Clinical and audiological aspects of stapes surgery otosclerosis

de Bruijn, A.J.G.

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Chapter 1

General Introduction.
1. Otosclerosis

1.1 Definition
Otosclerosis is a term used to describe a localised disease of the bone derived from the otic capsule and characterised by alternating phases of bone resorption and formation. Mature lamellar bone is removed by osteoclasts and replaced by osteoblasts into bone of a greater thickness, cellularity and vascularity. The characteristic lesion is a deposit of new bone with a different fibrillar and cellular pattern which is laid down at certain sites in the temporal bone. The site of predilection of otosclerotic foci is the region of the otic capsule, located between the cochlea and vestibule and just anterior to the footplate of the stapes. This region is associated with the globuli interossei or so-called embryonic rests. The term “otosclerosis”, introduced by Politzer in 1894, refers to the final inactive stage of the process where the bone is sclerotic or hardened. The term “otospongiosis”, first used by Siebenmann in 1912, refers to the active and vascular stage of the process and is more accurate from a pathological point of view as it indicates that an active lesion may be present. The term “otospongiosis” was initially widely used in Europe. However, the term “otosclerosis” was used in the UK and in North America and is at present adopted in the English language world literature to define the pathological changes.

1.2 History
It has been credited to Vasalva who, in 1741, has been the first to report a description of ankylosis of the stapes revealed during autopsy on the body of a patient who was believed to be deaf. In his textbook of 1868, Toynbee confirmed the link between unilateral deafness and ankylosis of the stapes in his pioneering studies of the ear when he found 136 specimens of ankylosis of the stapes footplate out of a total 1149 dissections of diseased ears. Politzer gave the first account of the histopathology of the condition in 1894. In his temporal bone sections through the stapes and oval window niche and adjacent labyrinthine wall, he showed that clinical otosclerosis did not present a diseased condition of the mucosa of the tympanic cavity, as had been previously held, but was a primary disease of the labyrinthine capsule with circumscribed bony deposits and tissue changes, leading to a progressive ankylosis of the stapes in the oval window niche and a progressive conductive deafness. Politzer's work was of fundamental importance as he demonstrated, for the first time, that the stapedial ankylosis was not secondary to “chronic middle ear catarrh”, but was the result of a primary disease of the labyrinthine capsule.

As Politzer, Siebenmann did also a lot of work on temporal bone sections, which led him to realise that the underlying pathological condition of stapes ankylosis is not a sclerosis but a growth process of the labyrinthine bone. Therefore, instead of otosclerosis, he proposed the name otospongiosis in 1912, which referred to the active and vascular stage of the process. However, it was not until 30 years later, when the fenestration operations were performed on a wider scale allowing direct inspection of the oval window in the living patient, that the concept of a “chronic catarrhal condition”, causing secondary fixation of the stapes footplate, was finally abandoned.
1.3 Aetiology

Many theories about the aetiology have been postulated. However, despite intensive research, the true nature about the origin of otosclerosis remains unclear. The theories that are currently considered as relevant, are concerned to more than one cause but it is not fully understood how they are related with each other. Several of these aetiological theories are related to the presence of the numerous embryonic cartilaginous rests (globuli interossei) scattered through the enchondral layer of the otic capsule.

Causse\(^8,9\), Chevance\(^10\) and their co-workers suggested that an auto-immune response forms a trigger for otospongiotic changes by enzymatic processes in these embryonic rests of cartilage. According to this theory, also named the enzymatic concept, a disequilibrium between certain enzymes and anti-enzymes in the microfoci of otosclerosis gives rise to variable clinical features. Conductive hearing loss occurs when the focus has a localisation in the region of the stapes footplate. When proteolytic enzymes reach the inner ear, sensorineural hearing loss may develop, and even vertigo may occur.

Causative factors from auto-immune responses are supported by studies which find elevated antibody levels to type II collagen (present in the embryonic cartilage rests) in patients with otosclerosis.\(^11-13\) Elevated levels of antibodies to type II collagen were also found in perilymfe.\(^14\)

Another indication for an auto-immune process being a factor in the aetiology of otosclerosis, is that certain human leucocyte antigen (HLA) determinants are more frequently found in patients with otosclerosis. The HLA is that region in the chromosomal system which plays an important role in the immunological response. It is well known that an association exists between the presence of certain HLA genotypes and certain autoimmune diseases. Several studies have shown different HLA antigens in a higher or lower frequency in patients with otosclerosis compared to normal individuals.\(^15-18\) However, despite these findings, there are also studies showing no relation between otosclerosis and HLA antigens.\(^19-21\)

A genetic component in the aetiology has been found by several studies, and, at present time, there is more or less consensus about the way otosclerosis is inherited; autosomal dominant with incomplete penetrance.\(^22-24\) At present, efforts are made with genetic linkage to find the gene on which otosclerosis is located. Tomek et al.\(^25\) studied a multi generational family and completed a genetic analysis. The results indicate strong evidence that chromosome 15q is involved in the development of otosclerosis.

Several studies found a relation between the onset or progression of conductive hearing loss and pregnancy.\(^26,27\) Therefore, endocrine factors have been suggested to play a role in the acceleration of the pathological changes and in the preponderance of females over males in clinical otosclerosis.

A viral participation in the aetiology of otosclerosis has been suggested by several investigators who found a possible relationship between prior infection with the measles virus and subsequently the development of clinical otosclerosis.\(^28-31\) With the more recently introduced, sensitive technique of polymerase chain reaction (PCR), Niedermeyer and Arnold\(^30\) found evidence for viral measles RNA sequences in active otosclerotic foci.
Furthermore, IgG anti-measles virus antibodies were detected in the perilymph of otosclerotic subjects supporting the hypothesis that a local immune response within the inner ear would be provoked by these viruses.

1.4 Epidemiology
In describing the prevalence of otosclerosis a distinction has to be made between clinical (with symptoms) and non-clinical otosclerosis, or histological otosclerosis. The latter form is a common condition in white adults and occurs in about 5 to 12% \(^32\) Clinical otosclerosis is less common than the asymptomatic histologic form. Several studies are undertaken in the past to obtain prevalence and incidence figures. Altmann et al. \(^37\) combined the data of Engström \(^38\) and Guild \(^32\) which showed that 6 out of 601 white individuals of 10 years and older whose temporal bones had been obtained at autopsy had stapedial ankylosis from otosclerosis. This prevalence of stapes fixation of 1% has generally been accepted as the prevalence of clinical otosclerosis among those of white European population. \(^39\) Hinchcliffe \(^40\) found a prevalence of 1.1% on the basis of otological and audiometric examination in 2 random samples of the British population. More recently, a large population study of adults in the UK reported a prevalence for presumptive clinical otosclerosis of 2.1%. \(^41\) Presumptive clinical otosclerosis in this study was defined as an ear with a normal appearance of the ear drum and with an air-bone gap (ABG) of 15 dB or greater over 0.5, 1, and 2 kHz. Several other prevalence studies showed lower figures. Gristwood and Venables \(^39\) found an overall prevalence of 4.70/1000 in the age category 30-49 years and a figure of 6.89/1000 in the age category 50-69 years among whites in South Australia. These prevalence figures are in agreement with the prevalence figures among Caucasoids in the USA found by Pearson et al. \(^42\) who reported a prevalence of 3.44/1000 and 7.70/1000 in the age groups of 30-49 and 50-69 years, respectively. Recently, Sakihara and Parving \(^43\) found an overall prevalence of 1.41/1000 in Denmark with an increase in the prevalence as a function of age from 0.22/1000 to 3.53/1000 in the elderly. However, it should be noted that these prevalence figures must be considered as underestimates of the true prevalence, as not all clinical otosclerosis in the area was included.

In clinical practice, otosclerosis is seen more often in women than men, and a sex ratio of 2:1 has been noted by many authorities. \(^39,41,42\) It has been suggested that the prevalence is probably the same in both sexes, although, due to hormonal influences, otosclerosis is likely to advance more rapidly in women. This is confirmed in histological reports where bilateral otosclerosis is more frequent in women and in clinical population reports where the incidence was similar but women were three times more likely to have an ABG of 30 dB or greater. \(^34,41\)

There is general agreement that the prevalence of otosclerosis varies enormously in different racial groups and populations. Otosclerosis is most frequently encountered in the Caucasian races. It appears to be uncommon among Mongoloid races \(^44,45\) and extremely rare in Negroid races. \(^37,46\)
1.5 Clinical features

The typical clinical features of otosclerosis are gradually increasing hearing losses at both sites, most frequently occurring between the third and fifth decade, and often associated with the presence of paracusis and tinnitus. Although usually both ears are affected by otosclerosis, often there is an asymmetry in hearing loss with one ear showing a greater conductive hearing impairment, and this ratio is usually maintained. Unilateral otosclerosis occurs in approximately 15 to 30% of patients.\(^{47}\) The deafness can remain confined to one ear, or the second ear may become affected later. Tinnitus is a common symptom which may disappear spontaneously. However, in many cases it continues unabated and may become louder as the hearing loss progresses. The origin of tinnitus is not clear but it may be the result of cochlear degeneration or an abnormal degree of vascularity within the labyrinthine capsule.\(^{47}\) Vertigo is not a prominent feature although some patients may experience a slight and transitory giddiness. Cause suggested that they are possibly the result of the action of toxic enzymes, which are liberated by the lesion, on the vestibular labyrinth.\(^{8,9}\)

The typical appearance during otoscopic examination is that of an intact ear drum. The "flamingo flush", or "Schwartz sign" is the result of vascular bone on the promontory, or prominent blood vessels in the submucosal layer of the mucous membrane of the promontory. It is an uncommon phenomenon, but when it is seen, it may indicate active disease which might progress rapidly.\(^{48}\)

Tuning fork tests are of help in establishing the type of hearing loss. However, audiometric testing is the most important method for hearing evaluation. Basic audiologic tests include the determination of pure tone thresholds for air-conduction (AC) and bone-conduction (BC), the assessment of the speech reception threshold (SRT) and the maximum speech discrimination score (MSDS) for a list of phonetically balanced words. The hearing thresholds measured with pure-tone audiometry depends on the stage of the otosclerotic changes. In the earliest stages, stiffness of the annular ligament produces a low frequency conductive hearing loss with normal BC thresholds. The AC curve shows a stiffness tilt sloping upward to the higher frequencies, and there is a minor ABG in the lowest frequencies. As stapedial fixation develops the ABG becomes more pronounced, the AC being elevated over all frequencies to produce a flat curve at about 50 to 60 dB. The BC curve no longer accurately depicts the level of cochlear reserve but suffers a mechanical distortion known as the Carhart notch, which was first reported by Carhart in 1950.\(^{49}\) Gatehouse and Browning\(^{50}\) define this phenomenon as "an alteration in BC thresholds due to a decrease in the external and middle ear components of BC transmission leading to a depression of BC thresholds". The depression of BC thresholds is maximal at 2 kHz, and has therefore conventionally been labelled as "Carhart notch". However, because the phenomenon occurs over the whole frequency range from 0.5 to 2 kHz, Gatehouse and Browning\(^{50}\) consider the term "Carhart effect" as more appropriate.

In many cases of stapedial otosclerosis, a sensorineural hearing loss (SNHL) increases gradually, develops coincidentally with, or develops after the conductive hearing loss. A mixed or combined hearing impairment develops with elevation of both AC and BC thresholds, the AC curve more than the BC, so that the BC curve of the patient lies between the AC curve
and the normal threshold level. The AC curve represents the sum of the hearing losses produced independent by the stapedial fixation and by the cochlear lesion.

Acoustic impedance measurements using an electroacoustic impedance meter can provide valuable clinical information including assessment of middle ear pressure, shape and height of the tympanogram, and middle ear muscle reflexes. The typical shape of a tympanogram in an ear with otosclerosis is that of a normal ear type A configuration but with a reduced maximum compliance within the normal middle ear pressures of between \(-100 \text{ mmH}_2\text{O}\) and \(+50 \text{ mmH}_2\text{O}\) (type As). Stapedial reflex measurements show the absence of ipsilateral and contralateral reflexes for measurements with the probe in the otosclerotic ear.\(^{51}\)

High resolution computerized tomography (CT-scan) has proved its value by establishing the so-called cochlear otosclerosis.\(^{52,53}\) Fenestral otosclerosis is essentially a clinical diagnosis and only when it is severe, with complete obliteration of the oval window niche, it may be visible on CT-scans.

The differential diagnosis includes conditions having a normal or near normal tympanic membrane in association with a conductive or mixed hearing impairment. This includes congenital stapedial fixation, stapedial fixation from tympanosclerosis, osteogenesis imperfecta and Paget's disease, traumatic ossicular discontinuity, the fixed malleus-incus syndrome and congenital cholesteatoma.\(^{48}\)

### 1.6 Non-surgical treatment of otosclerosis

#### 1.6.1 Hearing aids

Although nowadays the choice of treatment in cases of otosclerotic deafness is stapes surgery, there are circumstances that surgery is not desirable or possible and sometimes patients do not give informed consent for surgical treatment. In those cases the prescription of a hearing aid forms a good alternative. In the majority of patients with conductive or mixed hearing losses due to otosclerosis, the modern hearing aid, currently also available with digital technique, gives good results. The profoundly deaf may be assisted by high-powered bone conductors or vibrotactile devices that provide additional clues for speech reading. In those cases in which normal rehabilitation with hearing aids is not possible due to recurrent otitis externa, and also in the rare cases of bilateral fenestration cavities, a bone anchored hearing aid (BAHA) can be considered.\(^{54}\)

Although a more natural hearing can be obtained after stapes surgery, the advantages of surgery always have to be related to the disadvantages, and especially in bilateral cases the risk of severe SNHL, which can occur many years after surgery, has to be taken into consideration.

#### 1.6.2 Treatment with sodium fluoride

The thought that sodium fluoride, given in moderate to high dosis, might influence favourably the otosclerotic bone lesions, was first presented by Shambaugh and Scott in 1964.\(^{55}\) The idea was that sodium fluoride might promote the change of actively expanding otosclerotic lesions to more dense, inactive lesions. This stimulation of the natural tendency of otosclerotic lesions to become recalcified and inactive would result in a stabilisation of the
progressive sensorineural deafness, a reduction of tinnitus and an improvement of mild vestibular symptoms.\textsuperscript{55}

Several concepts are published in the last 35 years about the mode of action of sodium fluoride. In animal experimental studies, Petrovic and Shambaugh\textsuperscript{56} demonstrated the influence of sodium fluoride on bone metabolism, reducing osteoclastic bone resorption as its major effect, and at the same time promoting osteoblastic bone formation. Bretlau et al.\textsuperscript{57} reported on an evaluation of sodium fluoride treatment using scanning transmission electron microscopy (STEM) together with an energy dispersive x-ray element analyser. The results showed that using the calcium/phosphorus ratio as an indication for bone maturity, sodium fluoride could stabilise otosclerotic lesions, particularly the spongiotic type with unstable mineralisation, in retaining calcium relative to phosphorus. The results supported the view that sodium fluoride promotes recalcification and reduces the activity of otospongiotic bone.

Causse and co-workers suggested an anti-enzymatic action of sodium fluoride.\textsuperscript{58,59} When the balance between trypsin and $\alpha_1$-antitrypsin is disturbed in favour of trypsin, this results in an increase of trypsin values that damage the hair cells, leading to cochlear deterioration. It was shown that sodium fluoride can act directly on trypsin and reduces the levels of protease inhibitors, such as $\alpha_1$-antitrypsin and $\alpha_2$-macroglobulin. Causse et al.\textsuperscript{59} stated that the overall reduction of enzymatic rates by inhibition favours the stabilisation of the otosclerotic process and consequently arrest or slowing down the cochlear deterioration caused by an alteration of Corti cells attributable to active proteases. These findings were supported by other authors.\textsuperscript{60} In a clinical report on a large series of over 5000 patients treated with sodium fluoride, Causse et al.\textsuperscript{58} found that medical treatment with sodium fluoride results in a stabilisation of cochlear deterioration in the majority of cases having a progressive cochlear component. Only a few patients showed an improvement in sensorineural hearing. Furthermore, it was found that sodium fluoride seems to slow conductive hearing loss by stapedial fixation but cannot release stapedial fixation.

Despite these findings, the administration of sodium fluoride remains a matter of controversy, partly due to the unknown toxic effect of long-term medication with sodium fluoride.\textsuperscript{61} Furthermore, most studies showing favourable results are not randomised and only a few studies are carried out as double-blind randomised, placebo-controlled trials.\textsuperscript{62,63}

2. STAPES SURGERY FOR OTOSCLEROSIS

2.1 Evolution of otosclerosis surgery

2.1.1 The first stapes surgery era (1876–1900)

The historical development of surgery for conductive hearing losses due to otosclerosis is fascinating. The first attempts to correct hearing losses have to be interpreted in relation with the limited and confused knowledge the pioneers had of the physiology and pathology of the ear. There was no accurate way to measure hearing loss or to examine the ear adequately before, during, and after operation. The diseases of the ear, especially chronic otitis media and otosclerosis, were not well understood.\textsuperscript{64}
It was Professor Kessel of Jena who was one of the first otologists to use animal experimentation to gain information to guide treatment in humans in the mid 1800's. In 1876, Kessel mobilised the stapes in a young woman with inactive chronic suppurative otitis media with no drum, malleus, and incus. There was some immediate hearing improvement. Subsequently, Kessel removed the stapes in humans, with, according to him, "some improvement in hearing and no serious complications". The first mobilisations of the stapes and stapedectomy operations by Kessel were soon followed by other leading otologists of those days. Despite the initial enthusiasm and advances in stapes surgery, the inevitable happened. Because of the many failures, together with some serious complications, like meningitis, and even death, leading otologists of those days, like Politzer and Siebenmann, condemned stapes surgery for otosclerosis as useless and dangerous. Professor Kessel, nowadays contemplated as the father of stapes surgery, had fallen into disrepute and in 1900 he was publicly censured for unscrupulousness.

2.1.2 The fenestration era (1910–1960)
The fenestration era began as conditions started to improve for otologic microsurgery. The pathology of otosclerosis was better understood by the establishment of temporal bone pathology laboratories in otologic centres in Europe and the United States. Progress was made with the magnification and electrical illumination of the operation field. The term "fenestration" was used by Jenkins in 1913. He suggested that it would be better to detour the sound vibrations around the obstruction in the oval window by making an opening in the bony lateral semicircular canal and covering it with a skin flap from the ear canal. Initially, Holmgren from Sweden gave great impulse to the fenestration technique by introducing the operation microscope. He continued with the closed fenestration technique of Jenkins but he covered the fenestra with mucoperiosteum. Sourdille, a French student of Holmgren, developed the three-stage procedure named "tympano-labyrinthopexy." At the first stage a mastoidectomy was performed, at the second stage an external ear canal skin flap was created and at the third stage a fistula was made in the horizontal canal which was subsequently covered with the canal skin flap. This alteration of technique was an important improvement over the closed fenestration operation of Jenkins and Holmgren as sound could enter the perilymph space directly without the obstruction of the ear drum and bony canal wall.

It was Lempert who modified the three stage technique into a one stage technique, and who performed the operation with the electric drill rather than hammer and chisel. He published his results in 1938 in a famous paper entitled "Improvement in Hearing in Cases of Osteosclerosis: A New One Stage Surgical Technic". Later, Shambaugh Jr., the first pupil of Lempert, published his results with the fenestration operation. Soon the procedure gained world wide acceptance. The results with the Lempert modification of the fenestration operation were better, with a lasting hearing improvement to the 20 to 25 dB level in approximately 50% of the patients in expert hands.
2.1.3 The second stapes surgery era (1952 – present)

In 1952 Rosen\textsuperscript{72} published a preliminary report to describe a simple method of palpation of the stapes to determine whether it was partially or completely fixed. Rosen found that rocking palpation of a fixed stapes rendered the footplate more mobile with improvement of hearing. Sequentially, Rosen developed a procedure based on the premise that ankylosis was not necessarily permanent or irreversible. His relatively simple procedure, which resulted in better maintained hearing, could easily be followed by fenestration, should success not be achieved by stapes mobilisation. In 1953 Rosen\textsuperscript{73} published his results in a series of patients, on whom he performed mobilisation of the stapes, with improved hearing. After some time, the mobilisation technique became generally accepted and the fenestration became reserved, more or less, for those in whom mobilisation has failed.\textsuperscript{74,75}

In 1954, the first bypass procedures were begun. This type of surgery for otosclerosis was based on the concept that the otosclerotic focus of bone might be bypassed entirely, rather than being mobilised and left free to reankylose. It was Shea Jr. who rediscovered the stapedectomy procedure and reconstructed the transmission system with a prosthesis. In 1956 he did his first stapedectomy operation with insertion of a prosthesis with the same shape and size of a normal human stapes.\textsuperscript{64} This stapes replica was made out of a newly discovered material named tetrafluoroethylene or Teflon which was believed to be inert in the body. Shea used this Teflon prosthesis over a vein graft in a 54 year-old woman with extensive oval window otosclerosis. The woman’s hearing immediately improved.

Shea's contribution to otosclerosis surgery soon proved to be a start of a new revolution in the surgical treatment of otosclerosis, much as Lempert's fenestration has done many years before. Numerous modifications in the stapedectomy technique have been made over the years by numerous contributors. Furthermore the era of pistons had arrived, and soon many surgeons modified their techniques from total stapedectomy to partial stapedectomy and to stapedotomy using a wide range of different prostheses.

2.2 Current techniques of stapes surgery

Currently stapes surgery consists of the traditional stapedectomy in which the footplate is totally or near-totally removed, and stapedotomy in which a small fenestra is made through the central portion of the footplate. Another technique is to remove a part of the footplate which is also called partial stapedectomy\textsuperscript{76} or partial platinection.\textsuperscript{77} Some surgeons use a soft tissue graft, like vein, perichondrium or fascia, to cover or fill the oval window in order to limit the loss of perilymph and prevent subsequent fistula formation. Either operation can be performed under general or local anaesthesia, depending on availability and the surgeon's choice. An increasing number of surgeons prefer local anaesthesia supplemented by intravenous sedation because, with the patient awake, immediate hearing improvement can be noted intraoperatively as well as vestibular and other pathology reflected by complains of dizziness.

The standard approach for surgery is endaural. More exposure can be obtained by an endaural incision. After a tympanomeatal flap is created the middle ear can be exposed. In most cases bone from the posterior-superior scutum has to be removed, until the oval window can
be inspected with the facial nerve superiorly and the pyramidal process posteriorly. After the ankylosis of the stapes is confirmed, one of the three above mentioned techniques can be applied. Many surgeons believe that a small hole (the so called safety hole) should be placed in the centre of the footplate prior to removal of the suprastructure to avoid a negative suction effect on the vestibule if the footplate is removed with the suprastructure. Furthermore, if a stapes gusher is encountered, the preserved suprastructure can help support connective tissue over the central hole.

With regard to the indication for surgery, many surgeons feel that the ABG should be at least 15 dB and there should be a SDS of 60% or more for a good hearing improvement. Previously, there was some concern regarding the performance of stapes surgery in children. It is clear now that stapes surgery in children can be accomplished quite safely.78,79

2.3 Stapes replacement prostheses
A variety of different prostheses are available today for reconstruction, although a few surgeons prefer to use autolog material for reconstruction, like the posterior crus over a tissue sealed oval window after total removal of the footplate. All prostheses differ in size, shape and weight.80 The mass of the implant is important, because it will affect the transmission of lower and higher frequencies. Most of the prostheses consists of a piston or cylinder that projects into the oval window. The main difference in the design of stapes replacement prostheses are at the point of connection with the incus. The attachment to the incus consists of either a loop that surrounds the incus or a cup into which the lenticular process fits. Most loops are made of thin stainless steel or platinum ribbon that hooks onto the incus. Some prostheses have a ring composed of Teflon that require opening the ring prior to placement. Most prostheses are available in a variety of different lengths. Some prostheses are manufactured at a standard length and require the surgeon to trim them to the desired length. The shaft diameter of most prostheses ranges from 0.3 to 0.8 mm at the attachment of the oval window.

A stapes replacement prostheses must be composed of a biomaterial which is well tolerated by the middle ear with no reactions at both the oval window and the incus. The biomaterials currently most often used are a Teflon type polymer, stainless steel, and platinum.81 Some prostheses contains a combination of materials, like the combination of a Teflon piston shaft with a stainless steel loop in the Fisch piston or Schuknecht piston or the combination of a platinum shaft and a stainless steel loop in the McGee piston.80 If the incus is not available for reconstruction, an incus replacement prosthesis can be used like the Shea malleus handle prosthesis.82,83

3. Evaluation of hearing results

3.1 History of assessing hearing
In the days of the first stapes surgery era (1876-1900) mechanical acouometers were developed to produce controlled quantifiable stimuli in order to establish hearing in a more reliable way before and after surgery. Of these the best known were those of Itard (1821) and of Politzer
The first electrical audiometers stemmed from the invention of the telephone by Alexander Graham Bell in 1876, which opened the way for the development of sound generators with variable output intensities. Of the first electrical audiometers there were two which have attracted most attention; that of Hartmann (1878) and that of Hughes (1879). The latter was the first who made the audiometer commercially available. However, despite the technical improvements in establishing the amount of hearing loss, initially the first electrical audiometers were not well adopted in the relatively new speciality of otology which was more concerned with the more urgent and widespread problems of ear infections and their often fatal consequences.

After World War I, the invention of electronic valves was the next major advance in the development of electrical audiometers. During the late 1920s a lot of work had been conducted on BC vibrators by a variety of researchers, and from the early 1930s most audiometers were fitted with both AC and BC transducers. These modifications helped to facilitate the acceptance among otologists. From that time the basis principles, embodied in the new design of the standard clinical audiometer, have remained essentially unchanged to the present day.

At the time that the fenestration procedure was promoted by Lempert, the three frequencies of 0.5, 1, and 2 kHz were selected for hearing evaluation because they were the prime frequencies involved in hearing conversational speech. In those days, BC measurement above 2 kHz was unreliable due to technical limitations of the audiometers; therefore, higher frequencies could not be used in computing the ABG. Subsequently, when speech audiometry was developed, it was found that these pure tones correlated well with SRTs.

Later, in the 1950s the electronic valves were replaced by transistors and integrated circuits in audiometers were widely used in the 1970s. In the late 1970s the microprocessor was introduced in the audiometry devices and it has evolved to the present state of audiometry with the fully computer controlled audiometer.

### 3.2 Methods for hearing evaluation

How should hearing results be reported following stapes surgery? This is a question that every author who wants to report about hearing outcome after this type of surgery has to deal with. By reviewing the literature, a wide diversity of methods, criteria and parameters can be found used in the reports to summarise the audiologic results as well as many different statistical tests for their analysis. What method to be used depends on the primary purpose of the study. If technical factors are being evaluated, such as surgical procedure, type of prosthesis, use of a specific biomaterial, or surgical skill, closure of the ABG is probably most important. The comparison of pre- and postoperative BC, including higher frequencies, allows postoperative cochlear damage to be assessed. In addition speech discrimination tests are also good indicators of cochlear reserve. If the interest is in absolute hearing function, AC threshold levels are critical, although they will depend upon patient selection as much as technical skill. In general, pure-tone AC thresholds at speech frequencies correlate well with the SRT assessed by speech audiometry. In reporting the hearing results also other factors have to be taken into account with regard...
to the patient's perspective as the aim of stapes surgery is to reduce auditory disability. In general, the degree of disability is determined by the status of the better hearing ear. The ideal of bilateral normal hearing is not always attainable, and so, in advising patients regarding surgery for hearing gain, it is important not forget the contribution of the other ear. Smyth and Patterson assessed the requirements for patient's benefit by drafting the “Belfast Rule of Thumb”. They concluded that for significant benefit to be achieved the postoperative AC average over the speech frequencies must be close to 30 dB or the interaural difference reduced to less than 15 dB. The “Glasgow Benefit Plot” devised by Browning et al. is a valuable elaboration of the forenamed requirements. The degree of auditory disability can be assessed with the criteria of the American Medical Association.

### 3.3 Standardisation in reporting hearing results

At the time that stapes surgery was performed on a larger scale, many otologists realised that they needed generally accepted recommendations in order to report audiologic data in a more uniform way. Why is standardisation in the reporting of hearing results so important? Shea summarised the problem when he discussed 30 years of stapes surgery. He noted that "the diversity of techniques and lack of uniformity of reporting results make it difficult to agree on the best method to reconstruct the sound conducting mechanism after the stapes has been removed. Technique, materials, patient selection, type of procedure, type of prosthesis, and pathological condition are all factors that may affect results of middle ear reconstruction, but particularly the lack of standardisation in reporting hearing results has made comparison across studies difficult in the past."

Initially, the standard classification system, developed by a committee of the American Academy of Ophthalmology and Otolaryngology for surgery of chronic ear infection in 1965, was embedded for the evaluation of hearing results after stapes surgery. It required an observation period for at least 1 year and thresholds at the speech frequencies 0.5, 1, and 2 kHz were involved in computing ABG. Although it was not clearly stated in the classification system, it was a common use at that time to calculate the postoperative ABG by taking the difference between the postoperative AC and preoperative BC. This method of computing postoperative ABG was also recommended by The Committee on Nomenclature in Chronic ear Disease and the Otosclerosis Study Group in 1971. Independently from the American otological working groups, the Committee of Nomenclature of the Japan Society of Clinical Otology developed there own guidelines in 1987. According to these guidelines the average should be taken over the frequencies at 0.5, 1, and 2 kHz either by the “dividing by three method” or the “dividing by four method”. In the former method the hearing level at each frequency is counted equally, while in the latter method the hearing level at 1 kHz is counted twice to stress the importance of this frequency in speech recognition. The observation period should be at least 6 months and a successful result was obtained if one of the following was achieved: (1) an ABG closure of 20 dB or less, (2) a postoperative AC threshold within 40 dB, or (3) a gain in AC of more than 15 dB. The guidelines of the Japan Society of Clinical Otology (currently The Otological Society of Japan) did not give definite recommendations about how to establish postoperative ABG either with preop-
more recently, the Committee on Hearing and Equilibrium of the American Academy of Otolaryngology - Head and Neck Surgery\textsuperscript{97} published guidelines to evaluate hearing results after ossiculoplasty and stapes surgery. The guidelines have been established for two levels: level 1 provides for the uniform reporting of summary data and level 2 provides for the uniform reporting of raw data. The Committee encourages to report raw data from each individual case to enable more detailed analytical studies and meta-analyses in the future. With regard to level 1, the Committee recommended guidelines that had and still have an important impact in many otological societies. One of the major change in the recommendations is that the mean of thresholds at the frequencies 0.5, 1, 2, and 3 kHz should be used to form a four-frequency PTA in stead of the traditionally three-frequency PTA at 0.5, 1, and 2 kHz. Another important change is that the Committee advises to compute postoperative ABG by the method of taking the postoperative AC and BC thresholds instead of the traditionally method by taking the postoperative AC and preoperative BC. The latter method commonly results in a higher success rate after stapes surgery because ABG overclosure due to the Carhar effect is taken into account.\textsuperscript{98} In this study we will evaluate different methods for hearing evaluation for a large set of clinical data.

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