NATIONAL LYMPHEDEMA NETWORK

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The NLN Mission Statement

The mission of the NLN is to create awareness of lymphedema through education and to promote and support the availability of quality medical treatment for all individuals at risk for or affected by lymphedema.

The NLN is dedicated to:

- promoting research into the cause, prevention and treatment of lymphedema;
- securing adequate insurance coverage for medically necessary, safe and effective treatment;
- expanding the number and geographical distribution of lymphedema treatment facilities and certified therapists.

To achieve these goals, the NLN disseminates information about lymphedema to health care professionals so they can appropriately counsel their patients on its avoidance, and prescribe safe, effective treatment for those affected by this condition. The NLN also provides this information to the general public.

Pediatric Lymphology: Diagnosis And Treatment

By Ethel Foeldi, MD, and Guenter Klose, CI, CLT-LANA

Lymphatic diseases such as lymphedemas of the extremities, genitalia, or the head and neck, as well as chylous effusion (collection of lymph fluid in one specific area of the body) of the abdomen or thorax may be present at birth (congenital), or develop without obvious cause later in childhood or adolescence (primary). A more detailed classification of primary lymphedema is omitted, as it was part of an earlier *LymphLink* article on the management of childhood and adolescent lymphedema by Joseph Feldman, MD (April 2004) *[Reprints available through the NLN]*. Special attention must be given in the diagnosis of lymphatic diseases in children and adolescents.

Malignancies in children are rarely the cause of disease of the lymph vascular system, whereas malignancy always must be considered as a reason for the development of LE and/or chylous effusion in adults.

In patients who present with primary lymphatic diseases, it is especially important to check for additional congenital malformations. For example, lymphatic diseases manifesting as lymphedemas of the extremities *(Photo 1)* can be combined with lymphostatic protein-losing enteropathy (lymphedema of the small bowel which causes protein loss through the stool), lymphangioma (a benign tumor representing a congenital malformation of the lymphatic system, or chylous effusion. In addition, combination forms with venous and/or arterial malformations also exist but these are not discussed in this article.

Additional congenital malformations were found in 4% of children and adolescents (n=532) treated for lymphedema at the Foeldi Clinic. It is noteworthy that in these 4% of cases in multiple extremities, the genitalia as well as the face including the conjunctiva, were also often involved in the lymphedema.

DIAGNOSIS

Lymphedema of the extremities can be diagnosed through clinical examination (history, inspection and palpation). Lymphangioscintigraphy (LAS) is a lymphatic function diagnostic test, which can be used to confirm the diagnosis of primary lymphedema. LAS is used when the results of the clinical examination are questionable. Ultrasound examination is important if chylous effusion is suspected. Needle aspiration in patients with chylous effusion will show elevated levels of triglycerides and lymphocytes in the extracted chylous (lymph) fluid. In rare cases, examination by MRI, MR-Angiography or laparoscopy is necessary. Lab examination of the stool is

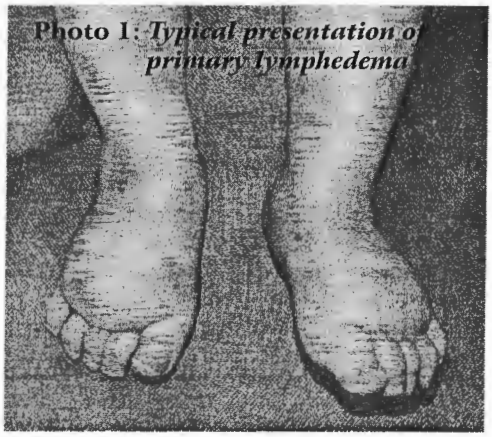

Photo 1: *Typical presentation of primary lymphedema*

helpful if protein-losing enteropathy is suspected. Diarrhea is common after the ingestion of a fatty meal in patients with protein-losing enteropathy and a blood sample may show hypoproteinemia (low protein concentration in the blood), decreased calcium level, or elevated TSH (thyrotrophic stimulating hormones). Thickening of the intestinal wall may be evident in ultrasound. An endoscopy with biopsy of small bowel should be mentioned as part of further diagnostics, if necessary.

Continued on page 2

In This Issue

Pediatric Lymphology...

Continued from cover page

DIAGNOSIS:

Lymphedema:
History
Clinical assessment

Chylous effusions:
Clinical assessment
Ultrasound
Needle aspiration of chylous fluid (triglyceride, calcium, lymphocytes)
MRI
If necessary MR-Angiography, Laprascopy

Protein-losing Enteropathy
Stool
Hypoproteinemia, Calcium, Blood test, TSH
Thickening of the intestinal wall (Ultrasound, MRI)
If necessary endoscopy with biopsy

prognosis of primary lymphedema is very good if adequate Complete Decongestive Therapy (CDT) is provided to the patient. After successful CDT, therapeutic selfcare protocols will be part of patient's daily routine into adulthood. Furthermore, the parent or primary caregiver must be educated about the patient's condition and learn how to provide the proper psychological support and lifestyle (e.g., diet, garment use, etc.).

In combined congenital malformations, when timely and adequate therapy is lacking, severe complications can develop that can even lead to the patient's death. The most common severe complications include sepsis with multi organ failure, meningitis, endocarditis and pneumonia.

DIET

A long term MCT-diet (MCT = medium chain triglyceride) is necessary in case of chylous effusion and protein-losing enteropathy. In severe cases, children may need substitution of protein, calcium and/or fat-soluble vitamins.

SURGERY

Surgical intervention is not part of the management of uncomplicated primary LE, however, it may be indicated in patients with large lymphangiomas. Surgery should also be considered in patients who developed lymphangioma without additional lymphedema; nevertheless, the technique and the timing of the surgery need to be planned carefully.

THERAPY:

Lymphedema of the Extremities:
Complete Decongestive Therapy (CDT)

Chylous Effusion; Protein-losing Enteropathy:
MCT-Diet, CDT, if necessary, surgery

Lymphangioma:
If necessary, surgery

COMPLETE DECONGESTIVE THERAPY (CDT)

In CDT 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, i.e., the responsibility for the home self-care program should be taught to the child in accordance with his/her age and ability.

MANUAL LYMPH DRAINAGE (MLD)

The initial treatment sessions with a CDT certified therapist may be approached by making the child's acquaintance (playing) and by teaching basic strokes of MLD such as stationary circles and effleurage (in the direction of the normal lymph transport) to the primary caregiver. Special attention should be given to the MLD trunk treatment (e.g., abdomen) because of the importance to "clear" the central regions of the body prior to extending the treatment to the extremities *(Photo 2)*. These MLD techniques can then be performed as often as necessary in short sessions even when the child is playing or sleeping. Only qualified therapists should use special techniques, such as fibrous tissue techniques (manual techniques used to soften indurated, proliferated connective tissue). After an initial series of 10 to 15 sessions with a certified LE therapist, the treatment should be the responsibility of the primary caregiver. A CDT certified therapist or knowledgeable physician should monitor the progress of the home treatment continuously.

Photo 2: ***MLD–parents receive instructions from therapist during treatment***

Photo 3: ***Bandaging of small toes***

BANDAGING

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 CDT therapist will have to be somewhat creative and considerate when applying the first bandages. Padding materials are required to avoid pressure sores, blisters and irritations of the skin. *The pressure exerted by the bandage has to be introduced gently and is increased over a comfortable period of time (Photo 3).*

Caution: Overeager parents and therapists 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.

COMPRESSION GARMENTS

Compression garments can be made for babies and toddlers, however, the application is often difficult or even impossible during the first few years of life. It is realistic to assume that the garments can be tolerated when the child reaches the age of two. These garments must be custom-made for each child. Flat-knitted fabrics seem to be the most effective and comfortable products. Measuring and fitting for such small garments often create 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. Keeping in mind that the ease of donning and the comfort of the child will determine the compliance with wearing the garment. After the initial decongestion has been achieved and the child is transitioned into the home care phase, a compression sleeve or alternative compression bandage may also be considered as an alternative to the compression bandaging at night.

SKIN CARE AND EXERCISE

Skin care, especially the application of a moisturizing lotion is very important because the skin has a tendency to become dry with the frequent use of bandages and/or garments. Meticulous skin and nail care will help to avoid the development of infections such as cellulitis. Mycosis of the foot (athlete's foot), eczema, and contact dermatitis need to be addressed if they occur. Exercise is also an important component of CDT. For small children with lymphedema, no special exercise program is required as long as the child is otherwise developing normally. The young patient with lymphedema should also monitor ancillary problems such as weight control, avoidance of tight clothing, and temperature extremes.

GENETICS AND LYMPHANGIOGENESIS

In recent years, researchers have been able to link some of the hereditary forms of primary lymphedemas to particular chromosomes and genes. In addition, lymphangiogenesis (the growing of new lymphatic vessels) has been investigated as future treatment for lymphedema. More time and research will be necessary in order to gain a therapeutic benefit from these studies.

CONCLUSION

Primary (congenital) lymphedema is a chronic and progressive condition that deserves serious medical attention. The diagnosis of uncomplicated extremity lymphedema can be established clinically whereas special testing is needed in more involved cases, e.g., chylous effusion, protein-losing enteropathy, etc. A cure for lymphedema is not available, but a patient who follows the recommended program of CDT as part of a daily routine will find great relief from the symptoms. With good adherence, the patient will be less prone to developing complications (cellulitis infections) and most importantly, avoid the progression of the lymphedema/lymphatic disease. ❐

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