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Research Article

The Effect of Apert Syndrome on Histology and Mineralization of Enamel and Dentin in Deciduous and Permanent Teeth

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Abstract

Aims: To analyze the histology, mineralization, and ion contents in deciduous and permanent teeth of Apert syndrome (AS), in comparison to match-paired teeth from normal children.

Methods: Deciduous and permanent teeth of a child with Apert syndrome and their match-paired normal teeth were examined under a light microscope and using an energy dispersive X-ray spectrometer program under a scanning electron microscope, the ion component of enamel and dentine were compared.

Results: The morphology of the enamel and dentin of Apert syndrome teeth was similar to normal. Postnatal traumatic lines were observed in the enamel of deciduous and permanent teeth of Apert syndrome.

In AS teeth, the enamel and the dentin contained larger concentrations of calcium and phosphate than normal teeth. In addition, the ratio between calcium and phosphate in AS enamel and dentin was very high in comparison to normal teeth.

Conclusion: AS affects the mineralization of enamel and dentin. It caused abnormal mineral content in comparison to match paired normal teeth. The traumatic lines observed in both deciduous and permanent teeth implicate that severe traumatic episodes occurred during early childhood.

These findings show that AS also affects the mineralization of teeth in addition to the known oral motor challenges.

Keywords: *Apert Syndrome; Enamel; Dentin; Mineralization; Traumatic Lines*

Introduction

Apert syndrome, or acrocephalosyndactyly type 1, is a rare, congenital craniosynostosis condition, resulting from a missense mutation in the gene encoding for fibroblast growth factor receptor 2. It is characterized by three specific clinical features: brachycephalic skull, midface hypoplasia, and limbs abnormalities (syndactyly of hands and feet). The prevalence is 15.5/1,000,000 births. The disease accounts for about 4.5% of all cases of craniosynostosis [1,2].

Craniofacial deformities include cone-shaped calvarium, flat forehead, proptosis, hypertelorism, and short nose with a bulbous tip. Intraoral findings include high arched palate with pseudocleft (bilateral swelling in the palatine processes), maxillary transverse and sagittal hypoplasia with concomitant dental crowding, and skeletal and dental anterior open bite [3].

The oral cavity is characterized by teeth impaction, severe crowding, delayed eruption, thick gingiva, ectopic eruption, high caries prevalence, and multiple tooth agenesis or supernumerary teeth. In addition, class III malocclusion and posterior crossbite with slight midline deviation were observed [4].

Surman., *et al.* conducted a histological assessment of the dental hard tissues, that revealed an intact enamel and dentinal structure, but some irregularities were noted in the region of the dentino-enamel junction (DEJ), which could affect the caries progression and also make dental management more difficult [5].

In previous research, the histological and quantitative differences in enamel and dentin were found in different syndromes, including down syndrome, cerebral palsy, Angelman syndrome, and familial dysautonomia [6-8].

Materials and Methods

12 extracted teeth from a child with genetically confirmed Apert syndrome and 12 match-paired teeth from normal children were examined.

The teeth examined were: one deciduous maxillary canine, two first and two-second maxillary deciduous molars, two mandibular first and two-second deciduous molars, one mandibular permanent canine, and two mandibular first premolars. The teeth were extracted during regular dental treatment due to the delayed eruption and impaction of permanent teeth or after orthodontic evaluation.

The parents and children gave their verbal consent to donate the teeth to the clinic.

The teeth were invested in epoxy resin (Epofix Resin) and sliced along a buccolingual plane parallel to the long axis of the tooth using an Isomet 1000. The canines and premolars were sliced through the cusp tip and the deciduous molars were sliced through the mesial cusps. The sections were then examined using a light microscope (BestScope) with an Axiocam camera (Zeiss Microscopy GmbH) and a scanning electron microscope (SEM), (FEI, Quanta 200, Eugene, OR) under high vacuum mode, without coating. Figure 3 was taken using SEM in order to observe the DEJ. All other figures were taken using the light microscope. Using an energy dispersive X-ray spectrometer (EDS), ion analyses were carried out for enamel and dentine. The location of the ion analyses was identical in Apert and normal teeth- prenatal and postnatal enamel and dentin in deciduous teeth (Figure 1) and enamel and dentin in permanent teeth (Figure 2). The relative ion concentrations of calcium, phosphate, oxygen, carbon, and nitrogen were determined from a rectangle with a minimum of 8000 counts. The ratio of calcium to phosphate (Ca/P) was calculated for enamel and dentin in both AS and normal teeth.

Results

The DEJ morphology, as observed by SEM was similar in both Apert and normal teeth (Figure 3). In Apert syndrome, traumatic lines were observed in the postnatal enamel of deciduous teeth and permanent teeth (Figure 4-6). No traumatic lines were observed in the normal teeth (Figure 7 and 8). The traumatic lines differ from growth lines (Stria of Retzius) in thickness and in order to observe

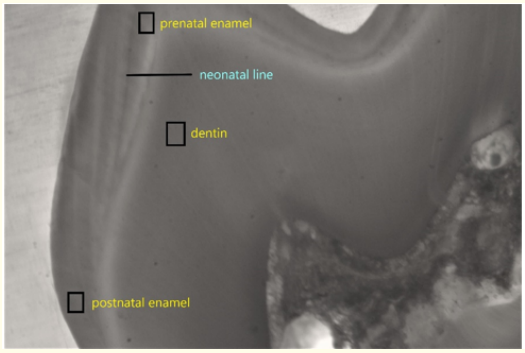


Figure 1: Location of ion concentration measurements in deciduous teeth.

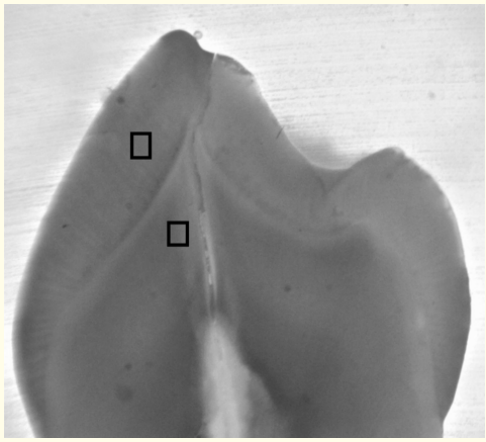


Figure 2: Location of ion concentration measurements in permanent teeth.

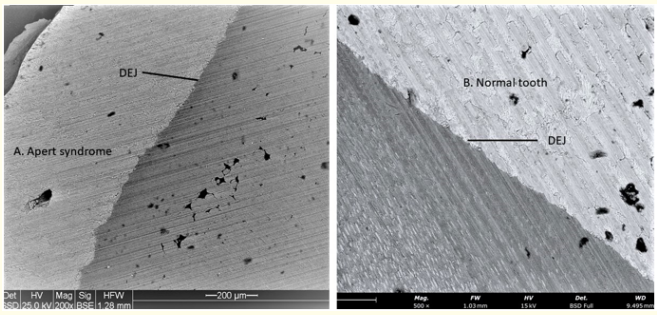


Figure 3: Dentino-enamel junction (DEJ) in Apert syndrome and normal teeth.

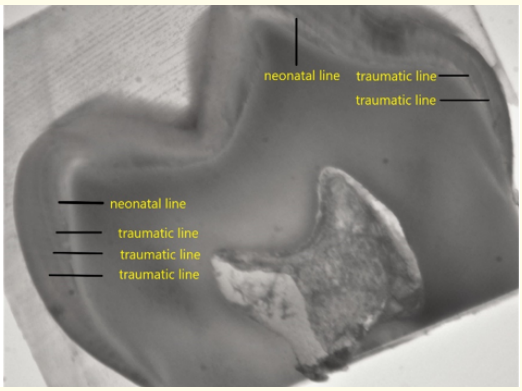


Figure 4: Traumatic lines in Apert syndrome second mandibular deciduous tooth.

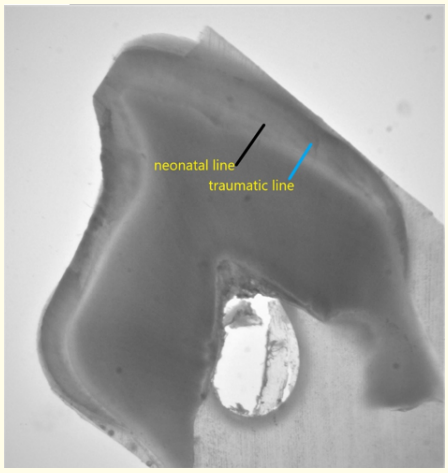


Figure 5: Traumatic line in Apert syndrome upper deciduous canine.

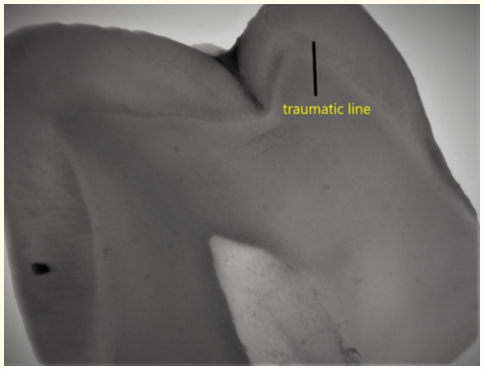


Figure 6: Traumatic line in Apert syndrome lower first pre-molar.

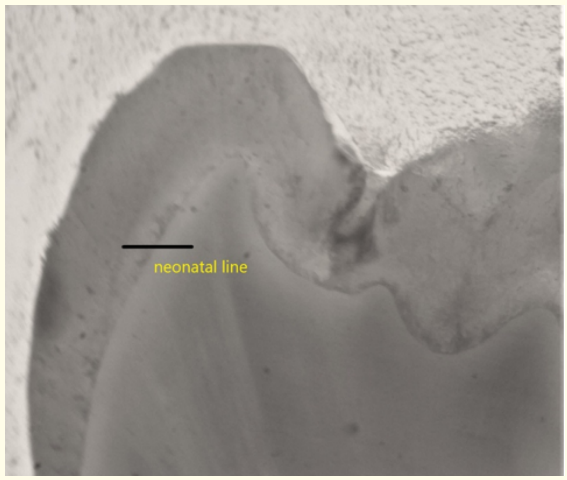


Figure 7: No traumatic line in normal lower second deciduous molar.

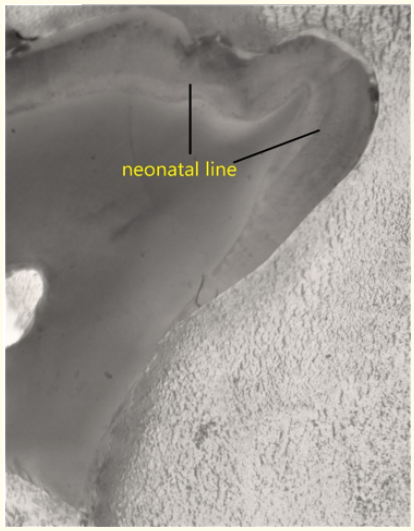


Figure 8: No traumatic line in normal lower first deciduous molar.

them SEM magnifications are required. The traumatic line can be observed under light microscope, similar to neonatal line. Table 1 shows the ion concentration in pre and postnatal enamel of deciduous teeth in AS and normal. In normal teeth there is a significant difference between pre and postnatal enamel: the concentration of calcium and phosphate in prenatal enamel are higher and the concentrations of oxygen and carbon are lower. In the AS teeth, the differences between pre and postnatal enamel are very small and the concentration of ions is similar. The main differences between

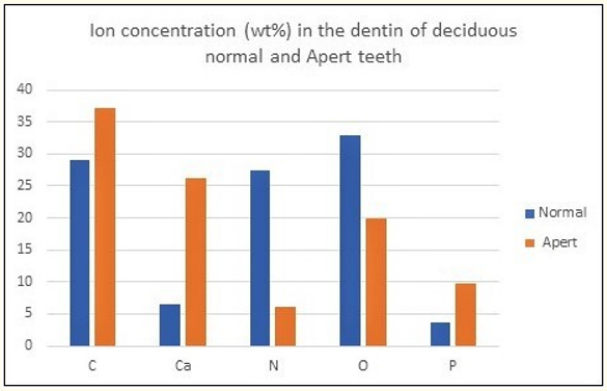
AS deciduous teeth and normal are in calcium concentrations in both pre and postnatal enamel and phosphate in postnatal enamel.

	Apert Syndrome		Normal	
	Post-Natal	Pre-Natal	Post-Natal	Pre-Natal
C	13.426	11.94	12.88	5.73
O	24.15	23.892	42.87	35.88
P	18.19	18.2	16.096	18.67
Ca	44.24	45.89	28.04	39.7225

Table 1: *Ion concentration in pre- and postnatal enamel in AS and normal deciduous teeth.*

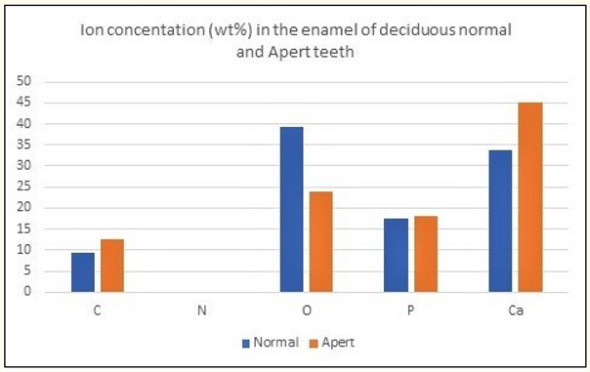
Note: C=Carbon, O=Oxygen, P=Phosphate, Ca=Calcium.

Graph 1-4 show the relative ion concentration in wt% in the enamel and dentin of Apert teeth in comparison to normal teeth.



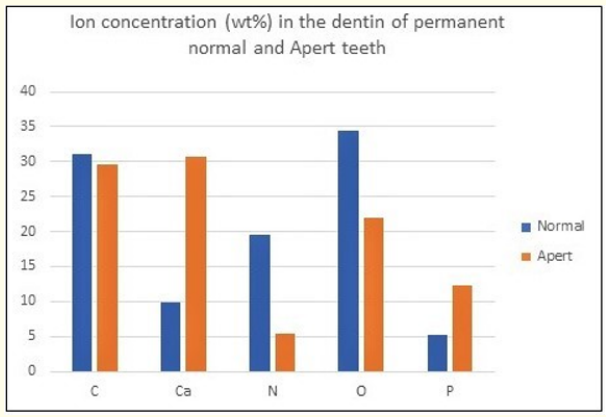
Graph 1: *Ion concentration (wt%) in the dentin of deciduous normal and Apert teeth.*

Note: C=carbon, Ca=calcium, N=Nitrogen, O=Oxygen, P=phosphate.



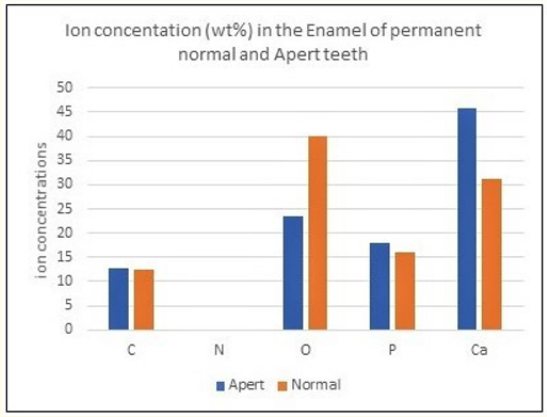
Graph 2: *Ion concentration (wt%) in the enamel of deciduous normal and Apert teeth.*

Note: C=carbon, Ca=calcium, N=Nitrogen, O=Oxygen, P=phosphate.



Graph 3: *Ion concentration (wt%) in the dentin of permanent normal and Apert teeth*

Note: C=carbon, Ca=calcium, N=Nitrogen, O=Oxygen, P=phosphate



Graph 4: *Ion concentration (wt%) in the enamel of permanent normal and Apert teeth*

Note: C=carbon, Ca=calcium, N=Nitrogen, O=Oxygen, P=phosphate

In Apert deciduous teeth, calcium concentration was higher in the dentin and enamel at average concentrations of 26.56% and 45.07%, respectively, in comparison to 6.25% in the dentin and 33.88% in the enamel of normal teeth. Similar findings were found in permanent teeth with average calcium concentrations in the dentin and enamel in Apert teeth of 32.69% and 45.82%, respectively, in comparison to 14.4% in the dentin and 31.1% in the enamel of normal teeth.

In all Apert deciduous teeth analyzed, phosphate concentration was higher in the dentin and enamel at average concentrations

of 11.19% and in 18.19%, respectively, in comparison to 3.49% in the dentin and 17.38% in the enamel of normal teeth. Similar findings were found in permanent teeth with average phosphate concentrations in the dentin and enamel of 12.93% and 16.53%, respectively, in comparison to 7.57% in the dentin and 15.91% in the enamel of normal teeth.

In addition, the dentin in Apert teeth showed a lower concentration of nitrogen compared to normal teeth. In AS deciduous teeth the concentration was 5.66%, compared to 28.1% in normal teeth. In AS permanent teeth the concentration was 5.01%, compared to 16.25% in normal teeth.

Ca/P ratio in enamel and dentin was significantly higher in Apert teeth compared to normal teeth (Table 2).

Ca /P	Deciduous teeth				Permanent teeth			
	Enamel		Dentin		Enamel		Dentin	
	Apert	Normal	Apert	Normal	Apert	Normal	Apert	Normal
	2.477	1.935	2.463	1.820	2.560	1.925	2.527	1.901

Table 2: *Ca/P ratio in Apert syndrome deciduous and permanent teeth compared to normal teeth.*

Note: Ca/P=Calcium to Phosphate ratio.

Discussion

One of the most interesting features of AS primary molars was the postnatal traumatic lines observed in the enamel. The first hypoplastic line observed on the slice, from the DEJ outwards, will be the neonatal line, and the latter will be traumatic lines [7]. In AS, 100% of the primary teeth and one permanent pre-molar showed postnatal traumatic lines in the enamel compared with none in controls. Postnatal formation of the enamel of the first primary molars ends at the age of 6 months, primary canines at 9 months, second primary molars at 10 - 12 months, and first pre-molars at 5-6 years (crown completion time) [8]. This study shows that AS children may suffer from severe traumatic (hypoxic) events as early as during the first year of their life and in early childhood, and these traumatic events leave a very distinct mark on postnatal enamel the traumatic lines observed, which are periods of disturbed mineralization causing thicker than normal striae of Retzius, due to impaired function of the ameloblasts [9,10].

Regarding the mineral content, AS teeth contained higher concentrations of inorganic ions, lower concentrations of nitrogen, and the ratio between Ca/P was higher than normal. This may imply AS teeth behave different physical properties than normal teeth, for example, fewer collagen fibers, which leads to a decrease in elasticity of the tooth structure and higher brittleness, which may cause a higher risk for cracks/fractures.

Conclusion

Apert Syndrome affects mineralization of enamel and dentin. It caused abnormal mineral content in comparison to match paired normal teeth. The main differences between AS deciduous teeth and normal are in calcium concentrations in both pre and postnatal enamel and phosphate in postnatal enamel.

The traumatic lines observed in both deciduous and permanent teeth implicate that severe traumatic episodes occurred during early childhood.

These findings show that AS also affects the mineralization of teeth in addition to the known oral motor challenges.

Ethics Statement

This is an observational study of teeth that the parents and children agreed to leave in the clinic after the treatment. The Barzilai Medical University Center Research Ethics committee has confirmed that no ethical approval is required.

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