EVIDENCE BASED STATEMENT

DOMAIN 10, Statement 2

TOPIC: "lymphedema classification & staging"

SEARCH TERMS & SOURCES

(lymphedema) AND (classification) OR (staging)

INCLUSION CRITERIA

- Lower limb only
- Reviews
- Publication < 10 years, only ENG

SEARCH RESULT BEFORE - AFTER SELECTION

32 (before) - 9 (after selection)

AGREEMENT BETWEEN THE 2 REVIEWERS before DOMAIN WORKING GROUP DISCUSSION & FINALIZATION

(N. of papers triggering disagreement in inclusion/No of papers from the initial search

1/32

PERTINENT LITERATURE NOT IDENTIFIED BY THE LITERATURE SEARCH

1. The diagnosis and treatment of peripheral lymphedema: 2020 Consensus Document of the ISL. Lymphology. 2020;53(1):3-19.

2. Douglass J, Kelly-Hope L. Comparison of Staging Systems to Assess Lymphedema Caused by Cancer Therapies, Lymphatic Filariasis, and Podoconiosis. Lymphat Res Biol. 2019;17(5):550-556.

4. Gianesini S, et al. Global guidelines trends and controversies in lower limb venous and lymphatic disease: Narrative literature revision and experts' opinions following the vWINter international meeting in Phlebology, Lymphology & Aesthetics, 23-25 January 2019. Phlebology. 2019 Sep;34(1 Suppl):4-66.

3. Greene AK, Goss JA. Diagnosis and Staging of Lymphedema. Semin Plast Surg. 2018;32(1):12-16.

EVIDENCE BASED STATEMENT

Domain 10; Statement 2

IDENTIFIED REFERENCES

(from the most recent down)

1. Lee E, Biko DM, Sherk W, et al. Understanding Lymphatic Anatomy and Abnormalities at Imaging. Radiographics. 2022;42(2):487-505
2. Cirocchi R, Amabile MI, De Luca A, et al. New classifications of axillary lymph nodes and their anatomical-clinical correlations in breast surgery. World J Surg Oncol. 2021 Mar 29;19(1):93.
3. Gordon K, Varney R, Keeley V, et al. Update and audit of the St George's classification algorithm of primary lymphatic anomalies: a clinical and molecular approach to diagnosis. J Med Genet. 2020 Oct;57(10):653-659.
*4. The diagnosis and treatment of peripheral lymphedema: 2020 Consensus Document of the ISL. Lymphology. 2020;53(1):3-19.
5. Douglass J, Kelly-Hope L. Comparison of Staging Systems to Assess Lymphedema Caused by Cancer Therapies, Lymphatic Filariasis, and Podoconiosis. Lymphat Res Biol. 2019;17(5):550-556.
6. Greene AK, Goss JA. Diagnosis and Staging of Lymphedema. Semin Plast Surg. 2018;32(1):12-16
7. Zeltzer AA, Anzarut A, Hamdi M. A Review of Lymphedema for the Hand and Upper-Extremity Surgeon. J Hand Surg Am. 2018;43(11):1016-1025.
8. Connell FC, Gordon K, Brice G, et al. The classification and diagnostic algorithm for primary lymphatic dysplasia: an update from 2010 to include molecular findings. Clin Genet. 2013;84(4):303-14 .
9. Lee BB, et al.International Union of Angiology. Consensus Document of the International Union of Angiology (IUA)-2013. Current concept on the management of arterio-venous management. Int Angiol. 2013 Feb;32(1):9-36.

EVIDENCE BASED STATEMENT

Domain 10; Statement 2

TEXT FOR INCLUSION IN THE DOCUMENT
(300 words, not counting the references)
DOMAIN 10, Statement 2, TOPIC: "lymphedema classification & staging"

Different classification and staging systems are available for lymphedema, but there is not a globally recognized single method providing a comprehensive description of the condition.
[**Douglass J, Kelly-Hope L. Comparison of Staging Systems to Assess Lymphedema Caused by Cancer Therapies, Lymphatic Filariasis, and Podoconiosis. Lymphat Res Biol. 2019;17(5):550-556**]
[**Gianesini S, et al. Global guidelines trends and controversies in lower limb venous and lymphatic disease: Narrative literature revision and experts' opinions following the vWINter international meeting in Phlebology, Lymphology & Aesthetics, 23-25 January 2019. Phlebology. 2019 Sep;34(1 Suppl):4-66**].
Moreover, the used terminology can be confounding as not universally adopted.
[**Greene AK, Goss JA. Diagnosis and Staging of Lymphedema. Semin Plast Surg. 2018;32(1):12-16**]
The International Society of Lymphology (ISL) recognizes a 4 stages system:
0) LATENT lymphedema.
Swelling is not yet evident despite initial alterations in drainage and related symptoms. It can be transitory and persists years before it manifests itself clinically.
1) REVERSIBLE EDEMA
Accumulation of fluid of high protein content reverted by limb elevation. The edema becomes pitting.
2) Not-REVERSIBLE EDEMA
Involution toward fibrotic tissue with edema that is not usually resolved by limb elevation. Pitting is evident but can disappear in case of advanced fibrotic involution.
3) ELEFANTIASIS
Massive lymphatic compromission and fibrosis, associated with skin changes such as acanthosis.
A single person can present multiple stages at the same time, based on the different involvement of the lower limb parts.
As reported by the same ISL, this staging refers just to the physical appearance of the disease, not contemplating the pathogenetic mechanism, the genetic vs environmental compromission grade.
An attempt quantify the severity has been made also by proposing to use the limb volume excess compared to the contralateral limb:
MILD (>5%, < 20%)
MODERATE (>20%, <40%)
SEVERE (>40%)
There is not real agreement on these values, as other groups consider "minimal" a variation in between 5% and 10%, while in between 10% and 20% is considered "mild".
*[**The diagnosis and treatment of peripheral lymphedema: 2020 Consensus Document of the ISL. Lymphology. 2020;53(1):3-19**]
This staging system is biased by multiple factors influencing the same limb volume (vascular and musculoskeletal) and by the contralateral limb possible lymphatic compromission.
A classification advancement should take into consideration also possible complications (skin infections or ulceration) and quantify the impact of the disease on the patient quality of life.

EVIDENCE BASED STATEMENT

Domain 10; Statement 2

STATEMENT FOR PUBLIC EVIDENCE-BASED AWARENESS

DOMAIN 10, Statement 2

"**Lymphedema is a progressive disease whose stage must be precisely identified by an expert**"

4 SELECTED REFEREENCES

1. Douglass J, Kelly-Hope L. Comparison of Staging Systems to Assess Lymphedema Caused by Cancer Therapies, Lymphatic Filariasis, and Podoconiosis. Lymphat Res Biol. 2019;17(5):550-556
2. Gianesini S, et al. Global guidelines trends and controversies in lower limb venous and lymphatic disease: Narrative literature revision and experts' opinions following the vWINter international meeting in Phlebology, Lymphology & Aesthetics, 23-25 January 2019. Phlebology. 2019 Sep;34(1 Suppl):4-66.
3. Greene AK, Goss JA. Diagnosis and Staging of Lymphedema. Semin Plast Surg. 2018;32(1):12-16.
4. The diagnosis and treatment of peripheral lymphedema: 2020 Consensus Document of the ISL. Lymphology. 2020;53(1):3-19

identified LITERATURE BIAS

Lack of staging system intra and interobserver validation studies

SUGGESTED NEXT LINES OF RESEARCH

Validation of a homogeneous globally recognized lymphedema classification system, including a severity scale and quality of life measures