

Impact of a Psychodermatological Education Package on the Subjective Distress, Family Burden, and Quality of Life among the Primary Caregivers of Children Affected with Epidermolysis Bullosa

Abstract

Background: Epidermolysis bullosa (EB) has profound effect on the subjective distress, family burden, and quality of life (QOL) of the primary caregivers (PCG). Knowledgeable PCG can efficiently manage children with these skin diseases and also improve their QOL. **Objectives:** To assess the subjective distress, family burden, and QOL, to develop and assess the short-term effectiveness of a psycho-dermatological education package (PDEP) for the PCG of children with EB. **Methods:** In this interventional study, 30 PCG of EB were assessed for subjective distress, family burden, and QOL. PDEP, a structured educational tool explaining the disease and its care and stress management, was developed by the authors for the PCG and administered to them after one month of enrolment. They were reassessed after three months and compared with the baseline assessment scores. For comparison, 37 PCG of CI were also studied. **Results:** The mean age (years) of the subjects was 28.7 ± 6.7 for EB and 30.5 ± 4.6 for CI. The mean or median (range) baseline scores for subjective distress, family burden and QOL of PCG ($n = 20$) of EB were 8.4 ± 7.9 , 6.5 (0-30); 28.5 ± 17.5 , 24 (7-77) and 12.6 ± 6.7 , 11.5 (4-28) and for PCG ($n = 14$) of CI were 12 ± 4.3 , 38.9 ± 16.2 and 17.7 ± 3.6 respectively. The PDEP improved the QOL ($p = 0.01$), knowledge ($p < 0.01$) and practices ($p < 0.001$) for PCG of EB and it improved subjective distress ($p < 0.001$), QOL ($p < 0.01$) and knowledge ($p < 0.01$) for PCG of CI. **Conclusions:** PDEP is an effective educational tool in improving the QOL and knowledge of PCG, which in turn provides efficient management and psychological support to children affected with EB and CI. It should, therefore, be routinely used for educating the PCG of children with EB and CI.

Keywords: Congenital ichthyosis, epidermolysis bullosa, psychodermatology

Introduction

Epidermolysis bullosa (EB) is a group of inherited disorders, characterized by extreme skin fragility manifesting at or soon after birth. Minimal trauma and friction can cause extensive blistering and/or erosions in children with EB, resulting in a number of complications. In spite of extensive research on the molecular genetics of EB and clinical manifestations, a definitive treatment is still not available. Currently, the only treatment consists of supportive care, blister and wound management, and psychological support to the child and family.^[1,2]

The natural course of EB and its management have profound implications not only on children but also on the primary caregivers (PCG), with the greatest

impact seen in children with severe disease, irrespective of the clinical type.^[3] The subjective distress and family burden increase with the caregiver's perception of the disease severity and uncertainty about the cure of disease, which in turn impairs the quality of life (QOL) of PCG.^[3,4]

It is of great importance to address the psychosocial issues of PCG of children with EB in addition to providing good meticulous nursing care so as to allow healthcare professionals to develop appropriate care strategies not only for the children with EB but also for their PCG and family. An education program that provides accurate knowledge about treatment and management of EB to the PCG may improve the child's prognosis and treatment compliance as well as their

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QOL. A knowledgeable primary caregiver can make a significant impact on the QOL of EB children as most of these children are managed at home. Therefore, the present study was conceptualized to develop and assess the short-term effectiveness of a psychodermatological education package (PDEP) on the subjective distress, family burden, and QOL of PCG of children with EB. The present study is one of its kind in India, in which the first nurse led intervention for EB children and for their PCG was planned.

Methods

This interventional study was conducted in the outpatient department of Dermatology, AIIMS, New Delhi between January 2014 and December 2014. A convenient sample of 30 PCG of children (aged 0–5 years) affected with EB and for comparison 37 PCG of children (aged 0–5 years) affected with CI were selected.

Data collection

The Institutional ethics committee (IEC) approved the protocol. PCG were informed about the study and gave their written consent for participation in the study. The study subjects were initially assessed for their sociodemographic data, subjective distress, family burden, and QOL. They were also assessed for knowledge and practices related to the management of EB and CI. The subjective distress and family burden were measured using the standardized Post Graduate Institute Neuroticism-1 General Health Questionnaire (PGI N-1 GHQ, $\alpha = 0.86$)^[5] and Caregiver burden inventory (CBI, $\alpha = 0.83$)^[6] respectively. The QOL was assessed using the Family Dermatology Life Quality Index (FDLQI, $\alpha = 0.91$)^[7]. The authors developed the subject datasheet, knowledge and practice questionnaires, and a feedback form, which were validated by four experts. We translated these questionnaires and forms into Hindi and then back-translated them to English with the necessary corrections. We checked the questionnaires for reliability with retest method in six PCG with a gap of 1 week. Retest reliability quotient was 0.90 for knowledge and 0.80 for practice questionnaire.

For educated PCG, the questionnaires for subjective distress, family burden, and QOL were self-administered, while for illiterate PCG, interview method was used. The overall time taken for the initial baseline assessment for each PCG was approximately 1 h.

Based on the initial pilot study assessment of subjective distress, family burden, QOL, and knowledge in 10 PCG of EB and CI, we developed a PDEP. Information on PDEP was also gathered from an extensive literature review. The content validity of PDEP was checked and verified by informal discussions with five experts (Dermatology, Psychiatry, and Nursing). The feasibility of administering the PDEP was also carried out in the same pilot study and was found to be simple, practical, valid, and reliable.

The PDEP comprised interactive sessions for a duration of 1 h conducted by a nurse (MPA) delivered with the aid of structured material with flashcards, explaining about various aspects of EB and CI (anatomy and physiology of the skin, nature and type of disease, genetic inheritance, clinical presentation, diagnosis, treatment including care of skin, complications, main problems associated with the disease, common myths, and misconceptions) and also management of stress. The session for EB PCG also included a demonstration of handling a child with EB, dressing kit preparation, and care of blisters. PDEP was administered to the PCG of EB and CI children at 1 month after the initial data collection. Families belonging to the same region were called at a time but the group did not exceed two families. Besides PCG, other family members who were willing to be part of the package were invited, but only the PCG was assessed by posttest.

Telephonic booster sessions were given periodically to reinforce the PDEP and the PCG were also allowed to contact the research team telephonically or personally as per their convenience. All the PCG were reviewed at 3 months after the administration of the PDEP and were reassessed for the same variables [Figure 1].

A feedback form about the usefulness of PDEP was also taken from the PCG at the end of the study.

Data analysis

All statistical analysis was implemented on Stata 12.1 (Stata Corp, College Station, TX, USA). Both descriptive and inferential statistics were used for data analysis. Frequency (%), Median (Range) or Mean \pm SD were used as appropriate. The subjective distress, family burden, QOL, and knowledge scores of PCG were compared at baseline and follow-up assessment. Continuous variables were compared before and after administration of PDEP using Wilcoxon signed-rank test/ Paired “t”-test. P value <0.05 was considered significant.

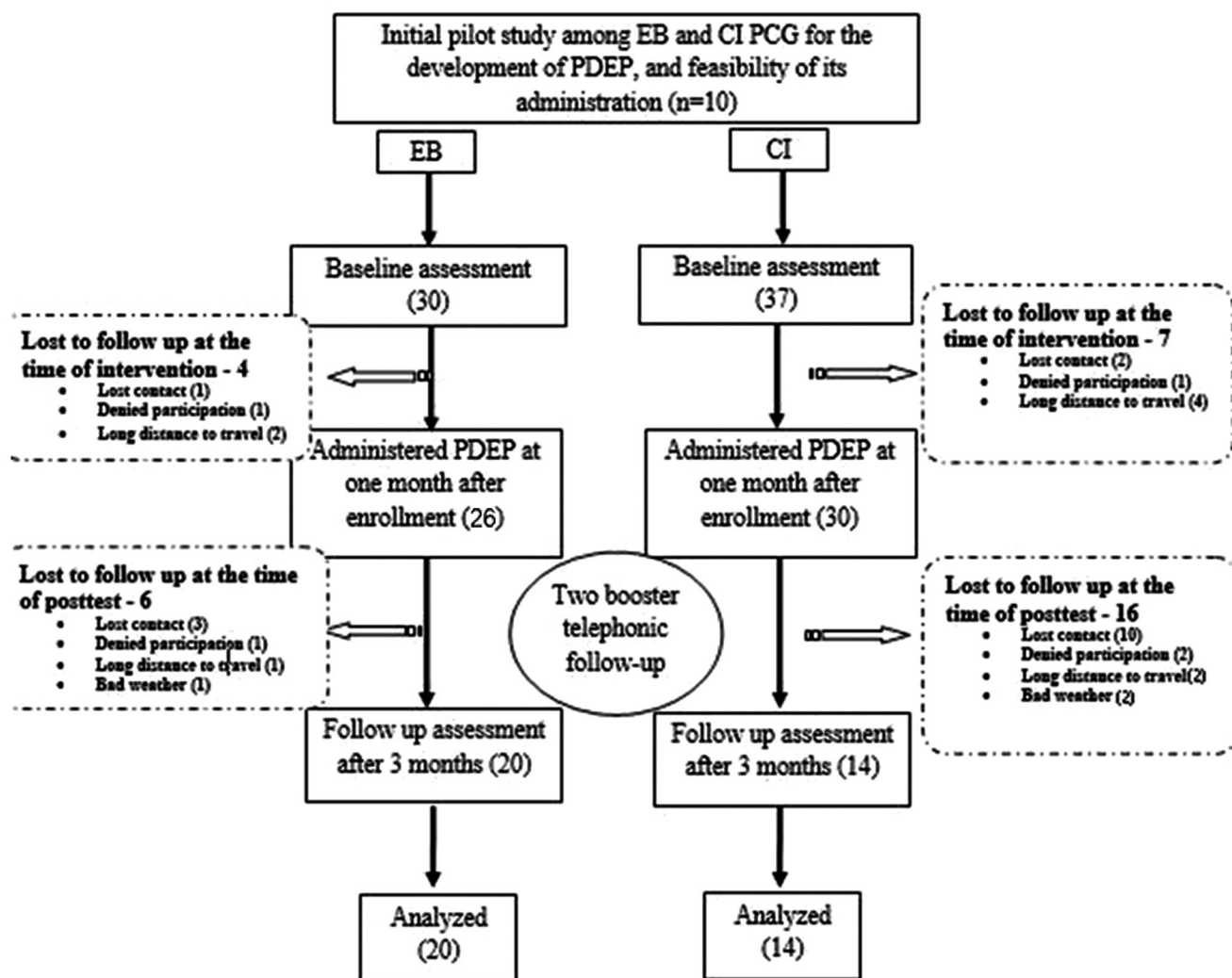
Results

Demographics

The mean age of the PCG (years) of EB was 28.7 ± 6.7 and CI was 30.5 ± 4.6 and the mean age (years) of children affected with EB was 2.9 ± 1.6 and 3.5 ± 1.1 for children affected with CI. Majority of the PCG were mothers and were not aware of their children’s medical condition. None of the PCG mentioned the presence of a similar disease in their family though other hereditary diseases were present. There was no medical illness among the PCG.

Subjective distress, family burden, and QOL

Significant improvement was observed in QOL after PDEP (12.6 ± 6.7 vs. 8.2 ± 3.5 , $P = 0.01$). There was also a significant increase in the mean scores for the knowledge (15.5 ± 4.0 vs. 17.8 ± 2.3 , $P < 0.01$) and



PDEP – Psycho-dermatological education package

EB – Epidermolysis bullosa

CI – Congenital Ichthyosis

Figure1: Flow chart depicting the study protocol

practices (9.1 ± 4.5 vs. 13.4 ± 2.4 , $P < 0.001$) of the PCG. Though the subjective distress and family burden score also decreased, they did not reach statistical significance [Table 1]. The results of the comparison group are given in Table 2.

Qualitative analysis of the feedback forms revealed that the PDEP was very useful and the PCG wanted to have the educational material with them for future reference, gained confidence in handling the child, and were hopeful about a possible definitive treatment for the disease in future.

Discussion

EB has a significant clinical and socioeconomic impact not only on the affected children but also on their families and/

or PCG. The problems associated with EB such as repeated blistering and wound care, disfigurements, deformities, and embarrassment in social gatherings have disastrous effects on the PCG. Several Investigators have evaluated the psychological problems, QOL, and family relationship of PCG with their EB children.^[3,8-12] No studies have evaluated the disease burden of PCG of EB in India. This interventional study was planned to develop a PDEP and to study its impact on the subjective distress, family burden, and QOL among the PCG of children with EB.

In the present study, the various aspects of QOL of PCG adversely affected were time spent on looking after the child (71%), emotional distress (59%), financial aspect (56%), effect on housework (45%), physical

Table 1: Comparison of pre and postintervention scores for subjective distress, family burden, QOL, knowledge, and practices, among PCG of EB

Characteristics	Before PDEP (n=20)	After PDEP (n=20)	P
Subjective distress			
Mean±SD	8.4±7.9	6.2±5.3	0.46
Median (min max)	6.5 (0-30)	3 (0-19)	
Family burden			
Mean±SD	28.5±17.5	26.9±13.3	0.70
Median (min max)	24 (7-77)	27 (9-49)	
QOL			
Mean±SD	12.6±6.7	8.2±3.5	0.01
Median (min max)	11.5 (4-28)	9 (0-14)	
Knowledge			
Mean±SD	15.5±4.0	17.8±2.3	<0.01
Median (min max)	16 (6-21)	18 (13-21)	
Practices			
Mean±SD	9.1±4.5	13.4±2.4	<0.001
Median (min max)	11 (0-16)	13 (8-16)	

Table 2: Comparison of pre and postintervention scores for subjective distress, family burden, QOL, and knowledge among PCG of CI

Characteristics	Before PDEP (n=14)	After PDEP (n=14)	P
Subjective distress Mean±SD	12±4.3	7.5±3.0	<0.001
Family burden Mean±SD	38.9±16.2	33.4±11.1	0.17
QOL Mean±SD	17.7±3.6	15.4±2.5	<0.01
Knowledge Mean±SD	7.7±2.1	16.5±2.1	<0.01

wellbeing (41%), job/study (41%), leisure activities (39%), social life (30%), people's reaction (30%), and personal relationships (30%). These findings suggest that EB imposed a heavy burden on the PCG. It was found that 18 PCG were requiring some form of respite care and 10 were in complete burnout, who were referred to the experts for further management.

We did not find any correlation between the QOL of PCG and the severity of the disease in our small sample size, which was in concordance with Tabolli *et al.*^[11] None of the PCG reported disturbed marital life as a result of caring for a child suffering from EB contradictory to that reported by Fine *et al.*^[9] who noticed that divorce was common among parents of children with EB. This may be partly explained by the cultural background and family support in India.

In the present study, many PCG were unaware of their children's disease or its natural course and complications. Lack of information about the disease can heighten the stress and anxiety of the PCG, further compromising their QOL. In such situations, an educational intervention like

PDEP can significantly play an important role in reducing the stress and improve their QOL.

In the present study, the PDEP was found to be an effective intervention in improving the knowledge ($p < 0.01$), practices ($p < 0.001$), and QOL ($p = 0.01$) among the PCG. Stevens *et al.* established a home nursing program to provide assistance to families or patients with severe EB.^[12] They perceived improvement in the QOL, a better provision of support and improved family life management after providing nursing care during dressing changes in their homes over a period of 2 years.

Though the subjective distress and family burden score of the PCG in our study also decreased, they did not reach statistical significance. This could be explained by the small sample size and also the fact that some of the patients were already following up with the EB team for some time. They could have been sensitized about the disease burden to some extent that resulted in nonsignificant observations.

In the comparison group, the PDEP improved subjective distress ($p < 0.001$), QOL ($p < 0.01$), and knowledge ($p < 0.01$) for PCG of CI. Ichthyosis also has a profound impact on the psychosocial aspect of the children as they have *fish*-like scaling all over the body and heat intolerance that forces them to avoid social gathering and playing. Many children avoid going to school as well and they tend to remain indoor most of the time. All these factors could seriously affect the QOL of their parents or their caregivers. Though both these genetic disorders have variations in their clinical presentations, complications, and natural course, it is worthwhile to note that developing a program for the PCG of both EB and CI definitely improves their QOL.

In summary, EB has a significant effect on the subjective distress, family burden, and QOL of the PCG. An effective educational tool like PDEP developed by us is crucial for the overall improvement in the QOL of PCG, which in turn will provide an efficient wound care management and other support to children affected with EB. The educational intervention in PDEP was simple, doable, and tailored according to their needs and provided clear unambiguous information about the care of EB. It also included an initial demonstration on nursing care, managing blisters using the available low-cost dressing materials, and discussion on day to day care of the child with the help of flashcards followed by practice sessions in front of the investigators.

Limitations of the Study

- Single setting, sample of convenience, and absence of control group within EB limit the generalizability of the findings.
- Some of the patients were already under regular follow-up and are sensitized about the disease to some

extent that may explain few of the nonsignificant observations.

- The study variables could not be assessed based on the disease types due to small sample size.
- The study could not be blinded as the same person was involved in the delivery of intervention and assessment.

Declaration of patient consent

The authors certify that they have obtained all appropriate participant consent forms. In the form, the participants have given their consent for their images and other clinical information to be reported in the journal. The participants understand that their names and initials will not be published and due efforts will be made to conceal their identity, but anonymity cannot be guaranteed.

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Nil.

Conflicts of interest

There are no conflicts of interest.

Note: Detailed Questionnaires / Screening Chart and such informations can be obtained on request to the corresponding author.

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