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Microscopic structure of dental hard tissues in primary and permanent teeth from individuals with Prader-Willi syndrome

Ronnaug Saeves[a,*], Randi Furseth Klinge[b], Steinar Risnes[b]

[a] TAKO-centre, Lovisenberg Diaconal Hospital, Pb 4970 Nydalen, 0440 Oslo, Norway
[b] Department of Oral Biology, University of Oslo, Oslo, Norway

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ABSTRACT

Objective: severe tooth wear, in terms of both erosive wear and attrition, is a significant problem in individuals with Prader-Willi syndrome (PWS). The purpose of the present study was to describe the structure of enamel and dentine in primary and permanent teeth from individuals with PWS.
Design: thirty-two primary and 10 permanent teeth representing 16 individuals with PWS were investigated in the study. The enamel surface was studied using scanning electron microscopy (SEM). The microscopic structure of enamel and dentine was studied using SEM, microradiography and light microscopy.
Results: the microscopic structure of enamel and dentine was found to be normal with the exception of a slight increase of interglobular dentine (IGD). Severe erosive defects were observed in primary teeth and also in permanent teeth with long exposure to the oral environment.
Conclusion: the erosive enamel defects in individuals with PWS seem more related to the factors in the oral environment than to enamel structure which appeared normal. The occurrence of IGD indicate deficient mineralization but is probably of minor clinical significance. Gastro-oesophageal reflux is worthy of further investigation in individuals with Prader-Willi syndrome.

1. Introduction

Prader-Willi syndrome (PWS) is the most common genetic human obesity syndrome, resulting from failed expression of paternally inherited genes on chromosome 15q11-13.

There are three main mechanisms resulting in PWS. The majority of individuals with PWS (70%) have a paternally derived deletion of 15q11-13, whilst maternal disomy 15 (UPD) occurs in 25% and the remaining 2–5% have imprinting defects (Cassidy et al., 1997Driscoll et al., 1992). The typical PWS deletion consists of two classes, type 1 and type 2 depending on the size and chromosome breakpoint position. When the genotype-phenotype relationships become clearer, it will be clinically important to subtype the deletion classes (Kim et al., 2011). Clinical diagnostic criteria have been developed, but as clinically overlapping disorders exist, the diagnosis must be confirmed by genetic testing (Cassidy and Driscoll, 2009). Due to the complexity of the genetics of PWS more than one test is usually necessary to confirm the diagnosis PWS and differentiate between the various forms. Epidemiological surveys estimate the population prevalence to be up to 1:52000 (Smith et al., 2003; Vogels et al., 2004; Whittington et al., 2001) and the gender ratio close to 1/1 (Akefeldt, Gillberg, & Larsson, 1991; Whittington et al., 2001).

The syndrome has a characteristic phenotype including severe neonatal hypotonia, early feeding problems, childhood onset hyperphagia, obesity, short stature associated with growth hormone (GH) deficiency, high pain threshold and intellectual disability (Gunay-Aygun, Schwartz, Heeger, O'Riordan, & Cassidy, 2001; Holm et al., 1993; Prader, Labhart, & Willi, 1956). The syndrome is recognized as the most common syndromal cause of obesity in children. Obesity can be controlled by diet restrictions. Growth hormone treatment improves growth, physical phenotype and body composition (Cassidy and Driscoll, 2009). Necdin is important for the differentiation of central and peripheral sensory neurons and is congenitally absent in PWS (Jay et al., 1997). A narrow forehead, almond-shaped eyes, down-turned corners of the mouth and a thin upper lip are characteristic facial features.

* Corresponding author.
E-mail addresses: ronnaug.saeves@tako.no, ingunris@gmail.com (R. Saeves), r.f.klinge@odont.uio.no (R.F. Klinge), steinar.risnes@odont.uio.no (S. Risnes).

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Varying degrees of oral motor dysfunction is common in individuals with PWS (Saeves, Asten, Storhaug, & Bagesund, 2011).

Decreased salivary flow rate (Bray et al., 1983; Hart, 1998; Saeves, Nordgarden, et al., 2012) and increased amounts of salivary ions and proteins have been reported in individuals with the syndrome (Hart, 1998; Saeves, Reseland et al., 2012). Unstimulated salivary flow in individuals with PWS has been identified as 30–38% of that in healthy controls (Hart, 1998; Saeves, Nordgarden, et al., 2012).

Dental caries, enamel defects and poor oral hygiene have been described in several case reports (Banks, Bradley, & Smith, 1996; Bazopoulou-Kyrkanidou and Papagiannoulis, 1992; Bots, Schueler, Brand, & van Nieuw, 2004; Scardina, Fuca, & Messina, 2007). Two recent surveys, however, reported more favourable oral health (Bailleul-Forestier et al., 2008; Saeves, Nordgarden, et al., 2012). Severe tooth wear, both erosive wear and attrition has also been reported in PWS (Bailleul-Forestier et al., 2008; Saeves, Espelid et al., 2012; Young et al., 2001).

To our knowledge no studies of dental hard tissues in teeth from individuals with PWS are currently available. The purpose of the present study was to describe the structure of enamel and dentine in primary and permanent teeth from individuals with PWS.

2. Material and methods

2.1. Tooth sample

The oral manifestations of 50 individuals with PWS (5–41 years) have previously been described in detail (Saeves et al., 2011; Saeves, Nordgarden, et al., 2012; Saeves, Reseland et al., 2012; Saeves, Espelid et al., 2012). For the present study, all participants were invited to send exfoliated primary teeth and extracted primary and permanent teeth. A total of 94 teeth were collected from 16 individuals (84 primary, 80 exfoliated and 4 extracted; 10 permanent, 9 extracted due to orthodontic treatment and 1 due to caries) (Fig. 1). Of these, 32 primary and 10 permanent teeth, representing all individuals were used in the present study. The majority of the teeth had been stored dry. Four extracted teeth, however, were fixed immediately in 5% formaldehyde.

The study protocol was approved by the Regional Committee for Medical Research Ethics and informed consent was obtained from all participants.

2.2. Scanning electron microscopy (SEM)

The crowns of 16 teeth (11 primary, 5 permanent) were cleaned by brushing under running tap water. The crowns were cut off from the root by a rotating diamond disc. After air drying for 5 days the crowns were mounted on aluminum stubs with cyanoacrylate glue, with the cut surface against the stub.

Eleven teeth (8 primary, 3 permanent) were embedded in Epoxy Embedding Medium (Sigma-Aldrich/Fluka). After cutting off the roots, segments of the crowns were cut longitudinally in facio-lingual direction. The median face of each segment was further grinded with water on 1200 grit silicon carbide paper (3 M® Company) and polished with 0.05 μm particle size Micro polish alumina powder (Buehler) in water against the back side of the grinding paper. After rinsing under running tap water, the specimens were dried with paper and etched for 45 s in 1% nitric acid (HNO_3) under constant stirring. After air drying for 5 days, the crown segments were mounted on aluminum stubs with a cyanoacrylate glue, the unground/unpolished aspect against the stub.

The specimens were sputter-coated with 30 nm gold palladium and observed in a scanning electron microscope (Philips XL 30 ESEM) operated at 12–15 kV.

2.3. Microradiography: light microscopy

Sections parallel to the long axes of 10 teeth (8 primary, 2 permanent) were cut on a Gillings-Hamco Thin Sectioning Machine. The sections were reduced in thickness to about 130 μm by grinding on abrasive papers. Microradiographs were produced in a Philips X-ray diffraction unit, excited to 20 kV at 20 mA,

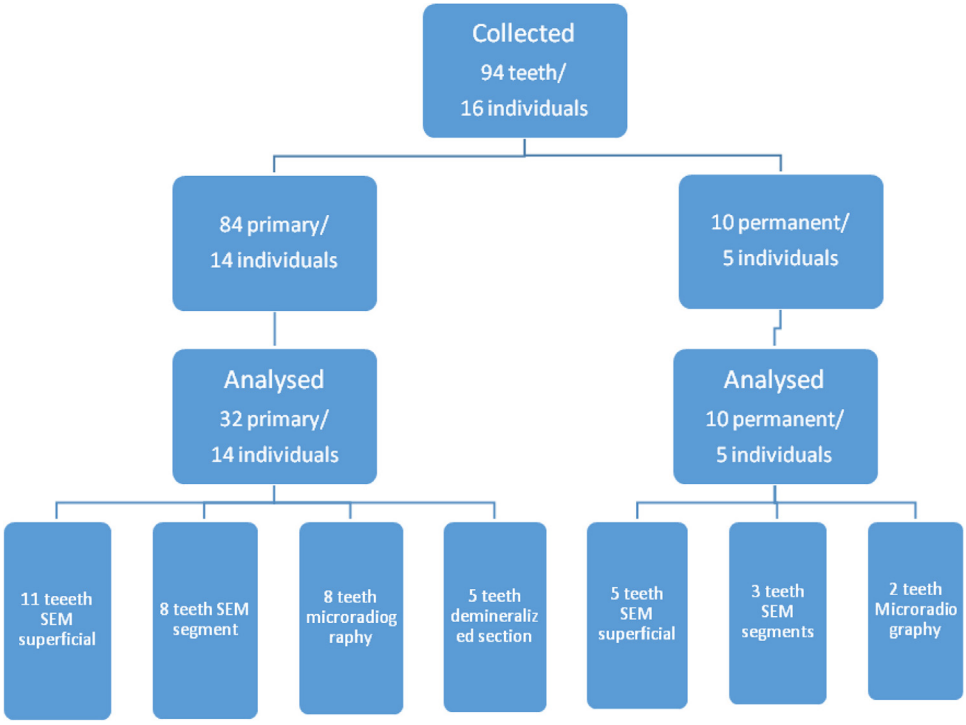

Fig. 1. Flow diagram showing number of teeth chosen for scanning electron microscopy (SEM), microradiography and light microscopy.