ABSTRACT

Osteogenesis imperfecta (OI), commonly known as "brittle bone disease",is a dominant autosomal disorder characterized by bone fragility and abnormalities of connective tissue with a wide spectrum of severity, raging from very mild bone fragility to lethal forms. It principally affects those tissues containing the main fibrilla collagen type I, e.g. bone and teeth. It also affects sclerae, joints, tendon, and skin. A variety of structural abnormalities of dentin have been described in dentinogenesis imperfecta. The different facial characteristics of the various types of OI are related to their severity. OI patients usually present with a triangular face, a protrusive bitemporal bone, and prominent frontal bone. Dental malocclusions are marked in many OI subjects and include a high incidence of Class III malocclusions, an anterior and/or posterior crossbite, and a posterior openbite.

Key words : Osteogenesis imperfecta, dentinogenesis imperfecta, malocclusion