

## Quality of Life of Patients with Pemphigus Vulgaris in Alexandria - Egypt

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**Abstract:** Pemphigus vulgaris (PV) is a life-threatening autoimmune disease resulting in the widespread denudation of mucous membrane and skin as well as severe impact on quality of life (QoL). The aim of this study was to assess the impact of PV disease on patient's quality of life. The study was conducted at the Dermatology Outpatient Clinic of the Main University Hospital, Alexandria, Egypt. This is a descriptive study. (100) adult PV patients showing up at the previously mentioned setting were recruited to the study. Sociodemographic and clinical data structured questionnaire and the World Health Organization Quality of Life (WHOQOL) – BREF sheet, were utilized for data collection. Results showed highly significant relationships noted between WHOQOL BREF scores and severity of PV disease in relation to general health, overall QoL, physical, psychological and environmental domains ( $\chi^2 = 14.010$  &  $P < 0.001$ ), ( $\chi^2 = 12.019$  &  $P < 0.001$ ), ( $\chi^2 = 8.518$  &  $P < 0.001$ ), ( $\chi^2 = 7.681$  &  $P < 0.001$ ), ( $\chi^2 = 3.681$  &  $P < 0.001$ ) respectively, while no significant relation was found between social domain and severity of disease ( $\chi^2 = 0.0555$  &  $P < 0.960$ ). The majority of the studied patients had poor quality of life in relation to all quality of life domains: physical, psychological, social and environmental domains (81, 68, 68 and 64 %) respectively. Conclusion: PV had a significant negative impact on the majority of patients' quality of life.

**Key words:** Pemphigus diseases • Pemphigus Vulgaris • Quality Of Life

### INTRODUCTION

Pemphigus is a group of potentially life-threatening autoimmune diseases characterized by cutaneous and mucosal blistering. Pemphigus can be categorized into several types such as pemphigus vulgaris, pemphigus vegetans, pemphigus erythematosus and pemphigus foliaceus [1-3].

Pemphigus vulgaris (PV) is a serious condition that has multiple clinical variants and causes painful blisters to develop on the skin and the lining of the mouth, nose, throat and genitals. The blisters are fragile and can easily burst open, leaving areas of raw unhealed skin that are very painful and increase risk of infection. However, it is not a contagious disease [4, 5].

In the case of PV, the immune system mistakenly produces antibodies against specific proteins in the skin and mucous membranes, known as desmogleins. These proteins form the glue that keeps skin cells attached and the skin intact. When desmogleins are

attacked, skin cells separate from each other and fluid can collect between the layers of skin, forming blisters that do not heal. In some cases, these blisters can cover a large area of skin [4, 6].

The cause of PV is still unknown, although the disease has attracted considerable interest [1]. Some triggers, including emotional stress, thermal burns, ultraviolet rays, certain medications, infections and genetic factors might be responsible for the disease provocation [4, 7]. There's currently no cure for PV, but treatment can help keep the symptoms under control [1, 8].

In general, PV affects people of all ages, including children, but most cases develop in older adults between the ages of 50 and 60 years. Also, it is equally distributed among genders [9]. It has a prevalence of around 30,000 cases in the USA and an incidence of 1–10 new cases per 1 million people per year [9, 10]. In Egypt, epidemiological studies were not carried out, despite the disease being relatively frequent [11].

Pemphigus vulgaris can negatively affect patient's physical, psychological and social health; also significantly impair the quality of life (QoL) of affected patients [12-14]. Since PV is a relapsing, difficult to treat illness, requiring long-term hospitalization and immunosuppressive treatment besides affecting the patient's appearance, it may inflict significant psychological trauma to the patients. Additionally, PV patients are prone to different psychiatric disorders such as profound depression and suicidal ideations [15].

Moreover, painful and non-healing oral erosions in PV patients can disturb sleep and make eating, drinking as well as brushing teeth very difficult which significantly reduce QoL [8, 16]. Also, genital lesions cause a burning sensation when the patient is urinating, especially in women [16, 17]. Various evidences reported an association between PV and other physical diseases such as dementia, epilepsy, Parkinson disease, Myasthenia Gravis, rheumatoid arthritis and cardiovascular diseases [18, 19]. Additionally, patients with PV have a higher risk of mortality from infections, especially septicemia and pneumonia, as well as cardiovascular diseases [20-22].

The disfiguring nature of the PV and especially the visible skin lesions negatively affect self-confidence and limit social capacity. Hence, it is conceivable that workplace life and work productivity are also affected. Pemphigus disease causes job loss. Losing a job from public or individual perspective is very important and has a significant impact on the individual's financial and social dimension [23].

Usually, occurrence of the PV disease makes the patient to always be alone. In addition, this disease affects social and sexual relationships and thus reduces QoL. In this regards, many studies reported that, single person loses the chance of marriage with this disease and the married patients divorce because their marital needs are not completed, secondary to genital erosions as well as lack of knowledge about the disease [16, 24, 25].

Evidence suggests that, an adverse drug reaction and financial burden associated with a long term treatment of PV are another challenge, reducing the patient's QoL. Care for patients with PV requires not only treating clinical manifestations, but it is also very important to identify and manage psychosocial aspects [2, 11].

As the essential objective of QoL is to ensure the highest level of well-being of people despite their disabilities, nurses have a crucial role in promoting patient comfort, education and planning nursing interventions aiming at improving patients' QoL through proper management. Additionally, nurses are in a unique position

to address the multidimensional impact of PV on patients' QoL by allowing them to verbalize concerns, teaching patients about the disease process, prevent disease complications, assisting patients to participate in their usual activities, encouraging patients to comply with their treatment and suggest lifestyle modifications that will potentially improve patients' QoL [26].

Improving QoL in pemphigus vulgaris patients is considered a very great challenge. Specific therapeutic and psychological strategies are needed to improve the negative impact of the disease on the general health. Literature review revealed limited studies assessing the impact of pemphigus vulgaris on patients' QoL among Arabs, especially Egyptians. Lack of those data necessitates the design of this study.

**Aim of the Study:** This study aimed to assess the impact of pemphigus vulgaris disease on patients' quality of life.

**Research Question:** What is the impact of pemphigus vulgaris disease on patients' quality of life?

## MATERIALS AND METHODS

### Materials

**Research Design:** A descriptive research design was used for the purpose of the study.

**Setting:** The Dermatology Outpatient Clinic of Main University Hospital, Alexandria, Egypt.

**Subjects:** A convenience sample of 100 adult PV patients attending the pre-mentioned health setting and meeting the following criteria, were included:

- Age: 20- 60 years old.
- Had a definite diagnosis of confirmed PV by the dermatologists.
- Absence of uncontrolled associated illnesses e.g. (hypertension, heart diseases, etc) or any other dermatology diseases.

**Sample Size Calculation:** Epi info -7 programs was used to estimate the sample size using the following parameters:

- Population size = 140
- Expected frequency = 50 %
- Acceptable error = 5%
- Confidence coefficient = 95 %
- Minimum sample size = 100 patients

**Tools:** Based on an extensive review of related literature; two tools were used for the purpose of data collection [11, 14, 15, 18 & 26-28].

**Tool I:** Socio demographic and clinical data structured questionnaire: This questionnaire was developed by the researchers to obtain information's about sociodemographic and clinical data of the studied patients. It consists of two parts as follows:

**Part I:** This part includes questions related to the socio demographic data of the studied patients (age, gender, level of education, occupation, marital status, area of residence and income).

**Part II:** It includes questions related to the clinical data of the studied patients (family history and onset of the disease, body surface area affected with the disease as well as severity of pemphigus vulgaris).

Severity of disease was determined by using Kumar's Scoring System [27], which includes the following:

- No mucosal and skin disease involvement.
- Mild disease (<10% body surface area (BSA) and only buccal mucosa, labiokingival, lingual, palatal, pharyngeal).
- Moderate disease (11-30% BSA and buccal and labiokingival, lingual, palatal or pharyngeal).
- Severe disease (>30% BSA and extensive oral erosions, i.e., >3 mucosal sites affected).
- The extent of affected area is calculated by rule of nine.

**Tool II:** The World Health Organization Quality of Life (WHOQOL) - BREF sheet: This tool was adopted from WHO [28] to measure how much the pemphigus vulgaris disease affected the patient's quality of life. It is a 26- item instrument consisting of four domains: physical health, psychological health, social relationships and environmental health. The tool also contains two items related to overall quality of life and general health.

- The physical health domain (7items) includes items on energy and fatigue, pain and discomfort, sleep and rest, mobility, activities of daily living, dependence on medical substances and work capacity.
- The Psychological domain (6 items) measures include body image and appearance, negative feelings,

positive feelings, self-esteem, thinking, learning, memory and concentration, spirituality, religious and personal belief.

- The social relationships domain (3 items) contains questions on personal relationships, social support and sexual activity.
- The environmental health domain (8 items) contains questions related to financial resources, freedom, physical safety and security, health and social care: accessibility and quality, home environment, opportunities for acquiring new information and skills, opportunities for recreation/leisure, physical environment and transportation.
- Each item is rated on a five point Likert scale and scored from one to five on a response scale. Answers of the studied patients were recorded, scored and then summed together. The total score was categorized into two levels as follows:
  - Poor quality of life: less than 60%.
  - Good quality of life: equal or more than 60%.

- Among the various instruments for measuring QoL in literature, the WHOQOL-BREF was chosen for this study because of the breadth of its domains and facets.

#### **Method:**

- Permission to collect data for the study was obtained from the directors and the responsible authorities of the previously mentioned study setting after explanation of the study purpose.
- Tool I was developed by the researchers based on a recent review of literature and tool II was adopted from WHO.
- Content and construct validity of the study tool were established by a jury of five experts in the fields of Medical Surgical Nursing and Dermatology. The necessary modifications were introduced accordingly.
- The tools reliability was tested using Cronbach's Alpha Coefficient Test ( $r=0.891$ ), which denotes high reliability.
- A pilot study was carried out involving 10% of the studied patients (who was not included in the study) to test clarity, feasibility and applicability of the developed tools. Based on the obtained results, the necessary modifications were introduced.

**Data Collection:**

- Every patient was interviewed individually after a brief explanation of the purpose of the study. The interview took around 20 minutes each. Data was collected over a period of 5 months starting from January to May 2017.

**Ethical Considerations:**

- Informed written consent for voluntary participation in the study was obtained from the pemphigus vulgaris patients after explaining the aim of the study. For illiterate patients, verbal explanation of the study purpose and patients' oral consents were secured.
- Subject's privacy and anonymity were assured.
- All patients were informed that they could withdraw from the study at any time if they wish not to continue.
- Data confidentiality was considered and respected.

**Statistical Analysis:**

- After the data were collected, they were coded and transferred into specially designed formats, so as to be suitable for computer feeding. Verification processes were carried out to avoid any errors during data entry.
- The suitable statistical program was utilized (IBM SPSS software package version 20.0) [29, 30] for both data presentation and statistical analysis of results.
- Qualitative data were described using numbers and percent and quantitative data were described using mean and standard deviation.
- Comparisons between different groups regarding categorical variables were tested using the Chi - square test. When more than 20% of the cells had an expected count less than 5, corrections for chi-square were conducted using Monte Carlo correction. The significance of the obtained results was judged at the 0.05% level.

**RESULTS**

Table (1) shows socio-demographic data of the studied patients. More than half of the studied PV patients (60%) had an age range between 50 and 60 years

Table 1: Distribution of the studied patients according to their sociodemographic data (n=100)

Sociodemographic data	No	%
Age		
20-	6	6.0
30-	14	14.0
40-	20	20.0
50- 60	60	60.0
Mean ± SD	43.38±7.45 years	
Gender		
Male	30	30.0
Female	70	70.0
Level of education		
Illiterate	20	20.0
Low education	65	65.0
High education	15	15.0
Occupation		
Administrative work	10	10.0
Manual work	25	25.0
Unemployed	65	65.0
Marital status		
Married	65	65.0
Divorced	5	5.0
Widow	15	15.0
Single	15	15.0
Residence		
Rural	60	60.0
Urban	40	40.0
Income (From patient's point of view)		
Sufficient	54	54.0
Insufficient	46	46.0

Table 2: Distribution of the studied patients according to their clinical data (n=100)

Clinical data	No	%
Family history		
No	90	90.0
Yes	10	10.0
Onset of disease		
≤ one year	10	10.0
> one year	90	90.0
Bo Body surface area affected		
Oral	35	35.0
Face and chest	15	15.0
Genital area	30	30.0
All over the body	20	20.0
Severity of the disease		
Mild	15	15.0
Moderate	35	35.0
Sever	50	50.0

with a mean of 43.38±7.45 years. The majority of the studied patients were females, had a low educational level and were unemployed (70, 65 & 65%) respectively. Additionally, the majority of the studied patients were married and rural residents and more than half of them had sufficient monthly income (65, 60 & 54%) respectively.

Table 3: Relationships of body surface area affected and onset of PV disease to WHOQOL-BREF scores (n=100)

Body surface area affected	WHOQOL-BREF scores				X <sup>2</sup>	P
	< 60 % Poor quality		≥ 60% Good quality			
	No.	%	No.	%		
Oral	30	30.0	5	5.0	32.61	<sup>MCP</sup> <0.0001*
Face and chest	12	12.0	3	3.0		
Genital	25	25.0	5	5.0		
All over the body	18	18.0	2	2.0		
Onset of PV disease						
≤ one year	8	8.0	2	2.0	76.82	<0.0001*
> one year	78	78.0	12	12.0		

Table 4: Relationships between severity of PV disease and WHOQOL-BREF scores of the studied patients (n=100)

WHOQOL-BREF scores	Severity of PV disease						X <sup>2</sup>	P
	Mild (n=15)		Moderate (n=35)		Sever (n=50)			
	No.	%	No.	%	No.	%		
General health								
• < 60 % Poor quality	10	10.0	25	25.0	30	30.0	14.010	<0.001*
• ≥ 60% Good quality	5	5.0	10	10.0	20	20.0		
Overall quality of life								
• < 60 % Poor quality	15	15.0	30	30.0	45	45.0	12.019	<0.001*
• ≥ 60 % Good quality	0	0.0	5	5.0	5	5.5		
Physical domain								
• < 60 % Poor quality	11	11.0	30	30.0	40	40.0	8.518	<0.001*
• ≥ 60 % Good quality	4	4.0	5	5.0	10	10.0		
Psychological domain								
• < 60 % Poor quality	15	15.0	18	18.0	35	35.0	7.681	<0.001*
• ≥ 60 % Good quality	0	0.0	17	17.0	15	15.0		
Social domain								
• < 60 % Poor quality	10	10.0	20	20.0	38	38.0	0.0555	0.960
• ≥ 60 % Good quality	5	5.0	15	15.0	12	12.0		
Environmental domain								
• < 60 % Poor quality	14	14.0	22	22.0	28	28.0	3.681	<0.001*
• ≥ 60 % Good quality	1	1.0	13	13.0	22	22.0		

Table (2) presents the distribution of the studied patients according to their clinical data. It was revealed that, the majority of studied patients (90%) had no family history of PV disease and had more than one year onset of disease. As regards the surface area affected, about two thirds of the studied patients had lesions in the oral and genital area. It can be noticed from the same table findings that (35%) had moderate degree of disease severity, whereas half of the studied patients (50%) had severe degree.

Table (3) displays the relationships of body surface area affected and onset of PV disease to WHOQOL-BREF scores. The table presents that, the majority of the studied patients who had poor QoL had oral (30 %) and genital (25%) areas of body surface area affected, whereas the

majority i.e. (78%) of them had a disease onset of more than one year. In the same context, high statistically significant relationships were detected between the body surface area affected, onset of PV disease and poor QoL of the studied patients where P<0.0001.

Table (4) shows relationships between severity of PV disease and WHOQOL-BREF scores of the studied patients. The table displays that, the majority of the studied patients had an overall poor quality of life in all domains: physical, psychological, social and environmental (81, 68, 68 and 64 %) respectively. Additionally, there were high statistical significant relations between patient's general health, overall quality of life, physical, psychological as well as environmental domains and the severity of disease where all p<0.001.

Table 5: Relationships between sociodemographic data of the studied patients and WHOQOL-BREF scores (n=100)

Socio-demographic data	WHOQOL-BREF scores				X <sup>2</sup>	P
	< 60 % Poor quality		≥ 60 % Good quality			
	No.	%	No.	%		
Age (years)						
20-	5	5.0	1	1.0	5.978	MCP=0.738
30-	10	10.0	4	4.0		
40-	10	10.0	10	10.0		
50-60	30	30.0	30	30.0		
Gender						
Male	20	20.0	10	10.0	13.503	0.039*
Female	50	50.0	20	20.0		
Educational level						
Illiterate	10	10.0	10	10.0	5.77	MCP =0.633
Low education	33	33.0	32	32.0		
High education	8	8.0	7	7.0		
Occupation						
Administrative work	8	8.0	2	2.0	2.513	MCP=0.497
Manual work	15	15.0	10	10.0		
Unemployed	38	38.0	27	27.0		
Marital status						
Married	35	35.0	30	30.0	1.55	MCP=0.459
Divorced/widow	9	9.0	11	11.0		
Single	8	8.0	7	7.0		
Residence						
Rural	40	40.0	20	20.0	10.41	MCP=0.789
Urban	30	30.0	10	10.0		
Income						
Sufficient	30	30.0	24	24.0	15.05	MCP=0.950
Insufficient	23	23.0	23	23.0		

Table (5) presents the relationships between sociodemographic data of the studied patients and WHOQOL-BREF scores. This table shows that, considerable numbers of the studied patients who had poor QoL were in the 50-60 years of age, were females, had low educational level, were unemployed, married, rural residents and had sufficient income (representing 30, 50, 33, 38, 35, 40 and 30%) respectively. No statistically significant relationships between QoL scores of the studied patients and all of their sociodemographic data was detected, except with the gender which had a statistically significant relation with QoL scores of the studied patients where P = 0.039.

### DISCUSSION

Pemphigus vulgaris is a serious chronic dermatological disease which can't be totally cured and will not usually settle without treatment. Therefore, much attention should be given by nurses for improving the QoL of patients with PV, thus better management of the disease could be secured [16, 31 & 32].

The main findings of the current study revealed that, the majority of the studied patients were between 50-60 years of age, females, with low educational level, married, unemployed and had rural residence. These findings were supported by Morsy *et al.* [32] who found similar findings in a previous study except for the marital status. Also, the majority of those patients were single. However, the present study findings are not in line with the British Association of Dermatologists leaflets [16] which indicated that, PV could affect males and females equally and at younger ages.

The current study findings showed that most of PV patients who had poor QoL were old unemployed married females. Also, the present findings revealed that there were no statistical associations between PV patients' QoL and their sociodemographic characteristics except gender. It was shown that the female sex was significantly correlated with poor quality of life of PV patients, since females are always preoccupied with their body images more than males. This finding was not supported by Ghodsi *et al.* [8] and Sung *et al.* [33] who found that QoL of patients with PV was not affected by sex of the patients

Regarding the clinical data of the studied PV patients, the results of the present study revealed that, the majority of patients had no family history of PV and very early onset of disease of more than one year. Moreover, most of the studied patients were found to have involved oral and genital areas as regards body surface area affected by the disease. Additionally, half of the studied PV patients had a severe degree of disease. These findings are consistent with Tamgadge *et al.* [1] who found that 70 - 90% of their cases had oral and genital mucosa as common sites of disease.

The present study demonstrated significant positive associations between poor QoL of the studied PV patients and body surface area affected as well as the early onset of disease. In the same line with our findings, Wardhana and Rusyati [7] who reported that, the majority of their studied patients had highly significant relations between poor QoL and body surface area affected especially in oral. However, contradicting to our findings, they cleared in their study that poor QoL of patients with PV was associated with recent disease onset of less than one year.

According to the current study main findings, there were significant negative impacts of PV on the majority of study patients' quality of life aspects as well as highly statistically significant associations between disease severity and patients' poor QoL. Moreover, the present study revealed that as the degree of disease severity is increased; as the studied patients with PV had poorer QoL in general health, overall quality of life and all of the WHO domains including physical, psychological and environmental except the social domain.

Similarly, Penha *et al.* [14] who found that, living with PV had affected the patients' general well-being; physical and psychological status and that moderate to severe cases were more affected than mild cases. However, the current study findings differ from their findings in that, they reported that, the majority of their studied patients had significant associations between their poor QoL and social relationships.

These results could be related to the disease restrictions on physical activity due to physical problems as painful blisters which might have led to restrictions on activities. No doubt poor psychological and emotional status might be related to the appearance of unattractive lesions on the skin, functional problems and disease chronicity. In the same context, Ghodsi *et al.* [8] reported that, higher level of anxiety and depression in these patients had a negative effect on QoL of PV patients.

On the other hand, the insignificant relations of PV WHOQOL-BREF scores to social domain could be attributed to that, the majority of the studied patients were unemployed females who had enough times can spend in social visits. Additionally, according to our Egyptian culture, the majority of people might prefer social relationships, even with having a disease like PV with an impaired body image. Thus, they might be socially interacted rather than being socially isolated. In addition, in our society, these patients could have a social support system which might encourage them to have social relationships and thus the studied patients had no affection on the social domain.

Dermatology nurses have a major role in PV patient education and planning nursing interventions aiming at reducing the consequences of PV and improving patients QoL. Also, they have the responsibility of encouraging PV patients to comply with their treatment and suggest lifestyle modifications that will potentially improve patients QoL.

## CONCLUSIONS

In conclusion, the present study findings document that PV had a significant negative impact on patients' quality of life. The majority of the studied patients had poor QoL in relation to general health, overall quality of life, physical, psychological and environmental domains. High statistical significant associations were found between severity of PV and all domains of patients' QoL except social domain.

### Recommendations:

- Standard of nursing care should be developed to improve the PV patients' QoL
- Rehabilitation programs are required to enhance PV patient's coping mechanisms and QoL.
- More research should be carried out in different areas in Egypt, to acquire more global understanding of the impact of PV on patients' QoL.

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