Abstract: Molar incisor hypomineralization (MIH) is defined as the developmentally-derived dental defect that involves 1 to 4 permanent first molars, frequently associated with similarly affected permanent incisors. The prevalence of MIH ranges from 3.6–25%. Several etiological factors are mentioned as the cause of the defect including prenatal, perinatal and childhood medical conditions that affect the developing enamel, while an underlying genetic predisposition could not be excluded. MIH presents as demarcated enamel opacities of different color in the affected teeth that occasionally undergo post eruptive breakdown due to soft and porous enamel, resulting in atypical cavities or even to complete coronal distortion. Accordingly the defect reveals serious clinical management problems attracting the attention of the dental profession. Children with MIH receive much more dental treatment than unaffected children. Affected molars usually require extensive treatment and might create serious problems for both patient and clinician, as they can frequently be difficult to anaesthetize and to restore adequately. Children at risk should be monitored very carefully during the period of eruption of their first permanent molars. Intensive individually prescribed preventive programs may postpone the initiation of the actual restorative treatment and reduce in long-term patient’s discomfort. Treatment planning should consider the long-term prognosis of these teeth. Advances in dental materials have provided clinical solutions in cases that were regarded as unrestorable in the past. In this review we would have a look at etiology, clinical presentation and treatment options of hypomineralized molar and incisor teeth.