A review of the heterogeneity of clinical lymphoedema scoring systems

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Key words

Lymphoedema, clinical scoring system, treatment efficacy, assessment

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Abstract

Background: Standardised staging of lymphoedema is crucial in accurately assessing treatment efficacy. This becomes challenging due to distinct scales for different locations, such as the upper extremity, lower extremity, and mediastinal areas, with heterogeneous modalities for measurement. Biometric measurements, such as limb circumference, tissue composition, and lymphoscintigraphy, can provide objective information about the severity and progression. **Aim:** To address challenges associated with standardised staging of lymphoedema by examining the heterogeneity scoring scales. Methods: MEDLINE, CINAHL, Embase, and Google Scholar were searched for lymphoedema scoring scales. **Results:** 690 studies were found in the initial search, with 86 included for full-text review. 31 were identified for data extraction. Of these 31 articles, 33 clinical scoring systems were identified: 10 for lower extremity lymphoedema, 6 for upper extremity, 2 for both upper and lower extremity, 9 for head and neck, and six general, non-specific scales. Common parameters included limb volume, skin changes, functional impairment, and pain. **Conclusion:** There are many clinical scoring systems for lymphoedema assessment; these systems reflect the condition's complexity, with varied focuses from physical measurements to psychological impacts. The plethora of systems available complicates consistent assessments, study comparisons, and uniform patient care, presenting a significant challenge to standardisation. Gaps in holistic assessment were noted, with limited systems addressing psychological well-being despite its significance in the condition's overall impact. A unified approach is necessary. Integrating patient feedback into this standardisation would ensure a comprehensive review addressing clinical and quality-of-life aspects.

The lymphatic system's primary function is to drain excess fluid from the tissues and return it to the bloodstream; when lymphatic drainage is impaired, fluid accumulates, causing the characteristic swelling of lymphoedema, a chronic condition most commonly affecting the arms and legs (Grada and Phillips, 2017; Greene and Goss, 2018; Keast et al, 2019; Azhar et al, 2020). Lymphoedema may be a result of congenital abnormalities, trauma, or infection, but more commonly, it is a post-surgical side-effect, especially after treatments for certain types of cancers (Unno et al, 2010; Azhar et al, 2020).

Lymphoedema is estimated to affect 90 million–250 million people globally, although this number is likely an underestimation due to variability in diagnostic criteria and missed clinical recognition (Rockson and Rivera 2008; Greene 2015; Keast et al, 2019; Torgbenuet al, 2020). Primary lymphoedema is rare, with 1 in 100,000 individuals affected. Secondary lymphoedema is more common, affecting approximately 1 in 1,000 Americans (Rockson and Rivera 2008; Greene 2015; Keast et al, 2019; Torgbenuet al, 2020).

In fact, 99% of lymphoedema is secondary (or acquired) lymphoedema, which is associated with higher morbidity, likely due to impaired compensation and comorbid conditions. In low- and middle-income countries, parasitic filariasis infection is the most common cause of lymphadenectomy. Lymph node radiation secondary to oncological surgery is the most common cause in high-income countries (Douglass and Kelly-Hope, 2019).

Lymphoedema is a significant cause of medical comorbidity, including chronic pain, functional impairment, recurrent infections, psychological distress and poor self-perception of body image (Greene 2015). Various clinical scoring systems have been developed to evaluate the severity and progression of lymphoedema (Greene and Goss, 2018). These scoring systems offer a structured approach to assess the extent of swelling, skin changes, functional impairment, and other clinical manifestations of the disease. Scoring systems can guide treatment decisions, monitor therapeutic outcomes, and facilitate standardised communication among healthcare professionals (Dambha-Miller et al, 2020).

However, a notable challenge in lymphoedema assessment is the heterogeneity of these scoring systems. Different scales prioritise various

Table 1. Inclusion and exclusion criteria	
Inclusion criteria	**Exclusion criteria**
All studies with clinical scoring systems for lymphoedema (regardless of type or location)	Studies with no lymphoedema clinical scoring system or classification
Both validated and unvalidated scoring systems	Parasitic lymph filariasis
Cohort studies, cross-sectional studies, randomised and non-randomised control trials, qualitative studies, literature reviews	Abstracts without a peer-reviewed manuscript, editorials, commentaries
Adult patients (age > 18 years)	Article not in English

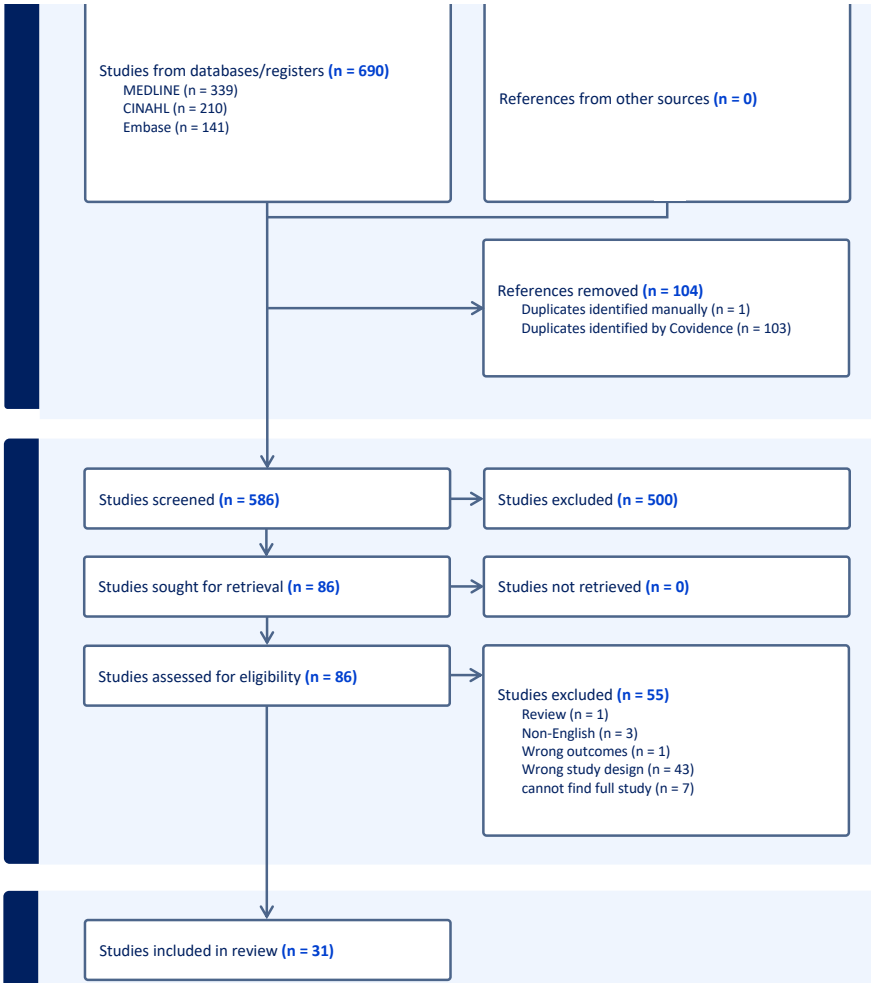

Figure 1. Preferred Reporting Items for Systematic reviews and Meta-analyses (PRISMA) flow chart displaying the screening process for included and excluded studies.

parameters, such as limb volume, skin thickness or functional outcomes. This variety means that there is no universal gold standard for assessing lymphoedema. As a result, the choice of a scoring system often depends on the clinical setting, the objectives of the assessment, and the preference of the healthcare professional.

To our knowledge, there is currently no published study that compares the various lymphoedema scoring systems. As such, we set out to delineate the diversity in the available clinical scoring systems and highlight the opportunities and challenges of such heterogeneity. As a secondary objective, we sought to review areas where unity can be achieved to allow for more actionable assessments.

Methods

The methods of the study were based on the Preferred Reporting Items for Systematic reviews and Meta-Analyses extension for Scoping Reviews guidelines (Tricco et al, 2018). The inclusion and exclusion criteria are shown in Table 1.

A medical subject librarian was consulted in the development of our search strategy, and searches included combinations of the following index terms: lymphoedema, clinical scoring system, assessment tool, the severity of illness index, and severity classification. Searches were conducted in Medline, Embase, CINAHL, and Google Scholar to obtain all relevant articles as of 1 June 2024, with no restrictions on publication dates.

Two independent reviewers (SS and KS) screened articles using Covidence, and discrepancies were resolved through a consensus discussion. Studies that met inclusion criteria were further assessed with a full-text screen. All articles that could not be screened for eligibility based on title and abstract were moved to the full-text screening stage. At the full-text screening stage, each excluded study was assigned a specific reason for exclusion. Reference lists of the included articles were reviewed for additional studies to screen. A spreadsheet was used to record set parameters from each scoring system by two independent reviewers (SS and KS) with conflicts resolved through consensus discussion.

Results

The literature search and screening process is presented as a PRISMA flow diagram [Figure 1]. The combined database searches yielded 690 records. After removing duplicates, 586 records underwent title and abstract screening; of these, 86 articles were reviewed in the full-text screening stage, 55 of which were excluded.

In the 31 included studies, 33 clinical scoring systems were described. Six described only upper-extremity lymphoedema [Table 2], 10 described only lower-extremity [Table 3], two described

both the upper and lower extremities [Table 4], six were non-specific or general scoring systems [Table 5], and nine were for head and neck [Table 6].

The most cited parameters in the scoring systems included limb volume (23 studies), skin changes (20 studies), functional impairment (19 studies), and pain (15 studies).

Historically, the gold standard for subjectively grading upper extremity lymphoedema (UEL) has been the International Society of Lymphoedema (ISL) grading system (Yamamoto et al, 2013; Wiser et al, 2020). This symptom-based scale has broad categories ranging from subclinical lymphoedema to lymphostatic elephantiasis (Wiser et al, 2020). This is a convenient way to stage patients on presentation but requires only an overall gestalt of the patient's presentation.

More objective measurements supplemented this system, including the volume or limb circumference difference between two limbs (Yamamoto et al, 2013; Kim et al, 2020). Although these measurements were convenient and more accurate than the ISL staging, they have the limitations of bilateral lymphoedema being more challenging to assess, and they are difficult to compare across individuals with different heights and BMIs.

Several alternative scales for UEL that rely on quantitative measurements have been suggested. The most notable quantitative scale is the UEL index suggested by Yamamoto et al (2013) which takes the circumference of five locations along the upper extremity and corrects for the patient's BMI. This scale has the notable benefit of being comparable across individuals despite differences in BMI. It may be a prudent scale to assess the efficacy of lymphoedema therapies in trials. It is also a technique that is easily accessible and adopted by providers. However, it is more time-consuming than the ISL grading or volume/circumference measurements, which limits its adoption in routine follow-up visits.

Lymphoscintigraphy and indocyanine green (ICG) have also been suggested for surgical planning for lymphoedema (Yamamoto et al, 2011; Yoon et al, 2020). Both appear to provide comparable ability to assess for the functional characteristics of the lymphoedematous limb but are significantly more specialised and less accessible to general practitioners and are not common in primary or urgent care settings.

The LEL index is analogous to the UEL scale. Both were proposed by Yamamoto et al and, consequently, have very similar benefits and drawbacks. Specifically,

Table 2. Upper extremity lymphoedema clinical scoring systems.

Scale	Staging criteria						Validated?
LENT/SOMA Lymphedema-Related Items (Cheville et al, 2003)	Grade 1 2–4 cm	Grade 2 4–6 cm	Grade 3 >6 cm	Grade 4 Useless arm			No
MRI Staging UEL 3 levels (forearm, elbow, upper arm; Kim et al, 2020)	Stage 0 No detectable fluid infiltration at any level	1 Circumferential fluid infiltration does not exceed 50% at any level	2 Circumferential fluid infiltration may exceed 50% at any level	3 Circumferential fluid infiltration exceeds 75% at all three levels			Yes (against ISL)
CLUE (Cancer- related Lymphedema of the Upper Extremity) Spinelli et al, 2019	Obscuration of anatomical architecture	Deviation from normal anatomical contour	Tissue texture	Oedema			Yes
Arm Dermal Backflow Stage (Yamamoto et al, 2011)	0 No dermal back flow pattern	1 Splash pattern around the axilla	2 Stardust pattern limited between the axilla and the olecranon	3 Stardust pattern exceeding the olecranon	4 Stardust pattern observed throughout the limb	5 Diffuse pattern and stardust pattern observed throughout the limb	Yes
Upper Extremity Lymphedema Index (Yamamoto et al, 2013)	Stage 1 < 130	2 130-150	3 150-170	4 >170			Yes
Lymphoscintigraphy severity scale (Yoon et al, 2020)	Stage 1 Mild lymphatic obstruction and collateral vessels without DBF signs.	Stage 2 Mild lymphatic obstruction with DBF signs appearing in the upper arm.	Stage 3 Significant lymphatic obstruction with DBF signs in the upper arm and forearm.	Stage 4 Lymphatic flow from the hand to LN around the clavicle is almost absent, DBF signs only in the forearm.	Stage 5 Lymphatic flow from the hand to the lymph nodes around the clavicle is absent. Signs of DBF are present only in the hand.		Yes

DBF = dermal back flow; ISL = International Society of Lymphoedema; LENT/SOMA: Late Effects in Normal Tissues – Subjective, Objective, Management, Analytic ; LN = lymph node

Table 3. Lower-extremity lymphoedema clinical scoring systems.

Scale	Staging criteria					Validated?
Leg Lymphedema Complexity Score (Bjork and Hettrick, 2020) – 11 domains	Comorbidities Skin integrity Mobility	Limb oedema Skin changes Pain/discomfort (2 weeks)	Tissue texture Fat disorders (lipoedema)	Lymphedema Life Impact Scale	Scars BMI	No
Calf oedema area by MRI (Wang et al, 2018)	Stage 0 TA 7,779.61 MA 5,423.64 WA 0.79	1 TA 7,387 MA 4,635 WA 320.96	2 TA 9,023.64 MA 4,620.49 WA 1,801.85	3 TA 14,020.09 MA 5,168.19 WA 6,564.56		No
American Physical Therapy Association Lymphedema Criteria (Cheville et al, 2003)	Mild < 3 cm interlimb circumference discrepancy	Moderate 3–5 cm	Severe >5 cm			No
Cheng lymphedema grading system (Cheville et al, 2003)	Grade 0 <9% circumference discrepancy	I 10–19%	II 20–29%	III 30–39%	IV >40%	No
MRI Volumes LEL (Lu et al, 2014)	Stage 0 VD 21 TTD 0.3 MTD 0.1 STTD 0.4	1 VD 208 TTD 8.1 MTD -0.2 STTD 8.4	2 VD 696 TTD 21.3 MTD 2.9 SSTD 18.3	3 VD 1597 TTD 38.4 MTD -1.0 SSTD 38.8		Yes
Ultrasound characteristics of LEL (Omura et al, 2021)	Staging determined by cutoff values in the quantitative ultrasonic characteristics of the skin layers (dermis and hypodermis)		Echogenic regions will decrease in thickness as the ISL stage progresses		The EAC in the dermis will overall decrease	No
Dermal backflow LEL (Shinaoka et al, 2022)	Stage 0 (Mildest) No defect	1 (Mild) PM or PL defect	2 (Severe) PM and PL defect	3 (Most severe) All defects		No
LEL Index (Yamamoto et al, 2011)	Stage 1: <250	Stage 2: 250–300	Stage 3: 300–350	Stage 4: >350		Yes
GDB stage based on ICG lymphography (Yamamoto et al, 2016)	Stage 0 No dermal backflow	Stage 1 Splash pattern around groin	Stage 2 Stardust pattern in groin/lower abdominal region	Stage 3 Stardust pattern extended to the genital region	Stage 4 Diffuse pattern with stardust pattern in the background	
LEC score (Yamamoto et al, 2013)	Stage 1 Score < 3.7		Stage 2 Score > 3.7			

BMI = body mass index; EAC = echo free area; GDB = grading dermal backflow; LEC = latency, edema, compression; LEL = lower extremity lymphoedema; MA = muscle area; MT = muscle thickness; PL = posterior lateral; PM = posteromedial; STTD = subcutaneous tissue thickness; TA = soft tissue area; TD = total thickness soft tissue; VD = volume difference; WA = water area.

both have increased robustness, depth of information, and are easily accessible. Consequently, both are time intensive to complete compared to ISL/limb circumference.

Interestingly, there was a greater variety of scales identified for LEL, such as calf oedema area/volume by MRI, qualitative features on ultrasound, lymphoscintography/ICG backflow measurements, the LEL index based on limb circumference correcting for BMI, and the Latency-Edema-Compression (LEC) score based on clinical factors (Cheville et al, 2003; Yamamoto et al, 2011, 2013; Lu et al, 2014; Yamamoto 2016; Wang et al, 2018; Bjork et al, 2020

Omura et al, 2022; Shinaoka et al, 2022).

It seems beneficial to have this robustness of data for assessing lymphoedema in clinical trials, as mentioned for the UEL index. Along with the proposed LEL index, there is an LEC score, which uses functional parameters such as latency period (time to develop lymphoedema), duration of oedema, period of compression therapy and number of cellulitis episodes per year (Yamamoto et al, 2013). The LEC score stratifies patients into more of a binary classification based on only clinical factors.

More specialised imaging methods have also been proposed for LEL. MRI results appear promising for measuring tissue areas/volumes in different ISL stages of lymphoedema, providing a quantitative supplement for categorising ISL stages (Lu et al, 2014). It remains an open question if MRI has utility in further stratifying patient populations with lymphoedema beyond the our ISL stages and if there is any clinical utility or predictive power to MRI measurement.

The ISL and GDB Stages based on ICG Lymphography addressed both UEL and LEL (Wang et al, 2018; Garza et al, 2019). These systems aim to provide a comprehensive understanding of the patient's lymphoedema status. Though

Table 4. Clinical scoring systems for upper and lower limb lymphoedema.					
Scale	**Staging criteria**				**Validated?**
International Society of Lymphology (Wang et al, 2019)	Stage 0 Latent or sub-clinical condition Swelling is not present apparent	Stage I Early accumulation of fluid which subsides with limb elevation. Pitting may occur	Stage II Limb elevation alone rarely reduces tissue swelling. Pitting is manifest	Stage III Pitting is absent Trophic skin changes are present.	Yes
GDB Stage based on ICG Lymphography (Yamamoto et al, 2016)	Stage 1 Many patent vessels Minimal DBF	Stage 2 Moderate patent vessels Segmental DBF	Stage 3 Few patent lymph vessels Extensive DBF	Stage 4 No patent lymph vessels Severe DBF	Yes

incorporating both extremities allows for a complete picture, it can potentially lead to an increase in the complexity of the scoring method, with decreased accuracy given the decreased specificity of the score when removing the region of lymphoedema as a consideration.

The same can be said for non-specific scoring systems, six of which were identified. The Common Toxicity Criteria (CTC) lymphoedema criteria and ISL scales are the most commonly used in practice (Cheville et al, 2003). The CTC takes multiple factors into consideration, including patient-reported symptoms and clinical features, including dermal changes, regions where lymphoedema is present, inter-limb discrepancies, obscuration of the genitals, lymph-related fibrosis, and phlemolymphatic cording (Cheville et al, 2003). While comprehensive, this score is not easily accessible or understood by those without previous experience in the field.

Scoring systems such as the British Lymphology Society Staging System group those affected into four categories based on risk factors: regional involvement, presence of malignancy, and limb volume (Honnor, 2006).

Other specialised imaging modalities, such as elastography, have been described. However, they have not been validated as stand-alone scoring tools or integrated into any pre-existing lymphoedema scoring system. Bioimpedance spectroscopy has also been described as a rating tool but is binary, non-specific, and not commonly used in practice (Ridner et al, 2018).

Several head and neck lymphoedema (HNL) scoring systems have also been described. One notable system is the Head and Neck External Lymphoedema and Fibrosis Assessment Criteria, which categorises scores by clinical signs, subjective symptoms, and functional impairment (Deng et al, 2015). The Secondary Quadrant Upper Lymphoedema criteria, guided by ISL guidelines, has multiple objective measures, including bioimpedance analysis, circumferential measurement, water displacement, perimetry and imaging (Levenhagen et al, 2017). The MD Anderson Cancer Center HNL rating scale simplifies categorisation into three levels based on visual assessments of lymphoedema and the presence or absence of pitting, similar to the Common Terminology Criteria for Adverse Events and Compression Class =scores (Deng et al, 2011).

Other scoring systems, not yet validated, include the ALOHA scale, which uses two unique metrics, MoistureMeter D and neck tape measuring systems (Nixon et al, 2014; Purcell et al, 2016). Using endoscopy, the Modified Patterson scale looks specifically at laryngeal and pharyngeal oedema in head and neck cancer patients (Starmer et al, 2021). This more subjective assessment depends on the user's comfort and skill level with endoscopy.

Discussion

The purpose of this study was to explore the range of clinical scoring systems for the evaluation of lymphoedema to identify areas where standardisation and unification could be achieved.

We identified 33 clinical scoring systems, targeting different regional areas affected by lymphoedema and focusing on varied parameters. While certain parameters, like limb volume, were universally recognised and incorporated, others, such as psychological distress and self-perception of body image, were only integrated in a subset of systems.

Further, classification systems fell into two predominant categories, scoring using a binary approach (present versus absent) versus grading systems with respective clinical signs with each grade. Regarding usability and clinical applicability, scoring systems with fewer parameters were reported to be more user-friendly and time-efficient in busy clinical settings. However, they might compromise on the granularity and comprehensiveness of the assessment. Conversely, while offering a thorough assessment, more detailed systems might be too cumbersome for routine clinical evaluations.

The diversity of clinical presentations of lymphoedema is represented in the heterogeneity of its scoring systems. As demonstrated in this study, a broad range of systems are currently in use and the challenge of selecting the system(s) that best align the objective and patient population falls on the clinician or researcher. While beneficial in capturing the nuanced presentations of lymphoedema, the diversity poses challenges for standardising assessments, comparing results across studies, and ensuring consistent patient care across different settings.

The heterogeneity in scoring systems, beyond reflecting the complexity of the disease, also underscores gaps in the collective understanding and approach to lymphoedema. While some systems are comprehensive in their assessment, capturing the condition's physical and psychological facets, others focus narrowly on specific clinical signs or symptomatology. This variation might lead to disparities in diagnosis, treatment, and long-term patient care. For example, only 12 of the 33 scoring systems incorporated an assessment of the patient's psychological well-being despite it being a significant comorbidity of lymphoedema.

This reveals a potential gap in the holistic assessment of patients with lymphoedema and highlights the need

Table 5. General lymphoedema clinical scoring systems.				
CTC v.3.0 Lymphedema Criteria (Cheville et al, 2003)				
Validated?	Yes			
Grade	1	2	3	4
Chyle/lymph leakage	Asymptomatic	Symptomatic	Symptomatic, interventional radiology or operative intervention indicated	Life-threatening complications
Dermal change	Trace thickening or faint discolouration	Marked discolouration or leathery skin texture on papillary formation	–	–
Head and neck	Localised to dependent areas, no disability	Localised facial or neck oedema with functional impairment	Generalised facial or neck oedema with functional impairment	Severe ulceration or cerebral oedema (tracheotomy indicated)
Limb	5-10% inter-limb discrepancy in volume or circumference at greatest point	>10-30% inter-limb discrepancy, obliteration of skin folds	>30% inter-limb discrepancy, lymphorrhoea, interference with ADL	Progression to malignancy, amputation indicated; disabling
Genital	Swelling or obscuration of anatomic architecture on close inspection	Readily apparent obscuration of anatomic architecture	Lymphorrhoea, interfering with ADL	Progression to malignancy
Viscera	Asymptomatic	Symptomatic; medical intervention indicated	Symptomatic and unable to aliment adequately or orally	Life-threatening consequences
Lymph-related fibrosis	Minimal-moderate redundant soft tissue unresponsive to elevation or compression	Marked increase in density and firmness without tethering	Very marked density and firmness with tethering affected ≥40% of oedematous area	–
Lymphocele	Asymptomatic	Symptomatic, medical intervention indicated	Symptomatic and interventional radiology or operative intervention indicated	–
Lymphatics (other)	Mild	Moderate	Severe	Life-threatening
Elastography (Erdogan Iyigun et al, 2019)				
Validated?	No			
Type	A	B	C	D
	No visible tissue swelling; palpable thickening and/or tightness of dermis Grade: Mild -visible soft tissue swelling on close inspection; Moderate - easily visible swelling that significantly alters normal tissue contours; Severe - extreme or massive tissue swelling	Visible soft tissue swelling; Involved tissues are soft to touch; Tissue swelling is reducible and fluctuates in severity Grade: Mild -visible soft tissue swelling on close inspection; Moderate - easily visible swelling that significantly alters normal tissue contours; Severe - extreme or massive tissue swelling	Visible soft tissue swelling; Involved tissues are firm to touch; Tissue swelling is non-reducible and persistent Grade: Mild, moderate, severe	Firm skin with increased density and decreased compliance in the absence of swelling Grade: Mild, moderate, severe

for a more comprehensive approach that considers both the physical and emotional ramifications of the condition.

Conclusion

Addressing the heterogeneity in lymphoedema scoring systems requires a two-pronged approach.

Firstly, there is a need for an evidence-based consensus among experts in the field. Collaborative efforts to synthesise the strengths of existing systems and address their gaps can pave the way for a more unified, comprehensive scoring method. This not only aids in standardising clinical assessments, but also ensures that research findings across different studies are comparable.

Secondly, the integration of patient feedback in refining these systems is crucial. Since lymphoedema impacts patients' lives on multiple fronts, patients offer invaluable perspectives on what dimensions of the disease are most pertinent to their quality

Table 5. (cont.)

International Society of Lymphology Staging System (Holcomb, 2006; Honnor, 2006; Wang et al, 2019; International Society of Lymphology, 2020, 2023)			
Validated?	Yes		
Stage 0	Stage I	Stage II	Stage III
Subclinical condition where swelling is not evident despite impaired lymph transport. May exist for months or years before oedema occurs.	Pitting may occur and is reversible. It may take up to a few hours of rest and elevation to reverse	Pitting occurs, and the hedeoma is not appreciably reduced with elevation of the affected limb. In late Stage II, the issue hardens and becomes fibrotic and pitting no longer occurs.	This stage is also referred to as elephantiasis. Pitting is absent. Skin changes, such as acanthosis, fat deposits, and warty overgrowths, may develop. Fluid may ooze from the skin due to high pressure in the lymphatic and venous vessels. It most commonly occurs in the legs and results from long-standing inadequately treated or untreated lymphoedema
British Lymphology Society Staging System (Honnor, 2006)			
1: People at risk	2: People with mild and uncomplicated oedema	3: People with moderate to severe or complicated oedema	4: People with oedema and advanced disease
People at risk are those with no clinical signs of swelling but with one or more of the following risk factors, know to be implicated in the development of chronic oedema: - Hereditary predisposition - Malignancy ± radiation or surgery - Chronic venous insufficiency - Filariasis - Trauma to lymph nodes and/or vessels - Chronic skin disorders	Oedema with excess limb volume <20% People have uncomplicated oedema if - It does not involve the trunk, head, genitals, digits - The limb is a normal shape - The subcutaneous tissue is predominantly soft and pitting - The skin on the affected part is healthy and intact - There is no arterial insufficiency - There is no known malignancy in the truncal quadrant affected by swelling	Excess limb volume >20% People have complicated oedema if they have any of the following: - Oedema of the trunk, head, genitals, or digits - The subcutaneous tissue is predominantly non-pitting and fibrotic - The limb shape is distorted - The skin on the affected parts is abnormal - There is active controlled malignancy in the truncal quadrant affected by the swelling - There is evidence of venous occlusion / arterial insufficiency or current acute cellulitis, all of which require a medical assessment - There is lymphorrhoea	People with advanced malignancy who have uncontrolled metastatic disease that will shorten their lives Oedema may be due to obstruction by the tumour or due to dependency or immobility. These may be compounded by hypoproteinaemia, renal and/or cardiac failure, and debility which may require assessment and treatment Common symptoms include: - Weeping and ulceration of the affected limb - Tension in the affected tissues - Impaired mobility - Impaired function - Impaired sensation - Heaviness of the affected part - Pain - Infection - Oedema affecting the trunk, genitals or face
Campisi's Lymphedema Clinical Staging System (Yamamoto et al, 2011a, 2011b, 2013)			
Validated?	Yes		
Stage 1	Stage 2	Stage 3	Stage 4
1A: No oedema with presence of lymphatic dysfunction 1B: Mild oedema, reversible with declivous position and night rest	Persistent oedema that regresses only partially with declivous position and night rest	Persistent oedema that continually becomes more severe (recurrent acute erysipeloid lymphangitis)	Fibrotic lymphoedema (with initial lymphostatic warts) and column-shaped limb Stage 5 Elephantiasis with severe limb deformation, scleroindurative pachydermatis, and widespread lymphostatic warts
Bioimpedence Spectroscopy (Ridner et al, 2018)			
Validated?	No		
Stage	L-Dex ≤7 No lymphoedema	L-Dex ≥7 Indicative of lymphoedema	

Table 6. Head and neck lymphoedema clinical scoring systems.

HN-ELAF (Deng at al, 2013)						
Validated?	Ongoing					
Grade	0	1	2	3	4	5
Signs	No visible oedema	No visible oedema	Visible tissue oedema, soft to touch; pitting or non-pitting, elevation may reduce oedema. Oedema comes and goes	Visible oedema, firm to touch; elevation does not reduce oedema. Oedema persistent	No visible oedema, mild-moderate firm tissues. A. History of oedema in areas of fibrosis B. No history of oedema in areas of fibrosis	No visible oedema, markedly firm tissues. A. History of oedema in areas of fibrosis B. No history of oedema in areas of fibrosis
Symptoms	None	Feeling uncomfortable in face/neck	Feeling uncomfortable/ pressure/tightness in the face/ neck	Feeling uncomfortable/ pressure/tightness in the face/ neck. Other sensation-related symptoms (e.g, hardness)	Feeling uncomfortable/ pressure/tightness in the face/ neck. Other sensation-related symptoms (e.g. hardness); pain/discomfort which may be worse on movement of affected area(s)	Feeling uncomfortable/ pressure/ tightness in the face/neck. Other sensation-related symptoms (e.g. hardness); pain/discomfort when moving or not moving affected area(s)
Functional impairment	None	None	Functional impairment dependent on affected areas	Functional impairment dependent on affected areas	Functional impairment dependent on affected areas	Functional impairment dependent on affected areas
MD Anderson (Nixon et al, 2014; Purcell et al, 2016)						
Validated?	Yes					
Level	0		1a	1b	2	3
	No visible oedema, but patient reports heaviness		Soft visible oedema; no pitting, reversible	Soft pitting oedema; reversible	Firm pitting oedema; not reversible; no tissue changes	Irreversible; tissue changes
ALOHA (Nixon et al, 2014; Purcell et al, 2016)						
Validated?	Yes					
Staging	MoistureMeterD (grading unspecified)			Neck tape measuring system		
Secondary Upper Quadrant Lymphedema (SUQL) based on Internal Society of Lymphology (Levenhagen et al, 2017)						
Validated?	Yes					
Level			0	I	II	III
	Staging Objective measures include bioimpedence analysis, circumferential measurement, water displacement, perometry and imaging		Subclinical state – peripheral swelling is not visible, but lymphatic transport is impaired. Symptoms and subtle tissue changes may be noted	Early onset of swelling that is visible and subsides with elevation. Pitting may be present.	Consistent volume change with pitting present. Elevation rarely reduces the swelling and progressive tissue fibrosis occurs	Skin changes such as thickening, hyperpigmentation, increased skin folds, fat deposits, and warty overgrowths occur. Tissue is very fibrotic and pitting is absent.

Table 6. Cont.

CTCAE (Deng et al, 2011)					
Validated?	Yes				
Grade	1	2	3	4	5
	Localised to dependent areas; no disability or functional impairment	Localised facial or neck oedema with functional impairment	Generalised facial or neck oedema with functional impairment (e.g. difficulty in turning neck or opening mouth compared to baseline)	Severe with ulceration or cerebral oedema; tracheotomy or feeding tube indicated	Death
ACS Lymphedema of Head and Neck (Deng et al, 2011)					
Validated?	Yes				
Stage	0	I	II	III	
	Swelling is local and does not affect regular functioning .	Swelling is local and affects regular functioning.	General swelling in the face or neck affects regular functioning (e.g., it may make it more difficult for a person to turn head or open and close mouth)	Swelling is severe and may accompany ulcers on the skin or brain swelling; the ability to eat is severely affected.	
CCL (Deng et al, 2011)					
Validated?	Yes				
Stage	0	I		II	
	No or minimal fibrosis (i.e., oedema pits on pressure and reduces with limb elevation)	Substantial fibrosis clinically (i.e., oedema does not pit and does not reduce with limb elevation)		Grade 2 plus elephantine (trophic) changes	
Stages of Lymphedema (Deng et al, 2011)					
Validated?	Yes				
Stage	0	I	II	III	
	Pathology: latency—focal fibrosclerotic tissue alterations Signs and symptoms: none Diagnosis: functional isotope lymphography	Pathology: reversible – high protein oedema, focal fibrosclerotic tissue alterations Signs and symptoms: pitting oedema; elevation reduces swelling and possibly "pain of congestion"	Pathology: spontaneously irreversible – extensive fibrosclerosis, proliferation of adipose tissue Signs and symptoms: brawny, hard swelling that does not recede with elevation	Pathology: elephantiasis – extensive fibrosclerosis, proliferation of adipose tissue Signs and symptoms: like stage II, invalidism	
Modified Patterson Scale (Starmer et al, 2021)					
Validated?	No				
	Location: Epiglottis, vallecula, pharyno-epiglottic folds, aryepiglottic folds, false vocal cords		Subjective rating of oedema: normal, mild, moderate, severe, not evaluable		

ACS = American Cancer Society; ALOHA = Assessment of Lymphedema of the Head and Neck; CCL = Clinical Classification of Lymphedema; CTCAE = Common Terminology Criteria for Adverse Events; HN-ELAF = Head and Neck External Lymphedema and Fibrosis Assessment

of life. By bridging clinical expertise with patient experiences, the field will be able to move towards a more standardised approach to lymphoedema assessment and care.

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