



	Experiment title: Mapping Texture in the Dental Enamel of Patients Affected by Morquio's Syndrome	Experiment number: 28 01 875
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Report:

Morquio's Syndrome (Mucopolysaccharide disease Type IV) is an autosomal recessive lysosomal storage disease affecting between 1:75,000 and 1:200,000 live births. The disease is characterised by the reduced activity of the enzymes N-acetylgalactosamine 6-sulphatase (MPS-IV A) and beta-galactosidase (MPS-IV B) resulting in the accumulation of keratin sulphate and chondroitin 6-sulphate (glycosaminoglycans) in connective tissue, the skeletal system and in teeth. Clinically the disease manifests after infancy and is associated with severe skeletal abnormalities, restrictive lung disease, aortic valvular disease and ultimately limits life expectancy to less than 30 years. MPS-IV is associated with structural defects of the dental enamel of both the primary and secondary dentition. The sporadic distribution of MPS-IV affected individuals frequently results in anecdotal case reporting and as a consequence there is limited understanding of the disease impact on amelogenesis and the resultant enamel structure. Furthermore, the crystallography, composition and microstructure of dental enamel may potentially give insight into the pathogenesis of MPS-IV as the affected enamel can be chronologically related to systemic events or medical interventions.

In two previous experiments on XMaS (28.01.732 and 28.01.750) we have successfully shown that it is possible to map the texture distribution in the hydroxyapatite of healthy enamel and also in teeth affected by amelogenesis imperfecta (AI). AI describes a common group of non-syndromic inherited defects of dental enamel formation that exhibit marked genetic heterogeneity and affect the amount, composition, density and structure of the enamel with clinical dental manifestations similar to mucopolysaccharide diseases.

The aim of this study was to use the same methodology, i.e. 2D synchrotron x-ray diffraction mapping techniques, to characterise the texture distribution in the enamel of hypoplastic teeth taken from Morquio affected individuals, and to compare this with teeth taken from the same odontological sequence in unaffected sibling controls.

We used 9 shifts of beamtime on XMaS to study the 2D texture distribution of the dental enamel from intact tooth sections of patients affected by Morquio's Syndrome. Teeth specimens were taken from the collection at The Diana Princess of Wales Children's Hospital in Birmingham, currently providing medical care for one

of the largest cohorts of Morquio Syndrome affected children in Europe and where currently a novel enzyme therapy is being trialled. Each tooth was sectioned into 500µm thick sections using a diamond blade cutter. Diffraction patterns were collected every 20µm at high resolution in order to obtain 2-D images of the enamel areas of interest in each tooth section.

So far, we have analysed data and created 2D texture maps from deciduous teeth taken from MPS IV (Morquio Syndrome) affected individuals, teeth taken from MPS II affected individuals (Hunter Syndrome), and teeth taken from unaffected siblings (healthy controls).

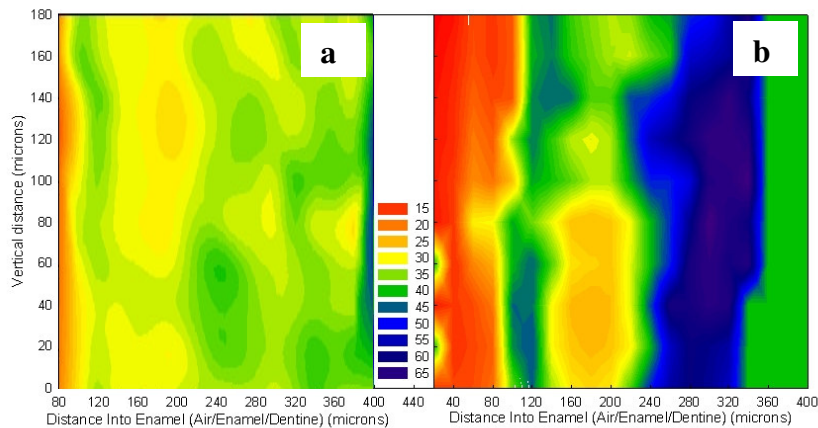


Figure 1 Texture distribution in a) deciduous enamel affected by MPS-IV and b) healthy deciduous enamel

Figure 1 shows the texture distribution of hydroxyapatite crystallites in deciduous enamel a) affected by MPS-IV and c) unaffected. There appears to be a marked difference in the texture distribution of enamel crystallites between enamel affected by MPS IV and healthy enamel. In healthy deciduous enamel (Figure 1b), we observe a steady decrease in crystal alignment when moving from the surface enamel towards the dentine interface (the amelodentinal junction or ADJ), as has been reported in the past by. However, in teeth affected by MPS-IV we have seen that the degree of texture only decreases very slightly when moving from the surface enamel to the ADJ (Figure 1a).

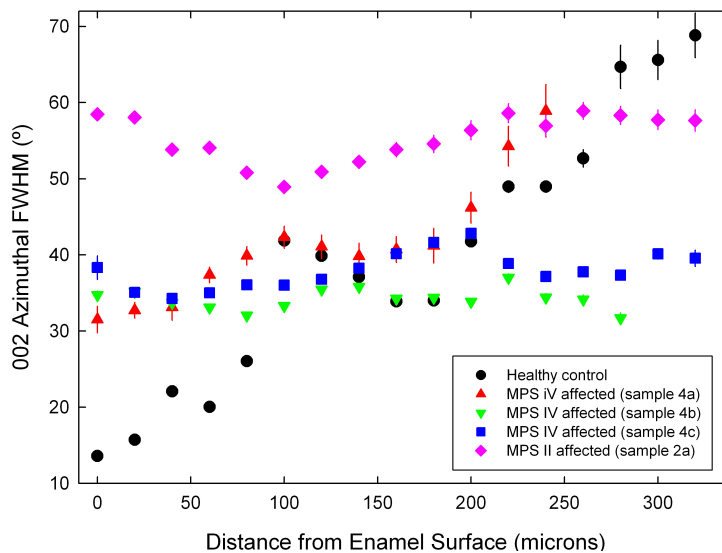


Figure 2 Texture plotted as a function of distance from the surface enamel towards the ADJ for various enamel specimens

Figure 2 shows the texture plotted as a function of distance from the enamel surface for three MPS IV affected individuals, one MPS II affected patient, and a healthy control. These data suggest that the degree of severity of symptoms in MPS individuals may be detected in different deviations from the “normal” texture distribution (black circles). Overall the degree of crystal orientation in surface enamel of MPS-IV and MPS II teeth is lower than expected perhaps a result of enamel malformation due to affected ameloblast cells.

Our findings will be presented at the Enamel VIII Conference in June 2011 and a paper is being prepared for submission to eCells and Materials (Open Access Journal).