




Case Cohort Review of 75 children with Epidermolysis Bullosa

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Background	Methods	Results	Discussions and Conclusions
<ul style="list-style-type: none">Epidermolysis bullosa is a genetic condition that affects connective tissue and can cause blisters in the skin and mucosal membranesThe prevalence of this disease in the United States is 8 per million as reported by the National Epidermolysis Bullosa Registry StudyIt can be hard for families to establish a dental home with dentists who feel confident in treating patients with EB and providing anticipatory guidance on oral healthMastication abilities and oral hygiene of those individuals with EB can be affected due to poor dentition, limited opening, oral and esophageal blistering, and strictures of esophagus	<p>Caregivers/Patients completed a 15-item oral health intake form in this IRB exempt study.</p> <p>Questions surveyed included demographics, dental history, primary nutrition, oral findings associated with EB and access to dental care.</p> <p>EB specific variables included; EB subtype, primary nutrition, presence of intraoral blisters, microstomia, ankyloglossia, enamel hypoplasia, crowding, oral habits, brushing habits, toothbrush type and toothpaste, how far they travel to appointments, and pain scale</p> <div><p>Evidence of extra-oral blistering, commissural fibrosis and microstomia often associated with dystrophic EB.</p></div> <div><p>Obliteration of hand folds with Recessive Dystrophic EB (RDEB) this can make oral hygiene significantly challenging</p></div>	<p>Data were collected from 75 patients over a three-year period. (2018-2021)</p> <p>Most patients had a diagnosis of Dystrophic EB (61%) with 73% of those having a recessive inheritance pattern.</p> <p>Nearly two-thirds (65%) of patients had exclusive oral nutrition with a significant number of dystrophic (<i>p=0.05</i>) having exclusive g-tube nutrition.</p> <p>Over half of patients (58%) reported weekly oral blistering, with the majority (37%) being on the tongue.</p> <p>56% were found to have some degree of microstomia and ankyloglossia (49%),and dental crowding (62%).</p> <p>8% reported moderate-severe dental pain, 57% reported having to travel >50 miles for dental access, with no difference between EB subtype (P=0.78)</p> <div><p>Opthalmic lubrication, pulse oximetry on ear and care with shearing forces on skin are among challenges of providing care to EB patients (photo used with permission)</p></div>	<ul style="list-style-type: none">The sample size in this study is the largest documented on epidermolysis bullosa and oral health findings.Our study shows that majority of individuals with epidermolysis bullosa must travel to a dentist to receive their dental careWhile most patients report oral nutrition, pain was a common featureChildren with EB need oral hygiene counseling to minimize the amount of intra-oral blistering and pain associated with daily brushingIt important for dentists to be knowledgeable and comfortable with treating children with epidermolysis bullosa to increase accessibility to careEpidermolysis bullosa is a complex medical condition that needs further research on how to reduce pain during daily oral hygiene
Objective	References		
<p>The purpose of this study was to review oral health findings and practices in a cohort of 75 subjects with Epidermolysis Bullosa (EB). Additionally, this study examined nutrition and pain impact.</p>	<ul style="list-style-type: none">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. 2010 Jun;146(6):635-40.Birge K. Nutritional Management of Patients with Epidermolysis Bullosa. J Am Diet Assoc 1995;95:575-579.Zidorio et al. Nutritional aspects of children and adolescents with epidermolysis bullosa: literature review. An Bras Dermatol 2015 Mar-Apr;90(2):217-23.Liversidge HM, Kosmidou A, Hector MP, Roberts GJ. Epidermolysis bullosa and dental developmental age. Int J Paediatr Dent 2005 Sep;15(5):335-41.		