

Study of Correlation of Severity and Quality of Life in Patients with Congenital Ichthyosis

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Abstract

Introduction: Congenital ichthyosis is a disorder of cornification of skin with various cutaneous symptoms. Quality of life is a subjective perception of patients regarding their disease status and treatment received. Dermatological disorders have poor quality of life associated with them as any other systemic disease. The present study was conducted to assess the quality of life among patients of congenital ichthyosis, so that suitable remedial measures may be devised based on the same.

Materials and Methods: It was a hospital based cross-sectional study with total 120 patients enrolled for study duration of one year. Childrens' Dermatology Quality Life Index (CDLQI) questionnaire was used to assess the quality of life among patients. Global disease severity scale and VAS scale for severity of ichthyosis was used along with recording of socio-demographic details.

Results: With a response rate of 87%, the sample size comprised of 104 subjects, aged between 0-15 years. Females outnumbered the sample by more than 2/3rd. 'Symptoms and feelings' was the most affected domain in the CDLQI. Itching, female gender and presence of scales were significant predictors of poor quality of life scores.

Conclusion: The present study shows the epidemiological correlation of patients with congenital ichthyosis and the predictors of poor quality of life score. Regular subjective assessment of quality of life among patients should be included in routine clinic visits, as counselling plays a very important role in these patients.

Key words: Congenital ichthyosis, Disease severity, Quality of Life

Introduction

Congenital ichthyosis is disorder of cornification associated with systemic symptoms.¹ There is production of abnormal quality and quantity of scales as well as abnormal thickness of stratum corneum with presence of skin inflammation. Treatment of ichthyosis revolves around hydration, lubrication and keratolysis. Various studies have been conducted in past regarding quality of life (QoL) in patients of ichthyosis.²⁻⁵ Quality of life is defined as "perspective of patient/subject towards one's position in life and how the patients sees his treatment or disease".⁶ The current study was done to know the quality of life of patients with congenital ichthyosis and the role of socio-demographic profile factors in these patients.

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Materials and Methods

This study was a hospital based cross sectional study. The study was conducted from June 2017 to May 2018 after taking approval from Institutional research and ethics committee and consent from the parents of subjects included in the study. Quality of Life (QoL) was measured using Childrens' Dermatology Life Quality Index (CDLQI) questionnaire for paediatric

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patients and Dermatitis Family Impact Questionnaire (DFI) for family members. CDLQI included 10 questions concerning 6 areas of the patients' lives: "symptoms and feelings", "daily activities", "leisure", "work", "personal relationships" and "treatment". For children less than 10 years of age the questionnaires were filled by the parents. Each question has four alternative responses: 'not at all', 'only a little', 'quite a lot' and 'very much' with corresponding scores as 0, 1, 2 and 3 respectively. The CDLQI score is calculated by summing up individual question resulting in maximum score of 30 and minimum score of 0. The greater the CDLQI score, higher the impairment in the quality of life of patients. The Dermatitis Family Impact Questionnaire contains 10 questions which measures influence of children's disease on different fields of life of their family members. The greater the score, higher the impairment in the quality of life of patients/parents. The assessment questionnaire was filled for the last one week prior to the visit to the hospital.

Inclusion criteria were patients with clinical diagnosis of congenital ichthyosis. A total of 120 patients were enrolled for the study. Global disease severity scale was also used to label the severity of disease as mild/moderate/severe/very severe. Sociodemographic details of the patients was asked when taking history which were age, income, education, occupation and distance to travel for physician compliance visits. Patients were also asked to fill in a questionnaire exploring their characteristics and the severity of their ichthyosis (erythema, scales, pruritus, 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 self-administered and filled by all patients (≥ 10 years old) with a confirmed diagnosis of inherited ichthyosis. For patients <10 years, parents were asked to fill the questionnaire. For illiterate patients, investigators read out the questionnaire and recorded the response. Two subgroups of patients were made to see the effect on QoL: CDLQI ≤ 10 (mild or moderate effect) and CDLQI > 10 (severe or very severe effect). The questionnaires were translated in local vernacular language (English/Hindi) and the translation was validated by an independent researcher.

Statistical analysis

Data was entered in Microsoft excel spreadsheet and analysed using SPSS (Statistical Package for Social Sciences) for Windows version 17.0. Categorical data was analysed using Chi-square test and normally

distributed quantitative data was analysed using independent sample t-test. Binary logistic regression analysis was performed to determine the impact of factors associated with a QoL impairment (CDLQI score > 10). Point of statistical significance was considered when p-value was less than 0.05 ($p < 0.05$).

Results

A total of 104 questionnaires were completed and returned (response rate:87%) Characteristics of the study population are described in Table 1. The study population was young in the age of 0-15 years comprising 68% and had female majority. On exploring the history of the patients, it was observed that majority of the parents of the cases had a consanguineous marriage (75.0%), which highlights the importance of need of creating awareness in society regarding genetic diseases. A positive history of collodion baby (17.3%) was observed and 9.6% cases reported history of erythroderma, bullous lesion, recurrent infections in the family. Most common form of ichthyosis diagnosed was ichthyosis vulgaris (64%), followed by non-bullous ichthyosiform erythroderma (32%) and lamellar form (11%) which gives a clue about the pattern of the disease.

Moderate to severe ichthyosis was present in about 2/3rd of the patients and most patients were regularly following a physician's advice. The mean score of CDLQI was 8.9 ± 5.5 (0–28). A CDLQI score > 10 was noted in 36% of the patients. Further the patients were segregated according to their diagnosis to see the effect of severity of disease on the domains of CDLQI. Most domains of CDLQI's areas were affected by ichthyosis, the strongest impairment being on "symptoms and feelings" (78% of patients), thereby highlighting the need for counselling of these patients, along with adequate bonding with the caregivers/peers. Patients with ichthyosis frequently complained of severe pruritus, scaling, tightness of skin, erythema and ocular problems and frequency varied according to the type of ichthyosis as depicted in Table 2. It was observed that most common complaints were pruritus and tightness of skin. Ocular complications were maximum in patients diagnosed with Lamellar Ichthyosis, the management of which should be given utmost importance in order to prevent long-term morbidity. Table 2 depicts the proportion of the patients affected with different diagnosis of ichthyosis. It was seen that ichthyosis had a large effect on the quality of life amongst majority of the patients. Maximum proportion of patients who were extremely affected by the disease were in the lamellar ichthyosis

group (66%) followed by Ichthyosis vulgaris group. Most of patients diagnosed with Lamellar ichthyosis were suffering in most of the domains. (Table 3) The items “daily activities”, “treatments”, “work”, “leisure”, and “personal relationships” were affected in 67%, 76%, 58%, 54% and 48% of the patients, respectively. Global disease severity is depicted in Figure 1.

All VAS scales for severity and female gender were significantly associated with a CDLQI score > 10 on univariate analysis. On binary logistic regression, itching was a significant factor influencing the value of the CDLQI score, followed by scales on the body and gender (female).

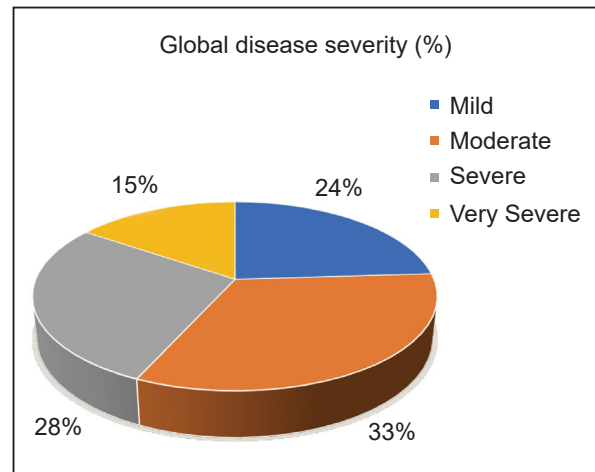


Figure 1: Pie chart showing distribution of subjects with global disease severity scale.

Table 1: Most common reported complaints amongst the patients diagnosed with different types of Ichthyosis

Type of ichthyosis	Pruritus	Scales	Tightness of skin	Erythema	Ocular problems	Total
Ichthyosis vulgaris	56 (90.3)	53 (85.5)	34 (54.8)	17 (27.4)	5 (8.1)	62 (100)
Non-bullous ichthyosiform erythroderma	14 (77.8)	15 (83.3)	9 (50)	12 (66.7)	1 (5.6)	18 (100)
Lamellar ichthyosis	10 (100)	10 (100)	10 (100)	7 (70)	4 (40)	10 (100)
X-lined Recessive	11 (78.6)	11 (78.6)	6 (42.9)	10 (71.)	0	14 (100)
Total	90 (86.5)	89 (86.5)	58 (55.8)	47 (45.2)	10 (9.6)	104 (100)

Table 2: Quality of life amongst the patients diagnosed with different types of Ichthyosis based upon the CDLQI scores

Type of Ichthyosis	No or small effect	Moderate effect	Large effect	Extremely large effect	Total
Ichthyosis vulgaris	2 (3.2)	5 (8.1)	40 (64.5)	15 (24.2)	62 (100)
Non-bullous Ichthyosiform erythroderma	1 (5.6)	1 (5.6)	12 (66.7)	4 (22.2)	18 (100)
Lamellar ichthyosis	0	1 (16.7)	3 (16.7)	6 (66.7)	10 (100)
X-lined Recessive	1 (7.1)	8 (57.1)	4 (28.6)	1 (7.1)	14 (100)
Total	4 (3.8)	19 (18.2)	57 (54.8)	24 (23)	104 (100)

Table 3: Distribution of patients affected according to different DLQI subsets

	Ichthyosis vulgaris (n=62)	Non bullous ichthyosiform erythroderma (n=18)	Lamellar ichthyosis (n=10)	X-lined Recessive (n=14)	Total (n=104; 100%)
Symptoms and feelings	34 (54.8)	20 (64.5)	10 (100)	11 (78.6)	81 (78%)
Daily activity	25 (40.3)	18 (58.1)	10 (100)	10 (71.4)	70 (67%)
Treatments	17 (27.4)	14 (45.2)	6 (60)	11 (78.6)	79 (76%)
Work/School	28 (45.2)	17 (54.8)	8 (80)	9 (64.3)	60 (58%)
Leisure	33 (53.2)	18 (58.1)	10 (100)	7 (50)	56 (54%)
Personal relationship	30 (48.4)	18 (58.1)	10 (100)	11 (78.6)	50 (48%)

Discussion

This study was conducted to see the effect of congenital ichthyosis on the quality of life of the patients and its correlation with severity of the disease. The response rate was fairly high (87%) with 104 subjects completing the questionnaires. The mean age of parents was 32.42 years with median age 30 years. Females were in majority with proportion of 56.7% whereas a study by Ghosh et. al revealed that 65% were males and rest 35% were females in their study.⁷ In our study, 74% of patients had regular follow-up visits to the physician whereas Isabelle et. al had reported physician visit of 87% which was higher as compared to our study. This might be due to residence of patients in far flung areas and lack of specialist dermatology physicians in the vicinity. More than two third of the subjects' parents had education higher than secondary. About 20% of subjects had monthly income of less than 10,000 rupees corresponding to lower literacy status (below primary) among them. Due to this, the long term prognosis and compromised skin status of their child was difficult for the family to comprehend.

In 1/3rd of patients, severe or very severe effect on QoL was seen which is very similar to Isabelle et. al. Previous studies have also reported the same results, in which CDLQI score was used to quantify the quality of life among the patients with ichthyosis.

Regarding various domains of Childrens' Dermatology Life Quality Index (CDLQI), "symptoms and feelings" was the most affected domain with a proportion of 78%. Other domains (daily activities, treatments, work, leisure, and personal relationships) were affected in 67%, 76%, 58%, 54% and 48% of the patients respectively. These results are similar to observations

reported by Isabelle et. al. who used DLQI⁸ as the tool for assessment of quality of life.

Female gender and VAS scale score for ichthyosis severity viz. erythema, scales, cutaneous pain, pruritus and ocular symptoms were significantly associated with CDLQI score more than 10 (CDLQI>10). On binary logistic regression, Itching (>5 on VAS score) had significantly higher odds (1.9 times) of CDLQI score more than 10. Female gender and presence of scales (>5 on VAS score) were also other significant predictors of CDLQI>10. Our observations are in accordance with Isabelle et. al.⁹ as it was the only study which studied the risk factors predicting the poor quality of life score on DLQI. Females are more prone to have poor quality of life score as females with skin lesions are looked down upon by the society and thereby are more prone to suffer from depression and anxiety. Itching and presence of scales also deteriorates the quality of life by disabling the patient resulting in loss of school days.

The strength of the study was that a validated tool was used to assess the quality of life among ichthyosis patients and factors associated with severity of ichthyosis were determined. Limitations include a small sample size.

Conclusion

The present study shows the epidemiological correlation of patients with congenital ichthyosis and the predictors of poor quality of life score. This study emphasizes on the need of incorporation of regular subjective assessment of quality of life as well as counselling of not only patients but also their caregivers, during the follow-up visits at the physician's clinic/ hospital.

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