

# Quality of life index evaluation in pemphigus vulgaris patients of Pakistan

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## Abstract

**Background** Pemphigus vulgaris is an immunobullous disorder characterized clinically by recurrent painful ulcerations in the mucous membranes and erosive blisters all over the body.

**Objective** The aim of this study was to evaluate quality-of-life and mental health status in Pakistani patients suffering from pemphigus vulgaris.

**Methods** A total of 40 patients with pemphigus were recruited from outpatient and indoor of Dermatology Unit-1, Jinnah hospital Lahore and Sir Ganga Ram hospital Lahore, from November, 2018 to June, 2019. They were assessed clinically, and then interviewed using urdu version of Dermatology Life Quality Index (DLQI) questionnaire.

**Results** Out of the 40 patients, there were 17 male and 23 female patients. No statistical correlation was found between demographic variables (age, sex, education level, rural/urban) and DLQI scores. However, there was positive correlation between marital status and DLQI score ( $P=0.012$ ) and negative correlation with chronicity of disease ( $P=0.018$ ). The average DLQI score was  $14\pm 7$ . Symptoms and embarrassment caused by disease were the most effected DLQI variables while sex life, sports and work were least effected.

**Conclusion** DLQI is important in deciding treatment regimen especially in young, unmarried and those with recent symptoms. It can alert the physician regarding depression and anxiety in a patient. Quality of life can thus be greatly improved.

## Key words

Pemphigus vulgaris, immunobullous, Quality of Life Index.

## Introduction

Pemphigus vulgaris is an autoimmune blistering dermatosis, characterized clinically by chronic, recurrent mucosal erosions and cutaneous blisters all over the body accompanied by pain, itching and burning in the lesions. It accounts for approximately 70% of all cases of pemphigus.<sup>1</sup> It equally affects both genders and mean age of

onset is usually between 50 and 60 years.<sup>2</sup>

The pain in erosions and body disfigurement due to generalized distribution of the disease exert a detrimental impact on physical and mental health of patients like inability to perform daily routine activities, functional limitation, low self-esteem and social isolation respectively.<sup>3</sup> Chronic and recurrent nature of disease add financial burden, anxiety, frustration and disinterest in continuation of their treatment. In extreme cases, they are rejected by their family leading to severe depression and suicidal tendencies.<sup>4</sup> Both disease and its treatment

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(steroids and immunosuppressive drugs) have significant effect on Quality of Life (QOL) in patients with pemphigus vulgaris.

World Health Organization (WHO) defines the term QOL as “individual’s perception of their position in life, in context of the cultural and value systems in which they live and in relation to their goals, expectations standards and concerns”.<sup>5</sup> The measurement of QOL gives the clinician a deep insight into patient’s overall health, a suitable mean to monitor disease activity and to evaluate the effectiveness of holistic care. Several assessment tools exist to quantify the impact of disease on QOL. Among these tools, Dermatology Life Quality Index (DLQI) is a skin specific measurement tool.<sup>6</sup> A 10-item DLQI was first designed by Finlay and Khan in 1994.<sup>7</sup>

Layegh P. evaluated the QOL in patients of pemphigus vulgaris by using DLQI questionnaire. He reported that DLQI score fell into the category of high disease impact on quality of life in 39.7% patients of pemphigus vulgaris while 7.7% patients were found to be in area of very high impact.<sup>8</sup> A significant reduction in the quality of life was observed in pemphigus patients in a study by Mayrhofer, and DLQI was suggested an appropriate indicator of evaluating the QOL at the time of diagnosis and during treatment.<sup>9</sup> Tabolli reported that pemphigus patients with a more chronic disease have a better quality of life compared to newly identified ones.<sup>10</sup> In a study by Paradisi *et al.*, a significant relationship was observed between severity of pemphigus and decreased quality of life, especially in female and elderly patients.<sup>11</sup>

Many researches reveal that autoimmune blistering dermatoses have negative impact on patient’s life in western countries<sup>12</sup> but there is no local data (considering the distinctive social

and cultural traditions of Pakistan) exclusively regarding pemphigus vulgaris. The aim of this study was to quantify the impairment of QOL in pemphigus vulgaris patients of Pakistan using Urdu version of the DLQI questionnaire.

## **Materials and Methods**

This cross-sectional study was conducted on 40 patients of either sex and age above 20 years with clinical and histopathological diagnosis of pemphigus vulgaris. The patients were enrolled from outpatient and indoor departments of Dermatology Unit-1, Jinnah Hospital Lahore and Sir Ganga Ram Hospital Lahore from November 2018 to June 2019. Approval from Ethical Committee of the concerned institutes was ensured.

After taking informed consent, demographical and clinical data were noted on pre-designed proforma. All the enrolled patients were interviewed to complete the DLQI questionnaire. It is a valid, simple and practical questionnaire designed to measure disability caused by pemphigus vulgaris. This questionnaire consists of 10 questions covering six different domains of QOL: symptoms and feelings (questions 1 & 2), daily activities (questions 3 & 4), leisure (questions 5 & 6), work/school (question 7), personal relationships (questions 8 & 9) and treatment (question 10).

Each question is scored on a 4-point scale; 0=not at all, 1=mild/ a little, 2=moderate/ a lot, 3=severe/ very much. DLQI total score is calculated by summing the score of each question resulting in a maximum of 30 and a minimum of 0 score. Higher the score, the more QOL is impaired.

Data was analyzed using SPSS version 20. Student’s t test was used for comparison of

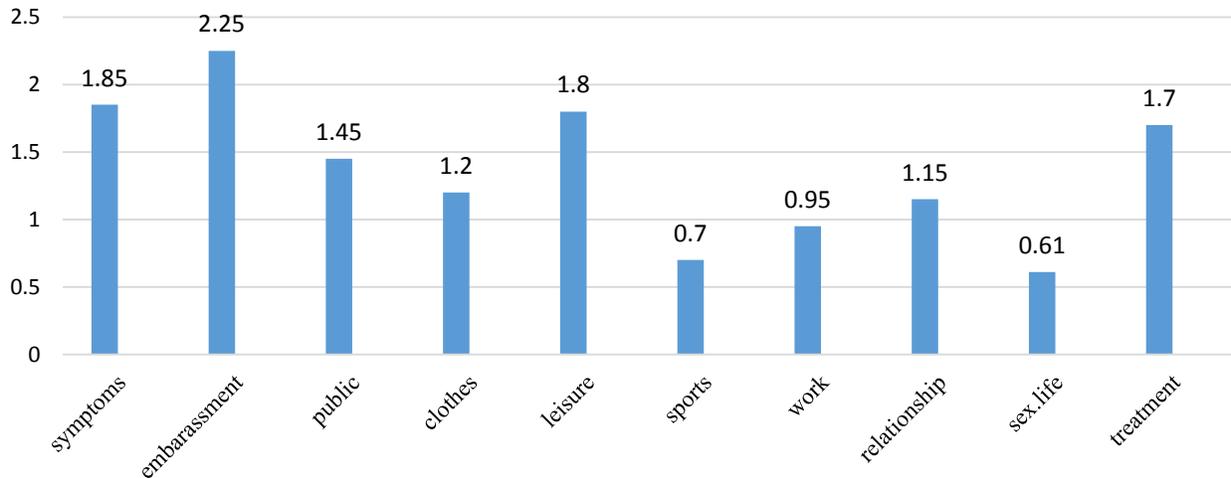
mean. A p value of <0.05 was considered significant.

**Results**

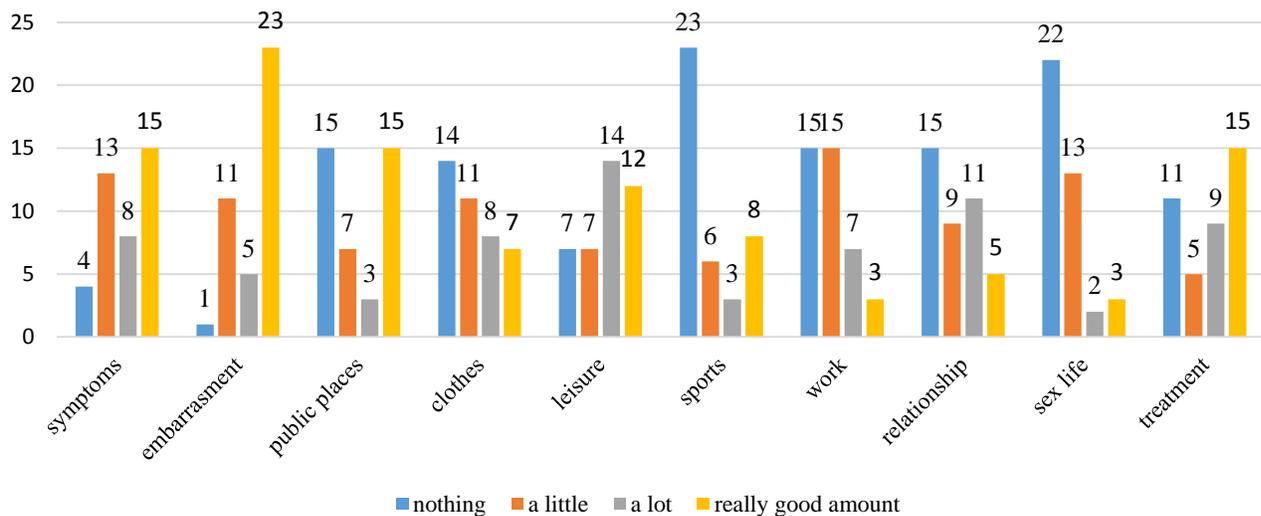
40 patients with pemphigus vulgaris were interviewed. The mean age of patients participating in study was 46±12yrs with clustering of patients in 51-60 age group. Out of 40, 23 were females.

No statistical correlation was found between demographic variables (age, sex, education level, rural/urban) and DLQI scores. However, there was positive correlation between marital status and DLQI score (P=0.012). A negative correlation was found between DLQI score and chronicity of disease (P=0.018)

The average DLQI score was 14±7. Symptoms and embarrassment caused by disease were the most affected DLQI variables while sex life, sports and work were least affected.



**Figure 1** Mean DLQI category scores in 40 patients.



**Figure 2** Scores of DLQI items for pemphigus vulgaris patients.

## Discussion

Dermatological diseases have a negative impact on patient's quality of life and mental health. Routine assessment of patient's quality of life is crucial in charting out treatment strategies. Despite this serious illness, no research regarding QOL assessment in pemphigus vulgaris patients has been conducted in Pakistan. This highlights the fact that most physicians in Pakistan only consider the physical aspect of disease and often undertreat pemphigus.

In our study, then mean DLQI score was  $14 \pm 7$ , which is comparable to the study done in Egypt<sup>13</sup> but more compared to studies conducted by Mayrshofer *et al.* ( $10.66$ )<sup>9</sup> and Ghodsi *et al.* ( $10.98 \pm 5.9$ ).<sup>14</sup> This underscores the fact that Pakistani patients have a strong emotional and mental component effected by disease. Out of demographic variables, only marital status showed correlation. This is expected due to cosmetic disfigurement caused by pemphigus, which creates fear and anxiety among unmarried regarding future proposals; Age and gender had no impact. This is consistent with studies conducted by Tabolli and morsya.<sup>13</sup>

DLQI questionnaire has been used to assess other dermatological conditions in Pakistan. On comparing with pemphigus, vitiligo ( $9.56$ ),<sup>15</sup> acne ( $7.84$ )<sup>16</sup> and psoriasis ( $11.5 \pm 4.81$ )<sup>17</sup> had lower scores. High mortality, painful oral and cutaneous ulcers, prolong course and aggressive treatment with its side-effects seem to be the reason for higher DLQI scores. This is consistent with work done by Mayrshofer and colleagues.<sup>9</sup>

In our study, patients ranked disease symptoms and feelings of embarrassment to be the most effected among DLQI categories. Symptom burden is attributable to mucosal involvement and blister fragility. Pain from erosive blisters

and oral ulcers causing poor alimentation lead to poor quality of life. Social embarrassment is caused by disfiguring cutaneous blisters. Contrary to this sports, sex life and work were least affected. This is in contrast to studies done in developed countries where work is affected to a greater degree. This is expected as 60% of our patients were females who work indoors compared to women in developed countries who tend to work in offices.

Chronicity of disease had negative correlation with DLQI scores, meaning the QOL is more impaired during early phase of disease. This is consistent with literature review.<sup>14</sup> Lack of knowledge about the disease, along with initial treatment related complications might explain the anxiety in patients regarding newly diagnosed disease. On the contrary, chronic patients are more mentally adapted, even though disease severity might increase over time and systemic treatments over time may impose serious side effects.

Patients reported treatment related side-effects to be quite bothersome. This is expected from prolonged steroids use. However, there was limitation in our study regarding this domain, whether duration of treatment or addition of adjuvant treatment had any impact. Similarly dosage of steroid use is also relevant. This has been observed in research done by Layegh and colleagues,<sup>18</sup> who showed in a sample size of 78 patients that disease chronicity has positive correlation with quality of life, while therapeutic regimen had no statistical effect. However, higher dosage of steroid were associated with poor quality of life.

Another domain that needed attention was surface area involved. Study of literature reveals that positive correlation exists between surface area involved by disease and DLQI scores.<sup>13</sup> This is evident in a Korean Study<sup>5</sup> where QOL

improved in patients who went into complete remission with little to no lesions. However, same response was not observed in patients with partial remission. This further validates our view that treatment response dictated by clinical improvement of lesions is not the same as patient's QOL improvement.

## Conclusion

Patient's subjective assessment of disease is as important as physician's objective assessment of disease. Our research being consistent with other studies, reveals that calculation of DLQI is important in deciding treatment regimen especially in young, unmarried and those with recent symptoms. Therapeutic response is greatly dependent on patient satisfaction. DLQI is an important tool that can alert the physician regarding depression and anxiety in patient and with prompt management, QOL can be markedly improved.

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