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| **Pathology Question:** |
| What is tooth hypomineralization and what occurs during tooth development to lead to this? |
| **Report:** |
| Tooth hypomineralization is an enamel defect that can be first clinically observed at eruption of the tooth. It is an important clinical differentiation to identify being that it has high potential to lead to rapid breakdown of tooth structure and carious disease (Garg). Enamel mineralization of tooth crowns occurs at the maturation stage of enamel-forming ameloblasts, therefore resulting in a qualitative effect to the enamel (Alsaleh 54). The development of the ameloblasts was disrupted therefore leading to lesser mineralized final enamel crown. The affected enamel presents with normal thickness with a smooth white, yellow or brown surface. It is also opaque and usually presents on the incisal or cuspal third of teeth with a distinct boundary to the adjacent normal enamel (Alasaleh 54). The permanent molars frequently have “extensive caries” and require “atypical restoration” (Alsaleh 54). Therefore, with hypomineralization, the quality of the enamel is affected and the opacity that can be clinically seen on the tooth’s enamel surface may have high potential to cause breakdown in the dentition.The other enamel defect that may also occur is enamel hypoplasia. This defers from hypomineralization in that the disturbance occurs during the enamel matrix formation when the secretion phase of ameloblasts is taking place (Garg). This defect affects the quantity of the enamel, is usually localized to one tooth, and is associated with trauma to the primary teeth if present on the permanent teeth (Alsaleh 55). Enamel is fully formed typically in the first eight years of an individual’s life for permanent teeth and only then; it cannot be repaired once the tooth has erupted. The initial calcification of enamel begins as early at three months old. Both enamel defects discussed above occur from depressed activity of the enamel-forming ameloblasts, which are typically fully active during these ages and especially at initial calcification (Garg). Although crowns are formed throughout the first decade of life, tooth hypomineralization is often due to a disturbance in tooth formation, specifically ameloblast function, between birth and the first year of life from a severe systemic event that could affect tooth development even if it were secondarily, for example a high fever would have potential to disrupt enamel formation in that time frame (Alsaleh 54). In other cases, such as the one being presented in this case, there may be other systemic factors that contribute to the enamel defects early in life.In regard to the patient case for this Rounds presentation, both enamel hypomineralization and hypoplasia are common oral findings of Alagille Syndrome (Bernivzei-Royko 479). As stated in the article by Dr. Bernivzei-Royko, “Dental manifestations are not a primary feature of the syndrome, but they invariably occur as a complication of the long-lasting cholestasis and are linked to hyperbilirubinemia. As a consequence of cholestasis during odontogenesis, enamel opacities, hypomineralization, and hypoplasia of tooth enamel can appear” (479). Cholestasis is an event that happens when the bile flow from the liver is blocked which then leads to hyperbilirubemia and thus jaundice. This high level of bilirubin causes a biliverdin, a green bile pigment, to be deposited in dental tissues, including enamel (Bernivzei-Royko 470). Therefore, it is very likely that in a patient presenting with enamel defects that also has Alagille Syndrome that the tooth hypomineralization was likely due to high levels of bilirubin that accumulated in the first few months of life, and potentially in utero as well if primary dentition is also affected. |
| **References:** |
| Alsaleh, Majd DDS MS. Enamel Development. Marquette University School of Dentistry. Oral Biology. Spring 2020. Berniczei-Royko A, Chalas R, Mitura I, Nagy K, Prssak E. Medical and dental management of Alagille syndrome: a review. Med Schi Monit. 2014;20;476-480. Published 2014 Mar 24. doi:10.12659/MSM.890577Garg N, Jain AK, Saha S, Singh J. Essentiality of Early Diagnosis of Molar Incisor Hypomineralization in Children and Review of its Clinical Presentation, Etiology and Management. Int J Clin Pediatr Dent 2012;5(3):190-196. |