Acta Derm Venereol 2014; 94: 344–346

SHORT COMMUNICATION

Factors Associated with Impaired Quality of Life in Adult Patients Suffering from Ichthyosis

Isabelle Dreyfus[1], Emmanuelle Bourrat[2], Annabel Maruani[3], Didier Bessis[4], Christine Chiaverini[5], Pierre Vabres[6], Khaled Ezzedine[7] and Juliette Mazereeuw-Hautier[1]

[1]Reference Centre for Rare Skin Diseases, Dermatology Department, Larrey Hospital, CHU Toulouse, FR-31059 Toulouse, Dermatology Departments, [2]Saint-Louis Hospital, AP-HP, Paris, [3]CHU Tours, [4]CHU Montpellier, [5]CHU Nice, [6]CHU Dijon, and [7]CHU Bordeaux, France. E-mail: dreyfus.i@chu-toulouse.fr
Accepted June 10, 2013; Epub ahead of print Oct 24, 2013

Inherited ichthyoses are genetic disorders of cornification characterised by scaling of various aspects and severity, often associated with erythroderma (1). The skin is usually uncomfortable, pruriginous and painful and no specific treatment is available. Impact on quality of life (QoL) was previously demonstrated (2–5). QoL is a dynamic process that can be defined as an individual perception of one's position in life (6). In order to improve patient's management, the aim of our study was to determine the impact of weighted factors associated with impaired QoL in adult ichthyosis patients.

MATERIALS AND METHODS

This was designed as a multicentre prospective study. Ethical review was not required by our institution. The procedures followed were in accordance with the Helsinki Declaration of 1975, revised in 1983. QoL was measured using the Dermatology Life Quality Index (DLQI) questionnaire, which included 10 questions concerning 6 areas of the patients' lives: "symptoms and feelings", "daily activities", "leisure", "work", "personal relationships" and "treatment" (7). Patients were also asked to fill in a questionnaire exploring their characteristics and the severity of their ichthyosis (erythema, scales, pruritus, cutaneous pain and ocular symptoms assessed using 5 Visual analogue scales (VAS) ranging from 0 to 10; global disease severity listed as mild/moderate/severe/very severe).

These questionnaires were anonymous and sent to all patients (≥ 16 years old) with a confirmed diagnosis of inherited ichthyosis (ichthyosis vulgaris was the only excluded form), who were either followed-up at the 11 French Dermatology Departments for rare skin diseases or affiliated to patient's support group, AIF. Patient's initials were checked to avoid duplicate responses. Two DLQI subgroups of patients were individualised regarding the effect on QoL: DLQI ≤ 10 (mild or moderate effect) and DLQI > 10 (severe or very severe effect) (8). A logistic regression analysis was performed to determine the impact of weighted factors associated with a QoL impairment (DLQI score > 10). The model included age, gender, ichthyosis's severity, cohabiting status, AIF membership status and academic degree. These categories were identified as key factors in the modulation of QoL in a recent qualitative study using focus groups (4). All these potential predictors of impaired QoL were firstly assessed individually, and odds ratio (OR), the corresponding 95% confidence intervals (CIs) and *p*-values were computed. The OR significance was determined by Wald chi-square test, and predictors with $p < 0.10$ were subsequently assessed using multivariate analysis with a forward stepwise selection procedure (multivariate analysis only concerned patients for whom all data were available, i.e. no missing information when pooling data together). Coefficient of determination R^2-adjusted represented the proportion of variability explained by the model. All statistical analyses were performed using STATA version 11.0.

RESULTS

A total of 171 questionnaires were completed and returned (response rate: 49%) among whom 158 were analysable. Characteristics of the study population are described in Table I. The study population was young and included a majority of females. Nearly 2/3 were suffering from moderate to severe ichthyosis (no subtyping made) and most patients were regularly followed by a physician.

The mean score of DLQI was 8.3 ± 6.5 (0–27). A DLQI score > 10 was noted for 31% of the patients. All the 6 DLQI's areas were affected by ichthyosis, the strongest impairment being on "symptoms and feelings" (86% of patients). The items "daily activities", "treatments", "work", "leisure", and "personal relationships" were affected in 77%, 62%, 59%, 55% and 45% of the patients, respectively.

Using univariate analysis (Table II), we found that all VAS scales and gender (female) were statistically associated with a DLQI score > 10. In addition, the fact that a patient lives alone seemed to be associated with DLQI score > 10. Multivariate analysis was performed on 133 of the 158 patients. After adjustment, in multivariate analysis (Table III), cutaneous pain emerged as the most significant factor influencing the value of the DLQI score, followed by scales and gender (female).

DISCUSSION

This study demonstrated a severe or very severe effect on QoL for 1/3 of the patients. It also identified a more

Table I. *Characteristics of the study population*

	All patients ($n = 158$)
Age, years, mean ± SD (range)	40 ± 19.1 (1–88)
Male/Female, %	44.9/55.1
Regularly followed by a physician, %	87.3
Member of the patient's support group (AIF), %	60.8
Global disease severity, %	
Mild	21
Moderate	36.3
Severe	30
Very severe	12.7
Cutaneous features (VAS/10)	
Score of erythema, mean ± SD (range)	2.75 ± 2.3 (0–8.9)
Score of scales, mean ± SD (range)	4.89 ± 2.9 (0–10)
Troublesome symptoms related to skin conditions (VAS/10)	
Score of pruritus, mean ± SD (range)	4.14 ± 2.9 (0–10)
Score of cutaneous pain, mean ± SD (range)	3.88 ± 3.2 (0–10)
Score of ocular troubles, mean ± SD (range)	3.31 ± 3.3 (0–10)

 doi: 10.2340/00015555-1710
 ISSN 0001-5555

Table II. *Determination of the factors specifically related to Dermatology Life Quality Index >10: results of the univariate regression analysis stepwise*

	Odds ratio (CI 95%)	*p*-value
Erythema (>5/10)[a]	2.83 (1.28–6.25)	0.01
Scales (>5/10)[a]	3.24 (1.58–6.66)	0.001
Pruritus (>5/10)[a]	3.61 (1.75–7.44)	0.001
Ocular troubles (>5/10)[a]	2.79 (1.34–5.83)	0.006
Cutaneous pain (>5/10)[a]	4.31 (2.09–8.9)	$<10^{-4}$
Gender (female)	2.10 (1.04–4.25)	0.039
Cohabiting status (cohabiting)	0.47 (0.22–1.01)	0.05
Age (>40 years old)	0.62 (0.30–1.25)	0.179
AIF membership status (affiliated)	0.56 (0.28–1.10)	0.09
Academic degree[b]	0.98 (0.48–1.99)	0.956

[a]VAS score >5. [b]French high-school diploma or more.

vulnerable population: women with severe scales and pain. These results are in accordance with our previous qualitative study (4) and with Kamalpour et al. (5), who investigated QoL of patients with ichthyosis using DLQI scores. Nevertheless, our methodology was quite different. Contrary to our study, Kamalpour et al. (5) did not differentiate between adults and children. Furthermore, they restrictively used correlation tools with no formal measurement of the risk accounting for severe impairment of QoL, i.e. generated OR. Our study is, to the best of our knowledge, the first study using univariate and multivariate logistic regression procedures in order to determine the impact of weighted factors associated with impaired QoL in adult patients with inherited ichthyosis.

Our study had several limitations related to the use of an anonymous and self-administered questionnaire: the characteristics of the non-responders were not available, the clinical form of ichthyosis the patient is suffering from could not be determined (although ichthyosis vulgaris was excluded) and the severity grading is not objective (especially for evaluation of scales and erythroderma). With regard to response rate, it is slightly lower than the mean response rate of 54% reported for published medical surveys (9).

Our studied population was not exhaustive but can be considered as representative since it comprised patients with various characteristics, either seen in hospital or members of AIF. Estimation of QoL was done through the DLQI, a validated and easy to use tool.

The factor identified as having the most important impact on QoL in our study was cutaneous pain. This is in accordance with the fact that "symptoms and feelings" was the most impacted item. Skin diseases are considered disabling mainly because of the aspect of the skin. Cutaneous pain is somewhat neglected, especially as analgesics are usually not effective. With regards to gender, its influence on QOL is not a common pattern, even if it was previously reported in hand eczema (10) and other chronic non-dermatological disease such as cardiac disease (11). In psoriasis, gender was demonstrated to influence QOL via higher anxiety and depression (12). Erythema, pruritus, ocular troubles and cohabiting status were supposed to be less important factors since they were only identified in univariate analysis but they have certainly to be considered in QOL impairment.

Table III. *Determination of the factors specifically related to Dermatology Life Quality Index (DLQI) >10: results of the multivariate regression analysis stepwise (*n = *133)*

	DLQI (R^2 = 15.2%)	
	Odds ratio [CI 95%]	*p*-value
Cutaneous pain	2.93 [1.23–7.01]	0.028
Scales	2.76 [1.11–6.82]	0.009
Women	2.71 [1.15–6.41]	0.023

In conclusion, we identified a more vulnerable population on which strategies to improve patient's management (ie. education programs) must be focused on. Evaluation of cutaneous pain should be included to ichthyosis assessment and research should turn towards novel drugs for scales and cutaneous pain reduction.

ACKNOWLEDGEMENTS

We thank the patients, M-C Verly and the support group AIF, A-C. Bursztejn, S. Mallet and S. Barbarot for their important contribution to this work.

REFERENCES

1. Oji V, Tadini G, Akiyama M, Blanchet Bardon C, Bodemer C, Bourrat E, Coudière P, et al. Revised nomenclature and classification of inherited ichthyoses: results of the First Ichthyosis Consensus Conference in Sorèze 2009. J Am Acad Dermatol 2010; 63: 607–641.
2. Gånemo A, Lindholm C, Lindberg M, Sjödén PO, Vahlquist A. Quality of life in adults with congenital ichthyosis. J Adv Nurs 2003; 44: 412–419.
3. Gånemo A, Sjödén PO, Johansson E, Vahlquist A, Lindberg M. Health-related quality of life among patients with ichthyosis. Eur J Dermatol 2004; 14: 61–66.
4. Mazereeuw-Hautier J, Dreyfus I, Barbarot S, Serrentino L, Bourdon-Lanoy E, Ezzedine K, et al. Factors influencing quality of life in patients with inherited ichthyosis: a qualitative study using focus groups. Br J Dermatol 2012; 166: 646–648.
5. Kamalpour L, Gammon B, Chen KH, Veledar E, Pavlis M, Rice ZP, Chen SC. Resource utilization and quality of life associated with congenital ichthyoses. Pediatr Dermatol 2011; 28: 512–518.
6. What's quality of life? The WHOQOL Group. World Health Organization Quality of Life Assessment. World Health Forum 1996; 17: 354–356.
7. Finlay AY, Khan GK. Dermatology Life Quality Index (DLQI) – a simple practical measure for routine clinical use. Clin Exp Dermatol 1994; 19: 210–216.
8. Hongbo Y, Thomes CL, Harrison MA, Salek MS, Finlay AY. Translating the science of quality of life into practice: what do dermatology life quality index scores mean? J Invest Dermatol 2005; 125: 659–664.
9. Asch DA, Jedrziewski MK, Christakis NA. Response rates to mail surveys published in medical journals. J Clin Epi-

demiol 1997; 50: 1129–1136.
10. Wallenhammar LM, Nyfjäll M, Lindberg M, Meding B. Health-related quality of life and hand eczema – a comparison of two instruments, including factor analysis. J Invest Dermatol 2004; 122: 1381–1389.
11. Emery CF, Frid DJ, Engebretson TO, Alonzo AA, Fish A, Ferketich AK, et al. Gender differences in quality of life among cardiac patients. Psychosom Med 2004; 66: 190–197.
12. Sampogna F, Chren MM, Melchi CF, Pasquini P, Tabolli S, Abeni D. Age, gender, quality of life and psychological distress in patients hospitalized with psoriasis. Br J Dermatol 2006; 154: 325–331.