



#### **EDS ECHO SUMMIT SERIES**

# Orodental manifestations of EDS

#### SPEAKER

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#### Enamel

96% anorganic hydroxyapatite

#### Dentine

mineral platelets embedded in a collagen fibre mesh (mainly I, also III and IV)

#### Gingiva

firmly attached to the periostum of the bone by collagenous fibers (I, III, IV, VII; V may be present in inflamed tissue)

#### **Pulp** soft connective tissue (collagen I and III)

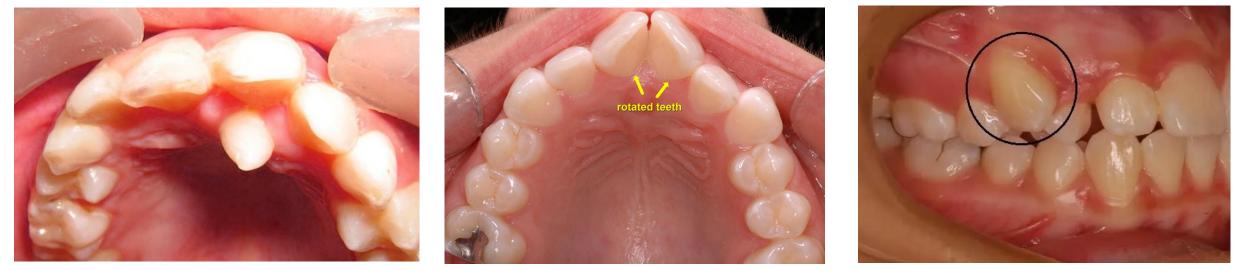
#### **Periodontal ligament**

collagen fiber bundles embedded in ground substance (I, III, XII) that span the distance between the bone and the tooth

#### Cementum

mineralized avascular hard connective tissue 50% collagen I

# Are these manifestations of EDS?



Supernumerary teeth

Rotated teeth

Tooth transposition

#### Single cases with hypermobile EDS and classical EDS

For references and sources see: Kapferer-Seebacher et al. Dental Manifestations of Ehlers-Danlos Syndromes: A Systematic Review. Acta Derm Venereol. 2020 Mar 25;100(7):adv00092.

# Is caries a manifestation of EDS?





# Dental manifestations with EDS

**Pulp calcifications** 

Hypoplastic roots

Classical EDS Periodontal EDS Hypermobile EDS

Classical EDS

Periodontal EDS

Hypoplastic roots

Vascular EDS

**Exceeding roots** 

Pope FM et al. J Oral Pathol Med 1992; 21; 418-21.; Hakki et al. J Dent Oral Biol. 2017; 2(15): 1090.; De Coster et al. Arch Oral Biol 2007; 52(2): 101-9.; De Coster et al. J Oral Pathol Med 2005; 34: 298–307; Lepperdinger et al. (under proposal)

# Dermatosparaxis EDS

<u>Malfait et al. 2004</u>: n = 3 (5 - 12 Jahre)

- Severe gingival hyperplasia in both jaws
- Hypermobility of the temporomandibular joint
- hypodontics, microdontics, opalescent teeth



Fragility of the skin, typical craniofacial features, lax skin, foldingof the palms, growth retardation

Malfait et al. The natural history, including orofacial features of three patients with Ehlers-Danlos syndrome, dermatosparaxis type (EDS type VIIC) AJMG 2004;131A:18–28

# Resistance to local anesthesia in people with EDS presenting for dental surgery

Schubart, Schaefer, Janicki, Adhkary, Schilling, Hakim, Bascom, Francomano, Raj. J Dent Anesth Pain Med 2019;19(5):261-70

Method

<u>Questionnaire:</u> 980 EDS + 249 non EDS respondents

"Have you ever had a problem with a local anesthetic injection not working adequately or properly?"

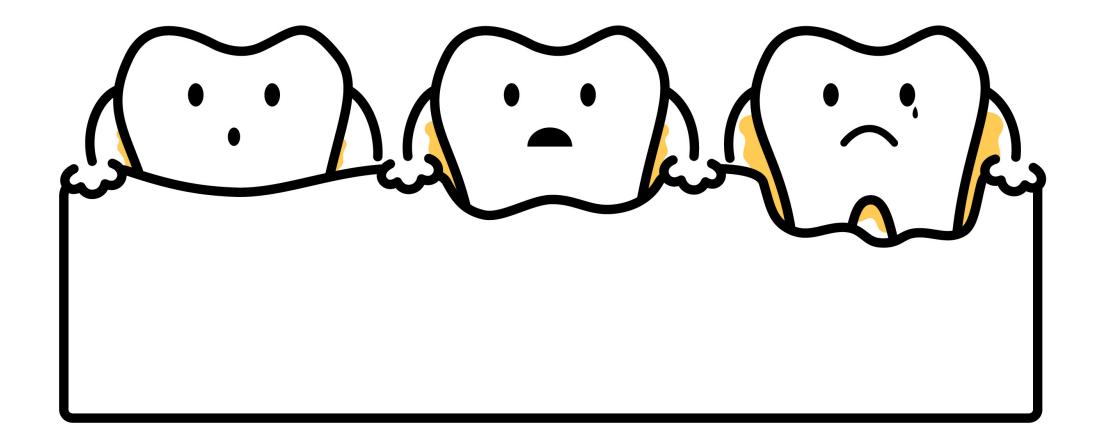
#### Results

**EDS:** 88% recalled inadequate pain prevention **non EDS**: 33% recalled inadequate pain prevention

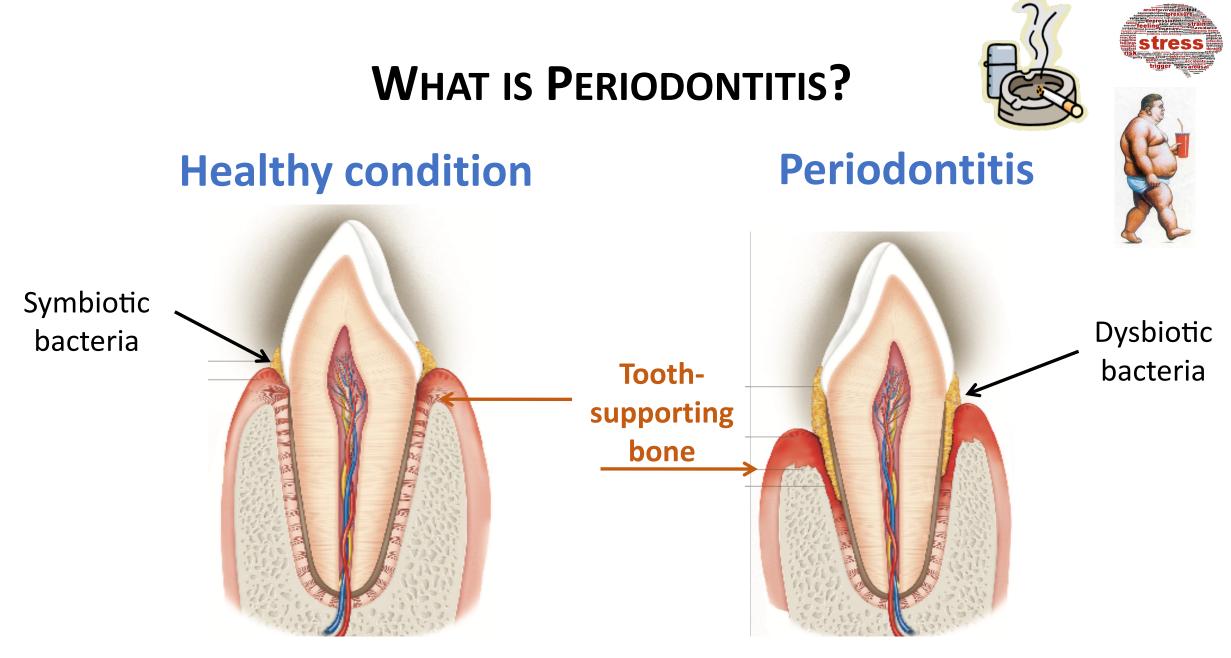
The agent with the highest success rate was articaine (30%), followed by bupivacaine (25%), and mepivacaine (22%).

# Oral health related quality of life





Periodontitis

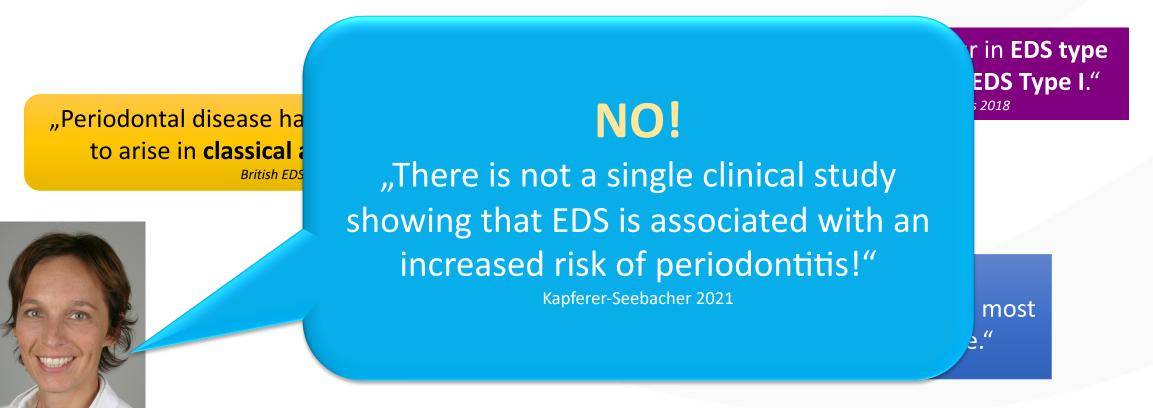


#### Mild immune reaction

**Inflammatory bone loss** 



# Is EDS associated with an increased risk of periodontitis?



Kapferer-Seebacher I, Lundberg P, Malfait F, Zschocke J. Periodontal manifestations of Ehlers-Danlos syndromes: A systematic review. J Clin Periodontol. 2017;44(11):1088-1100.



# Vascular Ehlers-Danlos Syndrome (n = 23)

Ferré et al. 2012:n = 17 vEDSn = 46 age-matched controls

Periodontitis: 23.5% in vEDS

45% in controls

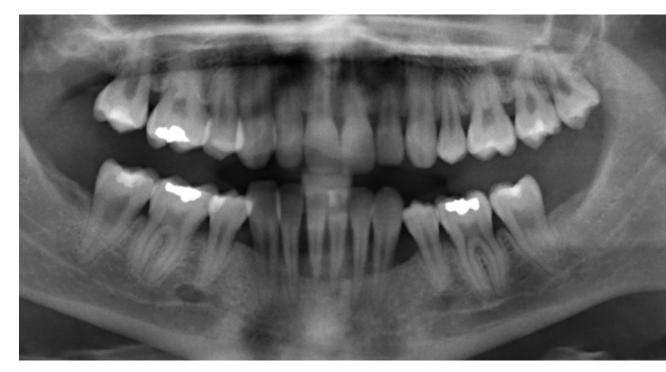
**De Coster et al. 2005:** n = 31; 6 of which with vEDS

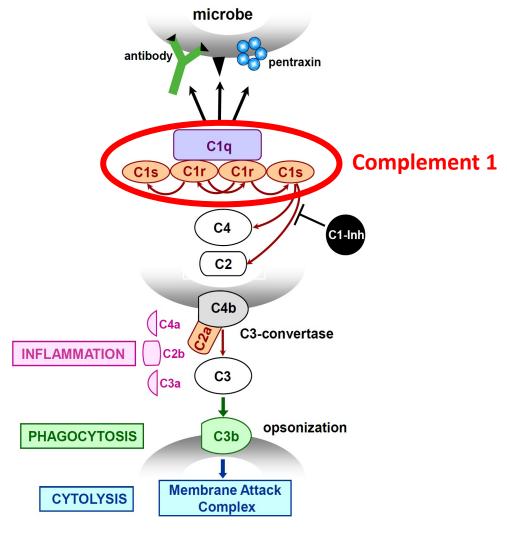
#### $\rightarrow$ No individuals with periodontitis

*Ferré FC et al. Oral phenotype and scoring of vascular Ehlers-Danlos syndrome: a case-control study. BMJ Open 2012;2:e000705. De Coster et al. Oral health in prevalent types of Ehlers-Danlos sydnrome. J Oral Pathol Med 2005; 34: 298–307.* 

# Periodontal EDS (formerly EDS VIII)

specific EDS subtype caused by mutations in C1R and C1S





Early severe periodontitis Age of diagnosis (mean): 14 years Complete tooth loss (mean): 20 years without periodontal treatment

Kapferer-Seebacher et al. Am J Hum Genet. 2016;3;99(5):1005-1014..

# Periodontal EDS (formerly EDS VIII)

### specific EDS subtype caused by mutations in C1R and C1S



Early severe periodontitis Age of diagnosis (mean): 12 years Complete tooth loss (mean): 20 years without periodontal treatment **Pretibial discolorations** 79% of affected Specific gum phenotype

Kapferer-Seebacher et al. Am J Hum Genet. 2016;3;99(5):1005-1014..



# **Healthy siblings**

# **Children with periodontal EDS**

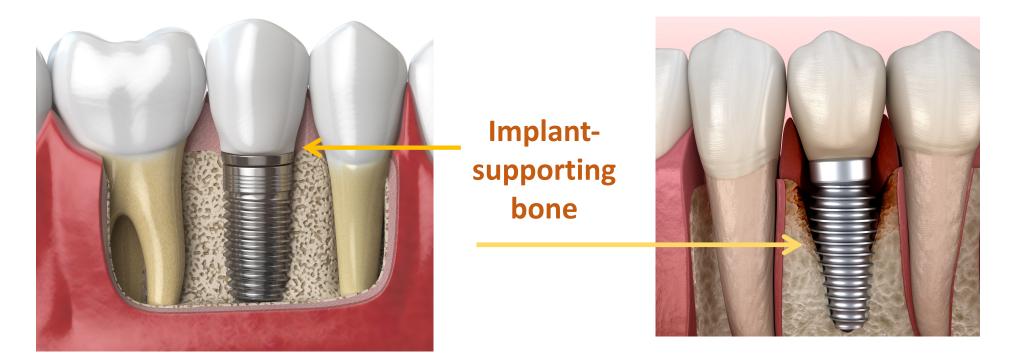


Kapferer-Seebacher et al. Genet Med. 2021 Feb;23(2):316-322.

# WHAT IS PERI-IMPLANTITIS?

# **Healthy condition**

# **Peri-Implantitis**

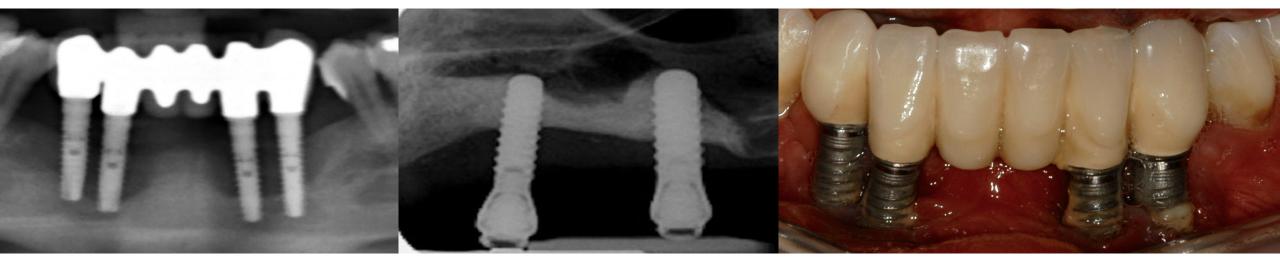


#### Mild immune reaction

**Inflammatory bone loss** 

# High risk of peri-implant disease in periodontal EDS. A case series.

Rinner, Zschocke, Schossig, Gröbner, Strobl, Kapferer-Seebacher. Clin Oral Implants Res. 2018;29(11):1101-1106.



Three individuals included

**There seems to be NO increased risk for implant loss with other EDS types** one study on five individuals with hypermobile or classical EDS *Jensen et al. 2012* 



# Conclusions

# All types of EDS

Resistance to local anesthetics

### **Periodontal EDS**

Early severe periodontitis Thin and fragile gums Pulp stones Increased risk of peri-implantitis

### **Classical EDS**

Pulp stones Shortened (hypoplastic) roots Vascular EDS

Exceeding root length

Striking lack of scientific evidence!



#### **Clinical studies on oral manifestations of various EDS types**

