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Lymphatic Clinical Severity Score (LCSS): Proposal of a New Clinical Method for the Appraisal of the Peripheral Lymphatic Diseases



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Abstract

An accurate clinical appraisal of lymphoedema patients is often a challenge because the lymphoedema disability scales commonly used in rehabilitation are not completely suitable to a proper classification of such subjects. Therefore, the choice of how charging the single lymphomatous patient to health public and private insurance systems worldwide is difficult. In order to address this issue, the authors defined and propose an evaluation system, called Lymphatic Clinical Severity Score (LCSS) that, analogously to the Venous Clinical Severity Score used in the assessment of venous chronic diseases, is aimed to assign the patient to a determinate clinical level. The LCSS score therefore results a useful parameter in evaluating whether to take in charge a patient is appropriate and how to do it. The LCSS has been successfully tested on 185 subjects with primary and secondary lymphoedema. The LCSS score determination can be repeated at the end of treatments and can be recalculated in the follow-up allowing a numerical temporal evaluation of the sickness.

Keywords: Lymphoedema; clinical assessment; Lymphatic Clinical Severity Score

Introduction

The need to synthetically classify the clinical seriousness level of patients with venous chronic disease suggested the development of a score system, the Venous Clinical Severity Score (VCSS) that corresponds to the sum of many different scores relative to the degree of the severity of the clinical findings that characterize each patient [1,2]. For some items, in the follow-up, it is possible to observe a decrease in the total score that reflects a clinical improvement and indicates a reduced, if still present, need for assistance. Lymphoedema patients present analogous classification problems and often public and private insurance systems dispute about the assignment of a patient to the regimen of assistance chosen (outpatients' department, Day Hospital, admission as an inpatient) [3-6]. Both primary and secondary lymphoedema are chronic disorders in which the interstitium is soaked by fluids with elevated protein concentration [4-8]. The high protein concentration in the interstitial fluid triggers the fibroblasts' transformation into fibrocytes with enhanced production of collagen resulting in tissue sclerosis.

This is the reason because lymphoedema induces an early fibrosis that compromises the full recovery of the oedema itself. These mechanisms facilitate both acute and chronic phlogosis [9] as well as dystrophic and dysplastic processes and, in some cases, trigger the genesis of sarcomas [10]. The evaluation scales commonly used in rehabilitation (Barthel, FIM, SMWT etc.) [11-14] are often unreliable to describe and emphasize all the psychosomatic aspects observed in lymphoedema patients and this pave the way to claims and contestations with reference to the charge of these subjects by public and private insurance systems. In order to address this issue and to provide an objective tool, the authors developed a score system, the Lymphatic Clinical Severity Score (LCSS), aimed to evaluate the peripheral lymphatic both primary and secondary disorders. LCSS, analogously to the Venous Clinical Severity Score (VCSS) already used in the assessment of venous chronic diseases, is aimed to assign the patient to a determinate clinical level. The LCSS has been successfully clinically tested.

Materials and Methods

Methods

Lymphoedema is often associated to a series of anatomicfunctional complications that result, sooner or later, in the corresponding clinical findings with different intensity levels. Among them, the most important are:

- a) Oedema
- b) Increase in tissue texture (consistency)
- c) Inflammation [15
- d) Cutaneous dystrophy [16]
- e) Recurrent dermato-lymphangio-adenitis (often erisipelalike)
- f) Pain [17]
- g) Lymph ulcerations [18]
- h) Joint involvement with functional impairment [16,17]
- i) Muscular hypotrophy [4,10,17]
- j) From all these findings the need to wear an elastic garment, at least in some hours of the day [1,2,4,16], could

spring out. This need must be considered as a tenth element in the evaluation of patient suffering from lymphoedema.

Analogously to VCSS rating criteria, in LCSS we assigned four possible severity levels to each of the ten previously described conditions. For each item the assigned score can ranges from 0 to 3 (Table 1). Particularly, the following score assignation criteria have been adopted (Table 2)

Table 1: Clinical Score According to Less.

LCSS	Severity of Each Item		
0	Absent/Not significant		
1	Mild		
2	Moderate		
3	Severe		

Table 2: Severity Score (Clinical History and Physical Examination) Assignment Grid.

Attribute	None = 0	Mild = 1	Moderate = 2	Severe = 3	
Lymphatic oedema	None	Occasional swelling with spontaneous regression	Permanent oedema	Elephantiasis with loss of normal morphology of the limb	
Tissutal consistency increase	None	Focal	Iinvolvement limited to distal third of limb	Involvement of the knee and proximal parts of the limb.	
Inflammation	None	Limited phlogosis involving less than third of limb	Diffuse phlogosis involving less than a third of a limb.	Diffuse phlogosis involving more than a third of a limb.	
Skin dystrophy	None	Limited to the marginal area	Diffuse involvement limited to less than a third of limb	Diffuse involvement interesting mor than a third of a limb.	
Recurrent Dermato- lymphangio-adenitis	None	History of only one episode	2 history of two episodes	History of more than 2 episodes	
Pain	Absent	Occasional. No restricton to daily activities nor need for drugs.	Moderate restriction of daily activities. Occasionally use of drugs	Severe limitation of daily activities or need for a regular use of drugs	
Lymph ulcerations	None	1 Lymph ulceration	2 Lymph ulcerations	More than 2 active lymph ulcerations	
Articular involvement (most important Joint of the limb)	None	Involving 1 great joint of the limb	Involving 2 great joints of limb	Involving the three great joints of limb	
Muscular Hypotrophy	None	Limited to marginal area	Diffuse, involving less than a third of the limb	Diffuse, involving more than a third of limb	
Compression therapy	None	Intermittent use of garment	Wears garment all the day	Full compliance to the garment	

- a) Oedema.0 = none.1 = occasional swelling with spontaneous regression. 2 = Permanent oedema. 3 = Elephantiasis with loss of normal morphology of the limb.
- b) Tissutal consistency increase. 0 = none. 1 = focal. 2 = involvement limited to distal third of limb. 3 = involvement of the knee and proximal parts of the limb.
- c) Inflammation. 0 = none. 1 = Limited phlogosis involving less than third of limb. 2 = Diffuse phlogosis involving less than a third of a limb. 3 = Diffuse phlogosis involving more than one third of a limb.
- d) Skin dystrophy. 0 = none. 1 = Limited to the marginal area. 2 = Diffuse involvement limited to less than a third of limb. 3 = Diffuse involvement interesting more than one third of a limb.
- e) Recurrent dermato-lymphangio-adenitis. 0 = none. 1 = history of only one episode. 2 = 2 history of two episodes. 3 = history of more than 2 episodes.

- f) Pain. 0 = Absent. 1 = Occasional. No restriction to daily activities nor needs for drugs. 2 = Moderate restriction of daily activities. Occasionally use of drugs. 3 = Severe limitation of daily activities or need for a regular use of drugs.
- g) Lymph ulcerations. 0 = none. 1 = 1 ulcer. 2 = 2 ulcers. 3 = more than 2 active lymph ulcerations.
- h) Articular involvement. 0 = none. 1 = Involving 1 great joint of the limb. 2 = Involving 2 great joints of limb. 3 = Involving the three great joints of limb.
- i) Muscular hypotrophy. 0 = none. 1 = Limited to marginal area. 2 = Diffuse, involving less than a third of the limb. 3 = Diffuse, involving more than one third of limb.
- j) Compression therapy. 0 = none. 1 = Intermittent use of garment. 2 = Wears garment all the day. 3 = Full compliance to the garment.

Patients

In order to assess the reliability of this score and assignation system 185 patients with lymphoedema of the limbs and/or of the external genitalia (81 M and 104 females, age range 0-77 y 72 with primary and 113 with secondary lymphoedema,) have been classified according to LCSS criteria after the collection of an accurate clinical history and a careful physical examination. In each case the findings were properly recorded in the Excell grid. The recommended therapeutic modality regimen springs, for every patient, from his/her LCSS total score and from the consequent attribution to a clinical level (Table 3). An exemple of LCSS attribution is illustrated in (Table 4) that shows how, in each patient, the LCSS corresponds to the sum of the ten scores.

Table 3: Loss and Consequent Therapeutic Indications.

Clinical Level	Total Score	Therapeutic Indications
Not significant	0-5 (Figure 1)	No Treatment
Mild	6-15 (Figure 2)	Consulting room
Moderate	16-22 (Figure 3)	Day Hospital
Severe	23-30 (Figure 4)	Admission as an inpatient

<u>Table 4</u>: Exemple of Card Fullfilling With Patient's Data, With Lcsss And Determination of Total Score.

Patient Name Examination Date		DOB			
Attribute		Scores			
		1	2	3	
Lymphatic oedema			2		
Tissutal consistency increase		1			
Inflammation		1			
Recurrent Dermato-lymphangio-adenitis			2		
Pain		1			
Lymph ulcerations	0				
Articular involvement (with special reference to the joint of the limbs)			2		
Muscular hypotrophy		1			
Compression therapy			2		
TOTAL	0	5	8	0	13

Notice: This subject's LCSS resulted = 13, with indication to a treatment as an outpatient

According to the final sum of score four clinical levels can be identified:

- a) Total score ranging from 0 to 5: no charge by the health insurance systems is justified; n = 19 pts
- b) Total score ranging from 6 to 15: these subjects are properly treated as outpatients; $n=47\,\mathrm{pts}$.
- c) Total score ranging from 16 to 22: these patients may be admitted to Day Hospital regimen; n = 59.
- d) Total score ranging from 23 to 30: these subjects can properly be treated as inpatients n = 74.

The patients assigned to the levels from 1 to 3 underwent a personalized decongestive integrated physical treatment of different

intensity and duration. In all cases a definitive elastic garment was prescribed. The final score was, in each subject, recalculated after the end of the treatment. The difference between the initial and the final total score resulted to be a suitable, numerical quantification of the improvement induced by the treatment. A comparison was also been performed, at enrolling, between LCSS level assignations and the clinical staging determination according to the ISL Consensus Document [10,11] that was also performed on all the 185 patients enrolled in our study (Tables 5 & 6).

<u>Table 5</u>: Staging According to The International Society of Lymphology (ISL).

ISL Clinical Stages	Corresponding Clinical Features		
0	No oedema but significant risk of its clinical appearance		
I	Oedema totally regressing by treatment (pitting oedema)		
II	Oedema with tissue fibrosis, only partially regressing by treatment (no pitting oedema)		
III	Elephantiasis with skin lesions and relapsing infections		

<u>Table 6</u>: Comparison Among ISL-Determined Clinical Stages and The Assigned Loss.

	Stage 0	Stage 1	Stage 2	Stage 3
LCSS Level 0	10	5	4	0
LCSS Level 1	9	21	23	32

Notice: That differences among LCSS levels and ISL stages were highly statistically significant in all cases (p<0.001 at chi square test).

Results

- a) According to the reported criteria the studied population of 185 patients has been classified in the following clinical levels:
- b) Clinical level 0 (total score ranging between 0 and 5): 19 patients (10.3%). These subjects were not assigned to any kind of treatment (Figure 1).



Figure 1: Exemple of a not significant lymphoedema of the left lower limb.

c) Clinical level 1 (total score ranging between 6 and 15): 85 patients (46%). These subjects were treated as outpatients (Table 3) and (Figure 2).



Figure 2: Exemple of patient with moderate lymphoedema of left lower limb.

d) Clinical level 2 (total score ranging between 16 and 22). 47 patients (25.4%). This group has been treated in Day Hospital (Table 3) and (Figure 3).



Figure 3: Exemple of patient with severe lymphoedema of the left lower limb.

e) Clinical level 3 (total score ranging between 23 and 30): 34 patients (18.3). These subjects have been admitted as inpatients in order to receive an integrated, intensive, decongestive physical treatment (Table 3) and (Figure 4).



Figure 4: Exemple of a patient with highly disabilitating lymphoedema.

At the end of the treatment the following total scores and changes were recorded:

- a. Patients assigned to level 0 obviously did not showed, any change.
- b. Patients assigned to level 1 showed in the post-treatment total score a mean decrease of 37% compared to the baseline.
- c. Patients assigned to level 2 showed a final total score mean reduction of 42.5% compared to baseline.
- d. Patients assigned to level 3 showed a final total score mean reduction of 38.4% compared to baseline.

Regarding the comparison between the LCSS level assignation and the ISL clinical staging no full concordance has been observed (p < 0.001). In particular we have founded:

- a. The ISL stage 0 corresponds to 0-2 levels of LCSS.
- b. The ISL stage I corresponds to 0-2 levels of LCSS.
- c. The ISL stage II corresponds to 0-3 levels of LCSS.
- d. The ISL stage III corresponds to 1-3 levels of LCSS and, vice versa:
- e. The 0 level of LCSS corresponds to 0-II ISL stages.
- f. The 1 level of LCSS corresponds to 0-III ISL stages.
- g. The 2 level of LCSS corresponds to 0-III ISL stages.
- h. The 3 level of LCSS corresponds to II-III ISL stages.

Discussion and Conclusion

LCSS represents a simple and reliable method of assignment of a patient to a determinate severity level and consequently to a defined assistance modality. Regarding the comparison between the LCSS level assignation and the ISL clinical staging it must be considered that LCSS does not intend to replace the ISL classification because this latter gives descriptive classes whilst LCSS refers to severity-parameters and disability degree (score) assessment. Therefore, LCSS will hopefully result a useful criterion to guide the patients' assignment choices of both private and public insurance health systems. Moreover, the comparison of the baseline and of the post-therapy LCSS scores will provide a useful way to assess the effectiveness of the adopted treatments. It is noteworthy that in the clinical history of the disease the LCSS may improve or impair. The modality of treatment must be chosen consequently to the updated clinical level. Obviously, there are irreversible scores, like the number of the previously suffered lymphangites, that will not decrease in the follow-up, even though therapeutic treatment.

Otherwise, it is important to underline that the score assigned to this finding reflects an irreversible anatomic-functional damage and, consequently weighs as partial component of the total score in an unaffected way in all subsequent clinical controls. The proposed method for the clinical assessment of peripheral lymphatic disease is inexpensive, reliable, safe and easy to perform. Analogously to VCSS, that successfully entered in the clinical practice long time ago, the LCSS is the expression of the severity of the sickness and of the clinical burden. Moreover, it is a simple key to the standardization

of the treatments and is a very useful tool in the assessment of the economic managing of lymphatic peripheral disease. It also represents a way to avoid claims about the therapeutic regimen chosen. The authors are therefore trustful that LCSS might gain a general acceptance by all people involved in lymphatic peripheral disease treatment.

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