatment and genital bandaging must be learned and performed by patient several times daily. The ready-made or custom-made special foam pads are inserted into the compression pantyhose or bandages for women. For male genital lymphedema, bandaging the penis and scrotum is not easy and often require cohesive bandages.

Isolated primary head and neck lymphedema are very rare and usually occur in combined with arm and upper trunk involvement. In the majority of facial lymphedema is unilateral. Compared to the secondary head and neck lymphedema after surgery and irradiation, primary patients have little functional problems such as limited mouth opening and neck motion etc. MLD sequence and compression methods with bandages or special compression garment are similar to the secondary lymphedema patients [18].

3. Intermittent Pneumatic Compression Pump (IPC)

The use of IPC for lymphedema therapy in decongestive phase and/or self-management phase of CDT or employed as stand-alone therapy is the most controversial issue. Some published reports support the use of IPC [19,20] but, others are not for patients with secondary lymphedema, especially after breast cancer therapy [21]. Major theoretical concerns about using IPC are generation of genital edema and development of a fibrosclerotic ring at the root of the extremity [22]. It should be avoided by careful observation and combined MLD. The American Cancer Society Working Group on the Diagnosis and Management of Lymphedema designated IPC as a potential adjunctive component of CDT. IPC is only effective when combined with low-stretch bandages or stockings to maintain edema reduction. It is still very hard to know whether a particular pressure, cycle, or cuff design is better than others. Still, the most popular pump system is non-graded sequential compression with 3 to 14 chambers. But the guidelines for patient and device selection for incorporating IPC into main therapeutic program continue to evolve. Patient factors include severity of lymphedema, involvement of the trunk or genital area, and the presence of complications that contraindicate the use of simple, classical devices [23]. Recently developed new devices that simulate manual massage and design improvements for area of coverage, ease of use, and sequence/actions may increase the therapeutic effect and patient compliance. By the way, IPC should form part of a long-term therapeutic program for patient with advanced lymphedema, particularly useful in elderly patients, patients in bed or patients with disabilities.

4. Special issues in the treatment of primary lymphedema

Primary lymphedema can occur in many sites such as leg, arm, face or genital area, but over 90% are leg lymphedema. Also, it is common to see patients with bilateral limb or multi-segment, more than one segment affected by lymphedema e.g. face and arms, or arms and leg and etc. Patients with primary lymphedema are

relatively younger compared to those with cancer therapy-related secondary lymphedema. Among those who visited my clinic, over 70% were un-der the age 30 and many of them were juvenile patients. Therefore, proper treatment planning, following medical care system model for chronic diseases that require lifetime medical care, is necessary. In addition to edema volume reduction or maintaining with CDT-based therapy, other therapeutic considerations such as the special shoe prescription including wide-depth shoe or heel lift to correct mild leg length discrepancy is required. When the degree of lymphedema is severe, functional impairment of arm or leg must be assessed regularly.

CONCLUSION

CDT-based therapy issues of primary leg lymphedema

According to the known epidemiologic data, about one third of leg primary lymphedema patients are bilateral distal leg involvements [9]. MLD sequence and compression therapy with short-stretch bandages for this type are similar to the treatment of chronic venous insufficiency or post-traumatic secondary lymphedema. Proximal parts of leg do not need to be applied of compression bandages or garments [11].

Primary leg lymphedema patients with involvement of thigh and trunk quadrant usually show extensive leg edema. Its treatment is similar to the treatment for secondary lymphedema.

According to long-term observational result in my experience, as similar to the results of other studies on the secondary lymphedema, the phase 1 CDT is also found to be effective on reduction excessive volume of the primary lymphedema patients (Figs. 1, 2). However, the amount of volume reduction in edema and long-term maintenance was relatively unsuccessful in primary lymphedema patients compared to that of secondary patients [3,4,23]. This might be due to occurrence of tissue change on the involved sites at the time of diagnosis and onset of treatment. Although, aggressive CDT based care with a strict prevention is warranted. Especially in case of children and adolescent patients, it is important to constantly educate them to understand their conditions and to manage themselves on their own. In order to overcome cosmetic and functional problems due to life-long condition, high compliance to CDT is required. Therefore, continuous education paired with professional psychological support must be emphasized above all.



Fig. 1. Primary lymphedema of bilateral distal legs before (**A**) and after (**B**) CDT.



Fig. 2. 15 years old girl with primary lymphedema of the left leg, mainly thigh involvement before (**A**) and after (**B**) CDT.

Conflicts of Interest

All authors declare no conflicts of interest.

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