Surgical management of otosclerosis and congenital stapes ankylosis

V.E. Kisilevsky

Print: Printpartners Ipskamp
Layout: Diny Helsper

ISBN:

Surgical management of otosclerosis and congenital stapes ankylosis.
Thesis Radboud University Nijmegen Medical Centre, Nijmegen.

Surgical management of otosclerosis and congenital stapes ankylosis

Een wetenschappelijke proeve
op het gebied van de Medische Wetenschappen

PROEFSCHRIFT

ter verkrijging van de graad van doctor
aan de Radboud Universiteit Nijmegen op gezag van de rector magnificus,
prof. mr S.C.J.J. Kortmann, volgens besluit van het College van Decanen in het
openbaar te verdedigen op
dinsdag 19 januari 2010
om 13.30 uur precies

door

V.E. Kisilevsky
geboren op 27 juni 1962
te Kharkov, Ukraine

Promotores: Prof. dr. C.W.R.J. Cremers

Copromotores: Jerry J. Halik, MD, FRCS(C), Markham –Stouffville Hospital / University of Toronto. (Canada)
John A. Rutka, MD, FRCS(C), University of Toronto (Canada)

Manuscriptcommissie:
Prof. dr. J. Keunen, voorzitter
Prof. dr. E. Offeciers, Med. Inst. St. Augustinus, Belgium
Prof. dr. M.A.W. Merkx

Contents

Chapter 1

Introduction

1.1

The definition, classification, etio-pathogenesis and histopathology of otosclerosis

Introduction

Stapes surgery is described as "the most challenging and rewarding procedure in otology", that achieved a "stage of perfection" and still remains an "elegant but precarious" operation.[1-3] Advances in understanding of the nature of otosclerosis were followed by refinements in treatment that started as empiric attempts of hearing improvement and evolved into precise surgical technique employing state-of-the-art technology. Medical treatments failed to restore hearing or to prevent deterioration of hearing impairment to date. Incomplete knowledge of biological mechanisms triggering otosclerotic process limited effective prevention of its disabling clinical effects. In this chapter we will review the evolution of otosclerosis studies and summarize the current knowledge of etiopathogenesis, patho-morphology and molecular biology of the disease.

Definition

Otosclerosis is a unique form of bony dysplasia exclusively affecting human temporal bone. The pathologic process is characterized by an abnormal bone remodeling in specific sites of predilection confined to the endochondral layer of otic capsule. Once started, the otosclerotic focus is usually progressed through three consecutive histopatological phases - spongiotic, fibrotic and sclerotic bone formation. Otosclerotic involvement of the vestibulo-stapedial joint, cochlea or vestibule defines an actual pattern of clinical presentation.[4-9]

History

When did the history of otosclerosis studies begin? The credit of the first description of stapes ankylosis as a cause of deafness is traditionally given to Antonio Valsalva, who related this condition to a "dry catarrh of the middle ear". Giovanni Morgagni, Valsalva's student, edited his writings and published it in the "Treatise of the human ear" in 1740, 17 years after Valsalva's death.[10] In 1864 Von Troltsch first used the term "sclerosis" to differentiate bony stapes ankylosis from a common middle ear catarrh.[11] Joseph Toynbee in his pathologic studies of temporal bones published in 1868 noted "that those disturbances of hearing, which formerly were classed under the name of nervous deafness, are caused in the majority of cases by ankylosis of the stapes with the oval window".[12] Adam Politzer coined the term "otosclerosis" as a primary disorder of the bony labyrinthine capsule in 1893.[13] This was a fundamental step in understanding the histo-pathology of stapes ankylosis, previously seen as a result of chronic catarrhal inflammation of middle ear mucosa. Defying beliefs that stapes fixation cannot be diagnosed in living patient, Politzer applied interpretation of tuning fork tests and patients' history of progressing hearing loss to a clinical diagnosis of otosclerosis.[14,15] Hermann Schwartze and Friedrich Bezold made an important contribution to clinical recognition of this condition, describing "Schwartze sign" and "Bezolds' triad", which became a basis for clinical diagnosis of otosclerosis in the pre-audiometry era. Friedrich Siebenmanns continued studies of histopathology of progressive hearing loss and suggested the term "otospongiosis" as more correctly reflecting osseous changes in the labyrinthine capsule.[16] Another proposed amendments to terminology included "ostitis chronica metaplastica" by Manasse, "osteitis rareficans stapedio-vestibulare" by Heimann, "capsulitis labyrinthica osteomalacia" by Ferreri and "otitis chronica metaplastica circumscripta" used by Bruhl, Denker and Kahler.[17] Currently the term "otosclerosis" is universally adopted in the English language world literature.[18]

Classification

The terminology used to refer to otosclerosis is variable hence diverse aspects have been considered in the proposed classifications.

Guild[19] differentiated ***histologic*** and ***clinical*** otosclerosis. The histologic form is defined as an asymptomatic disease that can be identified only by histomorphological examination. The clinical otosclerosis is characterized by a typical history of progressive conductive hearing loss due to otosclerotic involvement of vestibular-stapedial joint leading to stapes ankylosis. In 1985 Schuknecht and Barber[20] elaborated on the histologic variants and introduced grading system based on size, extent and histologic activity of otosclerotic lesion.

Politzer[21] distinguished ***typical*** tympanic and ***severe*** form of the disease based on its clinical course. The typical form is characterized by the gradual worsening of hearing over several years. In severe form hearing impairment progresses rapidly, leading to profound deafness.

Two clinical types of otosclerosis have been described according to the audiometric classification.[22] ***Type I*** is an equivalent of **tympanic** form according to Politzer and Siebenmann and concerns the patients with mild to moderate conductive hearing loss across all tested frequencies. ***Type II*** corresponds to **cochlear** or Lermoyez form and refers to patients with severe to profound mixed or sensorineural hearing loss with down-sloping type of audiometric curve.

Cochlear otosclerosis has been recognized as a separate entity to classify the cases of histologic involvement of the cochlea without stapes fixation that presents with a clinical picture of progressive pure sensorineural hearing impairment and radiological findings of areas of demineralization encroaching cochlear endosteum, recently demonstrated by the high resolution CT of temporal bone.[23-25]

The term *"**Far-advanced**"* otosclerosis has been introduced by House and Sheehy and is used to describe cases that clinically present with severe or profound deafness and demonstrate extensive otosclerotic involvement of the temporal bone intraoperatively. The histopathologic findings usually show massive otosclerotic process completely obliterating oval window and often involving round window and cochlea.[26-32]

The term *"**Juvenile**"* otosclerosis refers to the cases of early presentation of progressive conductive hearing loss during first or second decade of life and confirmed by intraoperative findings of massive involvement of stapedial footplate leading to ankylosis in absence of congenital ear malformations [33-35].

Karosi [36] recently suggested classification of stapes ankylosis that defined ***true otosclerosis*** as an inflammatory disease caused by persisted measles infection and referred to a non-specific degenerative stapes fixation as a ***pseudo-otosclerosis*.**

The radiologic classification has been proposed when the high resolution CT of the temporal bone has been incorporated into the investigation of hearing loss.[37] ***Fenestral*** and **retro-fenestral** or ***labyrinthine*** types of otosclerosis have been described.[38-40] The fenestral type refers to hypodence lesions of demineralized bone adjacent to the oval window area and /or impinging the stapes footplate.[41, 42] The retro-fenestral or labyrinthine/cochlear type is radiologically diagnosed in the presence of lucent areas of demineralization surrounding the cochlea`, described as a "double ring" or a "fourth turn" sign.[43-46]

Prevalence and Demographics

The reported prevalence of otosclerosis varies substantially. Toynbee, in one of the earliest large series of temporal bones dissections, found 136 cases of stapes ankylosis out of 1149 temporal bones (11.8%).[47] Other authors investigated 4080 skulls of ancient Europeans and identified only 8 (0.19%) cases of otosclerosis.[48] In the study of 1452 temporal bones by Hueb[49], the incidence of otosclerosis in Caucasians was 12.8%. The prevalence reported by Declau et al.[50], however, was only 2.5% in the series of 236 temporal bone specimens. The differences in histologic definition of otosclerosis and in inclusion criteria for temporal bone specimens were possible reasons of broad variations in the reported prevalence rates.

The importance of distinguishing between histologic and clinical findings was emphasized by Guild[19] in 1944. Guild found that histologic otosclerosis was about ten times more common than its clinical manifestations. While histologic form is clinically asymptomatic and can be diagnosed only by histomorphological examination of temporal bone, clinical otosclerosis is characterised by progressive conductive hearing impairment due to the stapes fixation. Summarizing observations of several authors who studied the demographics of otosclerosis, this condition predominantly affects Caucasian population with estimated prevalence of clinical otosclerosis 0.2% to 1%, male to female ratio about 1:2 and bilateral involvement in 70-80% of patients[51-60]**.** A correct interpretation of the prevalence requires distinguishing between histopathologic and clinical studies and attention to differences in race and gender distribution in the reported series.

Etiology

The current knowledge on otosclerosis allows to state that this is a heterogeneous disease with multiple plausible etiologies.[18,61,62] Several theories were proposed to explain its origins but the exact cause remains unknown. Review of the past theories helps to appreciate the progress made in understanding etiologic factors of this condition. Thus, such prominent otologists as Bezold, Korner and Schwabach related development of otosclerosis to inflammation caused by syphilis or tuberculosis. Citelli and Lermoyez noted similarities in bone histo-pathology between otosclerosis and rickets and gout and postulated a role of these diseases in the etiology of otosclerosis Traumatic etiology was suggested in two of his case reports by Politzer and was supported by Sercer[63] and later by Polanski.[64] None of these theories however, have passed the test of time and are remembered today to acknowledge the diversity of otosclerosis studies in the past. The major putative theories at present consider genetic factors, immunologic factors and viral infection.

Genetic factors in etiology of otosclerosis

The role of heredity has been originally inferred from demographic and epidemiologic observations and more recently investigated with DNA analysis techniques. Toynbee in 1861 was among the first investigators to notice a familial pattern of hearing loss.[65]. Family history has been also demonstrated in 52% of otosclerosis patients by Bezold, in 40% by Hammerschlag and Korner, and in 35% by Siebenmann.[17]. In more modern studies the similar distribution of inheritance has been demonstrated in 36% of females and 20% of males' offspring of parents with otosclerosis[66]**.** During a pre-DNA analysis era, research of mode of inheritance based on the Mendel's laws enabled to conclude that otosclerosis is autosomal dominant disease with incomplete penetrance of 30-40%. No evidence of association has been found between otosclerosis and eyes or hair color, or blood type group[67,68] .

The systematic screening of the human genome during the past decade enabled a significant progress in more than a century-old history of investigation of the heredity of otosclerosis. In 1998 Tomek et al.[69], described the first link between clinical otosclerosis and chromosome 15q, suggesting that the region in 15q25-26 could contain an otosclerosis gene. In 2001 the second gene has been localized on chromosome 7q34-36 by Van Den Bogaert et al.[70] In 2002 an existence of at least three genes has been predicted by these authors, based on the significant genetic heterogeneity revealed in nine families with otosclerosis.[71] Shortly after in 2002 Chen et al.[72] from the same group of Van Camp and Smith described the localisation of third gene (OTSC3) on chromosome 6p21-23. The OTSC3 area has been found to overlap the HLA region, adding the evidence basis to previously reported association between otosclerosis and HLA system. In 2004 Van Den Bogaert et al.[73] reported the fifth locus for otosclerosis, OTSC5, localised on chromosome 3q22-24, while fourth locus, OTSC4, has been reserved by the Human Genome Organisation Nomenclature Committee. Two years later in 2006 Brownstein et al.[74] identified new locus on chromosome 16q21-23 which has been designated OTSC4. Description of the sixth autosomal dominant locus on 6q13-16 region named OTSC7 has been published by Thys et al. in 2007.[75]

Although six different loci have been published to date, the identified intervals include several genes and no causative genes for otosclerosis have been cloned yet. Genetic studies of otosclerosis are hindered by a rarity of monogenic families segregating otosclerosis and by incomplete mode of penetrance, which varies from 25 to 40%[18, 67, 68]. Iliadou et al.[76] reported results of linkage analysis in a large pedigree with monogenic inheritance and excluded linkage to known loci (OTSC1, OTSC2, OTSC3 and OTSC5, as well as COL1A1 and COL1A2), suggesting that several otosclerosis susceptibility genes can be involved in the sporadic cases of otosclerosis representing multifactorial form of disease. Declau et al.[77] and Pauw et al.[78,79] outlined clinical findings in OTSC2, OTSC5 and OTSC7 families, demonstrating phenotype variations in each of these genotypes. Further insight in genetic factors influencing otosclerosis phenotype has been provided by Thys et al.[80] In this study transforming growth factor β1 (TGF- β1) gene was chosen as a primary candidate gene because of it involvement in chondrogenesis of otic capsule. TGF- β1 inhibits osteoclasts differentiation and therefore bone resorbtion caused by osteoclasts carrying CD61 surface antigen. The T2631 variant of TGF-β1 gene has been found to be under-represented in otosclerosis patients, supporting a hypothesis that it is protective against the disease. A variability of phenotypic expression of genetic forms of otosclerosis and a high proportion of sporadic cases imply the contribution of environmental factors in etiology of otosclerotic stapes fixation.

A genetic link to connective tissue disorders has been considered as a possible etiologic factor since the publication of electron microscopic findings of disrupted interwoven pattern of collagen bundles in active otosclerotic foci in 1970.[81]. Gene defect in the COL1A1, gene for type I collagen - the major collagen of the bone matrix, has been hypothesized contributing to the etiology of otosclerosis.[82]. The histopathologic and certain clinical similarities between otosclerosis and osteogenesis imperfecta (OI) have also stimulated an interest in the investigation of possible common genetic origins of these diseases. Mutations in genes for either type I or type II collagen (COL1A1 or COL1A2) have been found associated with dominantly inherited OI.[83]. The demonstration of association between the genes for type I collagen (COL1A1) and clinical otosclerosis led to the hypothesis of shared genetic etiology between otosclerosis and mild forms of OI.[84]. In 2002 McKenna et al.[85] noted that although similar COL1A1 expression has been found in fibroblasts cultured from patients with clinical otosclerosis and OI, COL1A1 mutations accounted only for a small proportion of otosclerosis cases, while the majority was related to other causes. In the most recent study in 2007 Chen et al.[86] reported that different regulatory regions of COL1A1 were found to be either protective or associated with the higher susceptibility to otosclerosis. The definitely causative otosclerosis genes still have not been cloned. While phenotype of mild forms of OI may mimic clinical otosclerosis, no conclusive evidence of shared genetic basis between otosclerosis and OI has been demonstrated to date.[87] The immunohistochemical analysis for five known types of collagen revealed high expression of collagen IV in the mucoperiosteal layer of otosclerotic bone and intense staining of otosclerotic foci with collagen I. These findings are in agreement with inflammatory reaction but distinctive from lesions in OI.[88]

Auto-immunity in etiology of otosclerosis

The otic capsule is a unique structure, being the only bone of human skeleton retaining remnants of cartilage tissue, known as “globuli interossei” in its endochondral layer. During the course of embryogenesis, 14 centers of ossification have been detected in the temporal bone starting from the sixteen week and completing development at the 24th week. Unlike other mature bones, otic capsule contains cartilaginous cells throughout life.[89]

Auto-immune response to collagen in the cartilaginous remnants of otic capsule as the etiopathogenetic mechanism of otosclerosis has been initially hypothesized in1984 based on the animal model.[90] In 1987 Yoo et al.[91] postulated the role of autoimmunity to type II collagen following detection of high levels of collagen II antibodies in serum and in perilymph of patients with otosclerosis. In 1988 Sorensen et al.[92], however, found no difference in circulating antibody concentrations between otosclerosis patients and normal controls. In the studies published from 1993 to 2002 several authors have confirmed the presence of serum antibodies against collagen II in patients with otosclerosis, supporting the hypothesis of collagen-targeting auto-immune mechanism.[93-97] Moreover, Lolov et al.[94] found that antibodies titres were related to the duration of clinical otosclerosis, in agreement with the pattern of auto-immune response to antigenic stimulation. Bujia et al.[95-97] documented high titres of antibodies to collagen II and IX, and chondrocytes in patients with otosclerosis, supporting that cartilage-specific autoimmunity against cartilaginous remnants in otic capsule plays a role in pathogenesis of otosclerosis.

An association between specific human leukocyte antigen (HLA) genotypes and several autoimmune diseases motivated studies of possible contribution of the HLA to the etiology of otosclerosis. In several case-control studies significant differences in levels of various HLA types have been detected between patients with otosclerosis and normal controls.[99-102] Gregoriadis et al.[98] demonstrated that patients with a family history of otosclerosis have significantly elevated levels of the HLA A11, Bw35 and B14. Similar findings were demonstrated by Miyazawa et al.[99], who found significantly higher frequency of HLA-Aw33 in otosclerosis patients compared with control group. Dahlqvist et al.[100] identified significantly lower frequencies of B40 antigen in 74 patients with otosclerosis. Similar results were reported by Svatko[101] who investigated 85 otosclerosis patients and found less frequent occurrence of B40, as well as B27 and Cw1 antigens. Singhal et al.[102] studied the relationship of HLA typing in 100 cases of surgically confirmed otosclerosis and detected significantly higher values of HLA types A9, A11 and B13 in otosclerosis patients compared with age and sex matched controls. Although several authors independently concluded that the presence of specific HLA may be related to an increased susceptibility to otosclerosis, other studies generated inconsistent results. Chobaut[103] and Majsky[104] demonstrated no difference in distribution of HLA-A, -B and –C in patients with otosclerosis and normal controls. Pedersen[105] reported similar results, noting however that they are not excluding the possibility that the errors in HLA system may be associated with a higher susceptibility to otosclerosis.

Viral infections

Inflammatory response to measles[106], mumps and rubella[107] viruses have been well studied since immuno-histochemical techniques made identification of specific viral antigens within active otosclerotic lesions possible. Lately, polymerase chain reaction enabled detection of measles virus genome in the otic capsule and footplate of patients with clinical otosclerosis.[108,109] In addition, elevated levels of anti-measles IgG in perilymph of patients with otosclerosis have been reported.[110] The testing of serum levels of anti-measles IgG has been reported to be highly specific and sensitive and was proposed for serologic diagnosis of otosclerotic stapes fixation in patients with conductive hearing loss.[111] While presence of measles antigens have been repeatedly demonstrated by several authors[112-114], the question of the cause of viral organotropism to otic capsule remains unresolved. Abundance of the receptors to measles virus antigen in otic capsule has been suggested as a possible explanation of the sole predilection of the otic capsule to persistent measles infection.[115] The lack of human cellular receptors of measles virus (the measles-binding protein (CD46)) in other mammals may explain the absence of animal model of otosclerosis, which is unique to a human temporal bone. Comparison of CD46 expression in otosclerotic and normal stapes footplates demonstrated increased numbers of osteoclasts with strong CD46 expression in active otosclerotic foci. Moreover, newly described defective CD46 isoforms detected in otosclerotic otic capsule, which may be responsible for the smooth virus replication, could further elucidate the organotropism of measles virus to the otic capsule.[116] Although numerous findings of measles virus in otosclerotic temporal bones prompted some authors to designate otosclerosis as "measles virus associated inflammatory disease"[107,117], others were unable to find evidence of this infection in stapes specimens from patients with otosclerosis.[118] The epidemiologic observations of decreasing incidence of otosclerosis since the introducing of measles vaccination programs in North America and Europe added an argument to viral etiology theory suggesting that the persistent measles virus infection is at least one of the triggering factors for the development of otosclerosis.[120]

Pathology

A very low rate of bone turnover in the otic capsule is the unique feature of a normal temporal bone, distinguishing it from the rest of the human skeleton. Unlike other mature bones, it contains remnants of unresorbed cartilaginous tissue, known as globuli interossei. In postcartilaginous development the otic capsule demonstrates a progressive inhibition of bone resorbtion toward inner ear spaces, forming a separate functional unit with almost completely absent growth and plasticity.[121,122] The areas of lack of osteoclastic and osteoblastic activity are centrifugally distributed around the oval and round windows and in proximity to perilymphatic spaces.[123] In patho-morphologic sense otosclerosis is a process of disinhibition of active bone remodeling localized to the area of otic capsule leading to resumption of arrested endochondral ossification in globuli interossei.

Once started, otosclerotic lesion evolves through spongiotic, fibrotic and sclerotic stages.[124,125] This sequence of events and their morphologic features are corresponding to the normal repair mechanism of compact bone. The earliest changes in otosclerotic process are believed to originate in globuli interossei with osteocyte-mediated lysis disrupting the interwoven collagen bundles.[126] This active or “spongiotic” stage of otosclerosis is characterized by sharply demarcated area of osteoclastic bone resorbtion with formation of large marrow spaces and intensive neovascularization. Hyperaemic blood vessels of the adjacent promontory mucosa can be observed through the tympanic membrane as a red blush known as the “Schwartze sign”. An objective increase in the blood flow to promontory in patients demonstrating Schwartze sign has been recently demonstrated using laser Doppler flowmetry[127]**.** The pronounced vascularity of active otosclerotic lesion is associated with a vigorous angiogenesis spreading through the newly formed marrow spaces. A new data from the microvessel density analysis of otosclerotic lesions has demonstrated that the extent of angiogenesis is the highest in clinical otosclerosis and is lower in histological otosclerosis and in the normal temporal bone.[128]

The histology of early otosclerosis is characterized by recruitment and activation of osteolytic osteocytes and osteoclasts. At the initial stage otosclerotic lesion expands to surrounding bone in serrated or finger like projections trough pre-existing vascular channels. Lysosome-containing histiocytes, found in the advancing edge of lesion, form characteristic basophilic zones showing an affinity to hematoxylin. This basophilic staining known as “blue mantles” has been first described by Manasse[129] in 1912 and is thought to be the earliest manifestation of otosclerosis[130,131]**.** With the expansion of otosclerotic focus a central resorbtion space is formed, rich with a cellular content of monocytes, macrophages, multinucleated osteoclasts and osteoblasts. Osteoblastic activity leads to formation

of new spongiotic bone with woven trabecular pattern, distinctive from a lamellar structure of the surrounding normal bone.
Fibrotic stage represents a healing phase following resorbtion cessation. The extracellular matrix containing disoriented collagen fibrils undergoes progressive fibrosis and calcification. This reorganization of spongiotic trabecular bone leads to formation of relatively avascular and acelullar dense sclerotic bone.
Sclerotic stage is characterized by scarcity of marrow spaces in inactive focus providing chalky-white gross appearance to remodelled otosclerotic bone.[4-9,132]

Specific areas most commonly affected by the otosclerotic process have been recognized as sites of predilection[20,49,133,134]. Initial otosclerotic foci within a temporal bone have been hypothesized to develop in proximity to the fourteen embryonic ossification centers in temporal bone, to the endolymphatic spaces and to the sources of blood supply[89, 135-137]. Fissula antefenestrum on the anterior rim of the oval window is the most common site of involvement. The second most common location is the round window niche. Apex and medial wall of cochlear labyrinth adjacent to the posterior wall of the internal auditory canal are sharing the third most common place of otosclerotic involvement. Other sites of primary involvement include stapedial footplate, posterior to the round window, anterior wall of the internal auditory canal and around the cochlear aqueduct[20, 49,133,134]. The expansion of otosclerotic focus from the fissula antefenestrum posteriorly toward the stapedial footplate causes gradual immobilization of the vestibulo-stapedial joint due to fibrosis and ossification of the annular ligament. This mechanism of stapedial ankylosis is responsible for the “on-off effect” seen on the impedance testing.[138,139] The degree of stapedial fixation is directly proportional to the severity of conductive hearing loss.[140-142] Anterior spread of prefenestral otosclerotic focus leads to invasion of cochlear endosteum affecting stria vascularis and annual ligament, subsequently contributing to sensorineural hearing impairment.[23,143-146] Studies that explored the association between a stage of histologic changes and clinical findings confirmed the correlation between the histologic stage and cochlear involvement.[49,51] Advancement of otosclerotic focus in medial direction affects partition between the basal turn of cochlea and vestibule and underlying otolitic organs, supposingly causing vestibular symptoms in otosclerosis.[147-149]

The biological mechanisms controlling remodeling of bone tissue include a system of genetically coded proteins, which are influenced by biochemical and hormonal stimuli. Major mediators of bone remodeling include cytokines, grown factors, leikotrienes, prostaglandins and neuropeptides[62,137,150-152]. Bone plasticity is a result of osteoclastic and osteoblastic activity maintaining continuous bone resorbtion and reformation. The balance between the osteoclastic and osteoblastic activity is regulated by a cascade of promoters and inhibitors. Tumor necrosis factor α (TNAα) and interleukins1α and 1β (IL1α and IL1 β) have been recently recognized as osteoclasts activators.[153] TNAα acts on osteoclasts precursors to produce macrophage colony-stimulating hormone (MCSF) and osteoprotegerin ligand (OPGL), leading to the formation and activation of osteoclasts.[154]

Osteoprotegerin (OPG) and transforming growth factor-β1 (TGF- β1) inhibit bone resorbtion by impairing osteoclasts differentiation and activation.[62,80, 55] Recent research has established that the cytokine system of osteoprotegerin (OPG), receptor activator of nuclear factor kB (RANK) and RANK ligand (RANKL) controls local bone remodeling.[156-159] RANK-RANKL interaction acts on osteoclasts precursors resulting in osteoclasts maturation and initiation of bone resorbtion. OPG competes for RANKL receptors on osteoclasts precursors, thereby inhibiting bone remodeling. A recent study demonstrated that OPG was produced in extremely high levels by type I fibrocytes of the spiral ligament and was secreted into the perilymph. Normal otic capsule has been found to contain levels of OPG by factor of 20 higher than other bones.[160] Moreover, the animal model of OPG deficiency exhibited features resembling histologic otosclerosis within the otic capsule.[161] Insight into the final common pathway of bone metabolism therefore brings contemporary knowledge of otosclerosis to the edge of understanding of biological mechanisms underlying this unique pathology of the human temporal bone.

References

1. Smyth GDL. Eighteen years experience in stapedectomy. Ann Otol Rhinol Laryngol 1978;87: 3-36.
2. House J W. The evolution of otosclerosis surgery. Clin N Am 1993;26:323-33.
3. Montandon PB. Teaching stapes surgery. Adv Otolaryngol 2007;65:353-60.
4. Hawke M. Otosclerosis. In: Diseases of the ear. Hawke M, Jahn AF, ed., Philadelphia: Lea & Febiger, 1987; 47-49
5. Schuknecht HF. Disorders of bone. In: . Schuknecht HF.Pathology of the ear.2nd ed. Malvern, PA: Lea & Febiger, 1993;365-414.
6. 6. Nager GT. Otosclerosis. In: Nager GT. Pathology of the ear and temporal bone. Baltimore: Williams & Wilkins, 1993; 943-1010.
7. Friedmann I, Arnold W. Pathology of the ear. Edinburgh London: Churchill Livingstone, 1993: 396-404.
8. Meyerhoff WL. Otosclerosis. In: Head and Neck Surgery. Bailey BJ, ed. Philadelphia: JB Lippincott,1993;1688-1701
9. Michaels L. The ear. In: Pathology of the head and the neck. Cardesa A, Slootweg P, ed., Berlin: Springler-Werlag, 2006; 252-3.
10. Valsalva AM. Tractatus de aure humana, add JP Morgagnus. MDCCXL Venice, Pitteri, 30-32.

11. The diseases of the ear, their diagnosis and treatment: a text-book of aural surgery in the form of academical lectures by Anton von Tröltsch; translated from the German and edited by D.B. St. John Roosa. New York: William Wood, 1864, pp 261,306.
12. Simpson JF. Joseph Toynbee-His contribution to otology. Proc Roy Acad Sci 1963;56:97-105.
13. Mudry A. Adam Politzer (1835-1920) and the description of otosclerosis. Otol Neurotol 2006; 27:276-81.
14. Beales PH. Otosclerosis-past and future. J Roy Soc Med 1979;72:553-61.
15. Milstein S. Otosclerosis. Am J Otol 1981;2:283-5.
16. Siebenmann F.Totaler knocherner verschluss beider labyrinthfester und labyrinthitis serosa infolge progressiver spongiosierung.. Verhanlungen Deutschen Otologischenn Gesellschaft 1912;6:267-83.
17. Puchkovski AM. Otosclerosis. In: Kopmaneyez SM eds. Diseases of the Ear. Kiev: Medgis, 1937; 956-91.
18. Menger DJ, Tange RA. The aetiology of otosclerosis: a review of the literature. Clin Otolaryngol 2003;28:112-20.
19. Guild SR. Histologic otosclerosis. Ann Otol Rhinol Laryngol 1944;53:246-67.
20. Schuknecht HF, Barber W. Histologic variants in otosclerosis. Laryngoscope 1985;95:1307-17.
21. Politzer A. A text-book of the diseases of the ear and adjacent organs, 5 th ed. Translation by Ballin M, Heller C. Philadelphia: Lea & Febiger, 1909.
22. Marres E, Huygen P, De Jong Brand O. The classification of audiograms in otosclerosis. J Otolaryngol ORL 1973;35:205-9.
23. Ghorayeb BY, Linthicum FH. Otosclerotic inner ear syndrome. Ann Otol Rhinol Laryngol 1978;87:85-90.
24. Hayashi H, Onerci O, Paparella MM. Cochlear otosclerosis. Otol Neurotol 2006;27:905-6.
25. Sakai O, Curtin HD, Fujita A, et al. Fenestral and cochlear otosclerosis: CT an MR findings. Am J Otolaryngol 2000;21:116-18.
26. House HP, Sheehy JL. Stapes surgery: selection of the patient. Ann Otol Rhinol Laryngol 1961;70:1062-8.
27. House WF. Oval and round window surgery in extensive otosclerosis. Laryngoscope 1959;69: 693-701.
28. Wiet RJ, Morgenstein SA, Zwolan TA, Pircon SM. Far-advanced otosclerosis. Arch Otolaryngol head Neck Surg 1987;113:299-302.
29. SheaPF, Ge X, Shea JJ. Stapedectomy for far-advanced otosclerosis. Am J Otol 1999;20:425-29.
30. Glasscock ME, Storper IS, Haynes DS, Bohrer PS. Stapedectomy in profound hearing loss. Laryngoscope 1996;106:831-3.
31. Shea JJ, Farrior JB. Stapedectomy and round window closure. Laryngoscope 1987;97:10-2.
32. Amedee RG, Lewis ML. Obliterative otosclerosis. Laryngoscope 1987;97:922-4.
33. Hajek EF. Juvenile otosclerosis. J Laryngol Otol 1961;75:621-39.
34. Robinson M. Juvenile otosclerosis. A 20-year study. Ann Otol Rhinol Laryngol 1983;92:561-5.
35. Huygen PLM, Cremers CWRJ, Verhagen WIM, Joosten FBM Camurati-Engelmann disease presenting as "juvenile otosclerosis". Int J Ped ORL 1996; 37: 129-141.
36. Karosi T, Konya J, Petko M, Szabo L, Pytel J, Jori J, Sziklai I. Two subgroups of Stapes fixation: otosclerosis and pseudo-otosclerosis. Laryngoscope 2005;115:1968-73.
37. Swartz JD, Mandell DW, Wolfson RJ, et al. Fenestral and cochlear otosclerosis: CT evaluation. Am J Otol 1985;6:476-81.
38. Rovsing H. Otosclerosis fenestral and cochlear. Radiol Clin N Am 1974;12:505-15.
39. Valvassori GE. Imaging of otosclerosis. Otolaryngol Clin N Am 1993;26:359-71.
40. Swartz JD, Harnsberger HR. The otic capsule and otodystrophies. In: Imaging of the temporal bone. 3rd ed. New York: Thieme, 1998; 240-317.
41. Mafee MF, Henrikson GC, Deitch RL, et al. Use of CT in stapedial otosclerosis.Radiology 1985;156:709-714
42. Swartz JD, Faerber EN, Wolfson RJ, et al.Fenestral otosclerosis: significance of preoperative CT evaluation. Radiology 1984; 151:703-707.
43. Valvassori GE, Dobben GD. CT densitometry of the cochlear capsule in otosclerosis. AJNR 1985;6:661-67.
44. Mafee MF, Valvassori GE, Deitch RL, et al. Use of CT in the evaluation of cochlear otosclerosis. Radiology 1985;156:703-708.

45. Valvassori GE. CT densitometry in otosclerosis. Adv Otolaryngol 1987;37:47-49.
46. Sakai O, Curtin HD, Hasso AN, Swartz JD. Otosclerosis and dysplasias of the Temporal bone. In: Head and Neck Imaging. Som PM, Curtin HD, eds. 4th ed. St. Louis: Mosby, 2003:1245-53.
47. Toynbee J. The diseases of the ear. London: HK Lewis, 1868.
48. Sakalinskas V, Jankauskas R. Clinical otosclerosis and auditory exostoses in ancient Europeans. Laryngol Otol 1993;107:489-91.
49. Hueb MM, Goycoolea MV, Paparella MM, Oliviera JA. Otosclerosis: the University of Minnesota temporal bone collection. Otolaryngol H&N Surg 1991;105:396-405.
50. Declau F, Van Spaendonck M, Timmermans JP, Michaels L, Liang J, Qiu JP, Van de Heyning P. Prevalence of otosclerosis in an unselected series of temporal bones. Otol Neurotol 2001;22:596-602.
51. Gros A, Vatovec J, Sereg-Bahar M. Histologic changes on stapedial footplate in otosclerosis: correlations between histologic activity and clinical findings. Otol Neurotol 2003;24:43-47.
52. Altmann F, Glasgold A, Macduff JP. The incidence of otosclerosis as related to race and sex. Ann Otol Rhinol Laryngol 1967;76:377-92.
53. Hall JG. Otosclerosis in Norway. A geographical and genetical study. Acta Otolaryngol 1974;324 (suppl):1-20.
54. Tshitularo MI, Joseph CA. Otosclerosis among South African indigenous blacks. E Afr Med J 2005;82:223-5.
55. Goto S, Omori Y. Otosclerosis in Japan. Nagoya J Med Sci 1957;19:147-54.
56. Pearson RD, Kurland LT, Cody DTR. Incidence of diagnosed clinical otosclerosis. Arch Otolaryngol 1974;99:288-91.
57. Levin G, Fabian P, Stahle J. Incidence of otosclerosis. Am J Otol 1988;9:299-301.
58. Tato JM, Tato JM Jr. Otosclerosis and races. Ann Otol Rhinol Laryngol 1967;76:1018-1025.
59. Browning GG, Gatehouse S. The prevalence of middle ear disease in the adult British Population. Clin Otolaryngol 1992;17:317-21.
60. Ohtani I, Baba Y, Suzuki T, Suzuki C, Kanu M, Deka RC. Why is otosclerosis of low prevalence in Japanese? Otol Neurotol 2003;24:377-81.
61. Gordon MA. The genetics of otosclerosis: a review. Am J Otol 1989;10:426-38.
62. Stankovic K, McKenna M. Current research in otosclerosis. Cur Opinion in Otolaryngol 2006;14:347-351.
63. Sercer A. Etiopathogenesis of otospongiosis and anthropological factors. Arch Italian Otol 1958; 69 (Suppl 34):1-92.
64. Polanski JF, Quispe MF, Felix F, Condon L, Portmann D. Post-traumatic otosclerosis: coincidence or trigger factor? Rev Laryngol Otol Rhinol 2008;129:57-9.
65. Toynbee J. Pathological and surgical observations on the diseases of the ear. Med Chir Trans 1861;24:190-205.
66. Shaaf T, Gapany-Gapanavicus B. The genetics of otosclerosis. Am J Hum Genet 1978;30:59-64.
67. Morrison AW. Genetic factors in otosclerosis. Ann R Coll Surg Engl 1967;41:202-237.
68. Morrison AW, Bundley SE. The inheritance of otosclerosis. J Laryngol Otol 1970;84:921-32.
69. Tomek MS, Brown MR, Mani SR, Ramesh A, Srisailapathy CR, Couke P, Zbar RI, Bell AM, McGuirt WT, Fukushima K, Willems PJ, Van Camp G, Smith RJH. Localization of a gene for otosclerosis to chromosome 15q25-q26. Hum Mol Genet 1998;7:285-90.
70. Van Den Bogaert K, Govaerts PJ, Schatteman I, Brown MR, Caethoven G, Offeciers FE, Somers T, Declau F, Coucke P, Van de Heyning P, Smith RJH, Van Camp G. A second gene for otosclerosis (OTSC2) maps to chromosome 7q34-36. Am J Hum Genet 2001;68:495-500.
71. Van Den Bogaert K, Govaerts PJ, De Leenheer EMR, Scatteman I, Verstreken M, Chen W, Declau F, Cremers CWRJ, Van De Heyning PH, Offeciers FE, Somers T, Smith RJH, Van Camp G. Otosclerosis: A genetically heterogeneous disease involving at least three different genes. Bone 2002;30:624-30.
72. Chen W, Campbell CA, Green GE, Van Den Bogaert K, Komodikis C, Manolidis LS, Aconomou E, Kyamides Y, Christdolou K, Faghel C, Giguere CM, Alford RL, Manolidis S, Van Camp G, Smith RJH. Linkage of otosclerosis to a third locus (OTSC3) on human chromosome 6p21.3-22.3. J Med Genet 2002;39:473-77.
73. Van Den Bogaert K, De Leenheer EMR, Chen W, Lee Y, Numberg P, Pennings RJE, Vanderstraeten K, Thys M, Cremers CWRJ, Smith RJH, Van Camp G. A fifth locus for otosclerosis, OTSC5, maps to chromosome 3q22-24. J Med Genet 2004;41:450-3.

74. Brownstein Z, Goldfarb A, Levi H, Frydman M, Avraham K. Chromosomal mapping and phenotypic characterization of hereditary otosclerosis linked to the OTSC4 locus. Arch Otolaryngol 2006;132:416-24.
75. Thys M, Van Den Bogaert K, Iliadou V, Vanderstraeten K, Dieltjens N, Schrauwen I, Shen W, Eleftheriades N, Grigoriadou M, Pauw RJ, et al. A seventh locus for otosclerosis, OTSC7, maps to chromosome 6q13-16.1. Eur J Hum Genet 2007;15:362-8.
76. Iliadou V, Van Den Bogaert K, Eleftheriades N, Aperis G, Vanderstraeten K, Fransen E, Thys M, Grigoriadou M, Pampanos A, Economides J, Iliades T, Van Camp G, Peterson MB. Monogenic nonsyndromatic otosclerosis: audiological and linkage analysis in a large Greek pedigree. Int J Ped Otolaryngol 2006;70:631-7.
77. Declau F, Van Den Bogaert K, Van De Heyning PH, Offeciers E, Govartes P, Van Camp G. Phenotype-genotype correlations in otosclerosis: clinical features of OTSC2. Adv Otolaryngol 2007;65:114-18.
78. Pauw RJ, De Leenheer EMR, Van Den Bogaert K, Huygen PLM, Van Camp G, Joosten FMB, Cremers CWRJ. The phenotype of the first otosclerosis family linked to OTSC5. Otol Neurotol 2006;27:208-15.
79. Pauw RJ, Huygen PL, Thys M, Van Camp G, Joosten FB, Cremers CW. Phenotype description of a Dutch otosclerosis family with suggestive linkage to OTSC7. Am J Med Genet 1007;143A:1613-22.
80. Thys M, Schrauwen I, Vanderstraeten K, Janssens K,, Dieltjens N, Van Den Bogaert K, Fransen E, Chen W, Ealy M, Claustres M, Cremers CWRJ, Dhooge I, Declau F, Claes J, Van De Heyning PH, Vincent R, Somers T, Offeciers E, Smith RJH, Van Camp G. The coding polymorphism T2631 in TGF- β1 is associated with otosclerosis in two independent populations. Hum Molec Genet 2007;16:2021-30.
81. Chevance L, Bretlau P, Jorgensen M, et al. Otosclerosis: An electron microscopic and cytochemical study. Acta Otolaryngol (Suppl)1970;272:1-68.
82. Thalmann I, Thallinger G, Thalmann R. Otosclerosis: a local manifestation of a generalized connective tissue disorder? Am J Otolaryngol 1987;8:308–16.
83. Sykes B, Ogilvie D, Wordsworth P, et al. Consistent linkage of dominantly inherited osteogenesis imperfecta to the type I collagen loci: COL1A1 and COL1A2. Am J Hum Genet 1990;46:293-307.
84. McKenna MJ, Kristiansen AG, Bartley ML, Rogus JJ, Haines JL. Association of COL1A1 and otosclerosis. Evidence for a shared genetic etiology with mild osteogenesis imperfecta. Am J Otol 1998;19:604-10.
85. McKenna MJ, Kristiansen AG, Tropitzsch AS. Similar COL1A1 expression in fibroblasts from some patients with clinical otosclerosis and those with type I osteogenesis imperfecta. Ann Otol Rhinol Laryngol 2002;111:184-9.
86. Chen W, Meyer NC, McKenna MJ, Pfister M, McBride DJ, Fukushima K, Thys M, Camp GV, Smith RJ. Single-nucleotide polymorphisms in the COL1A1 regulatory regions are associated with otosclerosis. Clin Genet 2007;71:406-14.
87. Moumoulidis I, Axon P, Baguley D, Reid E. A review on the genetic of otosclerosis.Clin Otolaryngol 2007;32:239-47.
88. Niedermeyer HP, Becker ET, Arnold W. Expression of collagens in the otosclerotic Bone. Adv Otolaryngol 2007;65:45-49.
89. Milroy CM, Michaels L. Pathology of the otic capsule. J Laryngol Otol 1990;104:83--90.
90. Yoo TJ. Etiopathogenesis of otosclerosis: a hypothesis. Ann Otol Rhinol Laryngol 1984; 93: 28-33.
91. Yoo TJ, Shea JJ, Floyd RA. Enchondral cartilage rests collagen-induced autoimmunity: a possible pathogenic mechanism of otosclerosis. Am J Otolaryngol 1987;8: 317-24.
92. Sorensen MS, Nielsen LP, Bretlau P, et al. The role of type II collagen autoimmunity in otosclerosis revisited. Acta Otolaryngol 1988;105:242-7.
93. Niedermeyer HP, Arnold W. Etiopathogenesis of otosclerosis. J Otolaryngol 2002;64:114-19.
94. Lolov SR, Edrev GE, Kyurkchiev SD, Kehayov IR. Elevated autoantibodies in sera from otosclerotic patients are related to disease duration. Acta Otolaryngol 1998;118:375-80.
95. Bujia J, Sittinger M, Arnold B, Wilmes E, Hammer C, Burmester G. Cartilage-specific autoimmunity in otosclerosis. HNO 1993;41:507-11.
96. Bujia J, Alsalameh S, Jerez R, Sittinger M, Wilmes E, Burmester G. Antibodies to the minor cartilage collagen type IX in otosclerosis. Am J Otol 1994;15:222-24.
97. Bujia J, Burmester G. The presence of antibodies directed against specific cartilaginous collagens in patients with otosclerosis. Acta Otolaryngol Esp.1993;44:277-80.

98. Gregoriadis S. Zervas J, Varletzidis E, et al. HLA antigens in patients with otosclerosis. Arch Otolaryngol 1982;108:769-771.
99. Miyazawa T, Tago C, Ueda H, Niwa H, Yanagita N. HLA associations in otosclerosis in Japanese patients. Eur Arch Otolaryngol 1996;253(8):501-3.
100. Dahlqvist A, Diamant H, Dahlqvist SR, Cedergren B. HLA antigens in patients with otosclerosis. Acta Otolaryngol 1985;100:33-35.
101. Svatko LG, Ismagilov SM. The significance of genetic markers in otosclerotic hearing loss. Vestn Otolaryngol 1994;5-6:13-15.
102. Singhal SK, Mann SB, Datta U, Panda NK, Gupta AK. Genetic correlation in otosclerosis. Am J Otolaryngol 1999;20:102-5.
103. Chobaut JC, Bertrand D, Raffoux C, Wayoff. HLA antigens in otosclerosis. Am J Otol 1982; 3:241-2.
104. Majsky A, Novotny Z, Fajstavr Y. HLA and otosclerosis. Tissue Antigenes 1982;20:306-7.
105. Pedersen U, Madsen M, Lamm LU, Elbrond O. HLA -A, -B, -C antigens in otosclerosis. J Laryngol Otol 1983;97:1095-7.
106. McKenna MJ, Mills BG. Immunohistochemical evidence of measles virus antigens in active otosclerosis. Otolaryngol Head Neck Surg1989;101: 415-421.
107. Arnold W, Friedman I. Immunohistochemistry of otosclerosis. Acta Otolaryngol Suppl 1990; 470:124-29.
108. McKenna MJ, Kristiansen AG, Haines J. Polymerase chain reaction amplification of a measles virus sequence from human temporal bone sections with active otosclerosis. Am J Otol 1996;17:827-30.
109. Karosi T, Konya J, Petko M, Szabo L, Pytel J, Jori J, Sziklai I. Two subgroups of Stapes fixation: otosclerosis and pseudo-otosclerosis. Laryngoscope 2005;115:1968-73.
110. Arnold W, Niedermeyer HP, Lehn N, et al. Measles virus in otosclerosis and the specific immune response of the inner ear. Acta Otolaryngol 1996;116:705-9.
111. Karosi T, Konya J, Petko M, Szabo L, Pytel J, Jori J, Sziklai I. Antimeasles Immunoglobulin G for serologic diagnosis of otosclerotic hearing loss. Laryngoscope 2006;116:488-93.
112. McKenna MJ, Mills BG. Ultrastructural and immunohistochemical evidence of measles virus in active otosclerosis. Acta Otolaryngol Suppl 1990;470:130-39.
113. Niedermeyer HP, Arnold W, Schuster M, et al. Persistent measles virus infection and otosclerosis. Ann Otol Rhiniol Laryngol 2001;110:897-903.
114. Karosi T, Konya J, Szabo L, Sziklai I. Measles virus prevalence in otosclerotic stapes footplate samples. Otol Neurotol 2004;25:451-56.
115. Karosi T, Jokay I, Konya J, Petko M, Szabo L, Sziklai I. Expression of measles receptors in otosclerotic, non-otosclerotic and in normal stapes footplates. Eur Arch Otolaryngol 2007;264:607-13.
116. Karosi T, Szalmas A, Csomor P, Konya J, Petko M, Sziklai I. Disease-associated novel CD46 splicing variants and pathologic bone remodelling in otosclerosis. Laryngoscope 2008; 118:1669-76.
117. Niedermeyer HP, Arnold W. Otosclerosis: measles virus associated inflammatory disease. Acta Otolaryngol 1995;115:300-303.
118. Grayeli AB, Palmer P, Tran BA, Huy P, et al. No evidence of measles virus in stapes samples from patients with otosclerosis. J Clin Microbiol 2000;38:2655-60.
120. Niedermeyer HP, Arnold W. Otosclerosis and measles virus-association or causation? J Otolaryngol 2008;70:63-9.
121. Sorensen MS. Temporal bone dynamics, the hard way. Formation, growth, modeling, repair and quantum type bone remodeling in the otic capsule. Acta Otolaryngol Suppl. 1994;512:1-22.
122. Sorensen MS, Jorgensen MB, Bretlau P. Drift barriers in the postcartilaginous development of the mammalian otic capsule. Eur Arch Otolaryngol 1992;249:56-61.
123. Frisch T, Sorensen MS, Overqaard S, Bretlau P. Predilection of otosclerotic foci related to the bone turnover in the otic capsule. Acta Otolaryngol Suppl. 2000;543:111-3.
124. Djeric D. Stapes pathology in otosclerosis: scanning electron microscopic examination. Adv Otolaryngol 2007;65:59-60.
125. Lim DJ, Robinson M, Saunders WH. Morphologic an immunohistochemical observations of otosclerotic stapes. Am J Otolaryngol 1987;8:282-95.
126. Davis GL. Pathology of otosclerosis. Am J Otolaryngol 1987;8:273-81.
127. Nakashima T, Sone M, Fujii H, et al.. Blood flow to the promontory in cochlear otosclerosis. Clin Otolaryngol 2006;31:110-15.

128. Jyung RW, Wacharasindhu C. Quantification of angiogenesis in otosclerosis. Laryngoscope 2005;115:811-16.
129. Manasse P. Die Ostitis Chronica Metaplastica der Labyrinthkapsel. Wiesbaden, J.F. Bergman,1912.
130. Weber MJ. The blue mantles in otosclerosis: a contribution to the pathology of the labyrinthine capsule. Ann Otol Rhinol Laryngol 1933;42:438-54.
131. Lindsay JR. Blue mantles in otosclerosis. Ann Otol Rhinol Laryngol 1974;83:33-41.
132. Linthicum FH. Histopathology of otosclerosis. Clin N Am 1993;26:335-57.
133. Nylen B. Histopathological investigations of the localization, number, activity and extent of otosclerotic foci. J Laryngol Otol 1949;63:321-27.
134. Guild SR. Incidence, location and extent of otosclerotic lesions. Arch Otolaryngol 1950; 52:848-52
135. Frisch T, Sorensen MS, Overgaard S, Bretlau P. Predilection of otosclerotic foci related to the bone turnover in the otic capsule. Acta Otolaryngol Suppl 2000;542:111-13.
136. Sorensen MS, Frisch T, Bretlau P. Dynamic bone studies of the labyrinthine capsule in relation to otosclerosis. Adv Otolaryngol 2007;65:53-8.
137. Chole RA, McKenna M. Pathophysiology of otosclerosis. Otol Neurotol 2001;22:249- 57.
138. Bel J, Causse JR, Michaux P et al. Mechanical explanation of the on-off effect (diphasic impedance change) in otoscpongiosis. Audiology 1976;15:128.
139. Love T, Stream R. The biphasic acoustic reflex: a new perspective. Laryngoscope 1978; 88:298.
140. Parahy C, Linthicum FH. Otosclerosis and otospongiosis: clinical and histiological comparisons. Laryngoscope 1984;94:508-12.
141. Hannley MT. Audiologic characteristics of the patient with otosclerosis. Otolaryngol Cl;in N Am 1993;26:373-87.
142. Cherukupally SR, Merchant SN, Rosowski JJ. Correlations between pathologic changes in the stapes and conductive hearing loss in otosclerosis. Ann Otol Rhinol Laryngol 1998; 107:319-26.
143. Lindsay JR, Beal DD. Sensorineural deafness in otosclerosis. Observations on histo-pathology. Ann Otolaryngol 1966;75:436-57.
144. Gussen R. Labyrinthine otosclerosis and sensorineural deafness. Pathologic findings of the spiral ligament. Arch Otolaryngol 1975;101:438-40.
145. Causse J, Chevance LG, Bretlau P, et al. Enzymatic concept of otospongiosis and cochlear otosclerosis. Clin Otolaryngol 1977;2:23-32.
146. Elonka DR, Applebaum EL. Otosclerotic involvement of the cochlea: a histologic and audiologic study. Otolaryngol Head Neck surg 1981;89:343-51.
147. Hawkins JE, Linthicum FH, Johnsson LG. Cochlear and vestibular lesions in capsular otosclerosis as seen in microdissection. Ann Otol Rhinol Laryngol 1978;87(Suppl 48):1-40.
148. Saim L, Nadol J. Vestibular symptoms in otosclerosis- correlation of otosclerotic involvement of vestibular apparatus and Scarpa's ganglion cell count. Am J Otol 1996;17:263-70.
149. Yang T, Young Y. Vestibular-evoked myogenic potentials in patients with otosclerosis using air- and bone-conducted tone-burst stimulation. Otol Neurotol 2006;28:1-6.
150. Adachi K, Chole RA, Yee J. Indomethacin inhibition of middle ear bone resorbtion. Arch Otolaryngol Head Neck Surg 1991;117:267-9.
151. Chole RA, Tinling SP. Adaptive bone modeling in the middle ear is substance-p dependent. Am J Otol 1998;19: 521-24.
152. Sherman EE, Chole RA. The effects of leucotriene and cyclooxygenase inhibition on adaptive bone remodeling in the middle ear. Otolaryngol Head Neck Surg 2000;123:1-8.
153. Kostenuk PJ, Shalhoub V. Osteoprotegerin: a physiological and pharmacological inhibitor of bone resorption. Curr Pharm Des 2001;7:613-35.
154. Abu-Amer Y, Ross FP, Edwards J, Teitelbaum SL. Lipopolysaccharide-stimulated osteo-clastogenesis is mediated by tumor necrosis factor via its P55 receptor. J Clin Invest 1997; 100:1557-65.
155. Janssens K, ten Dijke P, Janssens S., Van Hul W. Transforming growth factor-β1 to the bone. Endocr Rev 2005;26:743-74.
156. Rosa-Ranal M, De la Cruz DA, Lorena-Rubio Y, Larrea F. New paradigms in the regulation of bone metabolism. Rev Invest Clin 2001;53:362-69.
157. Liu X, Kirschenbaum A, Yao S, Levine AC. Cross-talk between the interleukin-6 and prostaglandin E2 signaling systems results in enhancement of osteoclastogenesis through

effects on the osteoprotegerin/receptor activator of nuclear factor-kB(RANK) ligand/RANK system. Endocrinology 2005;146:1991-98.
158. Nakamura M, Udagawa N, Nakamura H. Bone remodeling and mineral homeostasis.Clin Calcium 2006;16:46-53.
159. McKenna MJ, Kristiansen AG. Molecular biology of otosclerosis. Adv Otolaryngol 2007;65: 68-74.
160. Zehnder AF, Kristiansen AG, Adams JC , Merchant SN, McKenna MJ. Osteoprotegerin in the inner ear may inhibit bone remodeling in the otic capsule. Laryngoscope 2005;115:172-77.
161. Zehnder AF, Kristiansen AG, Adams JC , Kujawa SG, Merchant SN, McKenna MJ. Osteo-protegerin knockout mice demonstrate abnormal bone remodeling of the otic capsule and progressive hearing loss. Laryngoscope 2005;116:201-6.

1.2

A history of stapes surgery and the evolution of surgical techniques

Introduction

The contemporary state of the art of stapes surgery evolved during the 150 years of vigorous attempts of seeking the ways to restore an impaired hearing in otosclerosis patients. This eventful history has illustrated how the chance and coincidence shaped the minds of pioneers of otology, leading to the different methods of surgical restoration of hearing.

The actual ear surgery in humans was preceded by animal experiments and thoughtful observation of occasional traumatic events resulted in hearing improvement. In 1841 Joseph Toynbee described the case of short-lived restoration of hearing following loud noise exposure.[1] A first reference to the attempts of stapes mobilization for hearing improvement was mentioned by Prosper Meniere in 1842.[2] In 1878 Johannes Kessel reported the case of hearing improvement after severe head trauma, with post-mortem confirmed temporal bone fracture through the horizontal semicircular canal.[4] As a Docent of the University of Gratz, Kessel demonstrated that the removal of columella in pigeons does not cause permanent labyrinthine impairment.[5] Later, being a Professor of the University of Jena, Kessel performed stapes mobilisation and also tried removing the stapes, reporting "some improvement of hearing and no serious complications".[6] These early reports have been followed by publications of Boucheron[7], Miot[8], Moure[9] and Pottier[10] in France, Schwartze[11], Passow[12] and Lucae[13] in Germany and Faraci[14] in Italy, contributing to the collective experience in stapes mobilization gathered in Europe by the end of XIX century. At the same period of time, series of stapes surgeries have been reported in North America. Frederick Jack[15,16] carried out first stapes operation at the Massachusetts Eye and Ear Infirmary in June 1891. This was followed by series of 70 surgeries performed by Blake[17,18] who coined the term "stapedectomy" and used "incudostapedectomy" in cases requiring incus removal. Early enthusiasm from the occasional postoperative hearing improvement has been hampered however by overall poor surgical results and unfortunate cases of fatal complications. These inevitable failures have been caused by the limited knowledge of ear pathology causing hearing loss and crude surgical technique available at the time. The re-fixation of previously mobilized stapes prompted lack of the lasting hearing improvement. The absence of the proper grafting resulted in oval window fibrosis after stapes extraction and doomed early attempts of stapes removal to fiasco. Moreover, the dangers of violating of the inner ear space in pre-antibiotic era shaped an opinion against surgical treatment of otosclerotic stapes fixation. In 1900 at the International Congress of Otolaryngology, Siebenmann[19] was supported by Politzer and Moure in declaring stapes surgery as dangerous and useless. As Adam Politzer wrote further in 1901 "the unfavourable opinion as to the value of extraction of stapes in non-suppurative middle ear process, and in otosclerosis, has been confirmed by many authors", therefore providing his enormous authority against this procedure.[20]

Summarizing the overview of early stages of surgical treatment of otosclerosis, the two main branches in the evolution of surgical approaches can be recognized (Figure 1).

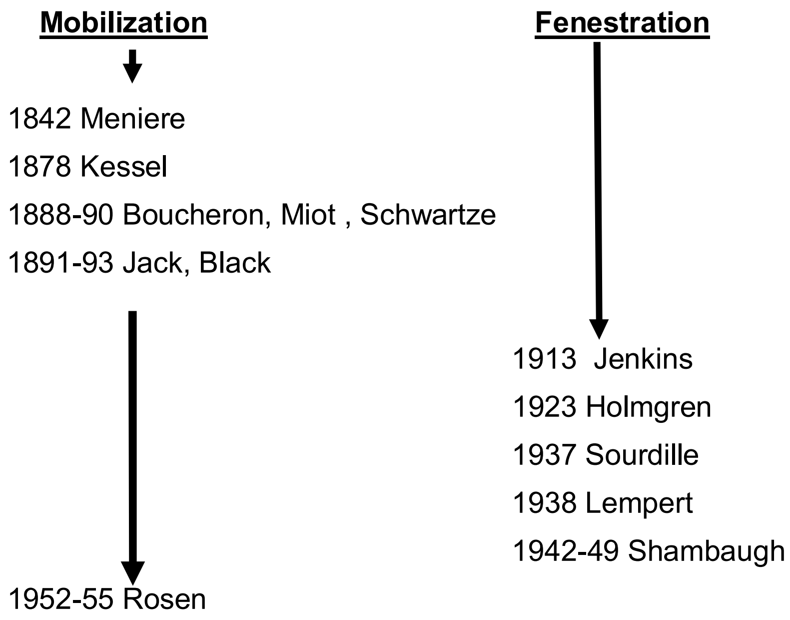


Figure 1. Evolution of early methods of surgical treatment of otosclerosis.

The idea of stapes mobilization could be possible originate in the observations of temporary hearing improvement due to direct tapping of the stapes with golden rods[2,3] by Meniere or Toynbee's description of accidental restoration of hearing following acute exposure to loud noise.[1] The concept of fenestration could possible emerge from the observation of improved hearing in case of traumatic temporal bone fracture through the labyrinth reported by Kassel.[6] The rudimentary knowledge of sound amplification function of the middle ear and vague understanding of mechanisms of hearing loss in otosclerosis until the beginning of XX th century precluded emerging of effective method of surgical correction of hearing loss in otosclerosis at the time. The growing body of knowledge concerning mechanisms of hearing loss in otosclerosis stimulated a new generation of otologists seeking another ways allowing sound stimulation of inner ear by by-passing an immobile stapes. It was not before 1913, when Jenkins[21] introduced fenestration operation, creating an opening in the horizontal semicircular canal and covering it with the skin flap from the external auditory meatus. Professor Gunnar Holmgren in 1923 modified the fenestration technique by using mucoperiosteal flap for the covering of fenestra in the horizontal semicircular canal, allowing substantial, although temporary, improvement of hearing.[22] Holmgrens' introduction of binocular surgical microscope[23], which replaced the first operational monocular microscope used by Nylen[24,25] was his another invention which has opened a new era in otologic surgery.[26] Retro auricular incision used by Jenkins, Holmgren and Nylen for creation of closed fenestration operation proved subsequent burying of the fenestra to the exterior, therefore limiting lasting maintenance of hearing gain. Maurice Sourdille recognized shortcomings of Holmgren's' technique and developed a three-stage exteriorized fenestration operation, called "tympanolabyrinthopexy", which he presented to the Otolaryngology Section of the New York Academy of Medicine in 1937.[27] Julius Lempert in 1938 made his contribution to the history of stapes surgery by introducing his one-stage fenestration procedure, first employing dental electrical burr for creation of the fenestra.[28] Lempert's innovative endaural approach for exteriorization of the ampulla of horizontal semicircular canal was based on his technique of endaural radical mastoidectomy, described in 1928 [29]**.** In 1945 Lempert published further modification of his technique, introducing the mobile cartilaginous stopple which was intended to simulate the movement of normal stapedial footplate and to prevented involvement of the perilymphatic space in postoperative inflammation of the tympano-meatal flap.[30] Lemperts' surgical skills and innovations used in his one-stage fenestration technique conquered the minds of finest otologists of the time, prompting wide acceptance of this technique as an effective treatment of otosclerosis which was previously deemed incurable.[31,32,33] Thus the opinion of uselessness of surgical treatment of otosclerosis held since the verdict of Siebenmann and Politzer in 1900 has shifted toward general acknowledgement of possibility of surgical correction of hearing loss. Although the fenestration technique enabled hearing improvement in otosclerosis, it never resulted in complete closure of air-bone gap. In addition, the life-long need for a management of radical mastoid cavity irreversibly affected patients' postoperative quality of life. The change of state of the mind concerning surgery as the preferred treatment of otosclerosis was probably the most important impact of the fenestration era on future developments in stapes surgery.

As history of science showed in the past, many scientific discoveries have happened by chance. Thus, re-introduction of the stapes mobilization followed the observation of accidental hearing improvement in the patient who underwent stapes palpation by Samuel Rosen.[34] The results of mobilization technique published by Rosen in 1953 seemed promising to otologists familiar with the typical outcomes of fenestration procedures.[35-37] Normal hearing often immediately achieved by stapes mobilization rendered popularity of this relatively simple procedure, performed under local anaesthesia via endaural approach. Hearing results of stapes mobilization were negatively affected by recurrent stapes ankylosis that made immediate hearing improvement unsustainable in a long term.

John Shea revolutionized stapes surgery by not only extracting ankylosed stapes, but also first performing immediate reconstruction of middle ear sound-conductive mechanism in 1955. This technique, evolved from the method of stapes mobilization re-introduced by Rosen[36,37] and early attempts of stapedectomy reported by Jack[38], has been presented at the Triological Society Meeting in Montreal in 1956, opening the modern era of stapes surgery.[39-41] The concept of stapedectomy with the immediate reconstruction of middle ear sound-conductive mechanism turned this method to the new operation of choice for otosclerosis rather than stapes mobilization or fenestration of bony labyrinth. In following decades massive body of information concerning complications[42-51] and long term results of stapedectomy[52-57] has been published. This enormous collective experience along with the publications of clinical and morphological findings at revision surgeries[58-63] resulted in numerous modifications to the original technique.[3,35,64-68] Every stage of the procedure has been revised and refined. A complete or nearly-complete removal of the stapedial footplate (the formal or “total stapedectomy”) has been replaced by “partial stapedectomy”[69,70] or “partial platinectomy”.[71] In pursuit of minimizing loss of perilymph and subsequent cochlear damage leading to sensory-neural deafness, a “small fenestra” stapedotomy technique has been advocated by Gordon Smyth in 1978.[72] The concept of “small hole” stapedotomy has been described earlier by Shea[41] in 1962 and gradually become a common practice following publications of Smyth[73], McGee[74], Bailey[75], Fisch[76], House[77] and others.[78,79]

A discussion concerned with the size of stapedotomy has been closely interwoven with the ongoing evolution of stapes prostheses. The prostheses of different materials, shapes, weights and dimensions have been painstakingly compared with scrupulous attention to their intra-operative handling properties, long-term stability and physical characteristics affecting postoperative hearing[80-88]. In the extensive study of phylogeny of the stapes prosthesis by Fritsch and Naumann[89] as many as 105 different prostheses have been reviewed and this number continues to grow following the evolution of the surgical technique. The problems of biocompatibility encountered with early designs and materials of prostheses gave rise to the “stapedotomy minus prosthesis technique” originated in the method of “anterior crurotomy mobilization” described by Fowler in 1960.[90] Silverstein further developed a "minimally invasive stapes surgery" approach without employment of artificial prosthesis, using mobilized posterior stapedial crura and preserved stapedial tendon in selected cases of mild otosclerosis.[91-93]

Technological advances have been continuously enhancing otologic surgical armamentarium and have been applied in the most critical steps of the surgical procedure. Thus, opening in the footplate which was originally made with sharp perforators has been recognized as one of the most traumatic steps of stapes surgery, associated with the risk of footplate fracture, floating footplate and excessive bleeding or perilymphatic leak.[94] This former cold steel stapedotomy has been replaced by use of microdrills[95] and later by laser beam application[96-102], allowing relatively atraumatic creation of stapedotomy with better preservation of high frequency hearing.[103,104]

Introduction of self-crimping prostheses and laser-assisted evaporation of stapedial superstructure allowed to revise and to reverse the order of surgical steps[73] which have been used in traditional stapedectomy.[41] With this modification, prosthesis is placed before removal of the stapes crura: by removing the posterior crus and stapedius tendon, access to the footplate is similar to that obtained during conventional stapedotomy, while the retained anterior crus provides better stability to the footplate and incus, reducing the risk of incus dislocation and providing additional protection against inadvertent footplate fragmentation.[105,106]

Exceptional hearing results of modern stapedotomy technique inspired Howard Houses' declaration in 1993 that "stapes surgery now reached a stage of perfection".[35] Rephrasing Joseph Sullivans' quotation of the speech before the Canadian Medical Association in 1942[33], it can be repeated although, that "Every good surgical technique undergoes continual stages of improvement and development. Why should the stapes surgery procedure be any exception to this fundamental principle of surgery?" The future directions in further refinement of stapedotomy technique will probably involve a laser-assisted endoscopic surgery[107] and robot-assisted technologies.[108] Although these high technologies can be helpful in overcoming technical difficulties of stapes surgery, progressing sensory-neural hearing loss in otosclerosis will limit abilities of surgical treatment and will require development of gene therapy to allow prevention and possible reversal of otosclerotic process.

References

1. Toynbee J. Pathological and surgical observations of the diseases of the ear. Trans Med Chir Soc Lond. 1841;24:190-196.
2. Meniere P. De L'exploration de l'appareil audif, ou recherché sur les moyens prospers a conduire au diagnostic des maladies de l'oreille. Gaz Med Paris 1842;10:114-117.

3. Hausler R. General history of stapedectomy. In: Otosclerosis and stapes surgery. Adv Otolaryngol. Karger, Basel 2007;65:1-5.
4. Heermann H. Johannes Kessel and the history of endaural surgery. Arch Otolaryngol 1969;90:652-658.
5. Kessel J. Ueber die durchschneidung des steigbugelmuskels beim menschen und uber die extraction des steigbugels, respective der Columella bie Thieren. Arch Ohrenh 1876;11:199-217.
6. Kessel J. Uber das Mobilisieren des Steigbugels durch Ausschneiden des Trommelfelles, Hammers und Ambosses bei Undurchgangigkeit der Tuba. Arch Ohrenheilkd 1878;13:69-88.
7. Boucheron E. La mobilisation de l'etrier et son procede operatoire. Union Med Paris 1888; 46:412-416.
8. Miot C. De la mobilisation de l'etrier. Rev Laryngol 1890;10:49-54.
9. Moure E. De la mobilisation de l'etrier. Rev Laryngol 1890;7:225-233.
10. Pottier A. Du traitement de la surdite consecutive a l'otite moyenne purulente. 1889 Paris; Ollier-Henry: 59.
11. Schwartze H. Lehrbuch der chirurgishen krankheiten des ohres. 1885 Stuttgart; Enke.
12. Passow K. Operative anlegung einer offnung in die mediale paukenhohlenwand bei stapes-ankylose. Verh Dtsch Otol Ges 1897;6:141.
13. Lucae A. Ueber operative entfernung des trommelfells und der beiden groesseren gehor-knochelchen bei sklerose der pauken-schleimhaut. Arch Ohrenh 1885;22:233-42.
14. Faraci G. Importanza acustica e funzionale della mobilizzazione della staffa; resultati di una nuova serie di operazioni. Arch Ital Otol 1899;9:209-221.
15. Jack F. Remarkable improvement of hearing by removal of the stapes. Trans Am Otol Soc 1891;5:284.
16. Jack F. Further observations on removal of the stapes. Trans Am Otol Soc 1893;5:494.
17. Black C. Middle ear operations. Trans Am Otol Soc 1892;5:306.
18. Black C. Operation for the removal of the stapes. Bost M&S J 1892;127:469.
19. Siebenmann F. Traitement chirurgical de la sclerose otique. Cong Int Med Sec Otol 1900; 13:170.
20. Politzer A. Lehrbuch der Ohrenheilunde fur praktische Aerzte und Studirende, 4 ed. Stuttgart, Enke 1901. English translation: Politzer A. A text-book of the Diseases of the Ear and Adjacent Organs for Students and Practitioners. 4th ed (Translation by Ballin M, Heller C) Philadelphia, Lea 1902.
21. Jenkins G. Otosclerosis: certain clinical features and experimental operative procedures. Trans17th Int Congr Med 1913;16:609-618.
22. Holmgren G. Some experiences in the surgery of otosclerosis. Acta Otolaryngol 1923;5:460-466.
23. Holmgren G. The surgery of otosclerosis. Ann Otol Rhinol Laryngol 1937;46:3-12.
24. Nylen C. The microscope in aural surgery, its first use and later development. Acta Otolaryngol Suppl (Stockh) 1954;116.
25. Dohlman G. Carl Olof Nylen and the birth of the otomicroscope and microsurgery. Arch Otolaryngol 1969; 6: 813-817.
26. Mudry A. The History of the Microscope for Use in Ear Surgery. Am J Otol, 2000;6: 877-886.
27. Sourdille M. New technique in the surgical treatment of severe and progressive deafness from otosclerosis. Bull NY Acad Med 1937;13:673.
28. Lempert J. Improvement in hearing in cases of otosclerosis: A new, one stage surgical technique. Arch Otolaryngol 1938;28:42-97.
29. Lempert J. Simple subcortical mastoidectomy. Arch Otolaryngol 1928;7:201-228.
30. Lempert J. Cartilaginous stopple. Arch Otolaryngol 1945;1:41.
31. Shambaugh G. Julius Lempert and the fenestration operation. Am J Otol 1995;16:247-252.
32. Beales P. Otosclerosis-past and present. J Royal Soc Med 1979;72:553-561.
33. Sullivan J. Indications for and results of the fenestration operation. Can Med Ass J 1945;53: 543-548.
34. Rosen S. Palpation of stapes for fixation: preliminary procedure to determine fenestration suitability for otosclerosis. Arch Otol 1952;56:610.
35. House H. The evolution of otosclerosis surgery. Otolaryngol Clin N Am 1993;26:323
36. Rosen S. Mobilization of the stapes to restore hearing in otosclerosis. N Y State J Med 1953;53: 2650.
37. Rosen S. Restoration of hearing in otosclerosis by mobilization of the fixed stapedial footplate. An analysis of results. Laryngoscope 1955;65:224-229.

38. Jack F. Supplementary report on a case of double stapedectomy operated upon ten years ago. Trans Am Otol Soc 1902;8:99-10.
39. Meltzer P, Lindsay J, Goodhill V, et al. Symposium. The operation for the mobilization of the stapes in otosclerotic deafness. Laryngoscope 1956;66:729-84.
40. Shea J. Fenestration of the oval window. Ann Otol Rhinol Laryngol 1958;67:932-51.
41. Shea J, Sanabria F, Smyth G. Teflon piston operation for otosclerosis. Arch Otolaryngol 1962;76:516-21.
42. Causse JB, Causse JR, Wiet RJ, Yoo TJ. Complications of stapedectomies. Am J Otol 1983; 4:275-80.
43. 43. Hough J, Dyer R. Stapedectomy. Causes of failure and revision surgery in otosclerosis. Otolaryngol Clin N Am 1993; 26:453-70.
44. Wiet RJ, Harvey SA, Bauer GP. Complications in stapes surgery. Options for preventions and management. . Otolaryngol Clin N Am 1993; 26:471-90.
45. Mann W, Amedee R, Fuerst G. Hearing loss as a complication of stapes surgery. Otolaryngol Head Neck Surg.1996; 115(4):324-8.
46. Pratt L. Postoperative complications associated with bilateral stapedectomy. Laryngoscope 1972;1: 1189-98.
47. Schuknecht H, Mendoza A. Cochlear pathology after stapedectomy. Am J Otolaryngol. 1981; 2(3):173-87.
48. Eviatar A, Jamal H. Cholesteatoma induced by stapedectomy. Arch Otolaryngol. 1983; 109(6):413-4.
49. Sale C. Bilateral deafness due to stapes surgery. Arch Otolaryngol. 1969; 90(4):467-9.
50. Belal A, Ylikoski J. Post-stapedectomy dizziness. A histopathologic report. Am J Otol.1982; 3(3):187-91.
51. Ferguson B, Gillespie, Kenan P, Farmer J. Mechanisms of cholesteatoma formation following stapedectomy. Am J Otol. 1986; 7(6): 420-4.
52. Smyth G.D.L, Hassard T.H. Eighteen years experience in stapedectomy. Ann Otol Rhinol Laryngol Suppl 1978; 5-6 87 :(Suppl 49)3-36.
53. Ginsberg I, Hoffman S, Stinziano G, White T. Stapedectomy-in depth analysis of 2405 cases. Laryngoscope 1978;88:1999-2016.
54. Sheehy JL, Nelson RA, House HP. Stapes surgery at the Otologic Medical Group. Am J Otol 1979;1:22—26.
55. Levy R, Shvero J, Hadar T. Stapedotomy technique and results: ten years' experience and comparative study with stapedectomy. laryngoscope 1990;100:1097-1100.
56. Somers T, Govartes P, Marquet T, Offeciers E. Statistical analysis of otosclerosis surgery performed by Jean Marquet. Ann Otol Rhinol Laryngol. 1994; 103(12):945-51.
57. Vincent R, Sperling NM, Oates J et al. Surgical findings and long-term hearing results in 3050 stapedotomies for primary otosclerosis: a prospective study with the otology-neurotology database. Otol Neurotol 2006; 27: S25-S47.
58. Feldman B, Schuknecht H. Experiences with revision stapedectomy procedures. Laryngoscope 1970: 80:1281-91.
59. Derlacki E. Revision stapes surgery: problems and some solutions. Laryngoscope 1985; 95: 1047-53.
60. Glasscock ME, McKennan KX, Levine SC. Revision stapedectomy surgery. Otolaryngol H&N Surg. 1987;96(2):141-48.
61. Palva T, Ramsay H. Revision surgery for otosclerosis. Act Otolaryngol (Stockh) 1990; 110: 416-20.
62. De La Cruz A, Fayad J. Revision stapedectomy. Otolaryngol Head Neck Surg 2000;12:728-32.
63. Lippy W.H, Battista R.A, Berenholz L, Schuring A.G, Burkey J.M. Twenty-year review of revision stapedectomy. Otol Neurotol 2003; 24:560-66.
64. Shea J. A personal history of stapedectomy. Am J Otol (Suppl)1998;19:2-12.
65. Shea J. Forty years of stapes surgery. Am J Otol 1998;19:52-55.
66. Shea J, Shea P, McKenna M. Stapedectomy for otosclerosis. In: Surgery of the ear. Glasscock-Shambaugh (ed), BC Decker, Hamilton 2003;57:517-31.
67. Goycoolea M. Surgery for stapes fixation. In: Atlas of otologic surgery. Paparella M, Nissen R (ed) WB Saunders Co, Philadelphia 1989;13:247-271.
68. Marquet T. "Stapedotomy" technique and results. Am J Otol 1985; 6:63-67.

69. Persson P, Harder H, Magnuson B. Hearing results in otosclerosis surgery after partial stapedectomy, total stapedectomy and stapedotomy. Acta Otolaryngol (Stockh) 1997; 117:94-99.
70. Moon C, Hahn M. Partial vs total footplate removal in stapedectomy: a comparative study. Laryngoscope 1984;94:912-15.
71. Cremers C, Beusen J, Huygen P. Hearing gain after stapedotomy, partial platinectomy, or total stapedectomy for otosclerosis. Ann Otol Rhinol Laryngol 1991;100:959-61.
72. Smyth G. The case for the small fenestra stapedotomy. trans Am Acad Ophthalmol Otolaryngol 1978;86:488-491.
73. Smyth G. Practical suggestions on stapedotomy. Laryngoscope 1982; 92:952-3.
74. McGee TM. Comparison of small fenestra and total stapedectomy. Ann Otol Rhinol Laryngol 1981; 90: 630-832.
75. Bailey T. Small fenestra stapedectomy. A preliminary report. Laryngoscope 1981;91:1308-1321.
76. Fish U. Stapedotomy vs. stapedectomy. Am J Otol 4:112-17, 1982.
77. House H, Hansen M, Al Dakhalil A, House W. Stapedectomy versus stapedotomy: comparison of results with long-term follow-up. Laryngoscope 2002;112:2046-50.
78. Rizer FM, Lippy. Evolution of techniques of stapedectomy from the total stapedectomy to the small fenestra stapedectomy. Otolaryngol Clin N Am 1993; 26:443-51.
79. Herzog J. 0.4 mm stapedotomy: a consistent technique for otosclerosis. Am J Otol 1991;12: 16-19.
80. Shea J, Homsy C. The use of proplast in otologic surgery. Laryngoscope 1974; 84:1835.
81. House H. The prefabricated wire-loop Gelfoam stapedectomy. Arch Otolaryngol 1962;76: 298-302.
82. Robinson M, Seltzer E. Stainless steel stapedial prosthesis, a preliminary report. Laryngoscope 1961;71:385-388.
83. McGee T. The stainless steel piston; surgical indications and results. Arch Otolaryngol 1965;81:34.
84. Lippy W, Burkey J, Schuring A, Berenholz L. Comparison of Titanium and Robinson stainless steel stapes piston prosthesis. Otol & Neurotol 2005;26: 874-877.
85. Grolman W, Tange R. First experience with a new stapes clip piston in stapedotomy. Otol & Neurotol 2005;26:595-598.
86. Zepeda-Lopez E, Bello-Mora A, Feliz-Trujillo M. Poststapedectomy hearing gain: comparison of a Teflon prosthesis with a Schuknecht-type wire-Teflon prosthesis. Ear, Nose, Throat J 2005;84:707-19.
87. De Bruijn A, Tange R, Dreschler W. Comparison of stapes prosthesis. a retrospective analysis of individual audiometric results obtained after stapedotomy by implantation of a gold and a Teflon piston. Am J Otol 1998;20:573-580.
88. Gjuric M, Rukavina L. Evolution of stapedectomy prostheses over time. Adv Otolaryngol 2007;65:174-8.
89. Fritsch M, Naumann I. Phylogeny of the stapes prosthesis. Otol & Neurotol 2008;29:407-415.
90. Fowler E. Anterior crurotomy with footplate fracture. Arch Otolaryngol 1960;71:296-304.
91. Silverstein H. Laser stapedotomy minus prosthesis: a minimally invasive procedure. Am J Otol 1998;19:277-82.
92. Silverstein H, Hester T, Rosenberg S, Deems D. Preservation of the stapedius tendon in laser stapes surgery.Laryngoscope 1998;108:1453-8.
93. Silverstein H, Jackson L, Conlon W, Rosenberg S, Thompson J. Laser stapedotomy minus prosthesis: absence of refixation. Otol Neurotol 2002;23:152-7.
94. Rothbaum DL, Roy J, Hager GD, Taylor RH, Whitcomb LL, Francis HW, Niparko JK. Task performance in stapedectomy: Comparison between surgeons of different experience levels. Otolaryngol Head Neck Surg 2003; 128: 71-7.
95. Gjuric M. Microdrill versus perforator for stapedotomy. Clin Otolaryngol 1990;15:411-13.
96. Perkins R. Laser stapedectomy for otosclerosis. . Laryngoscope 1980;90:228-41.
97. Palva T. Argon laser in otosclerosis surgery. Acta Otolaryngol (Stockh) 1987;104:153-7.
98. Cherini SG, Horn KL, Bowman CA, Griffin G. Small fenestra stapedotomy using a fiberoptic hand-held argon laser in obliterative otosclerosis. Laryngoscope 1990;100:1276-82.
99. Lesinski SG. Lasers for otosclerosis-which one if any and why. Lasers in Surg & Med 1990;10:448-57.

100. Vincent R, Gratacap B, Causse JB, Cherini S. Argon laser and Gherini-Causse Endo-Otoprobe in otologic surgery. ENT Journal 1996; 12:770-80.
101. Gantz B. Argon laser stapedotomy. Ann Otolaryngol Chir Cervicofac 1982;91:25-6.
102. Lesinski SG, Palmer A. Lasers for otosclerosis: CO2 vs. Argon and KTP-532. Laryngoscope 1989;99:1-8.
103. Raut V, Halik J. Argon laser assisted small fenestra stapedotomy for otosclerosis. Auris Nasus Larynx 2005; 32:11-15.
104. Kisilevsky V, Dutt S. Bailie N, Halik J. Hearing results of 1145 stapedotomies evaluated with Amsterdam Hearing Evaluation Plots. J Laryngol Otol 2009;26:1-7.
105. Szymanski M, Golabek W, Morshed K, Siwiec H. The influence of the sequence of Surgical steps on complications rate in stapedotomy. Otol Neurotol. 2007;28:152-6.
106. Lang EE, Waheed K, Colreavy M, Burns H. A retrospective review of stapes surgery following introduction of “reversal of the steps” technique. Clin Otolaryngol 2004;29:571-3.
107. Poe D. Laser-assisted endoscopic stapedectomy: A prospective study. Laryngoscope 2000;110: 1-30.
108. Rothbaum D, Roy J, Stoianovici D, Berkelman P, Hager G, Taylor R, Whiotcomb L, Francis H, Niparko J. Robot-assisted stapedotomy; micropick fenestration of the stapes footplate. Otolaryngol Head & Neck Surg 2002;127:417-26.

1.3

The rationale of this thesis

The rationale of this thesis emerged from a daily otology practice of preoperative patients counseling based on personal experience rather than averaged statistics published elsewhere. The ability to predict technical results of stapedotomy and to estimate the achieved benefit of surgery as close as possible to patient's perception required diverse methods of evaluation of surgical outcomes. In outset of this thesis, we aimed to answer the following questions concerned with the multi-dimensional evaluation of the results of stapes surgery:

1. What are the technical results of modified laser-assisted stapedotomy technique evaluated according to the AAO-HNS guidelines?
2. What are the results of each individual case in the large series of stapedotomies evaluated with the Amsterdam Hearing Evaluation Plots?
3. What are the functional results of unilateral stapes surgery evaluated with the Glasgow Benefit Plot?
4. What are the benefits of bilateral stapedotomy in achievement of normal and symmetrical hearing?
5. How the preoperative hearing status affects the surgical outcomes?
6. What are the technical results and the functional benefits of stapes surgery performed for the correction of severe and profound hearing loss?
7. What are the technical results and the functional benefits of stapes surgery performed for the correction of congenital conductive hearing loss?
8. What are the results of malleo-vestibulopexy technique used for correction of complex middle ear pathology?

Chapter 2

Primary Stapes Replacement

2.1

Hearing results of 1145 stapedotomies evaluated with Amsterdam Hearing Evaluation Plots

V.E. Kisilevsky
S.N. Dutt
N.A. Bailie
J.J. Halik

The Journal of Laryngology & Otology 2009; 123: 730-736

Abstract

Aims

To evaluate the hearing results of a large series of primary stapedotomies, according to American Academy of Otolaryngology, Head and Neck Surgery guidelines and Amsterdam hearing evaluation plots.

Study design

Retrospective chart review.

Methods

The charts for 1369 consecutive stapedotomy cases were reviewed; 1145 cases of primary stapedotomy were included. Raw data from the audiometric database were evaluated using Amsterdam hearing evaluation plots. The effect on outcomes of using different audiological parameters was analysed.

Results

A significant improvement was demonstrated in mean post-operative air conduction and speech reception thresholds, with no change in bone conduction. Air–bone gap closure of 10 dB or more was achieved in 82 per cent of cases. A 'dead ear' occurred in one patient (0.1 per cent).

Conclusion

This study reports the largest series of primary stapedotomies evaluated with Amsterdam hearing evaluation plots. This method enables visual identification of successful and unfavourable results, providing more accurate and detailed presentation of surgical outcomes.

Introduction

Surgical correction of conductive hearing loss in cases of otosclerosis is one of the most successful procedures in otology. Early reports on large series of stapedectomies demonstrated excellent results, with air–bone gap (ABG) closure of 10 dB or more in greater than 90 per cent of patients.[1–3] Since the introduction of stapedectomy by Shea in 1956, the procedure has been significantly refined. In addition to evolution of the surgical technique, the criteria for reporting the outcome of stapes surgery have been revised.

Closure of the ABG was initially considered to be themainmeasure of success for stapes surgery. Historically, ABG closure was calculated by comparing postoperative pure tone average (PTA) thresholds for air conduction with pre-operative bone conduction thresholds at only three frequencies (0.5, 1 and 2 kHz). This method has a number of flaws, in that post-operative deterioration in bone conduction at 0.5–2 kHz was missed, as was sensorineural hearing loss at high frequencies; it could also result in an artificially improved post-operative ABG by inclusion of overclosure in the calculation. In order to overcome these limitations, in 1995 the American Academy of Otolaryngology, Head and Neck Surgery committee on hearing and equilibrium recommended inclusion of the higher frequency of 3 kHz in the calculation of PTAs, and the use of post-operative bone conduction thresholds when calculating the post-operative ABG.[4] However, despite committee recommendations to report the change in bone conduction at 3 kHz for each case, this parameter remains underreported. The new guidelines also recommended reporting raw data along with summary datawhenever possible.While most of the literature on stapes surgery has been concerned with reporting summary statistics of surgical results, a few publications have provided detailed, raw data representing individual cases.[5,6]

In 2001, DeBruijn et al.[7] proposed awayof reporting results fromindividual cases – the Amsterdam hearing evaluation plots method. This innovation aimed to present results for each operated ear separately, with regard to pre- and post-operative bone conduction thresholds and the relationships between pre-operative ABG and gain in post-operative air conduction thresholds. Browning et al.[8] also introduced the Glasgow benefit plot for evaluation of bilateral hearing function in each individual case of middle-ear surgery. To the best of our knowledge, no large series of stapedotomies evaluated by these methods have been reported in the indexed, English language medical literature.

The objective of this study was to analyse the outcomes of 1145 stapedotomies, according to the current guidelines of the American Academy of Otolaryngology, Head and Neck Surgery committee on hearing and equilibrium for the evaluation of conductive hearing loss treatment, and simultaneously to evaluate these results using Amsterdam hearing evaluation plots. Results for the analysis of binaural hearing function by Glasgow benefit plots have been reported in a separate paper. A detailed analysis of hearing thresholds at 4 kHz has been performed in order to evaluate surgical trauma to the cochlea, since bone conduction thresholds at this frequency have been found to be the most sensitive indicator of inner-ear damage.[9]

Materials and methods

The charts for 1369 consecutive stapedotomy cases performed by one surgeon (JH, the senior author) from 1991 to 2006 were reviewed. Of these 1369 cases, 89 were revision stapedotomies and 48 were performed for congenital stapes fixation. Eightyseven cases were excluded from the study due to incomplete data. The audiometric results of 1145 primary stapedotomies for otosclerosis were included for evaluation. Six hundred and ninety-five patients (61 per cent) were women and 450 (39 per cent) were men. The mean age was 45 years (standard deviation (SD) 12 years), with a range of 16 to 84 years. The mean follow up was 16.4 months (SD 19.2 months), with a range of 0.25 to 117 months.

Stapedotomy was performed using the reverse order technique[10,11] via a per meatal approach. A modified Cawthorne (0.3 mm diameter) prosthesis was used until 2002, in 756 cases. Thereafter, a modified Causse (0.4 mm diameter) prosthesis was used, in 376 procedures. A 0.4 mm (for Cawthorne prostheses) or 0.5 mm (for Causse prostheses) fenestra was made in the footplate using Halik graduated perforators. Adjunctive use of an Argon beam laser (wavelength 512 nm), delivered via an EndoOtoprobe™ (Lumenis, inc. Salt-Lake City, UT, USA), was introduced in 2000. The laserassisted procedures were performed on 516 patients. A modification of the reverse order technique was made in conjunction with introduction of the laser: the Argon laser was used to vaporise the stapedial tendon and posterior crus of the stapes, prior to footplate fenestration. The laser was also used to begin the footplate fenestration by providing a ‘set-hole’ for the perforators. Fenestration was completed with graduated perforators. The laser also proved useful in dealing with adhesions.

Hearing outcomes were analysed according to the 1995 American Academy of Otolaryngology, Head and Neck Surgery committee on hearing and equilibrium guidelines.[4] Air conduction thresholds at 0.5, 1, 2, 3, 4, 6, 8 and 12 kHz and bone conduction thresholds at 0.5, 1, 2, 3 and 4 kHz, for operated and non-operated ears, were tabulated in a computer database. Mean pre-operative values for air and bone conduction pure tone averages and for ABG, at 0.5, 1, 2 and 3 kHz, were compared with corresponding post-operative values. The beginning of this study period pre-dated the 1995 American Academy of Otolaryngology, Head and Neck Surgery guidelines, and therefore 3 kHz results were not available for all patients. In such cases, 3 kHz air and bone conduction values were estimated by averaging results for 2 and 4 kHz. Post-operative bone conduction values were used to calculate the post-operative ABG. Postoperative ABGs were also calculated for the frequencies 0.5, 1 and 2 kHz and the frequencies 0.5, 1, 2 and 4 kHz, in order to evaluate the effects of these different audiometric parameters on the success rate. Mean pre- and post-operative air and bone conduction thresholds at 4 kHz were also assessed separately, in order to evaluate the impact of stapedotomy on hearing at high frequencies. Preand post-operative speech reception thresholds were also evaluated. The mean pre-operative hearing level, calculated as a PTA at the frequencies 0.5, 1, 2 and 3 kHz, was 54 dB (SD 15 dB), and the mean pre-operative ABG was 28 dB (SD 9 dB). One hundred and forty-four (13 per cent) patients were treated for severe or profound hearing loss (pre-operative hearing levels $\geq$71 dB).

In addition to reporting averaged results prepared from the mean values of several audiometric parameters, we also utilised Amsterdam hearing evaluation plots[7] to produce a visual presentation of the operative course of each individually operated ear. Pre-operative bone conduction PTAs were plotted against corresponding post-operative bone conduction PTAs, and pre-operative ABG values were plotted against air conduction gains, for each individual case, allowing data for individual patients to be recognised as plot points on a graph. Data were analysed with the statistical software program Statistica 6.0 (StatSoft, Tulsa, Oklahoma, USA). Statistical analysis was carried out for comparisons between pre- and post-operative values, using the paired t-test for the evaluation of air and bone conduction pure tone averages, speech reception thresholds and ABGs. The criterion selected for statistical significance was $p < 0.05$.

Results and analysis

Pure tone thresholds

A frequency-specific analysis of pre- and postoperative air and bone conduction levels demonstrated statistically significant improvement at all frequencies, except for bone conduction thresholds at 4 kHz and air conduction thresholds at 12 kHz (Table I). Figures 1 and 2 demonstrate pre- and post-operative audiograms reconstructed as box and whisker plots showing mean air and bone conduction values. This method allowed graphical presentation of a large amount of raw data, without hiding unsuccessful cases.

Table 1. Pre- and post-operative air and bone conduction by frequency.

Frequency (kHz)	Pre-op (dB)		Post-op (dB)		*p*
	Mean	(SD)	Mean	(SD)	
AC					
0.5	58.4	14.7	27.1	13.1	<0.001
1	57.0	14.9	28.2	13.7	<0.001
2	49.6	17.0	30.1	14.8	<0.001
3	50.2	18.4	31.0	16.5	<0.001
4	50.8	21.4	36.7	19.3	<0.001
8	56.1	24.5	50.4	24.6	<0.001
12	72.4	22.8	75.9	20.8	<0.001
BC					
0.5	21.2	10.9	19.3	10.5	<0.05
1	23.2	12.0	21.1	11.6	<0.001
2	30.5	12.8	25.8	13.3	<0.001
3	27.1	13.8	25.7	14.7	<0.05
4	23.9	16.3	25.3	16.3	NS

Preop - pre-operative; post-op - postoperative; SD - standard deviation; AC - air conduction; BC - bone conduction; NS - not significant

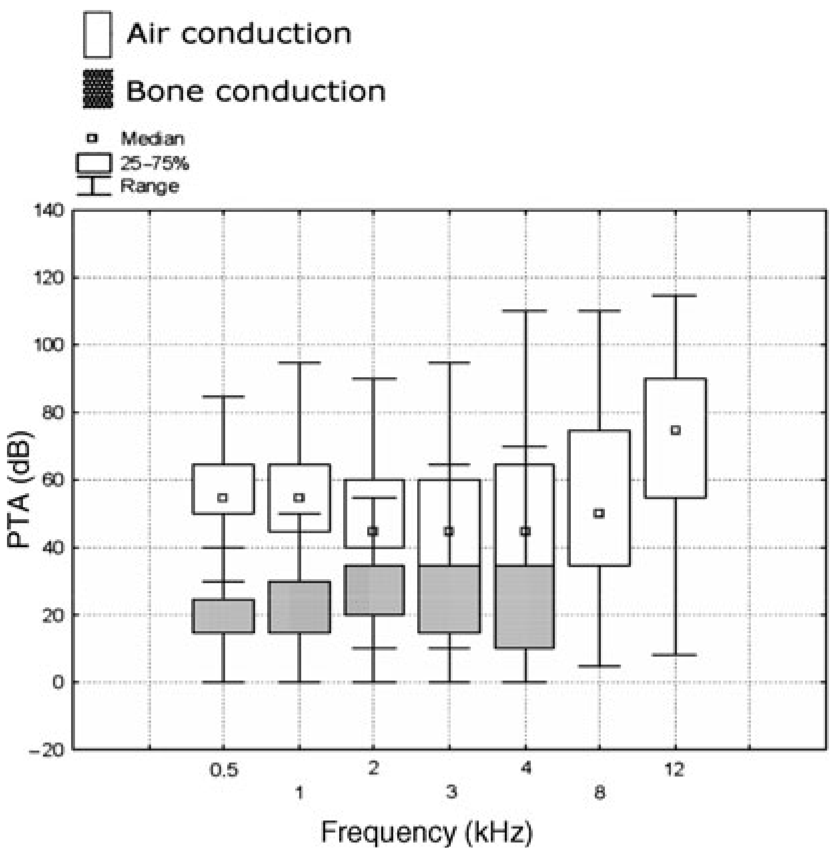


Figure 1. Reconstruction of pre-operative audiogram using mean air and bone conduction threshold at 0.5, 1, 2 and 3 kHz.

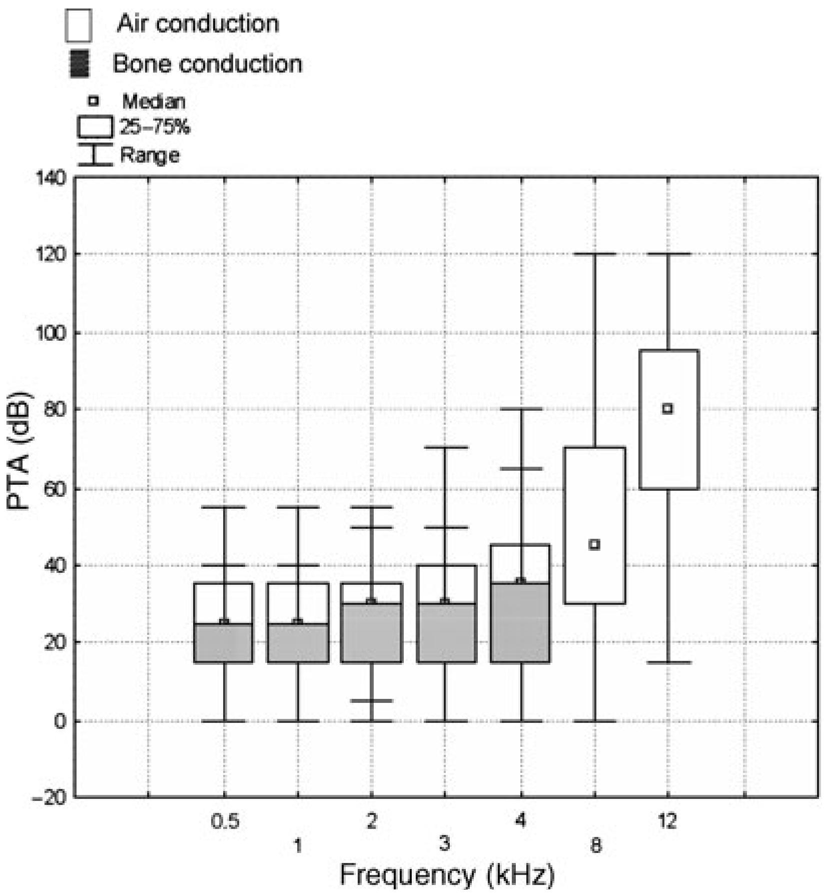


Figure 2. Reconstruction of post-operative audiogram using mean air and bone conduction thresholds at 0.5, 1, 2 and 3 kHz

Air and bone conduction, air–bone gap and speech reception thresholds

Figure 3 summarises the pre- and post-operative air and bone conduction thresholds and ABGs as box and whisker plots. Govartes et al.[12] have proposed combining, in one graph, box and whisker plots of several variables in order to enable detailed representation of a large amount of audiometric data. The median value is depicted as a central dot, the dispersion as a box and the range as whiskers. Outliers are depicted by separate dots beyond the whiskers. The post-operative, four-frequency average ABG was 7 dB (SD 6 dB), compared with 28 dB (SD 9 dB) pre-operatively ($p < 0.0001$). The mean postoperative speech reception threshold was 27 dB (SD 12 dB), compared with 51 dB (SD 14 dB) preoperatively (($p < 0.0001$). The mean post-operative bone conduction threshold was 25 dB (SD 11 dB), compared with 24 dB (SD 11 dB) pre-operatively ($p < 0.0001$).

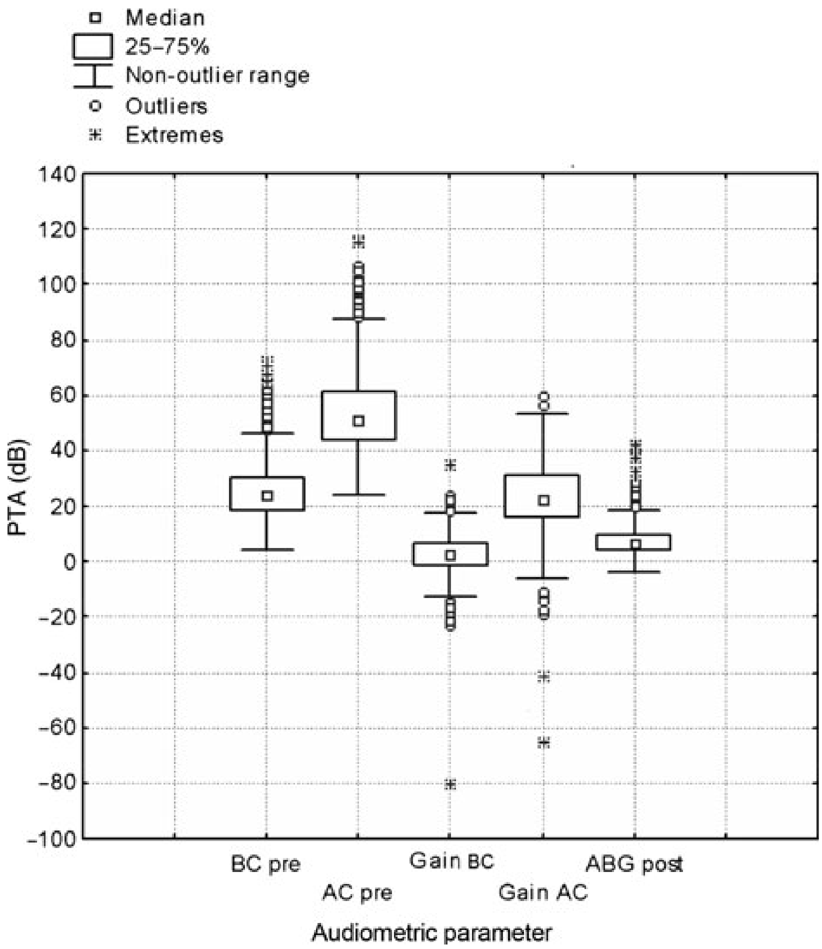


Figure 3. Main audiometric parameters presented as box and whisker plot. BC = bone conduction; AC - air conduction; ABG - air-bone gap.

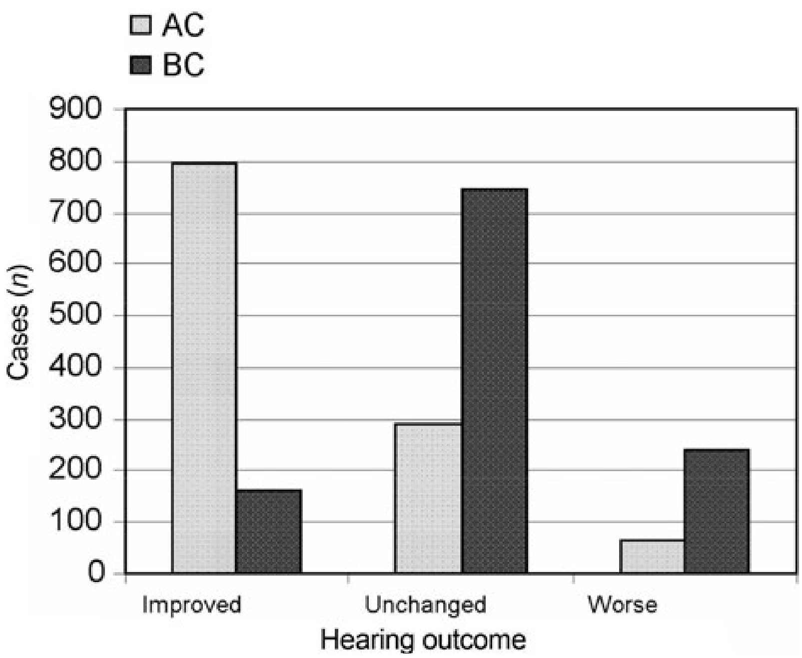


Figure 4. Hearing results at 4 kHz. Improved = >10 dB; unchanged = <u><</u>10 dB; worse = >10 dB; AC - air conduction; BC - bone conduction

Air and bone conduction at 4 kHz

Detailed analysis of hearing results at 4 kHz is shown in Figure 4. Air conduction thresholds improved in 793 (68 per cent) patients, remained unchanged in 288 (26 per cent) and worsened in 64 (6 per cent). Bone conduction thresholds improved in 160 (14 per cent) patients, remained unchanged in 744 (65 per cent) and worsened in 241 (21 per cent).

Air–bone gap at 0.5–2, 0.5–3 and 0.5–4 kHz

Inclusion of higher frequencies in the calculation of post-operative ABG did not influence ABG closure within 20 dB (Table II). Air–bone gap closure within 20 dB was achieved in about 97 per cent of cases, for any frequency combination. When 4 kHz was included in the ABG calculation, the success rate of closure to less than 10 dB fell from 82 to 75 per cent. Calculation of ABG at 0.5–2 kHz resulted in only a marginal increase in closure to less than 10 dB, rising to 83 per cent.

Table 2. Post-operative air-bone gap for the three common frequency combinations

ABG (dB)	0.5, 1 & 2 kHz	0,5, 1, 2 & 3 kHz	0.5, 1, 2 & 4 kHz
<u><</u>10	942 (82.3%)	937 (81.8%)	861 (75.2 %)
11-20	173 (15.1%)	178 (15.6%)	248 (21.7 %)
20	30 (2.6 %)	30 (2.6 %)	36 (3.1 %)

Hearing results assessment with Amsterdam hearing evaluation plots Plotting the pre- versus post-operative bone conduction thresholds, calculated according to the American Academy of Otolaryngology, Head and Neck Surgery guidelines as a four-frequency average at 0.5, 1, 2 and 3 kHz, enabled visualisation of each individual case at a glance (Figures 5 and 6). In Figure 5, every point below the lower diagonal line represents one of the 57 (5 per cent) patients who demonstrated a post-operative bone conduction threshold improvement of 10 dB or greater. Every point above the upper diagonal line represents one of the 35 (3 per cent) patients who demonstrated a post-operative bone conduction thresholds worsening of 10 dB or greater. In Figure 6, pre-operative ABG values are plotted against post-operative air conduction gain. Points below the lower diagonal line represent the 132 successful results with overclosure. Points above the upper diagonal line represent the 56 cases with unsuccessful outcomes caused by negative changes in air conduction thresholds or ABG ≥ 20 dB.

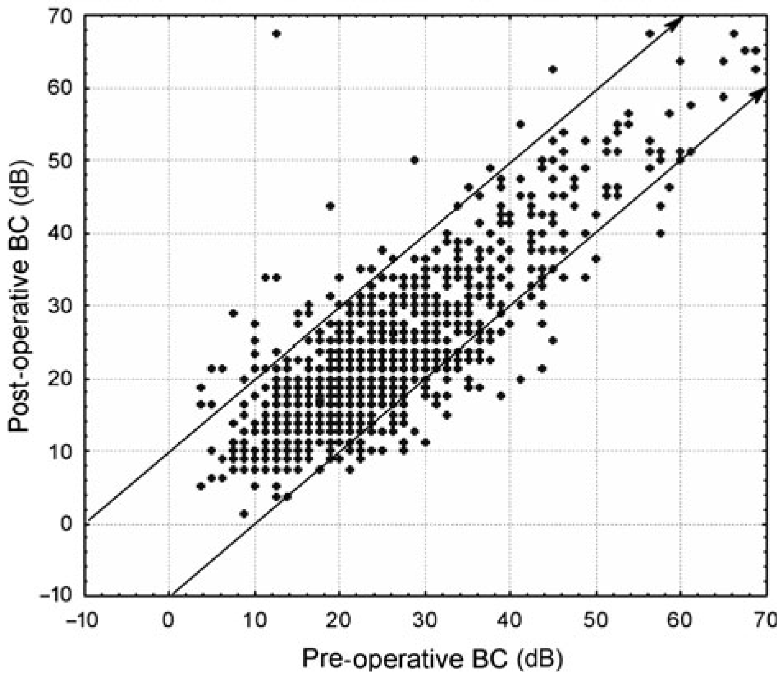


Figure 5. Amserdam hearing evaluation plot. Preoperative BC plotted against postoperative BC. BC - bone conduction

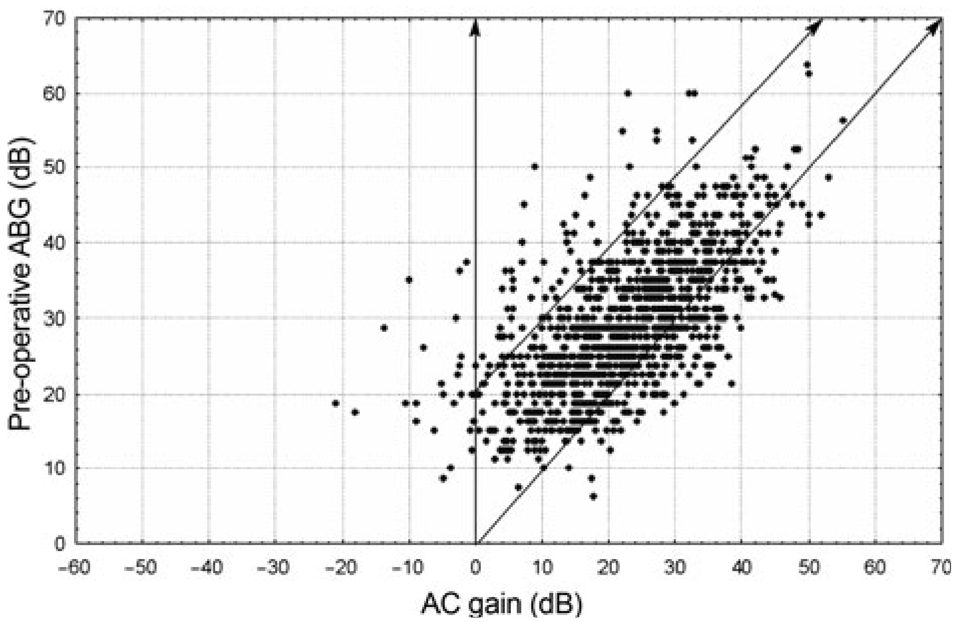


Figure 6. Amserdam hearing evaluation plot. Postoperative gain in AC plotted against the preoperative ABG. AC - air conduction; ABG - air-bone gap

Discussion

In the current era of evidence-based medicine, standardisation in reporting the outcomes of surgical intervention has become especially relevant. Adequate comparison of post-operative results requires uniform criteria of success.[13] Since the first reports of stapes surgery results, both the surgical technique and the audiometric criteria of success have undergone significant modification.[14,15]

In an attempt to avoid erroneous conclusions based on comparisons of different audiological parameters, two levels of guidelines were recommended by the American Academy of Otolaryngology, Head and Neck Surgery committee on hearing and equilibrium in 1995: summary statistics and raw data.[4] The recommendation to report summary statistics in terms of mean, SD and range was based on the assumption that these parameters are normally distributed. Audiometric data, however, are often not normally distributed.[12] Reporting the raw data along with the summary statistics, therefore, aims to avoid errors in the interpretation of the effects of intervention and to allow more detailed presentation of individual cases.[7] The parameters considered as summary statistics include the post-operative ABG and the number of decibels of ABG closure (both considered measures of technical success), as well as the post-operative air conduction thresholds (which correlate better with clinical success). In order to standardise reporting of the post-operative ABG, the use of post-operative rather than preoperative bone conduction thresholds has been recommended. In the past, the ABG was calculated by subtraction of pre-operative bone conduction thresholds from post-operative air conduction thresholds. This practice resulted in an underestimation of the post-operative ABG because of the phenomenon of overclosure.[16,17] Another weakness of this method is the inability to recognise postoperative changes in bone conduction; therefore, worsening in bone conduction thresholds caused by surgical trauma was underreported.[18,19]

Post-operative improvement in bone conduction thresholds in patients with otosclerosis was first described by Carhart.[20] The cause of this phenomenon was hypothesised to be a reduction of ossicular chain inertia as a result of stapes fixation. The range of the ‘Carhart effect’ has been estimated by different authors as 5–10 dB at 0.5 kHz, 10–20 dB at 1 kHz, 15–30 dB at 2 kHz and 5–20 dB at 4 kHz.[21,22] Blayney et al.,[23] in their dynamic model of stapedectomy, demonstrated a maximal response to sound stimuli of between 1.7 and 2.5 kHz. Stenfelt[24] recently investigated middle-ear ossicular motion with air and bone conduction stimulation, and also found that the inertia of the ossicular chain contributes to the perception of bone-conducted sound mostly between 1.5 and 3.5 kHz.

Bone conduction at 4 kHz seems to be less influenced by the inertia of the ossicular chain, and represents the highest frequency reliably measured by current audiometric equipment. These findings of middle-ear acoustic transfer provide the experimental basis for the frequencies chosen when reporting the results of stapes surgery. Improvement of bone conduction thresholds at 0.5, 1 and 2 kHz may be presumed to be influenced by the Carhart effect, and unchanged bone conduction at 4 kHz can be considered as primarily reflecting the effect of restored ossicular chain mobility, while worsening of bone conduction thresholds indicates sequelae of surgical trauma to the inner ear. Smyth and Hassard,[25] in their analysis of 18 years' experience in stapedectomy, found that success in ABG closure was highly frequency-dependent, being most prominent at low frequencies and disappearing at 4 kHz. In our series, bone conduction thresholds improved statistically significantly at 0.5, 1, 2 and 3 kHz, and worsened slightly (but statistically insignificantly) at 4 kHz.

These results are in agreement with those of other recently published, large series.[26,27] Other reports also found worsening of bone conduction thresholds following stapes surgery at 4 kHz.[28,29] Therefore, reporting changes in bone conduction at 4 kHz allows inter-series comparison of the degree of surgical trauma sustained.

Bone conduction thresholds based on averages might bias interpretation of surgical outcomes. In our study, detailed analysis of hearing results at 4 kHz demonstrated substantial differences between improvement rates for air conduction and bone conduction (Figure 3). Similar changes in pre- and post-operative bone conduction at 4 kHz were also reported by Coker et al.[30] Although the inclusion of 4 kHz data when calculating the PTA has been demonstrated to negatively affect success rates,[27] our results suggest that inclusion of high frequencies in PTA calculation, and reporting peri-operative changes in both air and bone conduction at all frequencies from 0.5 to 4 kHz, enables more accurate comparison of success rates and interpretation of reported outcomes.

The choice of frequencies for the calculation of PTA and ABG has been extensively debated. Historically, only the lower frequencies of 0.5, 1 and 2 kHz were used, because: (1) the ABG found in otosclerosis is most prominent at these frequencies; (2) these frequencies were considered the most important speech frequencies; and (3) the audiometer had technical limitations in achieving bone masking.[31,32]

Recognition of the importance of the higher frequencies for speech comprehension resulted in the American Academy of Otolaryngology, Head and Neck Surgery committee on hearing and equilibrium recommending, in 1995, the inclusion of 3 kHz for the calculation of air conduction and bone conduction PTAs.

This committee recognised the method of direct comparison between PTAs calculated using three (0.5, 1 and 2 kHz) or four (0.5, 1, 2 and 4 kHz) frequencies and PTAs calculated using 0.5, 1, 2 and 3 kHz, based on the findings of Goldenberg and Berliner.[33] Monsell,[14] referring to the same publication, also

concluded that the difference between averages of the three frequencies versus averages of the four frequencies would not significantly alter the results.
It is important to mention that Goldenberg and Berliner's study assessed the results of ossicular reconstructions and paediatric tympanoplasties, while audiometric data on bone conduction were available at all frequencies for only 228 patients. The difference between pre- and post-operative bone conduction thresholds in these authors' series was not statistically significant. Accordingly, the authors concluded that the definition of criteria for success had the greatest effect on the success rates of surgery, compared with the choice of frequency combinations. In a study which evaluated audiological data from 240 cases of stapes surgery, Berliner et al.[27] found the differences between pre- and postoperative bone conduction thresholds to be statistically significant at all frequencies, with the exception of 4 kHz. Inclusion of 4 kHz instead of 3 kHz in the ABG calculation resulted in lower rates of success (defined as an ABG closure within 10 dB). The difference between tympanoplasty and stapes surgery which involved inner-ear opening was suggested as an explanation for these contrasting findings. Similar results are demonstrated in our study in a larger group of patients: ABG closure was achieved in 937 (82 per cent) patients using a PTA calculated at 0.5–2 kHz, and in 861 (75 per cent) patients using a PTA calculated at 0.5–4 kHz. Therefore, comparisons of data sets using 0.5, 1 and 2 kHz or 0.5, 1, 2 and 3 kHz with data sets using 0.5, 1, 2 and 4 kHz appears to be more appropriate for evaluation of the results of tympanoplasty rather than stapes surgery.
The percentage of patients who demonstrate postoperative ABG closure to within 10 dB is most commonly used for reporting the technical success of stapes surgery. A review of recent studies that included higher frequencies for ABG calculation demonstrated that ABG closure to within 10 dB was achieved in 62 to 94 per cent of patients (Table III).[7,29,34,35]

Table 3. Results for ABG closure to within 10 dB in recent, comparable studies

Study	Pts (n)	ABG $\leq$ 10 dB (%)	Year
Vincent et al.[29]	3050	94	2006
Quaranta et al.[34]	2134	85	2005
Current study	1145	82	2009
De Bruijn et al.[26]	473	71	2001
Berliner et al.[27]	240	68	1996
Banjerjee et al.[35]	100	62	2002

It appears that the number of cases in any series affects the success rate. Several studies have demonstrated the existence of a learning curve for stapes surgery.[36–

[38] Independent reports by Hughes[39] and Yung et al.[40] found that 50 to 80 cases were required in order to achieve an ABG of 10 dB or better in 90 per cent of patients, and that rates of inner-ear damage appeared proportionally higher in their first 50 cases. A higher success rate reported in a larger series supports these findings. Therefore, ensuring that study numbers are appropriately large contributes to adequate interpretation of surgical outcomes, providing that the results were calculated for sequential data with no exclusion of unfavourable cases.

When summary statistics are presented in terms of means, the individual cases with poor outcomes have less influence on overall results.[41] Presentation of audiometric results using Amsterdam hearing evaluation plots enables the display of each individual stapedotomy outcome in a large series, with visual identification of successful and unsuccessful results. Schematically, every point below the upper diagonal line represents a successful outcome, and every point above this line represents an unfavourable result. As shown in Figure 6, the majority of the points are below the upper diagonal line representing 1052 (97.4 per cent) patients with a post-operative ABG within 20 dB. An improvement in post-operative bone conduction of more than 10 dB occurred in 57 of these cases (5 per cent) and is shown below the lower diagonal line. These results are similar to those reported by Vincent et al.,[30] who found overclosure in 98 out of 1672 (4 per cent) patients. The poor hearing outcomes indicated by either a post-operative bone conduction worsened by more than 10 dB (seen in 35 (3 per cent) patients), (figure 5) or a post-operative ABG of 20 dB or more (figure 6) (seen in 30 (2.6 per cent) patients), or both, are represented by points above the upper diagonal line. Different indicators of unfavourable outcomes, such as deterioration of bone conduction thresholds, negative changes in air conduction thresholds or an ABG of 20 dB or more, can be recognised by the specific area on the plot, enabling more detailed communication of hearing results.

- This study reports the largest series of primary stapedotomies evaluated with Amsterdam hearing evaluation plots.
- The study demonstrates successful hearing results with a low rate of complications. These results are comparable to those of studies using the same audiometric criteria.
- When reporting pre- and post-operative air and bone conduction thresholds, the inclusion of higher frequencies is important to enable adequate evaluation of hearing results and surgical trauma.

The technical limitation of Amsterdam hearing evaluation plots appears to be the resolution of the figure while presenting large series. In the Amsterdam hearing evaluation plots presented in De Bruijn and colleagues' study,[7] audiometric data from 451 stapedotomies can be clearly visualised.
In our series of 1145 cases, overlapping of points inevitably occurred due to the large number of cases involved.

Conclusion

This study reports the largest series of primary stapedotomies evaluated with Amsterdam hearing evaluation plots to date. It demonstrates successful hearing results with a low rate of complications. These results are comparable to those of other reports which applied the same audiometric criteria. When reporting pre- and post-operative thresholds, the inclusion of higher frequencies for air and bone conduction measurement is important in order to enable adequate evaluation of hearing results and surgical trauma. The use of Amsterdam hearing evaluation plots to display otological data allows visual presentation of unfavourable outcomes and therefore enhanced recognition of such data, compared with presentation as summary statistics alone. Amsterdam hearing evaluation plots represent a valuable adjunct to standard methods of evaluation of hearing outcomes following middle-ear surgery.

References

1. Shea J, Sanabria F, Smyth G. Teflon piston operation for otosclerosis. Arch Otolaryngol 1962;76:516–21
2. Shea J. Forty years of stapes surgery. Am J Otol 1998;19: 52–5
3. Sheehy JL, Nelson RA, House HP. Stapes surgery at the Otologic Medical Group. Am J Otol 1979;1:22–6
4. Committee on Hearing and Equilibrium guidelines for the evaluation of results of treatment of conductive hearing loss. Otolaryngol Head Neck Surg 1995;113:186–7
5. Tange RA, De Bruijn AJG, Dreschler WA. Gold and Teflon in the oval window. A comparison of stapes prostheses. In: Rosowski JJ, Merchant SN, eds. The Function and Mechanics of Normal, Diseased and Reconstructed Middle Ears. Hague: Kugler, 2000;258–9
6. Tange RA, GrolmanW, DreschlerWA. Gold and titanium in the oval window: a comparison of two metal stapes prostheses. Otol Neurotol 2004;25:102–5
7. De Bruijn AJG, Tange RA, Dreschler WA. Efficacy of evaluation of audiometric results after stapes surgery in otosclerosis. A method for reporting results from individual cases. Otolaryngol Head Neck Surg 2001;124: 84–9
8. Browning GG, Gatehouse S, Swan IRC. The Glasgow Benefit Plot: a new method for reporting benefits from middle ear surgery. Laryngoscope 1991;101:180–5
9. Smyth GDL. Immediate and delayed alterations in cochlear function following stapedectomy. Otolaryngol Clin North Am 1978;11:105–12
10. Smyth GDL. Practical suggestions on stapedotomy. Laryngoscope 1982;92:952–3
11. Fish U. Stapedotomy vs. stapedectomy. Am J Otol 1982;4: 112–17

12. Govartes PJ, Somers T, Offeciers FE. Box and whisker plots for graphic presentation of audiometric results of conductive hearing loss treatment. Otolaryngol Head Neck Surg 1998;118:892–5
13. Jackler RK. Comparability in reporting outcomes: a scientific imperative. Am J Otol 1996; 17:811–12
14. Monsell EM. New and revised reporting guidelines from the Committee on Hearing and Equilibrium. Otolaryngol Head Neck Surg 1995;113:176–8
15. Jahnke KP. Thoughts on the validity of the evaluation of methods in stapes surgery. In: Jahnke KP, ed. Middle Ear Surgery: Recent Advances and Future Directions. Stuttgart: Thieme, 2000;131–32
16. Sakai M. Proposal of a guideline in reporting hearing results in middle ear and mastoid surgery. Am J Otol 1994;15:291–3
17. Rosowski JJ, Merchant SN. Mechanical and acoustic analysis of middle ear reconstruction. Am J Otol 1995;16: 486–97
18. Moscillo L, Imperiali M, Carra P, Catapano F, Motta G. Bone conduction variation poststapedotomy. Am J Otolaryngol 2006;27:330–3
19. Monsell EM. Results and outcomes in ossiculoplasty. Otolaryngol Clin North Am 1994;27:835–40
20. Carhart R. Clinical applications of bone conduction audiometry. Arch Otolaryngol 1950;51:798–808
21. Austin D. Acoustic mechanisms in middle ear sound transfer. Otolaryngol Clin North Am 1994;27:641–54
22. Vartianen E, Karjalainen S. Bone conduction thresholds in patients with otosclerosis. Am J Otolaryngol 1992;134: 234–6
23. Blayney AW, Williams KR, Rice HJ. A dynamic and harmonic damped finite element analysis model of stapedotomy. Act Otolaryngol (Stockh) 1997;117:269–73
24. Stenfelt S. Middle ear ossicles motion at hearing thresholds with air conduction and bone conduction stimulation. J Acoust Soc Am 2006;119:2848–58
25. Smyth GDL, Hassard TH. Eighteen years experience in stapedectomy. Ann Otol Rhinol Laryngol (Suppl 5–6) 1978;87:3–36
26. De Bruijn AJG, Tange RA, Dreschler WA. Efficacy of evaluation of audiometric results after stapes surgery in otosclerosis. The effects of using different audiologic parameters and criteria on success rates. Otolaryngol Head Neck Surg 2001;124:76–83
27. Berliner KI, Doyle KJ, Goldenberg RA. Reporting operative hearing results in stapes surgery: does choice of outcome measure make a difference? Am J Otol 1996;17: 214–20
28. Perez-Lazaro JJ, Urquiza R, Cabrera A, Guerrero C, Navarro E. Effectiveness assessment of otosclerosis surgery. Acta Oto Laryngol 2005;125:935–45
29. Vincent R, Sperling NM, Oates J, Jindal M. Surgical findings and long-term hearing results in 3050 stapedotomies for primary otosclerosis: a prospective study with the otology-neurotology database. Otol Neurotol 2006; 27(suppl 2):25–47
30. Coker NJ, Duncan NO, Wright GL, Jenkins HA, Alford BR. Stapedectomy trends for the resident. Ann Otol Rhinol Laryngol 1988;97:109–13
31. Hannley MT. Audiologic characteristics of the patiënt with otosclerosis. Otolaryngol Clin North Am 1993;26: 373–87
32. Smith JB, Sullivan JA, McAskile K. The fenestration operation using minimal irrigation. Laryngoscope 1957; LXVII (suppl 7):643–60
33. Goldenberg RA, Berliner KI. Reporting operative hearing results: does choice of outcome measure make a difference? Am J Otol 1995;16:128–35
34. Quaranta N, Besozzi G, Fallacara RA, Quaranta A. Air and bone conduction change after stapedotomy and partial stapedectomy for otosclerosis. Otolaryngol Head Neck Surg 2005;133:116–20
35. Banerjee A, Hawthorne MR, Flood LM, Martin FW. Audit of stapedectomy results in a district general hospital. Clin Otolaryngol 2002;27:275–8
36. Backous DD, Coker NJ, Jenkins HA. Prospective study of resident-performed stapedectomy. Am J Otol 1993;14: 451–4
37. Burns JA, Lambert PR. Stapedectomy in residency training. Am J Otol 1996;17:210–13
38. Mathews SB, Rasgon BM, Byl FM. Stapes surgery in residency training program. Laryngoscope 2006;109: 52–3
39. Hughes GB. The learning curve in stapes surgery. Laryngoscope 1991;101:1280–4

40. Yung MW, Oates J, Vowler SL. The learning curve in stapes surgery and its implication to training. Laryngoscope 2006;116:67–71
41. Conrad GJ. “Collective stapedectomy” (an approach to the numbers problem). J Laryngol Otol 1990;104: 390–3

2.2

Bilateral hearing results of 751 unilateral stapedotomies evaluated with the Glasgow Benefit Plot

V.E. Kisilevsky
N.A. Bailie
J.J. Halik

The Journal of Laryngology & Otology, 2009, accepted for publication

Abstract

Aims

To evaluate bilateral hearing function after primary unilateral stapedotomy according to the 1995 American Academy of Otolaryngology, Head and Neck Surgery (AAO-HNS) guidelines and the Glasgow Benefit Plot (GBP). To analyze the effect of types of preoperative hearing impairment on postoperative hearing after stapedotomy.

Study design

Retrospective chart review.

Methods

The charts of 1369 stapedotomies performed by senior author (J.H.) from 1991 to 2006 were reviewed. 751 cases of primary unilateral stapedotomy were included. The hearing results were evaluated according to the 1995 AAO-HNS Committee on Hearing and Equilibrium guidelines and the Glasgow Benefit Plot criteria. Subgroups with preoperative unilateral, bilateral symmetric and bilateral asymmetric hearing loss were separately analysed.

Results

The most successful results in achievement of bilateral socially serviceable hearing were demonstrated in the group with unilateral hearing loss with 78% of patients having postoperative normal hearing. The achievement of bilateral socially serviceable hearing is highly correlated with the type of preoperative hearing impairment (r=0.74). A normal postoperative hearing levels are correlated with preoperative bone conduction (r=0.61).

Conclusion

This study represents the largest series of primary stapedotomies evaluated with the GBP. The bilateral postoperative hearing function depends on the type of preoperative hearing impairment. Preoperative bone conduction thresholds corrected for Carhart’s effect are useful for the prognosis of achievable post-operative air conduction.

Introduction

Since the introduction of the stapedectomy technique by Shea in 1956, the air-bone gap (ABG) closure and the improvement of air conduction (AC) thresholds in the operated ear were considered as main measures of success. Publications from early stapedectomy series reported success rates of more than 90 percent for ABG closure within 10 dB.[1,2,3] The aforementioned reports assured recognition of stapes surgery as a treatment of choice for the conductive hearing loss in otosclerosis and contributed to high expectation rate these patients. With the experience gathered over the three decades of modern stapes surgery, questions about the functional benefit of this procedure to patients started to arise. In 1985 Gordon Smyth[4] was the first otologist to question whether the patients and surgeons agreed on the benefits of stapedectomy. Smyth mentioned that the technical success of surgery did not necessarily translate to the improvement of patients' auditory status. Postoperative AC thresholds below 30 dB and an interaural difference less than 15dB were found to be more significant for patient satisfaction than the degree of ABG closure. Based on these findings, Toner and Smyth[5] coined the "15/30 dB rule of thumb" collectively known as the "Belfast rule of thumb" method for evaluation of the bilateral postoperative hearing. In 1991 Browning[6] suggested a new method for reporting benefits from middle ear surgery, based on plotting the preoperative and postoperative AC thresholds in both operated and non-operated ears. The proposed method, called the Glasgow Benefit Plot (GBP), employs patient groupings into the three major categories according to their preoperative hearing impairment (HI): unilateral (UHI), bilateral asymmetric (BAHI) and bilateral symmetric (BSHI) (Figure 1). The achievable aims of surgery are different in each group; therefore presentation of combined results would be misleading. Meaningful comparisons between different series are possible only if audiometric data of each group are evaluated separately.[6,7,8]

Bilateral hearing status remains rarely discussed in the reports on stapes surgery.[9] Our search of the indexed medical literature in Medline® and PubMed® databases demonstrated that among 3412 publications on the surgical treatment of otosclerosis to date, binaural hearing function as a keyword was used in only 83 (2%). The hearing in the non-operated ear was purposefully evaluated only in 21 out of these 83 studies; the remaining 62 publications were primarily focused on the results of bilateral surgeries.

In scarce publications that used the GBP for presenting results of middle ear surgery there is a contradicting distribution of surgical success among the groups.

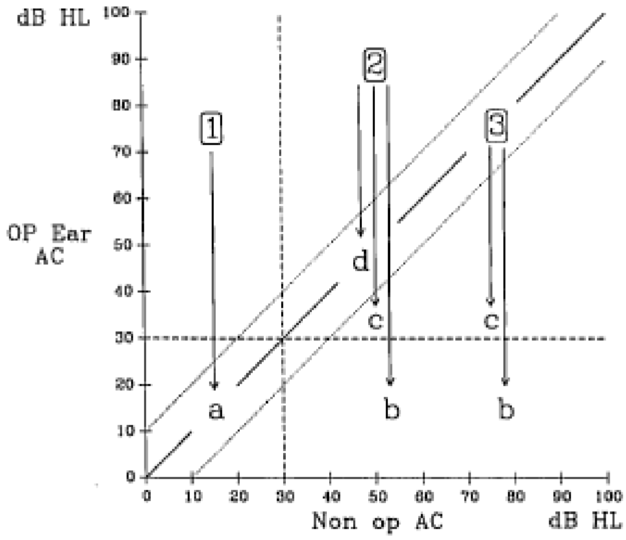


Figure 1. Potential changes from preoperative impairment group to postoperative impairment category. Group 1- unilateral HL, group 2- bilateral asymmetric HL, group 3- bilateral symmetric HL. Category a - bilateral serviceable hearing, b – unilateral serviceable hearing, c – bilateral asymmetric hearing impairment with operated ear converted to the better hearing ear, d – bilateral symmetric hearing impairment.

The adding up unilateral and bilateral cases, as well as results of primary and revision surgeries, or reporting results based on averages for whole series might be the sources of these inconsistencies. In this study we included only unilateral primary stapedotomy cases evaluating separately the three preoperative categories of UHI, BAHI and BSHI to avoid discrepancy. The results of 394 bilateral stapedotomies are analyzed in a separate study.

The aims of this study are twofold: a) to evaluate both technical and functional results of a large series of stapedotomies using the GBP and the AAO-HNS Committee on Hearing and Equilibrium guidelines[10] for reporting results of treatment of CHL, and b) to analyze the effect of types of preoperative hearing impairment on postoperative hearing levels after stapedotomy.

Patients and Methods

The charts of 1369 consecutive stapedotomies performed by one surgeon (senior author J.H.) from 1991 to 2006 were reviewed. Of the 1369 surgical cases, 89 were revision stapedotomies and 46 were performed for congenital stapes fixation. Eighty nine cases were excluded from the study due to incomplete data. The 1145 operations were primary stapedotomies in patients with otosclerosis. The audio-metric results of 751 unilateral stapedotomies were included in this study. 437

patients (58%) were women and 314 (42%) were men. The mean age was 46 years (SD 12 years).

The mean preoperative hearing levels calculated as a pure tone average (PTA) at frequencies 0.5, 1, 2 and 3 kHz was 54 dB (SD 14 dB), mean preoperative ABG was 29 dB (SD 9 dB). One hundred forty four (19%) patients were treated for severe or profound hearing loss (defined as preoperative hearing levels ≥ 71 dB).

Stapedotomy was performed using the reverse order technique[11,12] via a per-meatal approach. A modified Cawthorne (0.3mm diameter) prosthesis was used until 2002 in 409 cases. Thereafter, a modified Causse (0.4 mm diameter) prosthesis was used in 342 procedures. A 0.4 mm (for Cawthorne) or 0.5 mm (for Causse) fenestra was made in the footplate using Halik graduated perforators. Adjunctive use of the Argon beam laser (Lumenis Inc, Salt Lake City, Utah, USA), delivered via an EndoOtoprobe™ (Iridex Corp, Mountain View, California, USA), was introduced in 2000. The laser assisted procedures were performed on 343 patients. A modification of the reverse order technique was made in conjunction with the introduction of the laser: the Argon laser was used to vaporize the stapedial tendon and posterior crus of the stapes prior commencing the footplate fenestration. The laser was also used to initiate the footplate fenestration by providing a ‘set-hole’ for the perforators. The fenestration was completed with the graduated perforators.

The hearing outcomes were analyzed according to the 1995 AAO-HNS Committee on Hearing and Equilibrium guidelines.[10] Frequency specific AC thresholds at 0.5, 1, 2, 3, 4, 6, 8 and 12 kHz and bone conduction (BC) thresholds at 0.5, 1, 2, 3 and 4 kHz in operated and non-operated ears were tabulated in a computer database. As the beginning of this study period pre-dates the 1995 AAO-HNS guidelines, 3 kHz results were not available for all patients. In these cases 3 kHz AC and BC was estimated by average between 2 and 4 kHz allowing a uniform presentation of audiometric data in compliance with AAO-HNS guidelines. The mean preoperative PTA of AC, BC and ABG at 0.5, 1, 2 and 3 kHz were compared with corresponding postoperative values. Postoperative BC values were used to calculate postoperative ABG. In addition to reporting the results as mean values of AC, BC, ABG and AC gain, we utilized the GBP for visual presentation of bilateral hearing status in each individual case. Normal hearing was defined as an AC threshold ≤ 30 dB according to the original description by Browning[11] who adopted the “Belfast Rule of Thumb” for definition of socially acceptable hearing.

Symmetrical hearing was defined as the interaural difference between the operated and non-operated ear ≤ 10 dB as proposed by the GBP method. Based on these criteria, patients were grouped into three major categories according to their preoperative hearing impairment: group 1 (481 patients)-unilateral hearing impairment (UHI), group 2 (166 patients) - bilateral asymmetric hearing impairment (BAHI) and group 3 (104 patients)-bilateral symmetric hearing impairment (BSHI). Pre- and postoperative AC PTAs in operated ear were plotted on the vertical axis against corresponding AC thresholds in non-operated ear on the horizontal axis.

The database was analyzed with a statistical software program *STATISTICA 6.0*, StatSoft, Inc., Tulsa, OK, USA. Statistical analysis was carried out for comparisons between pre- and postoperative values using paired t-test for the evaluation of AC, BC, PTA, ABG, AC gain. The effects of type of preoperative hearing impairment and preoperative BC on achievement of postoperative symmetric and socially acceptable hearing of correlation were evaluated with Pearson test of correlation. The criterion selected for statistical significance was $p < 0.05$.

Results

Conventional Methods of Assessing Surgical Success

I. Closure of air-bone gap

Pre- and postoperative ABG for all the series, percentage of ABG closure for all the series, and separately for UHI, BAHI and BSHI are summarized in Table 1. A mean postoperative ABG of 7.5 dB was achieved in each group despite preoperative differences between the groups with the largest preoperative ABG of 32 dB in Group 2.

Table 1. Closure of air-bone gap

ABG	All unilateral n=751	Group 1 n=481	Group 2 n=166	Group 3 n=104
≤ 10 dB	602 (80%)	391 (82%)	134 (81%)	71 (74%)
≤ 20 dB	728 (97%)	469 (98%)	160 (96%)	99 (95%)
Preop	29(SD 9)	28 (SD 9)	32 (SD 10)	28 (SD 9)
Postop	7 (SD 6)	7 (SD 5)	8 (SD 6)	8 (SD 7)

ABG - air-bone gap, dB - decibel, SD - standard deviation,
Group 1 - unilateral hearing impairment,
Group 2 - bilateral asymmetric hearing impairment,
Group 3 - bilateral symmetric hearing impairment.

II. Improvement in air conduction thresholds

Postoperatively, significant improvement of AC thresholds was demonstrated in all groups ($p<0.001$). The group with BAHI had the most severe preoperative hearing loss, with average AC thresholds of 65 dB in operated ear and 40 dB in non-operated ear. Postoperative AC thresholds in this group were also poorer than in the UHI and BSHI groups, despite a greater AC gain of 27 dB, compared with 24 dB and 21 dB in UHI and BSHI groups, respectively (Table 2).

Table 2. Improvement in air-conduction (AC) PTAs (at 0.5, 1, 2 and 3 kHz).

AC	All unilateral	Group 1	Group 2	Group 3
	N=751	N=481	N=166	N=104
Preop dB	54 (SD 14)	50 (SD 12)	65 (SD 15)	54 (SD 14)
Postop dB	29 (SD 13)	26 (SD 10)	38 (SD 15)	33 (SD 14)
Gain dB	25 (SD 12)	24 (SD 11)	27 (SD 12)	21 (SD 12)
Unoperated ear	26 (SD 18)	13 (SD 8)	40 (SD 10)	53 (SD 15)

n - number of patients, dB - decibel, SD - standard deviation

III. Preoperative and postoperative BC

Preoperative BC thresholds were analyzed for identification of cases for which normal hearing is achievable. The highest proportion of normal cochlear reserve was found in UHI group in 411 (85%) of cases. The highest proportion of mixed hearing loss was found in BAHI group: 123 (74%) patients had BC thresholds ≥ 25 dB (Table 3).

Table 3. Pre- and postoperative bone conduction (BC) PTAs (at 0.5, 1, 2 and 3 kHz).

Preoperative BC	All unilateral	Group 1	Group 2	Group 3
	N=751	N=481	N=166	N=104
≤ 30 dB	567 (75%)	411 (85%)	81 (49%)	75 (72%)
≥ 25 dB	293 (39%)	123 (26%)	123 (74%)	47 (45%)
Preoperative dB	25 (SD 10)	22 (SD 8)	33 (SD 11)	26 (SD 10)
Postoperative dB	22 (SD 11)	20 (SD 8)	30 (SD 13)	25 (SD 11)

IV. The proportion of postoperative normal hearing is presented in Table 4. Analysis of correlation (Pearson test) have demonstrated that normal postoperative AC (Table 4) was found to be highly correlated withnormal preoperative BC (Table 3) , Pearson correlation coefficient r=0.7.

V. The distribution of postoperative symmetrical hearing is demonstrated in Table 5.
The type of preoperative hearing impairment was found to be highly correlated with postoperative symmetric hearing, Pearson correlation coefficient r =0.75.

Table 4. The proportion of postoperative normal hearing.

Postoperative AC	All unilateral	Group 1	Group 2	Group 3
	N=751	N=481	N=166	N=104
≤ 30 DB	507 (68%)	377 (78%)	71 (43%)	49 (57%)
% of achievable (pre BC ≤ 30 dB)	89%	92%	88%	65%

Table 5. Postoperative bilateral symmetrical hearing.

Interaural difference	All	Group 1	Group 2	Group 3
	N=751	N=481	N=166	N=104
Operated become better hearing ear	311 (41%)	106 (22%)	105 (63%)	100 (96%)
∆ ≤ 10 dB	351 (47%)	220 (46%)	91 (55%)	19 (18%)
∆ ≤ 15 dB	507 (68%)	262 (55%)	127 (77%)	37 (36%)

Glasgow Benefit Plot

Figures 2, 3 and 4 demonstrate the preoperative and postoperative AC PTA for the three preoperative hearing impairment groups.

1. *Postoperative normal hearing*
Postoperative AC PTA at 0.5,1,2 and 3 kHz less than 30 dB in operated ear was achieved in 507(68%) of all unilateral stapedotomies. It comprises 89% of patients with preoperative BC thresholds ≤ 30 dB allowing achievable serviceable hearing. Best results were demonstrated in the group with

unilateral hearing impairment (group 1). The less successful results for normal hearing levels were found in the group with bilateral asymmetric hearing impairment (group 2) (Table 4).

2. *Postoperative symmetrical hearing*

Postoperative bilateral hearing PTAs are summarized in Table 5. The operated ear became the better hearing ear in 41% of cases in this series but became the better hearing ear in 96% of the subgroup of patients with preoperative bilateral symmetric hearing impairment. Symmetrical hearing was obtained in 47% of all patients. For comparison, if the inter-aural difference of 15 dB or less is regarded as symmetrical hearing it was achieved in 68% of all patients. The most striking difference was found in BSHI group -, where a two-fold increase in postoperative symmetrical hearing was observed., In UHI group - postoperative hearing is found to be symmetric in only 55%; at the same time it is bilaterally normal in 78% of cases (Table 4). This apparent discrepancy can be explained by the fact that the mean AC PTA for the non-operated ear in this group was 13 dB(Table 2), therefore patients with postoperative hearing thresholds of 28< PTA AC≤30 dB have bilateral normal hearing despite an interaural difference formally exceeding 15 dB.

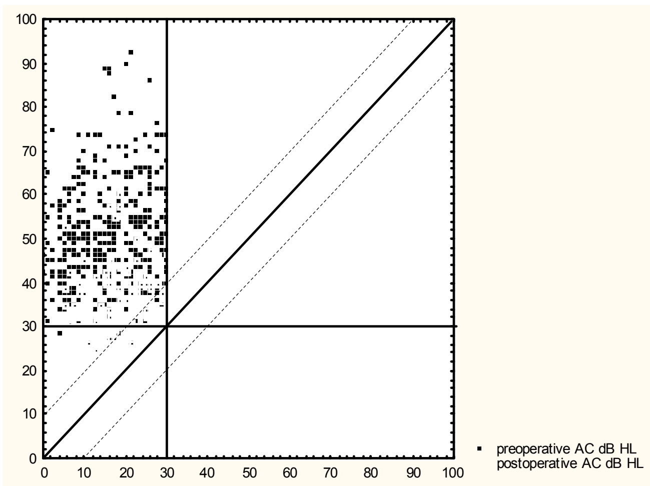


Figure 2. Results of preoperative group 1 (n = 481)
(Unilateral impairment with asymmetric thresholds).

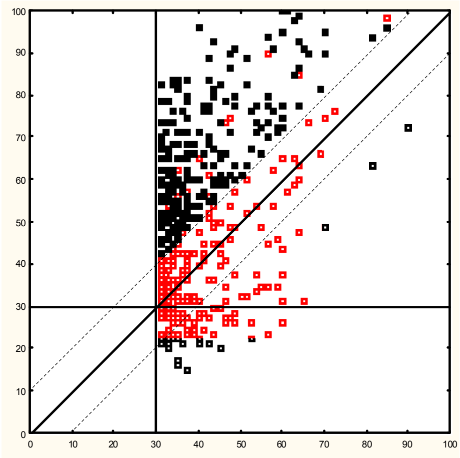


Figure 3. Results of preoperative group 2 (n =166)
(Bilateral impairment with asymmetric thresholds).

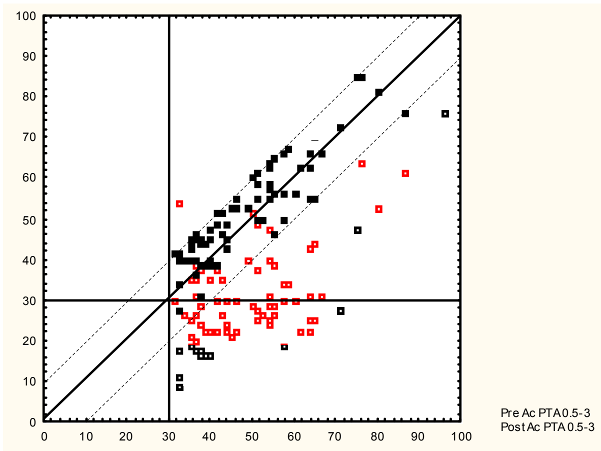


Figure 4. Results of preoperative group 3 (n =104)
(Bilateral impairment with symmetric thresholds).

Discussion

Stapes surgery is generally considered the most successful middle ear procedure for correction of a conductive hearing loss. This reputation is based on publications reporting stapedectomy success rates approaching 85-95% of cases.[1-3,13-15] In these studies, success is defined as the improvement of AC thresholds and ABG closure in the operated ear. This technical approach to the assessment of surgical outcomes does not consider overall hearing status based on bilateral auditory input. The GBP, created with the patient's bilateral hearing in mind, is considered a functional method for reporting benefits of middle ear surgery. This study employs the GBP in addition to technical methods for evaluation the results of stapedotomy. Preoperative and postoperative BC and AC

PTAs for both the operated and non-operated ear were calculated to evaluate the effect of types of preoperative hearing impairment on surgical success, as defined by GBP criteria for normal and symmetrical hearing.

To date, the GBP has been utilised in 19 studies indexed in Medline® and PubMed® database. Ten publications employed the GBP to analyse surgical success in chronic otitis media, tympanosclerosis, and otitis media with effusion. Nine studies used the GBP to investigate stapes surgery. The results of unilateral procedures were evaluated in four reports,[16-19] and results of bilateral surgeries in three.[20-22] Combined data from unilateral and bilateral cases were presented in one study;[23] and one publication discussed revision stapedectomy.[24]
The outcome of revision stapes surgery is usually less successful than that of primary stapedectomy.[25-27] In the case of bilateral disease, it is customary to operate first on the ear with worse hearing thresholds. Therefore, the chances for bilateral normal hearing in unilateral and bilateral surgery are substantially different and functional results between primary and revision, or unilateral and bilateral procedures cannot be compared directly. As Browning emphasised in his original paper[6], reporting the average results of an entire surgical series would make it impossible to calculate the differences between different patient populations within the series. Despite this, some authors use the GBP to evaluate the success of mixed series, thereby combining overall results of different patient populations. In order to circumvent the ambiguity that may arise from this method of evaluation, the present study considered outcomes of each preoperative group separately. This method is consistent with Browning's position, and uses the GBP as it was originally designed to be applied.

Technical results of previous unilateral stapedotomy series, assessed by conventional means, demonstrated ABG closure within 10 dB in 50% of cases in Browning's series[6], 65% in Huangs' series[16], and 74% of 509 cases reported by Persson et al.[23]. The recovery of AC PTAs demonstrated less variability: an AC gain of 28 dB was found by Browning[6], 27 dB by Persson[23], 29 dB by Huang[16], and 24 dB by Caces[19]. Results from this study indicate closure of the ABG within 10 dB in 80% of cases, and an AC PTA gain of 25 dB (SD 12 dB); which is equal or superior to other series evaluated by the same criteria. ABG closure within 10 dB was demonstrated in 82% of cases in UHI group, in 81% in BAHI group and in 74% in BSHI group (Table 1). Postoperative AC PTAs were better in UHI group (26 dB, SD 10 dB) than in BAHI group (38 dB, SD 15 dB) and BSHI group (33 dB, SD 14 dB)(Table 2). As was previously reported, clinical improvement correlates better with postoperative AC levels than with postoperative ABG values[28]. The present study establishes that the probability of normal postoperative AC thresholds is directly related to the type of preoperative hearing impairment, as defined by the GBP criteria. Superior outcomes are demonstrated in UHIgroup, in which 78% of patients achieved normal hearing in the operated ear. Forty three percent of BAHIgroup patients achieved postoperative thresholds better than 30 dB. A similar distribution of success rates, as evaluated by the GBP, was found by Szymanski[17], with 79% and 46% postoperative normal hearing in UHI group and BAHI group, respectively. Our findings are also consistent with Persson[23], who found normal hearing in 70% of UHI group and only in 36% of BAHI group. Similarly, Lundman et al.[28] reported severe postoperative hearing disability in about one third of otosclerosis patients with bilateral hearing loss, despite good results from unilateral surgery. The preoperative AC thresholds and proportion of mixed hearing loss is highest in BAHI group, intermediate in BSHI group, and lowest in UHI group. Therefore, knowledge of success rates in different hearing impairment groups may be helpful in preoperative counselling.

Pre-operative BC thresholds are usually considered the limiting factor for the achievable postoperative AC improvement.[29] The postoperative changes of BC thresholds in otosclerosis were initially described by Carhart[30] and Davis[31.] More recent reports demonstrated better postoperative results in patients with good preoperative cochlear reserve.[32] Moscillo evaluated variation in bone conduction thresholds following stapedectomy, and found greater improvement in bone conduction thresholds in younger patients; he concluded that normal BC thresholds are a prerequisite for successful surgical outcomes.[33] Ramsay found also that a preoperative mixed hearing loss resulted more frequently in profound SNHL postoperatively.[34] However, the controversy of using preoperative BC for prognostic purposes remains. The 1995 AAO-HNS guidelines, which use post-operative BC thresholds for ABG calculation, have been criticized because BC thresholds can change unpredictably.[35] For example, BC thresholds could either improve due to Carhart's effect, as a result of successful mobilization of the ossicular chain, or worsen, as a result of surgical trauma.[36-39] Limitations may exist for bone conduction audiometry owing to difficulties with masking, especially in cases of severe bilateral conductive hearing impairment.[40]

Results of our study demonstrated the best preoperative and postoperative BC thresholds in UHI group and the worst parameters in BAHI group, which more likely will require postoperative hearing aids (Table 3). This is in agreement with the highest proportion of mixed hearing loss in BAHI group and the highest

proportion of postoperative normal hearing in UHI group. Therefore, the combination of quantifying the Carhart's effect based on preoperative BC[36] with known incidence of SNHL in this series allows for reasonably accurate prediction of postoperative results. Further investigation of the predictive value of preoperative BC thresholds, using the GBP, is ongoing in our department. This study will compare potentially achievable and actual results of stapedotomy.
Many authors have tried to identify prognostic factors for success in middle ear surgery by analyzing preoperative audiometric data and operative findings. Gerard[41] reported the outcomes of 147 primary stapedotomies, with ABG closure within 10 dB in 60% of cases, but did not detect any factor that was predictive for postoperative hearing thresholds. Blakley found improved outcomes in patients with less severe preoperative hearing loss, and concluded that preoperative hearing thresholds predict postoperative hearing.[42] Ueda[43] investigated the preoperative ABG, and determined that smaller preoperative ABG results in better postoperative ABG closure. Therefore, if technical success in the operated ear only is the only consideration, better preoperative AC thresholds and smaller ABGs are associated with more successful surgical outcomes.

The patients' estimation of the benefits of surgery may not always match that of the surgeon. For example, a patient with the most severe hearing loss may have the largest functional benefit from surgery. Browning concluded that the magnitude of AC threshold change is more important for patient satisfaction than achievement of normal hearing in the operated ear. Patients with preoperative bilateral hearing impairment report greater benefit than those with unilateral hearing impairment provided the hearing thresholds of the operated ear are better relative to the non-operated ear. Following the analysis based on patients' reports of surgical benefit, Browning removed the horizontal 30dB line from the modified GBP.[44] Further modification of the GBP was recently reported by Schmerber[22], who proposed the Quartile Benefit Plot (QBP). The QBP includes the additional criteria of ABG closure within 10 dB, and absence of postoperative SNHL; which provides a more sensitive measurement of both technical and functional benefits of stapes surgery. From an historical perspective, increasingly comprehensive assessments of audiometric outcomes resulted in shrinking success rates. From 85-95% in series reported by the "fathers of stapes surgery"[1-3,13-15], to 78% when evaluated by the "Belfast rule of thumb", to 62% when assessed with the GBP[5], and finally to 39% when calculated by the QBP.[22] The imposition of more stringent criteria for evaluation of surgical results leads to lower success rates.

The definitions of technical success used by current methods, however, are limited by focusing on achievement of certain number of decibels of hearing level in one or two ears. Evaluation of the benefit of treatment is more complex and additional methods concerning quality of life aspects are needed to provide more balanced final answers.[45] Ramsay noted that even patients with excellent audiometric results frequently presented with symptoms of noise intolerance and sound distortion, negatively affecting their subjective estimate of surgical benefit.[46] This observation is also supported by Hall, who demonstrated that some patients with normal pure-tone thresholds after stapedotomy continue to experience significant hearing disability, due to abnormal binaural hearing function.[47] A recent study on quality of sound following stapedotomy found that audiometric improvement does not always correlate with an improvement in sound perception.[48] The study also determined that patients with postoperative AC PTAs <30dB demonstrated better handicap scores than those with AC PTAs >30dB. This is in agreement with the results of other studies, which demonstrated that improvement of postoperative thresholds resulted in better speech recognition scores and, subsequently, in better QOL.[49,50] Therefore, using a patient's preoperative impairment to predict their outcome, allows for more patients expectations oriented counselling. For practical purposes, the combination of conventional methods and preoperative BC PTAs in the GBP provides a working tool for the reasonably accurate prediction of postoperative hearing. This combination of technical and functional criteria allows also meaningful comparison of surgical techniques, and contributes to improved quality of future research.

Conclusions

In this study the largest group of 751 primary unilateral stapedotomies was simultaneously evaluated according to 1995 AAO-HNS guidelines and the GBP criteria. The addition of the GBP to conventional parameters provides detailed presentation of bilateral hearing in each individual case and visual identification of successes and failures. The best functional results are demonstrated in the group of unilateral hearing impairment. Socially acceptable hearing can be achieved in 78% of patients with unilateral CHL. Patients with bilateral symmetric hearing loss are more likely to benefit from second ear surgery for achievement of bilaterally serviceable and symmetrical hearing. Patients with bilateral asymmetric mixed HL will more likely require postoperative rehabilitation with hearing aids. The bilateral postoperative hearing function depends on the type of preoperative hearing impairment. Preoperative BC thresholds corrected for Carhart's effect can be used for the prognosis of achievable postoperative AC thresholds. Prospective study of preoperative audiometric parameters as prognostic factors will be of value.

References

1. Sheehy JL, Nelson RA, House HP. Stapes surgery at the Otologic Medical Group. Am J Otol 1979;1:22-26
2. McGee TM. Comparison of small fenestra and total stapedectomy. Ann Otol Rhinol Laryngol 1981; 90: 630-832
3. Hough JVD. Otosclerosis: the method of JVD Hough, in Gates GA (ed): Curr Ther in Otolaryngol-H&N Surg 1982;83:24, NY BC Decker 1982
4. Smyth GDL, Patterson CC. Results of middle ear reconstruction: do patients and surgeons agree? Am J Otol 1985; 6: 276-279
5. Toner JG, Smyth GDL. Comparison of methods of evaluating hearing benefit of middle ear surgery. J Laryngol Otol 1993; 107: 4-5
6. Browning GG, Gatehouse S, Swan IR. The Glasgow Benefit Plot: a new method for reporting benefits from middle ear surgery. Laryngoscope 1991; 101: 180-185
7. Browning GG. Reporting the benefits from middle ear surgery using the Glasgow Benefit Plot. Am J Otol 1993; 12: 135-140
8. Browning GG. Choice, advice and assessment of patients for ear surgery. JR Soc Med 1996; 89:571-576
9. Smyth GDL. Facts and fantasies in modern otology; the ear doctor's dilemma. J Laryngol Otol 1992; 106:591-596
10. Committee on Hearing and Equilibrium guidelines for the evaluation of results of treatment of conductive hearing loss. Otolaryngol Head Neck Surg 1995;113:186-7
11. Smyth G.. Practical suggestions on stapedotomy. Laryngoscope 1982; 92-8:952-3
12. Fish U. Stapedotomy vs. stapedectomy. Am J Otol 1982;4:112-17
13. Shea J. Forty years of stapes surgery. Am J Otol 1998; 19:52-55
14. Marquet J. Stapedotomy technique and results. Am J Otol 1985; 6:63-67
15. Shea J, Sanabria F, Smyth G. Teflon Piston operation for otosclerosis. Arch Otolaryngol 1962; 76: 516-21
16. Huang SC, Stanley RE. Stapedectomy at the Singapore General Hospital-use of functional hearing analysis. Singapore Med J1995; 36:158-162
17. Szymanski M, Siwiec H, Golabek W. Glasgow Benefit Plot for evaluation of stapedectomy results. Otolaryngol Pol 2003; 57:403-406
18. Subramaniam K, Eikelboom RH, Marino R, Atlas MD, Rajan GP. Patient's quality of life and hearing outcomes after stapes surgery. Clin Otolaryngol 2006; 31: 273-279
19. Caces F, Braccini F. Comparative study using AAO-HNS guidelines for conductive hearing loss and Glasgow Benefit Plot to evaluate results of stapes surgery for otosclerosis. Rev Laryngol Otol Rhinol 2007; 128: 47-53
20. Porter MJ, Zeitoun H, Brookes GB. The Glasgow Benefit Plot used to access the effect of bilateral stapedectomy. Clin Otolaryngol1995; 20:68-71
21. De Bruijn AJ, Tange RA, Dreschler WA. Evaluation of second-ear stapedotomy with the Glasgow benefit plot. ORL J Otolaryngol Relat Spec 1999; 61: 92-7
22. Schmerber S, Karkas A, Righini CA, Chahine KA. The Quartile benefit plot: a middle ear surgery benefit assessment scheme. Laryngoscope 2008; 118:1-6
23. Persson P, Harder H, Magnuson B. Using the Glasgow benefit plot as a prognostic instrument and for preoperative counselling in patients with otosclerosis. Otol Neurotol 2007;28 : 739-44
24. Magliulo G, Cristofari P, Terranova G. Functional hearing results in revision stapes surgery. Am J Otol 1997;18:408-12
25. Glasscock ME, McKennan KX, Levine SC. Revision stapedectomy surgery. Otolaryngol H&N Surg 1987;96:141-48
26. Palva T, Ramsay H. Revision surgery for otosclerosis. Act Otolaryngol (Stockh) 1990; 110: 416-20
27. Gros A, Vatovec J, Zargi M, Jenko K. Success rate in revision stapes surgery for otosclerosis. Otol Neurotol 2005; 26:1143-48

28. Lundman L, Mendel L, Bagger-Sjoback D, Rosenhall U. Hearing in patients operated unilaterally for otosclerosis. Self-assessment of hearing and audiometric results. Acta Otolaryngol 1999; 119: 453-8
29. De Bruijn AJ, Tange RA, Dreschler WA. Efficacy of evaluation of audiometric results after stapes surgery in otosclerosis. The effects of using different audiologic parameters and criteria on success rates. Otolaryngol Head Neck Surg 2001; 124:76-83
30. Carhart R. Clinical application of bone conduction audiometry. Arch Otolaryngol 1950; 51: 798-808
31. Davis H, Silverman D. Hearing and deafness. 3rd ed. New York: Holt, Rinehart and Winston, 1970: 105
32. Satar B, Sen D, Karahatay S, Birkent H, Yetiser S. Effect of cochlear reserve on postoperative outcome in otosclerosis. Eur Arch Otolaryngol 2007; 264:489-93
33. Moscillo L, Imperiali M, Carra P, Catapano F, Motta G. Bone conduction variation poststapedotomy. Am J Otolaryngol 2006; 27: 330-33
34. Ramsay HA, Linthicum FH. Mixed hearing loss in otosclerosis: indication for long-term follow-up. Am J Otolaryngol Am J Otolaryngol 1994; 15:536-9
35. Black B. Reporting results in ossiculoplasty. Otol Neurotol 2000; 24: 534-42
36. Cook JA, Krishnan S, Fagan PA. Quantifying the Carhart effect in otosclerosis. Clin Otolaryngol 1995; 20: 258-61
37. Vartiainen E, Karjalainen S. Bone conduction thresholds in patients with otosclerosis. Am J Otolaryngol 1992; 13 : 234-36
38. Smyth GDL, Hassard TH. Eighteen years experience in stapedectomy. Ann Otol Rhinol Laryngol 1978: 87:3-36
39. Soudry E, Sulkes J, Attias J, Naegris BI.Bone conduction in otosclerosis-operated versus non-operated ears. J Basic Clin Physiol Pharmacol 2007; 18:189-99
40. Giles M, Browning GG, Gatehouse SG. Problems in assessing the audiogram in patients with severe hearing impairment. J Laryngol Otol 1999;110: 727-31
41. Gerard JM, Serry P, Gersdorff C. Outcome and lack of prognostic factors in stapes surgery. Otol Neurotol 2008; 29:290-4
42. Blakley BW,Kim S, VanCamp M. Preoperative hearing predicts postoperative hearing. Otolaryngol Head Neck Surg 1998; 119:559-63
43. Ueda H, Miyazawa T, Asahi K, Yanagita N. Factors affecting hearing results after stapes surgery. J Laryngol Otol 1999; 113: 417-21
44. Browning GG. Do patients and surgeons agree? The Gordon Smyth memorial lecture. Clin Otolaryngol 1997; 22: 485-96
45. Meyer SE, Megerian CA. Patient's perceived outcomes after stapedectomy for otosclerosis. Ear, Nose & Throat J 2000; 79: 846-56
46. Ramsay H, Karkkainen J, Palva T. Success in surgery for otosclerosis: hearing improvement and other indicators. Am J Otolaryngol 1997; 18: 2-28
47. Hall JW, Grose JH, Pillsbury HC. Predicting binaural hearing after stapedectomy from presurgery results. Arch Otolaryngol Head Neck Surg 1990; 116:946-50
48. Tan FM, Grolman W, Tange RA, Fokkens WJ. Quality of perceived sound after stapedotomy. Otolaryngol-Head Neck Surg 2007; 137:443-4949.
49. Lippy WH, Burkey JM, Arkis PN. Word recognition score changes after stapedectomy for far advanced otosclerosis. Am J Otol 1998; 19: 56-58
50. Stewart MG. Outcomes and patient-based hearing status in conductive hearing loss. Laryngoscope 2001; 111:1-21

2.3

Functional results of 394 bilateral stapedotomies evaluated with the Glasgow Benefit Plot

V. E. Kisilevsky
N.A. Bailie
S.N. Dutt
J.J. Halik

European Archives of Otorhinolaryngology, 2009, accepted for publication

Abstract

Aims

To evaluate results of 394 bilateral stapedotomies according to the American Academy of Otolaryngology - Head and Neck Surgery (AAO-HNS) guidelines and with the Glasgow Benefit Plot (GBP). To analyze the benefit of a second-ear stapedotomy in achievement of normal and symmetrical hearing.

Study design

Retrospective chart review.

Methods

The charts of 1369 stapedotomies performed by senior author (J.H.) from 1991 to 2006 were reviewed. Results of 394 bilateral stapedotomies were included. The hearing results were evaluated according to the 1995 AAO-HNS Committee on Hearing and Equilibrium (CHE) guidelines and the GBP criteria. Success rates after 1 st and 2 nd ear surgery were separately analysed.

Results

ABG closure ≤ 20dB was demonstrated in 98 % of cases after both 1st and 2nd procedure. Postoperative AC gain was higher in the 1st ear surgery compared with the 2nd ear surgery in all groups of preoperative hearing impairment. As a result of 1st ear surgery, 142 (72%) patients changed into the category of unilateral hearing loss. As a result of 2nd ear surgery, 125 (64%) patients changed into the category of normal and symmetric hearing.

Conclusion

n this study the largest group of 394 primary bilateral stapedotomies was simultaneously evaluated according to AAO-HNS guidelines and GBP criteria. 1st ear surgery was more successful in achievement of normal hearing in the operated ear. 2nd ear surgery was more beneficial in providing symmetric hearing.

Introduction

Bilateral stapes surgery has been a source of controversy since the early implementation of a stapedectomy technique. Abnormal histopathological findings following bilateral stapedectomy and stapes mobilization have been well documented.[1-3] Immediate postoperative deafness and/or delayed sensori-neural hearing loss (SNHL) remain the most feared complications.[4,5] The prospect for doubling the risk of these complications is amongst the arguments against second ear surgery. Detrimental effects of bilateral vestibular loss also added to the case against second ear stapedectomy.[6] While technical results of stapedectomy have been evaluated in numerous studies, functional benefits of bilateral intervention were rarely reported. To date, the Glasgow Benefit Plot (GBP)[7] has been employed in three studies which evaluated results of bilateral stapedotomies in small groups of patients.[8-10] The aims of the present study were 1) to evaluate the results of a large series of 394 bilateral stapedotomies according to the 1995 American Academy of Otolaryngology - Head and Neck Surgery (AAO-HNS) guidelines criteria[11] and with the GBP and 2) to assess the benefits of a second-ear stapedotomy in achievement of binaural and symmetrical hearing.

Patients and methods

The charts of 1369 consecutive stapedotomies performed by one surgeon (senior author J.H.) from 1991 to 2006 were reviewed. Of the 1369 surgical cases, the 1145 operations were primary stapedotomies in patients with otosclerosis. Forty-eight stapedotomies were performed for congenital stapes fixation and 89 were revision stapedotomies. Eighty- seven cases were excluded from the study due to incomplete charts. Results of 751 unilateral cases are reported in a separate paper.[12] The audiometric results of 394 stapedotomies (197 patients) were included in this study. One hundred twenty three patients (62%) were women and 74 (38%) were men. The mean age was 43 years (SD 11 years) at 1st ear surgery and 45 years (SD 10 years) at 2nd ear surgery.

Stapedotomy was performed using the reverse order technique via a per-meatal approach.[13,14] A modified Cawthorne (0.3mm diameter) prosthesis was used until 2002 in 133 cases in 1st ear and in 95 in 2nd ear. Thereafter, a modified Causse (0.4 mm diameter) prosthesis was used in 60 procedures in 1st ear and in 98 procedures in 2nd ear. In eight cases different prostheses, other than Cawthorne or Causse, were used. A 0.4 mm (for the Cawthorne) or 0.5 mm (for the Causse) fenestra was made in the footplate using Halik graduated perforators. Adjunctive use of the Argon beam laser (wavelength 512 nm) delivered via an EndoOtoprobe ™ was introduced in 2000. The laser assisted procedures were performed in 134 1st ear surgeries and in 163 2nd ear surgeries.

The hearing outcomes were analyzed according to the 1995 AAO-HNS Committee on Hearing and Equilibrium (CHE) guidelines.[11] Frequency specific AC thresholds at 0.5, 1, 2, 3, 4, 6, 8 and 12 kHz and BC thresholds at 0.5, 1, 2, 3 and 4 kHz in operated and non-operated ears were tabulated in a computer database. As the beginning of this study period pre-dates the 1995 AAO-HNS guidelines, 3 kHz results were not available for all patients. In these cases 3 kHz AC and BC was estimated by average between 2 and 4 kHz. Mean preoperative pure tone average (PTA) of AC, BC and ABG at 0.5, 1, 2 and 3 kHz were compared with corresponding postoperative values. Postoperative BC values were used to calculate the postoperative ABG. In addition to reporting the results as mean values of AC, BC, ABG and AC gain, we used the GBP for visual presentation of bilateral hearing status in each individual case. The normal hearing was defined as an AC threshold ≤ 30 dB according to the original description by Browning[7] who adopted the "15/30 dB Belfast Rule of Thumb"[15] for definition of socially acceptable hearing. Symmetrical hearing was defined as an interaural difference between the operated and non-operated ear ≤ 10 dB as proposed in GBP method. Based on these criteria, patients were grouped into three major categories according to their preoperative hearing impairment: group 1 – unilateral hearing impairment (UHI), group 2 - bilateral asymmetric hearing impairment (BAHI) and group 3 -bilateral symmetric hearing impairment (BSHI). Before 1st ear surgery 8 patients had only a unilateral hearing impairment. These patients were treated with bilateral stapedotomy resulting from the development of hearing loss in the second ear during audiometric follow up after the 1st ear surgery. The BAHI group comprised 38 patients and BSHI group comprised 151 patients. Before 2nd ear stapedotomy, there were now 142 patients in UHI group, 45 patients in BAHI group and 10 patients in BSHI group. Pre- and postoperative AC PTAs in the operated ear were plotted on vertical axis against corresponding AC thresholds in the non-operated ear on horizontal axis.

The mean preoperative hearing levels (so called "threshold hearing") calculated as a PTA at frequencies of 0.5, 1, 2 and 3 kHz were 54 dB (SD 14 dB) in operated ear and 51 dB (SD 14 dB) in the non-operated ear at 1st ear surgery and 51 dB (SD 14 dB) and 29 dB (SD 11 dB) respectively at 2nd ear surgery. The mean preoperative ABG was 27 dB (SD 9 dB) at 1^{st} ear surgery and 25 dB (SD 8 dB) at 2^{nd} ear surgery.

The database was analyzed with a statistical software program *STATISTICA 6.0*, StatSoft, Inc., Tulsa, OK, USA. Statistical analysis was carried out for comparisons between pre- and postoperative values using paired t-test for the evaluation of AC, BC, PTA, ABG and AC gain. The criterion selected for statistical significance was p <0.05.

Results

Conventional Methods of Surgical Success Assessment

I. Closure of air-bone gap

The pre- and postoperative ABG and percentage of ABG closure were calculated for each series of 1^{st} and 2^{nd} ear stapedotomies as a whole and separately for the unilateral hearing impairment group (group 1,UHI), the bilateral asymmetric hearing impairment group (group 2, BAHI) and the bilateral symmetric hearing impairment group (group 3, BSHI)
(Tables 1, 2). The technical success in ABG closure ≤ 10dB was significantly higher in 1^{st} ear surgeries (90% compared with 80% in 2^{nd} ear, $p<0.05$). ABG closure ≤ 20dB was demonstrated in 98 % of cases after both 1^{st} and 2^{nd} procedure.

Table 1. Air-bone gap (ABG) closure at 0.5, 1, 2 and 3 kHz after 1^{st} ear stapedotomy.

	All 1^{st} ear stapedotomies	Group 1 UHI	Group 2 BAHI	Group 3 BSHI
	n=197	n=8	n=38	n=151
Postop ABG ≤ 10 dB	177 (90%)	8 (100%)	32 (84%)	137 (91%)
Postop ABG ≤ 20 dB	195 (99%)	8 (100%)	36 (95%)	151 (100%)
Preop ABG (dB) Mean SD	27 (±9)	29 (±6)	34 (±6)	26 ±8)
Postop ABG (dB) Mean (SD)	5 (±4)	7 (±2)	7 (±6)	5 (±4)

Group 1 - unilateral hearing impairment,
Group 2 - bilateral asymmetric hearing impairment,
Group 3 - bilateral symmetric hearing impairment.
n - number of patients, dB - decibel, SD - standard deviation.

Table 2. Air-bone gap (ABG) closure at 0.5, 1, 2 and 3 kHz after 2nd ear stapedotomy.

	All 2nd ear stapedotomies	Group 1 UHI	Group 2 BAHI	Group 3 BSHI
	n=197	n=142	n=45	n= 10
ABG ≤ 10 dB	157 (80%)	110 (78%)	37 (82%)	10 (100%)
ABG ≤ 20 dB	193 (98%)	142 (100%)	41 (91%)	10 (10%)
Preop ABG (dB) Mean (SD)	25 (±8)	24 (±7)	31 (±9)	18 (±3)
Postop ABG (dB) Mean (SD)	7 (±5)	7 (±4)	7 (±6)	5 (±3)

Group 1 - unilateral hearing impairment,
Group 2 - bilateral asymmetric hearing impairment,
Group 3 - bilateral symmetric hearing impairment.
n - number of patients, dB - decibel, SD - standard deviation.

II. Postoperative improvement in air conduction thresholds

Postoperatively, significant improvement of AC thresholds was demonstrated in all groups ($p<0.001$). The mean postoperative AC threshold in the operated ear was ≤ 30dB after both 1st and 2nd ear surgery, meeting the criteria of socially serviceable hearing (Table 3). Postoperative AC gain was better in the 1st ear surgery compared with the 2nd ear surgery in all groups of preoperative hearing impairment. Within the groups, the best mean postoperative AC thresholds were demonstrated in the category of unilateral hearing impairment (group 1) - the largest category in 2nd ear surgery.

Table 3. Improvement in air-conduction (AC) thresholds at 0.5, 1, 2 and 3 kHz.

AC (dB) Mean (SD)	All bilateral cases		Group 1		Group 2		Group 3	
	1st ear n=197	2nd ear n=197	1st ear n=8	2nd ear n=142	1st ear n=38	2nd ear n=45	1st ear n=151	2nd ear n=10
Preop	54 (14)	51 (14)	53 (6)	47 (9)	62 (11)	67 (16)	52 (14)	52 (19)
Gain	25 (9)	21 (10)	29 (6)	20 (9)	33 (8)	28 (11)	23 (9)	16 (5)
Postop	29 (11)	30 (12)	24 (3)	27 6)	29 (12)	29 (12)	29 (12)	36 (17)
Unoperated ear	51 (14)	29 (11)	23 (4)	24 (5)	44 (9)	41 (12)	52 (14)	45 (19)

n - number of patients, dB - decibel, SD - standard deviation.
AC thresholds in unoperated ear shown for comparison with postoperative AC thresholds.

Postoperative BC thresholds

Preoperative BC thresholds were analyzed for identification of cases with achievable socially acceptable hearing. Patients with unilateral hearing impairment have the most favourable proportion of normal cochlear reserve with BC levels ≤ 30 dB: 8 patients (100% of cases) in the 1st ear surgery and in 130 patients (92% of cases) in the 2nd ear surgery. The most unfavourable preoperative conditions with the highest proportion of mixed hearing loss and BC thresholds above 30 dB were found in patients with bilateral asymmetric hearing impairment (group 2) (Table 4). Postoperatively BC thresholds significantly improved in all groups ($p<0.05$), by 4 dB after the 1st ear surgery and by 3 dB after the 2nd ear surgery.

Table 4. Bone conduction (BC): mean pre- and postoperative thresholds at 0.5, 1, 2 and 3 kHz, proportion of patients with mixed hearing loss (preop BC ≥ 25dB) and preoperative cochlear reserve with achievable socially serviceable hearing (preop BC < 30 dB).

BC (dB) Mean (SD)	All bilateral cases		Group 1		Group 2		Group 3	
	1st ear n=197	2nd ear n=197	1st ear n=8	2nd ear n=142	1st ear n=38	2nd ear n=45	1st ear n=151	2nd ear n=10
Preop	27 (11)	26 (11)	23 (5)	22 (6)	27 (12)	36 (14)	26 (11)	35 (19)
Postop	23 (11)	23 (11)	18 (5)	19 (5)	23 (8)	33 (15)	24 (11)	31 (19)
Preop BC < 30 dB	145 74%	158 80%	8 100%	130 92%	24 63%	22 48%	113 75%	6 60%
Preop BC ≥ 25dB	87 44%	134 68%	4 50%	99 41 %	16 43%	29 64%	67 44%	6 60%

n - number of patients, dB - decibel, SD - standard deviation

Glasgow Benefit Plot

Figure 1 demonstrates schematic changes that can occur postoperatively according to the GBP criteria. Figures 2 and 3 demonstrate the AC thresholds change after 1st and 2nd ear stapedotomies. The most common type of hearing impairment among patients who were treated with bilateral stapedotomy was a bilateral symmetric hearing loss (77% of cases, 151 / 197). As a result of 1st ear surgery, 142 (72%) patients changed into the category of unilateral hearing loss. As a result of 2nd ear surgery, 125 (64%) patients changed into the category of normal and symmetric hearing.

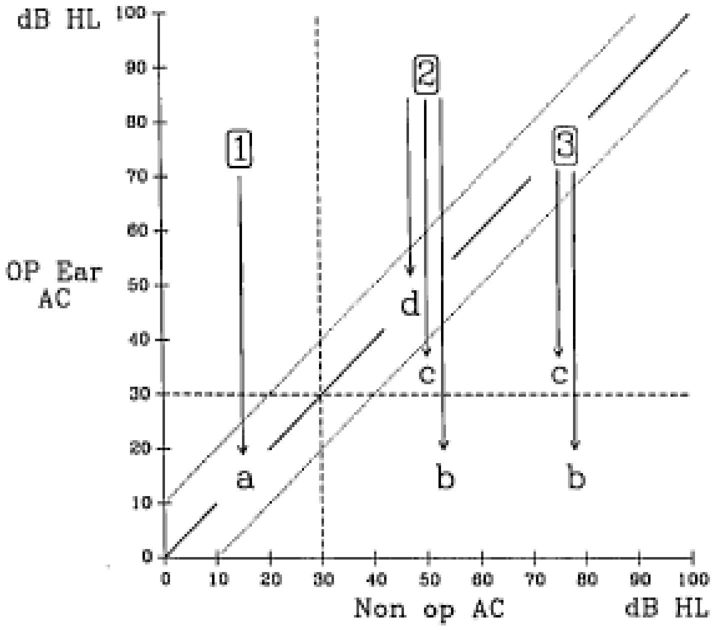


Figure 1. Potential changes from preoperative impairment group to postoperative impairment category.
Group 1- unilateral HL, 2- bilateral asymmetric HL, 3- bilateral symmetric HL.
Category a - bilateral serviceable hearing, b – unilateral serviceable hearing, c – bilateral asymmetric hearing impairment with operated ear converted to the better hearing ear, d – bilateral symmetric hearing impairment.

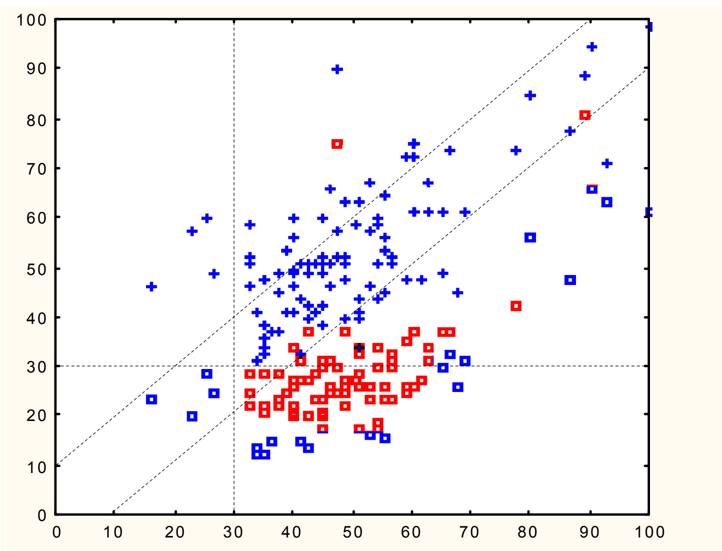


Figure 2. Results of first ear stapedotomy
▫ – postoperative AC PTA
+ – preoperative AC PTA
dB-decibel, AC-air conduction, PTA-pure tone average

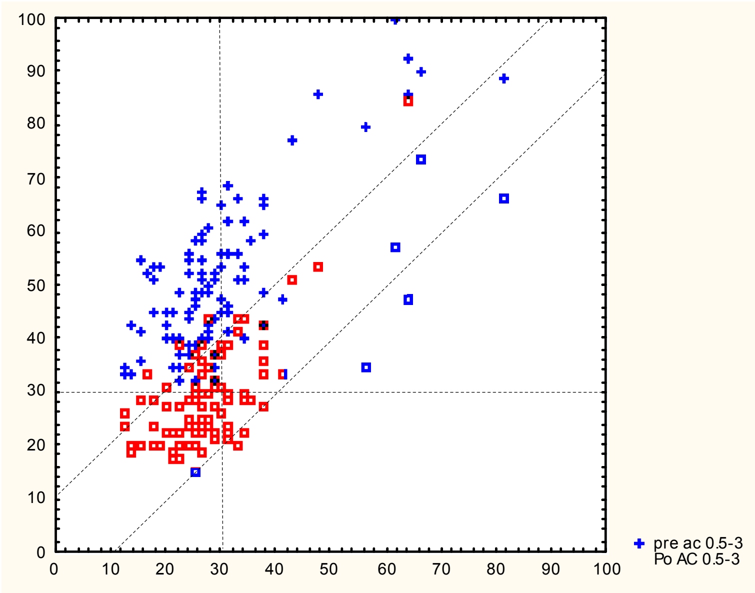


Figure 3. Results of second ear stapedotomy.
▫ – postoperative AC PTA
\+ – preoperative AC PTA
dB-decibel, AC-air conduction, PTA-pure tone average

1. Postoperative normal hearing

Postoperative AC PTA at 0.5,1,2 and 3 kHz ≤ 30 dB was achieved in 144 (73%) of 1st ear stapedotomies and in 125 (64%) of 2nd ear stapedotomies. The first surgery was more successful in achievement of socially serviceable hearing than the second ear procedure in all groups (Table 5). Ninety nine percent of patients with normal cochlear reserve (preop BC < 30 dB) achieved normal AC thresholds after the 1st ear stapedotomy and 79% of patients after the 2nd ear procedure.

Table 5. The proportion of postoperative normal hearing within the groups and proportion of cases with normal cochlear reserve (preop BC < 30 dB, Table 3) who actually achieved socially serviceable hearing

Postop AC (dB) Mean (SD)	All bilateral		Group 1		Group 2		Group 3	
	1st ear n=197	2nd ear n=197	1st ear n=8	2nd ear n=142	1st ear n=38	2nd ear n=45	1st ear n=151	2nd ear n=10
Postop AC < 30 dB % of group	144 73 %	125 64%	8 100%	104 73%	28 74%	17 38%	108 72%	4 40%
% of preop BC<30dB	99 % 144/145	79% 125/158	100% 8/8	80% 104/130	117% 28/24	77% 17/22	96% 108/113	66% 4/6

2. Postoperative symmetrical hearing

Postoperative bilateral hearing results are summarized in Table 6.

According to the GBP criteria requesting an interaural difference of less than 10 dB, symmetric hearing was achieved in 80% of cases after the 2nd ear stapedotomy. Using the “Belfast Rule of Thumb“and a less strict criteria for an interaural difference of less than 15 dB, symmetrical hearing was achieved in 95% of bilateral stapedotomies. The operated ear became the better hearing ear in 97% of cases after the 1st ear surgery and in 46% of cases after the 2nd ear procedure. This finding is not surprising given the mean AC thresholds of 50 (±15) dB in non-operated ear at 1st ear surgery and 29 (±11) dB at 2nd surgery.

Table 6. Postoperative symmetrical hearing: interaural difference (Δ) < 10 dB according to the GBP criteria of symmetrical hearing, interaural difference (Δ) < 15 dB according to the “Belfast Rule of Thumb “.

Interaural difference	All bilateral		Group 1		Group 2		Group 3	
	1st ear n=197	2nd ear n=197	1st ear n=8	2nd ear n=142	1st ear n=38	2nd ear n=45	1st ear n=151	2nd ear n=10
Δ < 10 dB % of group	18 9%	157 80%	8 100%	120 85%	2 5%	31 69%	8 5%	6 60%
Δ < 15 dB % of group	36 18%	187 95%	8 100%	136 96%	8 21%	41 91%	20 13%	10 100%
Operated side became the better hearing ear	191 97%	91 46%	4 50%	56 39%	36 95%	25 56%	151 100%	10 100%

Complications

In 1st ear surgery intra-operative complications were encountered in 9 cases. TM tears occurred in 4 patients and were repaired intra-operatively; stapedial footplate cracks or mobilization occurred in another 4 patients and the long process of incus was fractured in one patient.

In 2nd ear procedure 8 complications took place: stapedial footplate cracks or mobilization in 6 patients, a perilymphatic gusher repaired intra-operatively in one patient and an incidental incus mobilization occurred in one patient.

None of these complications resulted in immediate or delayed SNHL or persistent vestibular symptoms.

Discussion

Stapes surgery is currently considered the treatment of choice for otosclerosis. This technique has been proven to be highly successful in correction of the conductive component of hearing loss in an operated ear. The reputation of stapedectomy has been largely based on reports of high technical success measured by ABG closure in an operated ear. Otosclerosis, however, is a bilateral disease in the majority of cases.[16,17] Typically patients present with a slowly progressive hearing loss that is usually bilateral and often asymmetric.[18] Over the patients' lifetime the hearing loss inevitably evolves to mixed type due to combined effect of aging and progression of the disease itself. The adequacy of surgical management in this type of bilateral hearing loss in otosclerosis remains controversial.

The dilemma to operate or not on a second otosclerotic ear arises from difficulties in the assessment of benefit from a second ear procedure as well as the danger of unpredictable and potentially devastating complications. Early reports on the high technical success of stapedectomy were followed by descriptions of surgical complications.[19-21] The histopathological findings following bilateral stapedectomy included suppurative labyrinthitis, perilymphatic fistula, fibrous adhesions in utricle, bony fragments in saccule, and cholesteatoma formation.[6,22,23]

Risks of bilateral deafness or complete loss of vestibular function as delayed complications have driven opinion against a second ear stapedectomy.[23,25] Refinements of surgical technique however allowed for a reduction of complication rates reported in total stapedectomies with fat-wire prosthesis and Gelfoam sealing.[26,27,28] The currently preferred small fenestra stapedotomy, reverse order of surgical steps and laser-assisted fenestration has converted stapes surgery to a much safer procedure in experienced hands.[29-31]

Prevention of complications and improvement of postoperative hearing have been the main benefits from the numerous refinements in stapedectomy technique. While intra-operative complications appear better controlled, hearing improvement remains more difficult to interpret. Standard audiometry is helpful in documentation of changes in postoperative air and bone conduction in a operated ear, but its ability to describe bilateral hearing function in real life conditions appear limited.[32-34]

Historically, the literature on stapedectomy results was focused on technical success, measured as the degree of ABG closure. In the studies reporting 95-97% rates of ABG ≤10 dB, postoperative hearing function was rarely mentioned.[35-37] Notably, in one study the technical results of 1800 bilateral stapedectomies and the degree of ABG closure in relation to various operative findings was described in great details. No comments were found, however, on hearing thresholds following surgery.[38] In turn, in studies on the functional benefit of stapes surgery, ABG closure rates were not mentioned.[8,9]

In only a small number of publications have both technical and functional outcomes been reported. In these studies, an ABG ≤ 10 dB was achieved in 50%[7] to 74%[39] of cases. Unfortunately technically successful stapes surgery is not always beneficial in the patients' perception. Smyth[39] found that a post-operative hearing threshold below 30 dB or an interaural difference less than 15 dB are more important for patients' satisfaction than the actual ABG closure. Not withstanding the patients' main expectation from stapedectomy is an ability to hear equally well in both ears. A recognition that technically flawless stapedotomy will not necessarily result in increased patient satisfaction requires more functional measures of success. Smyth and Toner[15] coined the "15/30 dB Belfast Rule of Thumb" as a measure of benefit of the middle ear surgery. According to 15/30 dB rule, the aim of surgery should be postoperative AC thresholds within 30 dB and interaural difference less than 15 dB.

Emphasizing the importance of bilateral hearing for patient benefit from middle ear surgery, Browning[7] proposed the Glasgow Benefit Plot (GBP) - a new method for evaluation of hearing as a bilateral function. The GBP includes evaluation of AC PTA in both the operated and non-operated ear. A 30dB criterion of socially serviceable hearing, adopted from the "Belfast Rule of Thumb", was set as a desirable goal of surgery. The definition of symmetric hearing was set as 10 dB of interaural difference. The concept of grouping patients to three major categories of unilateral, bilateral symmetric and bilateral asymmetric hearing impairment allows for the preoperative assessment of chances for achieving symmetric or normal postoperative hearing.

The results of unilateral and bilateral stapedotomies evaluated with the GBP are vary according to audiometric parameters used in the calculations. In some studies PTA values used for plotting the GBP were calculated on a three frequency average of 0.5,1 and 2 kHz[8,9], while others[40], including our study, used four-frequency average according to the AAO-HNS CHE guidelines for reporting results of middle ear surgery. Inclusion of the 3 kHz frequency in the calculation of PTA has been demonstrated to decrease the percentage of patients achieving postoperative normal hearing[41], and subsequently, the success rate evaluated with the GBP. Another difference in the methodology is the definition of symmetric hearing. In the original description of the GBP[7] Browning used an interaural difference of 10 dB as a criterion of symmetric hearing. Other authors[15,18] used a 15 dB difference criteria whereas some studies defined symmetrical hearing as an interaural difference of ≤20 dB.[9] To evaluate the effect of these different criteria of symmetrical hearing on a success rate, we calculated the percentage of patients with a postoperative interaural difference of ≤10 dB and ≤15 dB (Table 5). After 1st ear stapedotomy symmetrical hearing was achieved in 9% of patients if GBP criteria were used and in 18% according to the “Belfast Rule of Thumb”. 2nd ear stapedotomy provided 80% of patients with symmetrical hearing by the GBP definition and 95% of patients according to the “Belfast Rule of Thumb”.

Unilateral surgery has been found to be satisfactory in providing postoperative normal and symmetric hearing in patients with unilateral hearing impairment.[42,43] Success rates within 71-79% have been reported in studies using the GBP in the assessment of unilateral stapedotomies.[44,45]

The results from our series[12] of 751 unilateral stapedotomies in patients with unilateral or bilateral hearing loss demonstrated that bilateral normal and symmetric hearing was achieved in 78% of 481 cases with unilateral hearing impairment compared with 70% of 509 cases reported by Persson[40] and 80.5 % of 80 cases in De Bruijn’s study.[9] In patients with bilateral symmetric hearing impairment in our series[12], serviceable hearing in the operated ear was achieved in only 43% of cases, while postoperative thresholds were symmetric in 77% of cases in this group. In cases with asymmetric impairment, symmetric postoperative hearing was achieved in only 36% of cases, even though the operated ear was converted to better hearing ear in 96% of these cases. Lundman[46] also found that unilateral surgery was insufficient in providing normal and symmetric hearing function in patients with bilateral impairment.

The results of the present study demonstrate that following 1st ear surgery the operated ear was converted to the better hearing ear in 97% of cases and serviceable hearing in operated ear was demonstrated in 99% of patients with achievable normal thresholds (preoperative BC ≤30 dB) (Tables 2, 4). As aforementioned, however, symmetrical hearing was achieved in only 9-18% of cases after 1st ear surgery (Table 5). The 2nd ear procedure provided symmetric hearing within 15 dB in 95% and normal hearing in 64% of cases. Second ear surgery nevertheless appears to be less successful in achievement of postoperative thresholds below 30 dB. The results seem to be affected by a higher proportion of mixed hearing loss in the second otosclerotic ear: 67% of patients had mixed hearing loss before 2nd ear surgery compared with 44% before 1st surgery (Table 3).

Our results indicated that preoperative BC thresholds and the type of preoperative hearing impairment define whether a bilateral normal hearing can be achieved postoperatively. In patients with mixed and asymmetric hearing loss, bilateral stapes surgery remains beneficial in facilitating postoperative fitting of bilateral hearing aids. The results of our study support the conclusions based on the evaluation of smaller series[8,47] in that 2nd ear stapedotomy contributes to achievement of bilateral serviceable hearing in patients with bilateral hearing impairment.

The low rate of intra-operative complications, similar in 1st and 2nd ear surgery, with no detrimental postoperative effect on cochlear and vestibular function, appears reassuring and speaks to the safety of bilateral stapes surgery. The complication rate in our series is similar to results recently reported by Albrecht[48] and Kujala.[49] These findings have lead some authors to suggest even performing simultaneous bilateral stapes surgery in selected patients.[50].

Conclusions

In this largest study of 394 primary bilateral stapedotomies results has been simultaneously evaluated according to AAO-HNS guidelines and GBP criteria. The addition of the GBP to conventional parameters allowed comparison of the benefits of 1st and 2nd ear surgeries in the achievement of bilateral socially serviceable and symmetric hearing. We conclude that in our experience first ear surgery was more successful in the achievement of normal hearing. Second ear procedure was more beneficial in providing symmetric hearing in this series.

References

1. Reddy J, Sataloff J, Liu J. Histopathological changes following bilateral stapedectomy.Arch Otolaryngol. 1966; 84(2):165-9.
2. Ferguson B, Gillespie, Kenan P, Farmer J. Mechanisms of cholesteatoma formation following stapedectomy. Am J Otol. 1986; 7(6): 420-4.
3. Altmann F, Basek M, Hough J. Otosclerosis with bilateral stapes mobilization. Histological and clinical considerations. . Arch Otolaryngol. 1960; Aug; 72:147-62.
4. Sale C**.** Bilateral deafness due to stapes surgery. Arch Otolaryngol. 1969; 90(4):467-9.
5. Faye-Lund H, Stangeland N, Rohrt T. Long-term results of bilateral stapedectomy. J Laryngol Otol. 1984; 98:247-54.

6. Belal A, Ylikoski J. Post-stapedectomy dizziness. A histopathologic report. Am J Otol.1982; 3(3):187-91.
7. Browning GG, Gatehouse S, Swan IR. The Glasgow Benefit Plot: a new method for reporting benefits from middle ear surgery. Laryngoscope 1991; 101: 180-185.
8. Porter MJ, Zeitoun H, Brookes GB. The Glasgow Benefit Plot used to access the effect of bilateral stapedectomy. Clin Otolaryngol Allied Sci.1995; 20(1):68-71.
9. De Bruijn AJ, Tange RA, Dreschler WA. Evaluation of second-ear stapedotomy with the Glasgow benefit plot. ORL J Otolaryngol Relat Spec. 1999; 61 (2): 92-7.
10. Schmerber S, Karkas A, Righini CA, Chahine KA. The Quartile benefit plot: a middle ear surgery benefit assessment scheme. Laryngoscope 2008; 118(14):1-6.
11. Committee on Hearing and Equilibrium guidelines for the evaluation of results of treatment of conductive hearing loss. Otolaryngol Head Neck Surg 1995; 113(3):186-3.
12. Kisilevsky V, Bailie N, Dutt S, Halik J. Post- stapedotomy bilateral hearing function evaluated with the Glasgow Benefit Plot. 2008 (in submission).
13. Smyth G. Practical suggestions on stapedotomy. Laryngoscope 1982; 92-8(1):952-3
14. Fish U. Stapedotomy vs. stapedectomy. Am J Otol 4:112-17, 1982.
15. Toner J, Smyth G. Comparison of methods of evaluating hearing benefit of middle ear surgery. J Laryngol Otol. 1993; 107:4-5.
16. Shea J, Shea P, McKenna M. Stapedectomy for otosclerosis. In: Surgery of the ear, By Glasscock M, Gulya A (ed), BC Decker, 2003.
17. Hawke M. Otosclerosis. In Diseases of the Ear, by Hawke M, Jahn A (ed), Gower Medical Publishing, 1987.
18. Emmet J. Physical examination and clinical evaluation of the patient with otosclerosis.Otolaryngol Clin N Am 1993;26(3): 353-9.
19. Schuknecht H, Mendoza A. Cochlear pathology after stapedectomy. Am J Otolaryngol. 1981; 2(3):173-87.
20. Pratt L. Postoperative complications associated with bilateral stapedectomy. Laryngoscope 1972;1: 1189-98.
21. Mann W, Amedee R, Fuerst G. Hearing loss as a complication of stapes surgery. Otolaryngol Head Neck Surg.1996; 115(4):324-8.
22. Eviatar A, Jamal H. Cholesteatoma induced by stapedectomy. Arch Otolaryngol. 1983; 109(6):413-4.
23. Von Haacke N, Wilson J, Murray J, Duvall E. Cholesteatoma following stapedectomy. J Laryngol Otol. 1987; 101(7):708-10.
24. Ludman H, Grant H. The case against bilateral stapedectomy, and problems of post-operative follow-up – from the King's College Hospital series. J Laryngol Otol. 1973; 87(9):833-43.
25. Smyth G, Kerr A, Singh K. Second-ear stapedectomy – a continued controversy. J Laryngol Otol. 1975; 89:1047-56.
26. Hough J, Dyer K. Stapedectomy: causes of failure and revision surgery in otosclerosis. Otolaryngol Clin N Am 1993;26(3): 453-70.
27. Glasscock ME, McKennan KX, Levine SC. Revision stapedectomy surgery. Otolaryngol H&N Surg. 1987;96(2):141-48.
28. Smyth GDL, Hassard TH. Eighteen years experience in stapedectomy. Ann Otol Rhinol Laryngol (Suppl) 1978 (5-6); 87:3-36.
29. House H, Hansen M, Al Dakhalil A, House W. Stapedectomy versus stapedotomy: comparison of results with long-term follow-up. Laryngoscope 2002;112:2046-50.
30. Vernick D. Laser applications in ossicular surgery. Otolaryngol Clin N Am 1996;29(6): 931-41
31. Szymanski M, Golabek W, Morshed K, Siwiec H. The influence of the sequence of surgical steps on complications rate in stapedotomy. OLtol Neurotol 2007;28: 152-6.
32. Black B. Reporting results in ossiculoplasty. Otol Neurotol. 2000; 24: 534-42.
33. Giles M, Browning GG, Gatehouse SG. Problems in assessing the audiogram in patients with severe hearing impairment. J Laryngol Otol. 1999;110: 727-31.
34. Schmerber S, Karkas A, Righini CA, Chahine KA. The Quartile benefit plot: a middle ear surgery benefit assessment scheme. Laryngoscope. 2008; 118(14):1-6.
35. Shea J. Forty years of stapes surgery. Am J Otol 1998; 19:52-55.
36. Sheehy JL, Nelson RA, House HP. Stapes surgery at the Otologic Medical Group. Am J Otol 1979 ;1(1):22-26
37. Somers T, Govartes P, Marquet T, Offeciers E. Statistical analysis of otosclerosis surgery performed by Jean Marquet. Ann Otol Rhinol Laryngol. 1994; 103(12):945-51.

38. Daniels R, Krieger L, Lippy W. The other ear: findings and results in 1800 bilateral stapedectomies. Otol Neurotol. 2001; 22(5):603-7.
39. Smyth GDL, Patterson CC. Results of middle ear reconstruction: do patients and surgeons agree? Am J Otol 1985; 6: 276-279.
40. Persson P, Harder H, Magnuson B. Using the Glasgow benefit plot as a prognostic instrument and for preoperative counselling in patients with otosclerosis. Otol Neurotol. 2007; 28 (6): 739-44.
41. Berliner KI, Doyle KJ, Goldenberg RA. Reporting operative hearing results in stapes surgery: does choice of outcome measure make a difference? Am J Otol 1996; 17(4):214-20.
42. Subramanian K, Eikelboom RH, Marino R et al., Patient's quality of life and hearing outcomes after stapes surgery. Clin Otolaryngol 2006; 31(4): 273-279.
43. Caces F, Braccini F. Comparative study using AAO-HNS guidelines for conductive hearing loss and Glasgow Benefit Plot to evaluate results of stapes surgery for otosclerosis. Rev Laryngol Otol Rhinol 2007; 128 (1-2) : 47-53.
44. Huang SC, Stanley RE. Stapedectomy at the Singapore General Hospital-use of functional hearing analysis. Singapore Med J.1995; 36(2):158-162.
45. Szymanski M, Siwiec H, Golabek W. Glasgow Benefit Plot for evaluation of stapedectomy results. Otolaryngol Pol 2003; 57(3):403-406.
46. Lundman L, Mendel L, Bagger-Sjoback D, Rosenhall U. Hearing in patients operated unilaterally for otosclerosis. Self-assessment of hearing and audiometric results. Acta Otolaryngol.1999; 119(4): 453-8.
47. De Bruijn AJ, Tange RA, Dreschler WA, Grolman W, Schouwenburg. Bilateral stapedotomy in patients with otosclerosis: a disability oriented evaluation of the benefit of second ear surgery. Clin Otolaryngol.1998; 23:123-7.
48. Albrecht C, Fonesca A, Bernardes G. "Second ear" stapedotomy: is it safe? Acta Otolaryngol Belg. 2004; 58(2):109-11.
49. Kujala J, Aalto H, Ramsay H, Hirvonen T. Simultaneous bilateral stapes surgery- a pilot study. Acta Otolaryngol.2007; 127:1255-8.
50. Kujala J, Aalto H, Ramsay H, Hirvonen T. Simultaneous bilateral stapes surgery.Acta Otolaryngol.2008; 128(4):347-51.

Chapter 3

Stapes Surgery in Special Situations

3.1

Results of stapedotomy in otosclerosis with severe and profound hearing loss

V.E. Kisilevsky
N.A. Bailie
J.J. Halik

Journal of Otolaryngology – Head & Neck Surgery 2009; 39: in print

Abstract

Objective

To evaluate the technical and functional results of 144 stapedotomies in patients with severe to profound hearing loss (SPHL) and to compare the success rate in this group with that of 1001 primary stapedotomies for mild to moderate hearing loss (MMHL).

Study design

Retrospective chart review.

Setting

Tertiary referral center.

Methods

The charts of 1369 consecutive stapedotomy cases were reviewed. One hundred twenty-seven patients (144 stapedotomies) with SPHL were included. The outcomes were analyzed according to the 1995 American Academy of Otolaryngology – Head and Neck Surgery (AAO-HNS) Committee on Hearing and Equilibrium guidelines and evaluated with the Amsterdam Hearing Evaluating Plots (AHEP) and the Glasgow Benefit Plot (GBP).

Results

The magnitude of hearing improvement was greater in patients with SPHL compared with the patients with MMHL. The mean gain in air conduction in patients with SPHL was 32 (± 13) dB compared with 23 (± 11) dB in the series of 1001 primary stapedotomies for MMHL. Air-bone gap closure ≤ 10 dB was achieved in 63% of cases and ≤ 20 dB in 90% of patients with SPHL, compared with 82% and 97% respectively in patients with MMHL.

Conclusions

SPHL in otosclerosis is not infrequent, comprising 13% of all primary stapedotomies in this series. The magnitude of hearing improvement post stapedotomy is greater in SPHL compared with MMHL. Symmetric hearing could be achieved in two-thirds of patients, and normal hearing can be achieved in selected cases.

Introduction

Surgical intervention for severe to profound hearing loss (SPHL) owing to otosclerosis is controversial as a higher proportion of these patients have a mixed hearing loss, with insufficient cochlear reserve to allow them to dispense with the use of a hearing aid post-operatively.[1-6]

Poor cochlear reserve in this group of patients is also felt to increase the risk of post-operative sensorineural hearing loss and poor speech discrimination.[7] In addition, the inclusion of patients with severe hearing loss in the calculation of audiometric results of stapedotomy has been found to negatively affect the success rate in these series.[8] As a result, knowledge of the pre-operative hearing levels and the proportion of patients with SPHL in reported series is important for the adequate interpretation of surgical outcomes. The aims of this study were to evaluate the technical and functional results of 144 stapedotomies in patients with SPHL and to compare the success rate in this group with that of 1001 primary stapedotomies for mild to moderate hearing loss (MMHL).

Patients and methods

The charts of 1369 consecutive stapedotomies performed by one surgeon (J.J.H., the senior author) from 1991 to 2006 were reviewed. Of these 1369 procedures, 1145 were primary stapedotomies in patients with otosclerosis, 89 were revision stapedotomies and 48 were performed for congenital stapes fixation. Eighty-seven cases were excluded from the study owing to incomplete data. Of 1145 primary procedures, 144 were performed on 127 patients who had SPHL with pre-operative air conduction (AC) pure-tone average (PTA) ≥ 71dB. The audiometric results of these 144 stapedotomies were included in this study. Sixty-nine patients (54%) were women and 58 (46%) were men. The mean age was 55 years (SD 12 years; range from 25 to 84 years). A bimodal age distribution is demonstrated in Figure 1. Fifty-one patients (40%) had a family history of conductive hearing loss due to otosclerosis. Fifty patients had other complaints: 40 patients (31%) had tinnitus, 6 patients (5%) complained of occasional dizziness and 4 patients (3%) had both.

Stapedotomy was performed using the reverse order technique via a per meatal approach.[9,10] A modified Cawthorne (0.3 mm diameter) prosthesis was used until 2002 in 100 cases. Thereafter, a modified Causse (0.4 mm diameter) prosthesis was used in 44 procedures. A 0.4 mm (for the Cawthorne prosthesis) or 0.5 mm (for the Causse prosthesis) fenestra was made in the footplate using Halik graduated perforators.

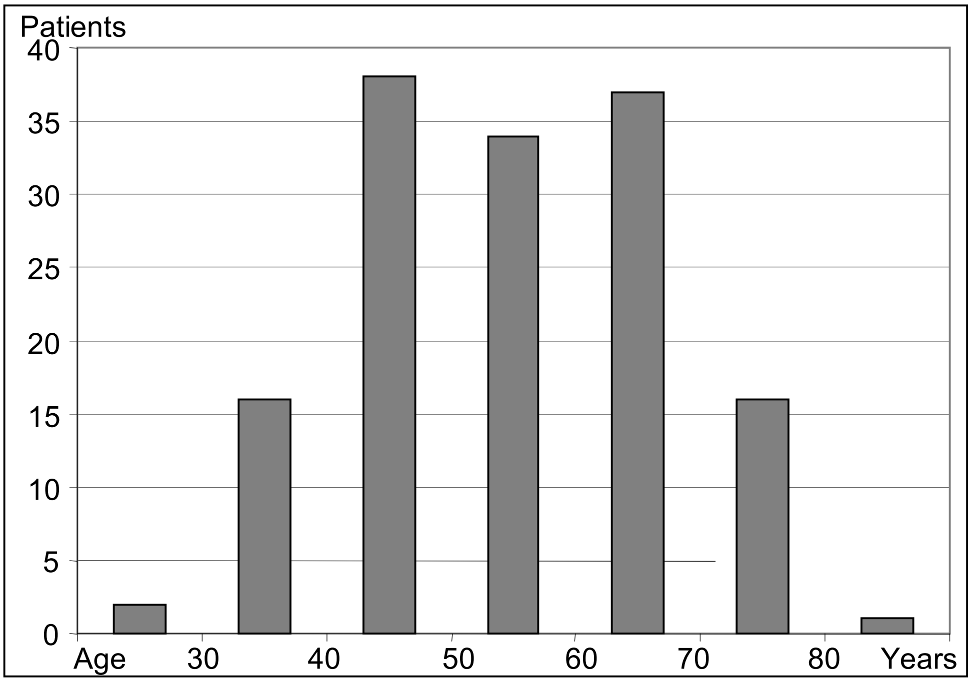


Figure 1. Age distribution in the group of 144 cases with severe to profound hearing loss. Data represent number of ears.

As part of practice-based learning of the senior author, adjunctive use of the Argon beam laser (Lumenis Inc, Salt-Lake City, UT, wavelength 512 nm), delivered via an EndoOtoprobe, was introduced in 2000. Laser-assisted procedures were performed in 77 cases. A modification of the reverse order technique was made in conjunction with the introduction of the laser: the Argon laser was used to vaporize the stapedial tendon and posterior crus of the stapes prior to footplate fenestration. The laser was also used to begin the footplate fenestration by providing a ‘set-hole’ for the perforators. The fenestration was completed with the graduated perforators. This reduced surgical trauma and technical difficulties related to manual fenestration. The hearing outcomes were analyzed according to the 1995 American Academy of Otolaryngology - Head and Neck Surgery (AAO-HNS) Committee on Hearing and Equilibrium (CHE) guidelines.[11] Frequency-specific AC thresholds at 0.5, 1, 2, 3, 4, 6, 8, and 12 kHz and bone conduction (BC) thresholds at 0.5, 1, 2, 3 and 4 kHz in operated and nonoperated ears were tabulated in a computer database. The beginning of this study period predated the 1995 AAO-HNS CHE guidelines; therefore, 3 kHz results were not available for all patients. In such cases, 3 kHz AC and BC values were calculated as an average of 2 and 4 kHz, allowing a uniform presentation of audiometric data according to the AAO-HNS guidelines. The mean pre-operative PTAs of AC, BC and air-bone gap (ABG) at 0.5, 1, 2, and 3 kHz were compared with corresponding post-operative values. Post-operative BC values were used to calculate post-operative ABG.

The mean pre-operative parameters are summarized in Table 1. The mean AC hearing level was 82 dB (SD 10 dB) in the operated ear and 51 dB (SD 24 dB) in the non-operated ear. The distribution of the severity of hearing loss is demonstrated in Figure 2. There were 115 ears (80%) with severe hearing loss (71 dB ≤ PTA < 90 dB) and 29 ears (20%) with profound hearing loss (91 dB ≤ PTA < 120 dB).

Table 1. Comparative pre-operative parameters in the group with severe to profound hearing loss (SPHL) and the group with mild to moderate hearing loss.

	Severe/profound HL (n = 144)	Mild/moderate HL (n = 1001)
	Mean (SD)	Mean (SD)
Age (yr)	55 (± 12)	43 (± 11)
AC PTA (dB)	82 (± 10)	49 (± 9)
BC PTA (dB)	46 (± 14)	23 (± 7)
BC PTA ≥ 30 dB	91 % (131/144)	15% (147/1001)
ABG (dB)	36 (± 11)	26 (± 9)
SRT (dB)	76 (± 12)	48 (± 10)

ABG - air-bone gap; AC - air conduction; BC - bone conduction; HL - hearing loss; PTA - pure tone average; SRT - speech reception threshold.

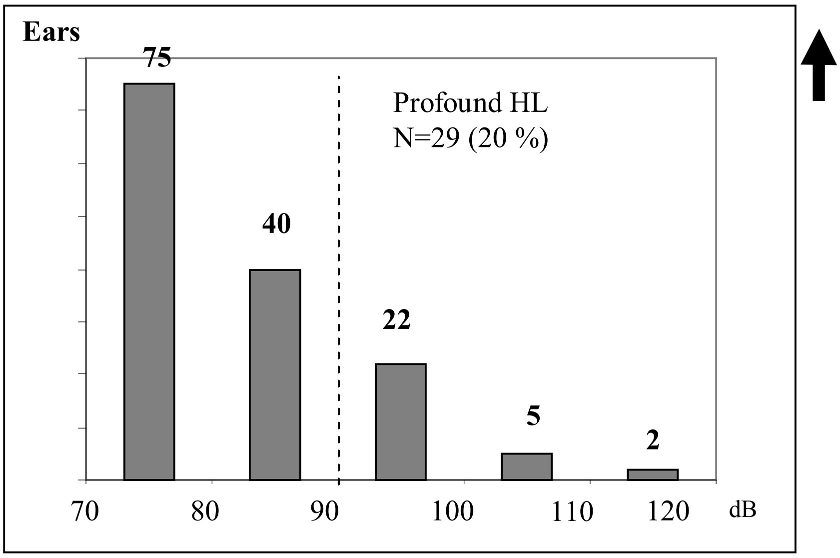


Figure 2. Pre-operative distribution of ears according to severity of hearing loss (HL).
71-90 dB = severe hearing loss; 91-120 = profound hearing loss.
dB - decibel;HL - hearing loss; n - number of ears

In addition to reporting averaged results prepared from the mean values of AC, BC, ABG, and speech reception thresholds (SRTs) in the operated ear, we used the Glasgow Benefit Plot (GBP)[12] to evaluate bilateral post-operative hearing function. Although, technically, there was no control group that did not undergo intervention, audiometric data from the nonoperated ear were gathered, analyzed, and reported to allow comparison with the operated ear and to appreciate the benefits of surgical treatment. The GBP was also useful in assessing the type of pre-operative hearing impairment. Normal hearing was defined as an AC threshold ≤ 30 dB according to the original description by Browning and colleagues, who adopted the "15/30 dB Belfast Rule of Thumb" for the definition of socially acceptable hearing.[13] Symmetric hearing was defined as an interaural difference between the operated and non-operated ear ≤ 15 dB.
Based on these criteria, the ears of patients were grouped into three major categories according to their pre-operative hearing impairment: group 1 (n = 26), unilateral hearing impairment (UHI); group 2 (n = 81), bilateral asymmetric hearing impairment, and group 3 (n = 37), bilateral symmetric hearing impairment. Pre- and post-operative AC PTAs in the operated ear were plotted on the vertical axis against corresponding AC thresholds in the non-operated ear on horizontal axis. The Amsterdam Hearing Evaluation Plots (AHEP)[14] was used for evaluation of post-operative changes in BC and visual presentation of AC gain in each case. Pre-operative PTAs of BC were plotted against corresponding post-operative BC thresholds, and pre-operative ABG values were plotted against AC gains, allowing data for each individual patient to be recognized as plot points on a graph. This addition of the GBP and AHEP meets AAO-HNS level I and level II guidelines for the uniform reporting of both summary and raw data.

Data were analyzed with the statistical software program Statistica 6.0 (StatSoft, Inc., Tulsa, OK). Statistical analysis was carried out for comparisons between pre- and post-operative values using the paired t-test for the evaluation of AC, BC, PTA, ABG, and AC gain. The criterion selected for statistical significance was $p < 0.05$.

Results

Conventional Methods of Assessing Surgical Success

Mean AC, BC, SRT

Significant improvement in all audiometric parameters (AC and BC thresholds, ABG, and SRT) was demonstrated post-operatively ($p < 0.001$) (Table 2). Even though the mean post-operative AC level was 51 (± 17) dB, the magnitude of hearing improvement was greater in patients with SPHL compared with patients with MMHL. The mean gain in AC in patients with SPHL was 32 (± 13) dB compared with 23 (± 11) dB in MMHL.

Table 2. Mean (SD) pre- and post-operative air conduction and bone conduction thresholds, air-bone gap, speech reception threshold, and air conduction gain at 0.5, 1, 2 and 3 kHz.

	AC	BC	ABG	SRT	AC gain
	Mean (SD)				
Pre-operative (dB)	82 (±10)	46 (±14)	36 (±11)	76 (±12)	-
Post-operative (dB)	51 (±17)	41 (±13)	10 (±8)	44 (±16)	32 (±13)

ABG - air-bone gap; AC - air conduction; BC - bone conduction; SRT - speech reception thresholds

AC improvement

Although all operated ears had SPHL pre-operatively, normal or socially acceptable hearing (mean AC ≤ 30 dB) was achieved post-stapedotomy in all 13 cases (13 of 144 or 9%) with the potential to achieve normal hearing (pre-operative BC PTA ≤ 30 dB). Post-operative MMHL in the operated ear was demonstrated in 114 (79%) cases whereas 17 (12%) ears remained in category of SHL (Figure 3).

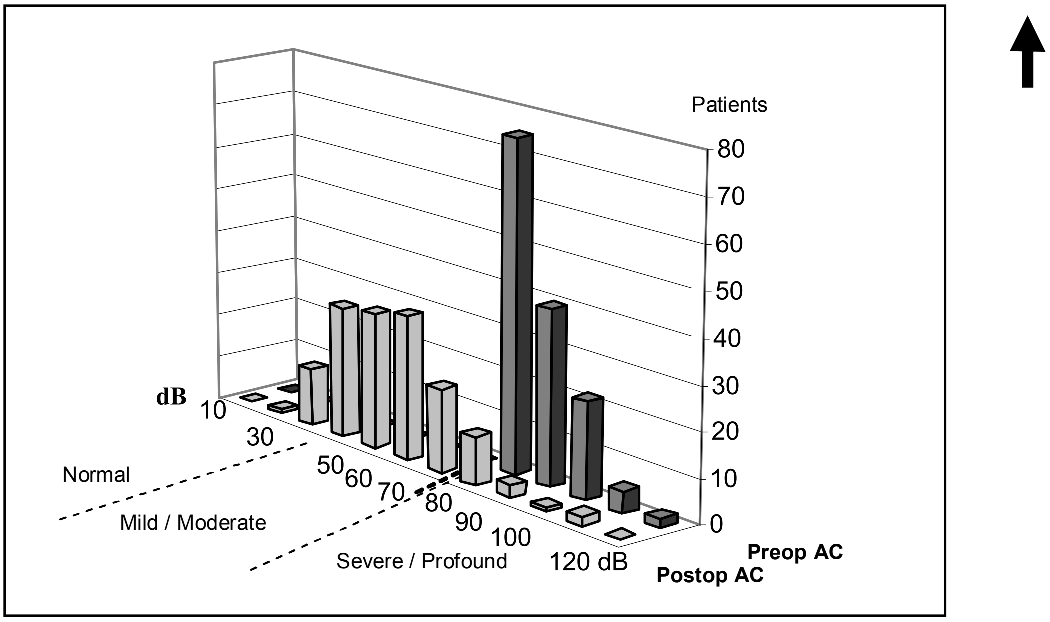


Figure 3. Air conduction (AC) threshold shift from pre-operative severe to profound hearing loss to post-operative mild to moderate and normal hearing.
AC - air conduction; dB - decibel.

ABG Closure

Pre- and post-operative ABG and percentage of ABG closure was calculated for the group with SPHL and compared with the group with MMHL (Table 3). The mean postoperative ABG of 10 (± 8) dB was demonstrated in the group with SPHL compared with 7 (± 5) dB in the group with MMHL. ABG closure ≤ 10 dB was achieved in 63% of cases and ≤ 20 dB in 90% of patients with SPHL, compared with 82% and 97% respectively, in the patients with MMHL.

Table 3. Comparative rates of air bone gap closure in the groups with severe to profound hearing loss and mild to moderate hearing loss.

Post-operative ABG (dB)	n of ears with SPHL	% of ears with SPHL	% of ears with MMHL
≤ 10 dB	90	63	82
≤ 20 dB	130	90	97
≥ 20 dB	14	10	3

ABG - air-bone gap; MMHL - mild to moderate hearing loss; SPHL - severe to profound hearing loss.

Hearing results assessment with AHEP.

Pre- and post-operative BC

BC thresholds remained within 10 dB of pre-operative value in 79 (55%) ears. These cases are represented by dots situated between two diagonal lines on Figure 4.

Improvement in BC > 10 dB was demonstrated in 63 (44%) ears represented by dots below the lower diagonal line. Worsening by more than 10 dB was observed in two cases (1%) and is represented by dots above the upper diagonal line.

AC gain and ABG closure ≤ 20 dB.

Successful results with ABG closure ≤ 20 dB were demonstrated in 130 (90%) ears. These cases are represented by dots situated below the upper diagonal line on Figure 5.

Overclosure, with gain in AC thresholds exceeding pre-operative ABG, was achieved in 34 (24%) of these cases, represented by dots below the lower diagonal line. Unfavourable results, with ABG > 20 dB, were demonstrated in 14 (10 %) ears and are represented by dots above the upper diagonal line.

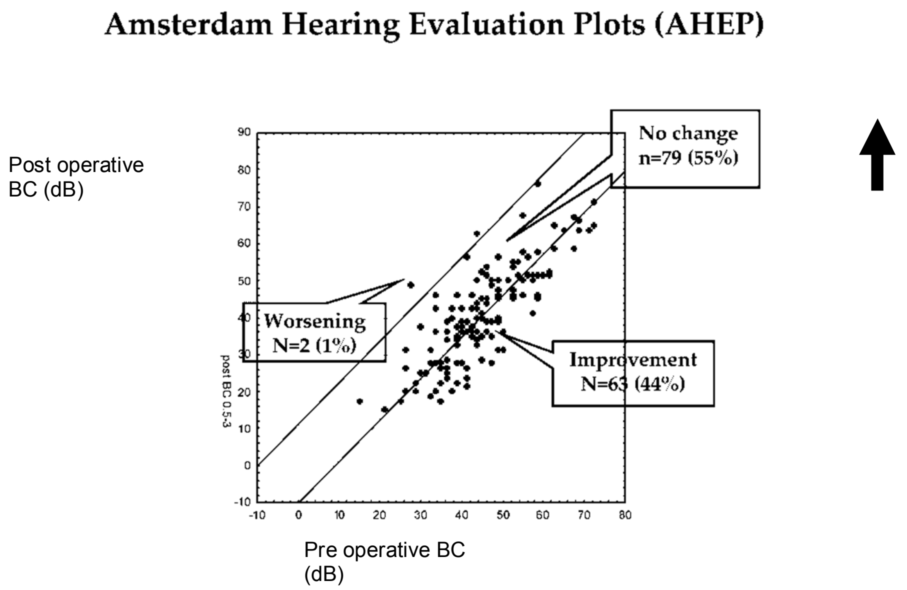


Figure 4. Pre-operative (x-axis) and post-operative (y-axis) bone conduction (BC) thresholds evaluated with the Amsterdam Hearing Evaluation Plots.

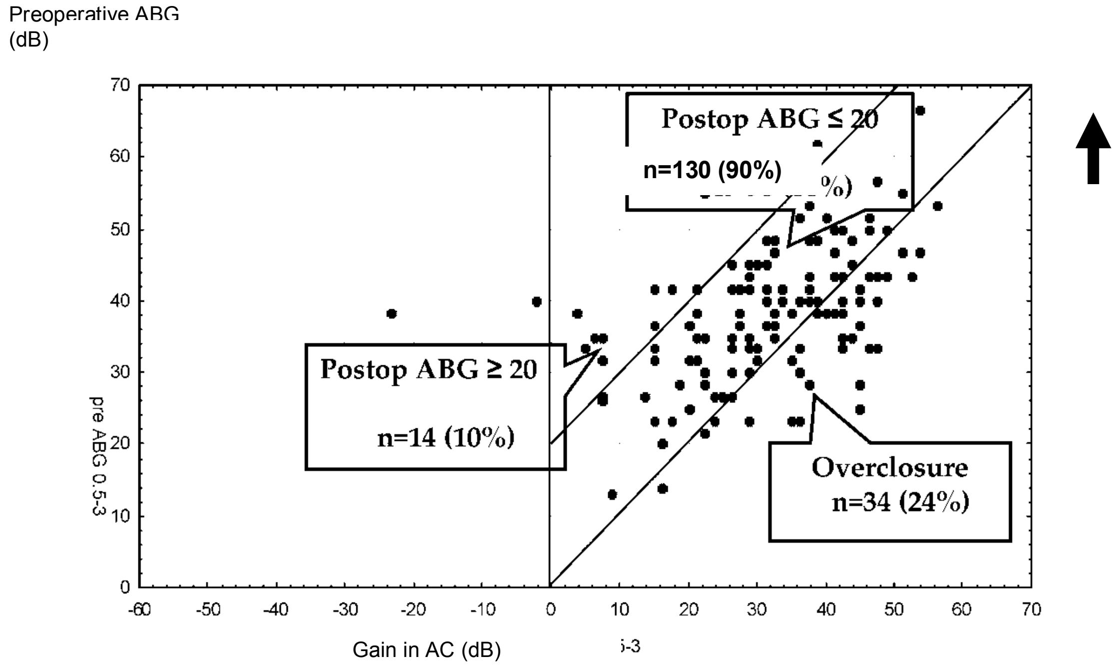


Figure 5. Evaluation of the gain in air conduction (AC) thresholds compared with pre-operative air-bone gap (ABG) with the Amsterdam Hearing Evaluation Plots.

Glasgow Benefit Plot

The most common type of pre-operative hearing impairment was a bilateral asymmetric HL in 81 cases (56%), followed by a bilateral symmetric hearing loss in 37 cases (26%) and a unilateral hearing loss in 26 cases (18%) (Figure 6). Postoperatively, the operated ear was converted into the better hearing ear in 65 (46%) cases. In the interpretation of these results, AC thresholds in the non-operated ear should be taken in consideration. The mean post-operative AC in the operated ear was 51 (± 17) dB and mean AC thresholds in the nonoperated ear were also 51 (± 24) dB. Symmetric hearing with an interaural difference of ≤ 15 dB was achieved in 63 (44%) cases. Normal or socially acceptable hearing (post-operative AC ≤ 30 dB) was achieved in the operated ear of 13 patients, that is, all those who had the potential to achieve normal hearing, with pre-operative BC thresholds ≤ 30 dB. Seventeen cases of 82 cases remained in a category of bilateral asymmetric hearing loss post-operatively (Figure 7).

Glasgow Benefit Plot (GBP)

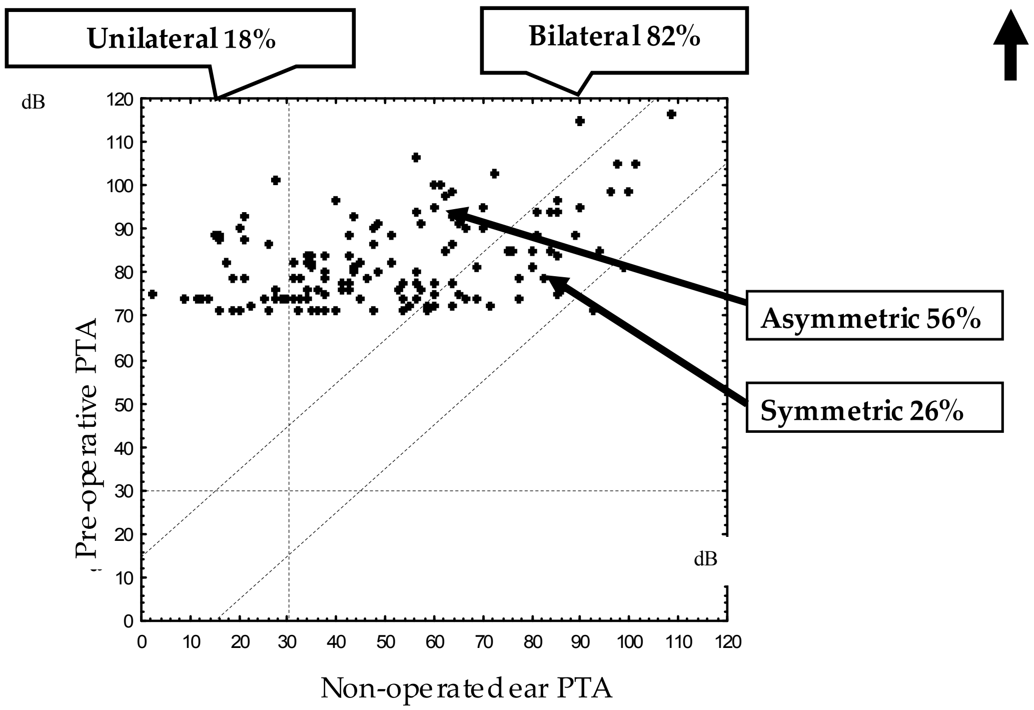


Figure 6. Pre-operative pure-tone average (PTA) of air conduction (AC) thresholds in operated (y-axis) and non-operated (x-axis) ears evaluated with the Glasgow Benefit Plot.

Additional post-operative symptoms

Nonauditory complaints are summarized in Table 4. Ten patients (7%) noticed temporary taste alteration; five patients (3%) reported occasional dizziness; and one patient complained of post-operative tinnitus. Tinnitus improved in seven patients, and pre-operative dizziness resolved in two patients.

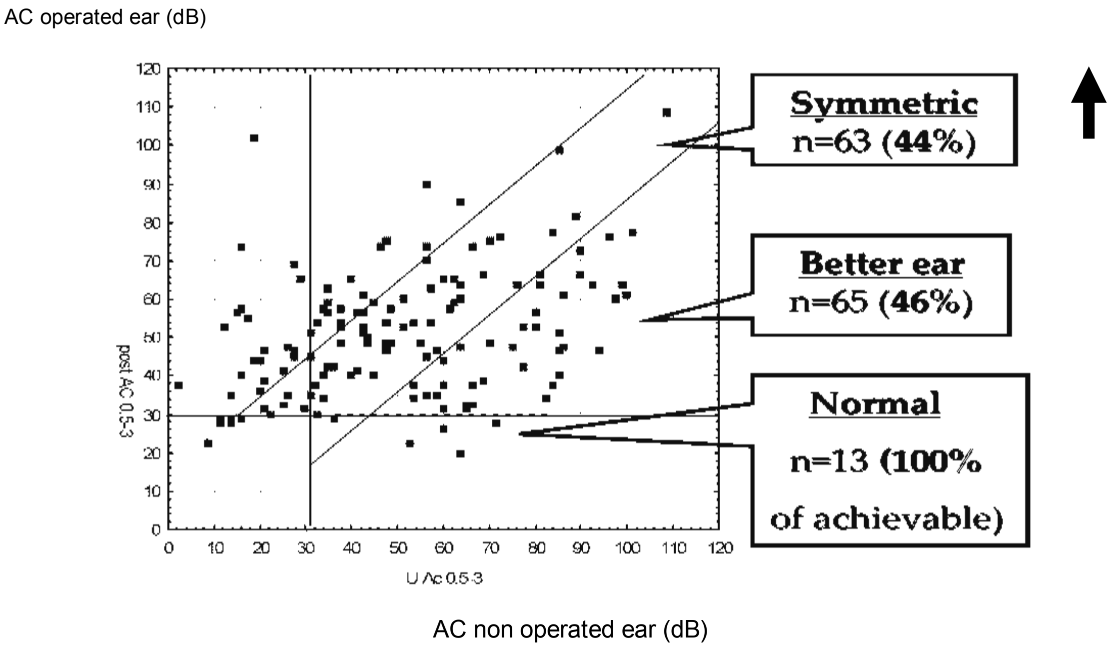


Figure 7. Post-operative pure-tone average of air conduction (AC) thresholds in operated (y-axis) and non operated (x-axis) easr evaluated with the Glasgow Benefit Plot.

Table 4. Additional symptoms.

	Pre-operative, n (%)	Improved, n (%)	New post-operative, n (%)
Tinnitus	40 (28%)	7 (18% 7/40)	1 (1%)
Vertigo	6 (4%)	2 (33% 2/6)	5 (3%)
Tinnitus and vertigo	4 (3%)	0	0
Taste alteration	0	0	1 (7%)

Data represent number of patients.

Discussion

Management of severe hearing loss in otosclerosis is challenging. The pre-operative diagnosis, choice of treatment and evaluation of surgical benefit remain controversial.

Audiometric presentation of advanced disease differs substantially from the typical audiologic characteristics of patients with early otosclerosis. The interpretation of the audiogram is difficult when there is a combination of SNHL in the better hearing ear and a mixed hearing loss in the second ear or a bilateral mixed SPHL.[15] Difficulties in bone masking in cases of bilateral asymmetric hearing loss often hinder the estimation of the actual ABG.[16,17] In patients with nonmeasurable AC thresholds or BC thresholds beyond the maximal output of audiometer, an ABG also cannot be reliably established.

The lack of typical audiometric signs of otosclerosis in these patients with a severe hearing loss requires additional diagnostic clues. Sheehy proposed several clinical diagnostic criteria, including a positive family history, a progressive pattern of hearing loss, a history of use of BC hearing aids, and evidence of an ABG from previous audiograms.[18] Shea and colleagues stated that the diagnosis of far-advanced otosclerosis, for the most part, could be easily made from the history and examination.[19] However, others have argued that the diagnosis of otosclerosis can only be confirmed by surgical exploration.[17,20]

Options for surgical treatment of advanced otosclerosis include stapedotomy or cochlear implantation (CI). Use of the middle ear implants in severe mixed hearing loss in otosclerosis has also been reported recently.[21] Decision making in the choice of treatment is often hindered by difficulties in diagnosis, quantification of sensorineural reserve and the controversies of treatment outcomes.

To help in the accuracy of the pre-operative assessment, Keith and colleagues reported the value of electrocochleography and promontory responses for confirmation of preserved cochlear function. [22]

Whereas Glasscock and colleagues noted that CI is too often discussed prior to consideration of its relevance,[23] Ramsden and colleagues suggested that CI may be the only effective treatment in cases of otic capsule involvement from the otosclerotic process.[24] Calmels and colleagues reported that objective and subjective results in the cochlear implant group were statistically better than in the stapedectomy group.[25] To the contrary, Khalifa and colleagues found that outcomes of stapes surgery are superior to CI. [26] Several other publications have suggested stapedectomy as a simpler, safer, and less extensive intervention with better chances for auditory rehabilitation compared with CI surgery.[19,23,27]

Previously, stapes surgery in advanced otosclerosis was believed to have a poorer prognosis than in the earlier stages of the disease. Several factors were reported as contributing to unfavourable outcomes. Shea and Farrior found that an obliterative footplate was frequently associated with bony round window closure.[28] Amedee and Lewis reported that advanced otosclerotic changes of the oval and round windows often required extensive drilling and were more frequently associated with excessive bleeding and post-operative SNHL.[29] Ramsay and Linthicum demonstrated that the presence of a mixed hearing loss at surgery increased the risk of profound cochlear loss.[6] The incidence of obliterative footplate in our series was two times higher in the group with SPHL compared with the group with MMHL (0.7% and 0.3%, respectively), but it was not accompanied by the higher complication rate that was reported in earlier studies. As mentioned by Glasscock and colleagues, who discussed the similar differences between the stapedectomy series from 1970 and 1994, the first and most important possible reason for improved outcomes is a superior surgical technique that was refined over time.[23]

Difficulty in measuring the benefit of surgery is one of the continued sources of controversy in the evaluation of treatment outcomes. Since unaided serviceable hearing was rarely achieved in previous reports, Sheehy stated that usual criteria for technical success reported as the degree of ABG closure are not feasible for evaluation of surgical results in advanced otosclerosis.[18] Keith and colleagues suggested that the purpose of stapedectomy in severe hearing loss is not necessarily the achievement of normal hearing, but in allowing a serviceable aided hearing.[22] The practical aim of surgery in this condition is to diminish the hearing impairment and facilitate post-operative amplification, rather than normal hearing, which is unattainable in the majority of these patients.[22] Glasscock and colleagues chose the post-operative ability to use a hearing aid as one criterion of success and concluded that a patient's satisfaction from restored ability using hearing aids is a more adequate measure of successful stapes surgery in profound hearing loss.[23]

Unaided serviceable hearing has rarely been achieved in previous reports. In the only study with a published detailed distribution of pre-operative BC thresholds, AC levels < 30 dB were reported in five of eight patients who had pre-operative BC thresholds < 30 dB, or 63% of what was potentially achievable.[27] Results from our series have demonstrated normal post-operative AC thresholds in 13 of 13 patients with pre-operative BC levels < 30 dB, or 100% of what was potentially achievable. Even though achievement of normal hearing is often impossible owing to poor pre-operative BC thresholds, 88% of patients in our series were converted from the category of SHPL to MMHL, or better, considerably facilitating their post-operative rehabilitation with hearing aids (Figure 3).

Amplification with hearing aids as an alernative to stapes surgery in otosclerosis was extensively debated in audiologic and otologic literature. The results of numerous studies can be summarized in the concise conclusion that the worse the hearing loss is, the more problems occur with fitting of hearing aids.[30-32.] This consensus, based on benefit surveys and quality of life studies, is also in agreement with extensive research underlying the latest advances in digital hearing aid technology. As was established, gain differences between nonspeech and speech signals are more pronounced in higher degrees of hearing loss. Attempts to achieve better speech recognition in patients with SPHL results in overamplifcation, which, in turn, is associated with greater loudness discomfort and poor sound quality, evaluated on 50, 65, and 85 dB input levels. Therefore, patients with 44 dB hearing levels poststapedotomy gain significant benefit in further hearing rehabilitation.[33,34] The unresolved challenges in hearing rehabilitation of SPHL with hearing aids also include greater feedback and occlusion effect.[35] Numerous disadvantages of this difficult for fitting hearing aids population resulted in agreement that for severe hearing losses, especially in the high frequencies, amplification is often ineffective.[36] Addressing the dilemma of use of hearing aids post-stapes surgery, Smyth and Hassard found that patients after small fenestra stapedotomy will require amplification on average 21 years postoperatively.[37] Frattali and Sataloff, discussing the management of profound deafness in the era of cochlear implants, advocated that otolaryngologists should not hesitate to offer stapes surgery to patients with far-advanced otosclerosis.[38] In our experience, patients with SHPL converted to MMHL by successful stapes surgery enjoy significant advantage of a better quality of hearing rehabilitation in general and speech recognition in particular. Postoperative gain in AC calculated for the four-frequency average was found to correlate with improvement in the SRT; therefore, better AC thresholds help fitting hearing aids and improve speech recognition in patients who still require postoperative amplification.[39]

Lippy and colleagues considered better speech understanding one of the main aims of stapes surgery for advanced otosclerosis.[40] The improvement of SRT's from 76 dB pre-operatively to 44 dB post-operatively in our group of patients can be seen as further evidence of the success of stapedotomy in the treatment of advanced otosclerosis. In studies concerning the results of stapedectomy for treatment of otosclerosis in the elderly, a high proportion of mixed hearing loss was found to correlate with age and advanced disease.[41-43] In our series, the mean age of patients with SPHL was 55 years, that is, 12 years older compared with the mean age of 43 years in the MMHL group (Table 1). The success rate in ABG closure in our study was also found to be lower in SPHL compared with MMHL (Table 3), in agreement with the similar findings reported by Persson and colleagues, Palva and Palva, Zaghis and colleagues.[8,20,44] However, Salvinelli and colleagues reported that despite greater post-operative ABG and AC thresholds in older patients with severe hearing loss, satisfaction from surgery in this group is higher when compared with younger patients with moderate hearing loss.[45] The differences in the surgical technique are that conventional stapedotomy with Cawthorne prosthesis(1991-1999), laser-assisted stapedotomy with Cawthorne prosthesis and laser-assisted stapedotomy with Causse prosthesis (2000-2001) have no significant effect on technical success.

Despite the higher proportion of mixed hearing loss and the older age of patients, only minor intraoperative complications were encountered in our series. No patients suffered severe cochlear damage. Post-operatively, 5 patients reported occasional dizziness; 10 patients noticed temporary taste alteration and 1 patient complained of new post-operative tinnitus. At the same time, pre-operative tinnitus improved in seven patients and pre-operative dizziness resolved in two patients (Table 4). Owing to the limitations of retrospective design, postoperative taste disturbance and dizziness may not be reported if they were not emphasized in the records. The dependence on clinical records is a common shortcoming of the lterature on advanced otosclerosis because a majority of publications that reported the results of stapedectomy are retrospective studies.[8,19,23,25,28,29,40-43,45]

The success rate in our series and low rate of complications supports other reports that stapes surgery is an effective treatment of severe hearing loss in otosclerosis.

Conclusions

Severe hearing loss in otosclerosis is not infrequent, comprising 13% of all primary stapedotomies in our series. The magnitude of hearing improvement post-stapedotomy is greater in SPHL when compared with MMHL. Improvement from SPHL to MMHL is possible in up to 88% of patients. Symmetric hearing could be achieved in two-thirds of patients, and even normal hearing can be achieved in selected cases. We conclude that, in our experience, the modified stapedotomy technique is a safe and effective treatment for advanced otosclerosis.

Acknowledgements: This work was made possible in part through the award of a TWJ Foundation fellowship to the second author.

References

1. Vartiainen E, Karjalainen S. Bone conduction thresholds in patients with otosclerosis. Am J Otolaryngol 1992;13:234-36.
2. Satar B, Sen D, Karahatay S et al., Effect of cochlear reserve on post-operative outcome in otosclerosis. Eur Arch Otolaryngol 2007;264:489-93.
3. Moscillo L, Imperiali M, Carra P et al., Bone conduction variation post-stapedotomy. Am J Otolaryngol 2006;27:330-33.
4. Johnson E. Hearing aids and otosclerosis. Otolaryngol Clin N Am 1993; 26:491-502.
5. Sanders J. The Successful hearing aid user. Otolaryngol Clin N Am 1978; 11: 187-93.
6. Ramsay H, Linthicum F. Mixed hearing loss in otosclerosis: indication for long-term follow-up. Am J Otolaryngol 1994;15:536-39.
7. Smith M, Hopp M. 1984 Santa-Barbara state-of-the-art symposium on otosclerosis. Results, conclusions, consensus. Ann Otol Rhinol Laryngol 1988;97:17-20.
8. Persson P, Harder H, Magnuson B. Using the Glasgow benefit plot as a prognostic instrument and for pre-operative counselling in patients with otosclerosis. Otol Neurotol 2007; 28:739-44.

9. Smyth G. Practical suggestions on stapedotomy. Laryngoscope 1982; 92:952-3.
10. Fish U. Stapedotomy vs. stapedectomy. Am J Otol 1982; 4:112-17.
11. Committee on Hearing and Equilibrium guidelines for the evaluation of results of treatment of conductive hearing loss. Otolaryngol Head Neck Surg 1995; 113:186-7.
12. Browning GG, Gatehouse S, Swan IR. The Glasgow Benefit Plot: a new method for reporting benefits from middle ear surgery. Laryngoscope 1991;101: 180-185.
13. Toner JG, Smyth GDL. Comparison of methods of evaluating hearing benefit of middle ear surgery. J Laryngol Otol 1993;107: 4-5.
14. De Bruijn AJG, Tange RA, Dreschler WA. Efficacy of evaluation of audiometric results after stapes surgery in otosclerosis. A method for reporting results from individual cases. Otolaryngol Head Neck Surg 2001; 124: 84-9.
15. Giles M, Browning G, Gatehouse S. Problems in assessing the audiogram in patients with severe hearing impairment. J Laryngol Otol 1996; 110: 727-31.
16. Nauton R. A masking dilemma in bilateral conduction deafness. Arch Otolaryngol 1960; 72:753-57.
17. House W, Glorig A. Criteria for otosclerosis surgery and further experiences with round window surgery. Laryngoscope 1960;69:693-701.
18. Sheehy J. Surgical correction of far advanced otosclerosis. Otolaryngol Clin N Am 1978;11: 121-3.
19. Shea P, Ge X, Shea J. Stapedectomy for far-advanced otosclerosis. Am J Otol 1999; 20: 425-29.
20. Palva T, Palva A. Surgery in profound hearing loss. Acta Otolaryngol 1968; 65: 565-9.
21. Venail L, Meller R, Deveze A, Tardivet L, Magnan J. New perspectives for middle ear implants: first results in otosclerosis with mixed hearing loss. Laryngoscope 2007; 117:552-5.
22. Keith R, Kereiakes T, Willging J, Devine J. Evaluation of cochlear function in a patient with far-advanced otosclerosis. Am J Otol 1992;13:347-9.
23. Glasscock ME, Storper IS, Haynes DS, Bohrer PS. Stapedectomy in profound hearing loss Laryngoscope 1996;106:831-3.
24. Ramsden R, Rotteveel L, Proops D, Saeed S, van Olphen A, Mylanus E., Cochlear implantation in otosclerotic deafness. Adv Otolaryngol 2007;65:328-34.
25. Calmels M, Viana C, Wanna G et al., Very far-advanced otosclerosis: stapedotomy or cochlear implantation. Acta Otolaryngol 2007;127: 574-8.
26. Khalifa A, El-Guindy, Efran E. Stapedectomy for far-advanced otosclerosis. J Laryngol Otol 1998;112:158-60.
27. Wiet RJ, Morgenstein SA, Zwolan TA, Pircon SM. Far-advanced otosclerosis. Cochlear implantation vs stapedectomy Arch Otolaryngol 1987;113:299-302.
28. Shea J, Farrior J. Stapedectomy and round window closure. Laryngoscope 1987; 97:10-12.
29. Amedee R, Lewis M. Obliterative otosclerosis. Laryngoscope 1987;97:922-4.
30. Stark P, Hickson L. Outcomes of hearing aid fitting for older people with hearing impairment and their significant others. Int J Audiol 2004; 43: 390-8.
31. Meister H, Lausberg I, Kiessling J, et al. Detecting components of hearing aid fitting using a self-assessment inventory. Eur Arch Otolaryngol 2005; 262: 580-6.
32. Metselaar M, Maat B, Krijnen P, et al. Self-reported disability and handicap after hearing aid fitting and benefit of hearing aids. Eur Arch Otolaryngol 2009; 266: 907-17.
33. Souza P, Tremblay K. New perspectives on assessing amplification effects. Trends Amplif 2006; 10: 119-43.
34. Henning RW, Bentler R. Compression-dependent differences in hearing aid gain between speech and non-speech signals. Ear Hear 2005; 26: 409-22.
35. Chung K. Challenges and recent developments in hearing aids. Trends Amplif 2005; 8: 125-64.
36. Turner CW. Hearing loss and the limits of amplification. Otol Neurotol 2006; 11 Suppl 1: 2-5.
37. Smyth G, Hassard T. Hearing aids post stapedectomy: incidence and timing. Laryngoscope 1986; 96: 385-8.
38. Frattali M,Sataloff R. Far-advanced otosclerosis. Ann Otol Rhinol Lryngol 1993;103: 433-7.
39. De Bruijn AJG, Tange RA, Dreschler WA. Efficacy of evaluation of audiometric results after stapes surgery in otosclerosis. The effects of using different audiologic parameters and criteria on success rates. Otolaryngol Head Neck Surg 2001;124: 76-83.
40. Lippy W, Burkey J, Arkis P. Word recognition score changes after stapedectomy for far advanced otosclerosis. Am J Otol 1998; 19:56-58.
41. Iurato S, Bux G, Mevoli S et al., Stapes surgery in elderly. Adv Otolaryngol 2007;65:231-36.

42. Lippy W, Burkey J, Fucci M et al., Stapedectomy in the elderly. Am J Otol 1996;17: 831-4.
43. Ayache D, Corre A, Van Prooyen S, Elbaz P. Surgical treatment of otosclerosis in elderly patients. Otolaryngol Head Neck Surg 2003;129:674-77.
44. Zaghis A, Del M, Ambrosetti U. Some observations concerning 200 stapedectomies: fifteen years post-operatively. Laryngoscope 1997;97:1211-13.
45. Salvinelli F, Casale M, Trivelli M, Di Peco V, Greco F. Otosclerosis and stapedoplasty in older adults. J Am Ger Soc 2002;50:1396-400.

3.2

The Markham Stouffville Hospital Experience with malleo-vestibulopexy

V. E. Kisilevsky
N.A. Bailie
S.N. Dutt
A. Safar
J.J. Halik

Journal of Otolaryngology – Head & Neck Surgery; 2009; 38:1-7

Introduction

Conductive hearing loss (CHL) caused by middle ear pathology can be successfully treated by surgical correction in many cases. The most common causes of CHL are chronic otitis media and otosclerosis. Congenital anomalies of ossicular chain and trauma to the middle ear are relatively rare. The potential for hearing improvement is dependent on the pathology impeding sound conduction. Stapes surgery for isolated stapes fixation is generally considered the most successful in correction of CHL.[1-4] The success rates of ossiculoplasty are greatly affected by the degree of middle ear pathology and can vary substantially.[5-8] The long process of incus and the incudo-stapedial joint have been shown to be the least robust region of the ossicular chain owing to their thin structure and poor blood supply.[9-11] Various surgical techniques and types of prostheses have been designed for incus interposition or replacement and applied with favourable results.[12-14] However, when more than one ossicle is missing or immobile, surgical outcomes are less successful.[15-16] Reported results of total ossicular replacement are rarely as good as those of primary stapedotomy and are less stable in the long term.[17-20] In a situation in which the stapes footplate is fixed and the incus is missing or abnormal, reconstruction of conductive mechanism is particularly challenging. The aims of this study were to analyze the hearing results of the malleovestibulopexy technique in the special situations encountered in revision stapedotomies and congenital middle ear malformations.

Patients and Methods

The charts of 1369 consecutive stapedotomies performed by one surgeon (senior author J.J.H.) from 1991 to 2006 were reviewed. Of the 1369 surgical cases, 89 were revision stapedotomies and 48 were performed for congenital stapes fixation. The diagnosis of congenital stapes fixation was based on a history of non-progressive CHL from early childhood and an intraoperative finding of hypoplastic or malformed stapes, incus and/or incudostapedial joint in the absence of an otosclerotic focus. A malleovestibulopexy (MVP) technique was employed for restoration of ossicular chain function in 24 cases. In 16 patients (1 bilateral case), MVP was used in revision stapes surgery, and in 7 patients MVP was employed for treatment of congenital middle ear anomalies. The combination of a missing or eroded incus with immobile footplate was found in 17 of 89 (19%) revision stapedotomies and in 7 of 48 (15%) of congenital anomalies. The audiometric results of 24 MVP procedures were included in this study. Sixteen patients (75%) were women and seven (25%) were men. The mean age was 44 years (SD 13 years), with a range from 13 to 71 years. The mean follow-up was 20 months (SD 18 months), with a range of 2 to 65 months**.** Three patients had a follow–up less than 12 months. One patient, with a follow-up of 2 months, was diagnosed intra-operatively with a combined congenital ossicular anomaly involving the hypoplastic incus, malformed stapes and incudostapedial joint. Postoperative audiometry demonstrated worsening of the Air Bone Gap (ABG) in this case. These results have been included to report all MVP cases without concealing unfavourable outcomes. Eighty-seven cases out of 1369 cases were excluded from the analysis as audiometric follow up was not available. The majority of these patients were referred from distant locations and have continued follow-up with their referring physician. MVP was not performed in any of these cases.

MPV was performed via a per meatal approach. A posterior tympanomeatal flap was elevated from 12 o'clock to the 6 o'clock position beginning approximately 5 to 7 mm from the tympanic annulus and reflected anteriorly. The chorda tympani was identified, mobilized and displaced inferiorly. Using a bone curette, the posterior-superior aspect of the bony tympanic annulus was curetted back to provide adequate exposure of the pyramidal process and gain access to the footplate, if not done previously, and strict haemostasis was secured. The ossicular chain was then inspected to identify incus erosion or malformation and palpated to ensure that the assumption of stapes fixation was correct. The tympanomeatal flap was extended anteriorly to gain exposure to the manubrium of the malleus. The tympanic membrane was dissected from the midportion of the handle of the malleus.

For congenital malformations, an argon laser (Lumenis Inc, Salt Lake City, UT) was used in conjunction with a 0.2 mm Otoprobe fibre-optic delivery system (Iridex Corp, Mountain View, CA) for stapedial tendon and superstructure removal and initiating the footplate fenestration. In revision stapedotomies, laser was beneficial in dealing with adhesions. Beginning with an initial power setting of 2000 mW and a pulse duration of 0.2 seconds, the stapedius tendon was vaporised. A series of laser pulses was then delivered to the posterior crus of the stapes, causing it to turn to ash and crumble easily. With the stapedius tendon and posterior crus of the stapes thus removed, wide access was gained to the stapes footplate. The laser power was adjusted to1500 mW and the pulse duration reduced to 0.1 seconds. A laser pulse was then delivered to the posterosuperior quadrant of the stapes footplate and repeated until perilymph just began to 'sweat' through the laser burn. The laser burn on the footplate provided a set-hole for a series of hand perforators. The stapedotomy was gently widened, beginning with a 0.3 mm perforator and increasing in 0.1 mm steps to a 0.6 mm perforator, which provided a final stapedotomy diameter of 0.5 mm. Suction on the open footplate was strictly avoided. A 6 x 0.4 mm Causse large-loop Teflon prosthesis or 5 X 0.3 mm Cawthorne prosthesis, modified such that the shaft length is reduced to 4 to 5 mm and with a small wedge cut from the ring, was then placed on the handle of the malleus with the distal end articulating through the fenestra in the stapes (Figure 1). Malleus manipulation was minimal and in no cases was the malleus dislocated or subluxed. The tympanic membrane was then replaced and a dressing was placed into the external auditory canal to be removed at the outpatient clinic the following day.

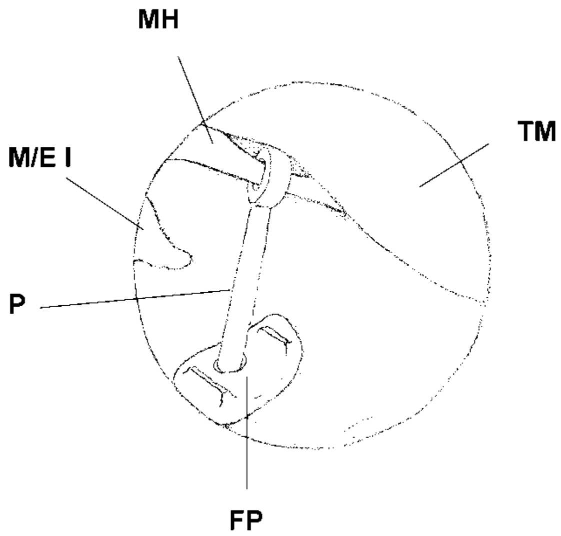


Figure 1. Prosthesis position on the footplate (FP) and malleus handle (MH). M/E I-malformed/eroded incus; P = modified Cawthorne-Causse prosthesis; TM – tympanic membrane

The hearing outcomes were analyzed according to the 1995 American Academy of Otolaryngology - Head and Neck surgery Committee on Hearing and Equilibrium guidelines (AAO-HNS CHE).[21] Frequency-specific air conduction (AC) thresholds at 0.5, 1, 2, 3 and 4 kHz and bone conduction (BC) thresholds at 0.5, 1, 2, 3 and 4 kHz in operated and nonoperated ears were tabulated in a computer database. As the beginning of this study period predates the 1995 AAO-HNS guidelines, 3 kHz results were not available for all patients. In these cases, 3 kHz AC and BC was estimated by average between 2 and 4 kHz, allowing a uniform presentation of audiometric data in compliance with AAO-HNS guidelines. The

mean preoperative pure-tone average (PTAs) of AC, BC and ABG at 0.5, 1, 2 and 3 kHz were compared with corresponding postoperative values. The archived charts of all consecutive stapes surgery cases were reviewed. Audiograms were accessed by the first and by the senior authors. The ABG was calculated by subtracting the postoperative BC PTA at the frequencies 0.5, 1, 2 and 3 kHz from postoperative AC PTA at the same frequencies. CHL was defined as ABG > 10 dB and a PTA BC ≤25 dB. Mixed hearing loss was defined as an ABG > 10 dB and a PTA BC ≥25 dB. Pre- and postoperative speech reception thresholds (SRT) were also evaluated.
The mean preoperative AC calculated as a PTA at frequencies 0.5, 1, 2 and 3 kHz was 54 dB (SD 14 dB), and for the mean preoperative BC was 21 dB (SD 11 dB). The mean preoperative ABG was 33 dB (SD 11 dB). One patient presented with severe hearing loss (preoperative PTA 84 dB) and one patient with profound hearing loss (preoperative PTA 95 dB). Mixed hearing loss with BC ≥ 25 dB was demonstrated in five patients, two with congenital ossicular malformations and three with revision surgery. In four patients, preoperative BC was worse than 30 dB. It is customary to define socially serviceable hearing as a PTA AC of ≤ 30dB. The BC thresholds provide the limit for possible improvement of AC; therefore BC thresholds >30dB preclude the possibility of postoperative socially serviceable hearing.
The database was analyzed with a statistical software program *STATISTICA 6.0*, (StatSoft, Inc., Tulsa, OK). Statistical analysis was carried out for comparisons between pre- and postoperative values using a paired t-test for the evaluation of AC, BC, SRT, and ABG. The criterion selected for statistical significance was $p < .05$.

Results

Air Conduction, Bne Cconduction, Air Bone Gap, Speech Reception Threshold

Significant ($p < .001$) improvement was demonstrated postoperatively in all audiometric parameters, including AC, BC, ABG and SRT (Table 1). One patient showed worsening of ABG; he was lost for follow-up 2 months after the surgery. Frequency-specific results are demonstrated in Table 2.
ABG closure ≤ 20 dB and ≤ 10 dB was achieved in 61% and 33% of all cases, respectively.
The mean ABG in a group (n = 20) with a follow-up of more than 12 months was 18.6 dB compared with 20.7 dB in a group of four patients with follow-up of less than 12 months.

Table 1. Pre- and postoperative AC, BC, ABG, and SRT at 0.5, 1, 2, and 3 kHz.

	AC	BC	ABG	SRT
Preoperative	54 (14)	21 (11)	33 (11)	53 (13)
Postoperative	39 (19)	19 (8)	20 (14)	35 (19)

ABG - air-bone gap; AC- air conduction; BC - bone conduction; SRT - speech reception threshold.

Table 2. Pre- and post-operative air and bone conduction by frequency.

	Frequency (kHz)	Pre-op Mean (SD)	Post-op Mean (SD)
AC (dB)	0.5	57.7 (14.1)	37.7 (23.8)
	1	55.0 (13.9)	37.5 (21.6)
	2	49.7 (16.7)	37.0 (18.9)
	3	52.5 (16.6)	42.9 (16.1)
	4	55.9 (18.9)	47.5 (18.1)
BC (dB)	0.5	18.8 (10.1)	18.8 (12.3)
	1	17.0 (12.3)	15.4 (8.9)
	2	24.5 (12.0)	20.6 (11.1)
	3	23.4 (12.4)	21.8 (10.1)
	4	20.4 (12.7)	22.9 (11.7)

AC - air conduction; BC - bone conduction.

Table 3. Postoperative Air-Bone Gap in revision and congenital groups.

	Postoperative ABG (96)		
Group	Mean (SD)	≤ 10 dB	≤ 20 DB
Revision	21.0 (13.1)	29%	53%
Congenital	16.6 (16.3)	57%	72%

ABG - air-bone gap.

The technical success of surgery measured by postoperative ABG closure was higher in a group of seven patients with congenital middle ear anomalies compared with a group that underwent revision surgery. ABG ≤ 10 dB was achieved in 57% of congenital anomalies and in 29% of revision procedures (Table 3).

Complications

None of the patients suffered sensorineural hearing loss (SNHL) postoperatively. One patient complained of transient vertigo spells induced by Valsalva manoeuvre; these symptoms resolved spontaneously after 1 month.

Nine patients complained of tinnitus preoperatively. Two patients reported resolution of tinnitus following postoperative hearing improvement. None reported it worsening.

Discussion

The main reasons for abnormal sound conduction are impaired mobility or loss of integrity of middle ear structures. Surgical methods for treatment of CHL are aimed at restoring the middle ear mechanism of impedance matching.

Restricted mobility of middle ear conductive mechanism is most commonly caused by otosclerosis. Surgical treatment of otosclerosis has evolved from initial attempts at stapes mobilization through fenestration techniques to stapedectomy and, more recently, to small fenestra stapedotomy. Current techniques of stapes surgery are highly successful in correction of the CHL caused by otosclerosis. Closure of an ABG < than 20 dB has been reported in 95 to 98% of cases.[22,23] Stapedotomy for tympanosclerotic stapes fixation is less successful than in otosclerosis and allows ABG closure within 20 dB in 58 to 70% of cases.[24-26] Success of stapes surgery in tympanosclerosis is dependent on the extent of tympanosclerotic ossicular involvement. Evaluating results of surgical treatment of tympanosclerosis, Tos and colleagues found that the best results were achieved in the group with stapes mobilization and an intact ossicular chain compared with any combinations of stapes mobilization or stapedectomy with ossiculoplasty. [27]

Otitis media (OM) with or without cholesteatoma is the most common cause affecting continuity of the ossicular chain. Historically, the surgical treatment of OM was intended to control chronic ear discharge and to prevent complications, and no attempts were made to reconstruct the deficit caused by destructive disease or surgery. Tympanoplasty as a method to restore the integrity of the middle ear conductive mechanism was first reported by Zollner in 1951[28] and by Wullstein in 1952[29]. Numerous modifications in ossiculoplasty techniques have been made with the aim of better and stable hearing results. Incus interposition with an auto- or homograft allowed ABG closure within 20 dB in two-thirds of cases[30,31] but has a failure rate of 15 to 20%.[32] To overcome the tendency to refixation of interimposed ossicles and to improve the stability of the assembly, Shea and Homsy in 1974 introduced the alloplastic Plastipore prosthesis for partial ossicular replacement (PORP) and total ossicular replacement prosthesis (TORP).[33] Trials of different prosthesis materials, including plastics, ceramics, and metals, have been attempted during the past decades. With the technological advances employed in ossicular prosthesis design and materials, the success of middle ear reconstruction remains mainly dependent on the extent of ossicular damage. Thus, Sasaki and colleagues found that the results of type III tympanoplasty were significantly superior to those of type IV. [34] In other studies of prognostic factors in ossiculoplasty, only absent malleus was found to be a significant unfavourable factor affecting postoperative ABG, regardless of the type of prosthesis.[35,36]

As a rule, given similar Eustachian tube function and condition of the middle ear mucosa, better hearing results were reported when only one ossicle was replaced compared with multiple ossicles or total ossicular chain reconstruction. Jackson and colleagues in 1983 achieved ABG closure within 20 dB in 49% of cases using PORP compared with 43% of cases when TORP was used. [37] A similar proportion between success rates of PORPs and TORPs was reported recently by Truy and colleagues[38], Yung[17] and Schmerber and colleagues.[18] According to these recent studies, ABG closure within 20 dB was achieved in 77 to 61% of cases with PORPs and in 38 to 52% of cases with TORPs.

Established surgical methods for treatment of common middle ear diseases meet with much less success in cases of combined middle ear pathologies. The situation in which one segment of the ossicular chain is missing and another is immobile requires considerable creativity and remains challenging, even for the experienced otologist. Revision stapedotomy is generally less successful than primary stapes surgery, not least because of the frequent combination of incus erosion with an immobile footplate.[39-41] Incus erosion was the second most common cause of failure of primary stapedectomy (after prosthesis malfunction) in series of revision stapedectomies reported by Farrior and Sutherland[42], Glasscock and colleagues[43] and Derlacki[44] and the most common finding in the series of revision stapedectomies published by Langman and Lindeman.[45] In addition to erosion, incus dislocation or fracture of the long process may also happen during either primary or revision surgery.[46] Otosclerotic involvement of the incus itself has also been reported.[47] When no useful incus is present, application of the standard technique of stapes prosthesis placement is impossible. Chronic otitis media can also result in a similar combination of ossicular erosion and fixation caused by tympanosclerosis.[25] In addition, congenital stapes ankylosis associated with ossicular chain anomalies has been described in Klippel-Feil syndrome[48], Pfeiffer's syndrome[49], lacrimoauriculodentodigital syndrome[50], osteogenesis imperfecta[51], dyschondrosteosis[52], and a more recently recognized autosomal dominant inherited syndrome with congenital stapes ankylosis.[53] Many authors have emphasized that multiple sites of ossicular pathology present a perplexing surgical problem that is difficult to solve.[37,54,55]

Several methods have been reported for reconstruction of the sound conductive mechanism in cases in which the incus is absent or inadequate and the stapes footplate is fixed. Shea proposed two-stage stapedectomy and ossiculoplasty in 1976 and later a one-stage technique using a malleus attachment piston prosthesis.[56,57] Sheehy in 1982 described stapedectomy with an incus replacement prosthesis.[58] Feldman and Schuknecht in 1970 introduced a malleus to oval window prosthesis placement in revision stapedectomies.[59] Schuknecht and Barley later modified this in 1986 using a malleus grip prosthesis.[60] Although the details of the surgical techniques differ, the common aim of these proposed methods was achievement of connection between the malleus handle and the vestibule exposed by stapedectomy or stapedotomy, or malleovestibulopexy (MVP), also known as malleostapedotomy. In the case of more extensive pathology, where both the incus and the malleus are missing, Berenhoilz and Lippy proposed total ossiculoplasty with footplate removal,[61] whereas Cremers described a method of myringochorda-vestibulopexy.[62] Evaluating the results of MVP in revision stapes surgery, Fisch and colleagues reported an ABG closure within 20 dB in 63% of cases and within 10 dB in only 14% of cases.[63] Similarly, Farrior and Sutherland found that the hearing results of revision stapedectomy are not as good in the case of malleus-incus erosion as they are in case of a displaced prosthesis.[42] Huttenbrink suggested that MVP techniques potentially carry more risk of inner ear damage because of large amplitude of pressure-induced displacement of the malleus.[64] Tange, however, was able to demonstrate ABG closure within 20 dB in 88% of cases in which the incus was inadequate after previous stapedectomy or owing to congenital malformations, using a malleus attachment piston Teflon prosthesis.[54] Hashimoto and colleagues also found that the outcomes of malleostapedotomy are superior to those of total stapedectomy with ossiculoplasty in the treatment of incudostapedial joint discontinuity with stapedial fixation.[65] In a small series of five patients treated with a malleus to oval window wire piston technique, Kohan and Sorin reported ABG closure within 20 dB in all cases.[66] ABG reduction from 36 dB to 13 dB after surgery has been reported in six patients treated with MVP by Dalchow and colleagues.[67] Follow-up in this report, however, was limited to 3 months only.

Analysis of the results in our study demonstrated success rates similar to those reported in other compatible series.[17,18] These short–term audiometric results of MVP in our series are equal or superior to results of TORP ossiculoplasty reported in literature[37,38], in which a comparable degree of ossicular pathology has been described. This study is weakened owing to the relatively small number of cases, which diminishes the power of the statistical analysis. Isolated congenital ossicular chain anomalies requiring the use of MVP are rare. As the number of primary stapes surgery procedures declines worldwide, revisions with MVP are also becoming less frequent. This limits the potential for mastering this technique and reporting surgical results to few highly specialized otology centres. The size of recently published series reporting variation of this technique varies from 6 to 10 patients in some studies[40,61,67] to 82 patients reported by Fisch and colleagues.[63] Therefore, reporting early postoperative results of a particular modification of the MVP technique, employing laser-assisted stapedotomy with a modified self-crimping prosthesis, provides data enabling further comparisons with other studies. No cases of SNHL ("dead ear") were associated with this technique in our series. One patient demonstrated short-term vestibular symptoms. In our opinion, MVP offers another useful procedure in the armamentarium of the otologist dealing with complex and ossicular pathology.

Conclusions

Complex ossicular chain pathology is not uncommon, comprising 19% of revision stapedotomies and 15% of congenital middle ear malformations in our series. When surgical options for middle ear reconstruction are limited owing to stapes immobility combined with an absent or eroded incus, MVP offers a useful alternative, enabling good hearing results. In our experience, the laser-assisted MVP technique is associated with low risk of postoperative SNHL and vestibular symptoms. Therefore, MVP is a valuable surgical option in cases for which traditional ossiculoplasty is not feasible. Further follow-up is required to test the long-term stability of MVP assembly.

References

1. Shea J. Forty years of stapes surgery. Am J Otol 1998; 19:52-55.
2. Sheehy JL, Nelson RA, House HP. Stapes surgery at the Otologic Medical Group. Am J Otol 1979; 22-26.
3. Marquet T. Stapedotomy technique and results. Am J Otol 1985; 6:63-67.
4. Smyth GDL, Hassard TH. Eighteen years experience in stapedectomy. Ann Otol Rhinol Laryngol 1978: 87:3-36.
5. Sismanis A. Tympanoplasty *in* Glasscock -Shambaugh *Surgery of the ear.* Glasscock M, Gulya A (ed) BC Decker 2003.
6. Chole R. Ossiculoplasty with presculpted banked cartilage. Otolaryngol Clin N Am 1994; 27: 717-26.
7. Schuring A. Ossiculoplasty with semibiologic and composite prostheses. Otolaryngol Clin N Am 1994; 27: 747-57.
8. Bojarb D, Causse J, Battista R. Ossiculoplasty with composite prostheses. Overview and analysis. Otolaryngol Clin N Am 1994;27:759-76.
9. Nager G, Nager M. The arteries of the human middle ear, with particular regard to the blood supply of the auditory ossicles. Ann Otol Rhinol Laryngol 1953;62:923-49.

10. Mikhovich PG. Blood supply of the human incudo-stapedial joint. Vestn Otorinolaringol 1975; 2:28-31.
11. Alberti P. The blood supply of the long process of the incus and the head and neck of the stapes. J Laryngol Otol 1965;79:964-70.
12. Wehrs R. Incus interposition and ossiculoplasty with hydroxyapatite prostheses. Otolaryngol Clin N Am 1994; 27:677-88.
13. McElveen J. Ossiculoplasty with polymaleinate ionomeric prostheses. Otolaryngol Clin N Am 1994;27:777-84.
14. Kyrodimos E. Sismanis A. Santos D. Type III cartilage "shield" tympanoplasty: an effective procedure for hearing improvement. Otolaryngol- Head & Neck Surg 2007;136:982-5.
15. Austin D. Avoiding failures in the restoration of hearing with ossiculoplasty and biocompatible implants. Clin N Am 1982;15:763-71.
16. Wehrs R. Homograft tympanoplasty. Clin N Am 1982;15:781-93.
17. Yung M. Long-term results of ossiculoplasty: reasons for surgical failure. Otol Neurotol 2006; 27:20-6.
18. Schmerber S. Troussier J. Dumas G. Lavieille JP. Nguyen DQ. Hearing results with the titanium ossicular replacement prostheses. Eur Arch Oto-Rhino-Laryngol 263:347-54, 2006.
19. Sasaki T. Xu A. Ishimoto S. Ito K. Yamasoba T. Kaga K. Results of hearing tests after total middle ear reconstruction. Acta Oto-Laryngol 2007;127:474-9.
20. Vincent R, Sperling NM, Oates J et al. Surgical findings and long-term hearing results in 3050 stapedotomies for primary otosclerosis: a prospective study with the otology-neurotology database. Otol Neurotol 2006;27:S25-S47.
21. Committee on Hearing and Equilibrium guidelines for the evaluation of results of treatment of conductive hearing loss. Otolaryngol Head Neck Surg 1995;113:186-22.
22. Shea J, Sanabria F, Smyth G. Teflon Piston operation for otosclerosis. Arch Otolaryngol. 1962;76:516-21.
23. Hough J, Dyer R. Stapedectomy. Causes of failure and revision surgery in otosclerosis. Otolaryngol Clin N Am 1993;26:453-70.
24. Gormley P. Stapedectomy in tympanosclerosis. A report of 67 cases. Am J Otol 1987;8:123-30.
25. Teufert K, De La Cruz A. Tympanosclerosis: long-term hearing results after ossicular reconstruction. Otolaryngol H&N Surg 2002;126:264-72.
26. Vincent R, Oates J, Sperling N. Stapedotomy for tympanosclerotic stapes fixation: is it safe and efficient? A review of 68 cases. Otol Neurotol 2002;23:866-872.
27. Tos M, Lau T, Arndal H, Plate S. Tympanosclerosis of the middle ear: late results of surgical treatment. J Laryngol Otol 1990;104:685-9.
28. Zollner F. Die Radikal-Operation mit besonderem Bezug auf die Horfunktion. Ztschr Laryngol Rhinol Otol 1951;30:104.
29. Wullstein H. Funktionelle Operationen im Mittelohr mit Hilfe des freien Spaltlappen-Transplantates. Arch Ohren-Nasen-u-Kehlkopfh 1952;161:422.
30. O'Reilly R, Cass S, Hirsh B, Kamerer D, Bernat R. et al. Ossiculoplasty using incus interposition: hearing results and analysis of the middle ear risk index. Otol Neurotol 2005; 26:853-858.
31. Al-Qudah M, Dawes P. Malleus-stapes assembly: experience with two prostheses. J Laryngol Otol 2006;120:736-9.
32. Austin D. Avoiding failure in restoration of hearing with ossiculoplasty and biocompatible implants. Otolaryngol Clin N Am 1982;15:763-72.
33. Shea J, Homsy C. The use of proplast in otologic surgery. Laryngoscope 1974;84:1835.
34. Sasaki T, Xu A, Ishimoto S, Ito K, Yamasoba T, Kaga K. Results of hearing tests after total middle ear reconstruction. Act Oto-Laryngol 2007;127:474-9.
35. De Vos C, Gersdorff M, Gerard J. Prognostic factors in ossiculoplasty. Otol Neurotol 2007; 28:61-7.
36. Yung M, Vowler S. Long-term results in ossiculoplasty: an analysis of prognostic factors. Otol Neurotol 2006;27:874-81.
37. Jackson C, Glasscock M, Schwarber M, Nissen A, Christiansen S, Smith P. Ossicular chain reconstruction: the TORP and PORP in chronic ear disease. Laryngoscope 1983;93:981-8.
38. Truy E, Naiman A, Pavillon C, Abedipour D, Lina-Grande G, Rabilloud M. Hydroxyapatite versus titanium ossiculoplasty. Otol Neurotol 2007;28:492-8.

39. Gros A, Vatovec J, Zargi M, Jenko K. Success rate in revision stapes surgery for otosclerosis. Otol Neurotol 2005;26:1143-48.
40. Magliulo G, Cristofari P, Terranova G. Functional results in revision stapes surgery. Am J Otol 1997;18:408-12.
41. De La Cruz A, Fayad J. Revision stapedectomy. Otolaryngol Head Neck Surg 2000;12:728-32.
42. Farrior J, Sutherland A. Revision stapes surgery. Laryngoscope 1991;101:1155-61.
43. Glasscock M, McKenna K, Levine S. Revision stapedectomy surgery. Otolaryngol Head Neck Surg 1987;96:141-48.
44. Derlacki E. Revision stapes surgery: problems and some solutions. Laryngoscope 1985;95: 1047-53.
45. Langman A, Lindeman R. Revision stapedectomy. Laryngoscope 1993;103:954-58.
46. Golabek W, Szymanski M, Siwiec H, Morshed K. Incus subluxation and luxation during stapedectomy. Ann U M Curie 2003;58:302-5.
47. Escada P, Capucho C, Chorado M, Da Silva J. Otosclerosis of the incus. Otol Neurotol 2007; 28:301-3.
48. Sakai M, Miyake H, Shinkawa A, Komatsu N. Klippel-Feil syndrome with conductive deafness and histological findings of removed stapes. Ann Otol Rhinol Laryngol 1983; 92:202-6.
49. Cremers CW. Hearing loss in Pfeiffer's syndrome. Int J Ped Otolaryngol 1981;3:343-53.
50. Ensink R, Cremers C, Brunner H. Congenital conductive hearing loss in the lacrimo-auriculo-dento-digital syndrome. Arch Otolaryngol Head Neck Surg 1997;123:97-9.
51. Armstrong B. Stapes surgery in patients with osteogenesis imperfecta. Ann Otol Rhinol Laryngol 1984;6:634-5.
52. De Leenheer E, Oudesluijs G, Kuijpers-Jagtman A, Rappold G, Sengers R, Cremers C. Congenital conductive hearing loss in dyschondrosteosis. Ann Otol Rhinol Laryngol 2003; 112:153-8.
53. Teunissen B, Cremers C. Congenital stapes and incus ankyloses. Autosomal dominant inherited syndrome with congenital stapes ankylosis. Laryngoscope 1990;100:380-84.
54. Tange R. Ossicular reconstruction in cases of absent or inadequate incus, congenital malformation of the middle ear and epitympanic fixation of the incus and malleus. J Oto-Rhino-Laryngol Rel Spec 1996;58:143-6.
55. Lippy W, Schuring A. Solving ossicular problems in stapedectomy. Laryngoscope 1983;93:: 1147-50.
56. Shea J. Stapes fixation in chronic middle ear disease. Laryngoscope 1976;86:230-2.
57. Shea J. Malleus Teflon piston attachment prosthesis. Laryngoscope 1983;93:989-991.
58. Sheehy J. Stapedectomy: incus bypass procedures. A report of 203 operations. Laryngoscope 1982;92:258-62.
59. Feldman B, Schuknecht H. Experiences with revision stapedectomy procedures. Laryngoscope 1970:80:1281-91.
60. Schuknecht H, Bartley M. Malleus grip prosthesis. Ann Otol Rhinol Laryngol 1986;95:531-4.
61. Berenhoilz L, Lippy W. Total ossiculoplasty with footplate removal. Otolaryngol Head Neck Surg 2004;130:120-24.
62. Cremers C. Myringochorda-vestibulopexy: a new method for total replacement of the ossicular chain. Am J Otol 1994;15:445-48.
63. Fisch U, Acar G, Huber A. Malleostapedotomy in revision surgery for otosclerosis. Otol Neurotol 2001; 22:776-85.
64. Huttenbrink K. Biomechanics of stapesplasty: a review. Otol Neurotol 2003;24:548-59.
65. Hashimoto S, Yamamoto Y, Satoh H, Takahashi S. Surgical treatment of 52 cases of auditory ossicular malformations. Auris Nasus Larynx 2002;29:15-18.
66. Kohan D, Sorin A. Revision stapes surgery: the malleus to oval window wire-piston technique. Laryngoscope 2003;113:1520-4.
67. Dalchow C, Dunne A, Sesterhenn A, Teymoortash A, Werner J. Malleostapedotomy: the Marburg experience. Adv Oto-Rhino-Laryngol 2007;65:215-21.

3.3

Hearing results of stapedotomy and malleo-vestibulopexy in congenital hearing loss

V. E. Kisilevsky
N.A. Bailie
S.N. Dutt
J.J. Halik

International Journal of Pediatric Otorhinolaryngology, 2009 Sep 30. Epub ahead of print

Abstract

Aims

To analyze hearing results of surgical treatment of hearing loss associated with the congenital stapes ankylosis with or without malformations of ossicular chain.

Study design

Retrospective chart review.

Methods

The charts of 1369 stapedotomies performed by senior author (JH) from 1991 to 2006 were reviewed. In 40 cases operative findings were consistent with isolated congenital stapes fixation or associated with middle ear malformations. The modified stapedotomy technique was employed in 33 cases and malleo-vestibulopexy was used in 7 cases. Operative findings were standardized according to Cremers' classification. The outcomes of 40 surgeries were analyzed according to the 1995 AAO-HNS Committee on Hearing and Equilibrium guidelines. High frequency hearing results on 4, 8 and 12 kHz were reported in addition to standard frequencies. Results of stapedotomies and malleo-vestibulopexies were calculated separately. Surgical complications were described.

Results

The mean postoperative air conduction (AC) was 33 dB, bone conduction (BC) 22 dB and speech reception thresholds (SRT) 31 dB. Closure of the air-bone gap (ABG) to within 10 dB was achieved in 24/40 (60%) of cases. Lack of improvement was observed in 3/40 (8%) patients. In 26/32 (81%) of cases with potential for bilaterally serviceable hearing it was achieved. In 24/40 (60%) of cases symmetrical hearing with interaural difference of less than 10 dB was demonstrated.

Conclusion

Significant hearing gain in patients with congenital stapes ankylosis makes surgical treatment a valuable adjunct or an alternative to hearing aids in selected cases.

Introduction

Conductive hearing loss secondary to isolated congenital middle ear malformations is rare. The reported occurrence varies from 0.5 to 1.2%.[1,2] Its clinical and audiometric presentation frequently mimics more common middle ear pathology, such as otosclerosis and fixation or discontinuity of ossicular chain caused by ear infections. Therefore, preoperative recognition of congenital nature of conductive hearing loss can be difficult.[3]
Even if congenital cause is properly diagnosed, the decision to operate on the ear with ossicular malformation is controversial. Indications for surgical treatment of congenital conductive hearing loss are diverse and no uniform criteria are universally accepted. Concerns regarding increased risk of cerebrospinal fluid otorrhea or sensory-neural hearing loss were discussed by several authors.[4-6]
Multiple concomitant pathologies of the middle ear and malformed ear canal or otic capsule may negatively affect postoperative hearing. Reported results of surgery for congenital conductive hearing loss vary significantly from 15% to 76% of ABG closure to within 10 dB.[7-9] It has been a common practice to report the surgical outcomes as percent of ABG closure calculated for four–tone average of 0.5, 1, 2 and 3 kHz. Surgical trauma, however, is most likely to cause threshold elevation in the higher frequencies.[10] To the best of our knowledge, no reports have been published in the indexed otologic literature regarding the effects of surgery on high frequency hearing in congenital middle ear pathology.

The aim of this study was evaluation of outcomes of surgical treatment of hearing loss caused by congenital ossicular malformations with special focus on effects of class of malformation and of type of surgery on hearing results, post-operative high frequencies hearing and post-operative symmetrical hearing.

Patients and methods

The charts of 1369 consecutive stapedotomies performed by one surgeon (senior author JH) from 1991 to 2006 were reviewed. Of the 1369 surgical cases, 1145 primary stapedotomies were performed for otosclerosis, 89 were revision stapedotomies, 40 surgeries were performed for congenital stapes ankylosis and 8 for osteogenesis imperfecta. In 87 cases out of 1369 post-operative audiometric follow-up was not available. These were the charts of patients referred from distant locations, which have continued being seen by their referring physicians post-operatively and have not been returned for post-operative audiometry in our institution. In none of these cases congenital problems have been identified, therefore they have been excluded from the study due to incomplete data. Forty cases with operative findings consistent with isolated congenital stapes fixation or combined ossicular chain malformations were included. There were 21 female and 17 male patients; bilateral surgery was performed on 3 patients. In one case of bilateral stapedotomy post-operative results from one ear were missing and the results of this surgery were excluded from the analysis. The patients' age ranged from 6 to 71 years with the mean age 39 (S.D. 14.6).

Stapedotomy was performed in 33 cases and malleo-vestibulopexy technique was employed in 7 cases. The diagnosis of congenital stapes ankylosis was based on a history of non-progressive conductive hearing loss from early childhood and an intra-operative finding of hypoplastic or malformed stapes, incus and/or incudo-stapedial joint in the absence of an otosclerotic focus.

The severity of pre-operative hearing loss and type of ossicular abnormalities were evaluated. Intra-operative findings were tabulated and reported according to Cremers' classification.[11] A correlation between class of congenital anomaly and hearing results was examined. Intra- and post-operative complications were described.

The hearing outcomes were analyzed according to the 1995 American Academy of Otolaryngology-Head and Neck surgery (AAO-HNS) Committee on Hearing and Equilibrium guidelines.[12] Means of pre-operative pure tone average of air conduction (AC), bone conduction (BC) and air-bone gap (ABG) at 0.5, 1, 2 and 3 kHz were compared with corresponding post-operative values. As the beginning of this study period pre-dates the 1995 AAO-HNS guidelines, 3 kHz results were not available for all patients. In these cases 3 kHz AC and BC was calculated as average between 2 and 4 kHz, allowing a uniform presentation of audiometric data in compliance with AAO-HNS guidelines. Post-operative BC values were used to calculate postoperative ABG. Mean pre- and post-operative AC thresholds at 4, 8 and 12 kHz were studied separately. Pre- and post-operative speech reception thresholds (SRT) were also analyzed. Post-operative AC thresholds were compared with non-operated ear for the evaluation of symmetrical hearing.

The surgical intervention included endo-meatal elevation of the tympano-meatal flap, curettage of postero-superior bony overhang for adequate exposure of the incudo-stapedial joint and oval window area, inspection of ossicles and assessment of ossicular chain mobility. Upon the confirmation of stapes fixation the reverse order technique for small fenestra stapedotomy was employed. This was consisted of creating 0.5 mm fenestra in the footplate using graduated hand perforators. The prosthesis was then placed on the incus with the distal end articulated through the fenestration. The stapedial tendon than was cut, the incudo-stapedial joint was separated, posterior crus was cut and anterior crus with capitulum of stapes down fractured and removed. Since 2000 an argon laser (Lumenis Inc., Salt Lake City, Utah, USA) used in conjunction with a 0.2-mm OtoProbe™ fiber-optic delivery system (Iridex Corp, Mountain View, California, USA) has been employed in the procedure. The argon laser was used for vaporizing the stapedial tendon, posterior crus of the stapes and commencing the footplate fenestration. The fenestration was completed with the graduate perforators. The laser also proved useful in dealing with adhesions. In one patient with stapes fixation combined with malleal head fixation early in the series stapes mobilization with stapedotomy was attempted to divide the bone between the incudo-malleolar complex and the epitympanic bone. This was replaced by a malleo-vestibulopexy with excision of the malleus had from the level superior to the insertion of the tensor tympani. Tympanic membrane was elevated off the malleal neck, tensor tympani was divided and malleal neck was cut then with nipper. A modified Cawthorne (0.3 mm diameter) or modified Causse (0.4 mm diameter) prosthesis (Medtronic Xomed Jacksonville, FL, USA) was used in stapedotomy procedure.

Eighteen patients underwent laser-assisted stapedotomy and 15 were treated prior to the year 2000 without the laser. Twenty-five ears received the modified Cawthorne prosthesis and 8 ears the modified Causse prosthesis. All Causse prostheses recipients underwent a laser-assisted procedure. Among Cawthorne prostheses recipients, 15 ears were treated by conventional stapedotomy and 10 ears underwent laser-assisted surgery.

Statistical analysis was carried out for comparisons between pre- and post-operative values using paired *t*-test for the evaluation of air and bone conduction pure tone averages, SRT and ABG. The influence of type of surgery and piston were evaluated with ANOVA multiple regression test. The criterion selected for statistical significance was $p < 0.05$.

Results

Intra-operative findings were tabulated according the Cremers classification of congenital middle ear anomalies.[11] An isolated stapes fixation (Class 1) with superstructure deformity was found in 22 cases. Class 2 anomaly associated with incudo-malleal deformation was demonstrated in 15 cases. A malformed ossicular chain with mobile stapes footplate (Class 3) was found in two cases: one patient had discontinuity of incudo-stapedial joint due to hypoplastic incus and one patient had malleus epitympanic fixation. Class 4 anomaly was encountered in one case: the facial nerve canal was found completely dehiscent with the nerve resting against the stapedial crura (Table 1).

Table 1. Intra-operative findings according to Cremers' classification* of congenital middle ear anomalies.

Class	Anomaly	Number of cases	Sub classification
1	Stapes fixation	22	
2	Stapes fixation with associated incudo-malleal anomaly	15	
3	Anomaly of ossicular chain but mobile stapes footplate	2	Ossicular discontinuity *n*=1 Epitympanic fixation *n*=1
4	Congenital aplasia/dysplasia of the OW or RW	1	Abnormal Facial canal *n*=1

* Teunissen E, Cremers C. Classification of congenital middle ear anomalies. Report on 144 ears. Ann Otol Rhinol Laryngol 1993; 102:606-11.

Table 2. Summary of audiometric outcomes.

		Mean pre-operative values (dB)					Mean post-operative values (dB)				
		PTA		ABG	HF PTA	SRT	PTA		ABG	HF PTA	SRT
Category	n	AC	BC		AC		AC	BC		AC	
Primary stapedectomy	1145	54	24	28	59	51	29	25	7	53	27
All Congenital anomalies	40	53	23	30	61	52	33	22	11	55	31
Stapedotomy group	33	54	24	30	59	52	32	22	9	53	30
Malleo-vestibulopexy group	7	51	22	31	69	53	39	21	17	65	35

PTA - pure tone average (0.5, 1, 2, 3 kHz); AC - air conduction; BC - bone conduction; ABG - air-bone gap; HF - high frequencies (4, 8, 12 kHz); SRT - speech reception thresholds; dB - decibel; n - number of ears

AC, BC, ABG, SRT

The main pre-operative and post-operative audiometric parameters of the group of primary stapedotomy for otosclerosis, whole group with congenital malformations and separately calculated audiometric data for the group of stapedotomy for congenital malformations and malleo-vestibulopexy (MVP) are summarized in Table 2. The mean post-operative AC PTA in congenital group as whole was 33 dB (S.D. 17.1) compared with 53 dB (S.D. 16.9) pre-operatively (p<0.001). Mean post-operative SRT have been also improved significantly (p<0.001) from 52 dB (S.D. 14) pre-operatively to 31 dB (S.D. 15) post-operatively post-operative BC thresholds were 22 dB (S.D. 12), not significantly changed when compared with

pre-operative BC of 23 dB (S.D. 11.7). A strong correlation was found between AC PTA and SRT pre- (r 0.89) and post-operatively (r 0.92).
While no statistically significant difference was found in pre-operative hearing between the group of stapedotomy and MVP, technical results measured by post-operative ABG and AC gain were significantly better ($p<0.001$) after stapedotomy compared with MVP.

Percentage of ABG closure

In the congenital ankylosis group, the post-operative ABG closure to within 10 dB was achieved in 24/40 (60%) of cases compared with 82% in the 1145 cases of primary otosclerosis (Table 3). The post-operative ABG of 20 dB or less was found in 34/40 (85 %) of congenital cases and ABG > 20 dB in 6/40 (15%). The mean post-operative ABG was 11 dB compared with 30 dB pre-operatively.

The separate analysis of surgical success between stapedotomy group and MVP group, measured as a percent of ABG closure, demonstrated significantly higher success rate in the stapedotomy group (Table 4).

Table 3. Success rates in ABG closure in congenital stapes ankylosis group compared with the primary stapedotomy group.

ABG	Primary stapedotomy for otosclerosis group, *n* = 1145	Congenital stapes ankylosis group, *n* = 40	Significance
≤ 10 dB	82% (937)	60% (24/40)	$p < 0.05$
≤ 20 dB	97% (1115)	85% (34/40)	$p < 0.05$
>20 dB	3% (30)	15% (6/40)	$p < 0.001$

ABG - air-bone gap; n - number of ears; dB - decibel

Table 4. Success rates in ABG closure in stapedotomy group compared with the malleo-vestibulopexy group.

ABG	Stapedotomy group, *n* = 33	Malleo-vestibulopexy group, *n* = 48	Significance
≤ 10 dB	20/33 (60%)	4/7 (57%)	$p < 0.05$
10dB<ABG≤ 20 dB	9/33 (27%)	1/7 (14%)	$p < 0.001$
>20 dB	4/33 (12%)	2/7 (28%)	$p < 0.001$

ABG - air-bone gap; n - number of ears; dB - decibel

High frequencies hearing results

The mean pre-operative high frequencies average (HF PTA) at 4, 8 and 12 kHz in whole congenital group was 61 dB. The mean post-operative HF PTA was 55 dB.

While the mean post-operative gain at high frequencies was 6 dB, it decreased toward the higher frequencies from 13 dB at 4 kHz to 3 dB at 12 kHz (Table 5).
The separate analysis of high frequencies hearing results in stapedotomy group and MVP group demonstrated significantly higher gain at 4 kHz in the stapedotomy group with no statistically significant difference in HF PTA gain (Table 6).

Table 5. High frequencies hearing results in congenital group as whole.

	4 kHz	8 kHz	12 kHz	HF PTA
Pre-operative (dB)	56	60	69	61
Post-operative (dB)	43	55	72	55
Gain (dB)	13	5	3	6

HF PTA - high frequencies pure tone average at 4, 8 and 12 kHz; dB - decibel

Table 6. High frequencies hearing results in stapedotomy group and malleo-vestibulo-pexy group.

	4 kHz	8 kHz	12 kHz	HF PTA
Stapedotomy group				
Pre-operative (dB)	56	58	61	59
Post-operative(dB)	42	53	66	53
Gain (dB)	14	5	-5	6
Malleo-vestibulopexy group				
Pre-operative (dB)	51	72	89	69
Post-operative (dB)	43	67	88	65
Gain (dB)	8	5	1	4

HF PTA - high frequencies pure tone average at 4, 8 and 12 kHz; dB - decibel

Laser versus non-laser stapedotomy

Post-operative hearing results were compared between laser-assisted and non-laser stapedotomies. The mean post-operative AC threshold in laser stapedotomy was 33 dB compared with 36 dB without the use of the laser. The mean post-operative BC was 21 dB in laser stapedotomy compared with 26 dB in the non-laser group (p 0.04). Post-operative ABG was 9 dB and 12dB respectively in laser versus non-laser group (not significant). Post-operative SRT were 31 dB in laser and 32 dB in non-laser group (not significant). HF PTA at 4, 8 and 12 kHz in laser stapedotomy was 54 dB compared with 60 dB in case of mechanical fenestration (p 0.044). Therefore, the advantage of laser assisted technique seems to be the better preservation of BC thresholds and better post-operative high frequencies hearing.

Hearing results by class of malformation

The pre-operative and post-operative audiometric data separately calculated for each class of malformation are summarized in Table 7. While the absolute values of mean post-operative AC, BC and SRT were better in cases with Class 3 malformation, the difference in pre-operative hearing status between the groups resulted in higher AC, SRT and ABG gains in the case with Class 4 malformation. A small number of cases with Class 3 and Class 4 malformations, however, hinder a meaningful comparison of surgical results with Class 1 and 2 malformations. No statistically significant difference in hearing outcomes was found between the groups with Class 1 and Class 2 malformations.

Table 7. Hearing results by class of malformation.

		Mean pre-operative values (dB)					Mean post-operative values (dB)					
	N	PTA AC	HF 4,8,12 kHz	BC	ABG	SRT	PTA AC	AC gain	HF 4,8,12 kHz	BC	ABG	SRT
Total	40	53	61	23	30	54	33	20	55	22	11	31
Class 1	22	53	57	23	30	52	35	18	52	23	11	32
Class 2	15	52	64	22	30	58	34	18	60	21	13	34
Class 3	2	46	68	18	29	48	16	30	53	13	3	18
Class 4	1	85	95	50	35	75	48	37	75	44	4	35

PTA AC - pure tone average on 0.5,1,2 and 3 kHz air conduction; HF - high frequencies average on 4, 8 and 12 kHz; BC - bone conduction; ABG - air-bone gap; SRT - speech reception thresholds.

Post-operative serviceable hearing

Thirty-two out of 40 32/40 (80%) patients had pre-operative BC PTA ≤ 30 dB. In case of complete post-operative closure of air-bone gap this parameter provided potential to achieve a serviceable hearing without amplification, defined as AC PTA≤30dB.[13] Twenty-six out of these 32 (81 %) patients actually achieved this hearing level.

Post-operative symmetrical hearing

Evaluation of post-operative binaural hearing demonstrated that symmetrical hearing with interaural difference of less than 10 dB was achieved in 24/40 (60%) patients and less than 20 dB in 34/40 (85%) patients. In 21/40 (53%) of cases an operated ear became a better hearing ear.

Surgical failures

While the results of a whole group demonstrated significant improvement of hearing post-operatively, surgical failure was observed in three patients. In one case, obliterative otosclerosis and congenitally malformed stapes superstructure was encountered in 41-year-old female patient. Post-operative hearing test demonstrated no significant change in air and bone conduction thresholds and CT scan showed an ossified vestibule. In the second case, a remnant of persistent stapedial artery was present in 26-year-old male patient. A failed attempt to remove a rigidly fixed malformed superstructure led to elevated post-operative bone conduction thresholds. In the third case, a 51-year-old male patient presented with a class 2 malformation. His stapes footplate was rigidly fixed, the malleus handle was foreshortened and long process of incus was thread-like in calibre. A malleo-vestibulopexy was performed on awaked sedated patient under local anaesthesia and upon the placement of the 5 x 0.3 mm Cawthorne prosthesis patient noted immediate improvement of hearing. Post-operative audiometric follow-up, however, demonstrated increased air and bone conduction thresholds compared with pre-operative audiograms.

Discussion

Isolated middle ear malformations as a cause of congenital hearing loss are rare. The reported occurrence of middle ear anomalies varies from 0.5 to 1.2 %.[1,2] In our series of 1369 stapedectomies, 40 cases with congenital stapes ankylosis represent 3% of all patients. This occurrence is similar to the 4% of occurrence rate reported by other authors.[14]

The pre-operative diagnosis of congenital conductive hearing loss is a difficult diagnostic task. Bilateral hearing loss and associated dysmorphic syndromes are usually extensively investigated due to a high suspicion index for congenital anomalies. In case of unilateral isolated conductive hearing loss, however, other more common middle ear pathologies are frequently assumed as the etiologic factor. A serous otitis media or recurrent acute otitis superimposed on underlying congenital middle ear malformation often contributes to misdiagnosis. Moreover, attempts of surgical management by ventilation tube placement without middle ear exploration may further delay the diagnosis. The dynamics of hearing loss (non-progressive versus progressive), history of ear infections or family history of conductive hearing loss should be considered in differential diagnosis of congenital stapes ankylosis, tympanosclerotic ossicular fixation or juvenile otosclerosis.[1,3,6-8]

The perplexing results of pre-operative assessment, especially significant conductive hearing loss with normal stapedial reflex should raise a suspicion for possible third window lesions, like X-linked gusher syndrome (also called DFN-3), large vestibular aqueduct syndrome, branchio-oto-renal syndrome.[15-17] Pre-operative high-resolution CT scan is required to rule out these potentially detrimental to hearing conditions. Temporal bone imaging may alert the surgeon, if it reveals dilated lateral portion of the internal auditory canal or abnormal communication between inner ear and intracranial space.[18]
The ability to predict the chances for post-operative hearing improvement and the risks of surgical complications is crucial for a decision to opt for ear surgery. Welling et al.[9] demonstrated in series of 66 pediatric stapedectomies, that surgical outcomes correlated significantly with diagnostic category.
In his series the best hearing results were achieved in the otosclerosis group (mean post-operative ABG 13 dB), closely followed by the group with isolated congenital stapes ankylosis (ABG 16 dB), while tympanosclerotic footplate fixation was associated with post-operative ABG of 25 dB and stapes ankylosis combined with other pathology with ABG of 32 dB.[9] De La Cruz[19] also found slightly poorer results in cases of congenital stapes fixation, when compared with otosclerosis. He attributed lower success rate in congenital group to higher incidence of coexisting middle ear anomalies.
The interpretation of intra-operative findings and their subsequent classification is often hindered by the diversity of descriptions found in surgical records.[6-9] The classification system introduced by Cremers[11] allows reliable inter-class comparisons of the surgical results. Of interest, the distribution of different classes of middle ear malformations in our series differs from the original group of 144 ears described by Cremers. A relatively smaller proportion of Class 2 anomalies in our series may be caused by the retrospective nature of our review of surgical reports. It is possible that some ears were erroneously classified as Class 1 malformations if associated incus anomaly was not emphasized in the medical chart. The limitations of a retrospective nature of this study may explain the fact that no syndromal etiolgy was detected in reported cases.

The results in our series with mean post-operative ABG 11 dB in Class 1 and 13 dB in Class 2 malformations (Table 7) demonstrate that good technical results can be achieved either in cases of isolated or combined congenital stapes fixation employing small fenestra stapedectomy or MVP techniques. The distribution of hearing outcomes according to the class of malformation, however, showed that the better post-operative hearing levels and higher gains in post-operative AC, SRT and ABG were achieved in Class 3 anomalies. Our results are approaching those of Cremers[20] who reported a mean gain of 31 dB after ossiculoplasty in a group with discontinuity of the ossicular chain. Funsaka[21] also achieved the best surgical results in a group of congenital incudo-stapedial joint separation compared with stapes ankylosis or ossicular chain fixation. A sensory-neural component of hearing loss (pre-operative BC > 20dB), which was found in 24/37 (65%) of Class 1 and 2 malformations, is a possible factor limiting the achievement of normal hearing in these cases. Since BC levels in Classes 1 and 2 remained unchanged post-operatively, they reflect a true sensory-neural component and not an artefact in BC measurement.

Class 4 malformation was identified in only one case in our series. Although a facial nerve canal was found completely dehiscent with a nerve resting against the stapedial crura, the performance of stapedotomy was found possible and resulted in significant hearing gain without surgical complications. Although in this case results were encouraging, Class 4 anomaly is considered by many as a class where no surgery should be performed. The decision to proceed with intervention depends on specific findings of complete oval window aplasia versus abnormal facial canal or persistent stapedial artery.

Cremers classification based on the largest number of ears with congenital malformations and refers to different surgical techniques available for middle ear reconstruction.[11] It allows reliable comparison between various surgical methods. We used the degree of ABG closure as a traditional measure of technical success of surgery. This enabled comparison between the results stapedotomy and MVP employed in our series (Table 4). Although technical results of stapedotomy seem to be superior to MVP, it should be mentioned that MVP was used in class 2 and 3 malformations only, as opposed to stapedotomy mostly used in isolated stapedial ankylosis. Therefore, the extent of ossicular pathology in MVP cases was negatively affecting hearing results. Historically, stapedotomy gained a reputation of the most successful technique for closure of ABG. According to the recent publications, it allows ABG closure to within 10 dB in 82-94% of patients with otosclerosis.[22,23] The reported success rate of ossicular chain reconstruction is usually lower than that of stapedectomy and varies widely between series (ABG closure to within 10 dB in 44-67%).[24,25] Ossiculoplasty for combined ossicular anomalies yields an ABG of less than 30 dB in about 70% of patients.[26] These variations are explained by large spectrum of ossicular chain pathology and subsequent different types of surgery and prostheses employed in middle ear reconstruction.

In case of congenital middle ear defects, diverse combinations of ossicular anomalies lead to even more variations in treatment outcomes. Some publications reported hearing results equal or approaching outcomes of stapedectomy for otosclerosis.[8,27] In these reports, however, pre-operative BC thresholds were used for calculation of post-operative ABG, thus underestimating real values of ABG in the case of post-operative improvement of bone conduction. We found that the percentage of patients achieving postoperative ABG to within 10 dB in our study is equal or superior to other congenital series where ABG was calculated using post-operative BC and AC levels (Table 8).

Table 8. The comparative success rate measured as a percent of ABG closure to within 10 dB.

Reference	Author	Year	N	ABG≤10dB
16	Cremers	1993	44	49%
15	De La Cruz	1999	50	59%
9	Welling	2002	21	24%
28	Massey	2006	25	48%
	Present study	2009	40	60%
8	House*	1980	13	76%
27	Lippy*	1998	47	91%

ABG - air-bone gap; dB - decibel; N - number of cases in series
* series that used pre-operative BC for calculation of post-operative ABG

The comparison of the surgical results in our group of congenital middle ear anomalies with those of stapedotomy for otosclerosis (Table 3) demonstrated superior outcomes in the later group. These finding are in agreement with the results reported by Massey et al.[28], who found that hearing outcomes of 25 stapedectomies performed for congenital stapes ankylosis were significantly worse when compared with the results of stapedectomy for otosclerosis. The mean post-operative ABG in this group was 12 dB and ABG closure to within 10 dB was achieved in 12 (48%) of cases. Bachor et al[29] investigated the correlation between surgical outcomes and pathological findings in a group of 12 children with stapes fixation. According to his findings, the difference in surgical outcome was related to the degree of footplate abnormality. Best hearing results were achieved in patients with juvenile otosclerosis when compared with combined ossicular malformations. The hearing results in our series are also similar to those reported by Albert et al.[30], who achieved mean post-operative AC thresholds of 15 dB and ABG of 10 dB in series of 28 stapedectomies and stapedotomies for congenital stapes ankylosis.

While post-operative AC PTA and the degree of ABG closure are important measures of technical success of surgery, the improved ability to understand conversation speech is the most important factor in patient's perception of surgical benefit.[13,31] The importance of high frequencies hearing for speech recognition and understanding speech in noise is well recognized.[32,33] The post-operative changes in high frequencies thresholds, along with BC levels, provide a sensitive measure of iatrogenic cochlear damage.[34,35] Historically, a cautious approach to surgical intervention in congenital stapes anomalies was based on high rates of reported surgical failures and increased risk of post-operative SNHL.[10,36,37] To the best of our knowledge, no perioperative hearing thresholds at 8 and 12 kHz or 4,8 and 12 kHz HF PTA have been previously reported in the results of surgery for congenital stapes ankylosis. The evaluation of pre-operative hearing status at higher frequencies demonstrated that the common pattern of audiometric configuration was a down sloping curve starting at 4-6 kHz with the mean PTA at 0.5,1,2 and 3 kHz 53 dB and mean HF PTA at 4,8,12 kHz 61 dB. Post-operative gain of HF PTA was 6 dB, although frequency specific analysis reveals decreased gain toward higher frequencies (Table 5, 6). Bearing in mind that despite of pre-existing sensory-neural component of HL in 25/40 (67%) of cases pre-operatively, BC levels on speech frequencies were preserved post-operatively; we feel that surgical treatment is a safe alternative to hearing aids in selected cases. Hearing gain achieved at high frequencies along with the closure of large ABG suggest also that even in cases requiring post-operative amplification surgical treatment offers a valuable adjunct to hearing aids.

The evaluation of postoperative bilateral hearing revealed that symmetrical hearing with interaural difference ≤10 dB was achieved in 60% of cases. In our opinion, these findings, along with the low complication rate, are encouraging to consider a second ear surgery in cases with bilateral asymmetric hearing impairment.

Conclusions

The outcomes of surgery for congenital stapes fixation are highly rewarding for patients, even though they are not as successful as the surgical results in otosclerosis. Isolated stapes fixation and ossicular discontinuity are associated with better surgical outcomes when compared with complex ossicular pathology. Preservation of cochlear reserve and improvement of high frequencies hearing is achievable in selected cases. It is possible to fulfil a potential for serviceable hearing in 81% of cases in this series. Significant hearing improvement in properly selected patients makes surgical treatment a valuable adjunct or alternative to hearing aids.

References

1. Raz Y, Lusting L. Surgical management of conductive hearing loss in children. Otolaryngol Clin N Am 2002;35: 853-75.
2. Stewart JM, Downs MP. Congenital conductive hearing loss: the need for early identification and intervention. Pediatrics 1993; 91:355-359.
3. Briggs R, Luxford W. Corrections of conductive hearing loss in children. Otolaryngol Clin N Am 1994 ;27:607-20.
4. Shea JJ. Complications of the stapedectomy operation. Ann Otol Rhinol Laryngol 1963; 72:1109.
5. Grant WE, Grant WJ. Stapedius tendon ossification: a rare cause of congenital conductive hearing loss. J Laryngol Otol 1991; 105:763.
6. De La Cruz A, Doyle K. Ossiculoplasty in congenital hearing loss. Otolaryngol Clin N Am 1994;27: 803.
7. Raveh E, Hu W, Papsin B, Forte V. Congenital conductive hearing loss. J Laryngol Otol 2002; 116:92-6
8. House J, Sheehy J, Antunez J. Stapedectomy in children. Laryngoscope 1980; 90:1804-9
9. Welling D, Merrell J, Merz M, Dodson E. Predictive factors in pediatric stapedectomy. Laryngoscope 2003; 113:1515-19.
10. Palva T. High-tone sensorineural hearing losses following chronic ear surgery. Arch Otolaryngol 1973; 98:176-8.
11. Teunissen E, Cremers C. Classification of congenital middle ear anomalies. Report on 144 ears. Ann Otol Rhinol Laryngol 1993; 102:606-11.
12. Committee on Hearing and Equilibrium. Guidelines for the evaluation of results of treatment of conductive hearing loss. Otolaryngol Head Neck Surg 1995; 113:186-7.
13. Browning GG, Gatehouse S, Swan IRC. The Glasgow Benefit Plot: A new method for reporting benefits from middle ear surgery. Laryngoscope 1991;101:180--185
14. Kuhn J, Lassen L. Management of congenital incudo stapedial anomalies in stapes surgery. Am J Neuroradiol 2006;27:694-9.
15. Cremers CW, Hombergen G, Scaf J, Huygen P, Volkers W, Pinckers A. X-linked progressive mixed deafness with perilymphatic gusher during stapes surgery. Arch Otolaryngol 1985; 111: 249-254.
16. Cremers CW. Audiologic features of the X-linked progressive mixed deafness syndrome with perilymphatic gusher during stapes surgery. Am J Otol 1985; 6: 243-246.
17. Merchant S, Rosowki J. Conductive hearing loss caused by third-window lesions of the inner ear. Otol Neurotol 2008; 29: 282-289.
18. Ceruti S, Stinckens C, Cremers CW, Casselman J. Temporal bone anomalies in the branchio-oto-renal syndrome: detailed computed tomographic and magnetic resonance imaging findings. Otol Neurotol 2002; 23: 200-207.
19. De La Cruz A, Angeli S, Slattery W. Stapedectomy in children. Otolaryngol Head Neck Surg 1999; 120(4):487-92.
20. Teunissen E, Cremers C. Surgery for congenital anomalies of the middle ear with mobile stapes. Eur Arch Otorhinolaryngol.1993; 250(6):327-31.
21. Funasaka S. Congenital ossicular anomalies without malformations of the external ear. Arch Otorhinolaryngol. 1979; 224(3-4):231-40.
22. Vincent R, Sperling N, Oates J, Jindal M. Surgical findings and long-term hearing results in 3050 stapedotomies for primary otosclerosis: a prospective study with the otology-neurotology database. Otol Neurotol (2006) 27:S25-S47.

23. Kisilevsky V, Dutt S. Bailie N, Halik J. Hearing results of 1145 stapedotomies evaluated with Amsterdam Hearing Evaluation Plots. J Laryngol Otol 2009;7:730-6.
24. Wehrs RE. Incus interposition and ossiculoplasty with hydroxyapatite prostheses. Otolaryngol Clin N Am 1994; 27(4):677-66.
25. Goldenberg RA. Ossiculoplasty with composite prostheses. Otolaryngol Clin N Am 1994; 27(4):727-45.
26. Kartush J. Ossicular chain reconstruction. Otolaryngol Clin N Am 1994; 27(4):689-707.
27. Lippy W, Burkey J, Shuring A, Rizer F. Short- and long-term results of stapedectomy in children. Laryngoscope 1998; 108(4):569-72.
28. Massey B, Hillman T, Shelton C. Stapedectomy in congenital stapes fixation: Are hearing outcomes poorer? Otolaryngol Head & Neck Surg (2006)134, 816-18.
29. Bachor. E, Just T, Wright C, Pau H, Karmody C. Fixation of stapes footplate in children: A clinical and temporal bone histopatologic study. Otol Neurotol 2005; 26:866-73.
30. Albert S, Roger G, Rouillon I, Chauvin P, Denoyelle F, Derbez R, Delattre J, Triglia J, Garabedian E. Congenital stapes ankylosis: study of 28 cases and surgical results. Laryngoscope 2006; 116:1153-57.
31. Smyth G.D., Patterson C.C. Results of middle ear reconstruction: Do patients and surgeons agree? Am J Otol 1985; 6 (3): 276-9.
32. Meyer SE. The effect of stapes surgery on high frequency hearing in patients with otosclerosis. Am J Otol 1999;20:36-40.
33. Martin MB, Rubinstein EN, Hirsch BE. High-frequency sensorineural hearing loss after stapedectomy. Otol Neurotol 2008;29:447-52.
34. Ballantyne J. Iatrogenic deafness. J Laryngol Otol 1970;84:967-1000
35. Raut V, Halik J. Argon laser assisted small fenestra stapedectomy for otosclerosis. Auris Nas Larynx 2005; 32 (1):11-15.
36. Von Haacke N. Juvenile stapedectomy. Clin Otolaryngol 1985;10:9-13.
37. Pappas DJ, Pappas DS, Hedlin G. Round window atresia with congenital stapes fixation. Laryngoscope 1998; 108(8): 1115-18.

Chapter 4

Surgical technique

4.1

Modified reverse order laser-assisted stapedotomy

V.E. Kisilevsky
N.A. Bailie
J.J. Halik

Laryngoscope, 2009, accepted for publication

Introduction

The evolution of modern stapes surgery, from stapedectomy through stapedotomy to step reordering, and more recently to the adjunctive use of laser has been driven by a desire to maximize technical success and minimize complications. The most serious complications of stapes surgery are sensorineural hearing loss (SNHL) or “dead ear”, most commonly from excessive footplate manipulation[1], and incus dislocation ,which may occure during prosthesis placement [2]. Prosthesis malfunction has been reported as the most common cause of delayed failure of primary stapedectomy.[3,4] Prosthesis-related failures can be corrected by a revision procedure; however, damage to the inner ear or the malleo-incudal joint is often irreversible. This article presents a surgical method that aims to minimize operative trauma. The surgical technique combines a modification of the reverse order technique [5,6], with the adjunctive use of argon beam laser and modified self-crimping prosthesis. In our experience, this technique provides better access to the footplate during fenestration than the reverse-order technique,while maintaining the advantage of better stability of the footplate and incus. Modification of the prosthesis facilitates placement without compromising hearing results.[7] Laser-assisted fenestration minimizes cochlear damage enabling preservation of high frequency hearing.[7,8]

Surgical method

I. *Middle Ear Exposure and Confirmation of a Diagnosis.*

Middle ear exposure is gained through the standard endo-meatal approach. The ossicular chain is inspected and gently manipulated to ensure the diagnosis of stapes fixation is correct, and to rule out malleus fixation. (Figure 1).

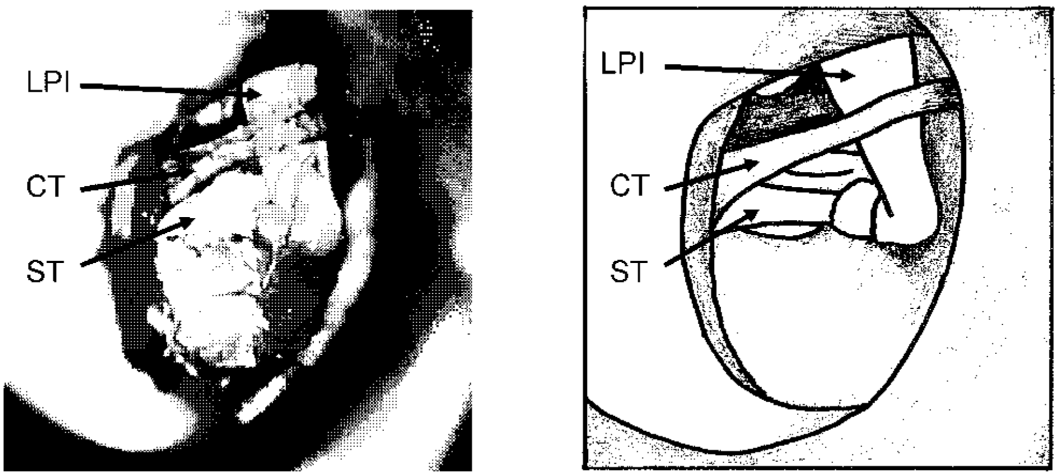


Figure 1. LPI: long process of incus; CT: chorda tympani; ST: stapedial tendon.

II. Approach to footplate: partial reverse order technique

An argon laser (Lumenis Inc, Salt Lake City, Utah, USA) is used in conjunction with a 0.2 mm OtoProbe™ fiber-optic delivery system (Iridex Corp, Mountain View, California, USA). Beginning with an initial power setting of 2000 mW and pulse duration of 0.2 seconds the stapedius tendon and the posterior crus are vaporized. Removal of the only the posterior crus of the stapes enable the same wide access to the footplate as by the conventional stapedotomy procedure whilst retaining the anterior crus provides stability to the footplate and incus, reducing the possibility of subluxation.

III. Creation of a fenestra: combination of Argon laser with manual perforators

The laser power is adjusted to1500 mW and the pulse duration reduced to 0.1 seconds. A laser pulse is then delivered to the postero-superior quadrant of the stapes footplate and repeated until perilymph just begins to 'sweat' through the laser burn (Figure 2). This provides a set-hole for a series of hand perforators. The stapedotomy is gently widened, beginning with a 0.3 mm perforator and increasing in 0.1 mm steps to a 0.6 mm perforator, providing a final stapedotomy diameter of 0.5 mm (Figures 3,4).

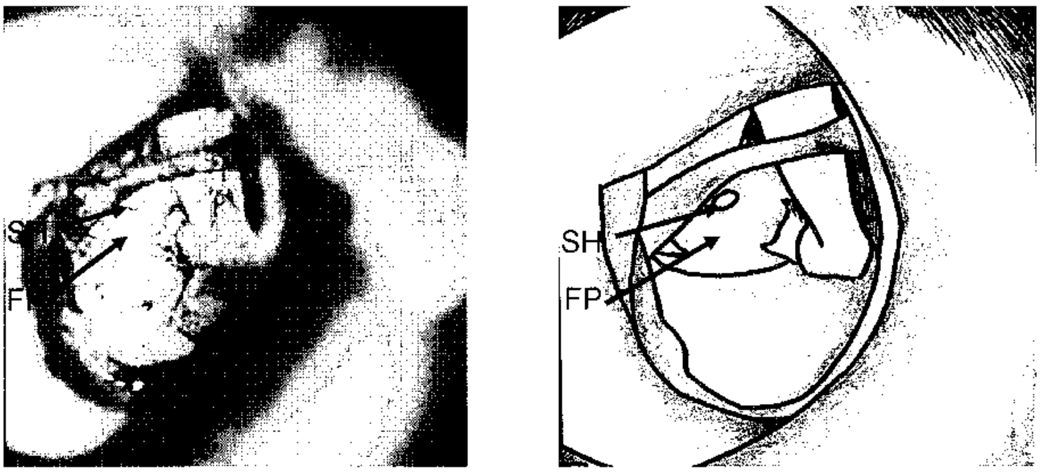


Figure 2. SH: set-hole, FP: footplate.

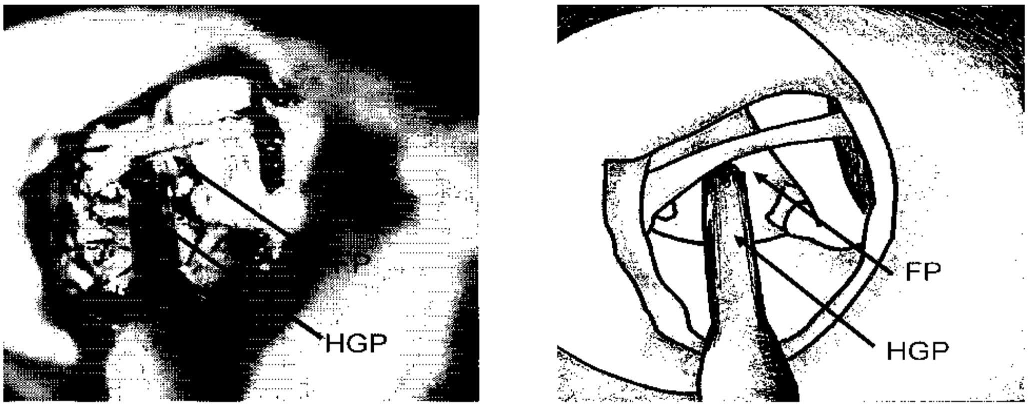


Figure 3. FP: footplate; HGP: Halik graduated perforator.

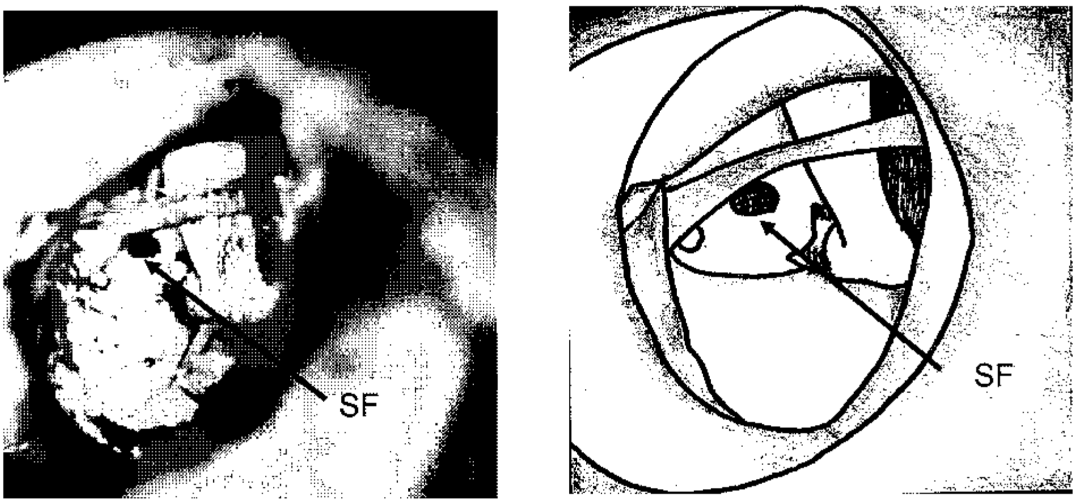


Figure 4. SF: small fenestra.

IV. Prosthesis modification and placement.

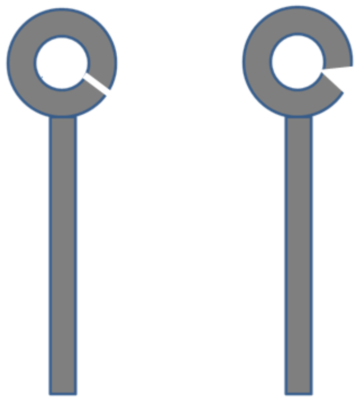
A modified Causse Teflon (0.4 mm diameter) prosthesis (Medtronic Xomed Jacksonville, FL, USA) is employed in this technique. The prosthesis is modified by creating a notch at 3 o'clock that allows an easy click-on attachment to the long process of incus, which provides a tight hold without crimping (Figure 5). This secure grip cannot always be achieved by the "memory" of the Teflon after the stretching of the prosthesis ring. The prosthesis is cut to an appropriate length, and is aligned with small middle ear alligator forceps while maintaining direct visualization of its distal and proximal ends. The prosthesis ring is maintained at a 90° angle to the long process of the incus to eliminate any distortion of the ring during application. The prosthesis is placed so that it simultaneously enters the footplate fenestra and engages the long process of incus (Figure 6). This reduces the risk of incus luxation and provides additional stabilization against the displacement of the footplate during the down fracture of the anterior crus.

V. *Superstructure removal.*

The incudo-stapedial joint is separated using a small right angle hook, anterior crus is down fractured toward the promontory. The superstructure is then gently removed, avoiding contact with the incus and prosthesis.

VI. *Closure*

Confirmation of restored mobility and continuity of the ossicular chain is achieved by gentle incus palpation, and any loose prosthesis attachment can be corrected by an adjustment of the ring position on the long process of incus. A secure position of the shaft within the fenestra is tested by gentle lateral pressure on the prosthesis with a small hook.



A B

Figure 5. A: original and B: modified Causse prosthesis.

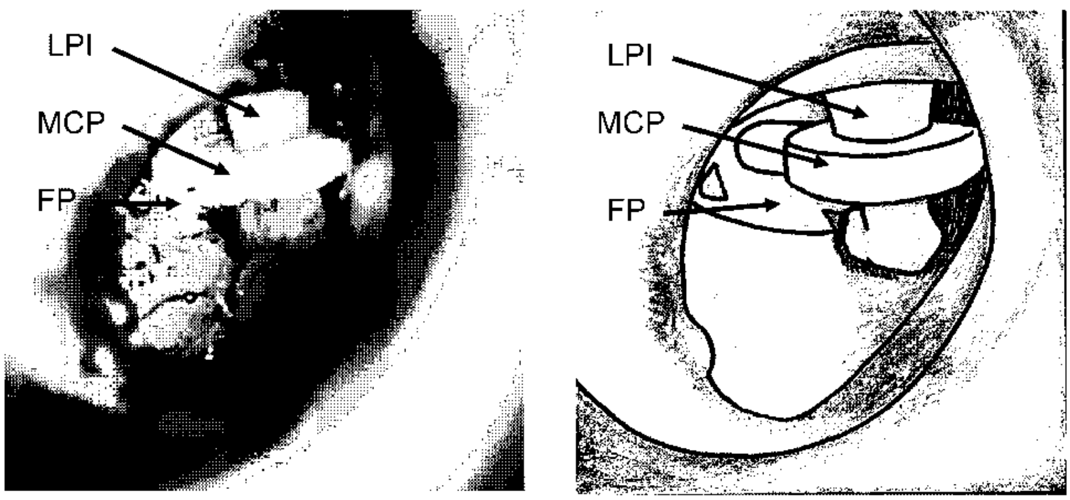


Figure 6. LPI: long process of incus; MCP: modified Causse prosthesis; FP: footplate.

Slight bending of the shaft ensures secure purchase within the fenestra. A precise fit of the fenestra to the diameter of the prosthesis reduces material used for the prevention of perilymph leak, and the natural blood clot around the shaft provides an adequate seal preventing postoperative fistula. The tympano-meatal flap is returned to its original position. The external auditory canal is lined with a strip of surgical rayon dressing and packed with a gauze ribbon soaked in Cortisporin ointment (Neomycin sulphate 3.5 mg, Polymyxin B sulphate 10.000U, Hydrocortisone acetate 5mg; Alcon Ltd, Hunenberg, Switzerland)) External ear canal packing is removed on the next postoperative day.

Discussion

Reviews of intra-operative complications and findings at revision surgery have driven the evolution from the original stapedectomy technique.[3,4,9-13] Footplate manipulation and prosthesis placement have been shown to occasion the most complications of surgical procedure.[2,3,14] Analysis of experience-based differences in surgical performance has led to a reduction in the surgeons' reliance on hand–held instruments during precise manipulations.[15-17] The employment of both laser assisted fenestration[18-21] and the development of self-crimping prostheses have reduced surgical trauma and technical difficulties of both manual fenestration and prosthesis crimping, respectively.[22-24] Recently, the advantage of reversed order technique in reducing operative complications rate has been demonstrated.[25,26]

The procedure described in this article combines the strengths of a number of previously described techniques.[5,6,19,24] and has emerged from practice-based learning of the senior author (JH).

This technique involves a modification of the reverse steps order technique, in which the prosthesis is placed before removal of stapes crura and incudo-stapedial joint disarticulation. By removing the posterior crus and stapedius tendon, access to the footplate is similar to that obtained during conventional stapedotomy; yet the retained anterior crus provides the same stability to the footplate and incus as provided by the reverse-order technique. In our experience, keeping the incudo-stapedial joint and anterior crus intact appears to provide additional protection against inadvertent footplate fragmentation or incus mobilization.

Initiating the fenestra with hand perforators or a microdrill can result excessive pressure in the vertical plane, which may carry the risk of inadvertent footplate mobilization or fracture. Lasers offer an advantage in this step, as the fenestration can be created without pressure on the footplate .Our preference for the Argon laser that the laser energy is delivered via a hand-held probe that has a similar feel to other middle ear instruments such as the Rosen's needle. Many surgeons perform laser-assisted stapedotomy with laser alone to fashion the stapedotomy; however, there have been concerns that this risks heating the perilymph and creating injury to structures of the membranous labyrinth.[27-30] The use of laser to simply attenuate the footplate so perilymph just begins to seep through obviates this risk to the labyrinth, while providing a set-hole for the perforators. The use of graduated perforators prevents both overheating of the vestibule and overexposure of the inner ear to laser heat or vibration from a micro drill. Another advantage is preventing contamination of the vestibule with char or bone dust created by the laser beam applied to the "rosette" area, or footplate drilling. This less traumatic opening of a vestibule, without the need for fragments removal, has been shown to reduce the risk of inner ear trauma and subsequent SNHL.[7,8]

The modification of the Causse Teflon prosthesis described in this article has provided reliable results and a low rate of piston-related failures. Normally, the ring of the piston would be stretched to allow it to be placed over the long process of the incus, which relies on elastic recoil of the material to secure the attachment. This technique may be problematic if elastic recoil has drawn the ring closed before it can be placed upon the stapes, or if the ring is over-stretched and it remains loose or falls off. Cutting a small wedge from the ring, such that the entire inner circumference of the ring is maintained, provides means for the ring to key into the long process of the incus without pre-stretching, while still maintaining a circumferential hold on the long process of the incus. The length of the piston shaft has been another source of controversy. Some authors assert that measurements should be made whilst other use a standard length, typically 4.5 mm. Our own use of a 5 mm prosthesis length was borne of advice given to the senior author from the late Gordon Smyth, who had switched to the use of a 5 mm prosthesis later in his career feeling that there may be a degree of late lateralization of the incus long process subsequent to division of the stapedius tendon (Gordon Smyth, personal communications). In our experience, this has not been associated with increased postoperative vertigo.[7,8] Traditionally, the results of stapes surgery were reported in relation to hearing thresholds at 0.5, 1 and 2 kHz and, more recently, this has been extended to include 3 KHz. It is well recognized, though perhaps not well publicized, that hearing at higher frequencies (>4 KHz) may remain poor after stapes surgery. Anatomically, the basilar region of the cochlea, which provides high frequency hearing, is the closest to the operative site during stapedotomy and it may be more susceptible to surgical trauma.[1,31-33] Thus, high frequency sensorineural hearing loss following stapedotomy may provide a measure of operative trauma. Evaluation of hearing results using this technique has demonstrated good preservation of high frequency hearing preservation and a low complications rate.[7,8] Technical success measured by air-bone gap closure is equal or superior to other recently published series.[6] The technique described in this article has been also proven useful in training of stapedotomy surgery, allowing the operation to be attempted in a series of controlled and increasingly difficult steps.

Acknowledgment

The authors wish to thank John Rutka MD, FRCS(C), and Ms. Maxine Armstrong, MSC, for their contributions in preparation of this article.

References

1. Smyth GDL. Immediate and delayed alterations in cochlear function following stapedectomy. Otolaryngol Clin N Am 1978;11:105-12.
2. Mathews SB, Hetzler DG, Hilsinger RL. Incus and stapes footplate stimulator. Laryngoscope. 1997; 107:1614-16.
3. Sheehy JL, House HP. Causes of failure in stapes surgery. Laryngoscope 1962; 72:10-31
4. Lippy WH, Battista RA, Berenholz L, Schuring AG, Burkey JM. Twenty-year review of revision stapedectomy. Otol Neurotol 2003; 24:560-66.
5. Smyth G. Practical suggestions on stapedotomy. Laryngoscope. 1982;92:952-953.
6. Fish U. Stapedotomy versus stapedectomy. Am J Otol 1982;4:112-17.
7. Kisilevsky V, Dutt S, Bailie N, Halik J. Hearing results of 1145 stapedotomies evaluated with Amsterdam Hearing Evaluation Plots. J Laryngol Otol 2009;26:1-7.
8. Raut V, Halik J. Argon laser assisted small fenestra stapedotomy for otosclerosis. Auris Nasus Larynx 2005; 32:11-15.
9. House JP. The evolution of otosclerosis surgery. Otolaryngol Clin N Am 1993; 26:323-35.
10. Fish U, Acar GO, Huber AM. Malleostapedotomy in revision surgery for otosclerosis. . Otol Neurotol 2001; 22:776-85.
11. Shea JJ, Shea PF, McKenna MJ. Stapedectomy for otosclerosis. In: Glasscock M.E, Gulya AJ, eds. *Glasscock-Shambaugh Surgery of the Ear.* 5th ed.Hamilton, Ontario: BC Decker Inc; 2003, pp 517-31.
12. Rizer FM, Lippy. Evolution of techniques of stapedectomy from the total stapedectomy to the small fenestra stapedectomy. Otolaryngol Clin N Am 1993; 26:443-51.
13. Cremers CWR, Beusen JMH, Huygen PLM. Hearing gain after stapedotomy, partial platinectomy, or total stapedectomy for otosclerosis. Ann Otol Rhinol Laryngol 1991;100:959-61.
14. Wiet RJ, Harvey SA, Bauer GP. Complications in stapes surgery. Options for preventions and management. Otolaryngol Clin N Am 1993; 26:471-90.
15. Burns JA, Lambert PR. Stapedectomy in residency training. Am J Otol 1996; 17:210-13.
16. Rothbaum DL, Roy J, Hager GD, Taylor RH, Whitcomb LL, Francis HW, Niparko JK. Task performance in stapedectomy: Comparison between surgeons of different experience levels. Otolaryngol Head Neck Surg 2003; 128: 71-7.

17. Mathews SB, Rasgon BM, Byl FM. Stapes surgery in a residency training program. Laryngoscope 1999; 109: 52-3.
18. Perkins R. Laser stapedectomy for otosclerosis. . Laryngoscope 1980;90:228-41.
19. Palva T. Argon laser in otosclerosis surgery. Acta Otolaryngol (Stockh) 1987;104:153-7.
20. Cherini SG, Horn KL, Bowman CA, Griffin G. Small fenestra stapedotomy using a fiberoptic hand-held argon laser in obliterative otosclerosis. Laryngoscope 1990;100:1276-82.
21. Lesinski SG. Lasers for otosclerosis-which one if any and why. Lasers in Surg & Med 1990;10:448-57.
22. Fritsch MH, Naumann IC. Phylogeny of the stapes prosthesis. Otol Neurotol 2008;29:407-15.
23. Wiet RJ, Kubek DC, Lemberg P, Byskosh AT. A Meta-analysis review of revision stapes surgery with Argon laser: Effectiveness and safety. Am J Otol 1997; 18:165-71.
24. Vincent R, Gratacap B, Causse JB, Cherini S. Argon laser and Gherini-Causse Endo-Otoprobe in otologic surgery. ENT Journal 1996; 12:770-80.
25. Szymanski M, Golabek W, Morshed K, Siwiec H. The influence of the sequence of Surgical steps on complications rate in stapedotomy. Otol Neurotol. 2007;28:152-6.
26. Lang EE, Waheed K, Colreavy M, Burns H. A retrospective review of stapes surgery following introduction of “reversal of the steps” techniqueClin Otolaryngol 2004;29:571-3.
27. Pauw BK, Pollak AM, Fisch U. Utricle, saccule, and cochlear duct in relation to stapedotomy. A histologic human temporal bone study. Ann Otol Rhinol Laryngol 1991;100:966-70.
28. Wong BJF, Neev J, van Gemert MJC. Surface temperature distributions in carbone dioxide, Argon and KTP(Nd:YAG) laser ablated otic capsule and calvarian bone. Am J Otol 1997; 18:766-72.
29. Lesinski SG, Palmer A. Lasers for otosclerosis: CO2 vs. Argon and KTP-532. Laryngoscope 1989;99:1-8.
30. Gantz BJ,Jenkins HA, Kishimoto S, Fisch U. Argon laser stapedotomy. Ann Otol Rhinol Laryngol 1982;91:25-6.
31. Martin MB, Rubinstein EN, Hirsch BE. High-frequency sensorineural hearing loss after stapedectomy. Otol Neurotol 2008;29:447-52.
32. Ballantyne J. Iatrogenic deafness. J Laryngol Otol 1970;84:967-1000.
33. Meyer SE. The effect of stapes surgery on high frequency hearing in patients with otosclerosis. Am J Otol 1999;20:36-40.

Chapter 5

General discussion
Summary and conclusions
Samenvatting
Acknowledgements
Curriculum Vitae
List of abbreviations

The possibility of surgical correction of impaired hearing in otosclerosis has inspired generations of otologists to seek better ways of achieving this goal.[1,2] A vast experience gathered during the modern history of stapes surgery enables implementation of the most refined technique, which evolved through numerous amendments made to the original stapedectomy.[3-7] The rising standards of care demanded critical appraisal of the traditional measures of surgical success.[8-10] An approach valuing the patient's satisfaction, rather than audiometric improvement, lead to development of more detailed and patient-oriented criteria of successful outcomes.[11-14] Concurrently, declining volume of stapes surgery over the past decades resulted in limited number of reports presenting a large series of patients. Only three reports concerning with the series of more than 1000 patients have been published in the indexed world medical literature during the past twenty years.[15-17] Moreover, diverse criteria used in reporting hearing results hinder valid comparison between the recently published small sets of otosclerosis patients.[18-21] The aim of this thesis was to evaluate functional hearing results of 1369 stapedotomies performed by a single surgeon in the period from 1991 to 2006. This extensive database allowed a separate analysis and comparison of hearing outcomes of unilateral[22,23] and bilateral[24] stapes surgery, as well as evaluation of the benefits of primary stapedotomy in patients with severe and profound hearing impairment[25], the magnitude of post-stapedotomy hearing improvement in congenital conductive hearing loss[26] and hearing results of malleo-vestibulopexy technique employed in special cases.[27] The distinctive feature of our methodological approach was an application of various tools of post-operative hearing assessment including Amsterdam Hearing Evaluation Plots[13], Glasgow Benefit Plot[14] and American Academy of Otolaryngology, Head & Neck Surgery guidelines for reporting results of the middle ear surgery[28], with the aim to achieve a more balanced and multi-dimensional comprehension of treatment outcomes.

In **chapter I** an introduction to the current concept of otosclerosis was presented. Review of pathologic and clinical studies of otosclerosis provided historical perspective for the contemporary state of knowledge of this unique pathology confined to human temporal bone. Evolution of the otosclerosis definition and classification linked to progress in understanding of the pathomorphologic changes was presented. Review of recent advances in genetics and immuno-histochemistry of otosclerosis enabled insight into its multi-factorial etiology, including role of heredity, auto-immune mechanisms and viral infection. Moreover, updated synopsis of molecular biology, pathomorphology and etiopathogenesis prepared for a discussion of current and future treatment options. In a second part of the introduction the evolution of surgical treatment of otosclerosis was presented. Refinements in surgical technique that resulted in various modifications were reviewed with a special emphasis on the changing vision of goals of treatment. A complete air-bone gap closure, considered by pioneers of stapes surgery as the main measure of successful surgery, has been recently incorporated in a broader system of criteria of success, focused on patient's perception of quality of sound and his ability to communicate, as a function of post-operative bilateral hearing. A pursuit of more inclusive approach to the evaluation of the surgical outcomes has inspired the studies, which were reported in the following chapters.[22-27]

In **chapter II** (2.1) the outcomes of 1145 primary stapedotomies were presented.[22] The objective of this study was to analyze the hearing results of stapes surgery reported according to the current (1995) guidelines of the American Academy of Otolaryngology, Head & Neck Surgery (AAO-HNS) Committee on Hearing and Equilibrium for the evaluation of results of treatment of conductive hearing loss[28], and simultaneously to evaluate these results with the Amsterdam Hearing Evaluation Plots (AHEP).[13] The effects of using different audiologic parameters on outcomes were analyzed. A detailed analysis of preoperative and postoperative hearing thresholds at 4 kHz was performed for evaluation of surgical trauma to cochlea assuming that BC thresholds at this frequency were the most sensitive indicator of the iatrogenic inner ear damage.[29-32] The audiometric results reported according to the AAO-HNS guidelines demonstrated significant improvement in mean postoperative air conduction (AC) and speech reception thresholds (SRT) with no change in BC in this series. A frequency-specific analysis of pre- and post-operative AC levels demonstrated statistically significant improvement at all frequencies, except AC thresholds at 12 kHz. The post-operative four-frequency average air-bone gap (ABG) was 7 dB (SD 6 dB) compared with 28 dB (SD 9 dB) preoperatively ($p<0.0001$). Closure of the ABG ≤ 10 dB was achieved in 82% of cases. The mean post-operative SRT was 27 dB (SD 12 dB) compared with 51 dB (SD 14 dB) preoperatively ($p<0.0001$). "Dead ear" occurred in one patient (0.1 %). In addition to reporting the averaged results according to the mean values of several audiometric parameters, raw data from the audiometric database were evaluated with the AHEP for a visual presentation of each individually operated ear. Addition of the Amsterdam Hearing Evaluation Plots (AHEP) to the assessment tools of large series of stapedotomies allowed graphic presentation of each individual case, enabling visual identification of successful and unfavorable results. As showed in AHEP graphs, the majority of the points lied below the upper

diagonal line representing 1052 (97.4 %) patients with postoperative ABG within 20 dB. Postoperative improvement of BC > 10 dB occurred in 57 of these cases (5%). The poor outcomes caused by worsening of post-operative BC by more than 10 dB were observed in 35 (3%) patients. An adjunct of AHEP allowed also a graphical identification of outliers providing more informed interpretation of summary statistics. Therefore, the application of the AHEP to large otologic database enabled easy identification of outliers and unfavorable outcomes that may not be recognized in the reports presenting audiometric data as summary statistics alone.
This study reported the largest series of primary stapedotomies evaluated with AHEP to date.

In **chapter III** (2.2) a bilateral hearing function following unilateral stapes surgery has been evaluated in the series of 751 primary stapedotomies.[23] A necessity of analysis of bilateral hearing function in interpretation of the results of unilateral stapes surgery has been emphasized by several authors.[11,14,42] Inclusion of preoperative and postoperative AC thresholds in both operated and non-operated ears in the assessment of benefit of surgery, as suggested by the Glasgow Benefit Plot (GBP) method[14], allows analysis of post-operative auditory improvement from the point of view of patients. The aims of this study were twofold: a) to evaluate both technical and functional results of a large series of stapedotomies using the Glasgow Benefit Plot (GBP)[14] criteria and the AAO-HNS Committee on Hearing and Equilibrium guidelines[28], and b) to identify prognostic factors for postoperative hearing levels after stapedotomy. For this purpose pre- and post-operative audiometric data calculated according to the guidelines of the AAO-HNS were analyzed using the GBP of normal and symmetrical hearing. Normal hearing was defined as an AC PTA threshold ≤ 30 dB, adopting the “Belfast Rule of Thumb”[12] for the definition of socially acceptable hearing. Symmetrical hearing was defined as an interaural difference of ≤ 10 dB, as proposed by the GBP method. Based on these criteria patients were grouped into three major categories according to their preoperative hearing impairment: unilateral, bilateral symmetric and bilateral asymmetric hearing impairment. The effects of type of preoperative hearing impairment and preoperative BC on achievement of postoperative normal and symmetric hearing were analyzed. The highest proportion of normal cochlear reserve was found in the group of unilateral hearing impairment. The highest proportion of mixed hearing loss was found in group with bilateral asymmetric hearing impairment. Postoperative symmetric hearing was found to be highly correlated with the type of preoperative hearing impairment. Normal postoperative

AC was found to be highly correlated with normal preoperative BC. The operated ear became the better hearing ear in 41% of all cases in this series but became the better hearing ear in 96% of the subgroup of patients with preoperative bilateral symmetric hearing impairment. Symmetrical hearing was achieved in 47% of all patients. Grouping of patients to categories of unilateral, bilateral symmetric and bilateral asymmetric hearing impairment has been found helpful for prognostication of postoperative hearing. Glasgow Benefit Plot, therefore, is a useful tool for establishing indications for a second ear stapedotomy. This study reported a largest series of primary stapedotomies evaluated with GBP up to the present time.

In **chapter IV** (2.3) the results of 394 bilateral stapedotomies were presented.[24] The aims of this study were to assess the benefits of a second-ear stapedotomy in achievement of binaural and symmetrical hearing. For this purpose the hearing results were evaluated using the GBP criteria and the success rates after 1st and 2nd ear surgery have been separately analyzed. A multi-dimensional approach in the reporting and the interpretation of surgical outcomes has been applied by using the AAO-HNS guidelines and GBP definitions of the types of pre-operative hearing impairment. A controversy regarding whether to operate or not on the second ear has been aroused from the concerns of doubling the risks of potentially detrimental to hearing complications[33-37], as well as from the difficulties in the assessment of benefits of a second ear procedure.[38-40] A rationale for bilateral stapedotomy, on the other hand, has been driven by pursuit of improving patient's quality of life, especially in the most crippled patients with bilateral mixed hearing loss, in whom a full rehabilitation after unilateral surgery or with hearing aids alone was not achievable.[41,42] The analysis of the results of bilateral stapes surgery demonstrated that the technical results after both 1st and 2nd procedure were identical with ABG closure ≤ 20dB achieved in 98 % of all cases. Evaluation of post-operative bilateral hearing function with the Glasgow Benefit Plot revealed that as a result of 1st ear surgery, 142 (72%) patients changed into the category of unilateral hearing loss, and as a result of 2nd ear surgery, 125 (64%) patients changed into the category of normal and symmetric hearing. Second ear surgery nevertheless appears to be less successful in achievement of postoperative thresholds below 30 dB. The results seem to be affected by a higher proportion of mixed hearing loss in the second otosclerotic ear: 67% of patients had mixed hearing loss before 2nd ear surgery compared with 44% before 1st surgery. Our results indicated that preoperative BC thresholds and the type of preoperative hearing impairment define whether bilateral normal hearing can be achieved

postoperatively. In patients with mixed and asymmetric hearing loss, bilateral stapes surgery remains beneficial in facilitating postoperative fitting of bilateral hearing aids. The results of our study support the conclusions that 2nd ear stapedotomy contributes to achievement of bilateral serviceable hearing in patients with bilateral hearing impairment. The low rate of intra-operative complications, similar in 1st and 2nd ear surgery, with no detrimental postoperative effect on cochlear and vestibular function, appears reassuring and speaks to the safety of bilateral stapes surgery.
The GBP used for the analysis of gains of 1st and 2nd ear stapedotomy appeared to be a helpful tool rendering preoperative identification of patients who will potentially benefit from the bilateral surgery. Comprehensive assessment of bilateral post-operative hearing, however, was a more complex task, which included parameters that could not be measured by the traditional audiometric tests alone. The scope of our further studies will be directed toward the functional aspects of speech intelligibility, using calculation and evaluation of Articulation Index (AI) as a prognostic tool for the achievable socially acceptable hearing. Recognition of importance of high frequency hearing for sound localization and understanding speech in noise will also require a separate study, which will be specifically focused on effects of stapes surgery on post-operative high and ultra-high frequencies thresholds.

In **chapter V** (3.1) the study was further focused on the most difficult for hearing dispensing patients with severe and profound hearing loss (SPHL).[25] Among 1369 stapedotomy cases reviewed, 144 surgeries had been performed on patients with pre-operative hearing thresholds of 71 dB or worse. The aims of this study were to evaluate the technical and functional results of 144 stapedotomies in patients with SPHL and to compare the success rate in this group with that of 1101 primary stapedotomies for mild/moderate hearing loss (MMHL). The outcomes were analyzed according to the 1995 AAO-HNS guidelines[28], and evaluated with the Amsterdam Hearing Evaluating Plots (AHEP)[13] and the Glasgow Benefit Plot (GBP).[14] In our series, severe and profound hearing loss in patients treated with primary stapedotomy was not rare comprising 13% of all cases. It appeared that the magnitude of hearing improvement post stapedotomy was greater in SPHL when compared with the MMHL. Even though achievement of normal hearing was often impossible due to poor pre-operative bone conduction thresholds, 88% of patients in this series were converted from the category of a severe or profound hearing loss to a mild or moderate hearing loss, or better, considerably facilitating their post-operative rehabilitation with hearing aids. Despite the higher proportion of mixed hearing loss and the older age of patients, only minor intra-operative complications were encountered, supporting our opinion that stapes surgery was a valuable adjunct to hearing aids for severe hearing loss in otosclerosis. When benefits of stapedotomy are weighted against the risks of this intervention in otosclerosis patients with severe or profound hearing loss (SPHL), it appears that more profound preoperative hearing impairment is associated with greater auditory improvement; therefore potential for post-operative gain seems to overweigh the risk. The mean gain in AC in patients with SPHL was 32 (± 13) dB compared with 23 (± 11) dB in the series of 1101 primary stapedotomies for MMHL. Even though the technical success in ABG closure in this group was lower, the change from SPHL to MMHL in this group of patients allows successful rehabilitation with hearing aids, which is probably more important for the improvement of quality of life than the degree of ABG closure. Analysis of the results using the Glasgow Benefit Plot demonstrated that symmetric hearing could be achieved in two thirds of patients, and even normal hearing can be achieved in select cases.

In **chapter VI** (3.2) the hearing results of the malleo-vestibulopexy (MVP) technique were presented.[27] This study reported special situations encountered in revision stapedotomies and congenital middle ear malformations, in which the reconstruction of conductive mechanism was particularly challenging since the traditional ossiculoplasty technique was not feasible. Correction of conductive hearing loss caused by complex ossicular pathology, encountered in congenital middle ear malformations or in revision stapedotomy, required flexible approach when standard ossiculoplasty techniques were not feasible. MVP has been used in cases of particularly unfavourable combination of the fixed stapes footplate with missing or abnormal incus. Despite the unfortunate preoperative middle ear situation, the short–term audiometric results in this series were equal or superior to results of ossiculoplasty or malleo-stapedotomy reported in literature, in which a comparable degree of ossicular pathology has been described.[43-52] In our experience, MVP technique was a valuable surgical option in cases with complex ossicular pathology, when standard ossiculoplasty could not be implemented. More research is required to evaluate the long-term stability of the malleo-vestibulopexy assembly.

In **chapter VII** (3.3) results of stapes surgery for conductive hearing loss secondary to congenital stapes fixation were reported.[26] Since the isolated congenital middle ear malformations were rare, we pursued a broad spectrum of research questions in order to maximize a scientific output of this study. Forty patients with operative findings consistent with isolated congenital stapes fixation or combined with ossicular chain malformations were included. Pre-operative factors, which have been evaluated, included severity of hearing loss, type of ossicular abnormalities and surgical technique used. Intra-operative findings were reported according to Cremers' classification.[53] The influence of the different types of ossicular pathology on hearing outcomes was examined aiming to recognize an association, if any, between the class of congenital anomaly and hearing results. Effects of surgery on the high- frequency hearing were also analyzed. Surgical failures were described. Assessment of postoperative bilateral hearing function with the Glasgow Benefit Plot demonstrated that 26/40 (65%) of patients achieved normal bilateral hearing and 24/40 (60%) had symmetrical hearing. From this analysis it appeared that even though postoperative normal hearing could not be achieved in all the cases, stapes surgery for congenital stapes fixation could be highly rewarding for patients. Nonetheless, it was not as successful as in the treatment of otosclerosis.

In **chapter VIII** (4.1) an evolution of surgical technique of stapedotomy employed in treatment of our patients from the year 1991 to present was described.[54] This method has been designed with the aim to minimize inner ear trauma. The surgical technique combined a modification of the reverse order technique[3,7,55-57] with the adjunctive use of argon beam laser[4,6,58-64] and modified self-crimping prosthesis.[64-68] Therefore, the procedure described in this thesis combines the strengths of several previously described techniques.[1-5,55,69-71] The employment of both laser assisted fenestration and the modification of self-crimping prostheses have reduced surgical trauma and technical difficulties relating to manual fenestration and prosthesis crimping. Furthermore, the advantage of reversed order technique in reducing operative complications rate has also been demonstrated.[7,22,56,63] In our experience, this technique provided better access to the footplate during fenestration than was traditionally enjoyed during the reverse-order technique, whilst maintaining the advantage of better stability of the footplate and incus. It appeared that keeping the incudo-stapedial joint and anterior crus intact during prosthesis placement provided additional protection against inadvertent footplate fragmentation or incus mobilization. The modification of prosthesis described in this paper facilitated placement without compromising post-operative hearing, thus providing reliable results and a low rate of piston-related failures. In addition, laser-assisted fenestration minimized cochlear damage enabling preservation of high frequency hearing.[22,63] The described technique has been also proven useful in training of stapedotomy surgery,

allowing the operation to be attempted in a series of controlled and increasingly difficult steps. Although we fully appreciated the advantages of this method, we also acknowledged its limitations, in particular a limited control during the fracture and removal of anterior stapedial crura. An employment of flexible middle ear endoscopy would possibly minimize the weakness of this step of the procedure.
At present time the stapes surgery as a method of hearing rehabilitation in otosclerosis competes with other technologies unseen at the time of emerging stapedectomy technique. The current alternative options include sophisticated hearing aids, implanted middle ear devices and cochlear implants.[72-77] This contemporary variety of different treatments required to reconsider the question whether stapedotomy is a too risky procedure to recommend as a first choice of treatment or as a bilateral procedure. The concerns regarding potential surgical complications are even sounder now considering a declining number of cases worldwide and consequent shrinking numbers of matured stapes surgeons.[78] An existence of a “learning curve” in mastering stapes surgery has explained the higher complications rate in hands of “occasional surgeons” reported by several authors.[79-83] Furthermore, the observed linear correlation between the hearing outcomes and the number of cases in published large series[15,16,22] supported the observation derived from a general surgical literature confirming that the large surgical volumes are associated with better operative outcomes.[84-89] The experience learned from the stapedotomy series presented in this thesis demonstrates very low incidence of severe complications: a “dead ear” occurred in one patient (< 0.1 %). The other complications encountered include a relatively mild and reversible balance disturbances and post-operative tinnitus, which did not affect overall patients’ satisfaction from the surgical results. In this aspect our outcomes are compatible with the other reported large series of stapedotomies[15-17] and support the view that stapes surgery is indeed a good option for correction of hearing impairment in otosclerosis especially if performed in a large volume otology center.

References

1. Shea J., Shea PF, McKenna MJ. Stapedectomy for otosclerosis. In: Glasscock M.E, Shambaugh G.E (eds). Surgery of the ear. BC Decker Inc 2003, pp 517-31.
2. House JP. The evolution of otosclerosis surgery. Otolaryngol Clin N Am 1993; 26:323-35.
3. Smyth GDL. Practical suggestions on stapedotomy. Laryngoscope 1982;92:952-3.
4. Perkins R. Laser stapedectomy for otosclerosis. . Laryngoscope 1980;90:228-41.
5. Rizer FM, Lippy. Evolution of techniques of stapedectomy from the total stapedectomy to the small fenestra stapedectomy. Otolaryngol Clin N Am 1993; 26:443-51.
6. Vincent R, Gratacap B, Causse JB, Cherini S. Argon laser and Gherini-Causse Endo-Otoprobe in otologic surgery. ENT Journal 1996; 12:770-80.

7. Szymanski M, Golabek W, Morshed K, Siwiec H. The influence of the sequence of Surgical steps on complications rate in stapedotomy. Otol Neurotol. 2007;28:152-6.
8. Sakai M. Proposal of a guideline in reporting hearing results in middle ear and mastoid surgery. Am J Otol 1994;15:291-93.
9. Monsell EM. New and revised reporting guidelines from the Committee on Hearing and Equilibrium. Otolaryngol Head Neck Surg 1995;113:176-8.
10. Jackler RK. Comparability in reporting outcomes: a scientific imperative. Am J Otol 1996;17: 811-12.
11. Smyth GDL, Patterson CC. Results of middle ear reconstruction: do patients and surgeons agree? Am J Otol 1985; 6: 276-279.
12. Toner JG, Smyth GDL. Comparison of methods of evaluating hearing benefit of middle ear surgery. J Laryngol Otol 1993; 107: 4-5.
13. De Bruijn AJG, Tange RA, Dreschler WA. Efficacy of evaluation of audiometric results after stapes surgery in otosclerosis. A method for reporting results from individual cases. Otolaryngol Head Neck Surg 2001;124:84-89.
14. Browning GG, Gatehouse S, Swan IR. The Glasgow Benefit Plot: a new method for reporting benefits from middle ear surgery. Laryngoscope 1991; 101: 180-185.
15. Vincent R, Sperling NM, Oates J, Jindal M. Surgical findings and long-term hearing results in 3050 stapedotomies for primary otosclerosis: a prospective study with the otology-neurotology database. Otol Neurotol 2006;27 (Suppl 2):25-47.
16. Quaranta N, Besozzi G, Fallacara RA, Quaranta A. Air and bone conduction change after stapedotomy and partial stapedectomy for otosclerosis. Otolaryngol Head Neck Surg 2006;133:116-20.
17. Daniels R, Krieger L, Lippy W. The other ear: findings and results in 1800 bilateral stapedectomies. Otol Neurotol. 2001; 22(5):603-7.
18. Jahnke KP. Thoughts on the validity of the evaluation of methods in stapes surgery. In: Jahnke KP, eds. Middle Ear Surgery: Recent Advances and Future Directions, Stuttgart: Thieme, 2000;131.
19. Berliner KI, Doyle KJ, Goldenberg RA. Reporting operative hearing results in stapes surgery: does choice of outcome measure make a difference? Am J Otol 1996;17:214-20.
20. Govartes PJ, Somers T, Offeciers FE. Box and whisker plots for graphic presentation of audiometric results of conductive hearing loss treatment. Otolaryngol Head Neck Surg 1998;118:892-95.
21. De Bruijn AJG, Tange RA, Dreschler WA. Efficacy of evaluation of audiometric results after stapes surgery in otosclerosis. The effects of using different audiologic parameters and criteria on success rates. Otolaryngol Head Neck Surg 2001;124:76-83.
22. Kisilevsky V, Dutt S, Bailie N, Halik J. Hearing results of 1145 stapedotomies evaluated with Amsterdam Hearing Evaluation Plots. J Laryngol Otol 2009;26:1-7.
23. Kisilevsky V, Bailie N, Dutt S, Halik J. Bilateral hearing function after stapedotomy evaluated with Glasgow Benefit Plot.(In submission).
24. Kisilevsky V, Bailie N, Dutt S, Halik J. Functional results of 394 bilateral stapedotomies evaluated with the Glasgow Benefit Plot. (In submission).
25. Kisilevsky V, Bailie N, Dutt S, Halik J. Results of stapes surgery in otosclerosis with severe and profound hearing loss. (In submission).
26. Kisilevsky V, Bailie N, Dutt S, Halik J. Small fenestra stapedotomy for congenital conductive hearing loss. (In submission).
27. Kisilevsky V, Bailie N, Dutt S, Safar A, Halik J. The Markham-Stouffville Hospital experience with malleovestibulopexy. Journal of Otolaryngology-Head & Neck Surgery 2009 (In print).
28. Committee on Hearing and Equilibrium guidelines for the evaluation of results of treatment of conductive hearing loss. Otolaryngol Head Neck Surg 1995; 113(3):186-3.
29. Ballantyne J. Iatrogenic deafness. J Laryngol Otol 1970;84:967-1000.
30. Smyth GDL. Immediate and delayed alterations in cochlear function following stapedectomy. Otolaryngol Clin N Am 1978;11:105-12.
31. Meyer SE. The effect of stapes surgery on high frequency hearing in patients with otosclerosis. Am J Otol 1999;20:36-40.
32. Martin MB, Rubinstein EN, Hirsch BE. High-frequency sensorineural hearing loss after stapedectomy. Otol Neurotol 2008;29:447-52.
33. Sale C. Bilateral deafness due to stapes surgery. Arch Otolaryngol. 1969; 90(4):467-9.

34. Ludman H, Grant H. The case against bilateral stapedectomy, and problems of post-operative follow-up – from the King's College Hospital series. J Laryngol Otol. 1973;87(9): 833-43.
35. Pratt L. Postoperative complications associated with bilateral stapedectomy. Laryngoscope 1972;1: 1189-98.
36. Mann W, Amedee R, Fuerst G. Hearing loss as a complication of stapes surgery. Otolaryngol Head Neck Surg.1996; 115(4):324-8.
37. Schuknecht H, Mendoza A. Cochlear pathology after stapedectomy. Am J Otolaryngol. 1981; 2(3):173-87.
38. Smyth G, Kerr A, Singh K. Second-ear stapedectomy – a continued controversy. J Laryngol Otol. 1975; 89:1047-56.
39. Porter MJ, Zeitoun H, Brookes GB. The Glasgow Benefit Plot used to access the effect of bilateral stapedectomy. Clin Otolaryngol Allied Sci.1995; 20(1):68-71.
40. De Bruijn AJ, Tange RA, Dreschler WA. Evaluation of second-ear stapedotomy with the Glasgow benefit plot. ORL J Otolaryngol Relat Spec. 1999; 61 (2): 92-7.
41. De Bruijn AJG, Tange RA, Dreschler WA, Grolman W, Schouwenburg PF. Bilateral stapedotomy in patients with otosclerosis: a disability-oriented evaluation of the benefit of second ear surgery. Clin Otolaryngol 1998;23:123-127.
42. Lundman L, Mendel L, Bagger-Sjoback D, Rosenhall U. Hearing in patients operated unilaterally for otosclerosis. Self-assessment of hearing and audiometric results. Acta Otolaryngol.1999; 119(4): 453-8.
43. Sheehy J. Stapedectomy: incus bypass procedures. A report of 203 operations. Laryngoscope 1982; 92:258-62.
44. Shea J. Malleus Teflon piston attachment prosthesis. Laryngoscope 1983;93: 989-991.
45. Schuknecht H, Bartley M. Malleus grip prosthesis. Ann Otol Rhinol Laryngol 1986;95: 531-4.
46. Cremers C. Myringochorda-vestibulopexy: a new method for total replacement of the ossicular chain. Am J Otol 1994; 15: 445-48.
47. Tange R. Ossicular reconstruction in cases of absent or inadequate incus, congenital malformation of the middle ear and epitympanic fixation of the incus and malleus. J Oto-Rhino-Laryngol Rel Spec 1996; 58:143-6.
48. Fisch U, Acar G, Huber A. Malleostapedotomy in revision surgery for otosclerosis. Otol Neurotol 2001; 22:776-85.
49. O'Reilly R, Cass S, Hirsh B, Kamerer D, Bernat R. et al. Ossiculoplasty using incus interposition: hearing results and analysis of the middle ear risk index. Otol Neurotol 2005;26:853-858.
50. Al-Qudah M, Dawes P. Malleus-stapes assembly: experience with two prostheses. J Laryngol Otol 2006; 120: 736-9.
51. Sasaki T. Xu A. Ishimoto S. Ito K. Yamasoba T. Kaga K. Results of hearing tests after total middle ear reconstruction. Acta Oto-Laryngol 2007; 127:474-9.
52. Dalchow C, Dunne A, Sesterhenn A, Teymoortash A, Werner J. Malleostapedotomy: the Marburg experience. Adv Oto-Rhino-Laryngol 2007; 65: 215-21.
53. Teunissen E, Cremers C. Classification of congenital middle ear anomalies. Report on 144 ears. Ann Otol Rhinol Laryngol 1993; 102:606-11.
54. Kisilevsky V, Bailie N, Halik J. Modified reverse order laser-assisted stapedotomy: How We Do It. (In submission).
55. Fisch U. Stapedotomy versus stapedectomy. Am J Otolaryngol 1982;2:112-17.
56. Lang EE, Waheed K, Colreavy M, Burns H. A retrospective review of stapes surgery following introduction of "reversal of the steps" technique. Clin Otolaryngol 2004;29:571-3.
57. Fiorino F, Barbieri F. Reversal of the steps stapedotomy technique with early removal of the posterior crus: early postoperative results. How we do it. Clin Otolaryngol 2008;33:359-69.
58. Palva T. Argon laser in otosclerosis surgery. Acta Otolaryngol (Stockh) 1987;104:153-7.
59. Cherini SG, Horn KL, Bowman CA, Griffin G. Small fenestra stapedotomy using a fiberoptic hand-held argon laser in obliterative otosclerosis. Laryngoscope 1990;100:1276-82.
60. Gantz B. Argon laser stapedotomy. Ann Otolaryngol Chir Cervicofac 1982;91:25-6.
61. Lesinski SG, Palmer A. Lasers for otosclerosis: CO2 vs. Argon and KTP-532. Laryngoscope 1989;99:1-8.
62. Wiet RJ, Kubek DC, Lemberg P, Byskosh AT. A Meta-analysis review of revision stapes surgery with Argon laser: Effectiveness and safety. Am J Otol 1997; 18:165-71.
63. Raut V, Halik J. Argon laser assisted small fenestra stapedotomy for otosclerosis. Auris Nasus Larynx 2005; 32:11-15.

64. Fritsch MH, Naumann IC. Phylogeny of the stapes prosthesis. Otol Neurotol 2008;29:407-15.
65. Raza A, Ali L, Ullah S, Iqbal S. Hearing improvement after stapedotomy using Teflon loop prosthesis. J CPS Pak 2006;16:659-61.
66. Kwok P, Fisch U, Strutz J. The crimping problem in stapes surgery. Adv Otorinolaryngol 2007;65:202-5.
67. Gjuric M, Rukavina L. Evolution of stapedectomy prosthesis over the time. Adv Otorino-laryngol 2007;65:174-8.
68. Huber A, Veraguth D, Schmid S, Roth T, Eiber A. Tight stapes prosthesis fixation leads to better functional results in otosclerosis surgery. Otol Neurotol 2008; 29:893-99.
69. Cremers CWR, Beusen JMH, Huygen PLM. Hearing gain after stapedotomy, partial platinectomy, or total stapedectomy for otosclerosis. Ann Otol Rhinol Laryngol 1991;100: 959-61.
70. Marquet J. Stapedotomy technique and results. Am J Otol 1985;6:63-67.
71. Causse JB, Causse JR, Parahy C. Stapedotomy technique and results. Am J Otol 1985;6:68-71.
72. Johnson E. Hearing aids and otosclerosis. Otolaryngol Clin N Am 1993; 26:491-502.
73. Lippy W, Burkey J, Schuring A, Rizer F. Stapedectomy in patients with small air-bone gaps. Laryngoscope 1997;107:919-922.
74. Venail F, Lavieille J, Meller R, Deveze A, Tardivet L, Magnan J. New perspectives for middle ear implants: first results in otosclerosis with mixed hearing loss. Laryngoscope 2007;117: 552-555.
75. Wiet RJ, Morgenstein SA, Zwolan TA, Pircon SM. Far-advanced otosclerosis. Cochlear implantation vs stapedectomy Arch Otolaryngol 1987; 113: 299-302.
76. Ramsden R, Rotteveel L, Proops D, Saeed S, van Olphen A, Mylanus E., Cochlear implantation in otosclerotic deafness. Adv Otolaryngol 2007;65:328-34.
77. Calmels M, Viana C, Wanna G et al., Very far-advanced otosclerosis: stapedotomy or cochlear implantation. Acta Otolaryngol 2007; 127: 574-8.
78. Conrad G. “Collective stapedectomy” (An approach to the numbers problem). J Laryngol Otol 1990;104:390-393.
79. Yung MW, Oates J, Vowler SL. The learning curve in stapes surgery and its implication to training. Laryngoscope 2006;116:67-71.
80. Rothbaum DL, Roy J, Hager GD, Taylor RH, Whitcomb LL, Francis HW, Niparko JK. Task performance in stapedectomy: Comparison between surgeons of different experience levels. Otolaryngol Head Neck Surg 2003; 128: 71-7.
81. Mathews SB, Rasgon BM, Byl FM. Stapes surgery in a residency training program. Laryngo-scope 1999; 109: 52-3.
82. Backous DD, Coker NJ, Jenkins HA. Prospective study of resident-performed stapedectomy. Am J Otol 1993;14:451-4.
83. Burns JA, Lambert PR. Stapedectomy in residency training. Am J Otol 1996; 17:210-13.
84. Morton R, Gray L, Tandon D, McIvor N. Efficacy of neck dissection: are surgical volumes important? Laryngoscope 2009;119:1147-52.
85. Chukmaitov A, Menachemi N, Brown S, Saunders C, Tang A, Brooks R. Is there a relationship between physician and facility volumes of ambulatory procedures and patient outcomes? J Ambul Care Manage 2008;31:354-69.
86. Pieracci F, Fahey T. Effect of hospital volume of thyroidectomise on outcomes following substernal thyroidectomy. World J Surg 2008;32:740-746.
87. Bell C, Hatch W, Cernat G, Urbach D. Surgeon volumes and selected patient outcomes in cataract surgery: a population-based analysis. Opthalmology 2007;114:403-4.
88. Tracey J, Zelmer J. Volumes and outcomes for surgical services in Canada. Healthc Q 2005;8:28-30.
89. Murray G, Teasdale G. The relationship between volume and health outcomes- a review. Scott Med J 2006;51:17-22.

Summary

This thesis is focused on the evaluation of the effectiveness of surgical management of otosclerosis and congenital stapes ankylosis. The design of this thesis has been shaped by the realities of the state of the art of contemporary otology: the demographical changes in otosclerosis population and the evolution of clinical and audiological management of otosclerosis. In demographical aspect, the declining volume of stapes surgery has resulted in scarce publications reporting a large series of patients and in shrinking numbers of experienced stapes surgeons. At the same time, the recent technological advances and the vast clinical experience have fostered emerging refined surgical technique and enabled highly successful technical results with a low rate of surgical complications. Considerable experience accumulated in our high-volume specialized otology center, with more than a thousand otosclerosis patients treated over the past decade, created a unique opportunity for a series of diverse retrospective studies united by a multi-dimensional methodological approach. At the outset of this thesis, we aimed to provide a multi-dimensional analysis of hearing results of the modified stapes surgery technique, incorporating both technical, surgeon-oriented parameters of success and more functional, patient-oriented criteria related to patients satisfaction. The pursuit to answer these scientific questions was driving the interest to perform this specific retrospective study.

An actual investigation of our surgical results has been preceded by an extensive literature search of the past and present of otosclerosis research. The overview of the history of otosclerosis and the evolution of stapes surgery has been summarized in chapter I “Introduction”, providing a historical background to the clinical and audiological aspects of contemporary otosclerosis management presented in this thesis. The scope and the extent of the “Introduction” was designed as a chapter of the textbook “Pathology of the ear” (Hawk M, Rutka J. edit. BC Decker Inc, in preparation) dedicated to otosclerosis.

The first part of the thesis (chapters 2.1, 2.2, 2.3) was concerned with the outcomes of primary stapedotomy. The second part of the thesis (chapters 3.1, 3.2, 3.3) has been dedicated to the evaluation of hearing results of stapes surgery in special situations. In the third part of the thesis (chapter 4.1) the modified surgical technique of reverse order laser-assisted stapedotomy has been described.

In **Chapter 2.1** the hearing results of a large series of primary stapedotomies have been presented. The charts of 1369 consecutive stapedotomies performed from

1991 to 2006 were retrospectively reviewed and 1145 cases of primary stapedotomy were included. Six hundred and ninety-five patients (61%) were women and 450 (39%) were men. The mean age was 45 years (SD 12 years), with a range from 16 to 84 years. The hearing outcomes were analyzed according to the 1995 AAO-HNS Committee on Hearing and Equilibrium guidelines. Frequency specific AC thresholds at 0.5, 1, 2, 3, 4, 6, 8 and 12 kHz and BC thresholds at 0.5, 1, 2, 3 and 4 kHz in operated and non-operated ears were tabulated in a computer database. Mean preoperative pure tone average of AC, BC and ABG at 0.5, 1, 2 and 3 kHz were compared with corresponding postoperative values. Postoperative ABGs were also calculated also for frequencies combinations of 0.5, 1, and 2 kHz and 0.5, 1, 2 and 4 kHz to evaluate the effects of these different audiometric parameters on success rate. Mean pre- and post-operative AC and BC thresholds at 4 kHz were also studied separately to evaluate the impact of stapedotomy on high frequency hearing. In addition to reporting the averaged results according to the mean values of several audiometric parameters, the raw data from the audiometric database were evaluated with the Amsterdam Hearing Evaluation Plots (AHEP) for visual presentation of each individually operated ear. The results of this study demonstrated significant improvement in mean postoperative air conduction (AC) and speech reception thresholds (SRT) with no change in BC. Closure of the air-bone gap (ABG) ≤ 10 dB was achieved in 82% of cases. “Dead ear” occurred in one patient (<0.1 %). An addition of AHEP enabled easy identification of outliers and unfavorable outcomes, providing detailed representation of surgical outcomes in each individual operated ear.

In **Chapter 2.2** bilateral hearing function in patients operated unilaterally for otosclerosis has been evaluated. 751 cases of primary unilateral stapedotomy were included in this retrospective study. The hearing results were evaluated according to the 1995 AAO-HNS Committee on Hearing and Equilibrium guidelines and criteria of symmetric and normal hearing of Glasgow Benefit Plot (GBP). Both operated and non-operated ears were tested at 0.5, 1, 2, 3, 4, 6, 8, and 12 kHz for air conduction (AC), and .5, 1, 2, 3 and 4 kHz for bone conduction (BC). The mean preoperative PTA for AC, BC, and ABG values at 0.5, 1, 2, and 3 kHz were compared with their corresponding postoperative values. Postoperative BC thresholds were used to calculate postoperative ABG values. In addition to reporting the results of AC, BC, and ABG gain, we utilized the GBP for evaluation of bilateral hearing status in each individual case. Normal hearing was defined as an AC PTA threshold ≤ 30 dB, symmetrical hearing was defined as an interaural difference of ≤ 10 dB, as proposed by the GBP method. Based on these criteria, patients were grouped into three major categories according to their preoperative hearing impairment: Group 1- unilateral hearing impairment (481 patients), Group 2 - bilateral asymmetric hearing impairment (166 patients), and Group 3- bilateral symmetric hearing impairment (104 patients). The technical results of stapedotomy were highly successful in all groups despite of the differences in preoperative hearing status. A mean postoperative ABG of 7.5 dB was achieved in all groups, as well as a significant ($p<0.001$) improvement of AC thresholds. Evaluation of bilateral postoperative hearing with the GBP demonstrated that achievement of bilateral normal hearing was most successful in the group with unilateral hearing impairment, with 78% of patients having postoperative normal hearing. The achievement of normal hearing has been found highly correlated with the type of preoperative hearing impairment ($r=0.74$) and with the preoperative bone conduction ($r=0.61$).

In **Chapter 2.3** the benefits of a second-ear stapedotomy have been analyzed based on the results of 394 bilateral stapedotomies evaluated with the GBP. The audiometric data from 197 patients have been reported according to the 1995 AAO-HNS Committee on Hearing and Equilibrium guidelines and the success rates after 1 st and 2 nd ear surgeries were separately analyzed. One hundred twenty three patients (62%) were women and 74 (38%) were men. The mean age was 43 years (SD 11 years) at 1st ear surgery and 45 years (SD 10 years) at 2nd ear surgery. The mean preoperative hearing levels calculated as a PTA at frequencies of 0.5, 1, 2 and 3 kHz were 54 dB (SD 14 dB) in operated ear and 51 dB (SD 14 dB) in the non-operated ear at 1st ear surgery and 51 dB (SD 14 dB) and 29 dB (SD 11 dB) respectively, at 2nd ear surgery. The mean preoperative ABG was 27 dB (SD 9 dB) at 1st ear surgery and 25 dB (SD 8 dB) at 2nd ear surgery. Statistical analysis was carried out for comparisons between pre- and postoperative values using paired t-test for the evaluation of ABG, AC, BC, PTA, and AC gain. The technical success in ABG closure ≤ 10dB was significantly higher in 1st ear surgeries (90% compared with 80% in 2nd ear, $p<0.05$). ABG closure ≤ 20dB was demonstrated in 98 % of cases after both 1st and 2nd procedure. Normal postoperative hearing was achieved in 73% of 1st ear stapedotomies and in 64% of 2nd ear stapedotomies. Postoperative BC thresholds improved by 4 dB after the 1st ear surgery and by 3 dB after the 2nd ear surgery. Symmetric hearing (interaural difference of less than 10 dB, according to the GBP criteria) was achieved in 80% of cases after the 2nd ear stapedotomy. The operated ear became the better hearing ear in 97% of cases after the 1st ear surgery and in 46% of cases after the 2nd ear procedure. The results of this study indicated that 2nd ear stapedotomy contributed to providing bilateral symmetrical hearing, when achievable, and that it was beneficial in facilitating postoperative rehabilitation with hearing aids in patients with mixed and asymmetric hearing loss. The low rate of intra-operative complications, similar in 1st and 2nd ear surgery, with no detrimental postoperative effect on cochlear and vestibular function, appears reassuring and speaks to the safety of bilateral stapes surgery in properly selected patients.

The **Chapter 3.1** was concerned with the evaluation of the results of stapes surgery in patients with severe or profound hearing loss (SPHL), defined as pre-operative AC PTA ≥ 71dB. This group was the most challenging for hearing rehabilitation due to the higher proportion of mixed hearing loss associated with poor cochlear reserve. Among 1369 charts retrospectively reviewed in this study, 127 patients (144 stapedotomies) have been identified and included in statistical analysis. The technical and functional results of 144 stapedotomies in patients with SPHL have been compared with the success rate of 1101 primary stapedotomies for mild/moderate hearing loss (MMHL). There were one hundred and fifteen ears (80%) with severe hearing loss (71 dB ≤PTA< 90 dB) and 29 ears (20%) with profound hearing loss (91 dB ≤PTA< 120 dB). Sixty-nine patients (54%) were women and 58 (46%) were men. The mean age was 55 (SD 12) years; range from 25 to 84 years. Fifty patients had other than hearing loss complaints: forty patients (28%) had tinnitus, six patients (4%) complained of occasional dizziness and four patients (3%) had both. The audiometric data were presented according to the 1995 AAO-HNS Committee on Hearing and Equilibrium guidelines, and evaluated with the Amsterdam Hearing Evaluating Plots (AHEP) and the Glasgow Benefit Plot (GBP). The results of this study revealed that the magnitude of hearing improvement was greater in patients with SPHL when compared to the patients with MMHL. The mean gain in AC in patients with SPHL was 32 (± 13) dB compared with 23 (± 11) dB in MMHL. The mean postoperative ABG of 10 (± 8) dB was demonstrated in the group with SPHL compared with 7 (± 5) dB in the group with MMHL. ABG closure ≤ 10 dB was achieved in 63% of cases and ≤ 20 dB in 90% of patients with SPHL, compared with 82% and 97% respectively in the patients with MMHL. Despite the higher proportion of mixed hearing loss and the older age of patients in SPHL group, only minor intra-operative complications were encountered in this series and it was concluded that stapes surgery is a safe and highly rewarding treatment for patients with advanced otosclerosis.

In **Chapter 3.2** the results of twenty-four malleo-vestibulopexy procedures (MVP) employed in revision stapedotomies or in cases of congenital conductive hearing loss were presented. The archived charts of all consecutive stapedotomy cases were reviewed. Of the 1369 surgical cases, 89 were revision stapedotomies and 48 were performed for congenital stapes fixation. The combination of missing or eroded incus with immobile footplate was found in 17/89 (19%) of revision stapedotomies and in 7/48 (15%) of congenital anomalies. The surgical options of traditional ossiculoplasty in these cases were limited and MVP technique has been employed. The hearing outcomes were analyzed according to the 1995 American Academy of Otolaryngology - Head and Neck surgery Committee on Hearing and Equilibrium guidelines. Frequency specific air conduction (AC) thresholds at 0.5, 1, 2, 3 and 4 kHz and bone conduction (BC) thresholds at 0.5, 1, 2, 3 and 4 kHz in operated and non-operated ears were tabulated in a computer database. ABG closure, AC gain and SRT improvement have been chosen as main treatment outcome measures. ABG closure ≤ 20 dB was achieved in 61% and ≤ 10 dB in 33% of all cases. ABG **≤** 10 dB was achieved in 4/7(57%) of congenital anomalies and in 5/17(29%) of revision procedures. Significant ($p<0.001$) improvement was demonstrated postoperatively in all audiometric parameters: AC threshold changed from 54 (SD 14) dB preoperatively to 39 (SD19) dB postoperatively, SRT changed from 53 (SD 13) dB preoperatively to 35 (SD19) dB postoperatively. No cases of SNHL (“dead ear”) were associated with this technique in our series. One patient demonstrated short-term self-limited vestibular symptoms. In our experience, MVP is a valuable surgical option in cases when traditional ossiculoplasty is not feasible. Further follow up will be required to test the long-term stability of MVP assembly.

In **Chapter 3.3** the results of stapes surgery for correction of congenital conductive hearing loss were presented. A retrospective review of 1369 consecutive stapedotomies revealed that 40 surgeries were performed for congenital stapedial anomalies. The diagnosis of congenital stapes fixation was based on a history of non-progressive conductive hearing loss from early childhood and an intra-operative finding of hypoplastic or malformed stapes or incudo-stapedial joint in the absence of an otosclerotic focus. Intra-operative findings were tabulated according the Cremers classification of congenital middle ear anomalies. The hearing outcomes were analyzed according to the 1995 AAO-HNS Committee on Hearing and Equilibrium guidelines. Means of preoperative pure tone average of air conduction (AC), bone conduction (BC) and air-bone gap (ABG) on 0.5, 1, 2 and 3 kHz were compared with corresponding postoperative values. Postoperative bone conduction values were used to calculate postoperative air-bone gap. Hearing results demonstrated significant ($p<0.001$) improvement of AC thresholds from 56 dB (SD 16.9) preoperatively to 34 dB (SD 17.1) postoperatively. BC thresholds remained unchanged: 24 dB (SD 11.7) and 23 dB (SD 12) respectively. Mean postoperative SRT was 31 dB (SD 15) compared with 53 dB (SD 14) preoperatively. The postoperative ABG closure to within 10 dB was achieved in 24/40(60%) of congenital cases compared with 82% of 1145 primary stapedotomies performed for otosclerosis. Overall better operative results were achieved in class 1 malformations. Postoperative mean AC PTA, high frequencies thresholds, BC and SRT were better in class 1 anomalies compared with class 2 and 4 combined. However, the differences between the groups did not reach statistical significance due to a small number of classes 2, 3 and 4 populations. Considering the rarity of isolated middle ear malformations causing a pure conductive hearing loss, the results of this series supplied valuable information on treatment outcomes of stapes surgery in this specific hearing impaired population.

In **Chapter 4.1** the modified surgical technique of reversed order laser–assisted stapedotomy was described. This chapter was prepared in “How I Do It” format, corresponding with the “Introduction” part which discussed the history and evolution of stapes surgery. A step-by-step description of the procedure was illustrated by intra-operative micro-photographs of the most important details, providing better appreciation of described modifications. In our experience, the presented technique enabled better access to the footplate during fenestration, whilst maintaining the advantage of better stability of the footplate and incus. Modification of the prosthesis facilitated placement without compromising hearing results. Laser-assisted fenestration minimized cochlear damage enabling preservation of high frequency hearing. The technique described in this chapter has been also proven useful in training of stapedotomy surgery, allowing the operation to be attempted in a series of controlled and increasingly difficult steps.

Based on the results of presented large series, we arrived to the conclusion that modified stapedotomy technique provides a valuable tool for safe and effective correction of hearing loss caused by stapes fixation. In experienced hands it allows achievement of very low rate of surgical complications, therefore supporting the suggestion that centralization of stapes surgery in the high volume otology centers might result in most beneficial outcomes for patients. The complexity of the comprehensive assessment of effects of stapes surgery is related to involvement of not only audiometric characteristics in operated ear, but also by dynamics of hearing impairment and non-auditory variables. Future research of postoperative quality of life and speech and sound perception will help to find more balanced answers related to these complex questions.

Samenvatting

Dit proefschrift is gericht op de evaluatie van de chirurgische behandeling van otosclerose en de resultaten van stijgbeugelvervangende chirurgie in andere bijzondere situaties. De opzet van deze proefschriftstudie wordt mede bepaald door de omstandigheden van de hedendaagse oorheelkunde, te weten: de demografische veranderingen in de otosclerose populatie, de ontwikkelingen in de audiologische mogelijkheden en de chirurgische mogelijkheden om otosclerose te behandelen. Het demografisch aspect is dat de geleidelijke afname in aantallen van stijgbeugelvervangende chirurgie er toe heeft geleid dat er nog maar zelden over grote series publicaties over de resultaten van stijgbeugelvervangende chirurgie verschijnen. Tegelijk neemt het aantal oorartsen af, dat een grote ervaring heeft met stijgbeugelvervangende chirurgie. Anderzijds hebben recente technieken ervoor gezorgd, dat de resultaten van de stijgbeugelvervangende chirurgie verder verbeterd zijn met een zeer laag percentage van complicaties. In ons oorheelkundig centrum zijn de afgelopen tien jaar tezamen meer dan 1000 otosclerose patiënten geopereerd. Dit verschafte een unieke mogelijkheid om een serie verschillende retrospectieve studies verricht met dezelfde methoden uit te voeren. De opzet van deze studies is om een analyse van de resultaten voor het gehoor van een gemodificeerde stijgbeugelvervangende operatie te verrichten, waarbij gelet werd op technische en aan de chirurg georiënteerde parameters die het succes kunnen beïnvloeden.

Voordat deze retrospectieve studies naar de uitkomsten van stijgbeugelvervangende chirurgie werden begonnen, werd de oude en meer hedendaagse literatuur over otosclerose bestudeerd. Een overzicht van de geschiedenis van otosclerose en van de ontwikkeling van de stijgbeugelvervangende chirurgie in de tijd wordt beschreven in **Hoofdstuk I** Inleiding. Dit plaatst de klinische en audiologische aspecten van de hedendaagse behandeling van otosclerose, zoals die in dit proefschrift beschreven zijn, tegen een historische achtergrond.

Het eerst deel van dit proefschrift (Hoofdstuk 2.1, 2.2, 2.3) is gericht op het beschrijven van de resultaten van de eerste stijgbeugelvervangende operatie in een patiënt. Het tweede deel van dit proefschrift (Hoofdstuk 3.1, 3.2, 3.3) gaat over de evaluatie van de resultaten van stijgbeugelvervangde chirurgie voor het gehoor. In het derde deel van dit proefschrift (Hoofdstuk 4.1) wordt een gemodificeerde chirurgische techniek voor de stapedotomie met behulp van een laser voorgesteld, waarbij de volgorde van de chirurgische stappen is veranderd.

In **Hoofdstuk 2.1** worden de resultaten voor het gehoor na een eerste stapedotomie operatie voor een grote serie operaties gepresenteerd. Retrospectief werden de gegevens van 1369 elkaar opvolgende stijgbeugel-vervangende operaties, verricht tussen 1991 tot 2006, uit de medische statussen verzameld. In 1145 gevallen ging het om een eerste stapedotomie in een patiënt. Zeshonderdvijfennegentig patiënten (695/1145 = 61%) waren vrouw en vierhonderdvijftig (450/1145 = 39%) waren man. De gemiddelde leeftijd was 45 jaar (SD 12 jaar) met een variatie in leeftijd van 16 tot 84 jaar. De evaluatie van de resultaten voor het gehoor werden verricht volgens de in 1995 uitgevaardigde richtlijnen van het AAO-HNS Committee on Hearing and Equilibrium. De resultaten van toonaudiometrie in het geopereerde en in het niet geopereerde oor werden in een computer data base opgeslagen. Voor de luchtgeleiding ging het om drempelwaarden gemeten bij 0.5, 1, 2, 3, 4, 6, 8 en 12 kHz. Voor de beengeleiding ging het om drempelwaarden bij 0.5, 1, 2, 3, 4 kHz. De gemiddelde preoperatieve luchtgeleidingsdrempels, de preoperatieve beengeleidingsdrempels en de luchtbeengeleidingscomponent voor 0.5, 1 en 3 kHz werden vergeleken met de postoperatieve uitkomsten. De postoperatieve luchtbeengeleidingscomponent werd evenzo berekend voor andere combinaties van toonhoogten te weten 0.5, 1 en 2 kHz en 0.5, 1, 2 en 4 kHz om het effect van deze verschillende parameters op het succes van deze operaties te leren kennen. De gemiddelde preoperatieve en postoperatieve luchtgeleiding- en beengeleidingsdrempels bij 4 kHz werden ook afzonderlijk bestudeerd om de betekenis van een stapedotomie voor deze hogere frequentie te leren kennen. In aanvulling op de beschrijving van de gemiddelde resultaten voor de eerder genoemde audiometrische parameters, werden de oorspronkelijke audiometrische gegevens uit de databank geëvalueerd met behulp van de Amsterdam Hearing Evaluation Plots (AHEP) om een visuele presentatie te verkrijgen voor elk geopereerd oor. De resultaten van deze studie toonden een opmerkelijke verbetering van de gemiddelde postoperatieve luchtgeleidingsdrempels en spraakverstaan resultaten zonder een verandering in de beengeleidingsdrempel. Het sluiten van de luchtbeengeleidingsdrempel binnen een waarde van 10 dB of kleiner werd in 82% van de geopereerde oren bereikt. Een volledig uitgevallen gehoor trad slechts na één operatie op (<0.1%). De toevoeging van de Amsterdam Hearing Evaluation Plot (AHEP) maakte het mogelijk de ooroperaties met een ongunstige uitkomst voor de luchtgeleiding of beengeleiding snel te herkennen, omdat voor elke operatie afzonderlijk deze resultaten grafisch apart worden weergegeven.

In **hoofdstuk 2.2** is de bilaterale postoperatieve functie van het gehoor na een stapedotomie op een van beide oren geëvalueerd. In deze retrospectieve studie werden 751 personen na een eerste eenzijdige stapedotomie betrokken. Deze gehoorresultaten werden volgens de 1995 AAO-HNS Committee on Hearing and Equilibrium richtlijnen en de criteria van de Glasgow Benefit Plot (GBP) voor een symmetrisch en een normaal gehoor gepresenteerd. Het gehoor van het geopereerde en het niet geopereerde oor werd gemeten voor de luchtgeleiding bij 0.5, 1, 2, 3, 4, 6, 8 en 12 kHz en voor de beengeleiding bij 0.5, 1, 2 en 3 kHz. De gemiddelde toonaudiometrische bevindingen voor de luchtgeleiding, de beengeleiding en voor de luchtbeengeleidingscomponent bij 0.5, 1, 2 en 3 kHz werden vergeleken met de daarmee overeenkomende postoperatieve waarden.

Postoperatieve beengeleidingsdrempels werden gebruikt om het postoperatieve luchtbeengeleidingsverschil te berekenen. In aanvulling op het beschrijven van de uitkomsten voor de luchtgeleiding en de beengeleiding alsook de lucht-beengeleidingscomponent werd de Glasgow Benefit Plot (GBP) gebruikt om het bilateraal functioneren van beide oren te evalueren voor iedere persoon afzonderlijk. Een normaal gehoor werd gedefinieerd als een verschil in de gemiddelde waarde voor de luchtgeleiding (Pure Tone Average) op beide oren beter of gelijk aan 30 dB. Een symmetrisch gehoor werd, conform de richtlijnen van de Glasgow Benefit Plot (GBP), gedefinieerd als een verschil van 10 dB of kleiner tussen de luchtgeleidingswaarden (PTA) voor beide oren. Conform deze criteria werden de patiënten ondergebracht in drie groepen in overeenstemming met hun preoperatieve gehoorverlies:

Groep 1: een eenzijdig gehoorverlies	(481 personen)
Groep 2: een beiderzijds asymmetrisch gehoorverlies	(166 personen)
Groep 3: een beiderzijds symmetrisch gehoorverlies	(104 personen)

De resultaten voor de verbetering van het gehoor bleken zeer succesvol voor elk van deze 3 groepen, ondanks het verschil in de preoperatieve gehoordrempels. Een gemiddelde postoperatieve luchtbeengeleidingscomponent van 7.5 dB werd in alle drie deze groepen bereikt. Een significante verbetering van de luchtgeleidingsgehoordrempel ($p \leq 0.001$) werd bereikt. Evaluatie van het gehoor voor wat betreft het bereiken van beiderzijds een normaal gehoor volgens de criteria van de Glasgow Benefit Plot (GBP) toont dat dit, met een percentage van 78%, het best gelukt is in Groep 1 met alleen een eenzijdig gehoorverlies. Het bereiken van een normaal gehoor blijkt in hoge mate gecorreleerd te zijn met het preoperatieve gehoorverlies (r = 0.74) en met de preoperatieve beengeleidingsdrempel (r = 0.61).

In **hoofdstuk 2.3** worden de resultaten, geanalyseerd met de Glasgow Benefit Plot (GBP), beschreven van een stapedotomie op het andere tweede oor. De audiometrische bevindingen bij 197 personen zijn bewerkt volgens de richtlijnen van de 1995 AAO-HNS Committee on Hearing and Equilibrium. De mate van succes van de stapedotomie op het eerst geopereerde oor en op het andere tweede oor werden afzonderlijk geanalyseerd. Honderddrieentwintig van de 197 personen (123/197= 62%) waren vrouw en 74 van de 197 (38%) waren man. De gemiddelde leeftijd was 43 jaar (SD 11 jaar) bij de eerste stapedotomie en 45 jaar (SD 10 jaar) bij de tweede stapedotomie op het andere oor. De gemiddelde preoperatieve luchtgeleidingsgehoordrempels berekend voor 0.5, 1, 2 en 3 kHz waren 54 dB (SD 14 dB) in het geopereerde oor en 51 dB (SD 14 dB) in het niet geopereerde tweede oor ten tijde van de chirurgie van het eerst oor. Ten tijde van de operatie op het tweede oor waren deze waarden respectievelijk 51 dB (SD 14 dB) en 29 dB (SD 11dB). De gemiddelde preoperatieve luchtbeengeleidingscomponent was 27dB (SD 9dB) ten tijde van de chirurgie op het tweede oor. Een statistische analyse werd uitgevoerd om een vergelijking te maken tussen de preoperatieve en de postoperatieve uitkomsten. De paired t-test werd hierbij gebruikt voor de evaluatie van de luchtbeengeleidingscomponent, de luchtgeleiding, de beengeleiding, het berekende gemiddelde voor de luchtgeleiding (Pure Tone Average) en de winst voor de luchtgeleidingsdrempel. Het bereikte resultaat voor het sluiten van de luchtbeengeleidingscomponent (≤ 10 dB) was significant hoger voor de stapedotomie op het eerst geopereerde oor (90%) vergeleken met 80% ($P< 0.05$) voor het als tweede geopereerde oor. Sluiting van de luchtbeengeleidingscomponent (≤ 20dB) werd bereikt in 98% van de operaties zowel voor de stapedotomie op het eerste oor als voor de stapedotomie op het tweede oor. Een normaal postoperatief gehoor werd bij 73% van de stapedotomieën op het eerste oor en bij 64% van de stapedotomieën op het tweede oor bereikt.

De postoperatieve beengeleidingsdrempels verbeterden 4 dB na de eerste oor stapedotomieën en 3 dB na de tweede oor stapedotomieën. Een symmetrisch gehoor (volgens de GBP criteria een verschil tussen de gehoordrempels van beide oren van ≤ 10 dB) werd in 80% van de gevallen na een tweede oor stapedotomie bereikt. Het geopereerde oor werd het beste horende oor in 97% van operaties op het eerst geopereerde oor en in 46% van de operaties op het tweede oor met een stapedotomie. Deze resultaten tonen dat stapedotomie op het tweede oor bijdroeg

aan het verschaffen van een binauraal horen zo bereikbaar en dat de tweede stapedotomie heeft bijgedragen aan het beter postoperatief kunnen revalideren van patiënten met (eerder) een gemengd en asymmetrisch gehoorverlies.
Het lage percentage van intra-operatieve complicaties zonder ernstige post-operatieve effecten op de functie van het binnenoor-cochlea en het evenwichts-orgaan, zowel bij een eerste als bij een tweede oor stapedotomie, is geruststellend en ondersteunt de opvatting dat een stapedotomie op het tweede oor een veilige en goede behandeling is in preoperatief goed geselecteerde patiënten.

In **hoofdstuk 3.1** worden de resultaten van stapedotomieën geëvalueerd bij patiënten met preoperatief een ernstig of zeer ernstig gehoorverlies, te weten een gemiddelde preoperatieve luchtgeleidingsdrempel (PTA) groter of gelijk aan 71 dB. Deze groep patiënten met grote gemengde gehoorverliezen en daarmee ernstige binnenoorverliezen is moeilijk te revalideren vanwege hun zo ernstige gehoorverlies. Voor 127 patiënten met 144 stapedotomieën uit de oorspronkelijke groep van 1369 stapedotomieën geldt dat zij preoperatief aan deze criteria voor een ernstig gehoorverlies voldeden. Een statistische analyse vond plaats.

De gehoorresultaten van deze 144 stapedotomieën werden vergeleken met de bereikte resultaten in de groep van 1101 eerste oor stapedotomieën voor milde/matige gehoorverliezen (MMHL). Er waren 115 oren (80%) met een ernstig gehoorverlies (71 dB ≤ PTA < 90 dB) en 29 oren (20%) met een zeer ernstig gehoorverlies (91 dB ≤ PTA <120 dB). Negenenzestig (69) van deze patiënten waren vrouw (69/115=54%) en 58 van deze patiënten (58/115=46%) waren man. De gemiddelde leeftijd was 55 jaar (SD12) ; de leeftijd varieerde van 25 tot 84 jaar. In totaal 50 van deze personen hadden ook andere klachten: veertig (40/115=28%) hadden oorsuizen, zes (6/115=4%) hadden zowel oorsuizen als af en toe duizeligheid. De audiometrische gegevens werden gepresenteerd volgens de richtlijnen van 1995- AAO-HNS Committee on Hearing and Equilibrium en verder geëvalueerd met de Amsterdam Hearing Evaluating Plots (AHEP) en de Glasgow Benefit Plot (GBP). De analyse van de operatieresultaten toont dat de grootte van de gehoorwinst groter was (32 ±13 dB) voor de groep met de ernstige gemengde gehoorverliezen (SPHL) vergeleken met de groep met de matige gehoorverliezen (MML). De gemiddelde luchtbeengeleidingscomponent was in de SPHL-groep 10 (±8) dB en in de MMHL-groep 7 (±5) dB. In de SPHL-groep werd een sluiten van de luchtbeengeleidingscomponent met ≤ 10 dB bereikt bij 63% en met ≤ 20dB in 90% van oren. Voor de MMHL-groep was dit respectievelijk ≤ 82% en 97%. Ondanks het hogere percentage gemengde gehoorverliezen en de hogere leeftijd in de SPHL-groep werden alleen matige/geringe peroperatieve complicaties in deze serie ervaren en de conclusie was dat stijgbeugel-vervangende chirurgie een veilige en een zeer succesvolle behandeling is voor patiënten met een vergevorderd stadium van otosclerose.

In **hoofdstuk 3.2** worden de resultaten voor 24 malleo-vestibulopexy operaties (MVP) uitgevoerd bij revisie stapedotomieën of in geval van een congenitale ketenanomalie. Deze gevallen werden verkregen uit de serie elkaar opvolgende stijgbeugelvervangende operaties.

Bij de 1369 geanalyseerde stijgbeugelvervangende operaties ging het in 89 gevallen om een revisie stapedotomie en in 48 om een aangeboren stapesankylose. De combinatie van een ontbrekend lange been van de incus met een immobiele voetplaat werd in 17/89 (19%) van de revisiestapedotomieën en in 7/48(15%) van de congenitale ketenanomalieën gevonden. De chirugische mogelijkheden van een ketenreconstructie bij een gefixeerde stijgbeugel cq stijgbeugelvoetplaat zijn zeer beperkt, zodat de malleovestibulopexie methode, ook wel malleostapedotomie genoemd, werd toegepast. De gehoorresultaten werden geanalyseerd volgens de richtlijnen van de 1995 AAO – HNS Committee on Hearing and Equilibrium. De luchtgeleidingsdrempels voor 0.5, 1,2,3 en 4 kHz en de beengeleidingsdrempels voor 0.5, 1,2,3 en 4 kHz in de geopereerde en niet geopereerde oren werden opgeslagen in het gecomputeriseerde databestand. Sluiting van de luchtbeengeleidingscomponent, de winst met de luchtgeleidings-drempel en de verbetering in het spraakverstaan werden gekozen als methoden om de resultaten van de gehoorverbeterende chirurgie te meten. Een luchtbeengeleidingscomponent ≤ 10 dB werd in 29/89 (33%) en van ≤ 20 dB in 54/89 (61%) van de 89 gevallen bereikt.

In de congenitale serie van 48 geopereerde middenooranomalieën werd bij zeven oooroperaties met behulp van een malleovestibulopexie een luchtbeengeleidings-component ≤ 10 dB bereikt bij 4/7 (57%) oren en ≤ 20 dB bij 5/7 (72%) oooroperaties.
De luchtgeleidingsdrempel verbeterde van 54 dB (SD 14) preoperatief naar 39 dB (SD19) postoperatief. De 50% spraakverstaanbaarheidswaarde verbeterde van 53 dB (SD 13) naar 35 dB (SD 19) postoperatief. In deze serie operaties werden als complicatie postoperatief geen dove oren vastgesteld.

Na een operatie werden eventuele vestibulaire klachten genoteerd, welke spontaan verdwenen. Naar onze mening is de malleovestibulopexie een waardevolle chirurgische optie in gevallen waarin een meer traditionele vorm van ketenreconstructie zoals de incudostapedotomie niet uitvoerbaar is. Over een langere tijd zijn vervolgcontroles nodig om de lange termijn resultaten van de malleostapedotomie te kunnen leren kennen.

In **Hoofdstuk 3.3** zijn de resultaten van stapesvervangende chirurgie vanwege een aangeboren gehoorverlies gepresenteerd. In 41 van de 1369 elkaar opvolgende stapesvervangende operaties was sprake van een congenitale stapesankylose. De diagnose congenitale stapesankylose was gebaseerd op de voorgeschiedenis van een niet progressief conductief gehoorverlies vanaf de eerste kinderjaren en de peroperatieve bevinding van een hypoplastische of misvormde stapes of een afwijkend incudostapediaal gewricht met het ontbreken van een otosclerotische focus. De intraoperatieve bevindingen werden ingedeeld volgens de Cremers classificatie voor congenitale middenooranomaliën. De resultaten van pre- en postoperatieve audiometrie werden geanalyseerd volgens de 1995 richtlijnen van het AAO-HNS Committe on Hearing and Equilibrium. De gemiddelden van de preoperatieve toonaudiometrie (AC), de beengeleiding (BC) en de luchtbeengeleidingscomponent (ABG) bij 0.5, 1, 2 en 3 kHz werden vergeleken met dezelfde postoperatieve metingen.

De postoperatieve beengeleidingswaarden werden gebruikt om de postoperatieve luchtbeengeleidingscomponent te berekenen. De uitkomsten van de post-operatieve luchtgeleidingsdrempels toonden een verbetering van de gemiddelde preoperatieve luchtgeleidingsdrempel van 56 dB (SD16.9) naar 34 dB (SD12) postoperatief. De preoperatieve en de postoperatieve beengeleidingsdrempels bleven onveranderd met respectievelijk 24 dB (SD 11.7) en 23 dB (SD 12). De gemiddelde postoperatieve 50% spraakverstaanbaarheidswaarde was pre-operatief 53 dB (SD 14) en postoperatief 31 dB (SD 15). Een postoperatieve luchtbeengeleidingscomponent kleiner dan 10 dB werd bij 58% van de 41 congenitale middenoren bereikt, terwijl dit percentage 82% was bij de primaire otosclerose operaties. Over het geheel genomen werden de beste resultaten bereikt bij de klasse 1 malformaties met alleen een geisoleerde stapes ankylose. De gemiddelde postoperatieve luchtgeleidingsdrempels, evenals de lucht-geleidingsdrempels voor de hogere frequenties, de beengeleidingsdrempels en voor het spraakverstaan waren beter voor de klasse 1 anomalieën in vergelijking met klasse 2 en klasse 4 anomalieën . Gezien de geringe aantallen in elk van deze groepen is er geen statistische significantie. Gepubliceerde series met een congenitale stapesankylose met beschrijving van de resultaten van stapes-vervangende chirurgie zijn zeldzaam, zodat deze bijdrage toch een welkome aanvulling op de huidige literatuur is.

In **Hoofdstuk 4.1** wordt de gemodificeerde techniek van de stijgbeugel-vervangende chirurgie met behulp van de laser beschreven. Immers bij deze techniek wordt de volgorde van de verschillende chirurgische stappen aangepast. Dit hoofdstuk is ingericht volgens de “How I do it” schrijfstijl en het correspondeert met de 2 hoofdstukken in het Hoofdstuk 1 Inleiding over “de geschiedenis van Otosclerose” en “de ontwikkleing van de stijgbeugelvervangende chirurgie”. Een stap voor stap beschrijving van deze operatietechniek is geillusteerd met intra-operatieve foto’s van de belangrijkste momenten in de operatie om de lezers te helpen bij het begrijpen van de beschreven volgorde in de verandering van deze techniek. Het is onze ervaring dat deze nieuwe techniek een ruimere toegang tot de voetplaat verschaft ten gunste van een optimale plaatsing van de stapedotomie opening in de voetplaat, terwijl de stabiliteit van de voetplaat en de incus gewaarborgd is. Een aanpassing van de toegepaste stijgbeugelvervangende prothese vergemakkelijkt de uitvoering van de operatie zonder nadelen voor de te bereiken gehoorverbetering. Stijgbeugelvervangende chirurgie met een laser om de stapedotomie opening te maken maakt de kans op binnenoorschade nog kleiner en is gunstig voor de gehoordrempel in de hoge tonen. De hier beschreven chirurgische techniek kan ook van nut zijn bij het opleiden van anderen in de techniek van stapedotomie, omdat de operatie hiermee weliswaar in een aantal moeilijke maar dan toch zeer gecontroleerde stappen ingedeeld wordt. Gebaseerd op de hier gepresenteerde resultaten van toch een zeer grote operatieserie zijn wij tot de conclusie gekomen dat deze gemodificeerde stapedotomie techniek een goede en veilige techniek is om een goede en effectieve verbetering van de gehoordrempel te bewerkstelligen. In ervaren handen leidt dit tot een zeer laag percentage operaties met binnenoorschade. Omdat een grote ervaring van de oorchirurg met stijgbeugelvervangende chirurgie leidt tot een grotere oorchirurgische vaardigheid, met als uitkomst minder oorchirurgische complicaties en betere resultaten voor het gehoor, wordt er hier voor gepleit om nog meer dan voorheen deze operaties te gaan concentreren in centra met een groot aantal van dergelijke operaties om aldus voor de patiënten de allerbeste resultaten te kunnen gaan bereiken.

Het op een passende en goede manier analyseren van de resultaten van stijgbeugelvervangende chirurgie is een complexe aangelegenheid. Aan die complexiteit wordt bijgedragen door de vele audiometrische bevindingen in het geopereerde en het andere oor, door een mogelijke progressie van het gehoorverlies in de tijd en nog andere niet audiometrische variabelen.

Toekomstige studies naar de waarde van stijgbeugelvervangende operaties zullen zich niet beperken tot de technische toonaudiometrische resultaten, maar zich meer en meer gaan richten op de verbetering van geluidswaarneming en het spraakverstaan in stille en geconditioneerde rumoerige situaties. Het meten van de verbetering van de kwaliteit van leven wordt een ander item. Dergelijke uitkomsten kunnen in de toekomst invloed hebben op de preoperatieve selectie van de al of niet te opereren oren.

Acknowledgements

The success and completion of this project became possible owing to contribution of incredible individuals and I extend sincere thanks to all of them with special thanks to:

MY RESEARCH SUPERVISOR Prof. dr. Cor Cremers for providing me with your expertise and guiding me through this process, I am deeply grateful for your unlimited support.

MY CLINICAL SUPERVISOR Dr. Jerry Halik for your tireless coaching and unlimited trust, for enabling my access to data that became the foundation of this thesis.

MY CLINICAL SUPERVISOR Prof. John Rutka for providing me with an opportunity to initiate this project and dedicating limitless time and generous resources that made possible this accomplishment.

MY COLLEAGUE AND CO-AUTHOR Dr. Sunil Dutt for your creativity and encouragement that set me up to succeed.

MY COLLEAGUE AND CO-AUTHOR Dr. Neil Bailie for your enlightening suggestions and meticulous assistance that refined and shaped this work.

MY FAMILY: My wife Mila for indefinite support, patience and love that allowed me to undertake this challenge. My sons Eli and Valery for joy and happiness that inspired this work and my Mom Svetlana for prayers that kept me going.

Curriculum Vitae Vitaly Kisilevsky MD

802-5418 Yonge St, Toronto, Ontario, Canada M2N 6X4
+1(416) 224-1636/ +1(416)519-3217, vitalykis@yahoo.com

2009	Sabbatical: May 4th- Medical Council of Canada Qualifying Examination Part I (Passed) October 29th- Medical Council of Canada Qualifying Examination Part II (Registered) TBA-PhD Thesis defense, The Radboud University Nijmegen Medical Centre
2008-06	Clinical Fellow in Otology/Neurotology at the Department of Otolaryngology, Head & Neck Surgery, University Health Network, University of Toronto
2005-04	Staff Otolaryngologist, Haemek Medical Center, Afula Expert Specialist, Israeli National Insurance Agency, Jerusalem; Workers Compensation board, Nazareth, Tiberias
2003-02	Research Fellow in Otology/Neurotology at the Department of Otolaryngology, Head & Neck Surgery, Toronto General Hospital, University of Toronto
2001-1999	ENT Specialist and Surgeon, Haemek Medical Center, Afula
1999	Board qualified in Otolaryngology- Head & Neck Surgery, Ministry of Health, Israel
1998- 92	Resident in Otolaryngology, Head & Neck Surgery, Haemek Medical Center, affiliated to the Rappoport Faculty of Medicine, Technion, Haifa
1990-86	Board qualified in Otolaryngology, Leningrad, Russia
1986-85	Resident in Otolaryngology, Clinical Hospital of Leningrad Medical University of Pediatrics, Leningrad, Russia
1985	Graduated with MD Degree from the Medical University of Pediatrics, Leningrad, Russia

List of abbreviations

ABG	Air-Bone Gap
AC	Air Conduction
AHEPs	Amsterdam Hearing Evaluation Plots
AAO-HNS	American Academy of Otolaryngology, Head & Neck Surgery
BC	Bone Conduction
BAHI	Bilateral Asymmetric Hearing Impairment
BSHI	Bilateral Symmetric Hearing Impairment
CHL	Conductive Hearing Loss
CT	Computerized tomography
dB	Decibel
GBP	Glasgow Benefit Plot
HL	Hearing Level
MHL	Mixed Hearing Loss
MMHL	Mild/Moderate Hearing Loss
PTA	Pure Tone Average
SD	Standard Deviation
SDS	Speech Discrimination Score
SNHL	Sensory Neural Hearing Loss
SPHL	Severe/Profound Hearing Loss
SRT	Speech Reception Threshold
UHI	Unilateral Hearing Impairment

Sponsors:
Nationale Hoorstichting / Sponsor Bingo Loterij
Ned. Ver. KNO-Heelkunde en Heelkunde van het Hoofd Hals gebied.