Summary

Introduction: Osteogenesis imperfecta (OI) is a genetic disorders of connective tissue. Hearing loss has been repeatedly reported as a major syndrome of osteogenesis imperfecta. The hearing loss in OI is predominantly of the conductive type. **Material and methods:** The family (mother and two daughters) suffering from osteogenesis imperfecta are presented. **Results:** Audiological examination is presented. It revealed various types of hearing loss in members of family (conductive, sensorineural, mixed). **Conclusions:** On this example variety of hearing problems in this disease are demonstrated. Etiology and classification of osteogenesis imperfecta, clinical proceeding and treatment are presented.