# Quality of life evaluation in patients with pemphigus vulgaris

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**Background:** Pemphigus is a rare autoimmune disease caused by autoantibodies against desmoglein. It clinically presents with painful blisters and erosions on the skin and mucous membranes. Few studies have been conducted on the quality of life in pemphigus patients which have all indicated the strong impact of the disease on emotional and physical status of the patients. According to evident differences in the culture and quality of life between our society and western societies, we assessed the quality of life in patients with pemphigus in the Northeast of Iran.

**Method:** This study was conducted on 78 pemphigus vulgaris patients who were visited or hospitalized in the dermatology clinics of Qaem and Imam Reza Hospitals in Mashhad, Iran. Data collection was done by the Dermatology Life Quality Index (DLQI) questionnaire in patients with pemphigus vulgaris. Statistical analysis was performed using SPSS 11.5 software with Chi-square, T-test and ANOVA statistical tests.

**Result:** The disease had a significant impact on the quality of life in the majority of patients (31 (39.7%) patients). Hospitalized and newly diagnosed cases and those receiving higher doses of steroid had lower quality of life. The patient's age, sex, education level, and therapeutic regimen did not have a significant effect on the quality of life.

**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 patients. Therefore, we will be successful in treating this disease only when psychological and social aspects of pemphigus are considered in addition to clinical improvement of the patients.

**Keywords:** autoimmune bullous disease, dermatology life quality index, pemphigus, pemphigus vulgaris

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## INTRODUCTION

Assessment of the quality of life has been increasingly considered to monitor quality improvement of services provided for patients with skin disease. Quality of life is assessed in dermatology for clinical, research, political and financial purposes. Although the impact of skin diseases on the patients' lives has been recognized since long ago, quality of life assessment tools have been recently considered in approaching patients suffering from chronic skin diseases. The majority of the patients with skin problems experience a chronic resistant course that affects their life quality and can be among the important factors predisposing to depression due to their chronic nature and impact on the individual's perception of their appearance. Many of these patients either do not begin their treatment due to depression or give it up due to frustration. Some patients have committed suicide due to depression and despair [1,2]. However, few reports have been published in this field. In a study by Terrab, there was a significant reduction in the average score in all components of the SF-36 questionnaire in the patient group when compared to healthy controls except for pain perception and overall health awareness change, with the highest variation in physical and emotional status. A significant reduction in the quality of life was observed in pemphigus patients in a study by Mayrshofer, and DLQI was suggested as an appropriate indicator in evaluating the quality of life at the time of diagnosis and during treatment [3,4].

Tabolli reported that pemphigus patients with a more chronic disease have a better quality of life compared to the newly identified ones [5]. In a study by Paradisi et al, a significant relationship was observed between the severity of pemphigus and decreased quality of life, especially in female and elderly patients. About 39.7% of the patients in this study suffered from anxiety and depression [6]. In a study conducted by Ghodsi, more than 77% of the pemphigus patients experienced anxiety and depression in the course of the disease [7]. In another study by Darjani, factors related to decreased quality of life included older age, long term disease, frequent hospitalization, lack of secondary studies, occupation, and treatment type [8]. As pemphigus is among the most prevalent diseases leading to hospitalization in hospital skin wards, and considering the high pressure of this disease on patients, rarity of the studies about the impact of this disease on the quality of life in patients, and the differences in the culture and quality of life between our society and western cultures, we decided to evaluate the quality of life in patients who suffer from this disease.

## PATIENTS AND METHODS

We first contacted Professor Finlay who designed the Dermatology Life Quality Index (DLQI) questionnaire for permission to use the Persian version of this index validated by a number of our colleagues in Shiraz University [9]. The questionnaire was then distributed among 78 patients with pemphigus vulgaris who were visited or hospitalized in the dermatology clinics of Qaem and Imam Reza Hospitals. The patients completed the questionnaire containing 10 questions, which had a minimum and maximum score of 0 and 30, respectively. A higher score indicated lower quality of life in patients, classifying them in five categories (Table 1).

**Table 1.** Classification of patients based on DLQI questionnaire index

| Group | Questionnaire Score | Disease impact on the quality of life |
|---|---|---|
| 1 | 1 - 0 | No impact |
| 2 | 5 - 2 | Little impact |
| 3 | 10 - 6 | Medium impact |
| 4 | 20 - 11 | High impact |
| 5 | 30 - 21 | Very high impact |

Patients with pemphigus vulgaris visiting the dermatology clinics of Qaem and Imam Reza hospitals participate in the study and they filled written consent to enter this assay. The goal of the study was explained to patients. Exclusion criteria included lack of sufficient knowledge or proper understanding of the concepts of the questionnaire to complete it and age below 16.

In this study, the collected data were assessed from two viewpoints. First, the demographic data including sex, age, disease duration, education level, classification of patients into two groups of recently diagnosed (those who had not been treated and were newly diagnosed with pemphigus vulgaris) and old patients, the type of the therapeutic regimen based on administration of systemic steroid alone or regimens including adjuvants (mainly azathioprine at 2-3 mg/kg/day), steroid dosage based on disease severity from 0.5-2 mg/kg/day (classified as follows to facilitate statistical analysis: 0.5-1, 1-1.5, and 1.5-2 mg/kg/day) were recorded. Second, the relationship between the above-mentioned variables was compared with the DLQI value calculated for patients. The data was analyzed using SPSS 11.5 software by ANOVA, t-test and chi-square statistical tests.

## RESULTS

Out of 78 patients with pemphigus vulgaris, 31 patients (39.7%) were male and 47 (60.3%) were female with a mean age of 46.98±13.48 years, ranging from 22 to 79 years. The most frequent age group was 51 to 60 years with 23 patients (29.5%). Twenty-five patients (32.1%) were recently diagnosed while 53 (67.9%) were old patients. The median duration of the illness in old patients was

**Table 2.** Frequency of patients in classified groups according to the DQLI rating scale

| Group | Frequency | Percent |
|---|---|---|
| 1 | 17 | 21.8 |
| 2 | 8 | 10.3 |
| 3 | 16 | 20.5 |
| 4 | 31 | 39.7 |
| 5 | 6 | 7.7 |

**Table 3.** The relationship between some variables in patients and their DLQI scores

| Variable | DLQI score (Mean ± SD) | P value |
|---|---|---|
| Sex | | |
| Male | 9.67 ± 6.97 | 0.701* |
| Female | 9.02 ± 7.57 | |
| Disease chronicity (month) | | |
| New patients | 12.72 ± 6.4 | 0.004* |
| Old patients | 7.66 ± 7.18 | |
| Treatment Regimen | | |
| Corticosteroids alone | 7.8 ± 7.5 | 0.139* |
| Corticosteroids & adjuvants | 10.33 ± 6.9 | |
| Corticosteroids dosage | | |
| 0.5-1mg/kg | 6.09 ± 5.1 | 0.001# |
| 1-1.5 mg/kg | 11.9 ± 6.7 | |
| 1.5-2 mg/kg | 12.87 ± 7.14 | |
| Educational level | | |
| Diploma | 9.8 ± 6.3 | 0.593 # |
| Masters | 10.05 ± 8.5 | |
| High degree | 7.1 ± 7.6 | |

*: t-test, #: ANOVA

16 months and between quartile ranges was 7-36 months.

The score of the DLQI questionnaire ranged from 0 to 28, with a mean score of 9.20±7.32. The quality of life score fell in group 4 of the table 1 in 31 patients (39.7%) showing that the disease severely affected them; a minority of the patients were in group 5 indicating very high impact of the disease on their lives. (Figure 1 and table 2)

Table 3 shows the relationship between the indices assessed in patients with the score of the DLQI questionnaire.

## DISCUSSION

One out of three individuals in the society experiences some kind of skin disease in their lifetime [10]. On the other hand, skin diseases can have different impacts on various dimensions of the individual's life [10]. Healthy skin is essential to the sense of physical and mental well-being.

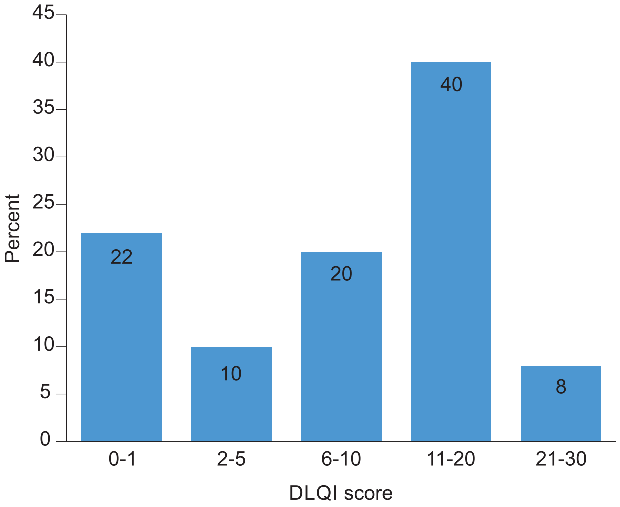

**Figure 1.** Frequency of patients in classified groups according to the DQLI rating scale

Healthy skin is also one of the important aspects of sexual attraction and self-confidence of the people. Therefore, any lesion on the skin is important for other people and will affect the individual [11]. As the term quality is an ambiguous one, the quality of life phrase can encompass nearly everything [12]. WHO has defined quality of life as "personal understanding of the status of life in the context of culture and value systems in which the individual lives in relation to goals, expectations, and standards" [13].

Some skin diseases such as eczema, acne, and pemphigus can change the appearance of the individual and have a destructive impact on their lives. Pemphigus has a special status among these chronic diseases. A significant decrease in the quality of life of pemphigus patients was observed in a study performed by Terrab et al [3] on 30 patients and 60 healthy controls using a general health questionnaire as well as another study by Mayrshofer in Germany conducted on 36 newly diagnosed pemphigus vulgaris patients [4]. In our study, the majority of the patients were in group 4 indicating a high impact of the disease on the quality of life while 7.7% of the patients were in group 5 implying a very high impact on the quality of life and nearly 20.5% were in the medium impact group. According to these results, it can be concluded that a high percentage of the patients in our study population lacked the indicators of good quality of life when compared to healthy individuals.

In view of the relationship between gender and the quality of life, no significant relationship was found in the studies performed by Tabolli [5], Ghodsi [7] and also Arbabi [13]. However, in a study by Paradisi [6], the quality of life index was found to be lower in women than men. In our study, no significant relationship was observed between the quality of life and gender. Considering the relationship of the quality of life and the patient's age, no significant relationship was found in our study as well as the studies performed by Tabolli [5] and Godsi [7] but in the studies by Paradisi [6] and Darjani, elderly people had a lower quality of life. This difference may be due to the small number of the elderly patients in our study. In our study and those conducted by Tabolli [5] and Ghodsi [7], patients with a more chronic disease had a better quality of life compared to newly diagnosed patients, which may be due to the onset of treatment, control of symptoms over time, and ability to accept the conditions, indicating the positive effect of treatment on improving the quality of life.

In the study by Darjani [8], it was observed that in patients who simultaneously took steroids and adjuvant, the quality of life was lower than other patients while we noticed no significant relationship. This reason for the difference could be that adding an adjuvant to the patients' regimen allowed us to taper the steroid dosage more rapidly, so the patients had a lower risk of steroid side effects that could in turn affect the quality of life.

Considering the relationship between the education level and the quality of life, no significant relationship was observed in our study as well as Tabolli [5] and Ghodsi [7] studies; however, in the study by Arbabi [14] on 414 patients with various skin diseases, it was observed that patients with higher education levels had a better quality of life. This discrepancy may be due to small number of patients with higher education in our study (8 Cases). Another index assessed in our study was the correlation between the administered steroid dose and the quality of life. Increased doses of steroid decreased the quality of life in our patients and it can be concluded that increased severity of the symptoms increases the steroid dose with a higher likelihood of drug sequelae in higher steroid doses.

Our study had a descriptive cross-sectional design and was related to a period in the course of the disease. The most important limitation of our study was the sampling method. If it was possible to conduct this evaluation in a more extended period, the study group could be assessed in better conditions in terms of disease extent and severity or dose equality of the administered drugs. The research tool in this study was DLQI designed by western researchers for their own culture and people and although validated by our colleagues in Shiraz University for the Iranian population, the need for a completely compatible tool with our culture and norms is felt. As the questionnaire was used for evaluation and some patients lacked the required education level to complete it, a percentage of the available patients were excluded that could interfere with the test results.

Pemphigus is not just a physical disease and has various aspects. Therefore, pemphigus treatment is successful only when the psycho-social aspects of the disease are considered in addition to clinical improvement of the patients. A good approach to achieve this goal is to determine the quality of life using appropriate standard tools consistent with Iranian society. Completion of this tool for outpatient or hospitalized patients and their follow-up in later visits and comparing the variations in the quality of life indexes over time can help us to choose appropriate treatment. The presence of an attending psychologist as a consultant to help patients to manage the mental and social aspects of skin diseases can be helpful, especially when the complexion of the individual is affected. Informing the relevant authorities and supportive organizations of the socio-mental problems of the patients with bullous or any other skin diseases and, if possible, organizing centers for emotional and economic support of the patients is suggested.

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