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- How epidermolysis bullosa (EB) affects quality of life
- Literature review on the impact of social support on quality of life (QoL) in people with EB
- Qualitative and quantitative associations between QoL and social support in people with EB

Epidermolysis bullosa: how social support affects quality of life

Key points

Epidermolysis bullosa (EB) is a group of rare genetic skin conditions with a far-reaching social impact

Studies into EB focus on finding a cure, resulting in psychosocial aspects being under-researched

Social support is seen as important, but poorly defined and measured

A bio-psychosocial approach to the management of EB should be at the centre of practice

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Abstract Measuring quality of life has become an increasingly important method of evaluating the effect of health and social care interventions. The rare genetic condition epidermolysis bullosa is known to have a deep social impact on people's quality of life, but most research into the condition is focused on its biomedical aspects and attempts to find a cure. A literature review has explored the relationship between social support and quality of life in people with epidermolysis bullosa. It concludes that, while more research is needed, social support does have a positive impact on quality of life and that professionals should adopt a bio-psychosocial approach to management.

Citation Butterworth S et al (2019) Epidermolysis bullosa: how social support affects quality of life. *Nursing Times* [online]; 115: 5, 48-51.

Epidermolysis bullosa (EB) is a group of rare genetic conditions that cause the skin to blister and tear at the slightest touch; indeed the skin is so fragile that it is commonly referred to as 'butterfly skin'. There is currently no cure, and severe forms of this long-term condition can be fatal.

Research into EB focuses on finding a cure, so there is a dearth of studies on its psychosocial impact. A PhD study conducted at the University of Chester and supported by DEBRA UK, the charity that supports individuals and families affected by EB, has explored the relationship between quality of life (QoL) and social support for people with EB. This literature review is the first part of a wider study of adults living with the condition, which is due to be completed in 2020. This article presents the results of the literature review conducted as part of the study.

Epidermolysis bullosa

A rare disease is defined as a condition that affects fewer than one in 2,000 people; DEBRA (www.debra.org.uk) estimates that there are around 5,000 people living with EB in the UK. Four main types of EB have been identified:

- EB simplex (the mildest form);
- Kindler syndrome;
- Dystrophic EB;
- Junctional EB (the most severe form; Fig 1) (Fine et al, 2014).

The more severe of these can be life limiting and cause significant disability (Dures et al, 2011).

At present there is no cure for EB and the need to find one dominates research in this field of practice. This means much of the research on EB is looking into the biomedical aspects of the condition. The social impact of living with EB – both for patients and their relatives and/or carers

– is far-reaching, as the condition may affect everything from activities of daily living, psychological wellbeing and social interactions to financial issues. However, research into the psychological and social effects of living with EB is limited.

Quality of life

The measurement of QoL, and health-related quality of life (HRQoL) in particular, has become an increasingly important method of evaluating health outcomes and the impact of interventions. HRQoL can be defined as a person's general subjective perception of the effects of illness and health interventions on the physical, psychological and social aspects of their life. The NHS Outcomes Framework highlights the need to enhance the QoL of people living with long-term conditions (Health and Social Care Information Centre, 2018).

A QoL scale should evaluate an individual's lived experience and not be based only on clinical criteria or the health professional's perspective (Bowling, 2001). The challenge for health professionals and researchers is to have a working definition of QoL that is appropriate for people with rare diseases and/or long-term conditions.

The research approach we adopted for our study was explorative in nature, as we sought to gain a better understanding of the experiences and perspectives of people living with EB.

Social support

There is a large body of evidence to suggest that social support affects health outcomes. However, social support is difficult to define and measure. One definition is that it consists of the social resources that a person perceives as being available and/or the social resources they actually receive. It is important to distinguish between 'perceived' and 'received' social support, as there is evidence that people who have a strong 'perceived' sense that social support is available cope better with stressful events than those who do not (Gottlieb and Bergen, 2010).

Social support is a complex notion conceptualised by many theories and models from different health and social care disciplines. According to House (1981), it can be categorised into four types:

- Emotional support: offering empathy, concern, affection, love, trust, acceptance, intimacy, encouragement or caring – also known as esteem support or appraisal support;
- Instrumental support: providing

Fig 1. Junctional epidermolysis bullosa



financial assistance, goods or services – also known as tangible support;

- Companionship support: providing shared social activities that give the person a sense of belonging;
- Informational support: providing advice and/or guidance.

Methods

Review question

When conducting a literature review, it is important to determine a focused review question, as it will underpin all aspects of the methodology. Formulating a review question should enable the researcher(s) to identify the components, inclusion and exclusion criteria, and search strategy for their literature review. The review question in our study was: What research evidence is there in the literature to indicate that there is a relationship between quality of life and social support for people living with the effects of the rare genetic condition epidermolysis bullosa?

Search strategy

Two literature searches were undertaken, one in August 2015 and one in September 2017. The first used the search terms: 'social support', 'epidermolysis bullosa', 'quality of life', 'social care', 'family' and 'psychosocial'. It identified 1,121 papers, which were screened to see if they met the following inclusion criteria:

- Paper had been written in (or translated into) English;
- Research had been undertaken in humans;

- Paper had been published after 1995;
- Paper included the search terms in its title and/or abstract.

Screening left 442 papers, which were assessed based on their full text; this narrowed down the selection to 17 papers.

The vast majority of papers identified in the search focused mainly on the biomedical features of EB, so a second search was conducted, which included papers published since 1967 and unpublished papers.

The second search identified 307 papers, 50 of which were suitable for full-text assessment. Of these 50, six were selected for inclusion in the review. There was no duplication between the 17 papers from the first search and the six from the second, so all 23 papers were used for the final synthesis.

Results

Content of papers

The 23 papers included in the final synthesis came from a range of countries, including Italy, the Netherlands and the US, with the largest number (nine) from the UK. Their publication dates showed there had been an increase in research exploring the psychosocial impact of living with EB in recent years. For the most part, the papers described qualitative studies with various designs, including questionnaires, interviews, surveys and literature reviews.

Adni et al (2012) explored the lived experiences of adults with a diagnosis of EB, including the impact of non-healing wounds on their daily lives and psycho-

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social status. They found very little research into the experiences of people living with EB, but identified six relevant themes:

- Coping;
- Pain;
- Perceptions;
- Emotional impact;
- Social impact;
- Support networks.

They concluded there was a need to adopt a multidisciplinary approach to the management of patients with EB.

Dures et al (2011) also conducted a study into the psychosocial impact of living with EB. It confirmed what all the other papers in our review said about research in this area being scarce. Their aim was to gain access to the experiences, feelings and social lives of people living with EB. They identified three main themes:

- Belief about containing the impact of EB;
- Understanding of the disease and its disabling effects;
- The way in which EB influences daily lives at intra-individual, inter-individual and sociocultural levels.

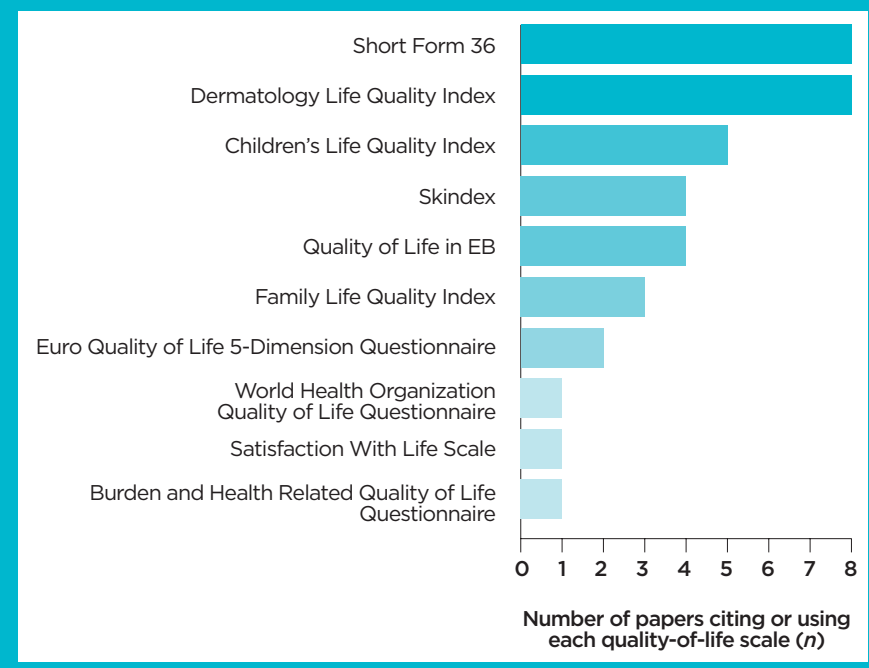
Maćik and Kowalska-Dąbrowska (2015) set out to determine how having a child with EB affects parents' lives and psychological wellbeing. They echoed other researchers' findings that, although many studies explore the effects of EB, most focus on the medical aspects of the condition. Their study was the only one in our literature review that gave a clear definition and measurement of social support, which was said to include four dimensions: informational, instrumental, appraisal and emotional. Their results indicated that:

- The support parents needed most from medical professionals was instrumental support;
- Parents of children who were more severely ill had more emotional support than parents of healthier children;
- Information support had the strongest correlation with perceived QoL.

Margari et al (2010) investigated psychiatric symptoms and QoL in 25 people with EB. They found that 80% experienced psychiatric symptoms and 82% experienced a deterioration in their QoL. The results also clearly demonstrated that the role of the family was crucial to participants' lives. The authors concluded that physical, psychological and social aspects are important in EB and a bio-psychosocial approach to EB management should be adopted.

Williams et al (2011) explored the experiences of young people with EB simplex, focusing on the nature of their personal perspectives. The authors based the

Fig 2. Quality of life scales cited or used in the 23 papers



rationale for their methodology on research cited by Eatough and Smith (2008), adopting an approach based on how people perceive and make sense of their life experiences. They identified three main themes:

- The self as different;
- Independence and dependence;
- Coping.

Participants in the study had the mildest form of EB, but reported feeling a strong sense of being different from their peers and being excluded from activities with those peers. In their accounts, they clearly made the point that their visible difference was one of the social factors that had the deepest impact on their lives.

Van Scheppingen et al (2008) explored the problems experienced by children with EB. They found that the visibility of the skin disorder brought about two social problems that children found difficult to deal with: staring and teasing.

Hubbard and Mayre-Chilton (2015) focused on how people with EB and how their families approached the issue of whether to have a gastrostomy tube placed and whether these tubes affected their QoL. In some people with EB, nutritional intake is compromised, which can result in malnutrition. A gastrostomy tube may help patients achieve an adequate nutritional intake. Four themes were identified:

- Importance of consent;
- Increase in control;
- Normalisation;
- Influence of family.

The researchers concluded that more research on QoL in patients with EB was needed so that, ultimately, patients' life experiences would have a bearing on clinical decisions. They suggested that patients should play an active role in shaping the care and support they receive, as these directly affect their lives.

QoL scales

The 23 papers included in our literature review cited or used ten QoL scales (Fig 2). Among them was the 17-item QoLEB questionnaire, an EB-specific QoL questionnaire, which was used or cited in four papers. Developed by Frew et al (2009), QoLEB is the first EB-specific QoL measurement tool. It takes into account the physical, psychological and social impact of EB. Frew et al (2009) suggested a holistic approach to medicine was extremely important in dermatology as many dermatological conditions have not only physical but also pronounced psychological and social consequences. Frew and Murrell (2010) said EB was a prime example of a dermatological condition with a profound impact on all aspects of health.

Sixteen of the 23 papers cited or used three QoL scales that were developed by Cardiff University:

- The Dermatology Life Quality Index (DLQI) – developed in 1994, this was the first dermatology-specific QoL instrument (Finlay and Khan, 1994); and it was used or cited in eight studies;

Table 1. Psychological and social dimensions of three QoL scales

| Dimension | Children's Life Quality Index | Dermatology Life Quality Index | Family Life Quality Index |
|------------------------|-------------------------------|--------------------------------|---------------------------|
| Symptoms and feelings | Yes | Yes | Yes |
| Daily activities | No | Yes | Yes |
| Leisure | Yes | Yes | Yes |
| Work and school | No | Yes | Yes |
| Personal relationships | Yes | Yes | Yes |
| Treatment | Yes | Yes | Yes |
| School holidays | Yes | No | No |
| Sleep | Yes | No | No |

- The Family Life Quality Index (FLQI) – used or cited in three studies;
- The Children's Life Quality Index (CLQI) – used or cited in five studies.

These three HRQoL scales include not only physical but also psychological and social dimensions (Table 1).

The World Health Organization Quality of Life (WHOQoL) Questionnaire (Skevington et al, 2004) includes social factors, as do the other QoL scales identified in this review, but it is the only one that also has a spiritual dimension (Box 1).

Discussion

There are important implications for practice emerging from this literature review. Although the need for social support was mentioned across the studies, the concept is not well defined and only one study – that by Maćik and Kowalska-Dąbrowska (2015) – defined and measured it.

The focus of much of research into EB is on the physiological features of the condition, rather than on psychosocial aspects. However, all ten QoL scales used or cited in the reviewed studies featured psychological and social dimensions. The drawback is that these various HRQoL scales did not use standardised dimensions in their evaluation of QoL, which makes comparison between studies, and generalisation of their outcomes, more difficult.

When health and social care professionals use HRQoL as an outcome measure, they need to be clear about definitions. Knowing that the medical model of health will focus on the physical features of EB, professionals need to consider the psychological and social factors that affect QoL in people with EB.

Our literature review found qualitative and quantitative associations between QoL

and social support. One key finding was that information support provided using good communication had the strongest correlation with people's perceived QoL (Maćik and Kowalska-Dąbrowska, 2015). This suggests that providing appropriate information and communicating it well is crucial to meet people's needs.

More research is needed into the impact EB has on the lives of people diagnosed with the condition and on the lives of their families and carers.

Conclusion

This literature review supports the idea that social support does have a positive impact on QoL, although more research is needed to refine the idea. We believe a multidisciplinary and bio-psychosocial approach to EB management, in which the perspectives of people living with EB are considered, should be at the centre of professional practice. **NT**

- DEBRA is the only UK charity providing care and support to people with epidermolysis bullosa (EB), and funding research into this potentially fatal condition. It is

Box 1. Dimensions included in the WHOQoL Questionnaire

- Physical health
- Psychology
- Level of independence
- Social relationships
- Environment (including financial resources)
- Spirituality/religion/personal beliefs

WHOQoL = World Health Organization Quality of Life

Source: Skevington et al (2004)

named after the founder's daughter, who had EB. For more information, go to www.debra.org.uk

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