

# The nursing assessment of pemphigus vulgaris ulcers

## ABSTRACT

**Introduction** Pemphigus vulgaris (PV) is a severe autoimmune bullous dermatosis resulting in the formation of intraepidermal blisters affecting the skin and mucous membranes. Epidemiologic data shows an incidence of 0.1–0.5 per 100,000 inhabitants per year, and mortality at almost 5–10%.

**Objective** The objective of this integrative literature review was to examine the classification/terminology of PV ulcers according to the description of skin lesions.

**Method** This is an integrative review of primary studies, series/clinical reviews or validation studies that describe or evaluate PV ulcers. Search strategies included relevant papers that were published between 2011–2019 using terms such as pemphigus, skin ulcer, dermatology, diagnostic, nursing assessment. The studies were selected for analysis after application of eligibility criteria and exclusion by duplicity.

**Results** The initial search resulted in 2,934 publications; 14 articles were eligible for analysis. The synthesis of the studies was organised as follows – 57.14% series/clinical reviews, 50% written by physicians, 64.29% level of evidence 4. The terminology used to describe PV ulcers included skin/mucosal erythema, new erythema, post-inflammatory hyperpigmentation, oral lesions, epithelialisation scabs, blisters, bullae, erosions, eroded areas, erosive exudative lesions, dry erosive.

**Conclusions** Studies with better levels of evidence are needed on this issue in order to determine the best way to describe the lesions using the dermatological glossary for nursing assessment.

**Keywords** nursing, dermatology, skin diseases, vesiculobullous, acantholysis

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## INTRODUCTION

Pemphigus vulgaris (PV) is a severe autoimmune bullous dermatosis in which the antibodies destroy the desmosomes, resulting in the formation of intraepidermal blisters that affect the skin and mucous membranes. PV occurs mainly between the 4th and 6th decades of life, affecting males and females with an incidence of 0.1–0.5 / 100,000 inhabitants / year and with a mortality rate of 5–10%. The disease distribution is universal, but most commonly affects people of Jewish ancestry<sup>3</sup>.

In PV, autoantibodies act predominantly on desmoglein 3 (Dsg3) which is expressed predominantly in the deeper layers of the epidermis<sup>2,3</sup>. Identifying the layer on which acantholysis occurs is a factor that assists in the diagnosis of bullous dermatoses. For example, it is possible to differentiate PV from pemphigus foliaceus by the site where acantholysis occurs, since pemphigus foliaceus affects the granular layer whereas PV affects the spinous layer<sup>2,3</sup>. These manifestations involve the formation of blisters with consequent ulceration and skin damage which can be devastating, affecting social interaction and even loss of employment<sup>4</sup>.

The impact of disfigurement associated with PV on patients' quality of life, self-image, family and social dynamics has been well documented<sup>4</sup>. In addition to cutaneous involvement, PV may involve mucosal tissue in the mouth, pharynx, larynx, nasal passage and ear canals (Figures 1 & 2). Areas affected by the disease can compromise normal breathing as well as the ability to speak and to eat to maintain adequate nutritional intake<sup>2,3</sup>.

The classification of PV has been the object of studies in recent years. The Commitment Index of Skin and Mucous in Pemphigus Vulgaris uses four different parameters to score the disease clinical status: (a) the number of blisters or eroded areas; (b) the size of blisters or eroded areas; (c) evidence of the Nikolsky sign (where sliding the finger firmly with pressure over the skin separates normal-appearing epidermis, producing an erosion); and (d) mucosal involvement and sepsis. The total score may vary from 0–100, and the patients are classified as follows: Class I score 0–30; Class II 35–65; Class III 70–100, meaning that the higher the score, the more critical the status<sup>1</sup>.

Pemphigus classifications tools, the Pemphigus Disease Area Index (PDAI) and the Japanese Pemphigus Disease Severity Score (JPDSS), were compared by Shimizu et al.<sup>5</sup>. PDAI measures skin and mucosal involvement by size and number of blisters in each anatomical region, and the score ranges from 0–263. JPDSS uses parameters for scoring: (I) the ratio of affected area of skin to the body's surface area; (II) the presence or absence of Nikolsky's sign phenomenon; (III) the number of newly developed blisters per day; (IV) the presence

or absence of oral lesions; and (V) the titer of pemphigus antibodies. Each parameter has a score ranging 0–3. In Shimizu et al.'s study<sup>5</sup>, the results show that PDAI more accurately reflects disease severity. The authors therefore propose the use of indexes to guide a uniform treatment according to grading criteria. Corticotherapy is the treatment of choice, and it can be associated with immunosuppressants if there is no improvement with isolated corticotherapy<sup>2,3</sup>.

Although considered a relatively rare disease, there is a need for nurses to recognise skin lesions associated with PV and communicate appropriate findings in order to help patients seek early treatment, evaluate disease progression, and monitor responses to treatment<sup>6</sup>. The purpose of this integrative review was to describe the taxonomy for the description and assessment of skin changes related to PV by nurses.

## METHOD

This integrative review was conducted to identify, analyse and synthesise studies that use qualitative, quantitative and mixed methods in this theme<sup>7</sup>. We have chosen the method described by Mendes<sup>7</sup> to guide the review which consisted of six stages: (1) formulation of the guiding question; (2) establishment of criteria for inclusion and exclusion of studies and data collection (search in the literature); (3) categorisation of studies; (4) evaluation of studies included in the review; (5) analysis and interpretation of data; and (6) synthesis of the knowledge evidenced in the articles analysed (presentation of the results)<sup>7</sup>.

### Formulation of the guiding question

The research question was – How are the lesions that characterise PV described in the literature in its definition and classification?

### Establishment of criteria for inclusion and exclusion of studies and data collection

The medical and nursing literature was searched from 2011–2019 in conjunction with a librarian to assist in answering the



Figure 1. Cutaneous lesions on trunk



Figure 2. Oral mucosa lesions

research question. Searches included Web of Science, LILACS, EMBASE, SCOPUS, PUBMED, BVS, CINAHL and COCHRANE with specific inclusion and exclusion criteria. To identify relevant publications, databases were searched using the following key terms – dermatology, pemphigus, skin ulcer, diagnostic and nursing assessment. The inclusion criteria included articles published in English, Spanish and Portuguese, peer-reviewed literature and consensus documents; the dates of publication were from 1 January 2011 to 31 December 2019. Commentary and editorials were excluded.

### Categorisation of studies

Selected studies were then categorised according to the six levels of evidence<sup>8</sup>:

- Level 1: evidence from meta-analysis of multiple controlled and randomised studies.
- Level 2: evidence from individual studies with experimental design.
- Level 3: evidence from quasi-experimental studies, time series or case-controls.
- Level 4: evidence from descriptive studies (non-experimental or qualitative approach).
- Level 5: evidence of case / experience reports.
- Level 6: evidence based on expert committee opinions, including interpretations of non-research based information, regulatory or legal opinions.

### Evaluation of studies included in the review

This step included the evaluation of the studies as well as data extraction. A standardised data collection form was used to extract the following information: authors; professional category of authors; title of the article; journal; year of publication; level of evidence; goals; methodological design; sampling detail; synthesis of information; evaluation/assessment of skin ulcers in pemphigus; methodology used to validate the instrument; description of the instrument; terminology used to characterise ulcers; and results and conclusions.

### Analysis and interpretation of data

The data evaluation stage included evaluating the quality of the primary sources using a specific methodological approach to determine the quality of the source. The data were evaluated and coded according to two criteria – the methodological rigour and the relevance to the topic of skin assessment. Studies were analysed and the rigour rated on a score from 0–4. The relevance to the topic was also scored and indicated, with 1 having no relevance to the topic and 2 indicating the article was relevant.

### Synthesis / presentation of results

The data analysis for the qualitative studies were reviewed and were systematically categorised, analysed and synthesised, and placed into distinct themes, patterns and relationships using a matrix method. The synthesis of the studies was organised into three axis: (1) characteristics of scientific publications on PV; (2)

terminology used to describe cutaneous lesions related to PV (Table 1); and (3) comparison of terminologies used in studies and dermatological description of skin lesions.

### Ethical and legal aspects

The research project was submitted and approved by the Research Ethics Committee / UNIFESP: 0450/2015 and followed the ethical and legal precepts of research with human beings according to Resolution 196/96 of the National Health Council.

## RESULTS

The initial search identified 2934 articles; 356 articles were included from review of the titles. After review of the abstracts, another 258 papers were not suitable for review since they did not meet the inclusion criteria; specifically, the excluded papers did not address PV and lesion assessment. A total of 58 articles were then selected for the study after exclusion of 40 duplicates. Of the 58 articles, 37 (63.79%) were primary studies, 17 (29.31%) were series/clinical reviews and four (6.90%) were validation studies.

A variety of terms were used in the literature to describe PV-related lesions. These are summarised below.

### Primary skin lesions

#### *Flat spots or maculae*

The first is flat skin lesions, including colour changes and blood-vascular stains. Nomenclature used were new erythema, erythema, minimal erythema, and marked erythema<sup>9–15</sup>. Erythema is defined as a red colour resulting from vasodilation that disappears by digital pressure or diascopy. Diascopy is a refinement in which a piece of clear glass or plastic is pressed against the skin while the observer looks directly at the lesion under pressure<sup>15,16</sup>.

#### *Solid formations / oedematous elevations*

Solid formations may include bullae and papulae (small superficial solid elevations of the skin) while oedematous elevations could be cutaneous lesions, scales or pustulae (defined elevation of the skin containing purulent fluid). These clinical features might be associated with a positive Nikolsky sign<sup>17</sup>.

#### *Pigment spots*

Nomenclature used in this section were post-inflammatory hyperpigmentation and hyperpigmented macules<sup>5,11,16, 18–20</sup>. Hyperchromia is defined as a spot of variable colour, caused by the increase of melanin or deposit of another pigment. The increase in melanin / melanodermic spots has a variable colour from light brown to dark bluish or black<sup>16,18</sup>.

#### *Liquid content*

Nomenclature used here were vesicles, blisters and bullous lesions<sup>1,5,9,10,12,13,20–55</sup>. Vesicles are defined as of circumscribed elevation, containing clear liquid, up to 1cm in size. The fluid, which is primitively clear (serous), may become cloudy (purulent) or red (haemorrhagic)<sup>16,18</sup>. A blister is defined as elevation containing clear liquid, greater than 1cm in size. The

fluid, which is primitively clear, may become yellowish-red or reddish, forming a purulent or haemorrhagic blister<sup>16,18</sup>. The term bullous lesions is used to refer to any liquid collections.

## Secondary skin lesions

### Thickness changes

Nomenclature used here are epithelial lesions, denuded epidermis and scar<sup>9–11,13,19,22,35,51,54,56–58</sup>. A scar is defined as a flat, salient or depressed lesion, without grooves, pores and hairs, movable, adherent or retractable. It associates atrophy with fibrosis and dyschromia. It results from the repair of destructive process of the skin. It can be: atrophic (thin, pleated, papyraceous); pitted (small holes appear); or hypertrophic (nodular, elevated, vascular, with excessive fibrous proliferation, with a tendency to regress)<sup>16,18</sup>.

### Tissue losses

Nomenclature used are erosions, eroded areas, erosive lesions, crust, ulceration, ulcer, ulcerative lesions, raw erosions and excoriations<sup>1,5,9–15,19–21,23–60</sup>. Erosion or exulceration is defined as superficial loss that affects only the epidermis. Crust is defined as a concretion of light yellow to greenish or dark red colour which forms in an area of tissue loss. It results from the desiccation of serosity (meliceric), pus (purulent) or blood (haemorrhagic), mixed with epithelial remains<sup>16,18</sup>.

Therefore, in view of the integrative review, it was possible to identify studies describing the dermatological alterations of PV ulcers according to criteria established by dermatological

glossaries. In order to illustrate the correlation of the identified terms and their corresponding study, the third analysis correlates the terms identified with the year, author category, study category and level of evidence, shown in Table 2.

## DISCUSSION

As evidenced by the results, studies in this area are scant and nurses have had little contribution. However, this is needed to highlight the consistency demonstrated between the terms used by authors and the terminology of elementary lesions.

To promote excellence in dermatological care it is necessary for nurses and health professionals to develop skills for the evaluation of cutaneous manifestations, especially skin lesions. The skin, as the largest organ of the human body, has protection as one of its numerous functions. Far beyond essentially dermatological affections, it can translate the person's general condition, signalling the presence of systemic disorders and sometimes presenting manifestations that are recognised as cutaneous markers of certain pathologies. However, the paucity of studies in PV has led to poor description and evaluation of skin lesions in PV. This may result in delayed diagnosis and suboptimal care and treatment<sup>6,20,29,36</sup>.

In fact, this subject points to the need for further investigation to compose a framework of knowledge based on scientific evidence in this area, so that dermatology nurses may develop an accurate consideration of the assessment of skin manifestations, especially skin ulcers. These professionals

Table 1. Terms identified grouped by level of evidence of the studies

Level of evidence	Terms used to describe lesions	
	Cutaneous	Oral mucosa
1	None	None
2	Erythema, new erythema, post-inflammatory hyperpigmentation, vesicles, blisters, erosions, erosive exudative, lesions, dry erosive lesions, eroded areas, crust, epithelial lesions	Oral lesions, ulceration, blisters, erosions
3	Blisters, erosion	–
4	Skin erosions, erythematous base, post-inflammatory hyperpigmentation, plaque, flictena, flaccid vesicles that easily break, vesicles, blisters, flaccid blisters, small, tense and ephemeral blisters, bullous lesions, lesions of the type blister, vesico-bullous lesions, pustule, ulcer, erosions, painful erosions, erosive lesions, dry erosive lesions, eroded and exudative ulcerated lesions, exulcerated lesions, reepithelialised lesions, scar, epithelial lesions	Vesicles, flaccid vesicles, flaccid vesicles that easily break, erosions, painful erosions, blisters, flaccid blisters
5	Blister-like injuries, vesico-bullous injuries, exulcerated injuries, crusts, erosive injuries, blisters, vesicles, excoriations, erythematous bullae, dry crusted erosions, flacid bullae, pink eroded friable crusted plaque, erythema, flacid blisters, denudation, erosive denuded epidermis, scattered flaccid and clear fluid-filled bullae, clustered flat-topped papules, scaly plaques, raw erosions, erosions covered by h(a)ematic crusts, postbullous erosions, hyperpigmented macules, scabby erythema, coalesced thick lesions, coarsegranular scabby and seepage purulent and h(a)emorhagic scars	Vesico-bullous injuries, ulcers, blisters, vesicles, excoriations, erosions, crusted erosions, enanthema, erosions covered with a whitish exudate
6	Vesicles that easily break, painful erosions, refractory erosions, contiguous erosions, erosive skin lesions, blisters, flaccid blisters	Ulcers, blisters and erosions

Images from the author's collection

Table 2. Terms identified in selected articles

Title / year	Category of authors	Study category	Level of evidence	Nomenclature	
				Skin	Oral mucosa
Clinical course of pemphigus in Thai patients (2011) <sup>9</sup>	Physicians	Primary	4	Vesicle, blister, erythematous base, healthy skin base, plaque, erosion, ulcer, pustule, scar	NI
Diagnosis and clinical features of pemphigus vulgaris (2011) <sup>28</sup>	Physicians	Series/ clinical reviews	4	Flaccid vesicles, painful erosions	Erosions
Validation of the cutaneomucous involvement index of pemphigus vulgaris (2011) <sup>1</sup>	Physicians	Validation	2	Blisters, eroded areas	Lesions
Nursing care evolution to the client with pemphigus: integrative literature review (2011) <sup>61</sup>	Nurses	Series/ clinical reviews	4	Bullous lesions, eroded painful lesions	NI
Severity score indexes for blistering diseases (2012) <sup>10</sup>	Physicians	Series/ clinical reviews	4	Exudative erosive lesions, dry erosive lesions, epithelial lesions, blisters, erosions, vesicles, new erythema	NI
Diagnosis and treatment of pemphigus (2012) <sup>21</sup>	Physicians	Series/ clinical reviews	4	Flaccid blisters, erosions, flaccid vesicles	Flaccid vesicles, erosions
Clinical and histological characterisation of oral pemphigus lesions in patients with skin diseases (2013) <sup>23</sup>	Physicians, dentist	Primary	4	NI	Erosions, ulcers, vesicles
Pemphigus: use of the Japanese severity index (2013) <sup>22</sup>	Unidentified	Validation	2	Blisters	Oral lesions
Nursing diagnoses in pemphigus vulgaris: a case study (2013) <sup>27</sup>	Nurses	Series/ clinical reviews	5	Blister-like lesions, vesico-bullous lesions, exulcerated lesions, crusts, eroded areas, erosive lesions	Vesico-bullous lesions, ulcers
Evidences related to the care of people with pemphigus vulgaris: a challenge to nursing (2013) <sup>29</sup>	Nurses	Series/ clinical reviews	4	Small, tense and ephemeral blisters, erosive and exudative ulcerated lesions	Lesions
Grading criteria for disease severity by Pemphigus Disease Area Index (2014) <sup>5</sup>	Unidentified	Primary	4	Erosions, flictena, new erythema, postinflammatory hyperpigmentation, blisters	Oral lesions
Diagnosis and classification of pemphigus and bullous pemphigoid (2014) <sup>26</sup>	Physicians	Series/ clinical reviews	4	Flaccid vesicles that break easily, painful erosions	
Japanese guidelines for the management of pemphigus (2014) <sup>24</sup>	Physicians	Series/ clinical reviews	6	Refractory painful eruptions, easily rupturing vesicles, painful erosions, contiguous erosions	Ulcers
Pemphigus disease activity measurements (2014) <sup>11</sup>	Unidentified	Validation	2	Exudative erosive lesions, dry erosive lesions, epithelial lesions, blisters, erosions, vesicles, new erythema, postinflammatory hyperpigmentation, erythema, crust, erosion	NI



Title / year	Category of authors	Study category	Level of evidence	Nomenclature	
				Skin	Oral mucosa
Familial pemphigus vulgaris occurred in a father and son as the first confirmed cases (2016) <sup>30</sup>	Physicians	Primary	5	Blisters	NI
Immune response in pemphigus and beyond: progresses and emerging concepts (2016) <sup>31</sup>	Physicians	Primary	5	Fluid-filled bullae, erosions and crusting	Erosions
Long-lasting localised pemphigus vulgaris without detectable serum autoantibodies against desmoglein 3 and desmoglein 1 (2016) <sup>12</sup>	Physicians	Primary	5	Erythema, blisters and erosions	Erosions
Mechanisms of disease: pemphigus and bullous pemphigoid (2016) <sup>32</sup>	Physicians	Series/ clinical reviews	5	Flaccid blisters, erosions.	Crusted erosions
Pemphigus vulgaris and amyotrophic lateral sclerosis (2016) <sup>33</sup>	Physicians	Primary	5	Flaccid blisters, erosions	Erosions
Pemphigus: pathogenesis to treatment (2016) <sup>34</sup>	Physicians	Series/ clinical reviews	5	Erosions, blisters	Erosions
Pregnancy-associated 'cutaneous type' pemphigus vulgaris (2016) <sup>35</sup>	Physicians	Primary	5	Crusted erosions and blisters, denuded epidermis, scattered flaccid and clear fluid-filled bullae	NI
The management of pemphigus ulcers: a challenge and learning opportunity for Brazilian nurses (2016) <sup>36</sup>	Nurses, physician	Primary	5	Vesicles, crusted lesions, blisters, ulcerative lesions, confluent strained blisters	Crusted lesions, ulcerative lesions
Analysis of serum cytokine profile in pemphigus (2017) <sup>37</sup>	Physicians	Primary	2	Blisters, erosions	Blisters, erosions
British Association of Dermatologists' guidelines for the management of pemphigus vulgaris 2017 (2017) <sup>38</sup>	Physicians	Series/ clinical reviews	6	Erosions or blisters	Erosions or blisters
Dew drops on spider web appearance: a newly named pattern of IgG4 deposition in pemphigus with direct immunofluorescence (2017) <sup>39</sup>	Physicians	Primary	4	NI	Erosions, flaccid blisters, vesicles
Increased miR-424-5p expression in peripheral blood mononuclear cells from patients with pemphigus (2017) <sup>40</sup>	Unidentified	Primary	3	Blisters	NI
Ocular involvement in pemphigus vulgaris – a retrospective study of a large Spanish cohort (2017) <sup>39</sup>	Physicians	Primary	5	Erosions	Erosions
Pemphigus vulgaris and pemphigus foliaceus: differences in epidemiology and mortality (2017) <sup>41</sup>	Physicians	Series/ clinical reviews	5	Blisters	Erosions
Pemphigus vulgaris persistently localised to the nose with local and systemic response to topical steroids (2017) <sup>62</sup>	Physicians	Primary	5	Erosive, crusted plaque	NI
Reviewing putative industrial triggering in pemphigus: cluster of pemphigus in the area near the wastewater treatment plant (2017) <sup>63</sup>	Physicians	Primary	5	Crusted erosions	NI
Dermatoscopy in diagnosis of cutaneous myiasis arising in pemphigus vulgaris lesions (2017) <sup>42</sup>	Physicians	Primary	5	Flaccid fluid-filled blisters, bullae, raw erosions	Painful oral erosions
Gastrointestinal cytomegalovirus disease in a patient with pemphigus vulgaris treated with corticosteroid and mycophenolate mofetil (2017) <sup>64</sup>	Physicians	Primary	5	Erosions covered by haematic crusts	Ulcer, erosions covered with a whitish exudate

Title / year	Category of authors	Study category	Level of evidence	Nomenclature	
				Skin	Oral mucosa
Pemphigus vulgaris induced by 5-aminolaevulinic acid-based photodynamic therapy (2017) <sup>13</sup>	Physicians	Primary	5	Scabby erythema, blisters, coalesced thick lesions, coarse granular, scabby and seepage	NI
A case of acute pemphigus vulgaris relapses associated with cocaine use and review of the literature (2018) <sup>43</sup>	Physicians	Primary	5	Blisters, vesicles, excoriations	Blisters, vesicles, excoriations
A case of pemphigus vulgaris associated with ulcerative colitis (2018) <sup>44</sup>	Physicians	Primary	5	Erythematous bullae, crusts	NI
An Oral Disease Severity Score validated for use in oral pemphigus vulgaris (2018) <sup>14</sup>	Physicians, dentist	Validation	2	NI	Minimal erythema, marked erythema, ulceration
Clinical and immunological features of pemphigus relapse (2018) <sup>45</sup>	Physicians	Primary	4	Erosions or blisters	Erosions or blisters
Diagnosis and management of pemphigus: recommendations of an international panel of experts (2018) <sup>46</sup>	Physicians	Series/clinical reviews	6	Blisters and erosions	Blisters and erosions
Mycobacterium tuberculosis and pemphigus vulgaris (2018) <sup>65</sup>	Physicians	Primary	5	Erosions	NI
Pemphigus vulgaris as the first manifestation of multiple myeloma: a case report (2018) <sup>47</sup>	Physicians	Primary	6	Bullous, erosive skin lesions	NI
Persistent B lymphocyte depletion after an ultralow dose of rituximab for pemphigus vulgaris (2018) <sup>66</sup>	Physicians	Primary	5	NI	Erosive lesions
Polycyclic annular presentation of pemphigus vulgaris with an eosinophil predominance in two pregnant patients (2018) <sup>48</sup>	Physicians	Primary	5	Vesicobullae, blisters, bullae, crusted lesions	Erosions
The significance of scalp involvement in pemphigus: a literature review (2018) <sup>56</sup>	Physicians	Series/clinical reviews	5	Erosions and crusted or scaly plaques	NI
Italian guidelines in pemphigus – adapted from the European Dermatology Forum (2018) <sup>49</sup>	Unidentified	Series/clinical reviews	5	Flaccid bullae with clear content, postbullous erosions	Erosions
Manejo estomatológico y sistémico de pénfigo vulgar: reporte de un caso (2018) <sup>20</sup>	Physicians, dentist	Primary	5	Bullous lesions, hyperpigmented macules	Ulcers
Cutaneous bacteriological profile in patients with pemphigus (2018) <sup>67</sup>	Physicians	Primary	3	Erosion	NI
Anterior scleritis in a patient of pemphigus vulgaris while on immunosuppressive treatment (2019) <sup>68</sup>	Physicians	Primary	5	Dry, crusted erosions	NI
Cerebral toxoplasmosis after rituximab for pemphigus vulgaris (2019) <sup>60</sup>	Physicians	Primary	5	NI	Erosions
Consensus on the treatment of autoimmune bullous dermatoses: pemphigus vulgaris and pemphigus foliaceus – Brazilian Society of Dermatology (2019) <sup>9</sup>	Physicians	Series/clinical reviews	6	Vesicles and flaccid blisters	Ulcers or eroded lesions
Koebner phenomenon: pemphigus vulgaris following Mohs micrographic surgery (2019) <sup>51</sup>	Physicians	Primary	5	Pink, eroded, friable, crusted plaque, red plaque with areas of erosion, flaccid bullae	NI
Pemphigus (2019) <sup>57</sup>	Physicians	Series/clinical reviews	5	Erosions and crusts, denudation	Erosions

Title / year	Category of authors	Study category	Level of evidence	Nomenclature	
				Skin	Oral mucosa
Pemphigus vulgaris possibly associated with application of a tissue expander in a patient with Crohn's disease and primary sclerosing cholangitis (2019) <sup>52</sup>	Physicians	Primary	5	Blisters, annular bullous lesions	NI
Recurrence of pemphigus vulgaris under nivolumab therapy (2019) <sup>53</sup>	Physicians	Primary	5	Erosions, blisters	Erosions
Refractory mucositis associated with underlying follicular dendritic cell sarcoma of the thymus: paraneoplastic pemphigus versus malignancy-exacerbated pemphigus vulgaris (2019) <sup>56</sup>	Physicians	Primary	5	Clustered flat-topped papules	Erosions
Scalp pemphigus vulgaris mimicking folliculitis decalvans: a case report (2019) <sup>54</sup>	Physicians	Primary	5	Erosions, papules, blisters	NI
Anesthetic management of a case of retroperitoneal tumor with pemphigus vulgaris with multiple comorbid conditions (2019) <sup>15</sup>	Physicians	Primary	5	Red lesions	NI
Pemphigus in the eastern region of Turkey (2019) <sup>55</sup>	Physicians	Primary	5	Flaccid bullae and vesicles	Erosions
Skin ulcers infected with conditional pathogenic strains treated with local hydrogen water packing in two pemphigus vulgaris patients: case reports with follow-up for 2 months (2019) <sup>19</sup>	Physicians	Primary	5	Erythema, ulcer, purulent and haemorrhagic scar	NI

NI: not identified in the publication

publish little on this subject, which corroborates with the lack of formulation of new evidence for their topical care. This lack of evidence makes nursing assistance to these people less secure, since a theoretical framework is essential for evidence-based care given the specificity and complexity of caring for these patients<sup>16,25</sup>.

The main researchers in this area, Brandão and Santos<sup>4,6,25,29</sup>, investigate this issue from the point of view of integral care, with a social-poetic approach and a nursing diagnosis that support the comfort and relief of pain among those affected.

Through clinical observation, the authors of this study concluded that traditional topical care generates high rates of hospital stay, pain and discomfort. This clinical observation was the trigger to build evidence of the best evaluation tools and descriptors and base this on the use of topical therapies that optimise the potential of healing by managing cofactors such as pH, thermoregulation, humidity, microbial load and adherence of the dressing<sup>25,36</sup>.

It can be observed that when the nursing descriptor was used, the literature search resulted in a much smaller number of articles in comparison to those searches without this descriptor. Although dermatological nursing is a recognised specialty in Brazil, the descriptors 'nursing in dermatology' or 'dermatologist nurse' or 'dermatological nursing' do not exist. Therefore, in search of studies, the descriptors 'nursing' and 'nursing assessment' were used.

Among the nomenclatures used in the different levels of studies, we can observe that all use the dermatological glossary to describe the lesions, with only a small variation with detailed description of the tissue such as exudative erosions. Despite the scarcity of studies written by nurses, it was possible to observe that the terminology used did not differ between professional categories<sup>25,29,36</sup>.

According to Sampaio and Rivitti<sup>3</sup>, skin lesion classification is like letters of the alphabet; just as letters compose words, through their combination they form morphological signs that allow us to "read" the cutaneous manifestations<sup>16,18</sup>. As shown in the results, the skin lesions, associated with complementary information, can help in the definition of diagnostic hypotheses, intervention decision and monitoring of healing evolution. The use of clinical descriptors to characterise skin lesions or ulcers standardises this information, making it more understandable for health professionals. In Table 3 examples of PV lesions related to the correspondent elementary skin lesion term are given.

Skin lesions are classified into six groups: colour changes; oedematous elevations; solid formations; liquid formations; thickness changes; and losses and repairs. This classification may be grouped into:

- Primary: flat spots or maculae (colour changes); solid (solid formations); oedematous elevations (rash, oedema); liquid content (liquid formations).








- Secondary: changes in consistency and thickness (changes in thickness); or loss of substance (tissue losses and repairs)<sup>16,18</sup>.

Diagnoses, interventions and nursing outcomes according to the North American Nursing Diagnosis Association (NANDA), Nursing Interventions Classification (NIC) and Nursing Outcomes Classification (NOC) for patients with PV were proposed by Pena et al.<sup>27</sup>. The analysis of the nursing diagnosis 'impaired skin integrity' and 'impaired oral mucous membrane' made it possible to perceive that the nurse assessment to arrive at this diagnosis does not require a specialised evaluation since the defining characteristics for such diagnoses involve only the rupture of the skin or mucosa, not requiring evaluation of thickness, tissue type or other characteristics assessed in a wound. In the proposed NOC outcome 'healing', in which one of the indicators used was granulation tissue, the knowledge about the tissue types that a wound can present is necessary. The proposed NIC intervention 'wound care' involves the decision about the intervention to be adopted, presupposing specific knowledge, skills and preparation in this area<sup>27</sup>. Considering the increasing performance of dermatology nursing as a specialty, these nomenclatures are becoming increasingly familiar to nurses.

As the nomenclature used to describe skin lesions becomes standard, whether secondary to PV or not, the interlocation between professionals tends to become more uniform, avoiding misunderstanding and resulting in safer patient care. However, in wound assessment there is still a great diversity in the terminology used to describe the skin damage, for example: slough and fibrin; wet necrosis and liquefaction necrosis; dry necrosis, 'scar' and pressure injury; granulation tissue, extra cellular matrix or viable tissue. This richness of terminologies causes doubt, within professionals consulting the records, often generating interpretations that differ from those of the person who registered them. In order to standardise the register of PV ulcers, we suggest the use of the elementary lesions terms – vesicles, blisters, erosions, ulcers and generalised crusts.

The chronic character and the cutaneous manifestation of PV steal from the patient their right to confidentiality. Frequently, the lesions are associated with contagion, impacting on social and familiar relations, and perpetuating stigma and isolation<sup>4</sup>. For Brandão, nursing actions become productive as they aim to reach the patient's holistic balance. Thus there is a constant search for alternatives that attend to the needs of people suffering from changes in skin integrity, and the development of technologies and techniques that promote healing in order to improve their quality of life<sup>4,27</sup>.

Table 3. Elementary skin lesion terms and corresponding examples of PV lesion

Terms used to describe lesions		Corresponding PV lesions
Elementary lesion term	Term used in studies	
Hyperchromia	Post-inflammatory hyperpigmentation, hyperpigmented macules	
Vesicles	Vesicles, blisters	
Erosion or exulceration	Erosions, eroded areas, erosive lesions, excoriations	
Crusts	Crust	
Ulcers	Ulceration, ulcer, ulcerative lesions	

## CONCLUSIONS

This study compared the terminology used to describe cutaneous ulcers in PV studies with the terminology used in describing elementary lesions. It also demonstrated that the publications authored by physicians were the most common ones, followed by those authored by nurses, and lacked uniformity in the description of the lesions.

This study demonstrated that the skin changes in PV can be classified as primary skin lesions (colour changes and liquid collections) or secondary skin lesions (changes in thickness and tissue loss). In view of the extensive dermatological glossary, lesions secondary to PV can be characterised as vesicles, blisters, erosions, ulcers and generalised crusts to support the practice of nursing in dermatology.

Lastly, it is concluded that studies with a higher level of evidence in this area are necessary in order to determine the best way to evaluate the lesions, as well as the nomenclature to be used for improved nursing care.

## CONFLICT OF INTEREST

The authors declare no conflicts of interest.

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