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The psychological functioning of children with Epidermolysis Bullosa (EB) and its relationship with specific aspects of disease

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Dear Editor

Epidermolysis Bullosa (EB) is a group of genetic conditions resulting in skin and mucosal membrane fragility. EB is characterised by chronic wounds and scarring, consequent functional limitations and high levels of pain. In its most severe forms, life expectancy is significantly foreshortened.

Research on psychological adjustment in EB is scarce. Existing studies are limited by small sample sizes and lack of robust psychological measurement. In the face of such a complex disease, there is little to guide psychosocial management.

In this study, we firstly described the psychological profile of young people with EB using the most robust, standardised psychological measures available. Secondly, the relationship between psychological functioning and the most common physical symptoms of EB: unhealed skin, pain and gastro-intestinal involvement as well as EB sub-type, was investigated. Thirdly, parental stress was measured due to the burden of daily care on parents and the proven association between parent and child mental health.

Participants were 81 patients at a UK National Centre of Excellence for paediatric EB and their parents which constituted 70% of the available population. Patients were between eight and 14 years of age due to age limitations of the psychological measures used. Participants were grouped into the EB sub-categories of Recessive Dystrophic; Dominant Dystrophic; Simplex and Other. The Other group was excluded from statistical analyses due to small numbers and the broad range of disease presentations. Child patient participants completed the Beck Youth Inventories (BYI)[1] which measures anxiety, depression, anger, self-concept and behavioural difficulties, the Children's Dermatological Life Quality Index (CDLQI)[2] and a pain rating.[3] Parents completed the Strengths and Difficulties Questionnaire (SDQ)[4] which generates six subscales: Emotional Symptoms, Conduct, Hyperactivity, Prosocial Behaviours, Peer Relationship Problems and a combined Total Difficulties scale; the Perceived Stress Scale[5] and

existing clinical tools to describe the area of skin unhealed and gastro-intestinal symptoms.[6] Differences between the EB group and the general population were analysed using a series of T-tests while associations between variables were analysed using Pearson's two-tailed correlational analyses.

The CDLQI for this sample showed significantly better health-related quality of life (mean Total Score=7.75 (SD=6.33) than reported by a paediatric sample from the Scottish EB Register (mean Total Score=16.2)[7] but significantly worse than a general paediatric dermatology sample (mean Total Score=5.1 SD=4.9).[2]

Parent-reported measures scored the EB group significantly higher than general population norms on Total Difficulties (mean=10.8, t=3.355, p<.001), Emotional Symptoms (mean=3.43, t=5.098, p< .001) and Peer Relationship Problems (mean=2.25, t=3.540, p<.001). Sixty-six percent of children with RDEB and 50% with EB Simplex scored in the clinical range for Emotional Symptoms and 50% of the RDEB group and 40% of the EBS group were in the clinical range for Peer Relationship Problems meaning that they are twice to three times more likely than the general population to have clinically significant difficulties in these areas. The DDEB group did not differ significantly from the general population. Patient-reported measures co-varied with parent-reported measures. However, they did not differ significantly from the general population on any psychological index.

Severity in all three symptom domains of unhealed skin, pain and gastro-intestinal symptoms was significantly correlated with parent-rated Emotional Symptoms. Pain was also significantly correlated with self-reported depression (r=.327, p=.004), anxiety (r=.289, p=.011) and self-concept (r=-.249, p=.031) while gastro-intestinal symptoms was also associated with self-reported depression (r=.235, p=.049) and behavioural difficulties (r=.425, p<.001).

Stress for both mothers (n=79) and fathers (n=45) was not significantly elevated compared to the general population. Mothers and fathers showed concordance in their

stress levels but only maternal stress was associated with one medical symptom index which was unhealed skin (which accounted for approximately 9% of variance) which had some face validity in that mothers are often responsible for daily skin care.

This study demonstrated that young people with more severe expressions of EB were more likely to have difficulties relating to anxiety, depression and peer relationships.

Clinically, this study can help to identify patients most at risk of psychological difficulty. Findings also highlight the importance of asking multiple informants about patient mental health because of the divergence in child- and parent-report. While parents reported normal stress levels, this does not obviate
responsibility from care teams to support parents with their additional burdens. Anecdotally, parents can under-report their struggles in order to not detract from their child's needs.

Further research is required to determine whether these findings are generalised to other paediatric EB groups. This sample was drawn from a highly specialist and well-resourced multi-disciplinary unit and some EB sub-types were excluded for statistical purposes.

Table

Table 1

Participant Demographics

		%	(n)
EB Sub Type	DDEB	37	(30)
	EBS	26	(21)
	RDEB	22	(18)
	Other	15	(12)
Gender	Female	52	(42)
	Male	48	(39)
Ethnicity	White British	78	(63)
	Asian	13.5	(11)
	Other	8.5	(7)
Age	Mean	10-4	
	Range	8-0 - 14-11	

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