International Journal of Women's Dermatology 4 (2018) 49–53

Contents lists available at ScienceDirect

International Journal of Women's Dermatology

Psychosocial impact of inherited and autoimmune blistering diseases☆,☆☆

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ARTICLE INFO

Article history:
Received 19 October 2017
Received in revised form 7 November 2017
Accepted 7 November 2017

Keywords:
Epidermolysis bullosa
autoimmune blistering disease
quality of life
self-esteem

ABSTRACT

Inherited and autoimmune blistering diseases are rare, chronic, and often severe disorders that have the potential to significantly affect patients' quality of life. The effective management of these conditions requires consideration of the physical, emotional, and social aspects of the disease. Self-esteem is integral to patients' ability to cope with their illness, participate in treatment, and function in society. This article discusses quality-of-life studies of patients with blistering diseases with a particular focus on self-esteem issues that patients may face.

Introduction

Blistering diseases encompass a heterogeneous group of congenital and acquired skin diseases of variable severity, and symptoms range from mild to life threatening. Many blistering diseases are chronic and burdensome afflictions with the capacity to considerably impinge on patients' quality of life (QoL). Physical symptoms including pain and itch, disfigurement due to blistering and scarring, reduced functional capacity that is imposed by disability, and the economic burden and side effects that are associated with treatment all contribute to the burden of these diseases (Sebaratnam et al., 2012).

Given the pervasive impact of blistering diseases on patients' lives, the emotional impact that is associated with these diseases is also significant. Patients' emotional health is particularly important to consider when evaluating individual disease burden because it is often independent of clinical severity and thus may be invisible to clinicians unless specifically sought out. Similar to other dermatological diseases, patients with blistering diseases can experience shame, poor self-image, and low self-esteem (Jafferany, 2007).

Self-esteem refers to an individual's sense of worth and is intrinsically tied to concepts such as self-confidence and body image (Mazzotti et al., 2011). Poor self-esteem can affect how patients cope with their disease, their outlook, adherence to treatment, and social functioning. Additional challenges that are faced by patients with blistering diseases include the rarity of these diseases, which results in poor awareness of the disease among the wider population, as well as the lifelong nature of blistering diseases with only variable treatment efficacy and limited supportive resources available.

During the past decade, there has been increasing interest in measuring the burden of blistering diseases in an effort to understand patients' experiences living with the disease and the subjective and objective effects of therapeutic interventions (Dures et al., 2011; Tabolli et al., 2009). A variety of generic and disease-specific QoL tools have been used to quantify disease severity and impact. These include generic measures such as the Medical Outcome Study 36-item Short-form Survey (SF-36), dermatology-specific measures such as the Dermatology Life Quality Index (DLQI), and disease-specific instruments and qualitative studies (Sebaratnam et al., 2012).

These tools have facilitated awareness and understanding of patients' perceptions of their disease and the impact of the disease on their wellbeing. Patients and clinicians often have different perceptions of QoL, and regular consideration of the physical, emotional, and social impact of the disease is vital to ensure the provision of holistic care. This article discusses QoL in patients with congenital and acquired (autoimmune) blistering diseases with a particular focus on the impact that living with these conditions has on patients' self-esteem as well as implications for future practice.

☆ Sources of support: This research did not receive any specific grant from funding agencies in the public, commercial, or not-for-profit sectors.
☆☆ Conflicts of interest: The authors have no conflicts of interest to declare.
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https://doi.org/10.1016/j.ijwd.2017.11.004

Inherited blistering diseases

Epidermolysis bullosa (EB) encompasses a group of inherited blistering diseases that are characterized by the blistering of the skin and mucous membranes after mild mechanical trauma. The main subtypes of EB are delineated on the basis of genetic and ultrastructural characteristics of disease and include EB simplex, junctional EB, and dystrophic EB. Recessive dystrophic EB (RDEB) is the most severe form of the disease with the greatest potential for internal organ damage, significant disability, and early death (Figs. 1a and b). Unfortunately, EB is a noncurable disease and the mainstay of current management is supportive treatment in the form of painful and time-consuming dressing changes. However, a number of new clinical trials and biological therapies offer hope, at last, of a treatment that could control EB.

Living with EB can exert a detrimental impact on the physical, emotional, and social health of afflicted patients with the burden of disease varying between the subtypes. A Scottish study of 116 patients with EB found that according to the DLQI, a validated dermatology-specific QoL tool, the impairment in QoL of patients with RDEB was greater than that of patients with any other skin disease previously assessed (Horn and Tidman, 2002). Children experienced a greater impact on their QoL compared with adults with higher average DLQI scores for both severe (22 of 30 in children; 18 of 30 in adults) and nonsevere (15 of 30 in children; 10.7 of 30 in adults) subtypes of EB. One contributor to this difference was the inability of children to participate in everyday activities due to the disease symptoms. This resulted in psychological and social sequelae and children reported feelings of isolation due to difficulties in engaging with peers.

A qualitative study that used semistructured interviews in 11 children with EB further demonstrated the frustration that children felt with the limitations that were imposed by EB. The inability to participate in common childhood activities such as sports was the third most prevalent concern after pain and itchy skin and contributed to affected children feeling different from their peers (van Scheppingen et al., 2008). This sense of being difference was exacerbated by the visibility of EB, and some children reported an awareness of a difference only after negative reactions or comments by others.

The highly visible nature of EB both in terms of blisters and dressing for treatment is particularly challenging for children at a time when concepts of self and body image emerge. Children are at a particular risk of internalizing negative reactions and therefore also at risk to develop body image-related distress and subsequent self-esteem issues (Jafferany, 2007). Indeed, one study suggested that restricted social interaction and increased psychological morbidity in patients with EB may cause adolescents in particular to be shy and introverted (Frew and Murrell, 2010).

In addition to social isolation, self-esteem may also be affected by the loss of autonomy that EB patients experience, particularly those patients with severe subtypes of the disease. A study of 425 patients with EB found that 73% of patients with severe forms such as junctional EB and recessive dystrophic EB required full assistance with activities of daily living (particularly walking and personal hygiene). In contrast, more than 90% of patients with less severe forms of EB such as EB simplex were fully independent (Fine et al., 2004).

However, clinical severity is often not associated with the severity of psychological symptoms in patients with EB, which is a phenomenon that is well-reported in other dermatological diseases. One study found that although 80% of patients with EB had subthreshold psychiatric symptoms such as anxiety and depression, symptoms were not correlated with disease severity (Margari et al., 2010) and family support and a multidisciplinary EB team along with patient advocacy groups such as the Dystrophic Epidermolysis Bullosa Research Association of America were important to help patients cope. Indeed, another study of a patient population with EB in Italy demonstrated that EB had the greatest impact on QoL in patients with higher perceived severity rather than clinical disease severity, with other contributors including higher psychological distress as measured by the General Health Questionnaire-12 in female and pediatric patients (Tabolli et al., 2009). These results mirror those from patients with significant burns, among whom self-esteem was found to be more affected by age and sex than by the size of the burns or the part of body that was burned (Bowden et al., 1980).

Interestingly, another study that developed an internationally validated, disease-specific, QoL tool for EB called the Quality of Life in EB questionnaire (QoLEB) demonstrated that patients with RDEB had better scores on the emotions subscale (measurement of patient-reported frustration, embarrassment, depression, and anxiety) compared with patients with less severe junctional EB (Cestari et al., 2016; Frew et al., 2009, 2013; Yuen et al., 2014). The authors postulated that this difference could be because patients with severe disease have well-developed coping mechanisms to adjust to their illness, with family and caregiver support and an individual positive outlook as key factors to develop emotional resilience (Dures et al., 2011; Frew et al., 2009).

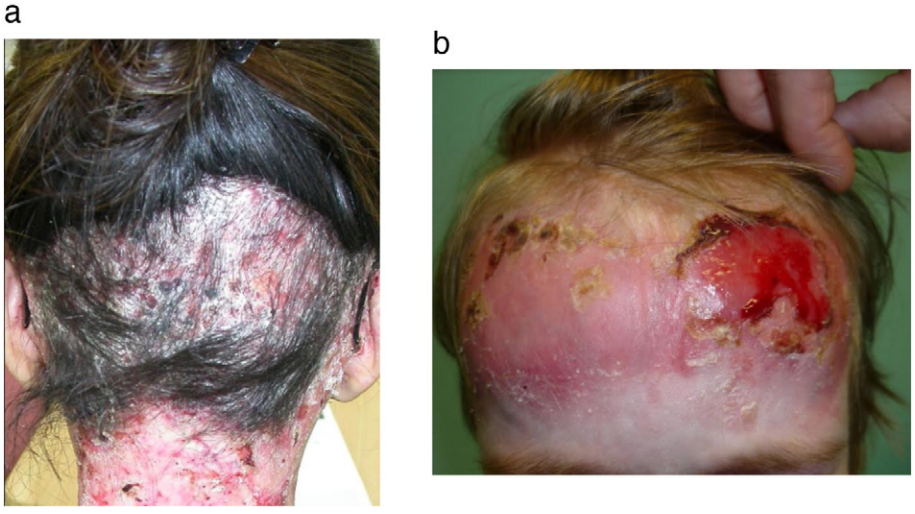

Fig. 1. (a) and (b). Recessive dystrophic epidermolysis bullosa with visible erosions and scarring alopecia that causes embarrassment and pain