

Treatments for hearing loss in osteogenesis imperfecta: a systematic review and meta-analysis on their efficacy

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Background

Hearing loss affects 70% of the people suffering from osteogenesis imperfecta (OI), a genetic disorder of the connective tissue mainly caused by mutations in the collagen type I. Hearing loss in OI has an early onset, is progressive, and can be conductive or sensorineural or mixed. There is no cure for OI and current treatments for its hearing loss rely on conventional surgical treatments for auditory impairments. However, these treatments outcomes may differ from those of the general population due to the underlying physiopathology of OI disorder characterized by bone fragility and soft tissue laxity. Here, we therefore present a systematic review and meta-analysis conducted on the efficacy of treatments addressing hearing loss in OI.

Methods

This study conforms to reporting standards of the Preferred Reporting Items for Systematic Reviews and Meta-analyses (PRISMA). Data sources used for this study includes PubMed, Web of Science, Scopus, and Google Scholar, from their inception to April 2020. We included published studies of OI patients undergoing a hearing loss treatment and assessment using pure tone audiometry with a specified length of follow up. We performed screening and data extraction, and discrepancies were resolved through consensus or adjudication. A random effects meta-analysis was conducted on 12 articles including OI patients that underwent stapes surgery. Articles including other treatments were excluded because of their small sample size (≤ 3 patients) or low number of articles on the topic. We conducted a meta-analysis on the proportion of patients with an air-bone gap (ABG) ≤ 10 dB. We further run sub-group meta-analysis to assess the efficacy of the two techniques to perform stapes surgery, stapedectomy and stapedotomy.

Results

Our search identified 953 studies, of which 32 were reviewed at full-text screening. Among them, 22 articles were about stapes surgeries, 8 about cochlear implants, 1 about bone anchored hearing aids and 1 about implantable hearing aids. We included 12 articles that met our inclusion criteria for meta-analysis, all of them were about stapes surgeries. The efficacy of stapes surgeries was assessed as the proportion of patients with a postoperative ABG ≤ 10 dB. Our results show stapes surgeries to have a success rate of 65.4% (95% CI, 55.1 to 75.1) in the OI population, with stapedectomy and stapedotomy being equally efficient, with a proportion difference between them of 5.43% (96% CI, -5.83 to 16.94, $P=0.35$).

Conclusions

Our meta-analysis indicate that the efficacy of stapes surgery has a scarce 65% success rate in the OI population compared to the 95% success rate in the general population. Overall, this study suggests that further studies are needed on hearing loss treatments for OI people. Particularly, the mechanisms of hearing loss in OI need to be determined in order to develop successful non-invasive treatment strategies.