Dentinogenesis imperfecta type III (DGI-III) is one of five distinct, hereditary disorders of dentin development affecting the teeth. Dentin is the hard, bone-like material that makes up most of a tooth and lies under the enamel serving to protect the soft, pulp tissue. These heritable dentin disorders may affect only the teeth or may be associated with the condition known as osteogenesis imperfecta. Whether this association is present is a major criterion in the classification of dentinogenesis imperfecta into three types. Dentinogenesis imperfecta type III is characterized by rapid erosion of the crowns in baby and permanent teeth. Dental pulp inside several teeth may be exposed. This pulp may be opalescent, smooth, and amber colored. Pulp chambers and root canals may appear very large on X-ray photos of baby teeth. Permanent teeth may have a reduction or even complete loss of the pulp chambers and root canals. Carriers of the gene for this disorder may have teeth that appear normal. However, upon examination their teeth have only an extremely thin ivory layer and an enlarged pulp chamber (shell teeth). Pitting of the tooth enamel may occur in the permanent teeth of patients. X-rays of the teeth are key to the diagnosis after a thorough family history and clinical examination.