Definitions and outcome measures for mucous membrane pemphigoid: Recommendations of an international panel of experts

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Mucous membrane pemphigoid encompasses a group of autoimmune bullous diseases with a similar phenotype characterized by subepithelial blisters, erosions, and scarring of mucous membranes, skin, or both. Although knowledge about autoimmune bullous disease is increasing, there is often a lack of clear definitions of disease, outcome measures, and therapeutic end points. With clearer definitions and outcome measures, it is possible to directly compare the results and data from various studies using meta-analyses. This consensus statement provides accurate and reproducible definitions for disease extent, activity,

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outcome measures, end points, and therapeutic response for mucous membrane pemphigoid and proposes a disease extent score, the Mucous Membrane Pemphigoid Disease Area Index. (J Am Acad Dermatol 2015;72:168-74.)

Key words: consensus; definitions; mucous membrane pemphigoid; outcome measures; severity score.

BACKGROUND

Mucous membrane pemphigoid (MMP) encompasses a group of autoimmune bullous diseases with a similar phenotype characterized by subepithelial blisters, erosions, and scarring of mucous membranes, skin, or both. It is associated with high morbidity and mortality, and without treatment patients can develop esophageal and laryngeal stenosis, strictures, and blindness.[1] Given the severe potential complications of MMP, effective treatment is required to delay and halt progression. Because of the rarity of this condition, however, large randomized controlled trials are lacking, and the evidence supporting these therapies is limited.[2] There has been an excellent consensus on the diagnosis of MMP,[1] but there exists a lack of clear definitions of disease stages, outcome measures, and therapeutic end points. With clearer definitions and outcome measures for MMP, it will be possible to directly compare the results and data from various studies using meta-analysis. Although ophthalmologists already developed a number of scoring systems for ocular MMP, a problem with these scores is that they combine activity with damage and are too complex for dermatologists to use. It was therefore also our intention to develop and propose a scoring system for MMP that would be practical for dermatologists who see these patients regularly to use to monitor response to therapy, which separated reversible activity from damage.

CAPSULE SUMMARY

- Currently there is a lack of common definitions of disease for mucous membrane pemphigoid and so it is difficult to make meaningful comparisons of small studies.
- These recommendations, which have been developed by international experts, provide appropriate definitions for the various stages of disease activity and therapeutic end points in mucous membrane pemphigoid.
- These definitions can be used in case series and clinical trials to compare the efficacy of treatments for mucous membrane pemphigoid.

PURPOSE

The purpose of this consensus statement is to provide accurate and reproducible definitions for disease extent, activity, outcome measures, end points, and therapeutic response for MMP. Using the same definitions of response and end points allows direct comparison of clinical trials and facilitates the analysis of these results in systematic reviews.

CONSENSUS METHODS

An international MMP definitions committee was organized in 2011. All experts in autoimmune bullous

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Abbreviations used:

BPDAI:	Bullous Pemphigoid Disease Area Index
MMP:	mucous membrane pemphigoid
MMPDAI:	Mucous Membrane Pemphigoid Disease Area Index
PDAI:	Pemphigus Disease Area Index

disease were invited to participate in the development of the MMP definitions. Experts in the field and those who had participated in previous consensus statements on pemphigus and bullous pemphigoid were invited.[3,4] The committee convened 8 times over 2 years to discuss and develop appropriate definitions. The meetings were held at the World Congress of Dermatology in Seoul, South Korea, in 2011 (D. F. M. and V. P. W.); European Academy of Dermatology and Venereology in Lisbon, Portugal, in 2011 (D. F. M. and B. M.); American Academy of Dermatology annual meeting in San Diego, CA, in 2012 (D. F. M. and V. P. W.); and Society for Investigative Dermatology in 2012 (V. P. W.). At each meeting, the minutes and issues at the previous meetings were discussed until a consensus on the definitions was made. The draft definitions and manuscript were electronically mailed to the entire committee for comments and discussion. The final consensus is the product of many meetings, discussions, and agreement. There is universal agreement in the committee about the definitions of end points, therapeutic responses, and treatment failures along with a score, termed "Mucous Membrane Pemphigoid Disease Area Index" (MMPDAI), for milder forms of MMP. For this method, the committee reviewed photographic examples from the group of patients with MMP, in particular the eyes and mouth, to discuss which areas to give weight to in the MMPDAI compared with the previous Bullous Pemphigoid Disease Area Index (BPDAI). It was decided to expand the score for the eyes to include both the left and right eye separately, which would give the eyes a weighting of 2 of 12 rather than 1 of 12 and to combine the tongue and floor of mouth as they are less often separately affected than other areas in the mouth. To further expand the weighting of the eyes, it was decided to use the quadrant system, as for the scalp, rather than counting individual lesions, which are difficult for dermatologists to visualize without a slit lamp. Because dermatologists typically cannot accurately assess other elements of the eyes, and the nasal, laryngeal, and esophageal mucosae, severe forms of MMP that may cause significant damage to these areas will require their own more detailed scores to evaluate each of these in a more specific manner.

THE CONSENSUS

Observation points

The end points are summarized in Table I.

Early observation end points

"Baseline" is defined as the day that MMP therapy is started by a physician.

"Control of disease activity" is defined as the time at which new inflammatory lesions cease to form and established lesions begin to heal. "Time to control of disease activity" (disease control; beginning of consolidation phase) is the time interval from baseline to the control of disease activity.

"Control of scarring" is defined as the time needed to control scarring progression.

"End of consolidation phase" is defined as the time at which no new lesions have developed for a minimum of 4 weeks and lesions and approximately 80% of inflammatory lesions have healed.

Intermediate observation end points

"Transient lesions" are new lesions that heal within 1 week or clear without treatment. "Nontransient lesions," however, are new lesions that do not heal within 1 week.

"Complete remission during tapering" is the absence of nontransient lesions while the patient is receiving more than minimal therapy.

"Long-term biologic therapy" refers to therapies given intermittently, for example, when rituximab is used for MMP, or intravenous immunoglobulin monthly.

Late observation end points

"Minimal therapy" in MMP corresponds to the following doses or less: dapsone 1.0 mg/kg/d; 0.1 mg/kg/d of prednisone (or the equivalent); minocycline 100 mg/d; doxycycline 100 mg/d; lymecycline 300 mg/d; topical corticosteroids once a day including fluticasone propionate suspension 400 μg/once a day; colchicine 500 μg/d; Salazopyrin 1 g/d; sulfapyridine 500 mg/d; sulfamethoxypyridazine 500 mg/d; and nicotinamide 500 mg/d.

"Minimal adjuvant therapy" (and/or maintenance therapy) is defined as the following doses or less: azathioprine (1 mg/kg/d) with normal thiopurine S-methyltransferase level; mycophenolate mofetil 500 mg/d; mycophenolic acid 360 mg/d; methotrexate 5 mg/wk; and cyclosporine 1 mg/kg/d.

"Ongoing biologic therapy" is characterized by the use of drugs such as rituximab.

Late observation end points of disease activity are identified as: (1) partial remission on minimal therapy; (2) complete remission on minimal therapy;

Table I. Definitions for mucous membrane pemphigoid

Early observation end points	
Baseline	The day that MMP therapy is started by a physician
Control of disease activity	The time at which new inflammatory lesions cease to form and established lesions begin to heal
Time to control of disease activity (disease control; beginning of consolidation phase)	The time interval from baseline to the control of disease activity
Control of scarring	The time needed to control scarring progression
End of consolidation phase	The time at which no new lesions have developed for a minimum of 4 wk, and lesions and approximately 80% of inflammatory lesions have healed
Intermediate observation end points	
Transient lesions	New lesions that heal within 1 wk or clear without treatment
Nontransient lesions	New lesions that do not heal within 1 wk
Complete remission during tapering	The absence of nontransient lesions while the patient is receiving more than minimal therapy
Minimal therapy	Dapsone $\leq$1.0 mg/kg/d; $\leq$0.1 mg/kg/d of prednisone (or the equivalent); minocycline $\leq$100 mg/d; doxycycline 100 mg/d; lymecycline 300 mg/d; topical corticosteroids once a day including fluticasone propionate suspension 400 μg/once a day; colchicine 500 μg/d; Salazopyrin 1 g/d; sulfapyridine 500 mg/d; sulfamethoxypyridazine 500 mg/d; nicotinamide 500 mg/d
Minimal adjuvant therapy (and/or maintenance therapy)	The following doses or less: azathioprine (1 mg/kg/d) with normal thiopurine S-methyltransferase level; mycophenolate mofetil 500 mg/d; mycophenolic acid 360 mg/d; methotrexate 5 mg/wk; cyclosporine 1 mg/kg/d
Long-term biological therapy	Refers to therapies given intermittently, for example, when rituximab is used for MMP, or IVIG monthly
Late observation end points	
Partial remission on minimal therapy	The presence of transient new lesions that heal without scarring within 1 wk while the patient is receiving minimal therapy for at least 2 mo
Complete remission on minimal therapy	The absence of new or established lesions while the patient is receiving minimal therapy for at least 2 mo
Partial remission off therapy	Presence of transient new lesions that heal within 1 wk without treatment while the patient is off all MMP therapy for at least 2 mo
Complete remission off therapy	Absence of new or established lesions while the patient is off all MMP therapy for at least 2 mo
Relapse/flare	Appearance of $\geq$3 new lesions a month (blisters, erosions) that do not heal within 1 wk, or the extension of established lesions in a patient who has achieved disease control

IVIG, Intravenous immunoglobulin; *MMP*, mucous membrane pemphigoid.

(3) partial remission off therapy; and (4) complete remission off therapy.

"Partial remission on minimal therapy" is the presence of transient new lesions that heal without scarring within 1 week while the patient is receiving minimal therapy for at least 2 months. "Complete remission on minimal therapy" is the absence of new or established lesions while the patient is receiving minimal therapy for at least 2 months. "Partial remission off therapy" is the presence of transient new lesions that heal within 1 week without treatment while the patient is off all MMP therapy for at least 2 months. "Complete remission off therapy" is the absence of new or established lesions while the patient is off all MMP therapy for at least 2 months.

MMP Disease Activity Index

Like the Pemphigus Disease Area Index (PDAI) and BPDAI,[3,4] the MMPDAI (Table II) measures

Table II. Mucous Membrane Pemphigoid Disease Area Index

Skin	**Activity**		**Damage**
Anatomic location	Erosion/blisters or new erythema		Postinflammatory hyperpigmentation or erythema from resolving lesion or scarring
	0 Absent **1** 1-3 Lesions, up to 1 lesion >2 cm in any diameter, none >6 cm **2** 2-3 Lesions, at least 2 lesions >2 cm diameter, none >6 cm **3** >3 Lesions, none >6 cm diameter **5** >3 Lesions, and/or at least 1 lesion >6 cm **10** >3 Lesions, and/or at least 1 lesion >16 cm diameter or entire area	No. of lesions if ≤3	**0** Absent **1** Present
Ears			
Forehead			
Rest of the face			
Neck			
Chest			
Abdomen			
Shoulders, back			
Buttocks			
Arms and hands			
Legs and feet			
Anal			
Genitals			
Total skin	**/120**		**/12**
Scalp	Erosion/blisters or new erythema	No. of lesions if ≤3	Postinflammatory hyperpigmentation or erythema from resolving lesion /scarring
	0 Absent **1** 1 Quadrant **2** 2 Quadrants **3** 3 Quadrants **4** Affects whole skull **10** At least 1 lesion >6 cm		**0** Absent **1** Present
Total scalp (0-10)	**/10**		**/1**
Mucous membrane	**Activity**		**Damage**
Anatomic location	Erosion/blisters		Postinflammatory hyperpigmentation or erythema from resolving lesion or scarring
Eyes (quadrants upper, lower, medial and lateral)*	**0** No erythema **1** Light pink **2** Moderate pink **3** Dark pink **4** Bright red add up quadrants	Subtotal	**0** absent **1** present
Left eye (0-16) x 0.625	**/10**	**/16**	
Right eye (0-16) x 0.625	**/10**	**/16**	
	0 absent **1** 1 lesion **2** 2–3 lesions **5** >3 lesions or 2 lesions >2 cm **10** entire area	Number lesions if ≤3	**0** absent **1** present
Nasal			
Buccal mucosa			
Palate			
Upper gingiva			
Lower gingiva			
Tongue/floor of mouth			
Labial			
Posterior pharynx			
Anal			
Genital			
Total mucosa	**/120**		**/12**

Total activity score: ☐ **Total damage score:** ☐

To complete the left-hand "Activity" column, look at each area in turn to see how many active blisters/erosions or quadrants are involved and mark the corresponding number in that row. For the right-hand "Damage" column, indicate the number/quadrants of scarring or postinflammatory lesions. Add these subtotals to yield a separate total activity score out of 250 and a total damage score of 21. The purpose would not be to add these together, as in studies the damage element may not decrease much, but the goal of treatment would be to decrease activity scores and hopefully not increase damage scores. *See Fig 1.

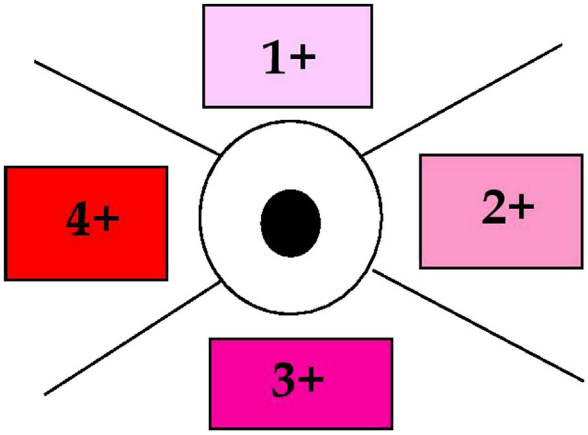

Fig 1. Diagram to illustrate how erythema is to be scored in different quadrants of each eye for the mucosal component of the Mucous Membrane Pemphigoid Disease Area Index. The degree of pinkness represents how high to score this parameter.

separate scores for activity involving the skin, scalp, and mucous membranes (Fig 1). There was much discussion about what should be involved in the MMPDAI and their respective weighting. It was noted that no consensus on how to stage MMP exists among specialists who treat MMP (eg, ophthalmologists; gastroenterologists; ear, nose, and throat surgeons; or dermatologists). Given the heterogeneity of the disease, separate tools were considered for mild MMP with oral, pharyngeal, nasal, genital, anal, and/or only inflammatory ocular lesions and severe MMP with laryngeal lesions, esophageal lesions, and/or ocular fibrosis. A few years ago, some of the authors developed and validated an outcome measure tool for pemphigus, termed the "Pemphigus Disease Area Index,"[3,5] then subsequently a modified tool for pemphigoid, called the "Bullous Pemphigoid Disease Area Index."[4,6] These tools have in common that about 45% of the score reflects skin involvement, 45% mucosal involvement, and 10% the scalp, measured in a different way to the rest of the skin. Each score has different weightings placed on the sites involved that reflect the propensity for those areas to be affected in that particular blistering disease, so that severities can be more easily distinguished, and for responsiveness to treatment to be measurable. In each condition, scarring sequelae (referred to as "damage") are scored separately from reversible disease activity and the two should not be combined.

The MMPDAI is applicable for milder forms of MMP. This tool is primarily for dermatologists who specialize in blistering diseases and who see patients with MMP regularly, but can also be used by other members of a multidisciplinary team for patients with MMP. The main purpose is its use in clinical studies for intervention and evaluation in MMP. It includes 2 columns, namely activity and damage, to separate active erosions and blisters from postinflammatory changes and scarring from resolving lesions. Active lesions are evaluated in each eye that have been divided into 4 distinct quadrants and airway scores (Fig 1) elicited depending on upper airway or posterior pharyngeal involvement. Other anatomic locations commonly affected by MMP were taken into consideration so that this score could differentiate between clinical responses in MMP. Some of the notable differences between BPDAI and MMPDAI include the addition of scarring to column "damage"; involvement of the forehead and shoulders, combination of legs and feet; and separation of anal, genital, and buttock involvement. Most notably, however, there is a separate section for scalp involvement and greater weight is given to the various mucosal surfaces. As the Brunsting-Perry form of MMP often includes the scalp and causes scarring alopecia, and this area of the body is difficult to conceal compared with other skin areas covered with clothing, it is more cosmetically disfiguring for patients. Hence, up to about 5% of the total score may be given for total scalp involvement.

Other activity scores for MMP or lichen planus with laryngeal lesions, esophageal lesions, and/or ocular fibrosis were evaluated for clinical relevance and ease of use.[7-9] Precisely scoring the ocular and laryngeal involvement would be ideal for monitoring and making therapeutic decisions. However, this excess detail had to be balanced with ease of completion in clinical and research settings for dermatologists and whether such detail would provide additional beneficial information to clinical decision making is currently uncertain.

The MMPDAI will undergo validation studies, similar to the PDAI and BPDAI.

DISCUSSION AND CONCLUSION

Because of the rarity and heterogeneity of MMP and paucity of randomized controlled trials, it has been difficult to compare the various proposed therapeutic options for MMP. This consensus paper with definitions of disease and response represents extensive discussion and agreement among experts of MMP. It provides a foundation for researchers and clinicians to develop studies with agreed upon end points so that results can be directly compared. It also provides a framework for other specialties such as ophthalmology and otolaryngology to develop a similar accurate scoring system to stage and measure the progress of MMP.

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