STAPES SURGERY IN OSTEOGENESIS IMPERFECTA: RETROSPECTIVE ANALYSIS OF 34 OPERATED EARS

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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 succesful 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

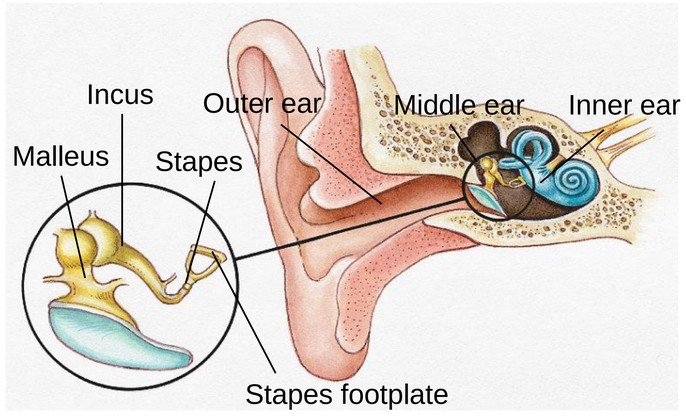


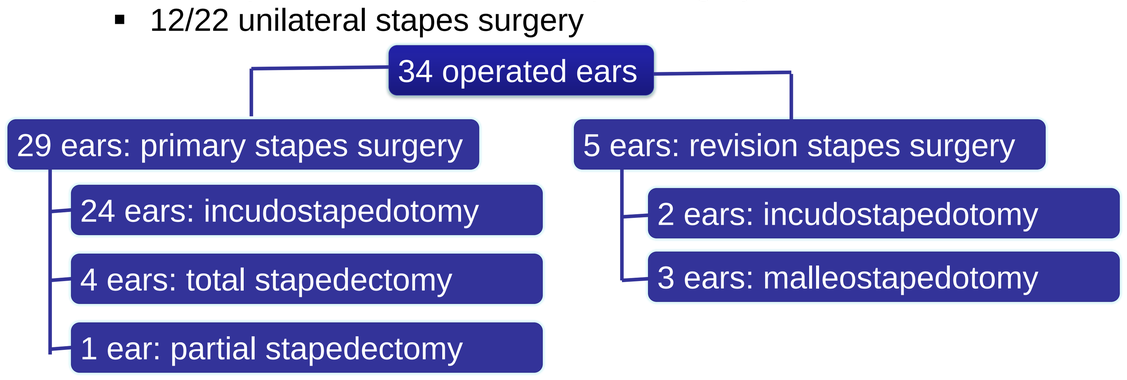
Figure 1. Cross-section of the ear, in which we distinguish the outer, middle and inner ear. The hearing loss in OI is most often caused by fixation of the stapes footplate, which hinders the transduction of sound waves to the inner ear.

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



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.

Figure 2. Mean preoperative audiogram (N=19)

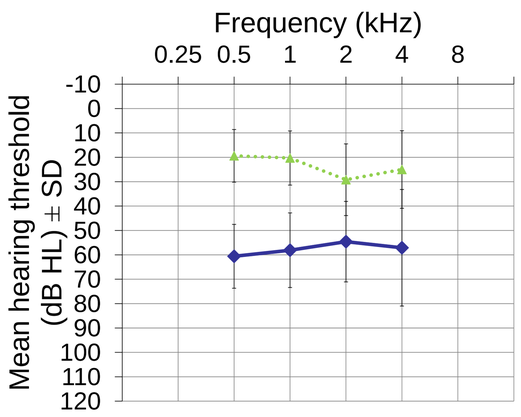


Figure 3. Mean short-term postoperative audiogram (N=19)

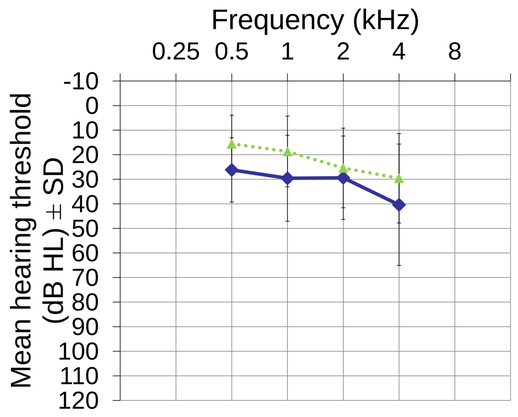


Figure 4. Mean long-term postoperative audiogram (N=19)

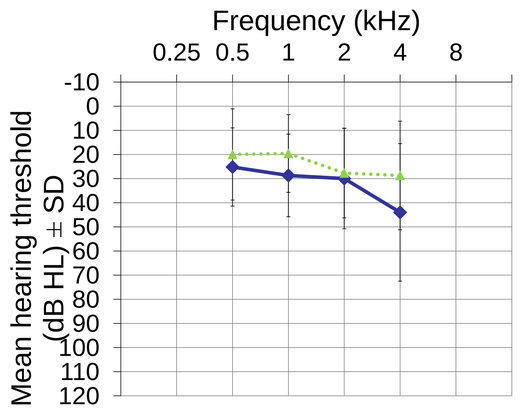


Table 1. Intraoperative findings in primary stapes surgery

	N (%)
Footplate	
- Fixed	29 (100)
- Thickened	20 (69)
- Brittle	8 (28)
- Thin	1 (3)
Stapes crura	
- Atrophic	13 (45)
- Fractured	1 (3)
Incus	
- Atrophic thinning	6 (21)
Middle ear mucosa	
- Hypervascularized	6 (21)
- Thickened	2 (7)
- High bleeding tendency	6 (21)

❑ 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.

Table 2. Intraoperative findings in revision surgery

	N
Prosthesis fixed in extremely thin and hard footplate	1
Fractured long process of the incus	3
Loose piston	1

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 succesful primary intervention.

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