Quality of Life and Economic Burden in Recessive Dystrophic Epidermolysis Bullosa

In Kyung Jeon

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The Graduate School, Yonsei University

Quality of Life and Economic Burden in Recessive Dystrophic Epidermolysis Bullosa

Directed by Professor Soo-Chan Kim

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This certifies that the Master's Thesis of
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ABSTRACT

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Background: Recessive dystrophic epidermolysis bullosa (RDEB) patients present blisters and erosions since birth, which cause pain, pruritus and various complications. RDEB affects quality of life (QoL) of patients in physical, emotional and social aspects. Besides, interminable dressings and supportive therapies imposes a significant economic burden on the EB family.

Objective: To assess the quality of life and economic burden in RDEB patients.

Methods: Sixteen RDEB patients were invited asking for their QoL and economic burden. Patients answered questionnaire consisting of visual analogue scale (VAS) on pain and pruritus, Sindex-29, Quality of Life in EB questionnaire (QOLEB) and their economic burden due to EB.

Results: Thirteen RDEB patients completed the questionnaire. Female patients presented higher VAS, QOLEB and total Skindex-29 scores than male patients. RDEB patients showed severe level of pruritus, which was more intolerable than pain. Mean VAS score on pain in RDEB was higher than oral lichen planus and postherpetic neuralgia, VAS score on pruritus was similar to chronic urticaria, atopic dermatitis and prurigo nodularis. Compared with other dermatologic conditions, RDEB patient were profoundly affected in all three scales of skindex-29. Mean “medical costs” in a month was 257.54 United States Dollar (USD) (±169.39) and mean “dressing costs” was 358.41 USD (±312.55), which was negatively related to patient age. QoL and economic burden were not significantly different among RDEB subtypes, BSA involved, hospitalization days and number of accompanying complications.

Conclusion: RDEB had profound impact on QoL and economic burden. Female patients and patient with more severe perceived disease severity presented worse QoL. Compared with other dermatologic diseases, RDEB showed severe symptoms and their QoL was seriously impaired. Most patients had economic burden, especially on preparing dressing materials, younger patients had more economic burden.

Key words: dressing, economic burden, quality of life, recessive dystrophic epidermolysis bullosa

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I. INTRODUCTION

Epidermolysis bullosa (EB) is a heterogeneous group of inherited skin disease characterized by blistering and erosion of the skin after minor mechanical trauma or friction. There are four major types of EB based on the different level of separation within the dermal-epidermal junction, detected by transmission electron microscopy and immunofluorescence mapping; epidermis in EB simplex (EBS), lamina lucida in junctional EB (JEB), dermis in dystrophic EB (DEB) and mixed in Kindler syndrome[1].

Recessive dystrophic epidermolysis bullosa (RDEB) is caused by mutations in the COL7A1 gene, and inherited in an autosomal recessive manner. RDEB severe generalized subtype (RDEB-sev gen), previously called a Hallopeau-Siemens type, and RDEB generalized other (RDEB-O) subtypes are two major subtypes of RDEB, presenting generalized blistering, nail dystrophy, ocular and oral involvement, contractures and severe deformities of hands and feet, and multiple extracutaneous impairments[1].

The physical impairments of RDEB patients affect quality of life (QoL) in physical, emotional and social aspects. Several studies on quality of life in EB have been reported in USA, Australia and Italy[2-5], but QoL study of EB patients in Asia has not been reported yet. Although promising cell based treatments are under investigation[6, 7], main treatment of EB is still rely on the supportive therapies; avoiding trauma, management of extracutaneous complications and wound dressing. Besides, interminable dressings and supportive therapies imposes a significant economic burden on the EB family. However, studies for measurement of economic burden in EB have not been reported yet[8]. In this study, we assessed the quality of life (QoL) and economic burden in RDEB patients for better understanding of disease burden.

II. MATERIALS AND METHODS

1. Study design

This cross-sectional, observational study was conducted with RDEB patients who were diagnosed at the Department of Dermatology of Gangnam Severance Hospital, Seoul, Korea. All diagnosis was confirmed by transmission electron microscopy, immunofluorescence mapping and mutational analysis. The study was approved by the institutional review board in Gangnam Severance Hospital.

Sixteen RDEB patients were invited to our investigation asking for their QoL and economic burden. Patients were explained about the questionnaires and written consent forms were taken before the survey. The documents were mailed to the patients who were not able to visit, and the completed questionnaires and the signed consent forms were returned by mail.

Patients answered questionnaires consisting of visual analogue scale (VAS) on pain and pruritus, Skindex-29, Quality of Life in EB questionnaire (QOLEB) and questions asking their economic burden for treatment. Information about the complications accompanied, involved body surface area (BSA), perceived disease severity of patient by patient global assessment (PGA), days of hospitalization due to EB in the last year was also obtained. For children under 7-year old, main caregivers were asked to take part in the survey to fulfill the questionnaires.

2. Measurement tools

A. Visual analogue scale

VAS has been demonstrated to be a reliable and valid method of pain and pruritus[9, 10]. Average degree of daily disease related pain and pruritus was assessed by a linear 10-score visual analogue scale, ranging from 0 (no symptom) to 10 (intolerable symptom). And we compared VAS scores of RDEB to other dermatologic diseases reported in the literature[11-15]. VAS differences of 6-10mm were considered as clinically meaningful[16].

B. Skindex-29

Skindex-29 is a reliable dermatology-specific instrument to measure QoL[17]. The Korean version of Sindex-29 has been translated and proved its value in evaluating QoL on various dermatologic diseases[18-21]. The questionnaire is made up for 29 questions consisting of three scales (symptom, functioning, emotional burden). Patients are requested to answer these questions concerning preceding 4-week period using 5 point scale. (from never =0, to all the time = 4). In each scale, the score is represented as a percentage of the highest score, from 0 to 100, and higher values indicate a poor QoL. And we also compared Skindex-29 scores of RDEB with other dermatologic diseases reported in the literature[22].

C. Quality of Life in EB questionnaire

It is the first disease specific QoL tool in EB, and is a valid and reliable measurement tool reflecting inability to perform certain tasks in EB patients[4]. Patients are asked to answer 17 questions using 4 point scale (from least = 0, to most impact = 3).

D. Patient Global Assessment

Subjective disease severity perceived by patients was assessed using PGA, which consists of 5 point scale (from very mild =0, to very severe = 4).

E. Economic burden

Average expenses for “dressing costs” and “medical costs” in a month were investigated. The “dressing costs” was defined as total expenses for preparing dressings, fixing materials, topical agents and medicines used during dressing change. The “medical costs” was defined as other direct and indirect medical expenses due to EB, excluding “dressing costs”.

F. Dressing burden

To clarify the burden of dressings, additional questions were presented; ‘Do you have economic burden to prepare dressings/fixing materials?’, ‘How often do you change your dressings?’, ‘How long does it take to change your dressings?’, ‘Are specialized dressings/fixing materials better in function than general dressings/fixing materials? ', 'What is the most important factor when you purchase dressings/fixing materials?'

3. Statistical analysis

Results are expressed as mean ± standard deviations. Statistical analysis was performed using SAS software (Version 9.1.3, SAS Institute Inc., Cary, NC, U.S.A.) using the Student's samples t-test, ANOVA and Spearman's rank correlation coefficient. P value < 0.05 was considered implying statistical significance.

III. RESULTS

Among 16 patients invited, 13 patients (81.3%) completed the questionnaires. Uncompleted questionnaires were not included in the data analysis.

1. Patient demographics and subjective disease severity

Basic patient characteristics are summarized in Table 1. Three patients were under 7-year old, and their main caregivers participated in the survey together. Nine patients (69.2%) were male and 4 patients (30.8%) were female. The mean accompanying complication numbers were 7. 77 ± 2.92, with a range of 3 to 12. According to the EB classification[1], seven patients (53.8%) were RDEB-sev gen, and 6 patients (46.2%) were RDEB-O. All patients had skin lesions involving more than 30% of BSA. Five patients (38.5%) were hospitalized more than 7 days due to EB in the last year, 1 patient (7.7%) less than 7 days, and 7 patients (53.8%) were not hospitalized at all. Eleven patients (84.6%) perceived their disease ‘very severe’ and 2 patients ‘severe’ (15.4%).

Table 1. Basic patient characteristics

Clinical variables	**Results**
Total cases	13
Age (years)	
Mean ± SD	21.57 ± 17.61
Gender, n(%)	
Male	9 (69.2)
Female	4 (30.8)
Complications	
Mean ± SD	7.77 ± 2.92
Range	3 - 12
EB subtype, n(%)	
RDEB, severe generalized	7 (53.8)
RDEB, generalized other	6 (46.2)
Area of body surface involved, n(%)	
>30%	13(100)
10-30%	0 (0)
<10%	0 (0)
Days of hospitalization due to EB in the last year, n(%)	
> 7	5 (38.5)
≤ 7	1 (7.7)
0	7 (53.8)
Perceived disease severity (PGA) , n(%)	
Very severe	11 (84.6)
Severe	2 (15.4)
Moderate	0 (0)
Mild	0 (0)
Very mild	0 (0)

2. QoL results

A. Visual analogue scale

Table 2 shows the VAS, QOLEB and Skindex-29 scores for different clinical variables. Female patients had higher VAS scores in pain and pruritus than male patients. Compared with RDEB-O, RDEB-sev gen had higher VAS score in pruritus, but had similar VAS score in pain. VAS scores in pain and pruritus were not significantly different among hospitalization days or between the perceived disease severities. Patient age, accompanying complication numbers had no correlation with VAS scores in pain or pruritus.

Compared with other dermatologic diseases, the mean pain scale was higher than oral lichen planus and postherpetic neuralgia (Fig. 1A). The mean pruritus scale was similar to chronic urticaria, atopic dermatitis and prurigo nodularis (Fig.1 B).

Table 2. Quality of life (QoL) results

Clinical variables		**VAS**		**QOLEB**	**Skindex-29**		
	n	**pain**	**pruritus**		**Symptoms**	**Emotions**	**Function**
Total cases	13	6.54±1.56	7.54±2.07	26.62±7.61	86.31±10.38	75.23±15.76	76.69±12.09
Gender							
Male	4	5.75±0.95	6.75±2.63	25.75±8.22	90.00±14.21	65.00±20.94	75.00±17.68
Female	9	6.88±1.69	7.88±1.83	27.44±7.78	84.67±8.72	79.78±11.51	77.44±10.00
Days of hospitalization due to EB in the last year							
0	7	6.57±1.39	7.14±2.27	25.14±7.35	80.71±10.67	79.14±10.38	77.71±12.22
≤ 7	1	6	10	25	91	94	88
> 7	5	6.60±2.07	7.60±1.82	29.80±8.70	93.20±5.72	66.00±19.02	73.00±12.81
EB subtype							
RDEB-sev gen	7	6.57±1.72	8.00±1.29	30.14±8.82	90.57±8.96	70.29±17.41	79.00±14.19
RDEB-O	6	6.50±1.52	7.00±2.76	23.17±3.76	81.33±10.35	81.00±12.57	74.00±9.65
Perceived disease severity							
Very severe	11	6.54±1.69	7.54±2.25	28±7.82*	86.72±10.39	78.54±13.97	79.54±10.82*
Severe	2	6.50±0.71	7.50±0.71	21*	84.00±14.14	57.00±15.55	61.00±1.41*

EB, epidermolysis bullosa, RDEB, recessive dystrophic EB, RDEB-sev, RDEB severe generalized, RDEB-O, RDEB generalized other. * $P < 0.05$

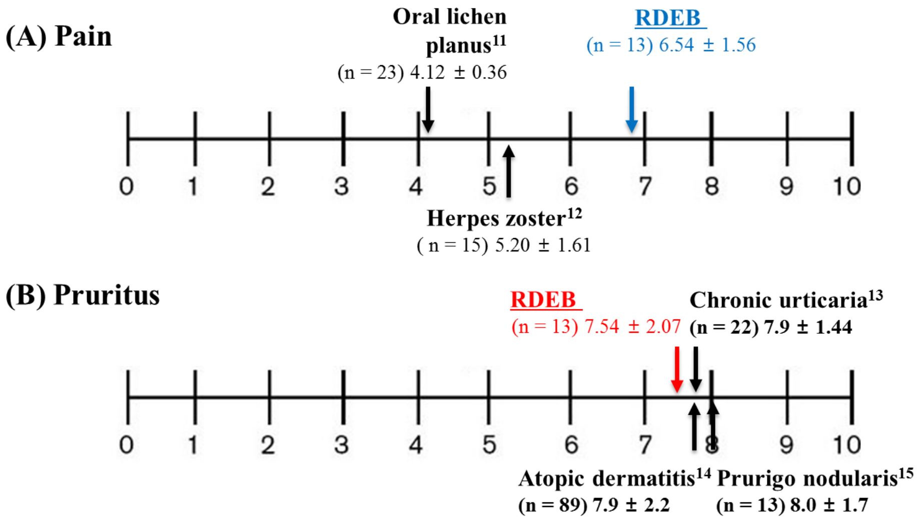


Figure 1. Visual analogue scale(VAS) in pain and pruritus. VAS differences of 6-10mm were considered as clinically meaningful. (A) The mean pain scale in RDEB was higher than oral lichen planus and hepres zoster (postherpetic neuralgia). (B) The mean pruritus scale was similar to chronic urticaria, atopic dermatitis and prurigo nodularis.

B. Skindex-29

The mean score was 86.31±10.38 in symptoms scale, 75.23±15.76 in emotions and 76.69±12.09 in functioning (Table 2). Male patients had higher scores in symptoms and female patients represented higher scores in emotions and functioning, but showed no statistical significances. Skindex-29 was not significantly different among the hospitalization days or between EB subtypes. In function scale, patients with perceived disease severity of ‘very severe’ reported significantly higher score than of ‘severe’ ($p < 0.05$). Patient age, accompanying complication numbers had no correlation with Skindex-29. Compared with other dermatologic conditions, RDEB patient were profoundly affected in all three scales of skindex-29 (Table 3).

Table 3. Skindex-29. RDEB versus other skin conditions

Clinical variables	**Sample size**	**Symptoms mean (SD)**	**Emotions mean (SD)**	**Function mean (SD)**
RDEB	13	86 (10)	75(16)	77(12)
Other skin conditions[22]				
Dermatomyositis	22	42 (25)	45 (27)	28 (29)
Vulvodynia	280	50 (17)	50 (20)	44 (22)
Psoriasis	44	42 (21)	39 (27)	23 (27)
Eczema	102	48 (23)	41 (27)	26 (26)
Acne vulgaris	63	30 (19)	41 (25)	16 (16)
Alopecia	7	31 (24)	27 (33)	14 (23)
Rosacea	29	33 (20)	33 (20)	16 (18)
Without skin disease	107	14 (2)	9 (13)	4 (8)

RDEB, recessive dystrophic EB.

C. QOLEB

Mean QOLEB score was 26.62±7.61 (Table 2), higher score was observed in female patients, patients with hospitalization days more than 7 days and RDEB-sev gen subtype, but showed no statistical significances. Patients with perceived disease severity of ‘very severe’ reported significantly higher score than of ‘severe’ ($p < 0.05$). Patient age, accompanying complication numbers had no correlation with QOLEB.

D. Economic burden

Table 4 shows the “medical costs” and “dressing costs” for different clinical variables. “Dressing costs” was greater than “medical costs”, the mean “medical costs” in a month was 257.53±169.39 United States Dollar (USD) with a range of 93 to 465 USD, and the mean “dressing costs” was 358.41±312.55 USD with a range of 93 to 930 USD. Patients with hospitalization days more than 7 days and RDEB-sev gen subtype spent more “medical costs” and “dressing costs” than other patients. “Medical costs” and the “dressing costs” were negatively related to patient age (Spearman rho = -0.68, p = 0.01 and Spearman rho = -0.56, p = 0.049), representing statistically meaningful correlation. Perceived disease severities and accompanying complication numbers had no correlation with economic burden. “Medical costs” and “dressing costs” had no correlation with VAS, QOLEB and Skindex-29.

Table 4. Economic burden of RDEB patients in a month

Clinical variables	**n**	**Medical costs (USD)**	**Dressing costs (USD)**
Total cases	13		
Mean ± SD		257.54 ± 169.39	358.41 ± 312.55
Range		93 - 465	93 - 930
Gender			
Male	4	267.40 ± 229.00	488.30 ± 411.60
Female	9	253.20 ± 152.70	300.70 ± 266.10
Days of hospitalization due to EB in the last year			
0	7	219.21 ± 148.79	273.69 ±298.32
≤ 7	1	46.50	93
> 7	5	353.40 ± 166.36	503.10 ± 304.21
EB subtype			
RDEB, severe generalized	7	298.90 ± 177.80	431.80 ± 299.80
RDEB, generalized other	6	209.30 ± 160.40	272.80 ± 332.00

EB, epidermolysis bullosa, RDEB, recessive dystrophic EB, USD, United States Dollar

E. Dressing burden

As regards the additional questions asking burden of dressings, 7 patients (53.85%) answered that they have economic burden on dressing materials “always”, and 3 patients (23.07%) answered “often”(Fig. 2A). Regarding dressing change frequency, 7 patients (53.85%) changed their dressing everyday, and 4 patients (30.77%) changed three times in a week (Fig. 2B). Seven patients (53.85%) answered that they require 1~2 hours to change their dressings, and 4 patients (30.77%) answered less than 1 hour (Fig. 2C).

Eleven patients (84.61%) answered that the specialized dressings are better or much better in function than general dressings (Fig. 2D). But price was the most important factor (38.46%) when they purchase dressings, followed by pain during dressing change (30.76%) (Fig. 2E). In fixing materials, 12 patients (92.30%) answered that the specialized fixing materials are better than general fixing materials (Fig. 2F). Price (38.46%) and fixation ability (38.46%) were two important factors when they select fixing materials (Fig. 2G).

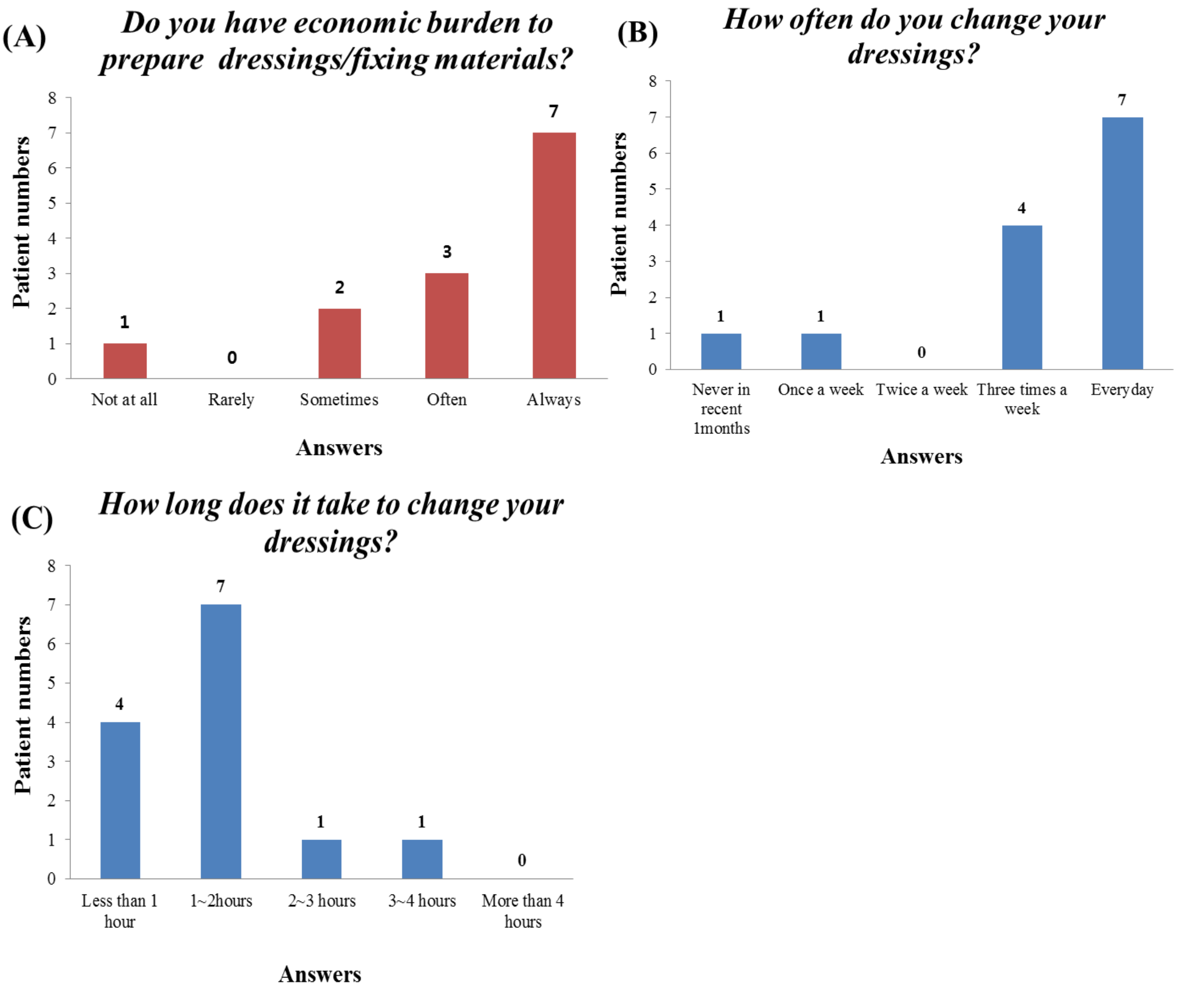
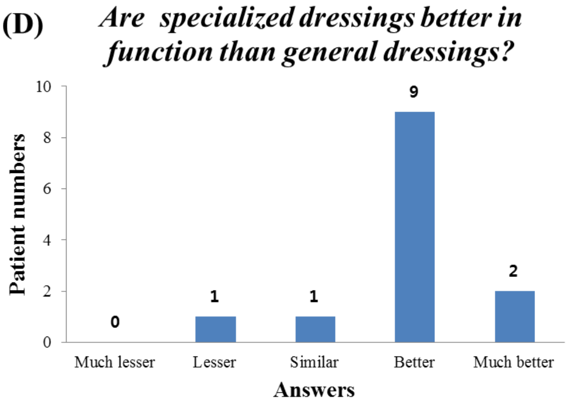
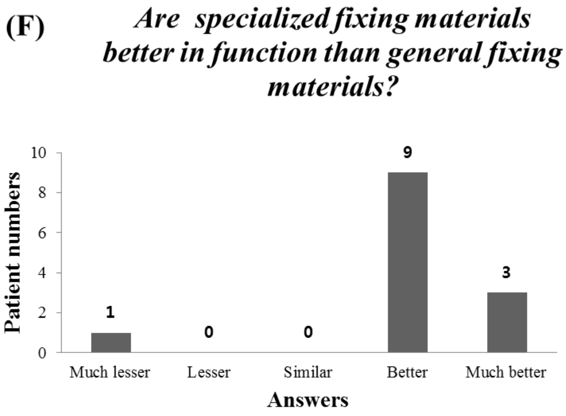
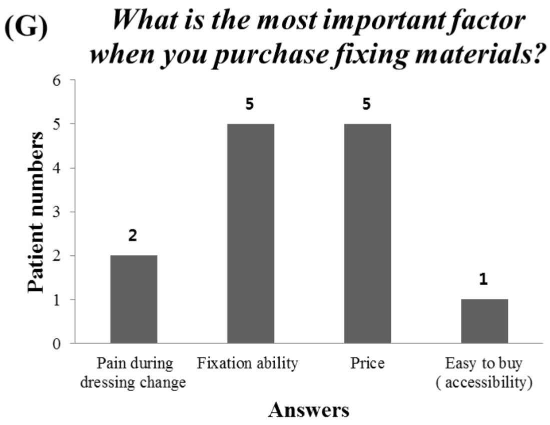


Figure 2. Questions and answers about economic burden and dressing materials. (A) Seven patients answered that they have economic burden on dressing materials “always”, and 3 patients answered “often”. (B) Seven patients changed their dressing everyday, and 4 patients changed three times in a week. (C) Seven patients answered that they require 1~2 hours to change their dressings, and 4 patients answered less than 1 hour.


(D) *Are specialized dressings better in function than general dressings?*


(E) *What is the most important factor when you purchase dressings?*


(F) *Are specialized fixing materials better in function than general fixing materials?*


(G) *What is the most important factor when you purchase fixing materials?*

(D) Eleven patients (84.61%) answered that the specialized dressings are better or much better in function than general dressings. (E) But price was the mot important factor when they purchase dressings, followed by pain during dressing change. (F) In fixing materials, 12 patients answered that the specialized fixing materials are better than general fixing materials. (G) Price and fixation ability were two important factors when they select fixing materials.

IV. DISCUSSION

Our study evaluated the QoL and economic burden of patients with RDEB. To our knowledge, this study was the first study assessing economic burden of EB and the first study evaluating QoL of RDEB patients in Asia.

QoL comprises of not only individual’s physical aspect, but also various aspects of emotional health, psychological and social well-being. While VAS can evaluate only symptom of the patients, Skindex-29 and QOLEB, one skin specific and the other EB specific tools can assess the various aspects of the patients. Skindex-29 can measure the psychological effects of the disease, changes to daily life and relationships with others. QOLEB can assess disease burden in daily life properly, because it takes into account specific problems experienced by individual patients[3].

Female gender is considered to be a correlated factor with QoL and psychological morbidity[8], which tendency has also been reported in EB and other dermatological diseases[5, 23]. Likewise, in our study female patients presented higher VAS, QOLEB and total Skindex-29 scores than male patients.

VAS score was more severe in females, but symptom scale in Skindex-29 was reported higher in males. The discordance between VAS and skindex-29 is no wonder, because VAS assesses the symptom based on the severity, whereas skindex-29 assesses the symptom based on the frequency. QOLEB also evaluates degree of pain based on frequency, Frew et al[4] mentioned that VAS would be more accurate tool for actual assessment of pain and pruritus. Besides, VAS is easy and quick to perform, and it could be useful during the consultation hours for accurate symptom management of EB patients.

The RDEB patients were suffering from severe pain compared to other dermatologic diseases reported in the literature, VAS score on pain was higher in RDEB than oral lichen planus and postherpetic neuralgia. Fine et al[24] reported that the severe level of pain was most often seen in JEB-Herlitz type and RDEB Hallopeau–Siemens type. But in our study, VAS score on pain was similar between RDEB-O and RDEB-sev gen.

Pruritus is a common symptom in the EB population, but accurate etiology of pruritus in EB patients is unknown; abnormal chronic skin inflammation, overheating caused by dressings, dry skin, healing wounds, weather are potential contributing factors[2, 25]. Pruritus has been reported to be the biggest concern of EB patients, followed by pain[2]. Likewise, in our study, RDEB patients showed severe level of pruritus, which was more intolerable than pain. VAS score on pruritus in RDEB was similar to chronic urticaria, atopic dermatitis and prurigo nodularis, however, pruritus in EB would be more influential because it usually starts since birth or early childhood and lasts lifelong, scarcely showing satisfactory response to the medications.

Compared with other dermatologic diseases, RDEB patients showed higher skindex-29 in all three scales, certainly revealing the disease burden of RDEB.

Compared with previous survey of EB patients in Italy[5], skindex-29 scale of RDEB was also higher in our study, because the previous survey seems to have enrolled more participants with milder phenotype than in ours; all patients in our study had skin involvement beyond 30%. Seven patients who had more than 30% skin involvement in the Italian study showed higher symptom scale than other patients, but our study still showed higher scores of skindex-29. One possible reason could be a different cultural background about symptoms, emotions and social relationships in Asia.

We hypothesized that patients with severe disease subtype, RDEB-sev gen, would show severely impaired QoL than other groups. However, RDEB subtypes represented no meaningful differences in QoL. In previous report[5], sindex-29 did not show significant differences between the types or subtypes of EB, which could be explained by heterogeneity within the disease[8]. However, in our study, other objective indexes (hospitalization days, number of accompanying complications) also showed no significant differences in QoL.

On the other hand, perceived disease severity of patient by patient global assessment was significantly associated with QOLEB scores and function scale in Skindex-29. Although establishing causal relationship was not possible in this observational, cross-sectional study, impairments in daily activity and social relationship might have influenced considerably when EB patient recognize their disease.

Assessment of the economic burden is an important aspect of this study. Most patients had economic burden on preparing dressing materials. “Dressing costs” was higher than “medical costs”, representing that the dressing was the main cause of economic burden in RDEB patients. In Korea, specialized dressing materials are not covered by insurance system. But exceptionally, for patients with JEB or DEB, 3 sheets of specialized dressings can be covered by insurance in a week. And patients with ‘rare and Incurable Disease’, including RDEB patients, patient’s responsibility consists 10% of total expenses. For instance, RDEB patient can buy 3 sheets of Mepilex lite® 20cm x 50cm, approximately for 14 USD in a week. But total “dressing costs” in a month was much more expensive, because RDEB patients usually need numerous sheets of dressing materials on the whole body, and most of them changed their dressings everyday or every other day, imposing ponderous economic burden.

The more skin lesion enlarges, the more economic burden on dressing occurs. Because all participants had skin lesions more than 30%, comparing “dressing costs” according to the involved BSA was not possible. Moreover, blisters and erosions can occur at any area of the skin of EB patients, investigating accurate involvement area was not feasible. BSA of infants or children is smaller than adults, thus they are expected to expense lesser than adults. But interestingly, younger patient expensed more “dressing costs” and “medical costs”. In infancy and early childhood, physical adjustment ability doesn’t develop fully, and self-induced mechanical trauma occurs more frequently, and dressings are hard to be maintained. And secondary complications due to skin barrier disruptions and immunologic immaturity might have increased the “medical costs” in younger RDEB patients. And older patients, in other words, who survived for a long time, are expected to have milder phenotype than others.

Patients used various kinds of general and special dressings/fixing materials, many of them answered that specialized dressings/fixing materials are better than general dressings/fixing materials in a functional aspect. But when they purchase materials for dressing change, not only functional aspect of the materials but also price of the materials influenced considerably on their choice.

There are several limitations in our study. First, there is a potential selection bias because our study was conducted in a single institute and our sample may not represent general EB population. Though, our institute is the only one institute that can perform laboratory diagnosis of EB, and almost all EB patients in Korea are referred to our institute. Secondly, a small sample size of this study also works as a limitation, though incidence of RDEB is very low. Thirdly, severe RDEB patients were over-represented, because serious patients were usually referred to our institute, actually, all patients participated in the survey had skin lesions involving more than 30% of body surface area and average 7 complications. Fourthly, main caregivers of children under 7-year old fulfilled the questionnaire. Their answers, which are not able to represent actual QoL of their children, were also included for the analysis. Finally, potential for recall bias exist in this questionnaire-based study.

V. CONCLUSION

In conclusion, RDEB had profound impact on QoL and economic burden. Female patients and patient with more severe perceived disease severity presented worse QoL. Compared with other dermatologic diseases, RDEB showed severe symptoms and their QoL was seriously impaired. Most patients had economic burden, especially on preparing dressing materials, and younger patients had more economic burden. Our study could help expanding understanding of QoL and economic burden in RDEB patient, and fulfillment of the patient’s need.

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ABSTRACT(IN KOREAN)

열성 이영양형 수포성 표피박리증 환자의 삶의 질과 경제적 부담

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전인경

열성 이영양형 수포성 표피박리증은 제7형 콜라겐의 유전적 결함에 의해 발생하는 질환으로, 환자들은 출생시부터 전신에 수포와 미란이 나타나며, 심한 통증과 소양감, 다양한 합병증이 동반된다. 이러한 신체적 고통은 환자들의 정신적, 사회적 측면과 삶의 질에도 큰 영향을 미친다. 삶의 질을 측정하는 것은 환자 입장에서 질병을 바라보고, 일상 생활에서의 기능적, 정신적 어려움을 인지하여 적절한 도움을 제공할 수 있다는 점에서 중요하다. 그 동안 미국, 호주, 유럽에서 환자들의 삶의 질에 관한 연구가 진행된 바 있으나 아시아에서는 보고된 바 없었다. 현재로서는 수포성 표피박리증의 근본적인 치료 방법이 없어 수포 예방이나 드레싱과 같은 보존적 치료를 평생 지속해야 하며, 이는 환자에게 커다란 경제적 부담을 가중시킨다. 하지만 아직까지 수포성 표피박리증 환자들의 경제적 부담에 대한 연구도 보고된 바 없었다.

본 연구는 한국인 열성 이영양형 수포성 표피박리증 환자들에게서 삶의 질과 경제적 부담에 대해서 연구해 보고자 하였다.

총 16명의 열성 이영양형 수포성 표피박리증 환자를 대상으로 설문 조사가 진행되었으며, 그 중 13명의 환자들이 설문 조사를 완료하였다. 피부 증상의 강도를 측정하기 위해 visual analogue scale을 사용하였고, 삶의 질을 측정하기 위하여 Skindex-29, Quality of Life in EB(QOLEB)를 사용하였으며, 질병으로 인한 한달 동안의 경제적 부담도 조사하였다.

본 연구를 통해 한국의 열성 이영양형 수포성 표피박리증 환자들의 삶의 질이 매우 저하되어 있다는 것과 치료로 인한 경제적 부담이 크다는 사실을 확인할 수 있었다. 여성 환자들은 남성 환자들보다 더 높은 VAS, Skindex-29, QOLEB 점수를 나타냈다. 환자들은 심한 소양감과 통증을 호소하였는데, 소양감을 통증보다 더 심하게 호소했다. 통증에 대한 평균 VAS 점수는 구강 편평 태선이나 대상포진 후 신경통에서 보고된 VAS 점수보다 높았고, 소양감에 대한 VAS점수는 만성 두드러기, 아토피 피부염, 결절성 양진에서 보고된 VAS 점수와 비슷하였다. 다른 피부과적 질환과 비교하였을 때, 열성 이영양형 수포성 표피박리증 환자들은 skindex-29의 증상, 기능, 감정 항목에서 모두 심각한 영향을 받고 있었다. 질병으로 인한 한달 평균 의료 비용은 257.54달러(±169.39)였고, 한달 평균 드레싱 비용은 358.41 달러(±312.55)였다. 경제적 부담은 환자 나이와 통계학적으로 의미 있는 연관성을 보여 환자 나이가 어릴수록 높게 나타났다. 환자들의 삶의 질과 경제적 부담은 질병의 아형이나 피부 침범 정도, 입원 일수, 동반된 합병증 개수와는 관련성이 없었다.

본 연구는 열성 이영양형 수포성 표피박리증에서 경제적 부담을 조사한 최초의 연구이며, 아시아에서는 처음으로 진행된 삶의 질 연구이다. 본 연구 결과는 열성 이영양형 수포성 표피박리증 환자를 진료하는 데 있어서 환자의 삶의 질과 경제적 부담에 대한 인식과 이해도를 넓히는 데 의의를 지닐 것으로 생각된다.

핵심되는 말 : 경제적 부담, 드레싱, 삶의 질, 열성 이영양형 수포성 표피박리증