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IECBS 2022 The 3rd International Electronic Conference on Brain Sciences
01-15 OCTOBER 2022 | ONLINE
Neuropsychology
Sciforum – 063422
brain sciences
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Reference models for children and young people with Epidermolysis bullosa

First case report on the challenges, rewards and limiting factors for successful academic and professional achievements

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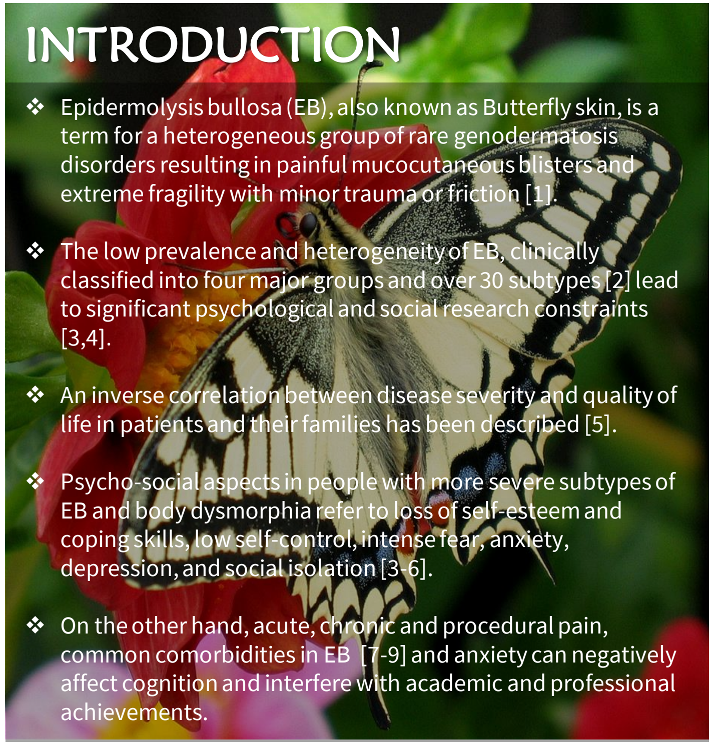

INTRODUCTION

- Epidermolysis bullosa (EB), also known as Butterfly skin, is a term for a heterogeneous group of rare genodermatosis disorders resulting in painful mucocutaneous blisters and extreme fragility with minor trauma or friction [1].
- The low prevalence and heterogeneity of EB, clinically classified into four major groups and over 30 subtypes [2] lead to significant psychological and social research constraints [3,4].
- An inverse correlation between disease severity and quality of life in patients and their families has been described [5].
- Psycho-social aspects in people with more severe subtypes of EB and body dysmorphia refer to loss of self-esteem and coping skills, low self-control, intense fear, anxiety, depression, and social isolation [3-6].
- On the other hand, acute, chronic and procedural pain, common comorbidities in EB [7-9] and anxiety can negatively affect cognition and interfere with academic and professional achievements.

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AIMS

Here, we present the first case report of a current international project that searches for reference models for children and young people with EB and the creation of a guidebook with different life stories of strategies and skills proven to help achieve academic/professional success despite EB's condition.

METHODS

Participants: Inclusion criteria was to be a patient with EB according to the consensus reclassification of EB [2], the will to participate, to be an adult or minor with informed consent from parents to be interviewed and to use the written and image material for scientific purposes and divulgation. Exclusion criteria was to be unable to be contacted or interviewed or to sign informed consent for the above issues.

Data recording: After informed consent, the participant was interviewed by two clinical researchers in Medical Psychology and Psychiatry from the Universitat Autònoma de Barcelona.

Data analysis: The transcript of the semistructured interview was assessed using qualitative content analysis.

RESULTS

After considering several candidates according to the inclusion/exclusion criteria, one adult (woman) with Recessive dystrophic epidermolysis bullosa (RDEB), the most severe form of EB [8] from DEBRA Mexico Monterrey, Mexico was contacted. The first participant to be involved in the project was Sonia Ortiz Romero, alias Mariposa de Cristal (Crystal butterfly). Several idiosyncratic characteristics made her eligible as the first case report: She can be considered an expert patient, as in terms of health education in chronic diseases, an expert patient can be defined as the one that is motivated, knows, searches and accepts help to care him/herself and is confident to be able to do it [10]. She presents RDEB involving the most complex clinical and psycho-social scenario. She played the role of expert patient as a member of Debra Mexico Monterrey with peers and schoolers in her city of Santiago Tilapa, and after receiving several recognitions, she was co-founder of ‘Son mis alas, I.A.P’ (‘They are my wings, NGO’), together with his brother Marcos Humberto. Her storytelling illustrates challenges, rewards, and limiting factors that make their Mexican association a successful case in helping other EB patients in Santiago Tianguistenco with their physical, psychological, and social well-being, as well as raising social awareness.

(Images use restricted to this scientific research work)

Sonia Romero, Mariposa de Cristal profile

‘Son mis alas’ team

25-31 October Awareness week campaign

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