

Use of Skindex-29[©] to assess quality of life in patients with port-wine stain

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Abstract

Objective To determine the effect of facial port-wine stain (PWS) on quality of life (QoL) in our population with skin type III to IV, using the Skindex-29[©] scoring system.

Methods A total number of 69 patients were enrolled in this study. Patients were examined clinically and demographic data was collected. Patients were provided with Skindex-29[©] questionnaire with three independent domains affecting patient emotions, symptoms and functioning. Questionnaire had to be completed by patients themselves or with the help of an interpreter. The specific scoring system was used with a scale from 1-100; higher score indicated a greater impact of skin disease.

Results Facial PWS had a moderately negative impact on QoL of the patient. Subscores calculated for symptoms were 8.17 ± 6.70 , emotions 25.72 ± 7.83 , and functions 21.00 ± 11.34 . Composite score was 19.53 ± 11.26 . These scores indicate that the greatest negative effect was on emotions, followed by functions and then symptoms. There was no significant difference in scores between the sexes. A greater area of involvement was significantly associated with higher emotional ($p=0.04$) and symptoms score ($p=0.003$). Deeper colour of PWS was associated with more symptoms ($p=0.01$).

Conclusion Our study indicates that facial PWS has negative impact on a patient's QoL, having more affect on emotions, followed by functions. Males and females were equally affected in all domains by PWS. Impact on emotions and symptoms is directly proportional to the size of PWS.

Key words

Port-wine stain (PWS), quality of life, Skindex-29[©].

Introduction

Port-wine stains (PWSs) are congenital, slow-flow, capillary vascular malformations, characterized by normal epidermis overlying abnormal plexus of dilated dermal blood vessels.¹ PWSs are present at birth, with incidence of 0.3% and do not disappear spontaneously.²

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In newborn babies, the lesions typically appear as light red macules and tend to grow proportionally with age. PWSs thicken and darken, to deep red or purple, due to progressive ectatic dilation of the blood vessels.^{2,3}

In the fourth decade of life, two-thirds of affected individuals develop soft tissue overgrowth and nodules, causing disfigurement, asymmetry, and spontaneous bleeding.^{4,5} Early intervention is often the optimal treatment of choice.^{3,6}

PWSs are generally considered to be asymptomatic but due to their location on the

face, they cause a significant cosmetic problem and may adversely affect a person's psychosocial well-being.⁷

Effects of skin disease on social well-being has been researched and debated over decades.⁸⁻¹⁴ It is impossible to separate impact of skin diseases from an individual's personal and social context.

The work of Finlay *et al.*⁹ in creating awareness among dermatologists about impact of skin disease on the patient's daily life, has been significant. They created the first dermatology disease-specific quality of index (QoL) index, Psoriasis Disability index, which was published in 1987 only after quite a few rejections as it was a different concept of looking at skin disease and so unfamiliar that it was viewed as being of no importance.⁸ They then developed Dermatology Life Quality Index (DLQI),¹⁰ which could be used across the board in different dermatological disorders. This sprouted development of other disease-specific indices,^{8,11-13} and further refinement.^{14,15}

Chren *et al.*¹⁶ first developed Skindex as a 62-question survey in 1996. It was then further refined into the Skindex-29 and then Skindex-16.¹⁷ The questionnaire tackles three domains of a patient's life: symptoms, emotions, and functions. It gives the opportunity to assess the burden of skin disease separately on different aspects of a patient's daily life.

The severity of skin disease is related both to its clinical extent (assessed using 'clinimetric measures') and its effects on a patient's quality of life (using 'psychometric' measures).¹⁵ Only then can we hope to understand the burden of skin disease on the patient.

Our objective was to assess the effect of facial PWSs on a patient's QoL using a validated, standardized, dermatology-specific instrument,

the Skindex-29. Also to record independent demographic and clinical factors which may influence it.

Methods

It was an observational and questionnaire-based study. 69 patients of PWS were enrolled in the study, after informed consent and data were recorded on a predesigned proforma.

Criteria for eligibility were age of 13 years or above, a facial PWS, and the ability to read, understand and answer the questionnaire independently, or if not literate, with a healthcare professional reading it out and recording the answer. Demographic data i.e. age, sex, relationship status, level of education, number of close friends, frequency of socializing, were recorded. Clinical data i.e. whether the PWS was unilateral or bilateral, percentage of body surface area involved (one palm = 1% total body surface area), colour and texture of the involved skin were noted. Patients were requested to fill out the Skindex-29 questionnaire (either in English or Urdu translation).

The main outcome measure of the survey was Skindex-29. It is a comprehensive questionnaire comprising of 29 scrambled questions tackling three domains of a patient's life. Some questions were related to symptoms (e.g. itching, burning, irritation, bleeding, sensitivity), others to emotions (e.g. worry, depression, shame, embarrassment, frustration, humiliation) and to functions (sleep, work and hobbies, social life, doing things, interaction with others).

Items were converted into a scale from 1 to 100. Domain score and a composite score were calculated by the recommended scoring procedures. Values proposed by Nijsten *et al.* were used to quantify QoL with Skindex-29. i.e.

very little, mild, moderate, severe, and very severe. Higher scores indicated a greater impact of skin disease on QoL.

Data were entered and analyzed according to SPSS version 20, and 'R'. Descriptive statistics were used to analyze the survey sample. Independent variables were demographics, including age, gender and educational level, socialization with others and clinical severity and treatment of PWS. Statistical analysis showing a *P* value of less than 0.05 was considered significant.

Results

A total number of 69 individuals were included in the study (**Table 1**). Their demographic data showed that majority were female (75.4%), ages ranged from 13 to 50 years and mean age was 23.6±7.32 years. A majority of patients presenting were graduates or had a professional degree (59.4%), only one patient (1.4%) was not educated, 48 (69.6%) were single, 14 were married (20.3%), while 7 were engaged or had proposals (10.1%).

Only 4 (5.7%) patients claimed to have no close friends, while 38 (55.1%) patients had one to three close friends. Socializing patterns showed a majority socialized up to once a week, 25 (36.2%) and 24 (34.8%) patients (cumulative 71%), and 15 (21.7%) patients socializing one to three times a week.

PWS disease characteristics (**Table 2**) showed that 66 were unilateral, only 3 were bilateral. Area of involvement (one palm = 1% of body surface area) was 1% or less in 46 patients, 2% in 14 patients and 9 patients had a greater involvement.

Skindex-29 scores showed (**Table 3**) that presence of facial PWS had a moderately

negative influence on QoL. Subscores for symptoms was 8.17±6.70, for emotions 25.72±7.83, and for functions 21.00±11.34. Composite score was 19.53 (S.D.=11.26). These scores indicate that the greatest negative effect was on emotions, followed by functions and then symptoms.

Univariate associations showed that there was no statistically significant difference in scores between males and females (**Table 3**). A greater area of involvement was significantly associated with higher emotional (*p*=0.04) and symptoms score (*p*=0.003). Deeper colour of PWS was associated with more symptoms (*p*=0.01).

Table 1 Port-wine stain patients demographics (n=69).

| | N (%) |
|---------------------------------|-----------|
| Age (years) | |
| 13-15 | 10 (14.5) |
| 16-20 | 16 (23.2) |
| 21-30 | 35 (50.7) |
| 31-40 | 6 (8.7) |
| >40 | 2 (2.9) |
| Sex | |
| Male | 17(24.6) |
| Female | 52(75.4) |
| Level of education | |
| None | 1 (1.4) |
| Primary | 12 (17.4) |
| Matric | 12 (17.4) |
| Some college | 3 (4.3) |
| Graduate or professional degree | 41 (59.4) |
| Relationship status | |
| Single | 48 (69.6) |
| Proposed | 3 (4.3) |
| Engaged | 4 (5.8) |
| Married | 14 (20.3) |
| Divorced | 0 (0) |
| Widowed | 0 (0) |
| No. of close friends | |
| 0 | 4 (5.7) |
| 1-3 | 38 (55.1) |
| 4-6 | 25 (36.2) |
| 7-9 or more | 2 (2.9) |
| Frequency of socializing | |
| <1 x per week | 25 (36.2) |
| 1 x per week | 24 (34.8) |
| 2-3 x per week | 15 (21.7) |
| 4-5 x per week | 4(5.8) |
| 6 or more x week | 1(1.4) |

Table 2 Port-wine stain characteristics (n=69).

| | N (%) |
|--------------------------------------------------|-----------|
| <i>Area of facial, head and neck involvement</i> | |
| 0.5% | 22 (31.9) |
| 1% | 24 (34.8) |
| 2% | 14 (20.3) |
| 3% | 6 (8.7) |
| >3% | 3 (4.3) |
| <i>Distribution</i> | |
| Unilateral | 66 (95.7) |
| Bilateral | 3 (4.3) |
| <i>Color</i> | |
| Pink | 19 (27.5) |
| Red | 38 (55.1) |
| Purple | 12 (17.4) |
| <i>Skin texture</i> | |
| Normal | 33 (47.8) |
| Papules | 25 (36.2) |

| | |
|----------------------------------|-----------|
| Nodules | 11 (15.9) |
| <i>Treatment period</i> | |
| <6 months | 4 (5.8) |
| <2 years | 34 (49.3) |
| <5 years | 21 (30.4) |
| >5 years | 10 (14.4) |
| <i>Age of starting treatment</i> | |
| <10 years | 2 (2.9) |
| 11-15 years | 18 (26.1) |
| 16-20 years | 15 (21.7) |
| 21-30 years | 30 (43.5) |
| >30 years | 4 (5.8) |
| <i>Satisfied with treatment</i> | |
| Yes | 67 (97.1) |
| No | 2 (2.9) |

Table 3 Univariate associations* of measures with Skindex-29 responses

| Variable | Emotions | | | | Symptoms | | | | Functioning | | | |
|--------------------|----------|------------|---------|----------------------|----------|------------|---------|----------------------|-------------|------------|---------|----------------------|
| | Beta | Std. Error | p Value | R ² value | Beta | Std. Error | p Value | R ² value | Beta | Std. Error | p Value | R ² value |
| Age (years) | 0.039 | 0.322 | 0.904 | 0.0002 | 0.092 | 0.178 | 0.605 | 0.0039 | 0.318 | 0.239 | 0.188 | 0.025 |
| Size of PWS (area) | 692 | 331.5 | 0.04 | 0.06 | 545 | 177.6 | 0.003 | 0.123 | 17.24 | 258 | 0.946 | 6.66 |
| Sex | | | 0.415 | | | | 0.155 | | | | 0.441 | |
| Education | | | 0.874 | | | | 0.424 | | | | 0.764 | |
| Friends | | | 0.917 | | | | 0.5 | | | | 0.0177 | |
| Socializing | | | 0.494 | | | | 0.114 | | | | 0.244 | |
| Color | | | 0.608 | | | | 0.00152 | | | | 0.244 | |

* Linear regressions/ANOVA models

Table 4 Comparison of Skindex-29 scores in port-wine stain (PWS) with other international studies and dermatological diseases.

| Diseases/authors | N | Symptoms | Emotions | Functioning | Composite |
|-------------------------------------|-----|-------------|-------------|-------------|-----------|
| CLE [18] | 178 | 41.3 (23.8) | 49.1 (27.8) | 28.4 (25.6) | 39.6 |
| Psoriasis [19] | 44 | 42.0 (21.0) | 39.0 (27.0) | 23.0 (27.0) | 34.7 |
| Acne Vulgaris [20] | 63 | 30.0 (19.0) | 41.0 (25.0) | 16.0 (16.0) | 29.0 |
| Vitiligo [21] | 245 | 13.9 (14.6) | 35.9 (23.6) | 16.7 (19.5) | 22.2 |
| Facial PWS, Hagen <i>et al.</i> [7] | 244 | 14.9 (18.4) | 34.4 (25.8) | 24.3 (22.3) | 24.6 |
| Facial PWS, Akbar <i>et al.</i> | 69 | 8.17 (6.70) | 25.7 (7.8) | 21.0 (11.3) | 19.5 |
| No skin disease [21] | 107 | 14.0 (12.0) | 9.0 (13.0) | 4.0 (8.0) | 9.0 |

CLE: cutaneous lupus erythematosus.

Discussion

Port-wine stains are congenital vascular malformations most commonly occurring on the face. Being highly visible, PWS may have a negative effect on health-related QOL and psychological adjustment in an individual. This study was conducted to assess the disease burden, by applying a validated, dermatology specific QoL instrument, the Skindex-29.

In our study, majority were females (n=52). This may reflect the greater concern PWS causes in females and their families, so greater efforts are made for treatment.

Skindex scores did not show a significant difference between the males and females, which indicates that males were as emotionally and functionally affected as females, contrary to

the study by Hagen *et al.*⁷ where females were more affected.

The mean age of presentation was 23.6 years, and a majority started treatment in the third decade of life (49% were over 21 years). Current recommendations are for earlier treatment, even in infancy.^{5,6} When treatment is delayed for months and years, hypertrophy with papules and nodules develop, making the PWS more refractory to treatment with lasers. It may also lead to a negative impact during school life, with bullying. Treatment should be started in infancy due to rapid response, which will ultimately improve the burden of disease in the patient. This study also indicates that we need to educate our medical personnel and the general population for early referral to specialized centres.

The level of education made no statistically significant difference to QoL scores, according to the survey. However, a greater number of graduates or professional people presented for treatment (59%). This may indicate that people with higher education are more aware of advanced treatments available.

Marital status analysis revealed that most of our patients were unmarried (69.6%). This may indicate that in our society, PWS may have a negative impact on chances of marriage, so prompts recourse to treatment.

Majority of patients (71%) only socialized up to once a week, and had one to three close friends (55%). Our people mostly live in joint family systems, so have close relationships and support within their families, this may reflect why they need less socializing. Skindex scores did not show a significant relationship between number of friends and emotional or symptom scores, but did show co-relationship with higher functioning scores.

Skindex-29 scores showed that patients with facial PWS had a significant negative impact on QoL, affecting emotions the most, followed by functions and symptoms. This correlates well with the study by Hagen *et al.*⁷, which showed similar effects (**Table 4**). Our results indicated less negative impact than that carried out in the USA. We need more studies in other diseases to define QoL scores in our population, and our country.

The QoL scores for emotions and symptoms were significantly more negatively affected with greater area of involvement; deeper colour of the lesion was significantly associated with greater symptoms. A deeper colour indicates a deeper tissue involvement, which has a greater tendency to hypertrophy, causes more symptoms and is less responsive to laser treatment.

In comparison to other dermatological diseases studied internationally, the quality of life scores in our study (composite score: 19.53) was lower than those without skin disease (composite score: 9), indicating a greater negative effect on QoL. Functioning (21.0) was more negatively affected than in acne vulgaris (16.0) and vitiligo (16.7), and similar to that of psoriasis (23.0). Emotional subscores showed a moderately negative influence (25.7), though less than Hagen *et al.*⁷ (34.4).

Conclusion

The presence of a PWS negatively affects the QoL of the patient, mostly in the emotional domain, followed by functions. This should encourage dermatologists to give consideration to QoL in highly visible skin diseases that are relatively asymptomatic, and provide necessary psychological support services to patients where needed. Health and public policy decision-making should give due importance to such aspects.

We need to spread consciousness towards earlier treatment of the condition, as our patients present rather late for treatment.

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