

Complex decongestive physiotherapy in paediatric patients with precox primary lymphoedema

Hanife Abakay, Hanife Doğan and Ayşe Güç

Key words

Body image, children, lymphoedema precox, quality of life, volumetric measurement

Hanife Abakay is Assistant Professor, Yozgat Bozok University, Sarıkaya School of Physiotherapy and Rehabilitation, Yozgat, Turkey. Hanife Doğan is Assistant Professor, Yozgat Bozok University, Sarıkaya School of Physiotherapy and Rehabilitation, Yozgat, Turkey. Ayşe Güç is Specialist Doctor, Kayseri City Hospital, Physical Therapy and Rehabilitation Hospital, Kayseri, Turkey.

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Primary lymphoedema results from congenital lymphatic damage that requires lifelong management of symptoms (e.g. swelling, pain, heaviness).

The accumulation of protein-rich fluid in the subcutaneous tissue as a result of the failure of the lymphatic system, which returns water and protein in the interstitial spaces to the blood circulation, is called lymphoedema (LO; Sander et al, 2002; Duygu et al, 2020).

The aetiological classification of lymphoedema is primary (idiopathic) and secondary lymphoedema (SLO; Duygu et al, 2020). With the exception of inguinal lymph node fibrosis, the aetiology of primary lymphoedema (PLO) is unknown. PLO is a disease that can manifest in the intrauterine period, is characterised by oedema starting from the dorsum of one or both feet, develops due to developmental anomalies in the lymphatic vessels, and can progress proximal to the lower extremity (Schook et al, 2011).

LO may occur as a result of hypoplasia, hyperplasia or aplasia in lymphatic vessels. When PLO is classified according to the age at which it occurs, it is divided into three

Abstract

Aim: The aim of this study was to investigate the effect of complex decongestive physiotherapy (CDP) on oedema, quality of life and body image in children with lymphoedema precox.

Methods: The study sample was 20 children (10 girls and 10 boys), with a mean age of 10.5 ± 1.1 years. The effectiveness of CDP was determined with a volumetric measurement. **Results:** After CDP treatment of the children with lymphoedema precox, the limb volumes decreased ($P < 0.05$), PedsQL total scores increased ($P < 0.05$), and body image scores decreased ($P < 0.05$). **Conclusion:** The use of CDP for the treatment of lymphoedema in children with lymphoedema precox was seen to decrease lower-extremity limb volume (and thus presumably oedema), improve body image scores and improve quality of life.

groups as LO congenitum, LO precox and LO tarda. LO precox is the most common type of LO (94%), developing before the age of 35 years, and showing autosomal dominant inheritance as a result of *FOX2* gene mutation. It generally emerges during puberty or pregnancy, and unilateral oedema is mostly seen below the knee in the lower extremities (Duygu et al, 2020).

Complex decongestive physiotherapy (CDP) is an evidence-based standard treatment in the treatment of LO (Ewertz and Jensen, 2011; Executive Committee, of the International Society of Lymphology, 2020).

The main aim of treatment is to reduce the extremity volume and return the LO to the latent stage by providing lymph drainage from the affected area to the healthy area using healthy lymphatic vessels and lymphatic pathways. CDP consists of four basic components of manual lymph drainage, skin care, compression therapy and exercise (Ewertz and Jensen, 2011).

In addition to the disease-related symptoms, the changes in body image and loss of some functions that occur in LO diminish quality of life (Finnane et al, 2011). Individuals with lower-extremity lymphoedema (LLO) can experience decreased mobility and psychological symptoms, such as addiction, social

isolation, stress, hopelessness, and increased pressure in social relationships, all of which are associated with decreased quality of life (Finnane et al, 2011). Studies in the literature have mostly focused on the effect of SLO on quality of life and body image in adult cancer patients (Teo et al, 2015; Jørgensen et al, 2021), while studies investigating the quality of life of PLO patients have also been conducted only on adults (Stolldorf et al, 2016).

Therefore, the aim of this study was to investigate the effect of CDP on volumetric measurements, quality of life and body image in paediatric patients with LO precox.

Materials and methods

The study was approved by the Kayseri City Hospital Clinical Research Ethics Committee (637/15.05.2022.). Written informed consent was obtained from each patient, and the study was conducted in accordance with the principles of the Declaration of Helsinki.

This study was conducted in a physical therapy and rehabilitation hospital. A total of 26 children were initially enrolled. Subjects were excluded if they had been treated for lymphoedema in the last year, or had a diagnosis of severe heart failure and/or arrhythmia, arterial disorders, kidney diseases or infection in the affected lower

Table 1. Demographic characteristics of children with primary lymphoedema.

	n(%)	Median (min-max)	Mean ± SD
Sex			
Female	10 (50%)		
Male	10 (50%)		
Age (years)		10 (9-12)	10,5 ±1,1
BMI (kg/m ²)			20.9±1.74
Affected extremity			
Right	8(40 %)		
Left	12 (60%)		

extremity.

Further exclusions were made of three children who lived outside the city and could not attend treatment regularly, two children with active infections during the treatment period, and one who had been treated with CDP 9 months previously. Thus the study involved 20 children.

Procedures

A record was made of the socio-demographic characteristics, physical characteristics (sex, dominant extremity, age, height, weight, BMI, existing and/or previous diseases, previous operations, LO history and classification) of the children who met the study inclusion criteria. Quality of life was assessed with the Pediatric Quality of Life Inventory (PedsQL 4.0) and body image with the Child’s Body Image Scale before and after CDP.

Circumference and volumetric measurements were used for the oedema evaluation.

Circumference measurement

With the patient in a semi-sitting position and the foot and ankle in the neutral position, circumference measurements were made at 5 cm intervals from the ankle lateral malleolus level to the proximal. The measurements were taken using a flexible, non-elastic, 7mm wide tape measure marked to 0.1 cm and recorded in centimetres. Leg-O-Meter was used for all measurements to ensure standardisation.

Volumetric measurement

The extremity volume was calculated using the frustum formula. The difference between both extremities (healthy and affected) was determined. Measurements were made twice, before and after the treatment. The frustum formula is:

$$V = [h \times (R1^2 + R1.R2 + R2^2)] / (12 \times \pi)$$

Where V=volume, h=range used in circumference measurement, R1=base circumference of conical segment, R2=upper circumference of conical segment. For ease of calculations, π=3 was used.

Complex decongestive physiotherapy

The children were treated in the Physical Therapy and Rehabilitation Department of Kayseri City Hospital. The treatment was carried out 5 days a week, for 4 weeks, for a total of 20 sessions, with each session lasting 60 minutes. All the measurements and evaluations were made twice, before starting the treatment at the first session and when the treatment was completed. The CDP was applied to the affected extremity of each child.

Pediatric Quality of Life Inventory

This scale was developed by Varni et al. (2005) to evaluate health-related quality of life in children. The inventory consists of 23 items in four subdimensions of physical functions (eight items), emotional functions (five items), social functions (five items), and school functions (five items). There are separate scales for children aged 5–7 years, 8–12 years, and 13–18 years. All the forms, including the family forms, have exactly the same items, and the only differences are in terms of items suitable for the child’s developmental stage and language which can be understood by a child of that age. The inventory is scored on a 5-point Likert scale, with higher total scores indicating a better health-related quality of life.

Evaluation of body image

Body image was measured with the Children’s Body Image Scale (BIS). The scale includes seven sex-specific images in anatomic positions, so there are separate

versions for boys and girls showing either seven male or seven female photographs. Each photograph represents a different BMI range, with the pictures ordered from A to G according to the increase in BMI. The children are asked to select the picture that they perceive as closest to their own body size and shape.

By subtracting the actual BMI value from the perceived BMI value, a score is calculated ranging from –6 to +6. A negative score indicates that the child perceives him/herself as smaller than he/she actually is, zero indicates the equivalence of perception and reality, and a positive score indicates that the child sees him/herself as overweight (Truby and Paxton, 2002; Saxton et al, 2009). The scoring was applied based on the age-relevant male and female BMI values.

Statistical analysis

Data were analysed statistically using SPSS 21.0. Continuous variables were stated as mean ± standard deviation, median (minimum–maximum) values, and categorical variables as number (n) and percentage. Conformity of the data to normal distribution was assessed with the Kolmogorov Smirnov test. The Wilcoxon signed-rank test was used to evaluate differences between dependent groups of data. A p-value <0.05 was accepted as the level of statistical significance.

Results

Evaluation was made of a total of 20 children with unilateral lower extremity LO, with a mean age of 10.5 ± 1.1 years (range, 8–12 years) and mean BMI of 20.9 ± 1.7 kg/m². The lower extremity involvement was right side in 40% and left side in 60%. The demographic characteristics are shown in Table 1.

The data showing the effect of CDP on volumetric measurement, PedsQL total and BIS values are given in Table 2. The volumetric measurement decreased from 4,016 ml before treatment to 3,821 ml after CDP treatment (p<0.05). The PedsQL total score was 75.4 points before treatment, and increased to 76.2 points after treatment (p<0.05). There was a statistically significant decrease in BIS values (p<0.05).

Discussion

The aim of this study was to compare the

Table 2. Comparison of before and after CDP outcomes in children with primary lymphoedema.

	Pre-CDP (n=20)	Post-CDP (n=20)	p-value
Volumetric measurement (ml)	4,016 (2,627–4,684)	3,821 (2,460-4,454)	0.000*
PedsQL score	75.4 (68.8–81.4)	76.2 (69,3-83,4)	0.000*
Body Image Scale	-1 (-4 to 0)	-2 (-4 to 0)	0.002*

*Wilcoxon test. CDP = complex decongestive physiotherapy; PedsQL = Pediatric Quality of Life Inventory

effect of CDP applied to children with LO praecox on the volume of oedema, quality of life and body image. To the best of our knowledge, this is the first study to examine the effect of CDP on quality of life and body image in paediatric patients with LO praecox.

The literature indicates CDP is an effective method at every stage of LO, because it reduces the volume difference between the healthy and affected limbs and improves quality of life (Kim et al, 2007; Lasinski, 2013). Noh et al. (2015) investigated the effectiveness of CDP in upper extremity LO, SLO and PLO. In a recent study by Abakay et al. in 2021, the effectiveness of CDP was compared in PLO patients with a mean age of 51.65 years and SLO patients, and the decrease in the amount of oedema was found to be similar in both groups. In terms of quality of life after CDP, a greater improvement was obtained in patients with SLO.

To the best of our knowledge, there are only three case reports in the literature which have investigated the efficacy of CDP on PLO patients (Kaya et al, 2010; Hwang et al, 2015; Kiloatar et al, 2017). Those three reports all showed that CDP is effective in reducing oedema; one of the cases was 2 years old (Hwang et al, 2015), and the other two cases were 61 and 62 years old, respectively (Kaya et al, 2010; Kiloatar, 2017). In a cohort study by Vignes et al. (2020), the effectiveness of CDP was demonstrated with scintigraphy in 222 PLO patients (mean age 45.8 years), but patients with congenital and lymphoedema praecox were not included.

In the current study, a mean reduction of 195 ml (4.85%) in the volume of the affected extremities was obtained in LO praecox patients, similar to the findings of other authors. However, this study showed for the first time that CDP is also effective in LO praecox patients. It can be considered

that it is important that LO praecox paediatric patients should be followed up and referred for treatment in the early period before they reach adolescence, and this will contribute to the prevention of problems that may occur during puberty.

There are many studies in the literature showing the negative effects of LO on quality of life and body image in breast and genitourinary cancer patients (Pusic et al, 2013; Kim et al, 2015; Penha et al, 2016). In a study of 1,054 patients with SLO, it was shown that LLO symptoms have a negative effect on quality of life and self-image (Carter et al, 2021). In another study comparing the quality of life symptoms of PLO and SLO patients, it was reported that patients with PLO had higher general health and quality of life than patients with SLO (Huggenberger et al, 2015).

The current study is the first to show that quality of life and body image perception can be improved by reducing the symptoms of LO in LO praecox patients. The body image score, which was -1 before CDP, decreased to -2 after CDP. This result, which can be attributed to the low body image awareness of young children, shows the need to recommend that parents monitor their children more closely. Reducing oedema symptoms in LO praecox will undoubtedly be beneficial for patients' quality of life and body image.

The main strength of this study was that the effectiveness of CDP was shown for the first time in paediatric patients with PLO, and the effectiveness of the LO treatment was evaluated with quality of life and body image questionnaires developed specifically for children. A limitation of the study could be said to be the small sample size. However, although this research was planned and presented as a pilot study, it can be thought to fill a gap in the literature. Another limitation may be that LO was not evaluated with a more objective

method such as bioimpedance analysis, tonometry and perometry. As these devices were not available in our clinic and the environmental measurement method is useful and practical, this evaluation method was selected.

Conclusion

The results of this study demonstrated that lower extremity volumetric measurements decreased with CDP applied in the treatment of lymphoedema in children with PLO, and improvements were determined in quality of life and body image. Nevertheless, there is a need for further, controlled studies in a larger population of different types of PLO to be able to confirm this significant effect of CDP on quality of life and body image.

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