

Scoring systems for Mucous Membrane Pemphigoid – A review of the literature

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Abstract

Mucous Membrane Pemphigoid (MMP) is a group of rare chronic autoimmune blistering diseases that predominantly affects one or more of the mucous membranes. Without treatment it can cause significant complications such as esophageal strictures, breathing difficulties, speech difficulties, laryngeal stenosis and blindness. Despite recent advances in targeted immunotherapy there have been no randomised controlled trials to date for MMP. Whilst this is partially a reflection of the rarity of the disease, it is also due to the lack of validated scoring systems. Validated scoring systems enable an investigator to effectively document response to treatment and communicate disease severity to peers. This article reviews and evaluates existing global and mucous membrane specific scoring systems for MMP. This article also proposes the creation of an Investigator's Global Assessment for MMP as a future area of research.

Why was the study undertaken? This scoping literature review was undertaken to provide an overview of the existing scoring systems for MMP. It critically analyses the strengths and weaknesses of the scoring systems and their validation studies, and points out any pathological associations with score results.

What does this study add? This study introduces the concept of an IGA score for MMP, and discusses its utility and value.

What are the implications of this study for the understanding of skin physiology and pathology and/or disease management? We hope this manuscript will aid in the creation of a validated IGA score, thus facilitating randomised controlled trials on MMP and FDA approval for MMP treatments.

Key words: Conjunctival Diseases (MeSH), Pemphigoid, Benign Mucous Membrane (MeSH), Severity of Illness Index (MeSH), Skin Diseases, Vesicobullous (MeSH), Validation Studies as Topic (MeSH), Non-Mesh Key Words, Outcome measure, Severity Score, Validation



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1. Introduction

Mucous Membrane Pemphigoid (MMP) is a group of rare chronic autoimmune blistering diseases (AIBD) that predominantly affect one or more of the mucous membranes [1–3]. The disease is characterised by erosions, subepithelial blisters, and scarring of the mucous membranes, skin or both [4–6]. Cases of

MMP which only affect one site, such as the eyes, are called mono-site MMP and are named after the area affected [2, 7]. Diagnosis can be made through perilesional biopsy and the detection of linear IgG, IgA or C3 along the epithelial basement membrane zone (BMZ) using direct immunofluorescence (DIF) [3, 8]. In Ocular MMP (OcMMP) a clinical diagnosis can be made as 44.7–55% of cases are negative on direct immunofluorescence (DIF) [7–10]. The pathogenesis of MMP is a type 2 hypersensitivity reaction against components of the hemidesmosome-epithelial membrane complex such as laminin and integrin by the antibodies such as IgG and IgA [6].

The oral and ocular mucous membranes are the most commonly affected sites in MMP. Whilst the exact statistics vary across sources, it is estimated that 80–90% of cases have oral involvement and that 50–70% involve the ocular mucosa [1, 7, 11–13]. Other commonly affected sites include the larynx, pharynx, esophagus, trachea and genitals [5, 12]. Only a quarter of patients with MMP have skin involvement [1, 13]. Severe MMP, which has a high risk of complications, often affects the pharynx, larynx, esophagus, trachea, genitals and conjunctiva [1, 7]. End stage complications of high risk MMP include esophageal strictures, breathing difficulties, speech difficulties, airway obstruction and blindness [1, 7, 12, 14]. Any form of laryngeal involvement is deemed a sign of poor prognosis, with 10.5% of patients with laryngeal MMP going on to require a tracheostomy [13, 15].

Abbreviations

ABQOL	Autoimmune Bullous Disease Quality of Life Questionnaire
ABSIS	Autoimmune Bullous Skin Disorder Intensity Score
AIBD	Autoimmune blistering diseases
BMZ	Basement membrane zone
BPDAI	Bullous Pemphigoid Disease Activity Index
BSA	Body surface area
COMDQ	Chronic Oral Mucosal Disease Questionnaire
DIF	Direct immunofluorescence
DLQI	Dermatology Quality of Life Index
FDA	U.S Food and Drug Administration
FDM	Fornix depth measurer
ICC	Intraclass correlation coefficient
IGA	Investigator's Global Assessment
IOC	Intraoral camera
MMP	Mucous Membrane Pemphigoid
MMPDAI	Mucous Membrane Pemphigoid Disease Activity Index
OcMMP	Ocular MMP
ODSS	Oral Disease Severity Score
OHIP	Oral Health Impact Profile
PDAI	Pemphigus Disease Activity Index
RCT	Randomised controlled trial
TABQOL	Treatment of Autoimmune Bullous Disease Quality of Life Questionnaire
UAT	Upper aerodigestive tract

2. Investigators global assessment scoring systems

At this point in time there have been no randomized controlled trials (RCTs) for MMP, so treatment is based on physicians' personal experience's [1, 12, 13]. Whilst this is partially due to the rarity of the disease, it is also due to the lack of validated scoring systems. A validated clinical grading system enables a researcher to effectively document the effects of disease, communicate disease severity to peers and determine the response to treatment [16, 17]. The aim of this study is to develop and validate a 5-point Investigators Global Assessment (IGA) for MMP and to compare it to the Mucous Membrane Pemphigoid Disease Area Index (MMPDAI). As such this literature review will provide an overview of available scoring systems for MMP (see Table 1). We are proposing an IGA score because it is the preferred scoring tool of the United States Food and Drug Administration (FDA) [17, 18]. An IGA uses clinical characteristics to globally analyze disease activity on a numerical scale at a single time point [17, 18]. The development and validation of an IGA is particularly important now, as it will enable researchers to conduct RCTs testing targeted immunotherapy treatments that are only newly available [19]. Targeted immunotherapy is desirable for MMP due to both its lower risk of side effects and greater efficacy [19, 20]. Currently MMP treatment includes topical corticosteroids, dapsone, methotrexate and tetracyclines as first line [7]. Additionally, anti-CD20 treatment using rituximab, and IVIG have also shown promising results in MMP in small cohort studies [1, 7, 12, 13, 20, 21].

Table 1. Summary of general scoring tools for MMP.

MMP Score	Advantages	Disadvantages
Proposed IGA Score		
Proposed IGA Score	Preferred scoring system of FDA.	Simplistic representation of MMP severity.
	Can represent both activity and damage.	
General Scoring Tools		
MMPDAI	Recommended by international guidelines.	Recommended specific scales be used for individual mucous membranes.
	Validated in oral MMP.	Poor interrater reliability for damage scores in oral sections.
	Correlates with UAT involvement.	Not validated in areas other than the oral cavity.
	Clinical score correlates with circulating IgA and linear IgE deposits on BMZ.	
	Differentiates between activity and damage.	
ABSIS	Recommended by international guidelines.	Assesses only oral mucosa - Tauber Score used in conjunction to assess ocular pathology.
	Validated in oral MMP.	Activity and damage scores combined.
	Assesses both subjective and objective aspects of disease activity.	Requires BSA calculation and evaluation of lesion type.
Setterfield Score	Clinical score correlates with IgA and IgG deposition on the BMZ and BP180 reactivity.	Activity and damage scores combined.
	Heavily weights mucosal involvement.	Not validated.
		Subjective measurements.

Table 2. Summary of commonly used mucous membrane specific scores for MMP.

ODSS for OMMP	Recommended by international guidelines.	Not initially designed for MMP.
	Validated in MMP.	No damage component.
Higgins Score for Laryngeal MMP	Specific to larynx.	Activity and damage score are combined.
Nash Score for Laryngeal MMP	Specific to larynx.	Based off symptomology.
Foster Score for OcMMP	Simplistic scoring system for OcMMP.	Insensitive to inferior fornix loss and new symblepharon.
	Stage four correlates with poor visual acuity.	Overlooks upper fornix shortening.
		Only score scarring. No activity component.
Mondino Score for OcMMP	Simplistic scoring system for OcMMP.	Cannot represent new symblepharon in the absence of inferior fornix shortening.
	More objective than Foster Score.	Insensitive to early disease stage.
		Overlooks upper fornix shortening.
		Only scores damage. No activity component.
		Requires slit lamp.
Tauber Score for OcMMP	Combines variables of Foster and Mondino score.	Overlooks upper fornix shortening.
		Only scores damage. No activity component.
		Requires slit lamp.
Cicatrising Conjunctivitis Assessment Tool for OcMMP	Composed of inflammation, scarring and morbidity score.	Not created specifically for MMP.
	Each individual component of the score is validated.	Requires slit lamp.
	Complies with current recommendations to use reference photographs to quantify conjunctival hyperemia.	

3. General scoring systems

3.1. Setterfield score

One of the earliest global scoring systems used for MMP is the unnamed multisite MMP score proposed by Setterfield [5]. This score is still occasionally used due to its emphasis on oral involvement though it has not been validated [22]. The Setterfield score is applicable to all MMP types, including severe cases of MMP, and heavily weights high risk areas and mucous membranes (only 14% of the score is for skin sites) [2]. The score has also been found to correlate with IgA, IgG and BP180 reactivity; patients with both IgA and IgG on the BMZ DIF and/or low BP180 reactivity correlated with high clinical scores [5, 22]. Despite this the score by Setterfield [5] lacks a distinct separation of activity and damage scores, which means a high score could indicate either a patient with active disease or a patient with controlled disease but severe complications. The score also includes subjective measurements such as mild, moderate, and severe in the larynx column.

3.2. Mucous membrane pemphigoid disease area index

The MMPDAI is a specific scoring system developed by an international panel of dermatologists who specialise in AIBD and is based on the existing and validated PDAI and BPDAI scores [4]. Whilst not validated in its entirety, the score is recommended for use in MMP by the German S2k Guidelines and European S3 Guidelines [2, 7].

The weighting of the scalp (4%), mucous membranes (48%) and skin (48%) have been altered in comparison to the Pemphigus Disease Activity Index (PDAI) and Bullous Pemphigoid Disease Activity Index (BPDAI) (10% and 45% respectively) to better represent the distribution of MMP [4]. The score also features a damage column to quantify post inflammatory changes and scarring, which most commonly cause complications in MMP [4]. Other merits include the ability to score the eyes without an ophthalmologist, and good intra-rater reliability [4, 11, 12]. The authors of the MMPDAI proposed an addendum in 2015 to account for the lack of a descriptor for lesions that are stable but erythematous to indicate activity [4]. The oral component of the score has been validated [11].

The MMPDAI has shown promising correlation with the pathological picture of MMP. Corti [23] found that MMPDAI score was associated with circulating IgA and linear IgE deposits on the BMZ and Endo [14] found that a high oral disease activity MMPDAI score correlated with upper aerodigestive tract (UAT) involvement. A case report of a patient with an oral disease MMPDAI score of 44, who developed acute respiratory failure secondary to laryngopharyngeal stenosis, also confirms this pattern [21]. Whilst individual case reports are a weak form of evidence, they are useful in reinforcing the results of Endo [14]. These are promising results as they suggest that an increased MMPDAI score correlates with high risk MMP, and thus a patient's potential for life threatening laryngeal scarring and stenosis. Overall, the papers by Endo [14] and Corti [23] have similar limitations: small sample sizes and the retrospective application of the MMPDAI to patient notes. It is not specified if the raters were aware of the severity of patients' MMP or if there were any serious complications which may influence their rating. Specifically, Endo [14]'s work fails to examine the correlations of MMPDAI scores with UAT in asymptomatic lesions and Corti [23] is limited by the low sensitivity of indirect immunofluorescence. It is also not specified if the reported scores in any of the above studies are combined activity and damage scores, or only activity scores.

Whilst the MMPDAI is one of the only generalised disease specific tools that can be used by members of a multidisciplinary clinical trial team, it is only proposed for mild MMP [2, 4]. Unlike in the Setterfield score, it is recommended that specific scales be used to quantify the degree of ocular, oral, laryngeal, esophageal or pharyngeal mucosa involvement in higher risk cases of MMP as this cannot be measured in detail by the MMPDAI [4]. Corti [23] had to estimate a patient with only esophageal involvement's MMPDAI as 10 and Ormond [11] found the oral sections of MMPDAI had poor interrater reliability for damage scores (prior to the erythema addendum). The MMPDAI could also benefit from review by oral medicine physicians, otolaryngologists and ophthalmologists with specialist knowledge of the effects of MMP outside of the skin, which may enable the score to be applicable to more specific cases of MMP.

3.3. Autoimmune bullous skin disorder intensity score

The Autoimmune Bullous Skin Disorder Intensity Score created by Pfützte [24] is recommended by the European S3 guidelines for MMP [2, 24]. It is a score reflecting clinical disease activity with a specific section for the assessment of oral mucous membranes [7, 11, 24]. There is also a subjective component in oral assessment which assesses a patients discomfort when eating and drinking [11, 24]. The types of foods in the subjective component can be swapped with foods that

are preferred regionally but are texturally equivalent to account for differences in patients' diets [24]. The ABSIS has been validated for oral MMP and found to have good interobserver reliability in oral MMP. This was better than the MMPDAI activity score which only showed satisfactory correlation, but not as good as the ODSS [11]. This score also uses the rule of nines to calculate body surface area (BSA) affected. Whilst the rule of nines is a quick tool to estimate BSA it often overestimates the area affected [25]. Rosenbach [26] found the ABSIS poorly represents small changes in disease activity at the low end of the disease spectrum in pemphigus. Although this study examined patients with pemphigus, a similar AIBD, it did not include patients with MMP. Therefore examining the validity and reliability of the ABSIS in MMP would be useful. A flaw of using the ABSIS for MMP is that it lacks a score for ocular involvement, which the authors propose can be combatted by using the Tauber score in conjunction [24, 27]. Despite this the ABSIS is not disease specific and has not been validated in multisite MMP [2].

4. Mucosal site-specific scoring systems

The mucosal sites are uniquely difficult to score and photograph. In MMP they are also higher risk areas, so accurate photography and validated scoring systems are vital [1, 7]. Visualising the conjunctiva is particularly challenging due to its three dimensional nature in which multiple portions are concealed from view [28]. In OcMMP eversion of the lid is often required to visualise fornix shortening and symblepharon, which can cause the tarsus to buckle and thus make assessment of the fornix inaccurate [29]. To aid in the objective measurement of fornix shortening fornix depth measurers (FDMs) such as those designed by Kawakita [30] and Williams [31] can be used. The FDM created by Williams [31] is superior to Kawakita [30] as its curved shape allows for the full extent of the upper fornix to be measured. A patient's measured fornix depth is then compared to age and race specific values to calculate an objective percentage shortening. To this date FDMs have only been validated in Caucasian and south Asian populations and thus there is a need for large scale population-based studies to validate them for the broader population [27, 29]. All ocular scoring systems discussed require a slit lamp, which is costly and may not be available outside of an ophthalmology or optometry practice.

Scoring and imaging of the intraoral area, esophagus and larynx can be aided by using pen cameras, intraoral cameras (IOCs) and endoscope type cameras. Pentapati and Siddiq [32] identified in a systematic review that IOCs can be used to for imaging oral mucosal conditions. Bradley [33] demonstrated the feasibility of using IOCs for diagnosis of oral conditions in a small pilot study on the diagnosis of dental patients through intraoral photographs in regional Northern Ireland. Despite this the Irish study was significantly limited due to its small sample size and lack of comparison to a control group [33]. The study also did not feature any MMP cases.

4.1. Oral

The oral disease severity score (ODSS) is a score used for chronic oral diseases that has been validated in MMP [11]. The ODSS is recommended for use in mono-site and predominantly oral MMP by both the European S3 Guidelines

and the German S2k guidelines for MMP [2, 7]. In the ODSS, which has previously been used in lichen planus and pemphigus vulgaris, 17 areas of the oral cavity are given a site score, area score and pain score which are then added to give a maximum score of 106 [11, 12]. This is more detailed compared to the division of the oral cavity into 7 areas in the MMPDAI and 11 in the ABSIS [4, 11].

Potential weaknesses of the ODSS include the combination of the subjective pain score and objective site and activity scores, which the authors of the ODSS feel provides a more comprehensive representation of disease experience [11]. This subjective component also does not represent a quality-of-life measure [11]. The score lacks a damage component, which despite the decreased propensity of the oral area for scarring is still concerning as most complications of MMP are linked to post inflammatory changes and scarring [1].

The ODSS was validated in a single point-of-time study of 15 patients with mild to moderate MMP, during which ten physicians scored each patient, and two physicians rescored their patients to test intra-rater reliability [11]. It was found the ODSS had good convergent validity with the ABSIS and MMPDAI and an intra-rater reliability intraclass coefficient (ICC) of 0.97 and 0.93. The study made many efforts to reduce bias, such as having a two hour wait time before patients were rescored and using specialists from multiple fields of medicine such as dermatologists and oral health specialists. Despite this, limitations include that none of the clinicians regularly used the ABSIS or MMPDAI, and only half were experienced using the ODSS.

4.2. Laryngeal

Methods of scoring laryngeal MMP have been proposed by both Nash [34] and Higgins [15] [2, 15, 34]. The Higgins scoring system is scored based on lesion type, site and airway patency, combining activity and damage stages [15]. In comparison the Nash scoring system is based on symptomology, due to the authors concerns that it would be difficult to routinely assess the larynx [34]. Whilst both scores exist neither have been validated and it is instead recommended that an otolaryngological version of the MMPDAI be used for clinical studies [2]. In pemphigus vulgaris a score combining number of lesions and lesion size has been used, which was shown to correlate with the PDAI, ABSIS and endoscopic examination [35].

4.3. Ocular

The Foster scoring system, followed by the Mondino and adapted Tauber tool are the most common scoring tools used in ocular MMP [8, 11]. Foster's score divides OcMMP into four stages based on general signs of cicatrisation whilst Mondino's scoring system only uses inferior fornix shortening [16, 36]. Whilst this score is more objective than the Foster score, it lacks sensitivity to early disease stages [37]. The Foster score is insensitive to inferior fornix loss or increases in symblepharons and the Mondino score fails to represent new symblepharons in the absence of fornix loss [37].

The Tauber scoring system was created to combat insensitivities in the Foster and Mondino scores [27]. This score follows the Foster stages but incorporates a sub score for inferior fornix shortening and horizontal symblepharon

involvement [37]. The Tauber score was validated in a retrospective cohort study of 125 eyes against the Foster and Mondino scores. All scores showed excellent interrater reliability, with the Tauber score best representing if MMP had advanced [37]. A systematic review and meta-analysis by Bocanegra-Oyola [8] recommend the Tauber system as it combines the variables in the Foster and Mondino scores, making it significantly more accurate.

The Foster, Mondino and Tauber tools can all be used retrospectively on clinical records due to their simplicity [29, 38]. This is particularly advantageous in rare disease such as MMP where most studies are retrospective. Stage IV of the Foster score has also been found to have a strong correlation with poor visual acuity (>6/60), whilst the Mondino score has been shown to have a poor correlation [16].

In a literature review of all available scoring systems for cicatricial conjunctivitis Ong [29] noted that the Foster, Mondino and Tauber tools overlook upper fornix scarring, which is also associated with complications threatening sight, and are largely qualitative measures based on a physicians clinical judgement. Other weaknesses include that the Foster, Tauber and Mondino scores only assess scarring, making them insensitive in detecting disease activity, and that approximation of inferior fornix shortening requires visualisation of the posterior lid surface [28, 38]. These scores also require lid traction, which can cause distortion of the lid which alters final scoring [28]. Despite this useful correlation, visual acuity is a poor measure of OcMMP progression as it cannot be assumed to be due to OcMMP as can instead be caused by a patients pre-existing comorbidities [38].

International guidelines recommend the validated cicatrising conjunctivitis assessment tool (CCAT) be used for ocular MMP [2, 7]. The CCAT validated by Ong [38] is composed of an inflammation, scarring and morbidity score. Unlike other scores discussed above the score assesses both the upper and lower fornix [38]. Similarly to the MMPDAI, the score divides the bulbar conjunctiva into four quadrants to better approximate conjunctival inflammation. All components of the assessment tool were individually validated, and only kept in the score if adequate agreement was shown [29]. The scarring and morbidity components, which are designed to represent the irreversible effects of the disease, had excellent intra-rater and interrater correlation [38]. The percentage of lower fornix shortening is also calculated using a FDM and age standardised fornix depth tables to provide a much more objective result than simply visualising fornix loss [27, 29]. Despite these strengths the tool still requires specialist equipment such as slight lamps and FDMs and only provides fornix depth tables for Caucasian populations. Since the CCAT validated in a mainly Caucasian population, further validation studies are required for diverse populations

In the CCAT, reference images are used to aid in scoring of conjunctival hyperemia, which is in line with current expert recommendations [6, 7, 38]. The creation of a visual scale to document hyperemia was first proposed Elder and Bernauer [16] in criticism of the Foster and Mondino scores. The detection of active inflammation is crucial in OcMMP as this allows the disease to be treated before vision threatening conjunctival scarring develops [29]. Despite this, Saw [39] showed progression of cicatrification wasn't always associated with less severe inflammation. It is thought these results occur because persistent mild inflammation in a patient with treatment resistant disease may cause cicatrification to progress [39].

Multiple other scoring systems have been proposed for OcMMP such as the score by Reeves [28] and Rowsey [40] but these have been excluded from this review due to infrequent use [28, 29, 40].

5. Quality of life measures

Whilst this report focusses on clinician and investigator reported scoring systems it is worthwhile to mention that there are no MMP specific patient scored quality-of-life measures [7]. The European S3 Guidelines on MMP recommend the use of the autoimmune bullous disease quality of life (ABQOL) questionnaire and treatment of autoimmune bullous disease (TABQOL) questionnaire as they are specific to AIBD and thus better encapsulate the impact of MMP [2, 41]. Versions of the ABQOL and TABQOL have been validated in North American, Polish, Japanese, Greek, Persian, Turkish, Arabic, Chinese, French, Malaysian and English populations with a diverse range of AIBD [42–52]. Both scores correlate highly with each other, the dermatology quality of life index (DLQI) and skindex-29 and weakly with the PDAI, BPDAI and MMPDAI [50]. The poorest correlation was in MMP due to the lack of specific questions about mucosal sites [50]. As such it has been suggested the ABQOL and TABQOL show no advantage over generic quality of life measures such as the DLQI or skindex-29 in diseases involving the mucous membranes [50]. Site specific scoring systems such as the chronic oral mucosal disease questionnaire (COMDQ), which is validated in English and Persian populations, and the oral health impact profile (OHIP) are also recommended [2, 41, 53, 54].

6. Conclusion

A general scoring system for MMP should represent all mucous membranes and areas affected in a way which represents their distribution in this disease specifically. As such the score should also be reviewed by specialists of multiple medical disciplines that treat MMP. The score should also be capable of quantifying activity as well as damage separately [27, 29]. These could be incorporated into one IGA score or divided into two IGA scores, to better represent the impact of blistering disease scarring, as has been done before in other AIBD scores such as the MMPDAI or EBDASI [55]. The activity score would be used to quantify the efficacy of treatment at suppressing the disease process and the damage score used to quantify the effects of post inflammatory changes. The damage score may even be used to investigate novel treatments for scar reduction. Due to the rarity of the disease an ideal score should also be easy to apply to retrospective photos of patients with MMP if required. Currently these authors, and a wider research group comprising specialists treating different forms of MMP, are in the process of developing and validating an MMP specific IGA score. This score aims to appropriately represent activity and damage and be applicable to all forms of MMP.

Additional information

Conflict of interest

The authors have declared that no competing interests exist.

Ethical statements

The authors declared that no clinical trials were used in the present study.

The authors declared that no experiments on humans or human tissues were performed for the present study.

Informed consent from the humans, donors or donors' representatives: Bellberry Limited.

The authors declared that no experiments on animals were performed for the present study.

The authors declared that no commercially available immortalised human and animal cell lines were used in the present study.

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Author contributions

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Data availability

All of the data that support the findings of this study are available in the main text.

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