Disease severity scores in dermatology: An update of the various indices

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Need for using disease severity indices: A need-based approach

Dermatology often deals with long-standing and chronic conditions which have important ramifications on the patient's day-to-day life. However, to assess the treatment outcomes and relief provided to patients, it is often necessary to measure the baseline disease severity, and compare it to post-treatment severity. Successful treatment often implies a reduction in quantifiable and somewhat objective scores of disease severity.

Characteristics of an ideal disease scoring index

The huge diversity of cutaneous conditions mean no single measure of disease severity will suffice all diseases–some diseases like atopic dermatitis are predominantly characterised by itch whereas others like pemphigus can have debilitating reduction in quality of life of the sufferer but have little to no itch. Thus, the characteristics of an ideal scoring index should be:

- Minimum inter-observer variation: There is little observer-to-observer variation in scores.
- Accurately captures how severe the disease is: It should properly reflect the disease severity, including all parameters that affect the sufferer's quality of life.
- Easy to calculate: It should be easy to calculate and preferably have little to no complicated calculations. It should be capable of being calculated bedside. For example: The Psoriasis Area and Severity Index (PASI) score for psoriasis is an easy to calculate score.
- Should not involve complicated investigations. For example, Modified Rodnan Severity Score (MRSS) for systemic sclerosis is an example where no invasive tools are used even though the disease in question is systemic.
- Capture both subjective & objective symptoms: For example, in Scoring Atopic Dermatitis (SCORAD) score for atopic dermatitis, itch and sleeplessness which are subjective symptoms are included, together with cutaneous plaques, which is an objective finding.
- Sensitive to even minor changes in disease activity.

Important scoring indices widely used in dermatology

Follicular-pilosebaceous unit disorders

Major diseases under this category include acne vulgaris, rosacea, hirsutism and hidradenitis suppurativa. Several scoring indices are used: A common theme among them is calculating the area involved by disease and the characteristic lesional morphologies, with increasing severity. Scoring indices for acne are amongst the most widely used and the most widely used is Global Acne Severity Score (GASS). For hidradenitis suppurativa, the Hurley staging score is used, and may have prognostic significance. For alopecia areata, Severity of Alopecia Tool (SALT) score is widely used, especially in research settings, although other scores like Alopecia Density Extent Index (ALODEX) and Lesional Area Density (LAD) score exist. A detailed analysis of these scores is presented in Table 1.

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Table 1: Disease severity scores for pilofolliculosebaceous disorders

Disease	Severity score	Remarks	Advantage	Disadvantage
Alopecia areata	i) SALT I and II[1]	For SALT score, the entire scalp is divided into 1% areas, each of which is again graded into 0–10 scale depending on the severity. The total scores are summed. Viz Table 2	i) Assessment of severity is essential for deciding response to treatment. ii) SALT score is a simple bedside technique which is often frequently used before initiating tofacitinib. No instruments/imaging involved. iii) Found to correlate well with clinical severity.	i) Somewhat tedious. ii) Time-consuming. iii) Not routinely used in clinical practice.
	ii) AASI[2]	Parameters considered: % of hair loss, hair pull test results, areas of no ostia/yellow dots. A weighted score is then calculated for each of left side, right side, top and back and then added. Viz Table 3	Found to correlate well, especially for hair of colour. More objective than SALT.	Labour-intensive. Steeper learning curve. Magnification required. Considerable inter-observer variation.
	iii) ALODEX score[3]	Measures alopecia as a % of scalp.	Useful for assessing treatment response.	Requires iPAD app. Time-consuming.
	iv) LAD score[3]	Combines lesional area with density of hair. A 100-point density scale was used.	Semi-objective measurement. Correlates with clinical severity.	Inter-observer variation.
Hirsutism	Ferriman-Gallwey score[4]	Used to grade hair over chest, abdomen, facial areas etc. across nine areas of the body, each area being 0–4.	Defines Hirsutism. Ferriman-Gallwey <8: Normal 8-15: Mild >15: Moderate to severe	May not exactly reflect the psychosocial burden.
Acne vulgaris	i) GASS[5]	Grading of affected areas as: 1: Comedone. 2: Comedones with occasional pustules on face. 3: Many comedones & inflammatory papules/pustules on face. 4: Nodules & cysts.	Relatively easy to calculate. Can be effectively used as a quick assessment tool.	May not reflect exact psychosocial burden–acne, being a disease of considerable psychosocial impact.
	ii) James and Tisserand scoring system[5]	Grade 1: Simple non-inflammatory acne ± papules. Grade 2: Comedones, papules & a few pustules. Grade 3: Severe inflammatory comedones. Grade 4: Cysts.	Easy to do Bedside tool Good inter-observer variation.	Not much affected by treatment response. Not useful at all to grade treatment response since treatment typically does not change from higher grade acne to lower grade of acne. Rather, the number of lesions are typically decreased.
	iii) Doshi Zaheer & Stiller systems[5]	Divides the chest, face and back into six areas and calculates a weighted score.	Easier to do. No instruments needed.	Lacks sensitivity; Has now been superseded by GAGS.
	iv) Hayashi *et al.* Photographic system (latest, in 2008)	Uses photographic method to grade score.	No complex calculations needed. Lesser inter-observer variation. Better suited for assessing response to treatment.	Needs cameras. Needs cameras.
	v) Leeds scoring index[5]	12 colours pictographic method for face & eight colour pictographic method for chest and upper back.	Easier to do. Conveys pictographic information. Correlates well with clinical improvement. Easier to administer.	Equipment intensive process.
	vi) Modified Cooks method	Uses photographic standards to grade acne with even grades: 0,2,4,6,8.	Does not have much inter-observer variation.	Equipment intensive.
Acne scars[6,7]	i) Lesional counting	Counts the number of lesions.	Easier to do.	Does not necessarily correlate with high psychosocial impact.
	ii) ECCA	ECCA is most commonly. used: The number of lesions are counted & each lesion is scored from 0–3 0: No scar 1: <5 scars 2: 5–20 scars 3: >20 scars	1. Helps in deciding modality of treatment. 2. Can help in assessing response to a therapy.	1. Subjective factors viz social impact not taken into account.

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Disease	Severity score	Remarks	Advantage	Disadvantage
	iii) Qualitative acne scar grading scale[7]	4 grades 0–4 based on whether scars are visible from 50 cm or they are hidden by beard and body hair.	Easy to do. Correlates well with disease severity, particularly psychosocial impact.	Subjectivity. Not routinely used.
Rosacea	Rosacea scoring index[8] Viz., Table 4	Scores symptoms like flushing, erythema, dryness, eye symptoms, nasal symptoms into absent, mild, moderate, severe.	Gives an idea about what therapeutic intervention is required. No investigations needed.	Tedious Photographic evidence better suited. Not used generally for day-to-day practice.
Hiadrenitis suppurativa[9]	1. Hurley staging system,	Hurley's stage is frequently used for measuring severity of disease depending on presence of i) sinus tracts ii) scars (sometimes bridging) iii) abscess.	Usually, Hurley's staging is the most widely used. Easy to do. Inexpensive.	Sometimes differentiation between a sinus tract and a non-healing ulcer is difficult. May not be fully reflective of the disease burden.
	2. MSS, 3. PGA, 4. HSSI, 5. AISI, 6. Canoui-Poitrine scoring system 7. HiSCR	It has three stages.	Assessing disease severity, and amenability to surgery is easier with this tool. Assessment for treatment response, HiSCR is preferred No investigations needed.	

SALT: Severity of alopecia tool, AASI: Alopecia areata severity index, ALODEX: Alopecia density extent index, LAD: lesional area density, GASS: global acne severity score, ECCA: Echelle d'Evaluation Clinique des Cicatrices d'acne, MSS: Modified Sartorius score, PGA: Physician's global assessment, HSSI: Hidradenitis suppurativa severity index, AISI: Acne inversa severity index, HiSCR: Hidradenitis suppurativa clinical response score

Table 2: Steps for SALT score for alopecia areata

1. Divide and label each area of the scalp into areas of 1% width
2. Rate each of these area on a scale of 0–100 of increasing severity
3. Sum up each of these scores to get total score

Maximum score: 100 **Minimum score: 0**

Table 3: Steps to score Alopecia Areata Severity Index (AASI)

1. Divide the entire scalp into four areas: A. Left B. Right C. Top D. Back
2. Calculate percentage (%) of Alopecia in each of A, B, C, D. Let them be called AA(L), AA(R), AA(Top), AA(Back) respectively
3. Perform hair pull test in each of A, B, C, D and grade as follows:
   0: Negative
   1: 10–20% of grasped hair comes out
   2: >20% of grasped hair comes out
   Perform this step five times in each of A, B, C, D and take the average for each of these four zones
4. Perform magnification test in each areas in 3x magnification in each of A, B, C, D and look for
   i) Yellow dots
   ii) Broken hair
   iii) Exclamation mark hair
   iv) Dystrophic vellus hair

   Next grade each of A, B, C, D as follows depending into what percentage each of A, B, C, D is affected by any of these findings listed above.
   0: Any of these findings are seen in 0% of each of A, B, C, D separately.
   2: Any of these findings are seen in <50% of each of A, B, C, D separately.
   4: Any of these findings are seen in >50% of each of A, B, C, D.

5. Add up scores of (3) and (4) for each of A, B, C, D to get SL index (severity of loss). Let them be called SL(L), SL(R), SL(Top), SL(Back). respectively.
6. Final Score = [AA(L)x SL(L) x 0.18] + [AA(R) x SL(R) x 0.18] + [AA(Top) x SL (Top) x 0.4] + [AA(Back)x SL(Back) x 0.2].

Table 4: Steps for calculating rosacea scoring severity index

Rosacea scoring severity index	
Primary features	Score each of these primary features as
A) Transient erythema	0: Absent
B) Non-transient erythema	1: Mild
C) Papules/pustules	2: Moderate
D) Telangiectasias	3: Severe
Secondary features	Score each of these secondary features as follows (except oedema)
i. Burning/stinging	0: Absent
ii. Plaques	1: Mild
iii. Dry appearance	2: Moderate
iv. Oedema	3: Severe
v. Ocular manifestations	For oedema, mark present/absent
vi. Phymatous changes	
vii. Peripheral location: absent/present	
Global rating	
A. Physicians global rating	
i. Erythematotelangiectatic	
ii. Papulopustular	
iii. Phymatous	
iv. Ocular	
B. Patients' global rating	

Papulo-squamous disorders

The most well-known disordet is psoriasis and PASI score is widely used, both for assessing severity and therapeutic response. Targeting a 90% reduction from baseline, so called PASI 90 is now a therapeutic goal across all modalities of treatment. Of note, the basic format for calculating PASI has been extrapolated and applied in lichen planus, pityriasis rubra pilaris and other conditions. For atopic eczema, SCORAD is used, while for hand eczema HECSI is used. Their detailed analysis is presented below, vide Table 5:

Table 5: Severity indices for Papulosquamous disorders

Disease	Scoring index	Brief comments	Advantages	Disadvantages
Psoriasis	i) PASI	Weighted score, included: I) Erythema. II) Scaling. III) Induration. Each of them is graded into 0–4 based on severity and their sum added (referred as "A"). Then area as % of region is graded into 0–6 (referred as "B") C = A × B. C is calculated over head-neck, upper limbs, trunk, lower limbs. Weightage factor is 0.1 for head neck, 0.2 for upper limbs, 0.3 for trunk, 0.4 for lower limbs. Final score is added across these four body regions.	i) Semi-objective. ii) Can help decide the modality of therapy: BSA >10% and PASI >12. are indications to start systemic therapy like Methotrexate, Cyclosporin, NB-UVB etc. iii) Gauze treatment response: PASI 75 denotes 75% reduction is PASI from baseline–used as a standard target while starting therapy. iv) PASI 90 is now the better standard of therapeutic response. v) Online ready-to-use calculators available.	i) Somewhat difficult to calculate bedside. ii) Difficult to perceive the degree of erythema in dark skin. iii) Inter-observer variation in grading. iv) Not to be done in erythrodermic or generalised pustular psoriasis of Von Zumbusch. v) May be falsely low in drug-induced psoriasis & treatment modified psoriasis. vi) No subjective symptoms like itch etc. included.
	ii) LS-GPS	Weighted evaluation and use of eight steps score.	Relatively accurate.	Very cumbersome; now rarely used.
	iii) National Psoriasis Foundation Severity Score[10]	Uses multiple domains viz induration of plaques, body surface area, global assessment.	Encompasses multiple domains. Takes into account the subjective component of patient defined itch.	Time-consuming.

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Disease	Scoring index	Brief comments	Advantages	Disadvantages
Psoriatic arthritis	psAId score[11]	12 domains were assessed with each domain scored 0–10.	i) Easy to do. ii) Reliable. iii) Can be used to monitor therapeutic response.	i) Slightly tedious. ii) More appropriately done by rheumatologists than dermatologists.
Palmoplantar pustulosis	ppPASI[12]	Modification of the PASI score. Scores erythema, scaling (desquamation) and pustules/vesicles. Both palms and both plantars are scored from 0 to 4. The extent of involvement of each region as defined in PASI is scored from 0 to 6. The total ppPASI score range from 0, to maximum of 72.	i) Semi-quantitative. ii) Used to gauze efficacy in RCTs. Example: Secukinumab's efficacy in PP was first identified using ppPASI 75 at week 16. NCT02008890. iii) Like PASI, ppPASI 90 is a therapeutic target in most developed nations.	i) Slightly complicated to calculate. ii) Many aspects of difficulties in day-to-day life not incorporated e.g., itch, pain etc. iii) Not very popular for routine clinical use. Mostly used for research purposes.
Nail psoriasis	NAPSI[13]	Divides the nail into eight quadrants. Each nail finding of Matrix & Bed is scored 1. Total score added. Viz., Fig. 1 & Table 17	i) Very easy to do. ii) Minimal inter-observer variation.	Still a research tool. Changes slowly in response to treatment.
PRP[14,15]	PASI A composite scoring index is under development.	Same as psoriasis.	In the absence of any specific scoring systems, PASI, with fair sensitivity, affords the best option to measure disease activity.	Non-specific–Many attributes of PASI are not directly observed or of limited value in PRP.
Lichen planus[16]	Lichen planus severity index[16] Viz Table 6	Five types of lesions– i) erythematous papule ii) violaceous papule, iii) violaceous plaque, iv) hyper-pigmented hypertrophic papule. v) Post-inflammatory hyperpigmentation. Total involved BSA determined and a BSA factor assigned. Area factor for each of these morphological lesions is calculated and multiplied with the respective multiplication factor. Sum of all the products gives the lesion severity score. Product of lesion severity score with the body surface area factor gives the final lichen planus severity score.	i) Developed in 2020 in India. ii) Found to be sensitive to changes in disease activity–good correlation. iii) Can guide decisions to switch over to systemic therapy.	1. Very tedious to calculate. 2. Subjective symptoms not considered–LP is very itchy. 3. Not applicable for Oral LP/ Nail LP. 4. Therapeutic targets not yet defined like PASI 75 or PASI 90.
Atopic eczema[17]	i) SCORAD and Objective SCORAD (O-SCORAD) viz Table 7 EASI[12,17]	A: Extent (0–100) B: Intensity (0–18) C: Subjective symptoms (0–20) Total Score: a/5 + 7B/2 + C. Total score: 0–103. viz Image 3. O-SCORAD has 0–83 range Like PASI, but the four parameters are erythema, induration, lichenification & scratching.	1. Captures subjective domains, objective domains. 2. Can be used to assess treatment response. 3.Versions available in Indian languages. 1. More objective. 2. Easy to do.	1. Very tedious and time-consuming. 2. Somewhat subjective. 3. May be dependent on level of education of respondents. Best suited for research settings. Better for research purposes.
Hand eczema[18-20]	HECSI	Six different symptoms (redness, scaling, vesicles, oedema, infiltration, and fissures), as well as the area involved. The total scores range from 0 to 360.	1. Captures objective & subjective responses. 2. May be used to guide therapy. 3. For RCTs, can offer excellent tool. 4. Found to have excellent inter-observer agreement & fairly responsive.	1. Time-consuming 2. Photographic evidence better suited for day to day management. 3. Further improvement can be done by incorporating 41 items.

PASI: Psoriasis area severity index, LS-GPS: Lattice system global psoriasis score, psAId: psoriatic arthritis impact on disease, NAPSI: Nail psoriasis activity severity index, PRP: Pityriasis rubra pilaris, EASI: Eczema area severity index, HECSI: Hand eczema severity index

Table 6: Lichen planus severity index

The Lichen Planus Severity Index

Step I: Calculate BSA and then assign one of the following factors:

0–10% → 1
10–30% → 2
30–50% → 3
50–70% → 4
70–90% → 5
>90% → 6

Step II: The number and percentage (%) of each of the morphological types of lesions is calculated.

Step III: Assign an AIF for each of the six types of lesions.

Step IV: Multiply area involvement factor with weightage factor for each of the six morphological types.

Step V: Add the scores of step IV for each type of lesion & calculate the total score.

Step VI: Multiply this total score obtained in Step V with the total BSA score calculated in step I.

BSA: Body surface area, AIF: Area involvement factor

Table 7: The SCORAD scoring sheet

A. Disease extent as % of body surface area	**____% of BSA involved**
B. Objective signs	Score each of (i) to (v) as
i) Papules/vesiculation	0: Absent
ii) Ooziness/crust	1: Mild
iii) Lichenification	2: Moderate
iv) Erythema	3: Severe
v) Xerosis	
C. Subjective signs	Score each of (i) and (ii) on a score of 0–10 of increasing severity
i) Pruritus	
ii) Loss of sleep	

Final Score: A/5 + 7 B/2 + C

Table 8: Steps to calculate PDAI

Step IA: Skin activity score:

In each of the below-mentioned areas, look for **erosions, blistering or new onset erythema** and then grade as follows:

0 → Absent
I → 1–3 lesions, at least **one of which** >2 cm in diameter and none >6 cm in diameter
2 → 2–3 lesions, at least two of which is >2 cm in diameter and none >6 cm in diameter
3 → >3 lesions, of any size but < 6 cm in diameter
5 → >3 lesions and/or at least one lesion > 6 cm in diameter
10 → >3 lesions and/ or at least one lesion > 16 cm in diameter or entire area

Area	Score
Ear	
Nose	
Face	
Neck	
Chest	
Abdomen	
Back and Buttocks	
Arm	
Hands	
Leg	
Feet	
Genitalia	
Total	_____/120

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Steps to calculate PDAI

Step II: Grade scalp as follows:

Look for erosions/ blisters/erythema and then grade as follows:

0 → Absent

1 → These changes are seen only in one quadrant

2 → These changes are seen in two quadrants

3 → These changes are seen in three quadrants

4 → These changes are seen in four quadrants

10 → Any lesion measuring >16 cm anywhere on scalp

Step III: Score each of the mucous membranes as follows:

0 → absent

1 → 1 lesion

2 → 2–3 lesions

5 → 3 lesions or two lesions > 2 cm

10 → entire area

Mucous Membrane	**Score**
Eyes	
Nose	
Buccal mucosae	
Hard palate	
Soft palate	
Upper gingiva	
Lower gingiva	
Tongue	
Floor of mouth	
Labial mucosa	
Posterior pharynx	
Anogenital mucosae	
Total score	/out of 120

Step III: Damage score

(Note: Not applicable to mucosae; Only for skin and scalp)

Score each of skin and scalp as follows:

0 → No residual damage

1 → Post-inflammatory hyperpigmentation or erythema vide Table 9

Area	**Score**
Ear	
Neck	
Face	
Nose	
Chest	
Abdomen	
Back & buttocks	
Arm	
Hand	
Feet	
Legs	
Genitals	/ out of 12

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Steps to calculate PDAI

Step IV: Scalp damage score

Score the scalp as follows

0 → if no damage seen

1 → if any damage from post-inflammatory hyperpigmentation or resolving erythema is seen in any quadrant anywhere on the scalp

Total Score = _____/2

Step V: All scores of Step I, II, III and IV are added to get the final objective score

Adapted from Boulevard C, Duvert LS, Picard-Dahan C, Kern JS, Zambruno G, Feliciani C et al. International Pemphigus study group. Calculation of cut-off values based on Autoimmune Bullous disease Severity Score (ABSIS) and Pemphigus Disease Area Index (PDAI) pemphigus scores for defining moderate, significant and extensive types of pemphigus. Br J Dermatol. 2016: 175(1):142-9

Table 9: Calculation of subjective score

Pemphigus Disease Activity Index

Step I: Skin (A)

In the skin, each area listed below is examined for the most dominant type of lesion and then a weighing factor is assigned

1.5 → Erosive, exudative areas

1 → Erosive, dry

0.5 → re-epithelized lesions

Area	Surface area involved	Weighing factor
Head-neck		
Right upper extremity		
Left upper extremity		
Trunk		
Right lower extremity		
Left lower extremity		
Genitals		
Total		

Step 2: The BSA and weighing factor are now multiplied.

Step 3: The sum is calculated to get a skin score.

Step 4: Mucous membrane score.

In each of these areas, give 0; if no lesion is present, else give 1.

Mucous membrane	Mucous membrane score
Upper gingival mucosa	
Lower gingival mucosa	
Tongue	
Right buccal mucosa	
Left buccal mucosa	
Hard palate	
Soft palate	
Upper labial mucosa	
Lower labial mucosa	
Pharyx	
Anogenital mucosa	
Total	

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Step 5: Sum up the total mucosal score to get a total mucosal score (TMS)
Step 6: Subjective symptoms: Ask the patient to rate the level of discomfort on a scale of 0–1.5 for each of these food items. Each of these food items is graded according to increasing hardness as follows. Multiply the level of food (1 through 10) with the difficulty score to get a subjective score:

Subjective Scoring:
0 → No problem in eating the food
1 → Bleeding/pain sometimes occur
1.5 → Bleeding/pain always occur

Food item	Food level (A)	Difficulty (B)	Severity score (A x B)
Water	1		
Soup	2		
Yoghurt (dahi)	3		
Custard	4		
Mashed potato/Scrambled egg	5		
Baked fish	6		
White bread	7		
Apple	8		
Whole grain bread	9		
Total			

Step 9: Add Skin Score + Mucous Membrane Score + Subjective score to get the total ABSIS

Table 10: SLEDAI 2K score

Seizure Psychosis Organic brain dysfunction Cranial nerve dysfunction Vasculitis Visual disturbance Lupus headache	Each of these, when present, is given a score of 8 (eight)
Arthritis Myositis Urinary casts Proteinuria (>0.5 g/24 hours) Pyuria (>5 WBC/HPF) Hematuria (>5 RBC/HPF)	When each of these is present, give a score of 4
Rash Alopecia Ulcers Pleurisy Pericarditis Low complement (C3, C4, CH50) Increased DNA binding by Farr Assay	Each of these, when present, gives a score of 2
Fever Thrombocytopenia Leukopenia	Each of these, when present, gives a score of 1
Total	

Add the scores to get the total SLEDAI 2K score

Table 11: MRSS Score

Area	Right	Left
Fingers		
Hands		
Forearms		
Upper arms		
Thighs		
Legs		
Feet		
Face		
Anterior chest		
Anterior abdomen		
Total		

In each of the above-mentioned areas, score skin tightness as follows:
0 → No thickening
1 → Mild thickening
2 → Moderate thickening
3 → Severe thickening

Infective conditions

Scoring indices are not widely used in infective dermatology, as these conditions are mostly acute and do not have long-term effects. Nevertheless, for some common dermatoses, like Tinea, which is assuming an unprecedented & epidemic proportions in India, ISATP scoring has been proposed vide Table 12.

Table 12: Disease severity scores for infective conditions

Disease	Severity index	Remarks	Advantage	Disadvantage
Tinea pedis[21,22]	ISATP. Alternative scoring index: AFSS.	Based on PASI. ISATP = 0.1 (Er + Es) A + 0.3 (Er + Es) A + 0.6 (Er + Es) A Er and Es where Er and Es represent redness & scaling.	1. Easier to calculate. 2. Useful to gauze effectiveness of new research molecules.	Not widely validated. Should not be used for clinical decision making till it is validated worldwide. Not useful for day-to-day practice.
Onychomycosis	OSI[22]	It is derived by multiplying the score for the area of involvement (range, 0–5) by the score for the proximity of disease to the matrix (range, 1–5). Ten points are added for the presence of a dermatophytoma or for greater than 2 mm of subungual hyperkeratosis. Mild onychomycosis = 1–5; moderate, 6–15; severe, 16–35.	1. Easy to calculate. 2. Can be used to assess response to treatment since it is highly resistant to therapeutic interventions.	1. Not routinely used in clinical practice.
Leprosy reactions	ENLIST ENL Score[23]	10-item list used to assess the severity of type 2 reactions.	1. Fairly objective. 2. Can be used to measure therapeutic responses.	1. Not routinely used. 2. Limited value in day-to-day practice since steroids are effectively used to manage ENL.

ISATP: Impairment & severity of tinea pedis, AFSS: Athlete foot severity index, OSI: Onychomycosis severity index

Bullous dermatosis

ABSIS score is widely used for all bullous dermatoses and is not specific for any one AIBD. For more specific scoring indices, PDAI for pemphigus (vulgaris and foliaceus and their variants), BPDAI for bullous pemphigoid and ODSS for any oral involvement. Following successful therapy, a quantifiable reduction in these scores is usually observed, with fair degree of clinical correlation. vide Tables 8-10 and 13

Table 13: Disease severity scores for Autoimmune blistering disorders

Disease	Severity index	Brief remarks	Advantages	Disadvantages
Pemphigus[24,25]	i. PDAI	**PDAI:** Total score 0–263 of which 120 for skin, 10 for scalp & 120 for mucosal activity. Moderate Activity: 0–15; Significant–15 to 45; Extensive: >45 Viz Table 8A-C	1. Used to define moderate, severe & extensive pemphigus. 2. Can help assess response to various modalities of therapy like pulse, rituximab etc. 3. More stress to objective findings (250/263). 4. Mucosa have a separate score.	1. May be time-consuming especially for beginners. 2. Assessing entire mucosae may not always be feasible (E.g., genital mucosae etc.). 3. Somewhat greater inter-observer variation.
	ii. ABSIS (pemphigus disease activity index)	**ABSIS:** 0–206. Out of it, 150 is for skin damage, 11 for mucosal damage and 45 for oral pain and bleeding. Table 9 A-B	1. Objective, lesser inter-observer variation 2. No instruments/ intensive equipment needed. 3. Shows variability with treatment response.	1. Very time-consuming. 2. Not proper for Indian settings–some foods like baked fish and beef steak are not commonly used in India.
	iii. POLIS[25]	Polis stands for pemphigus oral lesions intensity index. nine factors are considered, number of relapses, disease duration in weeks, number of relapses of oral lesions, persistence of oral lesions after subsisdence of cutaneous lesions, change in size of oral lesions development of new oral lesions in the past 1 week, difficulty in speaking.	1. Relatively sensitive measurement. 2. Also agrees well with clinical severity of oral lesions in pemphigus. Easy to do and bedside.	1. New score, not much known about it. 2. Somewhat reliant on patient history and prone to recall errors on the part of the patients.
Bullous pemphigoid	BPDAI.	Total 3 domains: 1. Number & size of bullous lesions–120 points. 2. Number & size of non-bullous lesions–120 points 3. Number & size of mucosal lesions–120 points	Easier score than PDAI & ABSIS. More sensitive & specific than ABSIS. Can be used to assess response. Separate scoring for bullous & non-bullous Pemphigoid	Time-consuming Few centres practice routine scoring for Bullous Pemphigoid patients. Scope & applicability for other Pemphigoid disorders like anti-laminin pemphigoid unknown.
Dermatitis Herpetiformis[26]	No specific Scale. Visual analogue scale may be used for itch	VAS consists of smiley facies, which may be used to measure itch.	Easy & not time-consuming.	Not specific to DH.
Mucous membrane pemphigoid[27]	ODSS. MMPDAI	ODSS is superior to MMPDAI.[26] ODSS: Sum of mucosal score and subjective scores.	Both emerging & developing tools. ODSS also applicable for oral LP. May help to choose therapy.	Not being very widely performed. Better performed with help of oral surgeons for oral lesions. Difficult to administer in Indian settings–some foods are unknown to Indians and vegetarianism also possess a problem as baked fish & Beef steaks are rarely used in India. Indian alternatives are being actively searched.

PDAI: Pemphigus disease activity severity index, BPDAI: Bullous pemphigoid disease activity index, ODSS: Oral disease severity score, MMPDAI: Mucous membrane pemphigoid disease activity index

Rheumatic dermatological diseases vide Tables 10, 11 and 12

Table 14: Disease severity scores for some rheumatological conditions

Disease	Score	Remarks	Advantage	Disadvantage
Systemic lupus erythematosus (SLE)[28,29]	i. SLEDAI Viz Table 9 ii) SLEDAI 2000	SLEDAI: 24 weighted clinical parameters based on 9 organ systems. All parameters within last 10 days.	SLEDAI is a strong predictor of Mortality in SLE. Extremely useful clinically.	1. SLEDAI does NOT capture worsening/improving symptoms rapidly. 2. Difficult to practice clinically
	ii. SELENA-SLEDAI ` SELENA	SELENA: Added items like alopecia, mucosal ulcers, and rash. SELENA considers 24 items across nine organ systems maximum score is 0–105	1. SELENA SLEDAI has a separate flare index to capture flares.	Somewhat time-consuming.
	iii. BILAG	Eight[8] organ systems are assessed and disease seveirt in each of them was assessed from A to E with A indicating most severe disease, requiring > 20 mg Prednisolone and E mild disease.	Appropriate to calculate organ-specific severity. Useful to assess response to therapy.	Time-consuming. Somewhat reliant on patient-provided history, which may not always be very accurate. Appropriately done by rheumatologist.
	iv. SLAM	Assesses disease activity in the last one month based on domain theory.	Captures current disease status; not much influenced by past flare.	Time-consuming. Better done by rheumatologists.
Systemic sclerosis[30,31]	MRSS Variations of MRSS include MRSS 5 (Restricted to five sites, Maximum score bing 10) And MRSS 17 (where 17 sites are scored) **viz** Table 9[29,30]	Each area is scored 0 for no thickening, 1 for mild, 2 for moderate & 3 for severe thickening across 17 sites. Back, scalp is not scored. Then score added. Range: 0–54.	1. Defines limited systemic sclerosis (MRSS <14) vs diffuse systemic. Sclerosis (MRSS >14) 2. Very useful bedside tool for measuring disease activity and therapeutic response.	1. Cannot differentiate between active and burnt-out disease. 2. Subjectivity involved.
Dermatomyositis	DSSI[31]	Recently developed. Consists of skin-specific manifestations.	Found to agree well with PGA scores.	Not yet widely validated. Implications & changes with response to therapy are not known.

SLEDAI: SLE disease activity index, SELENA: Safety of Estreogens in Lupus National Assessment, BILAG: British Isles Lupus Assessment Group, SLAM: systemic Lupus activity measure index, MRSS: Modified Rodnan severity score, DSSI: Dermatomyositis skin severity index

Table 15: VITIQOL Questionnaire

1. Do you feel bothered by the appearance of your skin?
2. Did you feel frustrated by the appearance of your skin?
3. Did you feel difficulty in showing affection due to your skin?
4. Did you feel difficulty in your daily activities on account of your skin?
5. While talking to someone did you worry what people might think about you?
6. Were you afraid that people might criticise you because of your skin?
7. Did you feel embarrassed or inhibited because of your skin?
8. Did the appearance of your skin affect the choice of your clothing?
9. Did you skin affect your leisure activities?
10. Did your skin condition affect your emotional well-being?
11. Did your skin condition affect your physical health as a whole?
12. Did your skin condition affect your personal appearance (haircuts/use of cosmetics etc.)?
13. Did your skin condition affect your sun protection behaviour in your leisure activities?
14. Did your skin condition affect your making new friends?
15. Did you worry about disease progression to other parts of the body?
16. Rate how severe your skin condition is on this scale of 0–10?

Note: The time frame for all these questions is the “Last one month

Table 16: SCORMA calculation sheet

SCORMA Index	
Scoring Mastocytosis Activity Index	
A. Extent	**Body surface area %_____**
B. Intensity i) Pigmentation ii) Vesiculation iii) Elevation/induration iv) Positive Darier sign	Each of these when present is given a value of 0–3 depending on the severity.
C. Subjective symptoms i) Flushing ii) Provocation factors if present iii) Diarrhoea iv) Bone Pain v) Pruritus	Each of these, when present is given a value of 0–10 depending on the severity.
Total Score	**A/5 + 5B + 2C/5 = _____**

Table 17: Nail Psoriasis Area Severity Index (NAPSI)

Step I: Divide each nail into four equal quadrants

Step II: In each quadrant, look for these signs of nail matrix findings. A score of 1 is given if for each quadrant if any of these signs are present, irrespective of the number of signs present.

i) Pitting
ii) Crumbling
iii) Red dots in Lunula
iv) Psoriatic Leukonychia

Step III: In each quadrant, look for these signs of nail bed findings. A score of 1 is given if for each quadrant if any of these signs are present, irrespective of the number of signs present.

i) Onycholysis
ii) Subungual hyperkeratosis
iii) Salmon patch/oil drops
iv) Splinter haemorrhage

Step IV: The scores for step II and step III are added to get total score for that nail. The procedure can then be repeated for all nails.

The scales used for disorders of pigmentation are tabulated below (Table 18)

Table 18: Disease severity scores for pigmentary disorders

Disorder	Index	Remarks	Advantages	Disadvantages
Vitiligo[31–34]	i) Viti QoL [viz Table 9]	Questionnaire based: 15 questions.	1. Specific to vitiligo. Indian versions are available. 2. May help decide treatment options.	Does not reflect disease progression. Usually, VitiQoL is preferred over VASI and VIDA.
	ii) VASI	For each body region, the VASI is determined by the product of the area of vitiligo in hand units (which is 1% per unit) and the extent of depigmentation within each hand unit–measured patch (possible values of 0, 10%, 25%, 50%, 75%, 90%, or 100%). The total body VASI is calculated by summing individual VASI scores for all regions.	Easy to calculate. Can help assess therapeutic response, especially to phototherapy.	Can be subjective. Does not take into account psychosocial factors. Reliant on patient given history of evolution of lesions, which may not always be reliable.
	VIDA vide Table 15	Score +4 - Activity of 6 weeks or less duration; +3 - Activity of 6 weeks to 3 months; +2 - Activity of 3 – 6 months; +1 - Activity of 6 – 12 months; 0 - Stable for 1 year or more; –1 - Stable with spontaneous repigmentation since 1 year or more	Easy to do. Reflects disease activity. Can assess response to therapy.	Significant recall bias may exist.
Melasma	MASI Modified MASI (M-MASI)	Involves assessment of pigment intensity, area & homogeneity. Modified MASI: Only area & darkness considered. Score ranges 0–24.	1. Shows excellent correlation to response to treatment. 2. Good tool for RCTs, i.e., research purpose	Not routinely done due to its tedious nature.

VASI: Vitiligo area severity index, MASI: Melasma activity severity index

Table 19: Disease severity scores for miscellaneous conditions

Disease	Severity score	Comments	Advantages	Disadvantages
1. Mastocytosis[35]	SCORMA vide Table 16	1. Consists of three parts: A = Extent B = Signs C = Subjective symptoms Net Score: A + B/5 + 2C/5 Viz Table 9	1. Easy to do. 2. No complicated investigations or invasive tests.	1. May not totally reflect the disease burden. 2. Not a substitute for imaging studies & other investigations. 3. Rarely done.
2. Cutaneous aging[36]	1. Glogau scale	Grade 1: No wrinkles. Grade 2: Dynamic wrinkles that disappear on stretching. Grade 3: Fixed wrinkles but can be camouflaged with makeup. Grade 4: Severe fixed wrinkles, tends to wear heavy makeup. 4: No makeup possible: Makeup cakes and cracks.	Very easy to do visual scale. Can be used to document wrinkle severity before rejuvenation techniques.	1. More objective indices needed. 2. Ill-suited for research.
3. Pruritus[37–39]	1. Visual analogue scale	Based on choosing the best depiction of the severity among a set of smiley icons by the patient.	Better suited for making a quick assessment of the degree of pruritus.	Cannot give much information viz degree of sleep loss etc. due to itch.
	2. The 5-D scale.	New scale that measures itch intensity on duration, degree, direction, disability & distribution. The subject marks specific checkboxes as mild, moderate and severe within each domain.	Tells about the degree of impairment in each domain. More informative and better suited for research purposes.	Somewhat time-consuming and rarely used for routine purposes.
	3. The 12-D scale.	Assesses pruritus severity across 12 items like sleep. disturbance last night, how much relief does scratching the lesions brings etc.	Can assess the psychosocial burden of chronic pruritus.	Time-consuming and not suited well for day-to-day care.

SCORMA: Scoring mastocytosis

General quality of life in skin disease measures

Apart from the ones mentioned above, there are general measures of quality of life, like SF 36 which has 36 questions pertaining to activities in day-to-day life, feelings of embarrassment at work, social and sexual life, and the necessity to wear specific clothes on account of skin disease. Others like Skindex 16, DQLI (Dermatological Quality of Life Index) and PGA (Physicians Global assessment) are widely available and downloadable from dedicated websites for day-to-day practice. They may be routinely used for all skin conditions, and although they may fail to reflect disease-specific manifestations, they may help in assessing the degree psychosocial burden due to skin disease. More work is needed for better-validated measures for rarer diseases like dermatitis herpetiformis which may have a tremendous impact on the sufferer's lives. These authors hope that the budding postgraduate residents will be acquainted with these scoring indices and incorporate them in their practice.

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Conflict of interest

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