Sensorineural Hearing Loss and Otosclerosis: A Clinical and Radiologic Survey of 437 Cases

YOUNG JE SHIN¹, BERNARD FRAYSSE¹, OLIVIER DEGUINE¹, CHRISTOPHE COGNARD², JEAN-PAUL CHARLET³ and ANNICK SÉVELY²

From the ¹Departments of Otolaryngology and ²Neuroradiology, Purpan Hospital, Toulouse, France and ³Department of Epidemiology, Hôtel-Dieu Saint Jacques, Toulouse, France

Shin YJ, Fraysse B, Deguine O, Cognard C, Charlet JP, Sévely A. Sensorineural hearing loss and otosclerosis. A clinical and radiologic study of 437 cases. Acta Oto Laryngol 2001; 121: 200–204. The aim of this study was to determine if a relationship exists between bone level thresholds and the extension of otosclerotic foci within the otic capsule. The study consisted of a retrospective case review in a university hospital. We included patients who underwent surgery for otosclerosis in our department and who had a CT scan prior to surgery. We analyzed the data charts and CT scans of 437 cases (386 patients). On CT scan, we distinguished patients with fenestral otosclerosis and/or with a pericochlear focus. A pericochlear focus could be extended (Group 2) or not (Group 1) to the cochlear endostium. Data for Groups 1 and 2 were compared with those for the control group of all patients for whom CT scan showed no cochlear focus (Group 3). Of the 437 CT scans, 399 were positive (91.3%). An anterior focus was reported in 305 cases (69.8%), a footplate thickening in 21 cases (4.8%) and both anomalies were encountered in 60 cases (13.7%). A pericochlear focus was reported in 53 examinations. This focus was extended to the endostium in 14 cases (26.4%) of the pericochlear foci). In Group 1, preoperative air conduction (AC) thresholds were significantly lower than in the control group ($p<0.05$). The air–bone gap was also significantly larger in Group 1 ($p<0.05$). Bone conduction (BC) thresholds were lower in Group 1 than in the control group but the difference was not significant. In Group 2, preoperative AC thresholds were significantly lower than in the control group ($p<0.05$). BC thresholds were also lower in Group 2 than in the control group and the difference was significant ($p<0.05$). As a result of this study, we assume that there may be a relationship between bone level thresholds and the radiological extension of otosclerosis within the otic capsule. Key words: bone conduction thresholds, cochlea, computerized tomography, deafness, otosclerosis, radiology, sensorineural hearing loss.

INTRODUCTION

Mixed hearing loss is commonly encountered in patients with otosclerosis. Based on clinical experience, most otologists agree that otosclerosis can damage the inner ear and cause progressive sensorineural hearing loss (SNHL).

Several theories have attempted to explain the cause of SNHL in otosclerosis and it is conceivable that more than one of these theories may have a role to play. In 1911, Siebenmann [cited in Schucknecht (1)] described the inner ear pathology of a 32-year-old woman with otosclerosis and suggested that these changes were due to an accumulation of the products of inflammation of the inner ear. The enzymatic theory has been proposed as one mechanism of SNHL. According to this theory (2), the proteolytic enzymes released by the otospongiotic–otosclerotic foci damage the inner ear. Spillage of enzymes from an active focus of otosclerosis into the cochlear fluid has been reported (3). These authors found a relationship between the amount of enzymatic activity of otospongiotic foci and the progression of SNHL. Another theory was the creation of venous shunts between the membranous labyrinth and the active otosclerotic bone. Ruedi (4) reported that pre-existing capillaries and veins from the labyrinth anastomose with the vessels of the otosclerotic focus. Venous congestion was therefore thought to cause cochlear hypoxia and hair cell malfunction.

Schuknecht (1) has demonstrated histologically that the inner ear structure most often affected by otosclerosis is the spiral ligament, which represents the area of attachment between the auditory canal and the endostium. Parahy and Linthicum (5) reported that the degree of SNHL was directly related to the amount of hyalinization of the spiral ligament. According to these authors, hyalinization occurs adjacent to active otospongiotic lesions. Once the endostium is involved by a focus, hyalinization can spread laterally from small channels through the endosteal bone.

Two histologic studies assessed the relationship between the degree of SNHL and size and involvement of the endostium by otosclerosis: no correlation was found in one study (6) and a good correlation in another (7). Radiologic studies have also assessed such a relationship. De Groot et al. (8) reported a positive correlation between CT findings and the degree of SNHL. In 1993, Vartiainen and Saari (9), who compared the CT results and audiograms of 40 patients, concluded that CT examination was of little value in predicting sensorineural hearing loss in patients with otosclerosis.
With regard to this controversial issue, the goal of this study was to investigate if a relationship exists between bone threshold levels and extension of the otosclerotic focus within the otic capsule.

MATERIALS AND METHODS

Inclusion criteria

We included patients who underwent surgery for otosclerosis in our department and who had had a CT scan prior to surgery. The diagnosis of otosclerosis, i.e. stapes fixation and visualization of a macroscopic focus of otosclerosis, had to be confirmed at surgery, thereby excluding minor malformations or tympanosclerosis of the footplate.

Population

We analyzed the data charts and CT scans of 437 cases (386 patients). The mean age of the population was 46.2 years (SD = 12.8). The female : male ratio was 1.84 (women: 64.8%; men: 35.2%). Unilateral deafness was reported in 109 patients (28%). Mean air conduction (AC) thresholds were at 22.4 dB, mean bone conduction (BC) thresholds at 52.4 dB and the mean air–bone gap was 30.0 dB for the whole population. AC and BC thresholds are reported from 0.5 kHz to 8 kHz in Table I.

CT scan

All patients referred to our institution for conductive or mixed hearing loss without patent otoscopic anomalies underwent a CT scan prior to surgery. We based our analysis on the report of the neuroradiologist at the center where the examination was performed. When in doubt, a neuroradiologist at our center made a new examination of the CT scan, which was used for our analysis.

Otosclerotic lesions were characterized by hypodensity of the otic capsule or footplate thickening. Foci were classified into four categories (10, 11): anterior focus (fenestral focus); per cochlear focus without or with endosteal extension; and footplate thickening.

Of the 437 CT scans, 399 were positive (91.3%). Anomalies were bilateral in 338 cases (84.7%) and unilateral in 61 cases (15.3%). An anterior focus was reported in 305 cases (69.8%), footplate thickening in 21 cases (4.8%) and both anomalies in 60 cases (13.7%). A per cochlear focus was reported in 53 examinations. This focus was extended to the endosteum in 14 cases (26.4% of the per cochlear foci).

Groups

We distinguished three groups in our study:

- Group 1. Patients with a per cochlear focus not extended to the endosteum on CT scan (n = 39);
- Group 2. Patients with a per cochlear focus extended to the endosteum on CT scan (n = 14);
- Group 3. Control group. This group comprised all patients without per cochlear involvement on CT scan (n = 384). Audiometric data for this group are reported in Table II.

Hearing loss

Audiometric testing was performed in double-walled sound rooms at our center. Hearing evaluations con-

<table>
<thead>
<tr>
<th>Table I. Audiometric data (dB HL) for the overall population (437 cases), with standard deviations in parentheses</th>
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<tbody>
<tr>
<td>0.5 kHz</td>
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<td>-------</td>
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<tr>
<td>BC</td>
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<tr>
<td>AC</td>
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<td>Air–bone gap</td>
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| Table II. Pre- and postoperative audiometric data (dB HL) for Group 3 (control group), with standard deviations in parentheses |
|-----------------|------|------|------|------|------|------|      |
|                 | Overclosure |
|                 | n   | BC   | AC   | Air–bone gap | 1 kHz | 2 kHz | 4 kHz | Mean  |
| Preop.          | 384 | 21.5 (11.2) | 51.1 (15.3) | 29.6 (9.3) | 3.4 (8.8) | 5.7 (9.1) | –0.2 (10.3) | 3 (7.5) |
| Postop.         | 384 | 18.7 (10.7) | 30 (13.6) | 11.3 (7.1) | 2.5 (11.3) | 6.3 (11.6) | –1.7 (12.2) | 2.4 (9.7) |
| 1 year          | 199 | 19.9 (12.3) | 31.4 (15.2) | 11.5 (7.1) | 2.5 (11.3) | 6.3 (11.6) | –1.7 (12.2) | 2.4 (9.7) |
Table III. Pre- and postoperative audiometric data (dB HL) for Group 1 (pericochlear focus without extension to the endosteum), with standard deviations in parentheses

<table>
<thead>
<tr>
<th></th>
<th>n</th>
<th>BC</th>
<th>AC</th>
<th>Air–bone gap</th>
<th>Overclosure</th>
<th>1 kHz</th>
<th>2 kHz</th>
<th>4 kHz</th>
<th>Mean</th>
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<td></td>
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</tr>
<tr>
<td>Preop.</td>
<td>39</td>
<td>23.4</td>
<td>56.1</td>
<td>32.8</td>
<td>23.4</td>
<td>4.6</td>
<td>8.3</td>
<td>3.3</td>
<td>1.7</td>
</tr>
<tr>
<td>Postop.</td>
<td>39</td>
<td>21.2</td>
<td>32.4</td>
<td>11.1</td>
<td>5.6</td>
<td>0.2</td>
<td>0.5</td>
<td>0.3</td>
<td>1.1</td>
</tr>
<tr>
<td>1 year</td>
<td>24</td>
<td>20.9</td>
<td>31.9</td>
<td>11.0</td>
<td>7.3</td>
<td>0.2</td>
<td>0.4</td>
<td>0.3</td>
<td>1.5</td>
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</tbody>
</table>

Table IV. Pre- and postoperative audiometric data (dB HL) for Group 2 (pericochlear focus with extension to the endosteum), with standard deviations in parentheses

<table>
<thead>
<tr>
<th></th>
<th>n</th>
<th>BC</th>
<th>AC</th>
<th>Air–bone gap</th>
<th>Overclosure</th>
<th>1 kHz</th>
<th>2 kHz</th>
<th>4 kHz</th>
<th>Mean</th>
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<td></td>
<td></td>
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<tr>
<td>Preop.</td>
<td>14</td>
<td>43</td>
<td>75</td>
<td>32</td>
<td>2.1</td>
<td>7.5</td>
<td>0.8</td>
<td>3.2</td>
<td>0.9</td>
</tr>
<tr>
<td>Postop.</td>
<td>14</td>
<td>40.5</td>
<td>53.3</td>
<td>20.1</td>
<td>7.5</td>
<td>2.8</td>
<td>0.4</td>
<td>0.3</td>
<td>1.1</td>
</tr>
<tr>
<td>1 year</td>
<td>8</td>
<td>47.8</td>
<td>62</td>
<td>20.6</td>
<td>0</td>
<td>0.0</td>
<td>0.0</td>
<td>0.0</td>
<td>0.0</td>
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</table>

The preoperative test was performed the day before surgery and was used as the baseline; the 2-month postoperative result served as the comparison test. When available, 1-year audiometric results were also reported. If there was no response to pure-tone thresholds by AC, the response was coded at 120 dB hearing level (HL). When there was no response to pure-tone thresholds by BC, the response was coded at 75 dB HL. Hearing results were reported in a manner as close to that described by the Committee on Hearing and Equilibrium for conductive hearing loss (12) as the retrospective nature of this study would allow.

Statistical analysis

Statistical analysis was performed using both classical bivariate and non-parametric tests according to distribution features. Those tests included one-way ANOVA, paired and unpaired t-test and Mann–Whitney’s U-test for comparing the values of a continuous variable between categories. The Spearman rank correlation test was used when a relation between two continuous variables was involved. The significance threshold was 0.05.

RESULTS

A pericochlear focus was found in 53 of 437 CT scans performed. This focus was extended to the endosteum in 14 cases.

Group 1 (pericochlear focus not extended to the endosteum) comprised 39 cases. Hearing results before and 2 and 12 months after stapedotomy are reported in Table III. Mean preoperative BC was 23.4 dB (SD: 11.7), mean preoperative AC was 56.1 dB (SD: 14.6) and mean air–bone gap was 32.8 dB (SD: 8.3 dB). Preoperative AC thresholds were significantly lower than in the control group (p < 0.05). The air–bone gap was also significantly larger in Group 1 than in the control group but the difference was not significant. The mean high-frequency PTA (1000, 2000 and 4000 Hz) improvements in BC scores from their preoperative level 2 and 12 months after surgery were 2.2 dB and 1.5 dB, respectively (vs 3 dB and 2.4 dB in the control group, respectively; p = NS).

Group 2 (pericochlear focus with extension to the endosteum) comprised 14 cases. Hearing results before and 2 and 12 months after stapedotomy are shown in Table IV. Mean preoperative BC was 43 dB (SD: 23.6), mean preoperative AC was 75 dB (SD: 14.6) and mean air–bone gap was 32 dB (SD: 9.9 dB). Preoperative AC thresholds were significantly lower than in the control group (p < 0.05). BC thresholds were lower in Group 2 than in the control group and this difference was significant (p < 0.05). The mean high-frequency PTA (1000, 2000 and 4000 Hz) improvements in BC scores from their preoperative level 2 and 12 months after surgery were 3.2 dB and 1.5 dB, respectively (vs 3 dB and −0.4 dB in the control group, respectively). Even though postoperative bone thresholds were lower in Group 2 than in the control group at 1 year, a statistical difference could not be confirmed due to the small size of the groups.

The round window was obliterated in 14 cases. This obliteration was associated with a pericochlear
focus in 8 out of 14 cases. Among these eight cases, the pericochlear focus was extended to the endosteum in five cases. Hearing results before and 2 and 12 months after stapedotomy for these 14 patients are reported in Table V. Mean preoperative BC was 40.2 dB (SD: 24.9), mean preoperative AC was 72.3 dB (SD: 30) and mean air–bone gap was 32 dB (SD: 8.4 dB). Mean BC and AC were significantly lower than in the control group \( (p < 0.05) \).

**DISCUSSION**

The relationship between endosteal involvement and degree of SNHL has not yet been clearly elucidated and remains controversial.

Elonka and Applebaum (13) reported a study of 29 otosclerotic bones with stapes footplate involvement: 1 focus of endosteal involvement or 2 or more foci of endosteal involvement. Analysis of audiometric data showed that the group of bones with 2 or more foci of endosteal involvement had a similar incidence of 45 dB SNHL (9 of 15; 60%) as the group with no endosteal involvement (5 of 8; 62%). The group with two or more foci had a greater incidence of \( \geq 60 \) dB SNHL (46%) compared with the groups with none (12%) or 1 focus (16%) involved. These authors found no, or only a slight relationship, between SNHL and the location and extent of otosclerotic endosteal involvement.

Schuknecht and Barber (6) could find no correlation between extent of endosteal involvement and BC thresholds. Lesions with endosteal involvement had a mean threshold level of 35.9 dB, as compared to 36.6 dB for non-involved ears.

Kwok and Nadol (7) reported that when otosclerotic ears with two or more sites of endosteal involvement were compared with otosclerotic ears with one or no sites of endosteal involvement, the ears with two or more sites of involvement had mild but significantly greater atrophy of the striae vasculares and spiral ligaments.

Goycoolea (14) reported mean threshold levels of 28.3 dB when the endosteum was not involved, 33.7 dB when one focus of endosteal involvement was present and 62 dB when two or more foci of endosteal involvement were present.

Finally, in a temporal bone study, Fraysse and Uziel (15) reported that the amount of SNHL was greater when the endosteum was involved.

Radiological studies have attempted to answer the question concerning the relationship between SNHL and extension of the otosclerotic lesions to the endosteum. De Groot et al. (8) reported that, in cases of BC impairment, the bony labyrinth appeared normal in about half the ears. In the other half, areas of bone resorption were present and a positive correlation was found between the degree of bone loss and the amount of BC threshold loss. In contrast, Vartiainen and Saari (9), by comparing the CT scans and audiograms of 40 patients, concluded that CT examination is of little value in predicting SNHL in patients with otosclerosis. Although these authors published their report in 1993, their study concerned CT scans performed in 1986 and 1987.

Diagnosis of endosteal involvement was not easy to obtain until technical progress allowed the production of higher definition images. Under these circumstances, otosclerotic images are specific, with a sensitivity of 91.3% (16). Some authors argue that large otosclerotic lesions can go undetected by CT. Thiers et al. (17) point out that large mature and sclerotic lesions of the otic capsule may be missed using this technique. However, use of densitometric readings of the otic capsule has improved the detection of such lesions (18). With the use of high definition images, our study reports a relationship between the degree of SNHL and the extension of the focus within the otic capsule. When a pericochlear focus exists on CT scan and is extended to the endosteum, BC thresholds are significantly lower. Visualization of a pericochlear focus without extension to the endosteum is not sufficient to attribute a SNHL to otosclerosis. However, when this focus is extended to the endosteum, one can correlate SNHL and otosclerosis. Another point of interest may be that, at 1 year, postoperative results of BC overclosure were lower in the population with extension to the endosteum. These patients may present a worsening of their BC levels after surgery.

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**Table V. Pre- and postoperative audiometric data (dB HL) of patients with round window obliteration, with standard deviations in parentheses**

<table>
<thead>
<tr>
<th></th>
<th>( n )</th>
<th>BC</th>
<th>AC</th>
<th>Air–bone gap</th>
<th>Overclosure</th>
<th>AC improvement</th>
<th>ABG closure</th>
</tr>
</thead>
<tbody>
<tr>
<td>Preop.</td>
<td>14</td>
<td>40.2 (24.9)</td>
<td>72.3 (30)</td>
<td>32 (8.4)</td>
<td>-2.9 (1.7)</td>
<td>5.7 (9.1)</td>
<td>14.5 (12.5)</td>
</tr>
<tr>
<td>Postop</td>
<td>14</td>
<td>43.3 (27.6)</td>
<td>60.9 (35.2)</td>
<td>17.6 (10.8)</td>
<td>-5.4 (22.2)</td>
<td>6.3 (11.6)</td>
<td>8.3 (13.2)</td>
</tr>
<tr>
<td>1 year</td>
<td>8</td>
<td>48.9 (27.1)</td>
<td>71.7 (36)</td>
<td>22.8 (1.6)</td>
<td>-5.4 (22.2)</td>
<td>6.3 (11.6)</td>
<td>8.3 (13.2)</td>
</tr>
</tbody>
</table>

\( ABG = \text{air–bone gap.} \)
and one may expect poorer results after surgery. However, we did not find a significant difference due to the size of the population involved and this issue should be studied in a larger population.

Round window obliteration may explain some cases of unsuccessful stapes surgery. Schuknecht and Barber (6) found the round window niche to be blocked in 7% of 118 ears with clinical otosclerosis, and Hueb et al. (19) in 36.11%, while this prevalence in our radiologic study was only 3.2%. Of these 14 cases, 8 were associated with extensive lesions within the otic capsule and, therefore, the degree of SNHL could not be interpreted as the sole effect of the round window blockage. This might explain why mean BC and AC were statistically more elevated than in the control group.

CONCLUSION

In this study, we have reported a significant relationship between the amount of air–bone gap and the extension of the lesion on CT scan, and another relationship between the degree of SNHL and the degree of endosteal involvement.

Ours is also the first radiological study to report such a correlation in a large population. We conclude that CT examination is of high value in predicting SNHL in patients with otosclerosis.

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REFERENCES


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Address for correspondence:
Young Je Shin
Service d’ORL
Hôpital Purpan
1, place du Dr. Baylac
FR-31059 Toulouse Cedex
France
Fax: +33 5 61493644
Tel: +33 5 61772401
E-mail: shin.y@chu-toulouse.fr