

# Epidermolysis Bullosa and Oral Health: Problems and Solutions

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Individuals with EB may have teeth with severely malformed enamel (enamel hypoplasia) and/or dental caries depending on the EB type and the presence of other risk factors for developing tooth decay. The enamel is usually normal in simplex and dystrophic EB types. Generalized enamel hypoplasia is typically limited to Junctional EB. Rarely individuals with non-Junctional EB types may have generalized enamel hypoplasia. Rampant dental caries occur in Junctional EB partly because of the enamel hypoplasia. Dental decay also is frequently seen in patients with severe recessive dystrophic EB. This excessive dental caries results from severe soft tissue involvement which leads to dietary changes (soft and high carbohydrate), increased oral clearance time (secondary to limited tongue mobility and oral scarring), and creates an abnormal tooth/soft tissue relationship. Oral involvement also reduces the ability to practice preventive measures directed at reducing caries.

It is currently recommended that all children have a dental home by one year of age. Because dental caries can form rapidly in individuals with recessive dystrophic and Junctional EB, it is even more critical that dental examinations begin by 1 year of age and be conducted at least twice a year. If caries becomes a problem, then more frequent visits (4 times a year) are indicated for preventive treatments and examination. Individuals with mild EB can be treated much as any other patient. The dentist should, however, be made aware of any history of mucosal fragility and oral blistering since dental therapy can precipitate oral lesions even in mildly affected patients. Many dentists are not familiar with EB. Therefore the patient or parent often must help educate the health care team. An altered approach to treatment may be required in individuals with enamel hypoplasia or rampant caries, extreme fragility of the mucosa and/or the presence of microstomia (a decreased oral opening size). Individuals with severe soft tissue involvement requiring multiple restorative and/or surgical procedures are often best managed with general anesthesia.

Preventing tooth decay is most challenging for individuals with severe mucosal involvement. In patients prone to oral blistering, oral hygiene may best be accomplished with a soft bristled, small headed toothbrush. Many small headed children's toothbrushes are available, some of which have special grip handles that may be helpful to individuals with hands involvement. Running the bristles under hot water prior to brushing makes them even softer. Parents need to brush children's teeth until about the age of 6 or 7 years because children lack the manual dexterity to properly clean their teeth. Parents should be very careful not to damage the gums or make the brushing experience negative and unpleasant. It is important, however, that the teeth be cleaned at least once a day preferably just prior to bedtime.

Talk with your dentist to make sure that your fluoride exposure is optimal. In addition to the systemic fluoride consumed by natural or artificially fluoridated water sources there are a variety of other fluoride delivery systems that may be beneficial. For individuals prone to developing cavities there are special high strength prescription fluoride toothpaste. Strongly flavored toothpaste (mint) may be irritating to the individuals with severe oral involvement, however, there are numerous non-mint flavors available. Bubble gum flavor is a big hit with children (and some adults). Non-alcoholic rinses with greater amounts of fluoride are available by prescription. Fluoridated toothpaste is recommended as a safe and highly effective way to reduce tooth decay when used appropriately. In children under the age of 3 a lateral smear or grain of rice sized amount is adequate to deliver the fluoride to the teeth. Children with certain forms of EB and marked soft tissue changes may continue to have problems rinsing or swishing and should continue to use only small amounts of fluoride toothpaste.

There are a variety of fluoride treatments that can be applied by dentists. The most common treatment for individuals with EB is the 5% Sodium Fluoride Varnish. This treatment is provided by your dentist and it is important that it be done at least twice a year to be effective. For individuals with high caries activity more frequent applications may be beneficial.

The diet constitutes major difficulty in caries control, and due to the complex systemic nutritional demands of individuals with severe EB types, this may be best managed with the assistance of a dietician. The effects of any diet planning should be considered with regards to dental health and tooth friendly foods (cheese, vegetables, fresh fruits) eaten as much as possible. Be careful of the less obvious cavity producing foods such as highly sweetened breakfast cereals, raisins and dried fruits. Cavity producing oral bacteria can ferment carbohydrates from a wide variety of foods. Other helpful hints are to rinse the mouth or drink water after eating if brushing is not possible. Chewing gum that is sweetened with Xylitol or Sorbitol can provide additional protection against tooth decay when chewed after meals and might be considered useful in individuals that can chew gum and are not able to clean their teeth adequately due to severe soft tissue involvement. Bottle or breastfeeding infants at bedtime can result in early childhood caries after the teeth are present (1 year of age). If nursing continues after 1 year, great care should be taken not to give the bottle with anything but water while the infant is going to bed. Virtually any bottle-fed liquid, except water, can cause tooth decay of the baby teeth.

Individuals with EB that develop tooth decay can have fillings and crowns. The type of restorations and the method of placing them will depend on the extent of tooth destruction and the severity of soft tissue involvement. In severely affected individuals these treatments may be best provided with the aid of a general anesthetic. Specific anesthetic protocols are used to prevent soft tissue damage and a safe anesthetic experience. While normal dental X-rays that are placed in the mouth may not be possible in EB cases having severe soft tissue fragility, information can be obtained using panoramic radiographs to evaluate the dentition and surrounding bone. Individuals with even the severest forms of EB can now maintain their natural dentition

providing them the ability to chew, eat a healthy diet and have a pretty smile. In the not too distant past, dental extraction was considered the treatment of choice for individuals severely affected with EB. Now we are able to prevent tooth decay, restore malformed enamel and help produce good alignment of the teeth.

The future looks even brighter as dental prevention continues to improve, new dental materials are stronger and more esthetic than ever and new technologies, such as dental implants, continue to improve. Dental health for all EB patients has become a reality, and that is really something to smile about.

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