Manifestasi klinis, aspek genetika molekuler dan management dentinogenesis imperfekta

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Abstrak

Dentinogenesis imperfecta (DGI) is an autosomal dominant disorder in which both the primary and the permanent teeth are affected. It occurs with an incidence of 1:8.000 live births. In DGI, the teeth are amber and opalescent, and the pulp chamber is obliterated by abnormal dentin. The enamel, although otherwise unaffected, tends to fracture, which leads to rapid attrition of dentin and marked shortening of the teeth. There are three types of DGI with similar dental abnormalities. Type I occurs in people with osteogenesis imperfecta, a genetic condition in which bones are brittle and easily broken. DGI types II and III occur in people without other inherited disorders than mutations mapped to the 6.6-cM D4S2691-D4S2692 interval at 4q21, which is the locus for the dentin sialophosphoprotein (DSPP) gene. It is now believed that the DGI types II and III may be the same disorder. This paper reviews clinical manifestation, aspects of molecular genetics, and management of DGI.