international wound journal

International Wound Journal ISSN 1742-4801

ORIGINAL ARTICLE

Understanding the outcomes of a home nursing programme for patients with epidermolysis bullosa: an Australian perspective

Louise J Stevens[1], Sue McKenna[2], Jennifer Marty[2], Allison J Cowin[3] & Zlatko Kopecki[2,3]

1 Brightsky Australia, Sydney, NSW, Australia
2 Dystrophic Epidermolysis Bullosa Research Association – Australia, Pittsworth, QLD, Australia
3 Regenerative Medicine, Mawson Institute, University of South Australia, Adelaide, SA, Australia

Key words
Epidermolysis bullosa; Home nursing; Wound management

Correspondence to

Z Kopecki, PhD
Regenerative Medicine
Mawson Institute
University of South Australia
Adelaide
SA
Australia
E-mail: zlatko.kopecki@unisa.edu.au

Stevens LJ, McKenna S, Marty J, Cowin AJ, Kopecki Z. Understanding the outcomes of a home nursing programme for patients with epidermolysis bullosa: an Australian perspective. Int Wound J 2014; doi: 10.1111/iwj.12394

Abstract

Epidermolysis bullosa (EB) consists of a spectrum of genodermatoses characterised by skin fragility and various degrees of skin and mucous membrane blistering. Minimal trauma and friction can cause extensive blistering in patients with EB, resulting in a number of complications. However, wound management is the main challenge for these patients because of a high risk of infection, fluid loss and potential development of aggressive squamous cell carcinoma (SCC). Indeed, patients with EB have an increased risk for developing skin cancers compared to the general population. In 2012, a home nursing programme was established in Australia to provide assistance to families or patients with severe forms of EB. Nursing care was provided to patients with severe EB during dressing changes in their homes over a period of 2 years. Both families of patients and nurses were surveyed periodically using a developed questionnaire to assess the benefits of this home nursing and its impact on the patients, their families and the nurses. Key findings included a perceived improvement in quality of life, a better provision of support and improved family life management. These findings are the first to highlight the benefits of this national home nursing programme for EB patients within Australia and demonstrate the continued need and benefit of home nursing for patients with severe skin blistering disorders.

Introduction

Caring for the needs of a person with EB takes its toll on the patient, his or her primary caregiver and the whole family (1). A survey of 22 parents of children and adults with EB, conducted by DEBRA Australia in 2010, revealed that 71% of primary carers of EB patients in Australia were mothers, irrespective of whether the patient was a child or an adult. The amount of respite care (not clinical nursing support) provided to these patients or families varied greatly across Australia (from 2 to 12 hours/week), depending on the state and location of families (unpublished communicated data). These findings highlighted a gap in clinical services in the community and suggested a need for home nursing services for patients with severe EB.

Wound care and dressing changes are an important part of the daily lives of patients living with EB. The greatest impact is experienced by patients with severe skin blistering, irrespective

Key Messages

- patients with the chronic, severe skin blistering condition epidermolysis bullosa (EB) benefit from home nursing as it results in improved quality of life, respite and provision of better support for patients or families with EB and improved family life management
- the aim of this investigation was to evaluate the effectiveness of a home nursing programme that was in place for 2 years and included 15 patients or their families and 25 community registered nurses from both metropolitan and rural regions of Australia
- the findings presented here cover a number of key considerations for the design of future programmes that will capture the true scale of both the need and the benefit of home nursing for patients with EB

doi: 10.1111/iwj.12394

of the clinical EB type (2). In patients with significant skin blistering, special considerations regarding both the patients and their wounds need to be given attention to facilitate optimal healing outcomes. Skin disadherence and poor wound reepithelialisation leads to the development of chronic, inflamed, recalcitrant wounds. These wounds cover large areas of the body and have a high likelihood of developing into scarred tissue and/or squamous cell carcinoma (SCC) (Figure 1) (3). Patients with severe EB have a much higher risk of developing aggressive SCC (2·5% by age 12 versus 1·35% to 2·7% lifetime risk in the general population) (4). The underlying principle for the management of EB wounds is to apply a non-adherent dressing to prevent pain and bleeding on removal and optimise the healing responses to improve the quality of life (5). Care for an individual diagnosed with EB is challenging and one of the most important nursing requirements is to educate individuals, carers and family members about appropriate wound care products and different dressing techniques (1). Good wound care management with effective dressing techniques is one of the most important factors in the management of EB; however, no single method has proved totally effective (6). A consensus approach to wound care in EB (7) and best practice guidelines (8) have been developed to help guide patients, families, carers and health professionals.

The latest reports from the Australian EB registry suggest that there are 243 EB patients in Australia, with a prevalence rate of 10·3 cases per million (9). However, based on worldwide prevalence figures, the number of individuals living with EB in Australia is expected to be closer to a thousand (10,11). In 2010, the Australian Federal Government launched a national dressing scheme called the National Epidermolysis Bullosa Dressing Scheme (NEBDS). This scheme is managed by a not-for-profit organisation, BrightSky Australia, and the NEBDS has improved the access to clinically appropriate dressings and education for over 140 eligible people with EB across Australia (11). Examples of some of the dressings available on the NEBDS Dressings Schedule, along with the recommendations for their use are listed in Table 1.

The objectives of this investigation were to ascertain the need for, and understand the impact of, a pilot home nursing programme for patients with severe EB and their families.

Materials and methods

Home nursing programme design, enrolment and analysis

The clinical investigations were conducted with approval from the SA Department of Health Human Research Ethics Committee, in accordance with the Declaration of Helsinki principles and with written informed consent. Interested participants (families or patients) were enrolled into the home nursing programme through DEBRA Australia. Inclusion criteria specified that only severe EB patients enrolled in the NEBDS were to be included in the study, irrespective of age, gender, location or EB subtype. Initially, three patients from South Australia were enrolled in a 3-month trial; however, the home nursing programme was expanded subsequently on a national level with 15 patients or families enrolled into the study (9 male, 6 female) from various locations across Australia (10 living in metropolitan areas and 5 living in rural Australia) (Figure 2) (Table 2). The average age of patients included in the home nursing programme was 14·8 years and all patients had severe EB, with four different main EB subtypes: EB Simplex (1), Junctional EB (4), Recessive Dystrophic EB (9) and Dominant Dystrophic EB (1) (Table 2). Community nurses who were registered nurses with previous dermatology experience were recruited from different nursing agencies across the country to provide home nursing services. Patients or families were involved in home nursing support during bath/dressing changes for EB patients for two visits per week, each lasting 3 hours.

Prior to the start of the home nursing programme, DEBRA Australia, together with BrightSky Australia, provided a 1-day EB educational training session for all nurses. Education was provided about the different subtypes of EB, the current best practice in EB wound care, and the different dressings available through the NEBDS. Recommendations on care and management of this chronic, complex condition were also given. A total of 25 nurses participated in the home nursing programme. The first meeting and in-home family visit by the nurse was facilitated by a DEBRA Australia national family support coordinator. The home nursing programme was monitored by DEBRA Australia and online surveys of families or EB patients and registered nurses were conducted at 3 months after the start of the home nursing programme, at 1 year and at 2 years after the start of the programme. Survey questions were developed by DEBRA Australia and were used for monitoring the home nursing programme as well as for assessing the need and effectiveness of home nursing for patients with severe EB. Statistical differences were determined using Student's *t*-test or analysis of variance. For data not following a normal distribution, the Mann–Whitney *U*-test was performed. A *P* value of less than 0·05 was considered significant. In all figures, the mean is presented as Mean ± SEM.

Results

Perception and participation in the home nursing programme

The survey response rate for patients or families and nurses was over 80%. The survey responses from all the patients or families and nurses enrolled in the programme at 3 months, 1 or 2 years following their initiation into the home nursing programme showed no significant differences regarding programme participation, key benefits of the programme or level of nursing care provided. Therefore, all survey data across the three time-points of assessment were compiled and analysed. The programme design allocated 6 hours of home nursing per week and, on average, patients or their families used 5·25 hours of home nursing per week (Table 2). Overall, the programme was well received, with 63·3% of patients or families and 90% of nurses wanting to continue being part of the programme (Figure 3). In addition, 90% of patients or families and 98% of nurses stated that the programme had met their medical and professional needs, respectively (Figure 2). Five families decided not to continue with the programme;

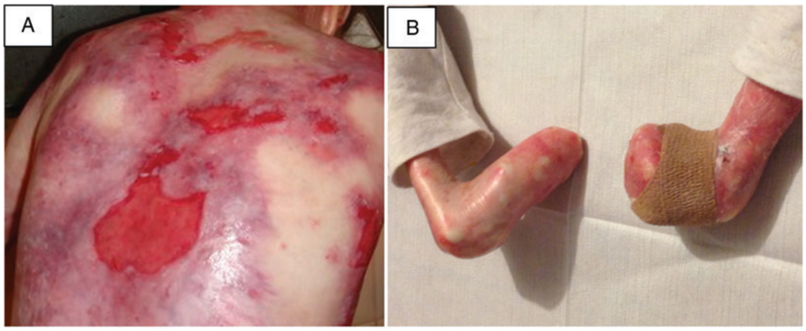

Figure 1 Clinical features observed in patients with severe epidermolysis bullosa (EB). (A) Lage chronic inflamed non-healing wound on the back of an recessive dystrophic EB (RDEB) patient, covering a large surface area. (B) Scarring and pseudosyndactyly in the RDEB patient, often associated with squamous cell carcinoma (SCC) development.

Table 1 Examples of some of the wound dressings available through the National Epidermolysis Bullosa Dressing Scheme (NEBDS) in Australia and their recommended use

Product category	Product name	Indication and/or wound characteristic	Wear time (days)	Rationale	Notes
Contact layer dressing – silicone	Mepitely	Moist wounds and for digits	3–4 days, dependent on condition of wound	Non-adherent soft silicone wound contact layer	There may be a risk of overgranulation when used for JEB
Contact layer dressing – non-silicone	Urgotul	Lightly exuding wound		Non-adherent wound contact layer, non-silicone – preferable for JEB	An option if silicone sensitivity is present Not recommended for very moist wounds due to problems with retention
Exudate transfer dressing	Mepilex transfer	Heavily exudating wounds	Dependent on level of exudate	For large amounts of exudate	To transfer exudate to secondary high absorbent dressing or for conformability, i.e. for digits, axillae and/or difficult to dress areas
Polymeric membrane	Polymem/Polymem Max[2]	Recalcitrant/chronic wounds Acute wounds First-line treatment for newborns with EB	Change frequently initially and then when strikethrough is noted	Cleanses the wound Another non-adherent option for those with sensitivity to silicone	Stimulates high levels of exudate and odour initially Not to be used in conjunction with other solutions, creams or gels
Foam dressings – silicone (thick/thin)	Mepilex/Mepilex Lite	Exudating wounds (for heavy exudate use Mepilex) or as protection for fragile skin	Up to 3–4 days, or when strikethrough is noted	If used as a secondary dressing, these dressings can be changed whilst leaving the primary dressing in place to protect the wound bed	Can be used as a primary dressing (caution with severe EB)
Hydrogels and dressings	Intrasite Conformable	Eroded blister sites. Neonates and infants	Change daily or when dry	May need urgotul as a primary contact layer for newborns with JEB	Useful for the nappy area in newborns. Can be used over nappy creams
Superabsorbant dressings	Releevo Eclypse	Protection Highly exuding wounds	Dependant on level of exudate	Absorption of excessive exudate	

Reprinted with permission from reference 8.

the reasons for this included some patients or their families feeling the programme to be too intrusive into family life, with the need for more consistency of care by the same nurse, as well as changes in family circumstances over time. Two of the patients or families that withdrew from the programme went on to receive other local funding support (which included nursing support), or support via the Federal Government National Disability Insurance Scheme (NDIS), which became available in some states during the course of the home nursing programme. Nurses who decided to leave the programme stated finding a full-time job or seeking a career change as reasons.

Quality of care provided and key benefits of the home nursing programme

To assess the level of nursing support provided and analyse the key benefits of the home nursing programme, all EB patients

Figure 2 Map of Australia illustrating the patient enrolment across the different states of Australia.

Table 2 Participants of the home nursing programme, outlining epidermolysis bullosa (EB) patient gender, age, location, clinical diagnosis and average weekly hours of home nursing utilised during the programme duration

Gender	Age	Metropolitan/ rural	Clinical diagnosis	Average weekly hours utilised
M	3	Metropolitan	SEB	4·0
M	14	Rural	JEB	6·0
M	6	Metropolitan	JEB	6·0
M	4	Metropolitan	JEB	4·5
M	32	Metropolitan	JEB	6·0
F	23	Metropolitan	RDEB	6·0
F	26	Metropolitan	RDEB	6·0
F	24	Rural	RDEB	4·0
M	5	Rural	RDEB	6·0
F	6	Metropolitan	RDEB	12·0
M	4	Rural	RDEB	3·0
M	4	Rural	RDEB	3·0
M	39	Metropolitan	RDEB	6·0
F	12	Metropolitan	RDEB	4·0
F	20	Metropolitan	DEB	2·23

SEB, EB simplex; JEB, junctional EB; RDEB, recessive dystrophic EB; DEB, dystrophic EB.

or families were asked to rate the quality of nursing care provided from excellent to poor. Different registered nurses involved in the programme had varying prior experience with EB patients and the care of complex patients in general. The majority of patients or families ranked the nursing care as excellent (33·2%) and very good (43·3%) (Figure 4). A trend towards a higher ranking was observed in the later assessments of the home nursing programme. Key benefits of the home nursing programme were highlighted by patients or families, where more than one response was taken into account. They included improved quality of life (36·6%), balance of family life with management of EB (or children with EB) (48·2%), improved level of support (88·3%) and other (45%) (Figure 5).

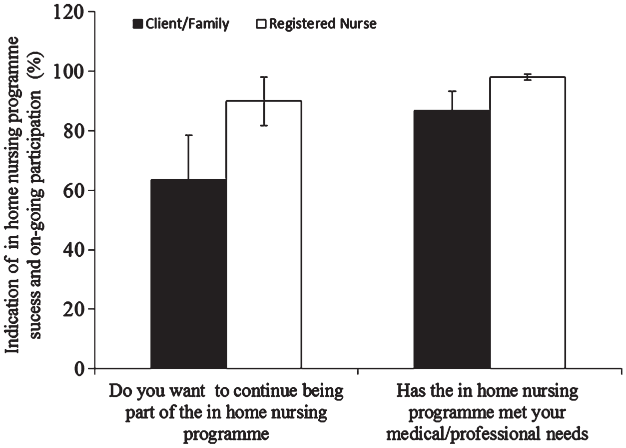

Figure 3 Evaluation of the home nursing programme's success and ongoing patient and nurse participation.

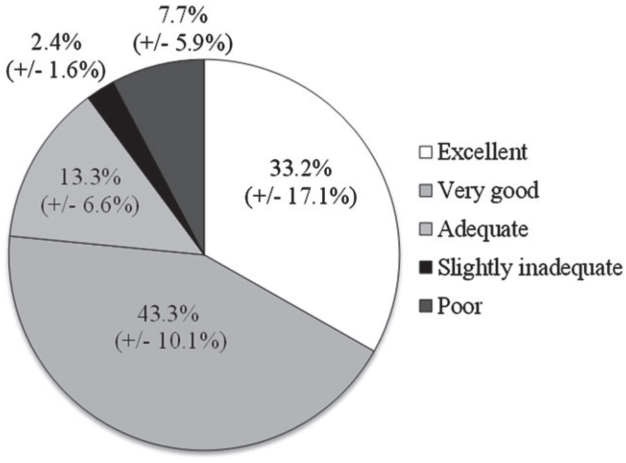

Figure 4 Patient evaluation of the level of nursing care provided during the home nursing programme.

Those who identified other key benefits further commented on the benefits of a registered nurse developing a care plan and screening tool for the development of SCC lesions. Parents also expressed great relief in having the support of professional nursing staff, whose skills could focus on managing recalcitrant wounds or referring patients to various specialist services if or when required.

Patient/family scale assessment of home nursing programme outcomes

An important aspect of this home nursing programme was the analysis and assessment of the programme outcomes, where all EB patients or their families were asked to analyse different programme parameters using a 5-level Likert scale of assessment (1 = strongly disagree, 2 = disagree, 3 = neither agree nor disagree, 4 = agree and 5 = strongly agree). On average, EB patients or their families agreed that they had developed a good relationship with their nurse (score 4·6), they were happy to receive help from a non-family member (score

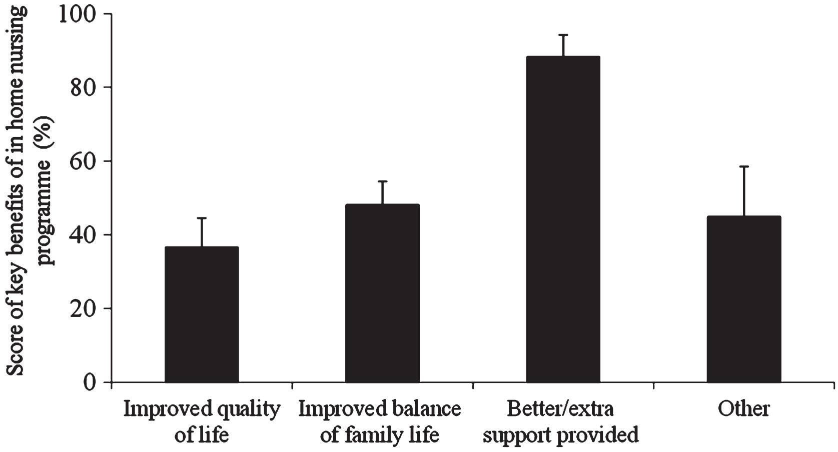

Figure 5 EB patient or family evaluation of the key benefits of the home nursing programme.

4·3) and that two 3-hour visits were adequate for the patient's needs (score 4·1) (Figure 6). In addition, EB patients or their families were neutral about the need for further training of the nurse (score 3·5) or the clear programme benefits of improved wound care and management (score 3·1) (Figure 6). This highlights that the benefits of the home nursing programme were more related to providing patients or primary carers some respite and support rather then necessarily improving wound care outcomes.

Discussion

Currently there is no cure for EB, nor is there any therapy that controls its associated symptoms completely. Pain is the universal feature of individuals living with EB and it is essential that this is addressed and treated effectively. Wound care for this group of patients is highly complex because the disease causes multiple wounds on various areas of the body and at different stages of healing. The management of exudate, malodour and critical colonisation poses complex issues and ongoing challenges. In addition, the selection of wound care dressings and patient or family practice or preference is often case-specific. A number of factors can further complicate the healing of blistered skin, including anaemia, malnutrition, infection, pruritus or SCC (12). Consequently, wound care for EB patients requires a multidisciplinary, coordinated approach that addresses individual-centred care whilst including nurses and primary caregivers (13). This programme aimed to address the need for increased support of EB patients in the community by providing professional registered nurses to assist in the management of wound care and dressing changes for the most severely affected individuals with EB in Australia.

The design of the home nursing programme allocated 6 hours of home nursing per week. It was found that the average weekly hours of home nursing utilised by the EB patient or family varied between families, depending on the severity of the condition, age of the patient and family circumstances, as well as the level of support available from family members other than the primary caregiver. It was acknowledged that patients' needs could indeed fluctuate over time, with the potential to affect the amount of home nursing required. Possible hospital admissions and other family circumstances were also acknowledged variables. However, the survey results suggest that assistance of 6 hours of home nursing per week during dressing changes was adequate time for patients with severe EB. Patients or families who had complex EB needs such as renal dialysis, tracheostomy or gastrostomy tube feeding also specified that additional overnight respite assistance or professional assistance would have been ideal; however, the majority of patients or families in this programme reported that their medical needs were met.

The programme nurses also reported that their professional needs were met thanks to their EB education and training prior to the exposure to complex EB patients. This training also helped them to overcome or accept the common difficulties and frustrations of caring for these patients with severe EB. These feelings resulted from the limited effectiveness of EB treatments, the chronic nature of the ongoing wound management challenges and the limited available resources, together with the perceived lack of expert professionals/service providers. This has been acknowledged by previous studies focusing on the impact of EB on health professionals providing EB care (14). In addition, studies examining the quality of health care for chronic conditions have shown that effective care for patients with chronic conditions cannot be delivered unless there are well-trained health professionals to ensure appropriate delivery (15). Importantly, building trust with the nurse involved has also been identified as a key factor in this home nursing programme, for allowing an effective collaborative relationship between the EB patient or family and the nurse, thereby leading to improved clinical outcomes and the provision of a better quality of care.

The complex care of an EB patient has a psychosocial impact on the individual, his or her primary caregiver and family as well as the nurses and health professionals involved in providing care. A number of studies have analysed these psychosocial effects and developed quality of life surveys specific to EB as the rarity of the disease, its lifelong and hereditary nature and disfiguring impact differentiate it from other chronic conditions (14,16–21). Collectively, these studies identified the most common problems reported by caregivers of patients with EB. They include time spent on care, emotional distress, effects on their physical well-being and increased expenditure

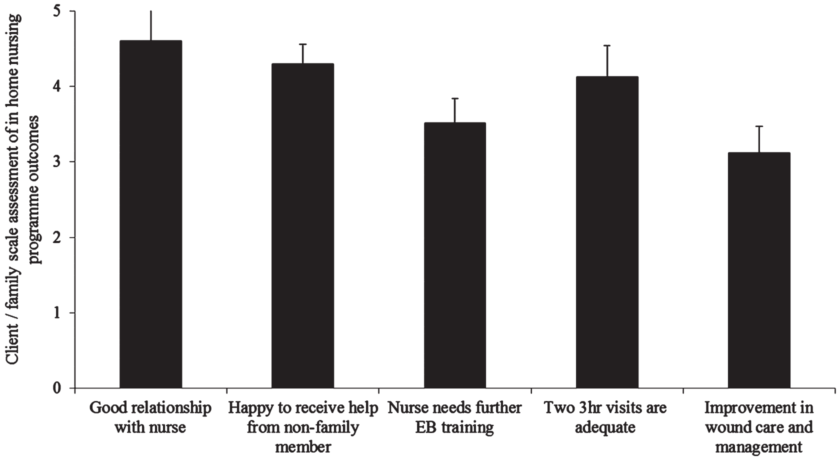

Figure 6 EB patient or family assessment of the home nursing programme outcomes.

(14,16–21). In addition, a qualitative study on families of children with EB identified the main problems experienced by parents to include the following: their children being different, their children's suffering and pain, their own feelings of uncertainty, restrictions on their employment or leisure time, difficulties in organising care or having time off and, lastly, the ignorance of care providers and their lack of skills (19–21). Similar findings were identified in this study, with families welcoming the additional help and, depending on family circumstances, being happy to continue with the programme. Some patients or families commented that 'it was great to get the respite and have a professional registered nurse observing for any suspicious skin/wound changes, focusing on hard-to-heal wounds'. Others also appreciated the 'development of care plans and referrals to specialists'. Increased support provided to primary caregivers allowed ease of mind, especially in relation to the screening of suspicious wounds for SCC development and subsequent referral to specialists. Lastly, in addition to offering a much needed respite to the primary caregiver, the home nursing programme provided the key benefit of facilitating the patient's independence from the primary caregiver. Indeed, studies presented in the National Institute for Health and Clinical Excellence public guidelines have clearly shown that enabling individuals to develop more control can facilitate positive behavioural changes in patients, thereby leading to an improvement in overall quality of life and better long-term health outcomes (22).

The survey highlighted the fact that even experienced nurses found professional development in paediatric or adult dermatology important and that there were significant challenges involved in adapting family or patient practices to the more advanced practices learnt from published guidelines on wound care for EB. Both families and nurses agreed that it takes time for professionals to get used to the families' routines, and the treatment options for individuals with EB. They also agreed that continual education of both the nurses and primary caregivers was essential, as well as their collaboration, to achieve the best clinical outcomes for the patients. In agreement with this finding, previous studies have highlighted the need for continual education of health providers and patients as well as a multidisciplinary integrated approach for the improvement of the health and quality of care of patients suffering from chronic conditions (23). Unsurprisingly, as judged by EB patients or families, the home nursing programme did not identify any significant improvements in specific wound care or management, as the primary caregivers often develop exceptional skills in not only coping with all the challenges that this condition presents, but also in managing the blisters, chronic wounds and complicated dressing change routines. Nonetheless, while assessment of wound care or management was not the primary focus of this programme, both families and nurses agreed on the need for home nursing for provision of respite and adoption of standardised advanced practices developed for EB patients.

A Cochrane review comparing specialist home-based services for children with acute and chronic illnesses identified that current research does not provide supporting evidence of reduction in access to hospital services or readmissions; however, home nursing was found to be a cost-effective means of reducing distress from hospital administrations, while enhancing primary care. In addition, this Cochrane review of studies established that home nursing leads to a significant reduction in the length of hospitalisation following admission (24). This suggests that implementation of home nursing programmes for patients with EB may also provide broader benefits to the community.

In summary, these findings support the need for home nursing for patients with EB, with key benefits including:

- The provision of much needed relief and support for primary caregivers
- A better balance of family life resulting in improved quality of life for patients living with severe EB
- The potential for earlier detection of SCCs and referrals to specialists
- A respite from the hospital system.

While this report has limitations with reference to the use of in-depth scales of assessment and the limited Australian population of individuals with severe EB, the responses to the survey clearly identify the need for, and the benefits associated with, the provision of home nursing support for this group of patients who live with a complex, lifelong condition.

Acknowledgements

The home nursing programme was supported by funding from DEBRA Australia Ltd and Little Heroes Foundation (Adelaide, South Australia). ZK was supported by the NHMRC Early Career Fellowship (#1036509). AJC was supported by the NHMRC Senior Research Fellowship (#1002009). The authors would like to acknowledge Simone Baird (DEBRA Australia) and Samantha Hay (Royal Children's Hospital, Brisbane) for their assistance with the home nursing programme and survey collection. The authors state that they have no conflict of interest to declare.

Author Contribution

All authors contributed to the manuscript preparation and have approved the final submitted and published versions.

References

1. Schober-Flores C. Epidermolysis bullosa: the challenges of wound care. *Dermatol Nurs* 2003;**15**:135–8, 41–4.
2. Kirkorian AY, Weitz NA, Tlougan B, Morel KD. Evaluation of wound care options in patients with recessive dystrophic epidermolysis bullosa: a costly necessity. *Pediatr Dermatol* 2014;**31:**33–7.
3. Lara-Corrales I, Arbuckle A, Zarinehbaf S, Pope E. Principles of wound care in patients with epidermolysis bullosa. *Pediatr Dermatol* 2010;**27**:229–37.
4. Shivaswamy KN, Sumathy TK, Shyamprasad AL, Ranganathan C. Squamous cell carcinoma complicating epidermolysis bullosa in a 6-year-old girl. *Int J Dermatol* 2009;**48**:731–3.
5. Dystrophic Epidermolysis Bullosa Research Association (DEBRA). *Epidermolysis bullosa: an outline for professional*. Berkshire: DEBRA, 2002.
6. Caldewell-Brown D, Gibbons S, Reid M. *Nursing aspects of epidermolysis bullosa: A comprehensive approach. Basic and clinical aspects*. New York: Springer-Verlag, 1992.
7. Pope E, Lara-Corrales I, Mellerio J, Martinez A, Schultz G, Burrell R, Goodman L, Coutts P, Wagner J, Allen U, Sibbald G. A consensus approach to wound care in epidermolysis bullosa. *J Am Acad Dermatol* 2012;**67**:904–17.
8. Denyer J, Pillay E. *Best practice guidelines for skin and wound care in epidermolysis bullosa*. London: International Concensus, DEBRA, 2012.
9. Kho YC, Rhodes LM, Robertson SJ, Su J, Varigos G, Robertson I, Hogan P, Orchard D, Murrell DF. Epidemiology of epidermolysis bullosa in the antipodes: the Australasian Epidermolysis Bullosa Registry with a focus on Herlitz junctional epidermolysis bullosa. *Arch Dermatol* 2011;**146**:635–40.
10. Kopecki Z, Murrell DF, Cowin A. Raising the roof on epidermolysis bullosa (EB): a focus on new therapies. *Wound Pract Res* 2009;**17**:76–82.
11. Stevens LJ. Access to wound dressings for patients living with epidermolysis bullosa – an Australian perspective. *Int Wound J* 2014;**11**:505–8.
12. Denyer JE. Wound management for children with epidermolysis bullosa. *Dermatol Clin* 2010;**28**:257–64 viii–ix.
13. Pope E, Lara-Corrales I, Mellerio JE, Martinez AE, Sibbald C, Sibbald RG. Epidermolysis bullosa and chronic wounds: a model for wound bed preparation of fragile skin. *Adv Skin Wound Care* 2013;**26**:177–88 quiz 89–90.
14. Dures E, Morris M, Gleeson K, Rumsey N. 'You're whatever the patient needs at the time': the impact on health and social care professionals of supporting people with epidermolysis bullosa. *Chronic Illn* 2010;**6**:215–27.
15. Epping-Jordan JE, Pruitt SD, Bengoa R, Wagner EH. Improving the quality of health care for chronic conditions. *Qual Saf Health Care* 2004;**13**:299–305.
16. Dures E, Morris M, Gleeson K, Rumsey N. The psychosocial impact of epidermolysis bullosa. *Qual Health Res* 2011;**21**:771–82.
17. Frew JW, Martin LK, Nijsten T, Murrell DF. Quality of life evaluation in epidermolysis bullosa (EB) through the development of the QOLEB questionnaire: an EB-specific quality of life instrument. *Br J Dermatol* 2009;**161**:1323–30.
18. Frew JW, Murrell DF. Quality of life measurements in epidermolysis bullosa: tools for clinical research and patient care. *Dermatol Clin* 2010;**28**:185–90.
19. Sampogna F, Tabolli S, Di Pietro C, Castiglia D, Zambruno G, Abeni D. The evaluation of family impact of recessive dystrophic epidermolysis bullosa using the Italian version of the Family Dermatology Life Quality Index. *J Eur Acad Dermatol Venereol* 2013;**27**:1151–5.
20. Tabolli S, Pagliarello C, Uras C, Di Pietro C, Zambruno G, Castiglia D, Sampogna F, Abeni D. Family burden in epidermolysis bullosa is high independent of disease type/subtype. *Acta Derm Venereol* 2010;**90**:607–11.
21. Tabolli S, Sampogna F, Di Pietro C, Paradisi A, Uras C, Zotti P, Castiglia D, Zambruno G, Abeni D. Quality of life in patients with epidermolysis bullosa. *Br J Dermatol* 2009;**161**:869–77.
22. National Institute for Health and Clinical Excellence. *Behaviour change at population, community and individual levels* (Public health guidelines). London: NICE, 2007:1–57.
23. Krause CM, Jones CS, Joyce S, Kuhn ME, Curtin K, Murphy LP, Krause CM, Boan B, Lucas DR. The impact of a multidisciplinary, integrated approach on improving the health and quality of care for individuals dealing with multiple chronic conditions. *Am J Orthopsychiatry* 2006;**76**:109–14.
24. Parab CS, Cooper C, Woolfenden S, Piper SM. Specialist home-based nursing services for children with acute and chronic illnesses. *Cochrane Database Syst Rev* 2013;**6**:CD004383.