

Epidermolysis Bullosa - Dental Considerations

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Important Aspects and Objectives of Dental Care

- Providing dental care that prevents the development of dental disease and minimizing the potential for EB complications
- Provide treatment strategies that provide optimal dental outcome
- Provide treatment strategies that enhance medical procedures
- Considering expectations of patient and family



Important Aspect of Epidermylosis Bullosa

- Understand EB
- Understand the signs and symptoms that may arise
- Understand the treatment risks and complications
- Be aware of others with experience in the area
- Be aware of resources of information



Oral Manifestations in the Epidermolysis Bullosa Spectrum

Consideration of four major EB groups:

- EB Simplex, localised generalised and Dowling- Meara
- EB Junctional Non –Herlitz, Herlitz
- EB Dystrophic Dominant and Recessive
- Kindler EB, Mixed EB

Oral Manifestations in the Epidermolysis Bullosa Spectrum

- Oral Blistering
- Oral Scarring
- Microstomia
- Enamel Defects
- Ankyloglossia



Epidermolysis Bullosa Oropharyngeal Severity (EBOS) score: A
multicenter development and reliability assessment
Fortuna et al JAAD 2013 VOL 68(1) 83-92

EB Simplex

- The genes associated with EB simplex all cause intraepidermal cleavage in the skin and are expressed by the oral mucosa.
- Oral Blistering occurs secondary to trauma or tissue manipulation.
- Localised and generalised will blister but heal without scarring
- Dowling – Meara can display some oral scarring
- Normal dental enamel and salivary flow

Junctional EB

- The proteins transcribed from the genes associated with Junctional EB are important in epithelial cell adhesion and the developing tooth bud.
- Oral blistering in both groups
- Enamel defects occur in both subgroups
- Microstomia occurs in Herlitz type EB
- Increased risk for dental caries
- Saliva flow seems normal in most individuals with junctional EB subtypes.

Dystrophic EB

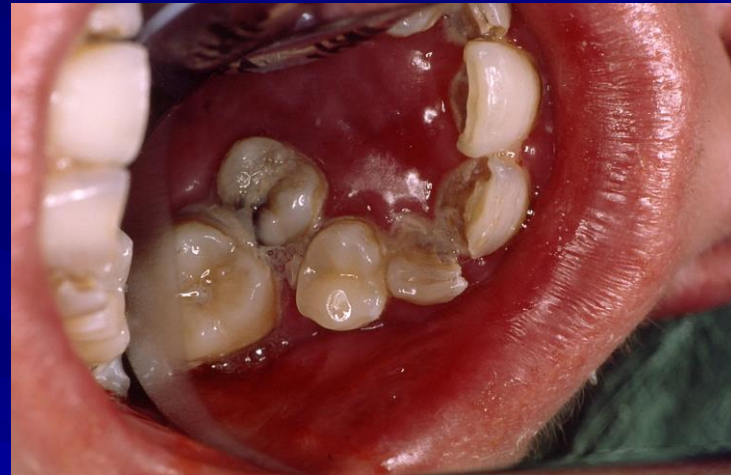
- The gene that is mutated in dystrophic EB is not expressed by the ameloblast
- Dominant DEB can have oral blistering and scarring
- Recessive dystrophic EB can have oral scarring with associated ankyloglossia, loss of vestibula architecture
- Microstomia occurs
- Due to changed oral anatomy there is reduced ability to clear food and consequently increased oral clearance time
- Apparent normal salivary flow rates

Kindler type EB

- Oral Blistering in neonates can be severe
- Severity reduces with age
- Individuals are at risk of developing periodontal (gum) disease during teenage years
- The Kindlin-1 protein is expressed by the epithelium that attaches the oral mucosa to the tooth consequently risk of periodontal disease
- Oral blistering, scarring and microstomia occur

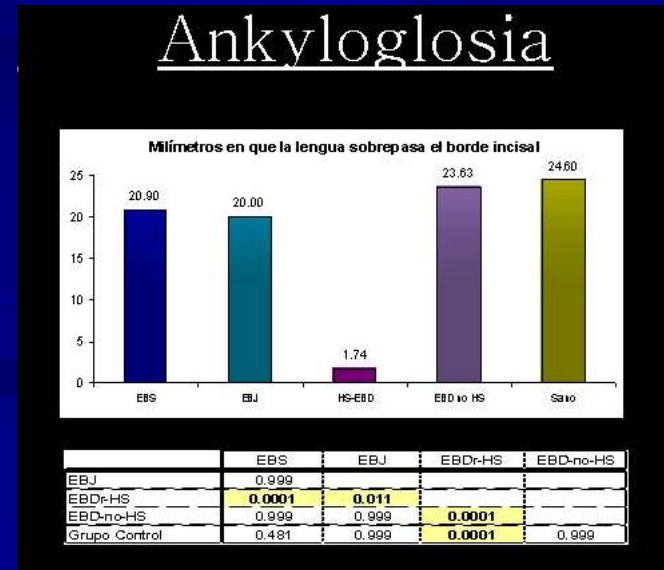
Oral Complications

- Changed oral anatomy
- Microstomia
- Ankyloglossia
- Diminished oral function
- Reduced saliva flow?
- Changed oral pathology



Changed Oral Anatomy

- Ankyloglossia
- Results in poor oral function in all movements
- Poor oral clearance



Changed Oral Anatomy

■ Loss of Vestibular Depth



Salivary Flow Reduced??

- Studies have shown that salivary flow is not reduced
- But with scarring of salivary duct it may be compromised?



Enamel Defects

- Amelogenesis Imperfecta
- Treatment options
 - Resin veneers in the young
 - Porcelain veneers in older individuals



Enamel Defects



Management of Microstomia

- Non Surgical Intervention
- Surgical Intervention



Management of Microstomia

Non-Surgical Management

□ Use of orthodontic appliances

Bite opening appliances



Image 6. Resin plug to improve mouth opening

Other suggestions include daily mouth opening stretching (See appendix 7.4), exercises with wooden spatulas,²⁶ or with devices such as a mouth trainer and threaded acrylic cone.



Image 7. Mouth opening exercises. Image 8. Mouth trainer



Image 9. Threaded acrylic cones.

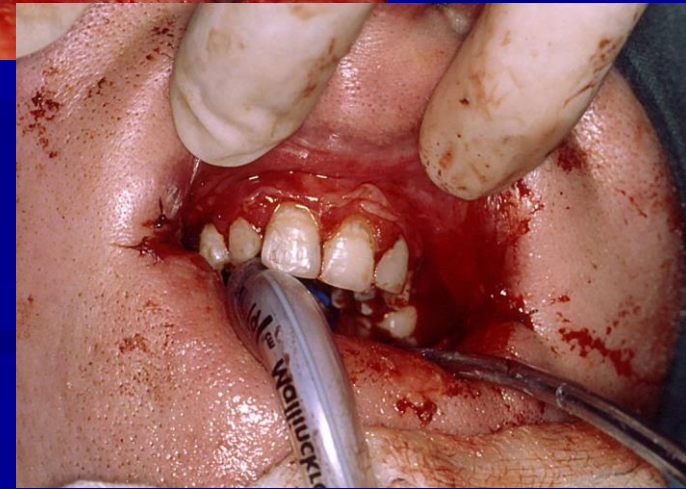
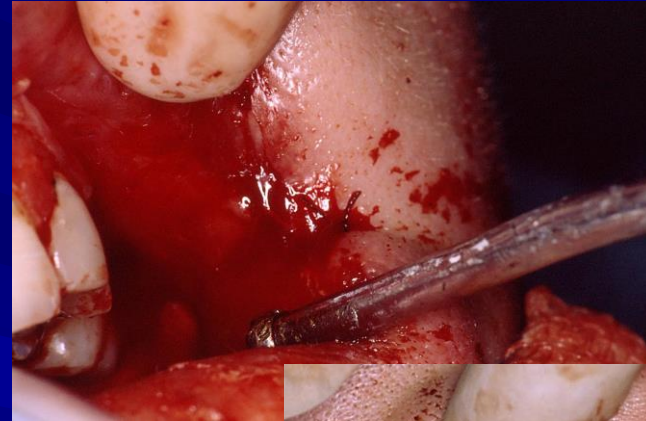
Management of Microstomia

Non Surgical Management



Management of Microstomia

■ Release of Strictures



Management of Microstomia

- Release of Strictures
- Use of Diode Laser



Management of Microstomia

- Release of Strictures
- Use of Diode Laser



Quality of Life

Today

Research

EBS:

- They were "Sometimes" concerned about problems with teeth and gums.
- "Seldom" they had troubles biting or chewing.

JEB:

- They were "always" concerned about problems with their teeth and gums.
- "Sometimes" they had troubles biting or chewing.

Severe generalized RDEB :

- "Often" they had troubles biting or chewing.
- "Seldom" they were able to swallow comfortably
- "Seldom" they were able to eat without feeling discomfort.

Other DEB subtypes:

- "Often" they had troubles biting or chewing.
- "Sometimes" they were concerned about problems with teeth and gums.





Oral Health Care for Patients with Epidermolysis Bullosa

Best Clinical
Practice Guidelines
October 2011



Best Clinical Practice Guidelines

■ The SIGN Guidelines were used

LEVELS OF EVIDENCE	
1++	High quality meta-analyses, systematic reviews of RCTs, or RCTs with a very low risk of bias
1+	Well conducted meta-analyses, systematic reviews, or RCTs with a low risk of bias
1-	Meta-analyses, systematic reviews, or RCTs with a high risk of bias
2++	High quality systematic reviews of case control or cohort studies High quality case control or cohort studies with a very low risk of confounding or bias and a high probability that the relationship is causal
2+	Well conducted case control or cohort studies with a low risk of confounding or bias and a moderate probability that the relationship is causal
2-	Case control or cohort studies with a high risk of confounding or bias and a significant risk that the relationship is not causal
3	Non-analytic studies, eg case reports, case series
4	Expert opinion

GRADES OF RECOMMENDATION	
<i>Note: The grade of recommendation relates to the strength of the evidence on which the recommendation is based. It does not reflect the clinical importance of the recommendation</i>	
A	At least one meta-analysis, systematic review, or RCT rated as 1++, and directly applicable to the target population; or A body of evidence consisting principally of studies rated as 1+, directly applicable to the target population, and demonstrating overall consistency of results
B	B A body of evidence including studies rated as 2++, directly applicable to the target population, and demonstrating overall consistency of results; or Extrapolated evidence from studies rated as 1++ or 1+
C	C A body of evidence including studies rated as 2+, directly applicable to the target population and demonstrating overall consistency of results; or Extrapolated evidence from studies rated as 2++
D	Evidence level 3 or 4; or Extrapolated evidence from studies rated as 2+

GOOD PRACTICE POINTS	
<input checked="" type="checkbox"/>	Recommended best practice based on the clinical experience of the guideline development group.

50 Guideline Developer's Handbook, NHS Scottish Intercollegiate Guidelines Network SIGN, Revised Edition January 2008

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Best Clinical Practice Guidelines

- Oral Care for Patients with Inherited Epidermolysis Bullosa
- Dental Treatment
- Anaesthetic Guidelines
- Series of Appendices



Oral Care for patients with inherited Epidermolysis Bullosa

- Early Referral
 - Education of parents
 - Early diagnosis of enamel abnormalities
 - Early diagnosis of dental caries
 - Early diagnosis of tooth crowding
- Oral Bullae and Ulceration
- Preventative Strategies
 - Oral hygiene: At home and professionally
 - Adjuvant therapies
 - Dietary modification
- Microstomia



Dental Treatment in EB

- Preventive, cleaning ,application of medicaments, early examination
- Restoration, fillings, crowns
- Orthodontic care, plates and braces if possible
- Periodontal therapy
- Oral surgery, tooth extraction including removal of third molars and vestibuloplasty



Oral Care and Dental Management for Patients with Epidermolysis Bullosa
Kramer S Dermatological Clinics 2010 Vol 28(2) 303-309

Preventive Strategies

- Must be age specific
- For young children then the emphasis is on reducing dental decay so that the child develops a pain free healthy aesthetic dentition
- For the older patient is to reduce the effect of tooth decay and also gum disease and ongoing effects of condition

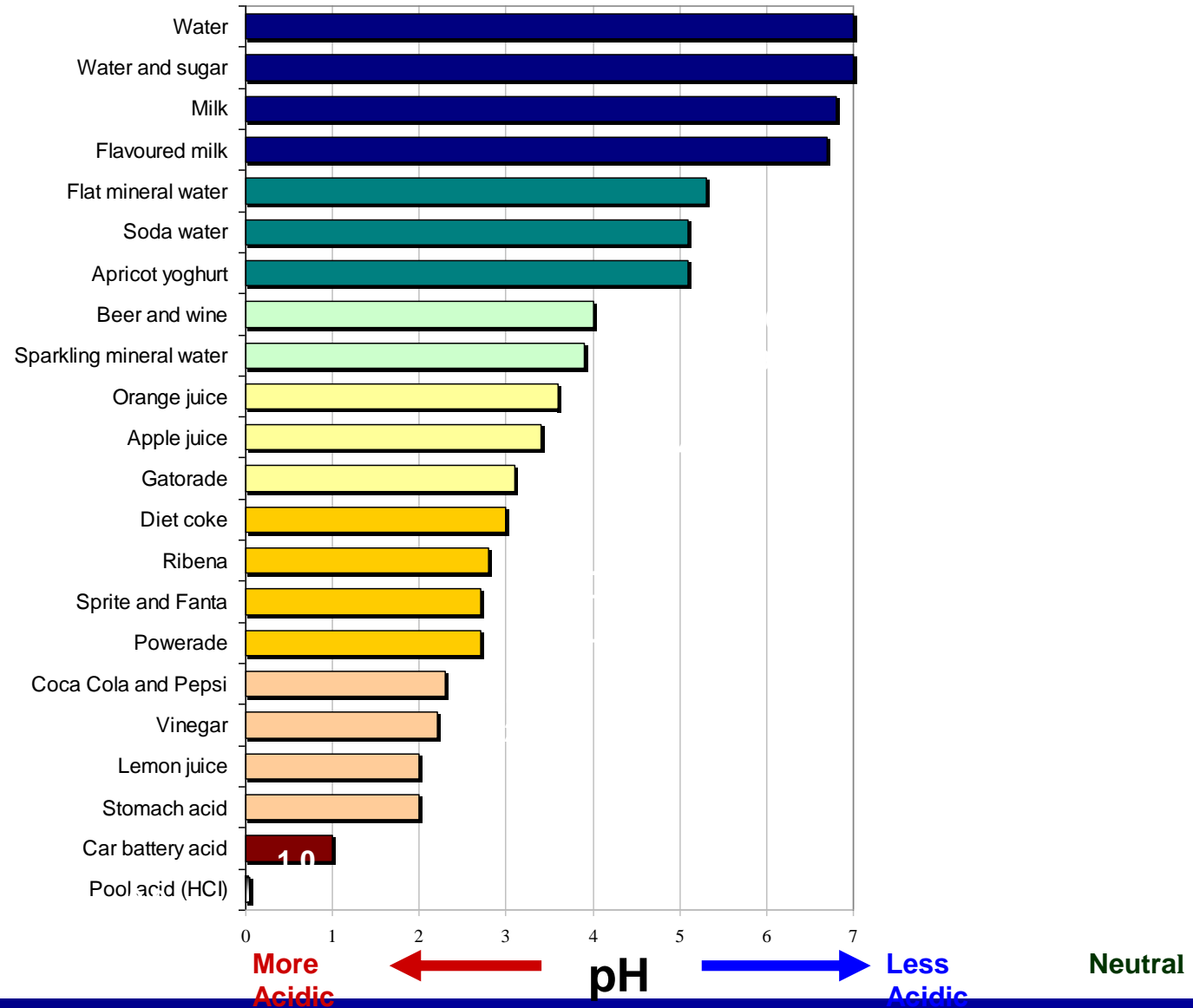
Preventive Strategies

- Reduction in the consumption of food containing refined carbohydrate
- Use of fluoride containing toothpaste
- Use of remineralizing agents
- Use of appropriate whether manual or electric toothbrush
- Regular dental reviews




Acidity (pH) of Common Drinks

The smaller the pH, then the stronger the acid.



Treatment Planning

- Evaluation of oral health; Which must include the gums ,teeth ,soft tissues
- Evaluation of treatment needs: number of fillings, extractions etc
- Extent of treatment required: complex or simple
- Understand the relationship of the medical condition to the treatment plan



3.8.5 Surgical Extractions

Contemporary oral health care is targeted at prevention of oral disease, but some patients still require extractions due to severe canes or the need for orthodontic care that involves severe dental crowding. Surgical/difficult extractions should be performed by an oral surgeon.

D When planning surgical extractions, especially if multiple extractions are needed, it is advisable to consult the patient's physician as profound anaemia could complicate the dental surgery.³⁸

D For multiple extractions it has been suggested to extract first the anterior teeth (i.e. from premolar to premolar) and then the molars to allow optimal access.³⁹

D An atraumatic technique should be used, making firm and safe mucosal incisions to prevent bullae formation.^{19,23}

D Haemostasis can be achieved with gentle pressure using gauze packs.^{9,41} These should be wet to avoid tissue adhesion.

Some authors have reported the extraction of healthy third or even second permanent molars in patients with severe generalized RDEB to improve or facilitate oral hygiene.^{2,40} There is controversy among different authors about this intervention. Severe tooth crowding,^{12,22,40} reduced alveolar arches secondary to growth retardation⁴⁰ and severe microstomia^{1,7,22,23,24,40,41,42} are described in patients with severe generalized RDEB, which would justify preventive extractions. However, nowadays most patients receive dietetic advice which optimises nutrition and growth. They receive orthodontic treatment (serial extractions) and are advised on exercises to improve microstomia. Therefore, preventive extractions of permanent molars need to be assessed very carefully on an individual basis.

Perioperative complications

Despite attempts to use as gentle manipulation as possible and all the special precautions, mucosal sloughing and blister formation has been reported after almost every surgical extraction in patients with severe RDEB.^{1,3,22,29,41} Blisters can arise at the angles of the mouth, lips, vestibule, tongue and any sites of manipulation, some measuring up to 4cm by 3cm.^{1,20} In some instances they might only be noticed by the patient or carer only on the second postoperative day.⁹



Image 16. Bullae, ulcers and mucosal sloughing after surgical extractions.

Postoperative complications

Despite the potential for extensive mucosal damage during surgery, postoperative complications are rare.^{3,20,23} Healing of the oral tissues occurs gradually after one to 2 weeks.^{1,6,21,41} Healing of the alveolar sockets seems to be uneventful.^{4,9} Nevertheless, there is a suggestion that scarring of the oral commissure can be accentuated after surgery.^{1,9} The use of postoperative antibiotics will depend on each individual case.

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Individuals with EB

Are RARE

Are BRAVE

And Deserve Optimum Dental Care

Development of the Guideline

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