
Malleus Ankylosis

Malleus Ankylosis: A Clinical, Audiometric, Histologic, and Surgical Study of 123 Cases

Robert Vincent, André Lopez, and Neil M. Sperling

Jean Causse Ear Clinic, Traverse de Béziers, Colombiers, France, and State University of New York Health Science Center Department of Otolaryngology, Brooklyn, New York, U.S.A.

Objective: Malleus ankylosis is a cause of conductive hearing loss that can be difficult to assess, particularly in association with otosclerotic stapes fixation. The aim of this study is to seek the clinical and audiometrical features unique to this pathology.

Study Design: The study design was a review of malleus ankylosis cases and an analysis of functional results after ossiculoplasty.

Setting: The study was performed at Jean Causse Clinic in France. **Patients:** The authors reviewed a series of 123 ears in 112 patients who underwent surgery for incudo-malleal ankylosis from January 1991 to September 1997.

Intervention: The surgical technique depends on the type of pathology encountered. In case of associated stapedial fixation, a stapedotomy with vein graft interposition and reconstruction with a total prosthesis will be performed during a same step.

Main Outcome Measures: Clinical evaluation, preoperative and postoperative audiometrical evaluation, operative findings, histologic examination, and postoperative functional results.

Results: In our series, a postoperative air-bone gap smaller than 10 dB was obtained in 77% of cases. These results confirm the possibility of managing both pathologies in a single surgical step. **Conclusion:** Incudo-malleal ankylosis remains an unusual pathology but should be systematically assessed during surgery and preferably after separation of the incudo-stapedial joint. A preoperative diagnosis is difficult to ascertain; some audiometrical features allow a suspicion. In this series, it is shown that otosclerosis can be responsible for ankylosis as seems to be confirmed by the two cases presented in this study. **Key Words:** Malleus ankylosis-Ossiculoplasty-Otosclerosis.

Am J Otol 20:717-725, 1999.

Malleus ankylosis is an unusual cause of conductive hearing loss. Recognized causes of malleus ankylosis include infection, trauma, or previous middle ear surgery, although some cases lack such a history. It is usually attributed to a congenital pathology, although it may be encountered in a middle ear without evidence of congenital deformity. The surgeon must systematically rule out the presence of an incudo-malleal ankylosis in all ears being explored for conductive hearing loss. Such fixation is more frequent in ears with otosclerotic ankylosis of the stapes and represents a significant cause of revision surgery in otosclerosis.

We review a series of 123 ears in 112 patients, operated on for incudo-malleal ankylosis and try to determine the clinical, and mainly

audiometrical, features unique to this pathology. This pathology represents a good indication for surgery, as it is shown by the analysis of the functional results in this series. Surgery is considered after a careful examination among which tuning fork testing is particularly important.

MATERIALS AND METHODS

One hundred twelve patients underwent surgery for incudomalleal ankylosis at Jean Causse Clinic from January 1991 to September 1997.

Clinical evaluation

A clinical evaluation consisting of the symptoms described by the patients, of their personal background and family history, and of the information provided by the clinical examination.

Preoperative and postoperative audiometrical evaluation

This audiometrical evaluation included: 1) audiometrical airborne gap calculated on the frequencies 0.5, 1, and 2 kHz; 2) bone conduction threshold calculated on the frequencies 0.5, 1, and 2 kHz; 3) bone conduction level at the frequency 4 kHz; 4) acoustic reflex tested contralaterally for both ears; 5) tympanogram.

Operative findings

This surgical evaluation was performed on 123 ears in 112 patients.

Histologic examination

The histologic examination included a histologic study of the head of the malleus and of the incus in 29 ears to seek the type and cause of incudo-malleal ankylosis.

Postoperative functional results

The postoperative functional results of 102 cases checked after a 6-month follow-up period were assessed. The results were assessed according to the usual method of the postoperative airborne gap calculated on 0.5, 1, and 2 kHz.

For the clinical evaluation and the preoperative and postoperative audiometrical evaluation, a comparison was made to a group of 112 patients who underwent stapedotomy (because malleus ankylosis is one of the main differential diagnosis of otosclerosis), looking for significant differences between the two groups.

CLINICAL EVALUATION

Sex-ratio

Of 112 patients operated on for malleus ankylosis from January 1991 to September 1997, there was a predominance of female patients (63% of cases compared to 37% males).

In the group of patients who underwent stapes surgery for otosclerosis, the sex-ratio was 70% female and 30% male.

Age

In the malleus ankylosis group, the average age was 50 years, ranging from 10 to 77 years. Eight pediatric cases were included in this series, all of which had a history of otitis media.

In the otosclerotic group, the average of age was 43 years, ranging from 13 to 75 years. There was one pediatric case (14 years).

Symptoms

Hearing loss was the primary symptom in 100% of cases in the two groups. In the malleus ankylosis group, it was associated with tinnitus in 18% of cases and with dizziness in 6% of cases. Three percent of the patients complained mainly of their tinnitus and 4% mainly of dizziness. In the otosclerotic group, hearing loss was associated with tinnitus in 38% of the cases and with dizziness in 4% of the cases.

Family history of deafness

For the malleus ankylosis group, in most of the cases (68%), there was no hearing loss in the family. There was a family history of hearing loss in 32% of the cases, among which 8% had a history of otosclerosis.

In the otosclerotic group, most of the cases had a family history of deafness (54%).

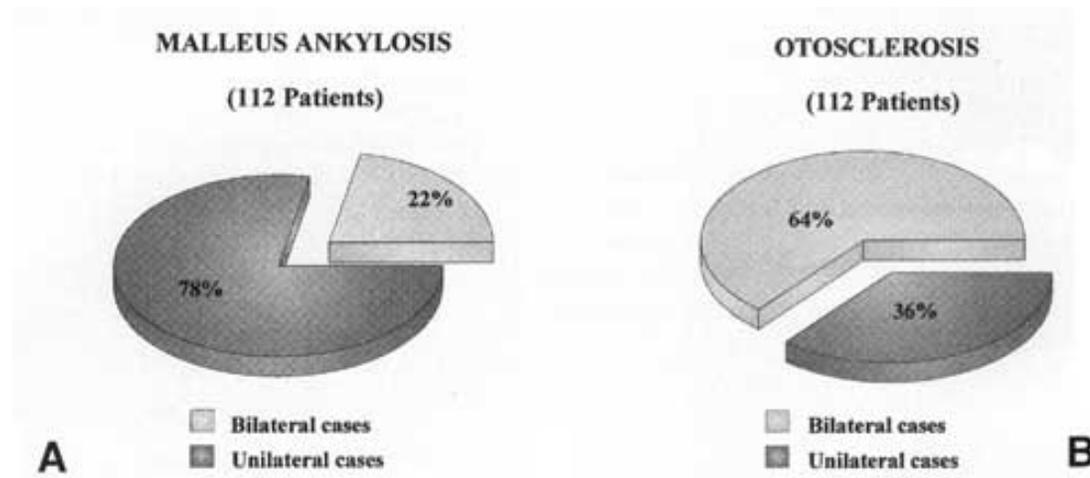


FIG. 1.

A: Malleus ankylosis group. The hearing impairment is mainly unilateral (78%).

B: Otosclerosis group. The hearing impairment is mainly bilateral (64%).

Otologic history

In the malleus ankylosis group, the otologic history of the patients was negative in 54% of the cases. A history of otitis media was found in 31 % of the cases. In 7% of the cases, the contralateral ear was operated on for otosclerosis. In 6% of the cases, the ipsilateral ear was previously operated on for otosclerosis. Iatrogenic causation could be questioned.

There was a history of trauma in 4% of the cases.

In the otosclerotic group, the otologic history of the patients was negative in 89% of the cases. A history of otitis media was found in 8% of the cases and trauma in 3% of the cases.

Otoscopy

For the malleus ankylosis group, in most of the cases, the tympanic membrane appeared normal (80%). There was a scarred eardrum in 8% of the cases. More rarely, there was a retraction pocket (4%), a tympanic perforation (2%), and one or several plaques of tympanosclerosis (6%).

In the otosclerotic group, the tympanic membrane was normal in most of the cases (99%).

AUDIOMETRICAL EVALUATION

In the group of patients who underwent surgery for malleus ankylosis, the hearing impairment was unilateral in 78% of the cases and bilateral in 22% of the cases (Fig. 1A).

In the group of patients who underwent stapes surgery, there was a minority of unilateral cases (36%). In most of the cases, the impairment was bilateral (64%) (Fig. 1B).

The incudo-malleal ankylosis seemed to be unilateral mainly.

Preoperative air-bone gap (0.5, 1, and 2 kHz)

The evaluation of the preoperative air-bone gap gave evidence of a difference between the two groups of patients:

- In the malleus ankylosis group, the air-bone gap was generally small; it was less than or equal to 20 dB in 55.5% of cases, with most of the patients having an air-bone gap between 18 and 20 dB. A larger air-bone gap, greater than 30 dB, was found only in 11 % of the cases (Fig. 2A).
- In the otosclerosis group, the proportions were reversed: in 87% of the cases, the air-bone gap was greater than 20 dB, with an air-bone gap greater than 30 dB in 41 % of the cases (Fig. 2B). This difference between the two pathologies seemed to confirm that malleus fixation results in a relatively small air-bone gap.

Bone conduction calculated on 0.5, 1, and 2 kHz

The study of the bone conduction calculated on those three frequencies allowed to assess the degree of preoperative sensorineural hearing loss. A new difference appeared clear:

- In the malleus ankylosis group, most of the cases (56%) demonstrated a sensorineural hearing loss greater than or equal to 20 dB with 22% demonstrating significant sensorineural hearing loss (hearing loss >30 dB) (Fig. 3A).
- In the otosclerosis group, preoperative sensorineural hearing loss was observed less frequently. A pure tone average of bone conduction greater than or equal to 20 dB, was found in 33% of the cases with a sensorineural hearing loss greater than 30 dB in 11 % of the cases. In most of the cases, the preoperative bone conduction was normal (5:20 dB loss in 67% of the cases) (Fig. 3B).

In our study, malleus ankylosis was also characterized by a clear impairment of the preoperative bone conduction level.

Preoperative bone conduction level at 4 kHz

A study of the bone conduction level at 4 kHz was particularly interesting for more precisely assessing the degree of preoperative sensorineural hearing loss in the two groups.

Looking at this feature, there was an obvious difference because the malleus ankylosis group showed a marked sensorineural hearing loss in this frequency with a hearing loss greater than or equal to 30 dB in 57% of the cases (Fig. 4A). In the otosclerosis group, the bone conduction loss at 4 kHz remained less than or equal to 20 dB loss in 43% of the cases (Fig. 4B). A sensorineural hearing loss at this frequency greater than 30 dB was seen in 29% of the cases.

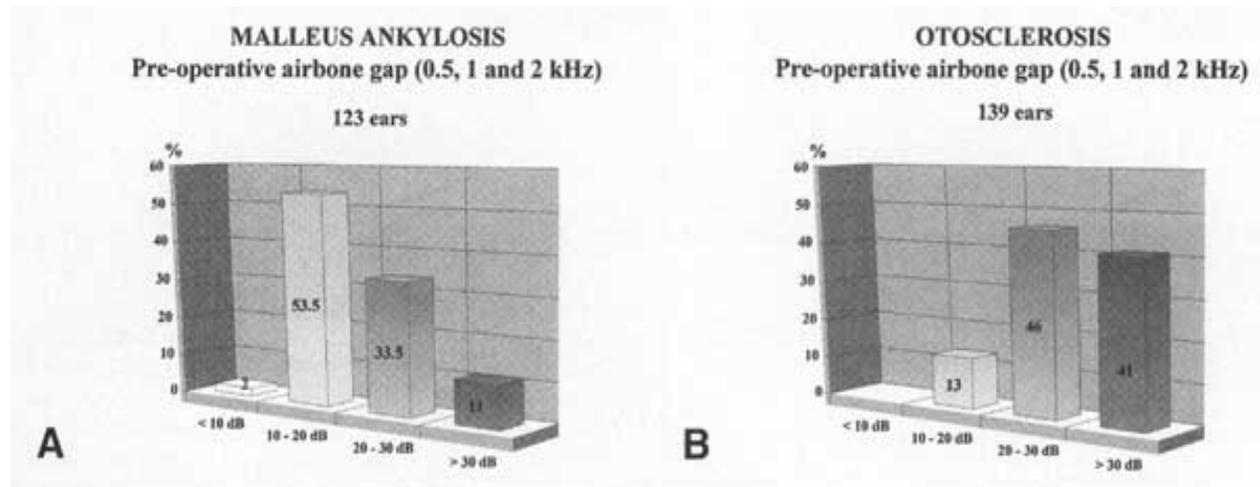
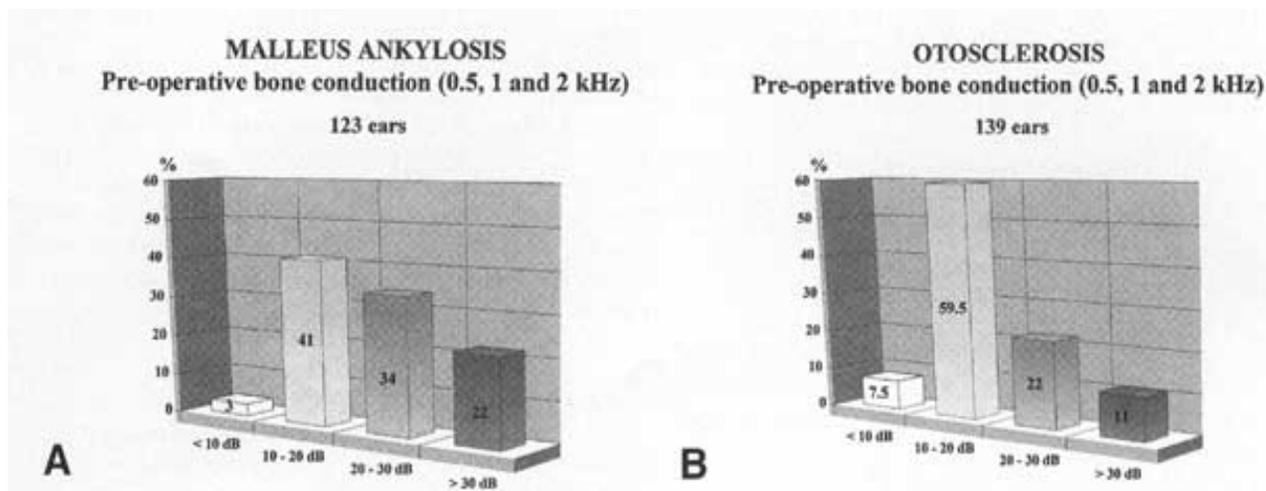


FIG. 2.

A: Malleus ankylosis group. Assessment of the preoperative air-bone gap (0.5, 1, and 2 kHz).

B: Otosclerosis group. Assessment of the preoperative air-bone gap (0.5, 1, and 2 kHz).

**FIG. 3.**

A: Malleus ankylosis group. Calculation of the preoperative bone conduction (0.5, 1, and 2 kHz).

B: Otosclerosis group. Calculation of the preoperative bone conduction (0.5, 1, and 2 kHz).

Acoustic reflex

Contralateral reflexes were absent in the impaired ear in 100% of the cases in the malleus ankylosis group and in the otosclerotic group. A marked difference between the two groups was noted in the cases in which the conductive hearing impairment was unilateral. This situation was found in 87 patients in the malleus ankylosis group and in 40 patients in the otosclerosis group.

In these two subgroups, the acoustic reflex was absent on the impaired side in all the cases. In contrast, on the normal ear, there was an important difference:

- In the malleus ankylosis group, the contralateral acoustic reflex was present in 65% of the cases (Fig. 5A).
- In the otosclerosis group, the contralateral acoustic reflex was absent in spite of the absence of hearing loss in 70% of the cases (Fig. 5B).

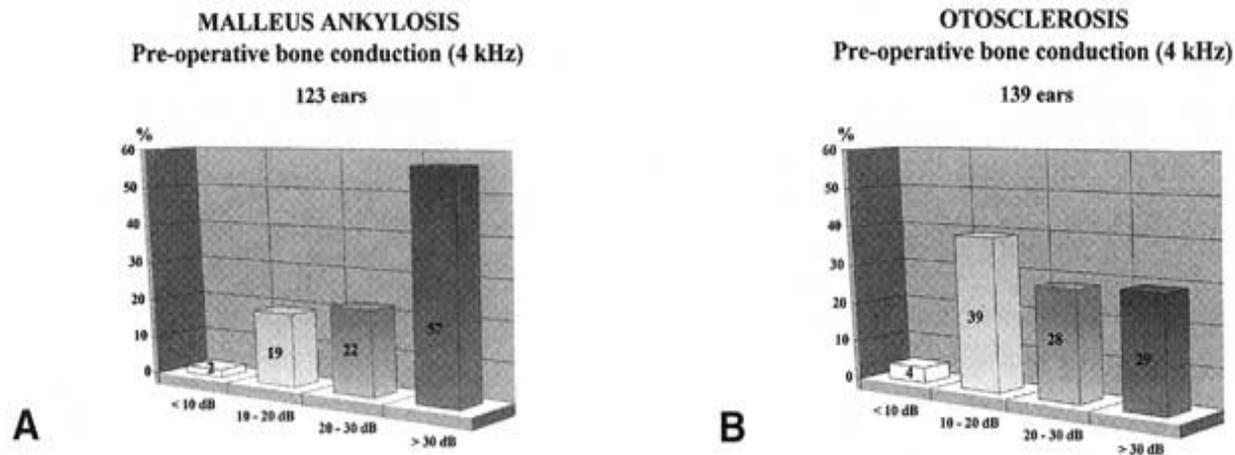


FIG. 4.
A: Malleus ankylosis group. Assessment of the preoperative bone conduction level at 4 kHz.
B: Otosclerosis group. Assessment of the preoperative bone conduction level at 4 kHz.

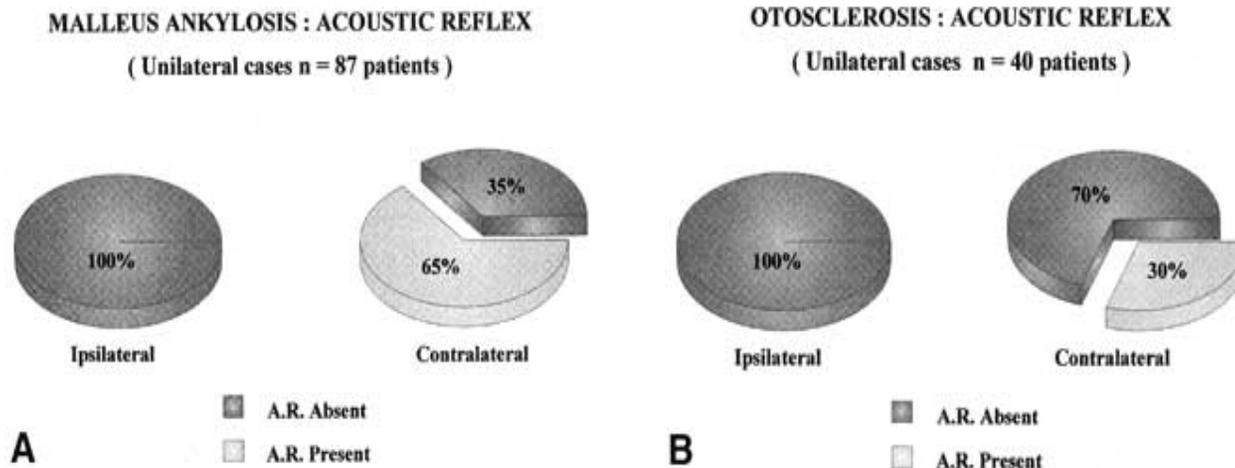


FIG. 5.
A: Malleus ankylosis group. Assessment of the preoperative acoustic reflex.
B: Otosclerosis group. Assessment of the preoperative acoustic reflex.

Tympanogram

The study of the tympanogram showed no difference between the two groups.

SURGICAL EVALUATION

This study was carried out on 112 patients with a malleus ankylosis of 123 cases operated on from January 1991 to September 1997.

It was a primary surgery in 86% of the cases and a revision surgery in 14% of the cases. In 6% of the cases, the malleus ankylosis was discovered during revision surgery for otosclerosis.

Surgical techniques

Based on the clinical findings, different surgical procedures were chosen. Ossieuloplasty was performed using an incus autograft (28%) or a partial prosthesis (incus replacement) (50%). A total prosthesis (incus and stapes replacement) was used in 12% of the cases when the stapes was also fixed (6%) or in case of revision surgery for otosclerosis (6%). Finally, in 10% of the cases, a mobilization of the incudo-malleal joint was performed. This technique is avoided because of the risk of recurrence of the ankylosis.

Description of surgical technique

The incudo-malleal joint mobility must be systematically assessed at the time of primary surgery, after the separation of the incudo-stapedial joint. Palpation with a 45° hook shows that the malleus is fixed. In case of malleus ankylosis, the incus is removed and the neck of the malleus is cut with the malleus nipper. The head of the malleus must be removed to avoid any recurrence of the ankylosis. The handle of the malleus remains mobile. Ossicular reconstruction depends on the status of the ossicular chain:

- When the stapes is present and mobile, an incus autograft or a partial prosthesis is used from the malleus handle to the stapes.
- In case of erosion of the stapes crura with a mobile footplate, a total prosthesis is placed from the malleus to the footplate.
- When the stapes is fixed, a laser stapedotomy is performed. A total prosthesis made of a pure and dense hydroxylapatite head and a 0.4-mm teflon shaft, or a malleus piston (0.4-mm teflon piston having no memory with an opening of the loop at 11 o'clock for the attachment to the handle of the malleus) is used.

This prosthesis is placed from the malleus to the stapedotomy.

A hand vein graft is used routinely for our stapedotomies.

Types of the fixation

It was assessed on primary cases (i.e., 106 ears). In most of the cases, ankylosis of the malleus was the only finding (78%). Ankylosis of the malleus and of the incus was found in 15% of the cases. Ankylosis of the incus alone was most rare (7%). An association with an otosclerotic stapes fixation was also found in 6% of these cases.

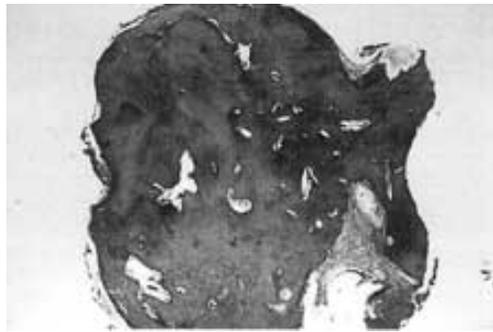


FIG. 6.
Histologic section showing bony fixation developing at the head of the malleus.

HISTOLOGIC STUDY

The histologic study was performed on 29 cases. The examination was done on the incus and on the head of the malleus to try to find the histologic nature of the ankylosis. A histologic cause was found in 15 cases (14 malleus and 1 incus). Bony fixation (exostosis) was the primary finding (12 cases) (Figs. 6 & 7). Tympanosclerosis was found in one case (Fig. 8). Two cases of otosclerosis were histologically confirmed. In one case, the ankylosis was caused by an otosclerotic bridge developed between the head of the malleus and the epitympanum. In the second case, the otosclerotic focus was present only inside the handle of the malleus. The study of these lesions using polarized light showed the characteristic aspect of the collagen.

Malleus otosclerosis: 2 cases

Case 1 was an 18-year-old woman with a unilateral mixed hearing loss of 40 dB. Her mother was operated on for otosclerosis. At the time of surgery, an otosclerotic ankylosis of the stapes was found associated with a malleus fixation. Ossiculoplasty was performed during the same procedure using a total prosthesis. The histologic study showed the presence of an otosclerotic focus limited to the handle of the malleus only, which did not appear to be responsible for the fixation of the head (Fig. 9). There were no otosclerotic foci in the incus but the incudo-malleal joint was remodeled by degenerative lesions.

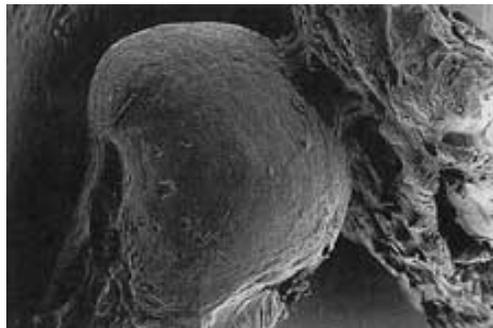


FIG. 7.

Electronic microscopy examination showing a huge outgrowth of bone fixing the head of the malleus to the tegmen.

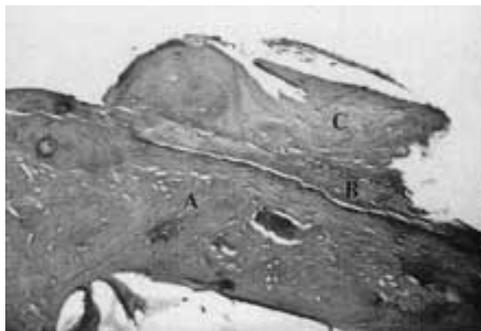


FIG. 8.

Histologic section showing tympanosclerosis, partially ossified, of the head of the malleus.

A: Ossicle (normal bone).

B: Tympanosclerotic tissue.

C: Newly formed bone in the tympanosclerosis.

Case 2 was a 45-year-old woman operated on for otosclerosis 2 years earlier with immediate failure that included persistence of a mixed hearing loss of 45 dB on the operated ear and 35 dB on the contralateral ear. An ankylosis of the head of the malleus was found at the time of revision surgery. The reconstruction was performed with a total prosthesis after the drilling of a new stapedotomy. The histologic analysis of the head of the malleus showed an otosclerotic focus responsible for the fixation of the head to the epitympanic wall (Fig. 10). The incus was normal.

Postoperative audiometric air-bone gap (0.5, 1, and 2 kHz)

Audiometric results were assessed on 102 ears checked after a 6-month period (Fig. 11). The average postoperative air-bone gap was 10 dB compared to 20 dB preoperatively.

The postoperative air-bone gap was closed to 10 dB or less in 77% of the cases.

Bone conduction average (0.5, 1, and 2 kHz)

The average postoperative bone conduction level was 20 dB for the same 102 cases. This indicates less overclosures than seen typically in otosclerosis where the average change in bone conduction is 5 dB. There was no sensorineural hearing loss in this series.

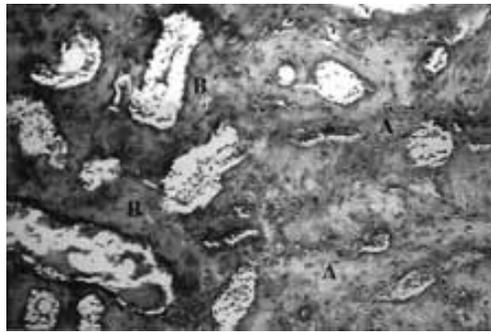


FIG. 9.
Histologic section showing an otosclerotic focus of the handle of the malleus.
A: Normal bone.
B: Otosclerotic focus (hypercellularity, basophily, and hypervascularization).

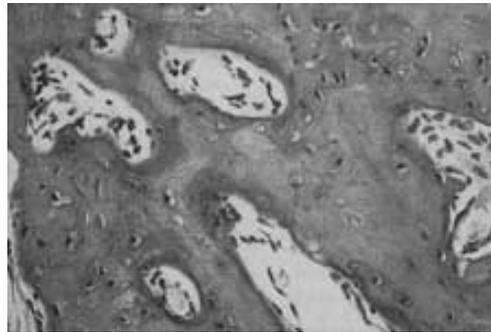


FIG. 10.
Histologic section showing an otosclerotic outgrowth of the head of the malleus. Evolution toward otosclerosis: decreasing vascularity of the cellularity and basophily

Bone conduction at 4,000 Hz

There was no improvement postoperatively at 4 kHz with preoperative and postoperative average being 40 dB.

DISCUSSION

Incudo-malleal ankylosis is an unusual pathology, which is most often diagnosed during exploratory surgery. It typically presents with conductive hearing loss with a normal tympanic membrane.

In our experience, malleus ankylosis represents 0.6% of our primary surgery cases and 3% of our cases of revision surgery for otosclerosis.

Its incidence during primary surgery for otosclerosis has been reported to be 0.4% by Sleenckx et al. (1), 1 % by Guilford (2), 1.6% by Powers et al. (3) and 1 % by Lippy et al. (4). At the time of revision surgery for otosclerosis, this incidence increases to 4.5% by Feldmann and Schuknecht (5) and 13.5% by

Sleecks (1).

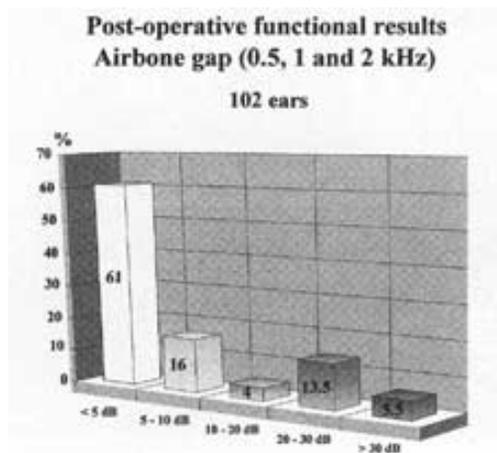


FIG. 11.
Postoperative functional results (102 ears checked after a 6-month period).

As quoted by Moon and Hahn (6), the first cases of malleus ankylosis were found in the late 19th Century. It was first described by Toynbee in 1860 who mentioned on a series of 1,013 temporal bones the possibility of deafness caused by a fixation of the malleus that he differentiated from a fixation of the stapes. He mentioned two cases of bony ankylosis of the malleus to the epitympanum, three cases of incus ankylosis and in three ears where the malleus was fixed by a ligamentous ankylosis to the epitympanic wall. In 1885, Schwarze (6) reported the possibility of isolated incudo-malleal joint ankylosis, without concomitant fixation of the stapes. In 1909, Politzer (6) reported the possibility of malleus ankylosis to the tegmen and ascribed it to a catarrhal adhesive process of the middle ear. Since then, several authors (2,7-11) stressed the necessity of awareness of malleus fixation during tympanoplasty and otosclerosis surgery.

In 1956, House (12) reported two cases of congenital fixation of the incus and one case of fixation of the malleus associated with stapes ankylosis. In 1962, Andersen et al. (13) described three cases of fixation of the malleus by an osseous connection to the posterior wall of the external ear canal. In 1963, Guilford (2) mentioned 16 cases of malleus ankylosis, in 13 of which the stapes was also fixed. In 1965, Goodhill (14) presented four cases of malleus ankylosis and introduced the notion of The Fixed Malleus Syndrome. In 1986, Martin (11) reported a series of 62 cases of malleus fixation.

Etiopathogenesis

Numerous etiopathogenic hypotheses have been suggested in the literature. These epitympanic fixations of the ossicular bones can occur without any history of otitis and seem more frequent in ears with otosclerotic stapes ankylosis than in the general population. They can occur as a consequence of head trauma but also without any apparent cause.

Congenital abnormalities have a great part in these etiopathogenic hypotheses because of a lack of full development of the epitympanic space. Altmann (15) observed in certain patients that the head of the malleus could be located close to the tegmen tympani involving chronic

mechanical irritations that may be at the origin of the creation of osseous connections responsible for a subsequent fixation. According to Sleecks et al. (1), an incomplete pneumatization of the middle ear in the fetus can be a factor of predisposition to an epitympanic fixation of the ossicular bones. For Sleecks et al., the fundamental cause of malleus fixation seems to be a lack of development of the epitympanic space leaving the head of the malleus and the head of the incus in close contact with the tegmen. This abnormality, associated with a lack of resorption of the Meckel cartilage, can create situations similar to that described by Altmann (15) on a series of 45 temporal bones in which a bony strip goes from the epitympanum to the head of the malleus. According to Ritter (16), a lack of resorption of the embryonal mesenchyme at the 7th month of fetal life can create a persistent bridge of mesenchyme likely to explain the persistence of the ossifications.

In a study on a series of temporal bones, Davies (17) frequently found bony projections, sometimes thick, extending from the lateral part of the anterior upper attic wall to the malleus. These projections could, in certain cases, come into contact with the head of the malleus.

According to Moon and Hahn (6), primary malleus ankylosis appears to result from ossification of the superior or anterior tympano-malleolar ligament. This condition occurs in elderly patients and is combined with varying degrees of sensorineural presbycusis. The association with an incus fixation, which is rare, may be explained by a concomitant arthritis of the incudo-malleolar joint. For Moon and Hahn, these situations are not caused by inflammatory changes nor by otosclerosis but may correspond to aging related changes.

In our study, a trauma history was found in 5% of the cases.

In their study on posttraumatic conductive hearing loss, Does and Bottema (18) described two cases of fixation of the malleus having required the section of the malleal neck and appearing to be the consequence of a fracture line extending through the tegmen. For Sleecks et al. (1), these posttraumatic fixations can only be seen in anatomically predisposed ears.

In our study, the histologic examination shows a fixation of the malleus by tympanosclerosis developed from the malleus head, without fixation of the ligaments (Fig. 8). Otitis history is found in 31 % of the cases and the tympanic membrane is abnormal in 21 % of the cases (tympanosclerotic plaques, retraction pockets, or scarred eardrum).

Malleus ankylosis can also occur after a history of chronic middle ear inflammation. Ojala (19) and Hilding (10) reported postinflammatory malleus fixations and attributed the fixation to the new bone formation from the surrounding epitympanic wall. According to Davies (17), tympanosclerosis may infiltrate the tensor tympani tendon. He advises sectioning the tendon routinely in cases of persistent malleus ankylosis after removal of the tympanosclerotic plaques. During the process of middle ear infection, the bony structures are particularly affected and can become remodeled. New bone may form because of osteoblast activity. This osteoid bone may then be gradually modified with an overall increase in size of the ossicles.

Incudo-malleolar ankylosis and otosclerosis

In our study, the presence of indisputable otosclerosis of the malleus was seen in one case and of the incus in another, and was thought to be responsible for the fixation of the ossicle in the second case. To our knowledge, no previous case of otosclerotic fixation of the malleus or incus has been published (Fig. 9).

In the 1960s, Guilford (2) described five cases (1 %) of malleus ankylosis on a series of 414 stapedectomies, and 16 cases of malleus fixation among which 13 cases were associated with a stapedial otosclerosis. In 1967, Guilford and Anson (20), on a series of 3991 stapedectomies, reported 30 patients with malleus ankylosis (an incidence close to 1%). Powers et al. (3) described 35 cases of malleus ankylosis, 27 of which were found to be combined with a stapedial otosclerosis. According to Goodhill (14), malleus ankylosis can be

associated with otosclerosis, but seems to be an isolated lesion.

Altmann (15) reported that it was not rare in case of stapedial otosclerosis to find a subluxation of the footplate in the vestibule with a twisting of the malleus and incus with subsequent arthritic changes in the joints. In 1960, Dietzel (21) reported on a series of 55 temporal bones with stapedial otosclerosis, 10 cases in which the head of the malleus was in an abnormally high position, in close contact with the tegmen. Davies (17) does not seem to agree with that because, on a series of 27 temporal bones with stapedial otosclerosis, he found 1 temporal bone when there was evidence of disorganization of the incudo-malleolar articulation and 8 temporal bones revealing obvious displacement of the footplate out of the oval window.

For Sleecks et al. (1), an abnormally high position of the head of the malleus is rarely found in nonotosclerotic ears; they agree with Altmann (15) for whom the high incidence of malleus ankylosis in otosclerotic stapedial fixations would occur as a result of "leverage" by the otosclerotic foci. Sleecks et al. (1) described radiographic findings suggesting an association of incudo-malleolar ankylosis and ossifications of the tympano malleolar ligaments with the tendency to exostosis, hyperostosis frontalis interna, and calcifications of the intracranial ligaments in patients with advanced otosclerosis. Sleecks et al. noted that ossicular bone is biochemically (22) and histologically (23) similar to the otic capsule, both being enchondral bone and having cartilaginous rests, and would therefore be expected to have similar pathologies.

Given the association of malleus ankylosis with stapedial otosclerosis, one might think that otosclerosis was the cause of the malleus fixation. This has yet to be demonstrated in spite of many histologic studies. In 1940, Covell (23) examined 46 temporal bones of patients showing stapedial otosclerosis and found typical otosclerotic lesions located inside the head of the malleus in one pair of temporal bones, but without any evidence of fixation of the malleus. In 1967, Powers et al. (3) observed 35 patients operated on for malleus ankylosis combined in 27 cases to stapedial otosclerosis. The fixations were related to exostosis developed from the head of the malleus, toward the anterior wall of the epitympanum. Histologically, the bony structures were normal, without any inflammatory or otosclerotic lesions. This histologic findings agree with the findings of this study (Fig. 7): 15 cases.

In 1974, Belal and Stewart (24) described an otosclerotic focus inside the incus (long process) and, more recently, Schuknecht (25) described a focus developed inside the incus and another one in the head of the malleus. Those three cases were found on a temporal bone and are not associated with any fixation of the incus or the malleus.

Preoperative diagnosis

In our series, the preoperative audiometrical evaluation carried out in comparing the two groups of patients gave evidence of a certain number of differences between the two pathologies. In malleus ankylosis, the hearing impairment appeared to be more unilateral and characterized by small air-bone gaps, associated with a fairly marked sensorineural hearing loss in the frequencies 0.5, 1, and 2 kHz, and particularly distinct in the 4 kHz. In otosclerosis, the hearing impairment was most often bilateral with a larger preoperative air-bone gap and a sensorineural hearing loss that was less frequent and less severe.

The study of the acoustic reflex tested contralaterally in unilateral impairments showed an interesting difference: in the case of unilateral malleus ankylosis (which was the most frequent) the acoustic reflex was present in the opposite side (normal ear) in most of the cases, whereas in otosclerosis the acoustic reflex appeared to be absent, even in the absence of hearing impairment.

According to our findings, it seems possible to suspect an incudo-malleolar fixation on the audiogram when the following features are present :

- Unilateral mixed hearing loss, usually nonprogressive

- Small air-bone gap, predominantly in the low frequencies
- Association with a sensorineural impairment in the high tones
- Acoustic reflex absent on the impaired ear but present on the contralateral ear.

In our experience, the last feature is particularly important. Tuning fork test is systematically performed to confirm the conductive component and to allow the decision for surgery.

In 1896, Dench (6) first described the normal movements of the malleus handle observed with the Siegle pneumatic speculum and described instances of restricted movements. Several authors (1,6,14) mentioned the interest of the Siegle otoscope to look for absence of movement of the manubrium, suspected in tympanosclerosis, congenital malformations, and most commonly if the patient has a history of otosclerosis surgery. Computed tomography (CT) scan can confirm the diagnosis, but the diagnosis is usually made at the time of surgery when palpating the incudo-malleal joint as it is done systematically after the incudo-stapedial joint separation.

The audiometric features of malleus ankylosis have been studied by numerous authors (1,6,11,26), but no unique patterns have been recognized. The tests may reflect otosclerosis demonstrating a hearing loss with significant retention of word recognition ability. In Moon and Hahn's study (6), speech discrimination and tympanometry were variable. In 15% of the cases reported, there was abnormally high tympanic membrane compliance. Moon and Hahn theorized that the tympanic membrane tends to become atrophic and more flaccid while continuously functioning against a fixed malleus handle. He also reported the absence of acoustic reflex in most cases, many of which returned to normal after repair. Moon and Hahn found small air-bone gaps rarely exceeding 30 dB with an average of 10 to 15 dB, associated most of the time with a marked sensorineural impairment. Goodhill (14) also found a small air-bone gap predominant in the low tones, associated with a sensorineural impairment, but with a tuning fork clearly lateralized toward the impaired ear. For Davies (17), the gap usually averaged less than 24 dB. Ritter (16) stressed the importance of tuning fork test to confirm a small conductive impairment in this type of pathology characterized by a sensorineural hearing loss on the higher frequencies. Powers et al. (3) also reported that isolated fixation of the malleus rarely leads to an air-bone gap greater than 30 dB.

In Sneeck et al.'s study (1), a fairly constant feature in cases of primary epitympanic ossicular fixation was an elevation of the bone conduction thresholds in the middle and high frequency range. Numerous studies regarding the transmission of sounds under different artificial conditions (26,27) demonstrated that fixation spots of the ossicular chain at any other place than the stapes footplate could produce a clinical picture close to the one of otosclerosis (pseudo-otosclerosis) of Goodhill (14) and can have various audiometrical expressions, including a large air-bone gap predominantly in the low frequencies often associated with a certain degree of sensorineural hearing loss in the high frequencies, a pure sensorineural hearing loss, or a small air-bone gap. Elpern et al. (26) studied experimentally on temporal bones the influence of nonotosclerotic ossicular fixations, namely on the bone conduction, creating artificially fixations in five different places: malleus, incudo-malleal articulation, incus, incudo-stapedial articulation, and stapedius tendon. Malleus and incus ankylosis lead to a hearing loss significantly greater than when the fixation involves other places and are close to the one found by Andersen et al. (27), after experimental fixation of the anterior crus of the stapes. For these authors, a significant difference does not exist experimentally between a malleus fixation and a stapes fixation.

Surgical techniques

In case of combined stapedial otosclerosis, we use during the same surgical step a total prosthesis as recommended by Sneeck et al. (1), Feldman (5), and Sheehy and Powers (28), with stapedotomy and vein graft interposition. Some authors (4,6) prefer to perform the operation in two steps: first the stapedectomy only and second the management of the malleus fixation. In our experience, the management of both

fixations in one step allows good functional results without particular risk of postoperative sensorineural hearing loss.

Simple mobilization of the incudo-malleolar articulation has been abandoned by most considering the risk of recurrence of fixation (3,11). More often, the incus and the head of the malleus are removed after sectioning the malleus neck with the malleus nipper. Reconstruction is performed using an incus or head of malleus autograft or a partial prosthesis.

RESULTS

In our series, a postoperative air-bone gap smaller than 10 dB was obtained in 77% of cases.

A total prosthesis was used in 15 stapedotomy cases (12%) as follows:

- Primary surgery-malleus ankylosis and otosclerosis of the stapes: six cases
- Revision surgery for otosclerosis-malleus ankylosis: eight cases
- Revision surgery for tympanoplasty-nonotosclerotic stapes fixation: one case.

Postoperative results appear to be satisfactory with a closure of the air-bone gap within 10 dB obtained in 12 cases. In two cases, the postoperative air-bone gap is closed between 10 and 20 dB and between 20 and 30 dB in one case. There was no sensorineural hearing loss.

These results confirm the possibility of managing both pathologies in a single surgical step.

All authors insist on the improvement provided by surgery, in spite of the small air-bone gap (1,3,6,11,14). Goodhill (14) reported the improvement obtained in speech audiometry as shown in our study. Powers et al. (3), on a series of 35 cases, obtained closure of the airbone gap to 10 dB in 87% of cases, with no cases of sensorineural hearing loss using a total prosthesis in a single stage. Sheehy and Powers (28) reported their results after having used a total prosthesis on 82 otosclerotic ears, among which 26 showed a malleus or incus ankylosis. They obtained a closure of the air-bone gap up to 10 dB in 60% of the cases and up to 20 dB in 84% of the cases, and reported three postoperative sensorineural hearing losses of 20 dB or more. Similar results were reported by Lippy (4) who never uses total prostheses.

CONCLUSIONS

Incudo-malleolar ankylosis remains an unusual pathology but should be systematically assessed during surgery and preferably after separation of the incudo-stapedial joint. A preoperative diagnosis is difficult to ascertain; some audiometrical features allow suspicion. The postoperative audiometrical results presented in this study appear to be satisfactory and are confirming the interest of surgery, even if the preoperative air-bone gap is often small. The surgical technique depends on the type of pathology encountered. However, in case of associated stapedial fixation, a stapedotomy with vein graft interposition and reconstruction with a total prosthesis will be performed during a same step. Several etiologies can be the cause of malleus ankylosis and have previously been described in the literature. Otosclerosis can be responsible for ankylosis as seems to be confirmed by the two cases presented in this study.

REFERENCES

1. Sleetcks JP, Shea JJ, Pitzer FJ. Epitympanic ossicular fixation. *Arch Otolaryngol* 1967;85:63-75.
2. Guilford FR. Panel on footplate pathology. Technique and prognosis. *Arch Otolaryngol* 1963;78:520-38.
3. Powers WH, Sheehy JL, House H. The fixed malleus head. *Arch Otolaryngol* 1967;85:73-7.
4. Lippy WH, Schuring AG, Ziv M. Stapedectomy for otosclerosis with malleus fixation. *Arch Otolaryngol* 1978;104:388-9.
5. Feldman BA, Schuknecht HF. Experiences with revision stapedectomy procedures. *Laryngoscope* 1970;80(8):1281-91.
6. Moon CN, Hahn MJ. Primary malleus fixation: diagnosis and treatment. *Laryngoscope* 1981;91:1298-307.
7. Goodhill V. Pseudo-otosclerosis. *Laryngoscope* 1960;70:122-8.
8. House H. Early and late complications of stapes surgery. *Arch Otolaryngol* 1963;78:606-13.
9. Proctor B. The development of the middle ear spaces and their surgical significance. *J Laryng* 1964;78:631-648.
10. Hilding DA. Postinflammatory fixation of the malleus. *Arch Otolaryngol* 1965;81:17-9.t?
11. Martin CH, Martin H, Prades JM, et al. Ankylose de la tête du marteau. Etiologie. Indications thérapeutiques et résultats. *Ann Otol Laryngol (Paris)* 1986;105:19-25.
12. House H. Diagnostic aspects of congenital ossicular fixation. *Trans Am Acad Ophthalmol Otolaryngol* 1956;60:787-90.
13. Andersen HA, Jepsen O, Ratjen E. Ossicular chain defects. *Acta Otolaryngol* 1962;54:393-402.
14. Goodhill V. The fixed malleus syndrome. *Trans Am Acad Ophthalmol Otolaryngol* 1966;70:370-80.
15. Altmann E. The finer structure of the auditory ossicles in otosclerosis. *Arch Otolaryngol* 1965;82:569-74.
16. Ritter FN. The histopathology of the congenital fixed malleus syndrome. *Laryngoscope* 1971;81:1304-13.
17. Davies DG. Malleus fixation. *J Laryngol Otol* 1969;82:331-5 1.
18. Does IES, Bottema T. Post-traumatic conductive hearing loss. *Arch Otolaryngol* 1965;82:331-9.
19. Ojala L. Pathogenesis and histopathology of chronic adhesive otitis. *Arch Otolaryngol* 1953;57:378-401.
20. Guilford FR, Anson BJ. Osseous fixation of the malleus. *Trans Am Acad Ophthalmol Otolaryngol* 1967;71:398-407.

21. Dietzel K. Befunde an den Gehörknöchelchen und den Mittelohrmuskeln bei Otosklerotischer Steigbügelgelfixation. *Arch Ohr Nase Kehlkopfheilk* 1960;176:655-9.
22. Maurer H. Vergleichende biochemische Knochenuntersuchungen bei der Otosklerose. *Ann Univ St?aarav* 1962;9:87-95.
23. Covell WP. The ossicles in otosclerosis. *Acta Otolaryngol* 1940;28(3):263-76.
24. Belal A, Stewart TJ. Pathological changes in the middle ear joints. *Ann Otol Rhinol Laryngol* 1974;83:159-67.
25. Schuknecht HF. *Pathology of the Ear 2nd ed.* Philadelphia: Lea and Febiger, 1993.
26. Elpern BS, Greisen O, Andersen HC. Experimental studies on sound transmission in the human ear. *Acta Otolaryngol (Stockh)* 1965;60:223-9.
27. Andersen HC, Hansen CC, Neergaard EB. Experimental studies on sound transmission in the human ear. *Acta Otolaryngol (Stockh)* 1963;56:307-17.
28. Sheehy JL, Powers WH. Incus replacement prosthesis in otosclerosis surgery. *Arch Otolaryngol* 1969;89:393-8.