CDP Treatment of Small Children and Infants with Primary (Congenital) Lymphedema

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Primary Lymphedema: Lymphedema generally is classified as primary or secondary. Primary lymphedema develops without an obvious underlying cause, and is the result of lymphatic dysplasia (malformation) which results in an accumulation of protein rich fluid in the interstitial tissues of the affected body part. In contrast, secondary lymphedema is most often the result of surgery and/or radiation treatment for cancer, trauma or infection. Primary lymphedema is reported to occur 87% in females; only 13% of all cases are in males. In most cases, the initial manifestations of primary lymphedema occur in individuals at about 17 years of age. Eighty-three percent of primary lymphedema appears before the age of 35 and is referred to as lymphedema praecox. Primary lymphedema which first appears after the age of 35 is called lymphedema tardum.

Congenital lymphedema is a form of primary lymphedema; it is present at birth or develops soon thereafter and can be either hereditary (familial) or sporadic (non-familial).

The hereditary lymphedemmas can be subdivided into two types: Type I is referred to as Nonne-Milroy-Syndrome (Milroy’s disease). In this type of familial, congenital lymphedema, the innate defect of the lymphatic system appears in the lower part of the body in the form of aplastic lymph vessels (aplasia = defective development or congenital absence). The lymphedema begins distal to the inguinal ligament (groin) and may be associated with dilated lymph vessels of the intestines which can lead to congenital chylascites and protein loss. Type II hereditary primary lymphedema is called Meige’s Syndrome. In this case, the lymphedema becomes manifest during puberty. It most often affects the lower extremities — rarely the upper extremities or the face. Other anomalies such as partial syndactylism of the toes (fusion of two or more toes), distichiasis (two rows of eyelashes), myopia (nearsightedness), yellow nails, vertebral anomalies, cerebro-vascular malformations, senso-neural hearing loss, cleft palate and bronchiectasia are often found in type II hereditary lymphedema. Hereditary (primary) lymphedema seems to be much less common than the sporadic (non-familial) primary lymphedema. (Figure 1).

Figure 1 – LYMPHEDEMA CLASSIFICATION

Pathology: Primary lymphatic dysplasia can be present as a variety of anomalies. For example: Hypoplasia of lymph vessels — the number of lymph vessels is reduced and the diameter of the existing lymph vessels is smaller than normal (most common form); hyperplasia of lymph vessels (lymphangiectasia) — the lymph vessels are bigger than normal and have insufficient valve function; aplasia (absence) of single lymph vessels or lymph capillaries — lymph node hypoplasia combined with fibrosis and groin lymphadenopathy in most cases (Kinmonth Syndrome). Any of these changes will reduce the transport capacity of the lymphatic system. Lymphedema arises when the transport capacity is not sufficient to remove the normal amount of proteins and water from the interstitium.

Diagnosis and Differential Diagnosis: Since primary lymphedema can be combined with other congenital anomalies and/or general angiodysplasia (malformation of blood as well as lymph vessel tissue), an expert physician assessment is important to identify all medical issues before treatment begins. The clinical diagnosis of primary lymphedema can usually be established without invasive (and expensive) testing. Lymphangioscintigraphy (LAS) is a lymphatic function test and can be used to confirm the diagnosis of primary lymphedema. With the results of the LAS, the expert physician or radiologist will be able to draw conclusions about the function of the remaining, poorly developed, or damaged peripheral lymphatic system. In general angiodysplasia, this test is required to assess the magnitude of lymphatic impairment. In uncomplicated extremity lymphedema with a typical medical history, the outcome of the LAS usually does not provide information that would change the treatment options for the patient. Also, if the patient decides or the parents of a small child with lymphedema decide to have Complete Decongestive Physiotherapy (CDP) to treat the lymphedema, the results of the LAS usually will not influence the treatment protocol. Ultrasound, Venous Doppler, Computer Tomography (CT scan), or Magnetic Resonance Imaging (MRI) are diagnostic tests that generally are not required in primary lymphedema of the extremities. These tests may be ordered by expert physicians in more complicated (combined) lymphedemas to subsequently provide safe treatment for each patient.

Therapy: Since lymphedema is a chronic and progressive condition, the patient faces a lifelong medical problem. When a child is born with lymphedema, the entire family is concerned. Treatment should be initiated as soon as possible but only after a thorough medical evaluation has been performed.

Complete Decongestive Physiotherapy (CDP): In CDP for small children, the focus of the treatment should be the education and training of the parents or other adult primary caregiver. The child also should be involved in the training. For example, the responsibility for the home maintenance program should be taught to the child in accordance with chronological and psychosocial guidelines. Some of these guidelines are outlined in the article, “Some Thoughts on Childhood Lymphedema,” by Pere M. Summers, OTR/L, which can be found in NLN Newsletter Volume 9, No. 1 (reprints are available).
The initial treatment sessions with a CDP certified therapist may be approached by making the child’s acquaintance (playing) and by teaching basic strokes of Manual Lymph Drainage (MLD), such as stationary circles and effleurage (in the direction of the normal lymph transport) to the primary caregiver. These MLD techniques can then be performed as often as necessary in short sessions even when the child is playing or sleeping. The frequency and duration of each session should be determined by the following parameters: The effectiveness of MLD must not be compromised, tolerance of the child, level of commitment of the primary caregiver (burnout!) etc. Special techniques such as fibrous tissue techniques (manual techniques used to soften indurated, proliferated connective tissue) should be used only by qualified therapists. After an initial series of approximately 10 to 15 sessions, the treatment should be the responsibility of the primary caregiver. The progress of these home treatments should be monitored continuously by a CDP therapist or knowledgeable physician.

Bandaging is indicated in most cases of primary lymphedema. A common problem in bandaging is the small size of the extremity and the pressure sensitivity of the delicate baby skin. The CDP therapist will have to be somewhat creative and tolerant when applying the first bandages. Padding materials are required to avoid pressure sores, blisters and irritations of the skin. Velfoam (material used in rehabilitation) often replaces the bulky conventional foam and provides adequate protection. The pressure exerted by the bandage has to be introduced gently and is increased over a comfortable period of time. Caution: Overeager parents and therapists sometimes must be reminded that the purpose of the bandage is to raise the compromised tissue pressure in lymphedema and that the bandage must never be used to “squeeze” the limb. Bandaging too vigorously may have an adverse effect on the lymphedema! In addition, it can cause tourniquet effects, or further compromise the already reduced transport of the lymph vessels. Bandaging may be the only way to restore the diminished tissue pressure after edema fluid has been evacuated through the techniques of MLD. Occasionally, it may be a good idea to give the child a rest and stop bandaging to avoid burnout! If swelling recurs, this normally can be controlled with the next MLD and bandaging session. For optimal compliance, the bandage has to be applied in a way that will not interfere with normal childhood activities (play) and development. Progress may be halted by the occurrence of spontaneous infections (lymphangitis) or normal childhood diseases such as influenza, chickenpox, etc.

The application of compression garments is often very difficult or even impossible during the first few years of life. It is realistic to assume that the first garment can be tolerated when the child reaches the age of two. These garments must be custom-made for each child. Flat-knitted fabrics such as the JUZO Elvarex seem to be the most effective and comfortable products (photo). Note that the JOBST Elvarex garment is not yet available for lower extremities here in the U.S., but is expected to be introduced in early 1998. Measuring and fitting for such small garments often creates a big challenge for the therapist. Compression class I (18-21 mm/Hg), or class II (25-32 mm/Hg) should be sufficient in most cases. Easy application of the garment and the tolerance of the child will determine the compliance.

Skin care is very important because skin has a tendency to become dry with the frequent use of bandages or garments. Mycosis of the foot (athlete’s foot), eczema, or contact dermatitis need to be addressed if they occur.

Exercise is an important component of CDP. For small children with lymphedema, no special exercise program is required as long as the child is otherwise developing normally. Should a special pediatric exercise program be necessary for conditions other than the lymphedema, it is the therapist’s responsibility to work this into the daily routine and advise the primary caregiver accordingly.

Ancillary problems such as weight control, the avoidance of tight clothing and temperature extremes also should be observed by the young lymphedema patient. In addition, any orthopedic and postural problems which may exist must be treated. In the case of lower extremity lymphedema, consultation with an orthopedic specialist is advisable. Splayfoot, which may lead to pain in the area of the metatarsal, often is observed in patients with lymphedema (one study reports that in a group of 50 randomly chosen patients with lower extremity primary lymphedema ranging from 13 to 70 years of age, 45 (90%) developed splayfoot in the swollen extremity).

Compression Pumps: Mechanical drainage via a multi-chambered, sequential compression pump is usually not effective and any results achieved are short-lived. Problems with the use of the compression pump are numerous and are not discussed in this article. In the past, only a small number of individuals reported good progress when using the pump in the treatment of their lymphedema. Nevertheless, whenever a pump is used, a knowledgeable lymphedema therapist or physician should monitor the patient frequently and check the adjacent trunk and genital areas for edema.

Surgery: Surgical interventions are often discussed for lymphedema. There is no safe or reliable surgical treatment available which restores the reduced transport capacity in primary extremity lymphedema. Resection (debunking) procedures introduce much scarring and the long-term results are often unsatisfactory. Lymphatic surgery is sometimes necessary, but should be reserved for special cases, e.g. chylos or other reflux syndromes.

Conclusion: Primary (congenital) lymphedema is a chronic, progressive condition that deserves serious medical attention. A cure for lymphedema is not available, but a patient or parent who follows the recommended program of CDP as part of a daily routine will find great relief from the symptoms. With good compliance, the patient will be less prone to developing serious complications (infections) and, most importantly, avoid the progression of the lymphedema.

References:
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