OSTEOGENESIS IMPERFECTA AND HEARING .

Kaija M.Kuurila, Reidar A.Grénman, Reijo K.Johansson, Ilkka I.Kaitila*
Department of Otorhinolaryngology-Head and Neck Surgery, Turku University Central Hospital, Turku; Department of Clinical Genetics, Helsinki University Central Hospital, Helsinki*, Finland

The principal symptoms of Osteogenesis imperfecta include bone fragility, blue sclerae, and, in about 50% of the patients, progressive hearing loss.
PURPOSE OF THE STUDY: To evaluate the frequency of hearing loss in adult patients with OI. METHODS: In a nationwide search 230 Finnish OI-patients were ascertained corresponding a prevalence of 1/22 000. The 204 patients participating the study (87%) were evaluated by a questionnaire, and 181 patients by clinical audiometry. This report focuses on audiometric evaluation of 133 adult patients. 91 patients could be typed according Sillence classification. Hearing loss was defined as pure tone average (PTA 0.5-2kHz) above 15 dB on patients less than 60 years of age and above 20 dB in patients older than 60 years.
SUMMARY OF THE RESULTS: The mean age of 50 men and 83 women was 40.4 years (17-87 years). 62.4% of the cases were familial, 83.5% had blue sclerae, 39% dentinogenesis imperfecta and all except one had had fractures. 29 patients (21.8%) had undergone ear surgery, bilateral in 41.4% of cases. In the non-operated 225 ears there was hearing loss in 46.2%. Hearing loss in 9.6% was conductive, in 44.2% mixed and in 33.6% sensorineural. 5.8% had anacusis and the rest 6.8% were unclassifieble. Hearing loss predominantly began after the age of 30 years, increased with age, and developed from conductive to mixed and sensorineural. In 84.6% of the cases PTA 0.5-2kHz was more than 30 dB. OI type I was most common, 75.8 % (69/91). 50% of the ears had hearing loss, which predominantly was from mild to moderate, and of mixed or sensorineural type. All 3 patients (3.3%)with type III had normal hearing. 19.8% (18/91) had OI type IV. 61% had normal hearing, and all the 6 patients with OI type IVA had normal hearing.The hearing loss in 6/10 patients with type IVB was predominantly of mixed type, and from mild to moderate. CONCLUSION: In adult patients with OI hearing loss was found in 46.2% of non-operated ears. Hearing loss progressed from conductive to mixed and sensorineural with age. It was most common in OI type I, and no hearing loss was found in type III and IVA.

Reference: Proceedings of the 7th International Conference on Osteogenesis Imperfecta. Montreal, Canada, 1999.