

SCORING SYSTEMS IN BULLOUS DERMATOSES

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Sir

These group of disorders are chronic blistering auto immune disorders characterized histologically by intra epidermal/sub epidermal blister formation. They have circulatory auto antibodies against distinct adhesion molecules of epidermis and BMZ. They are potentially life threatening, characterized by widespread blisters and erosions of skin and mucosa.

Pemphigus being clinically heterogenous group of disorder, comprehensive scoring systems are needed to assess the severity of disease for effective monitoring of treatment.

Pemphigus Disease Area Index (Tabl. I) integrates cutaneous with mucosal disease in well-defined anatomical locations, assesses number and sizes of lesions and also scores post-inflammatory hyperpigmentation of resolving lesions.

Numerous scoring systems (Tabl. II-IV) have been described in literature at different times to score disease activity of pemphigus.

SCORING SYSTEMS**ABSIS****(Auto immune bullous skin disorder intensity score)****SKIN INVOLVEMENT**

2 parameters

* The extent of affected area (calculated by rule of nine)

* Quality of skin lesions

% of BSA X weighing factor

Weighing factor

1.5- for erosive, exudative lesions, blisters, positive Nikolsky sign.

1 - for erosive, dry lesions

0.5- re epithelialized lesions

0-150 points

Compare scores from initial scores with treatment to monitor disease activity.

ORAL INVOLVEMENT

1st Mucosal score-extent of lesions (0-11)

1- upper gingival mucosa

2-lower gingival mucosa

3-upper lip mucosa

4-lower lip mucosa

5-left buccal mucosa

6-right buccal mucosa

7-tongue

8-floor of mouth

9-hard palate

10-soft palate

11-pharynx

Rating

0-no lesion

1-presence of lesion

2nd Mucosal involvement- severity of lesion (0-45)

1 point-if pain/bleeding always occurred

0.5 point-pain/bleeding occurred sometimes

0 point-no pain

Types of food -

Water -1*x

Soup -2*x

Yoghurt -3*x

Custard -4*x

Smashed potatoes/scrambled egg -5*x

Baked fish -6*x

White bread -7*x

Apple/raw carrot -8*x

Whole grain bread -9*x

Skin	Activity		Damage
PDAI-skin			
Anatomical location	Erosion/blister or new erythema	Number of lesions, if <3	Post-inflammatory hyperpigmentation or erythema from resolving lesions
	0 = Absent 1 = 1-3 lesions, up to one >2cm in any diameter, none >6cm 2 = 2-3 lesions at laest two >2cm, none >6cm 3 = >3 lesions, none >6cm 5 = >3 lesions, and/or at least one >6cm 10 = >3 lesions, and/or at least one >16cm or entire area		0 = Absent 1 = Present
Ears Nose Rest of face Neck Chest Andomen Back/buttocks Arms Hands Legs Feet Genitals Total skin	/120		/12
PDAI-skin			
Scalp	Erosion/blister or new erythema	Number of lesions, if <3	Post-inflammatory hyperpigmentation or erythema from resolving lesions
	0 = Absent 1 = In one quadrat 2 = Two quadrats 3 = Three quadrats 4 = Whole skull 10 = At least one lesion ?6cm		0 = Absent 1 = Present
Total scalp	/10		/1
PDAI-mucous membranes			
Anatomical location	Erosion/blister	Number of lesions, if <3	
	0 = Absent 1 = 1 lesion 2 = 2-3 lesions 5 = >3 lesions or 2 lesions >2cm 10 = entire area		
Eyes Nose Buccal mucosa Hard palate Soft palate Upper gingiva Lower gingiva Tongue Floor of mouth Labial mucosa Posterior pharynx Anogenital Total mucosa	/120		

Table I. PDAI (Pemphigus disease activity index).

Severity		Antibody Titer Level
Oral	Skin	
0 = Nil	0 = Nil	Dsg1 and Dsg3 levels as assessed by ELISA
1 = <3 Erosions	1 = <5 Erosions	
2 = 3-4 Erosions	2 = 5-20 Erosions	
3 = >10 Erosions	3 = >20 Erosions	

Table II. Harman's Pemphigus Grading.

Oral score
0 - No mucosal involvement
1 - Minimal disease (only bucal mucosa, labiokingival, lingual, palatal, pharyngeal)
2 - Moderate disease (bucal and labiokingival, lingual, palatal or pharyngeal)
3 - Severe disease (extensive oral erosions, i.e., >3 mucosal sites affected)
Skin score
0 - Quescent disease
1 - Minimal disease (<10% BSA)
2 - Moderate disease (11-30% BSA)
3 - Severe disease (>30% BSA)

Table III. Kumar's Scoring System.

Severity	Cutaneous involvement	Mucosal involvement
Mild (1+)	10% BSA, able to carry daily routine without discomfort or localisation to oral mucosa only	Only localised to buccal mucosa, no difficulty in swallowing or chewing
Moderate (2+)	10-25% BSA along with oral mucosal involvement, able to carry out daily routine with discomfort	Buccal and gingivolabial mucosal involvement, difficulty in solid food intake
Severe (3+)	25-50% BSA along with oral mucosal involvement, unable to carry out daily routine	Extensive oral mucosal involvement, difficulty in semisolid food intake
Extensive (4+)	50% BSA along with mucosal involvement, bedridden or has complications	Extensive oral mucosal involvement, other mucosal involvement, difficulty in swallowing liquids also (unable to take anything orally)

Table IV. Mahajan's Scoring System.