



Dental management of a case of severe epidermolysis bullosa in a pediatric patient.

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BACKGROUND

Epidermolysis bullosa (EB) is a rare genetic skin disorder categorized by blistering due to mechanical trauma. This disorder presents in 4 different types: dystrophic EB, EB simplex, junctional EB and Kindler Syndrome. The fragile blistering skin occurs on extra-oral tissue, but in extreme cases, may occur on internal mucosal tissues such as the mouth and the lining of the stomach. Common oral findings of the disease include microstomia, intraoral ulcerations and bullae formation, ankyloglossia, tongue atrophy, elimination of buccal and vestibular sulci, lingual depapillation and atrophy of the palatal folds¹.

CLINICAL PRESENTATION

A 7-year-old female patient with medical history significant for epidermolysis bullosa presents to Riley ED for influenza A and adenovirus. Patient has been followed by a pediatric dentist and parent was concerned that ulcers were secondary to infection of dental origin. Patient's symptoms include bilateral mouth and cheek pain as well purulent discharge occurring more than 1 month. Clinical exam revealed multiple ulcers on extraoral and intraoral tissues. The lesions were painful and oral tissues were very friable, making oral health care incredibly challenging. In addition, patients with severe EB have been shown to have a high caries rate due to severity of soft tissue involvement, diet which is high in sucrose due to provide necessary nutritional intake, and inability to physically remove dental plaque due to limited ability to use a toothbrush².



MANAGEMENT AND FOLLOW UP CARE

Patients with severe EB should have the following preventative and oral care instructions relayed to their primary caregiver:

1. Enhance oral clearance of food debris with increased fluid intake
2. Using a waterjet system to mechanically remove plaque and food from between and around the dentition
3. Multiple rinses with fluoridated mouthwash, prescription-strength fluoride mouth rinse, chlorhexidine have all been shown to help control caries
4. Utilization of neutral sodium fluoride varnishes
5. Sucralfate prescription, which has been shown to be an effective prophylactic and therapeutic agent³

REFERENCES

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