Background

- 50% of patients with osteogenesis imperfecta (OI) suffer from hearing loss
- Conductive hearing loss
  - onset 2nd to 4th decade
  - due to abnormal bone remodeling affecting the stapes footplate (middle ear)
  - evolves to a mixed hearing loss when the pericochlear bone and otic capsule become involved in the aberrant bone remodeling process
- Stapes surgery in OI:
  - high risk of complications (bleeding of mucosa, ossicular atrophy)
  - cannot eliminate the progressive sensorineural component (inner ear) of hearing loss
  - large series have shown successful hearing loss reduction in the majority of OI patients
- Research aims:
  - to report on the intra-operative findings in a Belgian-Dutch series of OI patients undergoing stapes surgery
  - to present the audiometric findings after the longest postoperative follow-up published to date

Methods

Surgery reports and preoperative, short-term and long-term postoperative audiometric results from 10 Belgian and 12 Dutch OI patients operated at different institutes were obtained retrospectively

Surgery:
- 10/22 patients had bilateral stapes surgery
- 12/22 unilateral stapes surgery
- 29 ears: primary stapes surgery
- 5 ears: revision stapes surgery
- 24 ears: incudostapedotomy
- 4 ears: total stapedectomy
- 1 ear: partial stapedectomy
- 34 operated ears

Audiometric evaluation:
- Preoperative audiometry: 34/34 ears
- Short-term post-operative audiometry (<12 months): 28/29 primary operated ears and 5/5 revision surgery ears
- Long-term post-operative audiometry (mean duration: 16 years; range: 1–37 years): 19/29 primary operated ears and 3/5 revision surgery ears

Results

- Primary stapes surgery
  Short-term postoperative audiometry revealed improved hearing and reduced air-bone gaps in 28/29 ears with primary stapes surgery. In the 19 ears with long-term postoperative audiometric follow-up hearing gain was still significant at the latest audiometric evaluation.

- Revision stapes surgery
  Short-term postoperative audiometry revealed significant hearing gain and air-bone gap closure in 5/5 ears with revision surgery. Long-term follow-up in 3 ears with revision surgery demonstrated maintenance of hearing improvement.

Discussion and Conclusion

- Beneficial hearing gain and reduction of air-bone gap in the large majority, continuing for several decades
- Our long-term results (mean duration: 16 years) reveal a better hearing gain preservation than reported in other series of equal magnitude but with a shorter follow-up period
- The need for revision surgery is most often caused by erosion of the long process of the incus, which asks for a new prosthesis attached to an alternative location.
- In contrast with previous reports, a good postoperative result is obtained in most of the ears with revision surgery after an initially successful primary intervention.