

the Child Dental Patient with

Dystrophic Epidermolysis Bullosa: Clinical cases

François Clauss, Marie-Cécile Manière, Arina Vinereanu

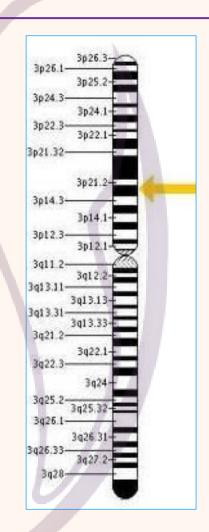






Clinical case 1

- Dina, 5 years-old
- Form of recessive dystrophic epidermolysis bullosa (DEB)
 - COL7A1 gene mutation (locus on chromosome 3)
- Early childhood caries



General clinical features

- Dermatological manifestations
 - Erosions and skin bubbles
 - Chronic infections
 - Retraction and acquired syndactyly
 - Plantar thickening
 - Risks of evolution towards carcinoma
- Ophtalmological manifestations
 - Corneal ulcerations
- Digestive manifestations
 - Oesophageal stenosis
 - Feeding difficulties (gastrostomy)
- Metabolic aspects
 - Impaired electrolyte and liquid balance
 - Growth delay
 - Hypermetabolism
 - Anemia

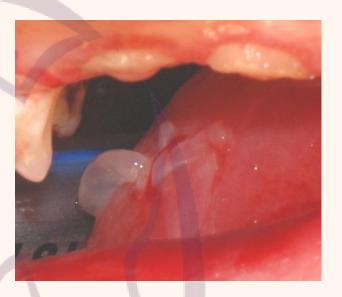




Schumann H., [Epidermolysis bullosa. An update]. Hautarzt, 2009. 60(8): p. 614-21

Oral clinical features

- Mucosal alterations
 - Erosions and bubbles
- Functional disorders
 - Retraction of oral mucosa
 - Ankyloglossia
 - Microstomia
- Extensive caries activity
 - Lesions on 12 primary teeth, with several pulpal necrosis linked to:
 - Hypercaloric and fractionated food
 - Toothbrushing difficulties
 - Reduced salivary secretion



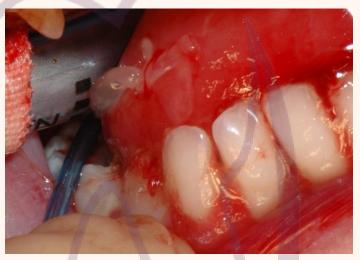
Tongue bubbles



Microstomia

Clinical and radiographic examination







Indication for a global rehabilitation under general anesthesia

- Early childhood caries
- Lack of cooperation
- Global rehabilitation in one session
- Access difficulties due to microstomia

Anesthesia management

- Anesthesic risks and difficulties associated to DEB
 - Risks of tracheal mucosa detachment
 - Microstomia
 - Metabolic alterations
 - Infectious risk
- Essential aspect in the anesthesia management
 - Reduction of the pressure points with the adhesive plasters

Pre-operative steps

- Preparation of the patient
 - Antibiotherapy
 - Corticotherapy
 - Topical anesthesia: EMLA®
 - Transfusion in case of anemia
- Preparation of the operating room
 - Protection mattress

Goldschneider, K., et al., Perioperative care of patients with epidermolysis bullosa: proceedings of the 5th international symposium on epidermolysis bullosa, Santiago Chile, 2008. Paediatr Anaesth. 20(9): p. 797-804

Monitoring

- Pulse oxymeter
 - Ear or digital captor
 - Plaster / Clip
- ECG
 - Classical patches
 - Lubrification agent : Surgilube®
 - Hydrocortisone application
 - Modified Patch
- Tensiometer





Recommandations aux anesthésistes de l'hôpital de Saint-Louis http://www.dar-saint-louis.fr/documents/epidermolyses.pdf

GA Induction

- Intra-venous induction
 - Propofol
 - Difficulties to find venous access
 - Stabilisation
 - Suture
 - Auto-adhésive elastic plaster (Coban®)
- Inhalation induction
 - Isoflurane, Sevoflurane
 - Reduction of the mask pressure





Tracheal Intubation

- Difficulties
 - Microstomia
 - Ankyloglossia
- Type of intubation
 - Naso-tracheal
 - Oro-tracheal is preferred
 - Intubation using fibroscopy useful
 - Lubrification of the intubation tube
- Stabilization of the intubation tube



Eyes protection

- Partial occlusion
- Vitamin A ophtalmological cream
- No adhesive plaster



Oro-dental management

- Principles
 - Lubrification agent on all the instruments
 - Reduction of iatrogenic trauma
- Disinfection of the oral cavity Prophylaxis
- Conservative treatments
 - Composite restorations: 53, 63, 74
- Prevention
 - Sealants: 75, 85
 - Topical fluoride application
- Surgery
 - Extraction of 55, 54, 52, 51, 61, 62, 64, 65, 84
 - Local hemostasis (Surgicel®)



Post-operative follow-up

- Immediate follow-up
 - Hospitalization during 48h
 - Antibiotherapy and analgesia
- 1 month follow-up
 - Topical application of fluoride varnish
 - Oral hygiene recommandations: use of a soft toothbrush + fluoride toothpaste
- Every 3 months
 - Prevention and control



Post-operative aspect of the lower lip



Clinical case 2

- M, 9 years old
- Recessive dystrophic epidermolysis bullosa (DEB)
- Referred by dermatologist for complex dental treatment



Clinical findings

General

Oral

- Dermatological manifestations
 - Erosions and skin bubbles
 - Chronic infections
 - Retraction and acquired syndactyly
- Digestive manifestations
 - Oesophageal stenosis
 - Feeding difficulties



Oral features

- Functional disorders
 - Ankyloglossia
 - Microstomia
- Extensive caries, linked to:
 - Hypercaloric semi-fluid food
 - Difficulties in performing oral hygiene routine
 - Low salivary flow
- Severe malocclusion



1st visit:

- 31, 41 glass-ionomer fillings (GC Fuji IX) after manual cavity preparation
- Difficulties encountered:
 - patient'scompliance: Frankl2→3
 - local anaesthesia not accepted
 - Severity of carious lesions
 - limited mouth opening



→ Need for GA

Panoramic X-ray needed!

Further treatment planning

- restorations on PT
- endodontics PT?
- multiple extractions of DT (DP?)
- regular check-ups
- secondary prevention

Difficulties encountered: Parents refused treatment under GA

Clinical case 3

D, age 1y 4m, Recessive DEB

- referred by dermatologist for preventive oral care

Clinical findings: - lesions on skin and oral mucosa

- signs of ECC

Anamnesis: semi-fluid food; night feeding; no brushing/cleaning





• 1st visit:

Parents were informed about the role and methods of efficient home oral care and feeding habits

CPP-ACP (casein phosphopeptide – amorphous calcium phosphate) cream (GC Tooth Mousse) was recommended

Need for regular check-ups even in the absence of symptoms, need for professional cleaning and topical fluoride application were explained

Need for getting used to the dental office setting and staff from young age were pointed out

Planning for next visits

• Difficulties encountered:

Young child, no cooperation, difficult examination No-show for next visits despite call from dental office

Dental treatment GOAL in EB

PALIATIVE

efficient PREVENTION



Oral care guidelines for parents of EB children

- First dental visit : age 3-6 m
- Soft non-cariogenic diet nutritionist
- Home Oral Hygiene:

Choose small, soft brush & assist brushing - minimize tissue damage; brush can be further softened by dipping it in warm water; bristles can be shortened if mouth opening is limited and does not permit reaching distal areas of the arches

Chlorhexidine 0.12% mouthwash

Fluoride toothpaste (>1000-1450 ppm even for young kids, appropriate amount – "grain" for <3y, "pea" for >3y

Please visit Parents'section of http://oscarpd.eu/ for video

- Daily mouth opening exercises
- Regular dental check-ups even in absence of symptoms