

Physiotherapy results in a baby with congenital lymphedema: a follow-up study

Türkan Akbayrak¹, İlkim Çıtak¹, Funda Demirtürk¹, Mintaze Kerem², İnci Akarcalı¹

Departments of ¹Connective Tissue Manipulation and ²Pediatric Rehabilitation, Hacettepe University School of Physical Therapy and Rehabilitation, Samanpazarı, Ankara, Turkey

SUMMARY: Akbayrak T, Çıtak İ, Demirtürk F, Kerem M, Akarcalı İ. Physiotherapy results in a baby with congenital lymphedema: a follow-up study. Turk J Pediatr 2002; 44: 349-353.

Physiotherapy results of a 6.5-month-old baby with congenital lymphedema in the lower limbs are presented in this study. Her motor developmental level and reflexes were evaluated and test results did not show any abnormal findings. Assessment of limb volume included circumferential and volumetric measurements, and photographs showing the changes in macroscopic view are presented. The physiotherapy program consisted of manual lymphatic massage, remedial exercises, multilayered inelastic compression bandaging, meticulous skin care and education of parents. The treatment lasted for 2.5 months, five days per week. Evaluations were done immediately after treatment and six months after treatment. The evaluations showed reduction in swelling after the treatment and this reduction continued during the follow-up period. It can be concluded that this physiotherapy program reduces the swelling in a baby with congenital lymphedema. Further studies are required in order to see the effectiveness of this therapy program in a greater number of subjects.

Key words: congenital lymphedema, manual lymphatic massage, remedial exercises, bandaging.

Lymphedema is the chronic swelling of one or more extremities resulting from a malformation of the lymphatic system, causing lymph stasis and accumulation of interstitial fluid^{1,2}.

Lymphedema can be classified as primary (idiopathic) or secondary^{1,3}. Primary lymphedema is related to in utero developmental defect and affects 1.15/100,000 persons younger than 20 years of age¹. Although in reality present at birth, it is described according to age at presentation: congenital (present at birth to 1 year), praecox (from 1-35 years), and tarda (occurring after the age of 35 years)². Primary lymphedema leads to disabling and disfiguring swelling of the extremities⁴.

Lymphedema, regardless of the etiology, is essentially incurable and can have a negative effect on physical and psychological well being⁵. Different therapy approaches exist for lymphedema which serve to contain swelling⁶. The objectives of treatment are to reduce swelling, restore shape, and prevent inflammatory episodes, e.g.

recurrent cellulitis⁶. There are essentially three main approaches to lymphedema treatment: physical therapy, drug therapy and surgery⁶. The principle of physical therapy is to reduce excessive capillary filtration and improve drainage of interstitial fluid and macromolecules from congested regions to normally draining lymph node sites⁶. This is achieved through a combination of compression, exercise and massage⁶.

This study presents the early and long-term physiotherapy results of a 6.5-month-old baby with congenital lymphedema in her lower limbs.

Case Report

The swelling of the lower limbs of the baby was first noticed by her parents at two months of age. She was examined by pediatricians and diagnosed as congenital lymphedema. She was then referred to Hacettepe University, School of Physical Therapy and Rehabilitation, Connective Tissue Manipulation and Pediatric Rehabilitation Units.

Physical characteristics of the subject were recorded (Table I). The subject was initially evaluated with regard to motor developmental and reflex levels as described by Bobath⁷, and the test results did not show any abnormal findings.

Evaluation of the limb volume consisted of volumetric measures using limb submersion in water, and circumferential measures of limbs at various points⁸⁻¹¹ (Table II). The evaluations were performed at first visit, after the 2.5-month treatment course and at the follow-up visit (6 months after treatment), in order to assess changes in the amount of swelling. Changes in macroscopic view of the lower limbs of the baby were followed with photographs (Figs. 1-3).

The physiotherapy program included manual lymphatic massage, multilayered inelastic

compression bandaging, remedial exercises and meticulous skin care^{6,12,13}. The treatment lasted for 2.5 months, five days per week. The mother was also educated about these methods and was advised to repeat them at home after each treatment session and during the follow-up period.

The pre-treatment, posttreatment and follow-up values of circumferential measurements are presented in Table II and volumetric measurement values in Table III. The values reflected reduction in swelling. This reduction can also be observed from the photographs (Figs. 1-3). Motor developmental and reflex levels were repeated after the treatment and at the follow-up visit. Test results of all evaluations showed no abnormal findings.

Table I. Physical Characteristics of the Subject During the Follow-Up Period

Physical Characteristics	Height (cm)	Weight (g)	Head circumference (cm)
First visit	69	8550	43
After treatment	71	8750	44
Six Months after treatment	75	9400	46

Table II. Circumferential Measurement Values (cm)

Measurement levels	Before treatment (R)	After treatment (R)	Control (R)	Before treatment (L)	After treatment (L)	Control (L)
Malleolus	14	14	14	15.5	15	13.3
M+2 cm	15.1	15	13.2	15.6	15	13.2
M+4 cm	18	16	15.5	18.5	17	15.2
M+6 cm	19.1	18	17	19.5	18	16
M+8 cm	18.5	18	17.3	18.6	17.5	17.5
M+10 cm	-	-	16.5	-	-	17
M+12 cm	-	-	16.5	-	-	16.5
M+14 cm	-	-	19	-	-	17.5
Knee	20	20	19.1	20.1	20	19
K+2 cm	22.5	21	19.1	21	21	19.2
K+4 cm	23	23	21	24	22.3	20.2
K+6 cm	25.5	23.6	22	25	23	21.2
K+8 cm	-	-	22	-	-	21.2
K+10 cm	-	-	22.5	-	-	22.5

M: Malleolus.

K: Knee.

R: Right lower limb.

L: Left lower limb.



Fig. 1. Macroscopic view of the subject before treatment.



Fig. 2. Macroscopic view of the subject after treatment.



Fig. 3. Macroscopic view of the subject six months after treatment.

Table III. Volumetric Measurement Values (ml)

Volumetric values	Right lower limb	Left lower limb
First visit	250	250
After treatment	200	175
Six months after treatment	125	125

Discussion

Studies about congenital lymphedema are usually related to the diagnosis, etiology, pathogenesis, evaluation and surgical management of the problem¹⁴⁻¹⁶. Recent foreign literature about physiotherapy results has usually been related to post-surgical lymphedema¹⁷⁻²².

The reported patient was diagnosed as congenital lymphedema. When the baby was referred to us, we planned a physiotherapy program in the light of the related literature. To our knowledge, there has not been any study performed in Turkey investigating the results of physiotherapy programs in such a case. We performed the physiotherapy program as outlined in the case report section and found improvement as shown in Tables II and III.

Ohkuma¹⁰ investigated the effectiveness of microwave and elastic dressing in 30 cases of primary and secondary lymphedema. He concluded in his study that treatment of lymphedema by microwave and elastic dressing appears satisfactory, particularly when given at an early stage of the disease. Although the case characteristics and therapy approaches of our study differed from the work of Ohkuma, we also had the advantage of starting therapy at an early phase, and this may be one of the factors of success in our case.

Ko et al,¹³ reported that complete decongestive physiotherapy, including manual lymphatic massage, multilayered inelastic compression bandaging, remedial exercises, and meticulous skin care, is a highly effective treatment for both primary and secondary lymphedema¹³. Our study included similar approaches for reducing the limb volume of the baby. We noticed that manual massage was well accepted by the baby, so this method can be used easily by therapists and parents. The reaction of the baby to bandaging was not as positive as it was to the massage; however, we advised the parents to use it as it was taught, while carrying the baby daily and during bed rest. Since the case was an infant, remedial exercises were presented as

playing activities and tactile stimulation was used for facilitation of the pumping activity of muscles. Skin care was important because the skin of the lower limbs was dry and stretched. We thus advised the parents to keep the skin moist and clean.

To our knowledge, this is the first study investigating a physiotherapy program result in such a case. Nevertheless, further studies are required in order to see the effectiveness of this therapy program in a greater number of subjects.

REFERENCES

- Smeltzer DM, Stickler GB, Schirger A. Primary lymphedema in children and adolescents: a follow-up study and review. *Pediatrics* 1985; 76: 208-218.
- Mandell GA, Alexander MA, Harcke HT. A multiscintigraphic approach to imaging of lymphedema and other causes of the congenitally enlarged extremity. *Semin Nucl Med* 1993; 23: 334-346.
- Wright NB, Carty HM. The swollen leg and primary lymphoedema. *Arch Dis Child* 1994; 71: 44-49.
- Ferrel RE, Levinson KL, Esman JH, et al. Hereditary lymphedema: evidence for linkage and genetic heterogeneity. *Hum Mol Genet* 1998; 7: 2073-2078.
- Marcks P. Lymphedema. Pathogenesis, prevention, and treatment. *Cancer Pract* 1997; 5: 32-38.
- Mortimer PS. Therapy approaches for lymphedema. *Angiology* 1997; 48: 87-91.
- Bobath B. *Abnormal Postural Reflex Activity Caused by Brain Lesions* (3rd ed). London: Heinemann; 1985: 137-195.
- Gerber LH. A review of measures of lymphedema. *Cancer* 1998; 83: 2803-2804.
- Pfalzer LA. Oncology: examination, diagnosis, and treatment. Physical therapy considerations. In: Myers RS (ed). *Saunders Manual of Physical Therapy Practice* (1st ed) Chapter V. Philadelphia. W.B. Saunders Company; 1995: 149-191.
- Ohkuma M. Lymphedema treated by microwave and elastic dressing. *Int J Dermatol* 1992; 31: 660-663.
- Campisi C. Lymphedema: modern diagnostic and therapeutic aspects. *Int Angiol* 1999; 18: 14-24.
- Rinehart-Ayres ME. Conservative approaches to lymphedema treatment. *Cancer* 1998; 83: 2828-2832.
- Ko DS, Lerner R, Klose G, Cosini AB. Effective treatment of lymphedema of the extremities. *Arch Surg* 1998; 133: 452-458.

14. Caner B, Günaydin E, Yalaz K, et al. Lymphoscintigraphic evaluation of primary congenital lymphedema. *Turk J Pediatr* 1990; 32: 117-121.
15. Hourihane JO, Bennet, CP, Chaudhuri R, Robb SA, Martin ND. A sibship with a neuronal migration defect, cerebellar hypoplasia and congenital lymphedema. *Neuropediatrics* 1993; 24: 43-46.
16. Irrthum A, Karkkainen MJ, Devriendt K, Alitalo K, Vikkula M. Congenital hereditary lymphedema caused by a mutation that activates VEGFR3 tyrosine kinase. *Am J Hum Genet* 2000; 67: 295-301.
17. Humble CA. Lymphedema: incidence, pathophysiology, management, and nursing care. *Oncol Nurs Forum* 1995; 22: 1503-1509.
18. Földi E. The treatment of lymphedema. *Cancer* 1998; 83: 2833-2834.
19. Rockson SG. Precipitating factors in lymphedema: myths and realities. *Cancer* 1998; 83: 2814-2816.
20. Földi E. Treatment of lymphedema and patient rehabilitation. *Anticancer Res* 1998; 18: 2211-2212.
21. Mortimer PS. The pathophysiology of lymphedema. *Cancer* 1998; 83: 2798-2802.
22. Ramos SM, O'Donnell LS, Knight G. Edema volume, not timing, is the key to success in lymphedema treatment. *Am J Surg* 1999; 178: 311-315.