

## Determination of Dermatology Life Quality Index criteria in Libyan pemphigus patients

### Abstract:

**Introduction:** The term "pemphigus" describes a group of uncommon autoimmune dermatoses of the mucocutaneous membranes caused by acantholysis, or the breakdown of cell-to-cell adhesion, which can result in the production of potentially fatal bullae and erosion. Based on their unique clinical characteristics and pathogenesis, several subtypes of pemphigus illness have been discovered. These subtypes include paraneoplastic pemphigus (PNP), IgA pemphigus, pemphigus vulgaris (PV), and pemphigus foliaceus (PF).

**Aim of the study:**

To measure the Dermatology Life Quality Index (DLQI) in Libyan pemphigus patients and assess the impact of the disease on their lives.

**Materials and methods:**

In this descriptive study, 25 patients with pemphigus attending Bullous Clinic, dermatology department, Jumhoria Hospital, Benghazi, Libya, over one year from January to December 2021, were enrolled.

All patients were registered in the clinic, and the diagnosis of pemphigus was confirmed by histopathology.

For every patient, the DLQI score was measured by filling out the questionnaire.

**Results:**

Of the 25 patients with pemphigus included in this study, 5 patients (20%) were male and 20 patients (80%) were female. The age ranged from 30 to 72 years, and the mean age was 43.64 years. The duration of the disease ranged from 1 to 408 months (mean: 83.12 months). Pemphigus foliaceus was encountered in 13 patients (52%). Concerning disease severity, 44% of cases presented with a severe illness. In 15 patients (60%), mucous membranes were not involved. Eight patients (32%) were treated with steroids (Prednisolone) and cytotoxic (Azathioprine) drugs. The mean DLQI scores for pemphigus vulgaris were 12.67 and for pemphigus foliaceus were 10.31. Dermatology Quality Life Index (DLQI) scores showed a large effect on life quality in 56% of the study cases and a moderate effect in 32% of them. **Conclusion:** Pemphigus has a substantial impact on Libyan patients' DLQI scores; individuals receiving systemic combination treatment were shown to have lower scores.

**Keywords:** Pemphigus, pemphigus vulgaris, Demographic features, Dermatology life quality index (DLQI), Libya.

### Introduction:

"Acantholysis, or the loss of cell-to-cell adhesion, is the cause of potentially fatal bullae and erosion development in a family of uncommon autoimmune dermatoses of the mucocutaneous membranes known as pemphigus. Pemphigus vulgaris (PV), Pemphigus foliaceus (PF), IgA pemphigus, and paraneoplastic pemphigus are among the several subtypes of pemphigus disease that have been recognized because of their unique clinical characteristics and pathogenesis" (1, 2). While pemphigus is a global condition, its geographic and ethnic distribution is disproportionate, with individuals of Ashkenazi Jewish or Mediterranean heritage having a much greater frequency (1, 2, 3, 4). Pemphigus affects all racial and ethnic groups, although it is more

common in certain populations (3, 4, 5). HLA-DRB1\*04 and HLA-A\*10 are two of the many unique HLA class II genes that are more common in Ashkenazi Jewish communities and may be the reason for the high incidence of pemphigus (6, 7, 8).

### **Aim of the study**

- 1) To study the demographic characteristics of pemphigus in Libyan patients in the city of Benghazi.
- 2) To assess the disease's impact on patients' lives and measure the Dermatology Life Quality Index (DLQI) in pemphigus patients.

### **Materials and methods:**

In this descriptive study, 25 patients with pemphigus who visited the Bullous Clinic, dermatology department, Jumhoria Hospital, Benghazi, Libya, over one year from January to December 2021, were included.

Each patient was exposed to a complete medical and drug history, followed by an examination according to the prepared proforma. All patients were registered in the clinic after the diagnosis of pemphigus was confirmed by histopathology.

The patients were grouped based on the type of treatment they received: monotherapy—either topical or systemic steroids (Prednisolone), cytotoxic medications (Azathioprine), or combination therapy.

For every patient, the DQLI score was measured by filling out the questionnaire which includes 6 items: symptoms and feelings (Questions 1 and 2), daily activities (Questions 3 and 4), leisure (Questions 5 and 6), and personal relationships (Questions 8 and 9), each with a maximum score of 6, work and school (Question 6), and treatment (Question 10), each with a maximum score of 3.

The scoring for each question is as follows:

Very much:	3
Alot:	2
A little:	1
Not at all:	0
Not relevant:	0
Unanswered:	0

Question 7 "prevented work or studying" scored 3.

DLQI is calculated by summing the scores of each question resulting in a maximum of 30 and a minimum of 0. The higher the score, the more quality of life is impaired.

### **Meaning of DLQI scores:**

- 0-1 = No effects at all on the patient's life.
- 2-5 = Small effect on patient's life.
- 6-10 = Moderate effect on the patient's life.
- 11-20 = Very large effect on the patient's life.
- 21-30 = Extremely large effect on the patient's life.

Statistical analysis:

Data were analyzed using the Statistical Package for Social Science (SPSS), version 23. Descriptive statistics: the mean, standard deviation, and median were estimated. Inferential statistics were applied as needed. Student  $t$ -tests were used to find the difference between the means of the groups and Chi-square ( $X^2$ ) to find the difference in the distribution of the categorical variables between the groups. P-value was considered significant when  $\leq 0.05$ .

### Results:

Among the 25 pemphigus patients included in this study, 20 patients (80%) were females and 5 patients (20%) were males. The age ranged from 30 to 72 years, and the mean age was 43.64 years. The duration of the disease ranged from 1 to 408 months (mean: 83.12 months) (Table 1). Pemphigus foliaceus was encountered in 13 patients (52%), and Pemphigus vulgaris in 12 patients (48%). Concerning disease severity, 44% of cases presented with a severe illness (Figure 1). In 15 patients (60%), mucous membranes were not involved (Figure 2). Eight patients (32%) were treated with steroid (Prednisolone) and cytotoxic drugs (Azathioprine), and 7 patients (28%) were treated with steroid (Prednisolone) only (Figure 3).

Dermatology Life Quality Index (DLQI) scores showed a large effect on life quality in 56% of the study cases and a moderate effect in 32% of them (Figure 4). The mean DLQI scores for pemphigus vulgaris were 12.67 and for pemphigus foliaceus were 10.31 (Table 2).

DLQI scores were lower in cases where pemphigus had only partial skin involvement or was generalized without mucous membrane involvement (mean score: 9.6 and 9.22, respectively) (Table 3).

The mean (DLQI) scores for various pemphigus therapies received indicated that patients receiving systemic steroids (Prednisolone) and cytotoxic drugs (Azathioprine) had a lower mean score (10.50), followed by systemic steroids (Prednisolone) alone (10.71) (Table 4).

### Discussion:

“Pemphigus, an autoimmune blistering disease that affects the skin and mucous membranes, adversely impacts patients' quality of life (QOL). While there are various QOL measurement tools that can be used in this disease, few studies have assessed how a patient's change in disease severity can affect their QOL. This study aims to identify which disease severity index correlates best with the change in QOL” (9, 10 11).

Patients with pemphigus usually fall between the ages of 50 and 60, yet depending on the patient's ethnic background and place of origin, the mean age at diagnosis might vary greatly (11, 12).

The average age at diagnosis in Western Asian countries, such as Kuwait, is 36.5 years, while in European countries, like Bulgaria, it is 72.4 years (13). The average age of the participants in our research was 43.6 years, and 80% of them were female.

“Significantly, reports of illness in pediatric patients—including those as young as 6 years old—have also been described” (14).

“In France, pemphigus vulgaris accounted for 73% of all cases; incidence increased with age; and the female-to-male sex ratio was 1:2”. (2). “In Tunisia, pemphigus foliaceus was more frequent (61%), the female-to-male sex ratio was 4:1, and the incidence rate was higher in young women” (2). “The incidence rate was 15.5 cases per million per year for pemphigus foliaceus among women aged 25 to 34 years and was even higher in some rural areas”. (2). In our patients,

pemphigus foliaceus is slightly more common than pemphigus vulgaris, and the majority of our patients were females (80%), with a mean age of 43.6 years.

“Quality of life (QOL) has been defined as “the individual’s perception of their position in life, in the context of the cultural and value systems in which they live and in relation to their goals, expectations, standards, and concerns” (15).

“The Dermatology Life Quality Index (DLQI) is the most commonly applied questionnaire to measure health-related quality of life (HRQoL) in dermatology” (16). “Since its development in the early 1990s, it has been used for over 40 different skin conditions worldwide”. (17, 18). “It is recognized as being useful in various health service settings, including primary care, day-case treatment, outpatient consultations, and inpatient care”. (19, 20).

“Although a previously deadly diagnosis because of skin barrier breakdown leading to infection, the advent of steroids and steroid-sparing agents has allowed pemphigus to be considered a less-fatal, chronic disease” (21). “In this study, 32% of patients were treated with steroids and cytotoxic drugs, and 28% were treated with steroids only. Therefore, assessing a pemphigus patient's quality of life (QOL) has become an important part of monitoring the disease” (3, 21).

Despite the limited literature on QOL in pemphigus (21–30), it is clear that this disease greatly affects patients' emotions, physical health, and social functioning (30, 31). The number of clinical trials in pemphigus has increased immensely over the past decade (32), creating a growing need for objective measurements in both QOL and disease severity to monitor improvement.

“The QOL of patients with pemphigus vulgaris was evaluated in one study of 27 German participants using a dermatology-specific instrument”. (33). “The dermatology-specific Dermatology Life Quality Index (DLQI) score for the study cohort averaged  $10.1 \pm 6.6$  compared with healthy individuals with a mean score of  $0.5 \pm 1.1$ , and the authors concluded that a diagnosis of pemphigus vulgaris conferred a large impairment in QOL. The study pointed out that DLQI Patients with mucosal involvement were found to have a higher DLQI, averaging  $10.4 \pm 7.3$ , compared with those patients with mucosal sparing, who averaged  $9.3 \pm 5.1$ , indicating a poorer QOL. Ghodsi et al. found a similar mean DLQI score of  $10.9 \pm 6.9$  in 61 newly diagnosed untreated PV patients in Tehran using the Persian DLQI” (16). “Our study showed that the mean DLQI among 25 patients was 11.4. The highest subscores were related to symptoms and feelings (2.8) and daily activities (2.2). Investigators found that the DLQI score was significantly increased in patients with severe disease and mucosal involvement”. [16]

“These results were consistent with our study, which found that DLQI in pemphigus patients with generalized skin involvement or mucous membrane involvement was more impaired than in those without mucous membrane involvement or localized pemphigus. Also, we found a higher DLQI in patients with pemphigus vulgaris than those with pemphigus foliaceus. Disease severity and extent of symptoms likely affect the ability to partake in daily activities and therefore result in lower QOL. A negative correlation between DLQI score and duration of disease was also noted, suggesting increased impairment in the initial stages of the disease. In 2013, Wysocynska et al. reported an average DLQI of  $4.0 \pm 5.9$  in a patient population mainly composed of patients with >5 years of disease” (34). “This is further supported by Tee CT et al.'s study population, which had a low median DLQI ( $3.0 \pm 8.0$ ), and the DLQI scores were not affected by gender, age, ethnicity, or duration of illness” (12).

“Patients likely undergo an adjustment period upon initial diagnosis, which affects QOL scores early on in the disease course. In 2015, a meta-analysis across four studies surrounding QOL in pemphigus patients found a mean DLQI of 12.0 (95% CI 11.1–12.9) with symptoms/feelings and

daily activities subscores most consistently affected”<sup>(35)</sup>. The mean DLQI scores in our study for different pemphigus treatments received showed patients who were on systemic steroids and cytotoxic drugs had a lower mean score (10.50), followed by systemic steroids (10.71).

**Conclusion:** Pemphigus is not just a physical disease and has various aspects. It is responsible for many changes in health related quality of life. In this study, pemphigus had significant effects on DLQI scores in Libyan patients and lower score scores were observed in treated patients with systemic steroids and cytotoxic drugs.

**Ethical approval and consent:** The study was approved by Jumhoria teaching hospital ethical committee - Every patient signed an informed written consent for acceptance of participation in the study.

**Competing interests:** Authors have declared that no competing interests exist.

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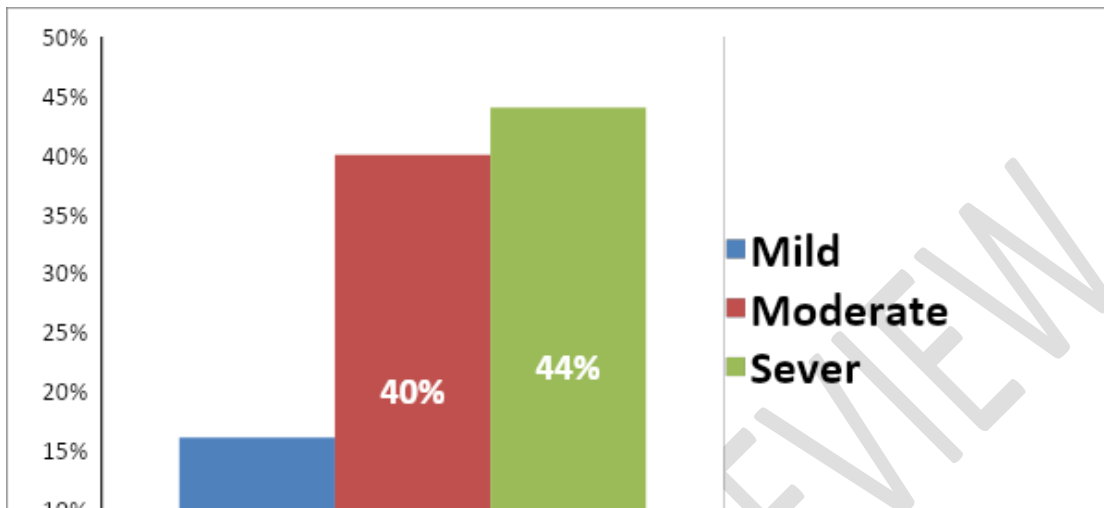
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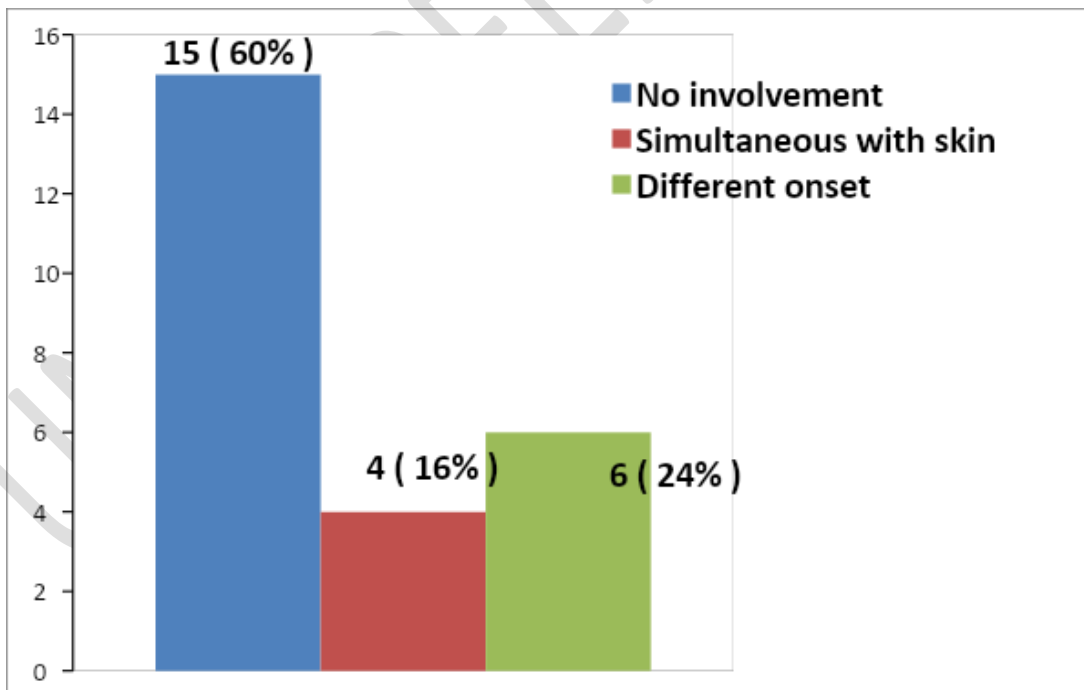
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**Table 1: Descriptive statistics of scale variables in the study**

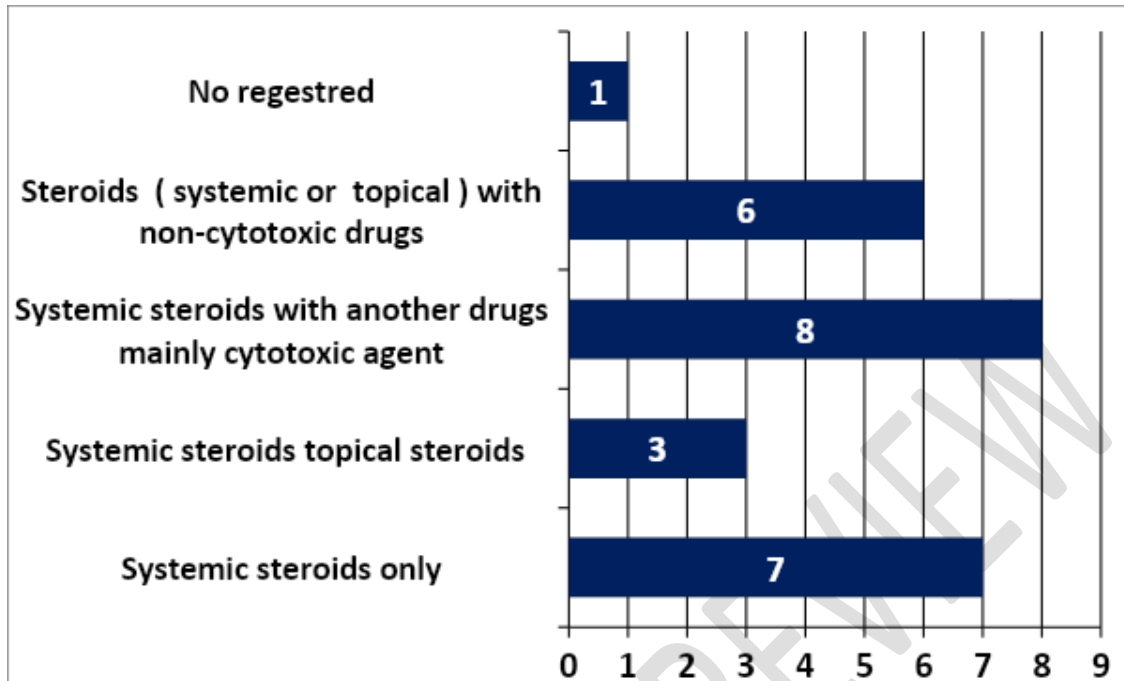
Parameter	Age of study case ( in years)	Duration of illness (in months)	Number of symptoms coexisting	Number of complication	Score of life quality index
Mean	43.64	83.12	5.72	4.16	11.44
Std deviation	10.404	96.782	2.208	2.749	5.432
Minimum	30	1	0	0	0
Maximum	72	408	9	10	19



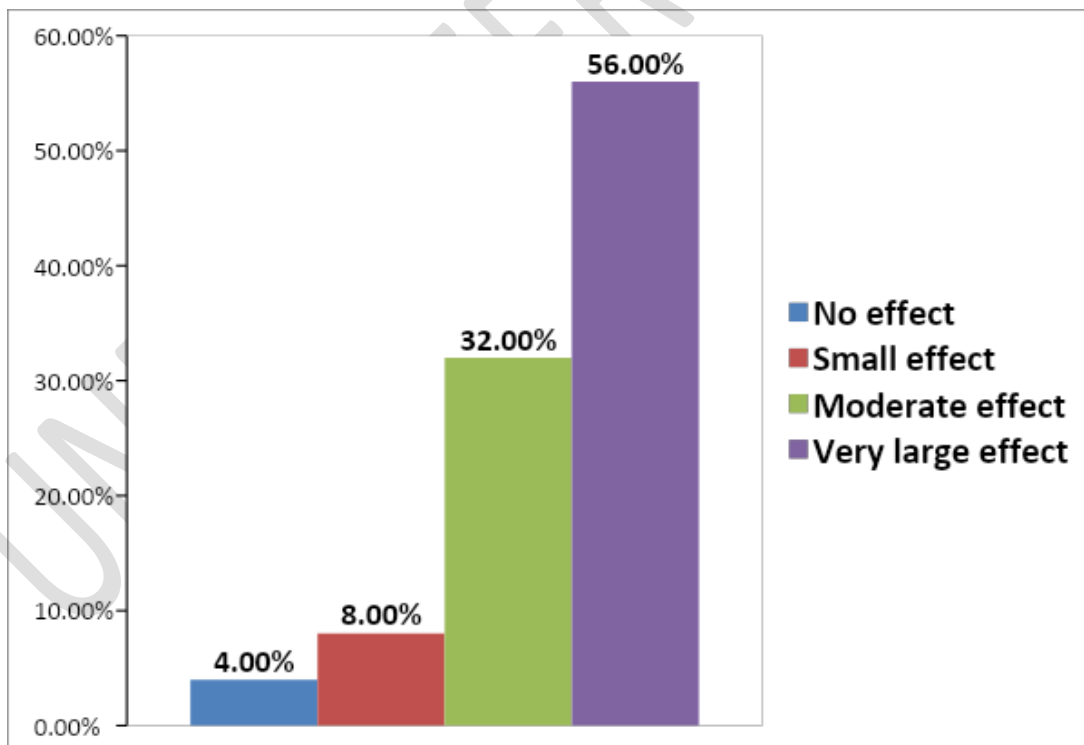
**Figure 1 : Severity of illness in study cases**



**Figure 2 : Mucous membrane involvement in disease in study cases**



**Figure 3 : Treatment used in the study cases**  
 (systemic steroid (Prednisolone), cytotoxic agent (Azathioprine))



**Figure 4 : Distribution of study cases on quality of life index score categories**

**Table 2: Descriptive parameters of score of life quality index in categories of pemphigus type**

Type of pemphigus	Frequency	Mean	Std. deviation
Pemphigus vulgaris	12	12.67	5.314
Pemphigus foliaceus	13	10.31	5.498

**Table 3 : Descriptive parameters of score of life quality index in site of involvement categories**

Distribution on the body	Frequency	Mean	Std. deviation	Minimum	Maximum
Generalized skin involvement	9	9.22	4.790	0	15
Generalized skin involvement with mucous membrane	5	16.60	1.517	15	19
Partial skin involvement	5	9.60	5.079	3	17
Partial skin involvement with mucous membrane	5	13.00	6.819	2	19
Total	24	11.63	5.468	0	19

**Table 4** : Descriptive parameters of score of life quality index in major treatment categories

Type of treatment	Frequency	Mean	Std. deviation
Systemic steroid	7	10.71	4.499
Systemic + topical Steroids	3	12.00	5.245
Systemic steroids + cytotoxic agent	8	10.50	7.231
Systemic or topical steroids + non-cytotoxic agent	6	14.17	3.371
Total	24	11.67	5.427