

The use of methods of radiation diagnosis to identify the morphological features of the teeth in patients with hypophosphatemic rickets

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Hypophosphatemic rickets (XLHR) is a rare hereditary disease, prevalence of which is about 1 in 20 000 newborns. The most prevalent type is inherited in an X-linked fashion and caused by mutation in the gene encoding phosphateregulating endopeptidase homolog (PHEX), predominantly expressed in osteoblast and odontoblast.

A linkage between hereditary hypophosphatemic rickets and dental alterations seems to be obvious as the pathways of bone and dental hard tissues mineralization are similar. One of the main findings in patients with known XLHR is multiple abscesses or sinus tracts associated with carious free teeth of the primary and the permanent dentition. Using microcomputer tomography in the experiment and cone-beam computed tomography in the clinic, we have identified such morphological features in XLHR patients as a very large pulp chambers with prominent pulp horns extending to dentine-enamel junction. The affected teeth are characterized by a thin enamel layer and porous dentine. Due to the irregular structured dentine, the enamel fissures may extend into the pulp horns exposing the pulp to a high risk of microbial invasion. This may be an explanation for the seemingly spontaneous pulp necrosis.