Outcomes of Middle Ear Surgery

Congenital minor ear anomalies and other challenging situations

Robert Vincent

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Outcomes of Middle Ear Surgery

Congenital minor ear anomalies and other challenging situations

ter verkrijging van de graad doctor aan de Universiteit Utrecht,
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PROMOTORES

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Chapter 1

Introduction

R. Vincent en C.W.R.J. Cremers

CONGENITAL ANOMALIES

Congenital anomalies of the middle ear and ear canal are subdivided into major anomalies and minor anomalies

In the major anomalies there is always a bony ear canal atresia present. In addition, there may be a malformation of the auricle. Anomalies of the ossicular chain are commonly seen in congenital ear canal atresia.

Congenital major ear anomalies

Altmann (ref. 1955, F. Altmann) was the first to propose an anatomical classification of ear anomalies. He divided them into 3 subclasses I-III according to the severity of the atresia. Later, Nager (ref. Nager GT, 1971) and Schuknecht (ref. Schuknecht HF, 1975) considered Altmann's classification helpful especially from the surgical point of view. Cremers et al. (ref. Cremers C. et al, 1988) modified the Altmann classification by splitting class II into class IIA and IIB. He showed that by applying the subclassification for class IIA the postoperative remaining airbone gap was less than 20 dB in 80% of all these operated ears. For the other higher subgroups, the hearing results remained less positive. The need for better treatment options therefore remained in the following years.

Over time, passive and active implantable bone conduction devices have become available. These are surgically applied, either percutaneously or transcutaneously. These devices have in recent years gained in effective output power widening the spectrum for their use. These bone conducting devices still require surgical intervention and are, therefore, not the method of choice in all our patient groups, such as the very young children.

Non-surgical applications have become available, of which the SoftbandTM Bone Conducting Device solution (ref. Verhagen et al, 2008) and more recently the adhesive solution (ref. Dahm et al, 2018; ref. R. Vincent et al., 2019) (AD HEAR) have proven to be effective. One of the major advantages of the last options above is the possibility to wear them for very early hearing support in the first months after birth. The importance of early rehabilitation of the hearing loss has been established in numerous publications, where it has been recognized that auditory stimulation is very important for the hearing maturation in unilaterally or bilaterally affected cases.

The surgical treatment options were only advisable for highly selected cases (Class I and IIa Altmann-Cremers classification). It is important to realize that these surgical interventions took place only after the age of about 8-10 years on average. The recent developments in hearing rehabilitation have considerably outdated the previous classical microsurgical treatment options for the major congenital ear anomalies.

Congenital minor ear anomalies

In the minor congenital ear anomalies, the congenital anomaly is restricted to the middle ear and doesn't involve the external auditory canal. A congenital partial bony plate still present in the tympanic membrane might be considered as the "embryological" demarcation between the major and the minor congenital ear anomalies.

In the rest of the first section of this thesis the focus will remain on minor ear anomalies and its various microsurgical treatment options. To place the published work from this thesis into perspective we feel that a short historic overview is in order.

Some historical notes on microsurgery of the middle ear and stapes surgery

In this PhD book various new and existing microsurgical techniques are described, however, for an informed understanding a historical perspective is helpful. In the 1950s micro-ear-surgery underwent an enormous development through the introduction of myringoplasty in 1952 by Zollner (ref. Zollner F, 1955) and by Wullstein (ref. Wullstein H, 1956) and by the introduction in 1958 of stapes replacement surgery by Shea (ref. Shea JJ, 1958) in patients with otosclerosis. In the era before the introduction of his new technique, fixation of the stapes in otosclerosis was treated by fenestration, predominantly of the horizontal semicircular canal or by re-mobilization of the stapes.

This fracturing of the otospongeotic plaque around the stapes footplate was performed with a tiny hammer and chisel. (ref. Tange RA, 2014) These techniques had both considerable risk of sensorineural hearing loss. In the latter case there was a high chance of refixation of the stapes due to the body's mechanism to repair bony fractures.

The history of stapedectomy (complete removal of the stapes) along with the development of different fenestration techniques (perforation of a part of the stapes footplate) is well known. Since John Shea's new technique was first described in the 1950s the introduction of various different surgical procedures for/of a stapedectomy have subsequently been reported. (Jahnke, 2004; Tange RA, 2014) Both the stapedectomy and the stapedotomy create an open access towards the very venerable inner ear structure to restore the sound transmission inward. As a response there has been much debate on whether to and if so how to seal the opening into the inner ear.

A vein graft employed in order to seal the opened oval window was part of the surgical technique as applied from the start of stapedectomy by John Shea (ref. Shea JJ, 1958) on May 1st, 1956. A Teflon stapes prosthesis was applied to replace the stapes.

Over time different materials were applied as alternatives for the Teflon material that Shea used. These include a steel-wire-adipose tissue prosthesis, a hollow polyethylene tube and, titanium or gold pistons. Most of these alternatives were shown to have disadvantages. In the 1980s full opening of the oval window (stapedectomy) was followed by sequentially smaller openings (stapedotomies).

The first change to a partial removal of the footplate (partial platinectomy) was then reduced into a smaller opening, stapedotomy. Refined microsurgical instrumentation finally included the adoption of laser technology which facilitated these new developments in more atraumatic surgery.

Stapedectomy and stapedotomy are almost exclusively performed to resolve the conductive (component of the) hearing impairment, resulting from a stapes footplate fixation. Sometimes there exist other deficits in the mobility of the ossicular chain exist, i.e. affecting the malleus and/or the incus. In the regular stapedotomy procedure, the piston is attached to the incus (incudo-stapedotomy), while in case the stapes replacing piston has to be attached to the malleus this is -named a malleostapedotomy. From the early start of stapes replacing surgery malleostapedotomy procedures have been performed and have been refined (Van Rompaey et al, 2011; Burggraaf et al, 2018).

Congenital minor ear anomalies

Mirko Tos's definition of congenital minor ear anomalies was: "The minor congenital malformations involve fixations and defects of the ossicular chain and malformations of the oval and round window regions with a normal eardrum and ear canal". (Ref. Tos M, 2000). Tos provided in this text a review and a survey of pathology and surgery of the various forms of minor congenital malformations as published in the first three decades after the introduction of tympanoplasty and stapes surgery (ref. Tos M, 2000).

Teunissen (ref. Teunissen E, 1992) stated earlier in the chapter Introduction of his PhD thesis on major and minor congenital ear anomalies that although after the introduction of stapes replacement surgery in 1958 in general this stapes replacement surgery found general application for the treatment of otosclerosis in the early part of 1960, it's subsequent application for the treatment of congenital stapes fixation took much longer.

In the 1960s many surgeons still preferred to perform fenestration and stapes mobilization, as had been performed previously, for congenital stapes ankylosis. (Teunissen, 1992; Shambaugh, 1952; Ombredanne, 1959; House 1958; Hough; 1958). Tange (ref. Tange, 2014) provided in his table F an overview of the literature for outcomes and applied type of fenestration surgery for otosclerosis. This started in 1935 and stopped around 1958. Barbara Hepworth gave a spectacular artists' impression of that era in the otology publication (ref. Booth J.B., 2000). An impressive quite early surgical series for congenital minor ear anomalies was published in a series of publications by Ombredanne from Paris between 1959-1968. (ref. Ombredanne 1959; 1960; 1962a; 1962b; 1964; 1966a; 1966b; 1968).

Classifications

A series of authors have contributed to the field of surgical findings and outcomes in the field of minor congenital ear anomalies as mainly listed by H. Thomeer (ref. Thomeer H, 2012) in his PhD thesis on congenital minor ear anomalies. In total 10 different classifications have been proposed for congenital minor ear anomalies (ref. Thomeer H, 2012). Named Henner classification 1956 (ref. Henner 1956), Ombredanne classification 1964 (ref. Ombredanne, 1964), Arslan and Giacomelli classification 1963 (ref. Arslan & Giacomelli, 1963), House classification 1969 (ref. House, 1969), Funusaka classification 1979 (ref. Funusaka, 1979), the Cremers classification 1993 (ref. Teunissen & Cremers C, 1993), the Charachon classification 1994 (ref. Charachon et al, 1994), the Park classification 2009 (ref. Park & Choung, 2009).

Tos (ref. Tos, 2000) used the Cremers classification when he republished this as is presented in his table 1. The Cremers subclassification for congenital minor ear anomalies was additionally split up by the Hear classification committee in 1998, as it was too by Tos (ref. Tos, 2000). (Table 2) The main classification in 4 classes was kept intact.

In this PhD thesis 3 original papers, published in 2016 (ref. Vincent et al., Otol Neurotol 2016;37; Vincent et al Laryngoscope 2016 pp 682- 688; Vincent et al. Laryngoscopy 2016 pp. 2552-25580), are included for class 1 & 2, class 3 and class 4.

A systematic analysis of the type, incidence and surgical outcome of anatomical anomalies according to the different classes of a generally accepted classification is helpful. These data inform the surgeon about how frequently any type of a stapes ankylosis (class 1 and 2) might be expected and about the probability that stapes replacing surgery might produce hearing improvement.

The preoperative counselling necessitates discussion of such data, including peri- and postoperative risks of permanent inner ear damage resulting in hearing loss and balance problems. The frequency with which a class 4 anomaly might be expected is helpful to discuss, guiding the surgeon to weigh the pros and cons of performing or declining surgery, based on the chances for hearing improvement.

It is relevant to have data available about the rate of success and about the variability and long-term success of such surgical outcomes.

Table 1. The Cremers classification of minor CONGENITAL anomalies of the middle ear and their relative surgical incidence in 144 ears operated on in the Nijmegen ENT clinic during the period from 1964-1990. (With the courtesy of E. Teunissen & C.W.R.J. Cremers, Annals of Otology Rhinology and Laryngology 1993;102:606-612 and with the courtesy of M. Tos. Surgical solutions for conductive hearing loss. Thieme, Stuttgart 2000).

Class	Main anomaly	Subclassification	%/144 ears
1	Isolated congenital stapes ankylosis (or fixation)		30.6
2	Stapes ankylosis associated with another congenital ossicular chain anomaly		38.2
3	Congenital anomaly of the ossicular chain but mobile stapes footplate	a. Discontinuity of the ossicular chain b. Epitympanic fixation	15.3 6.3
4	Congenital aplasia or severe dysplasia of the oval or round window	Aplasia Dysplasia Crossing (prolapsed facial nerve Persistent stapedial artery	6.9 2.1 0.7

Table 2. Air-bone gap 12 months after reconstructive surgery in 127 minor congenital ear anomalies. (With the courtesy of E. Teunissen & C.W.R.J. Cremers, Annals of Otology Rhinology and Laryngology 1993;102:606-612 and with the courtesy of M. Tos. Surgical solutions for conductive hearing loss. Thieme, Stuttgart 2000).

	No. of ears		Air-bone gap	Air-bone gap	Air-bone gap
Classification	per class	reconstructive surgery	< 10 dB (%)	10-20 dB (%)	>20 dB (%)
Class I	44	41	20 (49%)	9 (22%)	12 (29%)
Class II	55	51	20 (39%)	17 (33%)	14 (28%
Class III	31	31	9 (29%)	14 (45%)	8 (26%)
Class IV	14	4	1 (25%)	2 (50%)	1 (25%)

In the same way it is necessary to have data detailing the complications and mishaps in the outcome of such elective and rare surgical procedures.

To collect clinical data on the outcome of specific, usually genetic, syndromes causing a conductive congenital hearing loss is helpful for preoperative evaluation and patient counselling. In otology the end goal should be to be in a position to advise and treat the patient in a personalised manner, based on one's own actual surgical results and surgical capabilities. This personalised care approach, for the greater benefit of the individual patient, has become common practice in other specialties and is a necessary but challenging direction in otological practice. From this viewpoint it is considered highly relevant to contribute in this PhD thesis 3 original publications on the outcome of elective surgery for congenital minor ear anomalies.

The outcome of hearing improvement after exploratory tympanotomy in a series of 144 ears operated from 1986 to 1990 is shown with preoperative and postoperative data for each ear separately for classes 1, 2, 3 and 4.

Figures 1, 2, 3 and 4 (ref. Teunissen & Cremers, Annals of Otology Rhinology Laryngology 1993;102:606-612). The percentages of closure of the airbone gap < 10 dB, 10-20 dB and > 20 dB are shown for this series (ref. Teunissen & Cremers, 1993). (Figure 2). No temporary or permanent facial palsy had been noted postoperatively. No inner ear deafness has occurred as result of this surgery.

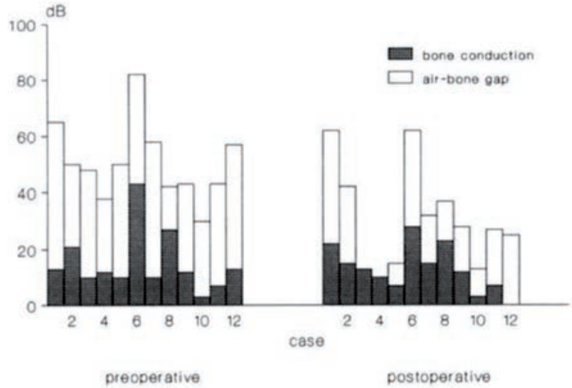

Figure 1. Preoperative and 1-year-postoperative individual hearing levels for bone and air conduction in 12 class I ears operated on between 1986 and 1990. (With the courtesy of E. Teunissen & C.W.R.J. Cremers, Annals of Otology, Rhinology and Laryngology 1993;102:606-612)

The outcome of exploratory tympanotomies of an additional consecutive Nijmegen series of 106 congenital minor ear anomalies from 1986-2001 and classified according to the Cremers classification (Teunissen & Cremers 1993) has been published additionally and separately for each of those 4 classes, and individually, applying the Amsterdam hearing plot. (Thomeer, 2012 PhD thesis). (ref. Thomeer et al, 2010; Thomeer et al, 2011; Thomeer at al 2012; Thomeer et al 2012).

To date the surgical outcome of a consecutive series of 250 ears with congenital minor ear anomalies has been documented and published.

Kisilevski et al have also reported on the outcome of ear anomalies following the Cremers classification (Kisilevski et al, 2009) (ref. Teunissen & Cremers, Ann Otol 1993).

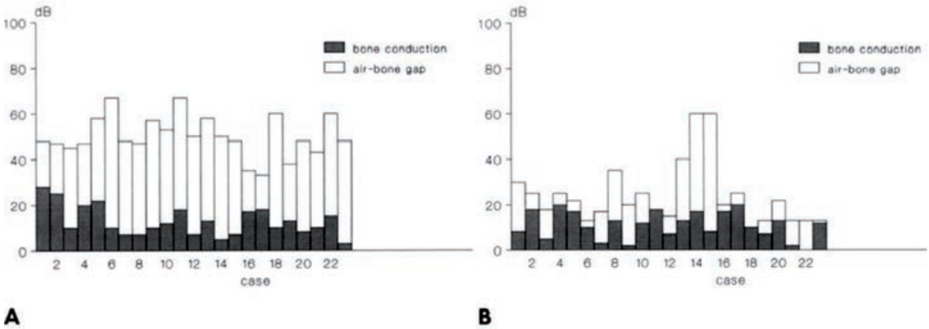

Figure 2. Individual hearing levels for air and bone conduction in 23 class 2 ears operated on between 1986 and 1990. A) Preoperative B) One year postoperative. (With the courtesy of E. Teunissen & C.W.R.J. Cremers, Annals of Otology, Rhinology and Laryngology 1993;102:606-612)

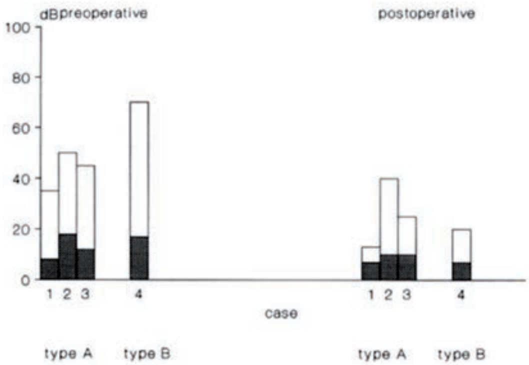

Figure 3. Preoperative and 1-year-postoperative individual hearing levels for bone air conduction in 4 class 3 ears operated on between 1986 and 1990. (With the courtesy of E. Teunissen & C.W.R.J. Cremers, Annals of Otology, Rhinology and Laryngology 1993;102:606-612)

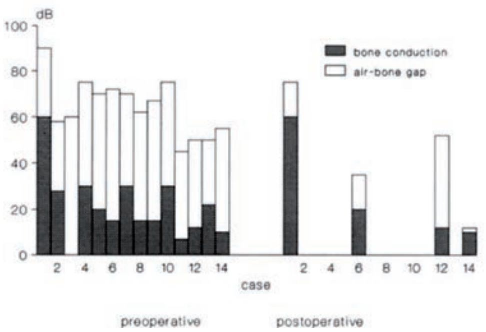

Figure 4. Preoperative and 1-year-postoperative individual hearing levels for bone and air conduction in 14 class 4 ears, including 4 ears with neo-oval window, operated on between 1964 and 1990. (With the courtesy of E. Teunissen & C.W.R.J. Cremers, Annals of Otology, Rhinology and Laryngology 1993;102:606-612)

Table 3. Proposal for future classification of minor congenital anomalies of the middle ear based on Cremers classification, modified and supplemented by Tos and the HEAR subcommittee. (With the courtesy of M. Tos, Surgical solutions for conductive hearing loss, Thieme Stuttgart 2000)

Class	Main anomaly	Subclassification
1	Isolated congenital stapes ankylosis (or fixation)	a) Footplate fixation 1. Normal stapedial arch 2. Monopodial stapedial arch 3. Monocrural stapedial arch Dysplastic arch b) Stapes supra structure fixation 1. Elongation of the pyramidal fixation 2. Stapes-pyramidal process bony bar 3. Stapes facial canal bony bar 4. More than one bone bar Dysplastic arch
2	Stapes ankylosis associated with another congenital ossicular chain anomaly	a) Discontinuity of the chain b) Epitympanic fixation c) Tympanic fixation 1. of the malleus handle (small atretic plate) 2. of the long process of the incus
3	Congenital anomaly of the ossicular chain but mobile stapes footplate	a) Discontinuity of the ossicular chain b) Epitympanic fixation c) Tympanic fixation 1. of the malleus handle 2. of the long process of the incus
4	Congenital aplasia or severe dysplasia of the oval or round window	Aplasia Dysplasia Crossing (prolapsed) facial nerve Persistent stapedial artery

* The stapedial tendon and pyramidal process may sometimes be missing in addition to other stapes deformities.

The need for an early bilateral hearing therapy

Patients with a unilateral or bilateral congenital minor ear anomaly present with a conductive or even mixed hearing impairment and are in need of hearing rehabilitation.

Several options to improve hearing are available, including conventional air conduction hearing amplification and osseointegrated partially implantable passive or active bone conduction devices with a percutaneous or transcutaneous application. Non-surgical alternatives for such bone conduction devices are the SoftbandTM application (ref. Verhagen et al., 2008) and the adhesive plaster (ADHEAR) (ref. Gawliczek et al., 2018).

Elective surgery with an exploratory tympanotomy is possible from about the age of 10 years or later, although Sterkers & Sterkers reported operating at an age from 4 to 8 years (ref. Sterkers & Sterkers 1988). Starting the needed hearing therapy with suitable hearing devices as early as possible will help to establish bilateral auditory stimulation and central auditory maturation in a timely manner.

Impact of syndromal diagnosis on the severity and type of the congenital minor ear anomaly

Part of the preoperative evaluation of surgery for congenital minor ear anomalies is to evaluate whether a specific genetic syndromal disease is involved (ref. Cremers & Teunissen, 1991 Int J Ped ORL). Consultation by a clinical geneticist might be helpful to establish a syndromal diagnosis. Some 70 or even more syndromal genetic diseases with conductive or mixed deafness as a main or relevant feature have been reported.

Although it is most valuable for the practicing otosurgeon to be aware of the otological information available in the literature (Tewfik & der Kaloustian, 1997; Toriello & Smith, 2013), only a limited number of publications on these syndromes (Table (ref. Thomeer PhD, 2012; Table 4) can be considered to be quite informative for the ear surgeon.

Table 4. Syndromal diagnosis in series of congenital ossicular chain anomalies. (With the courtesy of H. Thomeer, Congenital minor ear anomalies, PhD Thesis, Radboud University, Nijmegen 2012)

Syndrome	Subtype
Stapes gusher	Stapes gusher
Branchiogenic	Branchio Oto Renal Syndrome Treacher Collins Syndrome Preauricular sinus, external ear anomaly Hemifacial microsomia
Craniosynostosis	Crouzon Apert Saethre Chotzen Pfeiffer
Skeletal	Klippel Feil Wildervanck Frontometaphyseal Dysplasia Teunissen Cremers Syndrome Maxillofacial Dysostosis
Miscellaneous	Noonan Toriello Turner Lacrimoauriculodentodigital

The textbooks scarcely provide relevant oto-surgical information to guide the ear surgeon adequately. A targeted literature search might be needed for any specific oto-genetic syndrome involved.

We like to name only a few of these syndromes to help raising the awareness of the relevant and guiding information given by a timely syndromal diagnosis. Thomeer (2013) listed those already in his PhD thesis. (ref. PhD thesis Thomeer)

Stapes Gusher Syndrome

For the details of this syndrome we provide the full description as already published by Thomeer in his PhD thesis (Thomeer 2013). This syndrome shows specific inner ear anomalies at CT-scanning. Quite remarkable audiological features might also be present.

A rare complication of stapedotomy during exploratory tympanotomy is a gusher. This is an excess of perilymphatic and cerebrospinal fluid (CSF) that discharges from the stapedotomy opening, filling the middle ear and exuding through the external auditory meatus. It appears more frequently in the X-recessive hereditary syndrome with progressive sensorineural hearing loss (DFN3) (ref Cremers CW et al, 2002; Cremers CWR, 2007). The causative mutation lies in the *POU3F4* gene, which encodes a POU domain transcription factor (class 3, factor 4) (Cremers F et al, 2000; Schild C et al, 2011). The sensorineural component has been shown to be progressive, although a conductive component to the hearing impairment may also be relevant. The smaller (< 20 dB) conductive component in pure tone audiometry (PTA) may be due to a pseudo-conductive hearing component resulting from the inner ear anomaly and the so-called "third window phenomenon".(ref. Rosowski JJ et al, 2004) As a result of the X-recessive pattern of inheritance, most cases occur in male patients, who develop a progressive mixed hearing loss in childhood.

The widened access for CSF to the inner ear is considered to be the cause of the progressive sensorineural hearing loss. It is important to try to prevent the risk of encountering a gusher before embarking upon stapes surgery. This can be accomplished by performing a preoperative CT scan. This informs the surgeon about the anatomy of the inner ear and confirms the suspicion of a stapes gusher. This lateral widening of the internal acoustic canal is visible on CT scans. (Ref. Phelps PD et al, 1991; Talbot JM & Wilson DF 1991).

Treacher Collins Syndrome

The specific otosurgical guiding information as presented earlier by Thomeer (2013, PhD thesis) is repeated and extended here.

Overall, anomalies in the ossicular chain vary. However, a malformed stapes head with curved crura and a protruding facial nerve are often encountered, along with a dysplastic long process of the incus (ref.Marres HA, 2002; Marres HA et al, 1995; Marres HA et al., 1995). Moreover, the oval window can be partially covered by the facial nerve following an abnormal course. The characteristic curved crura or monopodial stapes may be in close contact with the facial nerve, interfering with good postoperative hearing outcomes. In general, hearing outcomes are moderate to poor due to the above described anomalous ossicular chain.

The unusual anatomical situation which can be encountered, typical for Treacher Collins Syndrome, is a markedly smaller than usual anteriorly placed and curved bony external acoustic canal, a protruding facial nerve in the external genu, and usually a much too short long process of the incus. A small curved but mobile stapes and a small but mobile handle of the malleus, might possibly favour performing a type of malleostapedotomy assisted by laser surgery. Marquet et al (1988) published personal drawings in their figure 33A and 33B to show the specific abnormal anatomy of the stapes and around the stapes in relation to the facial nerve in Treacher Collins Syndrome (Figure 5; Marquet et al, 1988).

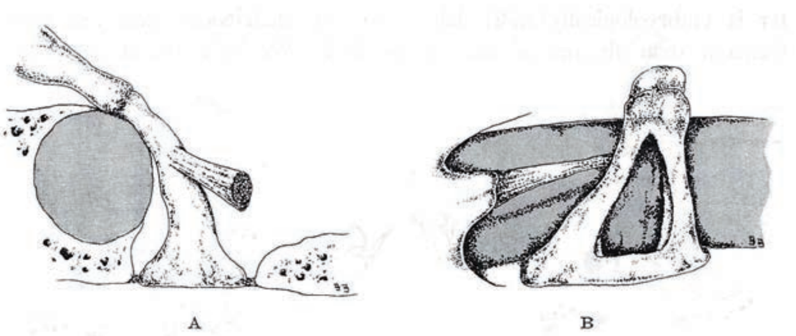

Figure 5. Specific abnormal anatomy of stapes and facial nerve in Treacher Collins Syndrome (With the courtesy of Marquet et al, Acta Oto-Rhono-Laryngologica Belgica 1988;42/2:123-302

Branchio-Oto-Renal (BOR-) Syndrome

The range of hearing impairment in the BOR-syndrome with an autosomal dominant pattern of inheritance may vary from almost none to a severe/profound mixed hearing impairment. (Cremers & Fikkers van Noord, 1980). The branchial arch syndrome shows anomalies at the level of the auricle, the external acoustic canal, the middle ear and the inner ear. Some hypoplasia of the cochlea is a regular finding at high resolution CT-scanning. (Kemperman et al., 2002) A widened vestibular aqueduct is not an unusual finding. As in Treacher Collins syndrome, another branchiogenic syndrome, the anomalies found in the middle ear may vary widely, including a range of variation in the dimensions of the middle ear. This can have a negative influence on the outcomes for hearing after an exploratory tympanotomy. (ref. Cremers C et al, 1981 ref. 43 Thomeer; ref. Cremers CW et al. 1993 (ref 45 Thomeer). The sensorineural component as part of the mixed hearing impairment may in time fluctuate or progress. (Kemperman et al, 2001; Stinckens et al, 2001)

The renal involvement in this syndrome can be severe. Bilateral agenesis of the kidney has rarely been reported. (ref. Widdershoven et al, 1983)

Several genes like EyA1, Six1 and Six5 are involved in the etiology of the BOR- syndrome. Mutation analysis could be included as part of a diagnostic clinical genetic work up.

Craniosysostosis syndromes

Congenital stapes ankylosis has been reported as a feature of these syndromes. Coincidentallyl a bony epitympanic occlusion of the malleus and incus could occur. There are only a few surgical reports available in the literature. (Ensink et al, 1996; Cremers, 1981 Int J Ped ORL).

The Klippel Feil Anomaly

The Klippel Feil anomaly is a condition characterized by shortness of the neck resulting from reduction in the number of cervical vertebrae or the fusion of multiple hemivertebrae into one osseous mass. The hairline is low, and motion of the neck is limited. This anomaly can be seen on its own but might also be a feature of a well delineated clinical syndrome.

A stapes gusher is reported to have occurred once during a stapedectomy procedure. It was the result of a too wide bony communication between the lateral part of the internal acoustic canal and the inner ear. High-resolution CT-scanning performed preoperatively will document such a specific inner ear anomaly in time.

Outcomes of exploratory tympanotomies for hearing have been reported (Cremers et al, 1984; van Rijn & Cremers, 1988; Strübbe et al, 1994; Strübbe et al, 1995).

Osteogenesis Imperfecta

Osteogenesis Imperfecta (OI) is a heritable connective tissue disorder predominantly characterized by bone fragility, blue sclera and hearing loss. Type I is the most common type of this syndrome and is inherited as an autosomal dominant trait. Up until the 1970s, the middle ear changes in Osteogenesis Imperfecta were considered to be a severe form of otosclerosis. (ref. Tos M, 2000). Osteogenesis Imperfecta is caused by a mutation in the gene *Col1A1* or the gene *Col1A2*. These genes encode elements of type 1 collagen, an important constituent of bone, skin, ligaments, tendons and blood vessels. The disease is associated with large phenotypic and genotypic variability. The fracture rate may vary from a few fractures occurring during puberty to hundreds of fractures over a lifetime. More than 1.000 different mutations causing OI have been identified (ref. Swinnen, 2012).

Hearing loss has been reported to manifest in about half of the OI population. The type of OI-related hearing loss may be conductive, sensorineural or mixed. The hearing loss characteristics such as the type of hearing loss, onset, progression, severity and symmetry show large variations among studies and among OI subjects (ref. Swinnen, 2012). The audiological phenotype lacks correlation with the genotype (ref. Hartikka et al, 2004; Stinckens et al, 2011 Orphanet etc.) Ocular manifestations in OI have been studied by phenotype-genotype analysis in mainly the same Dutch and Belgian OI families. For some ocular 'manifestations' phenotype-genotype correlations have been shown. One of the conclusions arising from this study is that blue sclera is not an obligatory feature for the diagnosis OI. (ref. Swinnen et al, 2019 Submitted).

Conductive hearing loss usually arises during the second to the fourth decade of life, and thereafter evolves to a mixed hearing loss. The underlying pathology is an abnormal bone remodeling process which also affects the oval window region within the temporal bone. The hearing loss severity and type correspond to the extent of the hypodense areas in the temporal bone visualized by computed tomography.

Schuknecht (1993 2nd edition) names the characteristic histologic temporal bone findings in Osteogenesis Imperfecta and the problematic situation seen on histological findings in the disease Otosclerosis and Osteogenesis Imperfecta. The credits have been given to Nager (ref 1988) and others (Zajtchuk & Lindsay, 1975; Brosnan et al, 1977; Sando et al, 1981), who clearly demonstrated the histologic independence of these two disorders. Georg Nager (1993) published the book "Pathology of the Ear and Temporal Bone" and extended the histologic evidence and clinical genetic views. Based on this evidence, Osteogenesis Imperfecta and Otosclerosis are considered to be two different diseases.

In spite of a higher risk of intraoperative complications and the continuing progression of the sensorineural hearing loss component, stapes surgery may still be considered as a valuable method of managing conductive and mixed hearing loss in Osteogenesis Imperfecta.

See also Tos (ref. Tos, 2000) in chapter 21 on conductive hearing loss due to Osteogenesis Imperfecta in his book on surgical solutions.

The outcomes of stapes surgery in Osteogenesis Imperfecta

Larger series on the outcome for stapes surgery were published by Shea and Postma in 1982 (ref. 1982), Pedersen in 1985 (ref. 1985) (ref. Cremers, Garretsen, van de Rijt & Swinnen in 1985, 1989, 1990, 1991, 2003, 2009, 2012), (ref. Cremers, 1985; Cremers and Garretsen, 1989; Garretsen and Cremers in 1990; Garretsen & Cremers, 1991; van der Rijt & Cremers, 2003) Kuurila et al, 2004; Vincent et al, 2005 and Swinnen et al, Swinnen et al, 2009; Swinnen et al., 2012).

Other contributions were added by Armstrong in 1984 (ref. Armstrong 1984), Dieler et al in 1997 (ref. Dieler et al, 1997) and Albahnasawy et al in 2001 (ref. Albahnasawy et al, 2001). Garretsen analysed the short and long-term results of stapes surgery in Osteogenesis Imperfecta in the 3 largest early series by Shea and Postma (1982) (ref. Shea and Postma, 1982), Pedersen and Elbrond (1983) (ref. Pedersen & Elbrond, 1983) and Garretsen and Cremers (1991) (ref. Garretsen & Cremers, 1991) and concluded that the results are poorer than in otosclerosis. This proves to be the case in all publications analyzing small or large series of O.I. patients.

One of the recent larger surgical series on O.I. is published in 2014 from France and is included as an original contribution (ref. Vincent et al, 2014) in this PhD thesis

OTOSCLEROSIS

Outcomes of stapedotomy in children and outcomes of revision stapedotomy.

In this PhD manuscript two published original articles are included on these topics. One is about the outcomes of stapes surgery for otosclerosis in children in a series of 41 consecutive cases (Vincent et al, 2016). The second one is an analysis of a prospective series of 652 revision stapedotomies (Vincent et al, 2010).

Otosclerosis in adolescence

The diagnosis of otosclerotic conductive hearing loss is usually unproblematic. Typically, there is a progressive loss of hearing, which occurs in young adults. This can be bilateral and symmetrical or unilateral, with a positive family history in about 30% of cases. (ref. Jahnke, 2014) In rare cases (in 1.5 – 3.0%) the otosclerosis hearing loss appears before and during adolescence, although a first slight hearing loss had already been noted before the age of 20 years in 15% of patients whose otosclerosis was first diagnosed when they were adults. (ref. Romanet et al, 1990; Robinson, 1983; Lippy et al, 1998)

Even in large series of otosclerosis patients, surgery before age 11-18 is rare. In the Bern statistics it concerned 6 patients (1%). A marked obliterative otosclerosis was present in three cases (ref. Wengen, 1992).

Tos (2000) reminded that obliterative otosclerosis is the most important factor predisposing to sensorineural hearing loss during stapedectomy. Special surgical techniques are needed in case otosclerotic foci are obliterating the oval window. Applying the stapedotomy technique, the results have improved, but they are still poorer than in non-obliterative otosclerosis.

Regarding outcome and series size, rather exceptional data are part of this PhD thesis. They concern series of congenital stapes ankylosis, Osteogenesis Imperfecta and a quite exceptional series of 41 consecutive cases with a primary stapedotomy in children with otosclerosis. (Vincent et al., 2016 Laryngoscope pp. 442-446) These series are unprecedented in the literature.

STAPES REPLACEMENT TECHNIQUES AND SPECIFICALLY THE CAUSSE STAPEDOTOMY WITH VEIN INTERPOSITION.

In chapter 7-16 of his book "Surgical Solutions for Conductive Hearing Loss" Tos (ref., 2000 Hfst. 7-16) depicted detailed, well illustrated overviews on stapes surgery for otosclerosis and on the many aspects involved in surgical management. Closure of the vestibule after a total footplate removal, covering the oval window with a pressed vein graft, as performed by Shea and Portmann, is illustrated in his figure 244a-b (Figure 6). The later 0.8 mm stapedotomy technique by Causse is illustrated in his figure 248a (Figure 7) (ref. Causse & Causse, 1985). The vein to seal off the vestibule is usually taken from the back of the hand (ref. Shea, 1958; Portmann, 1959) as was already published by Shea in 1958 and by Portmann in 1979.

Many other detailed illustrations of how to seal off the oval window niche after stapes replacement surgery as part of other techniques has been given as a historic overview. The gradual development from the stapes mobilization technique towards the total stapedectomy, partial stapedectomy, and from the partial platinectomy towards the presently preferred stapedotomy is well illustrated and documented.

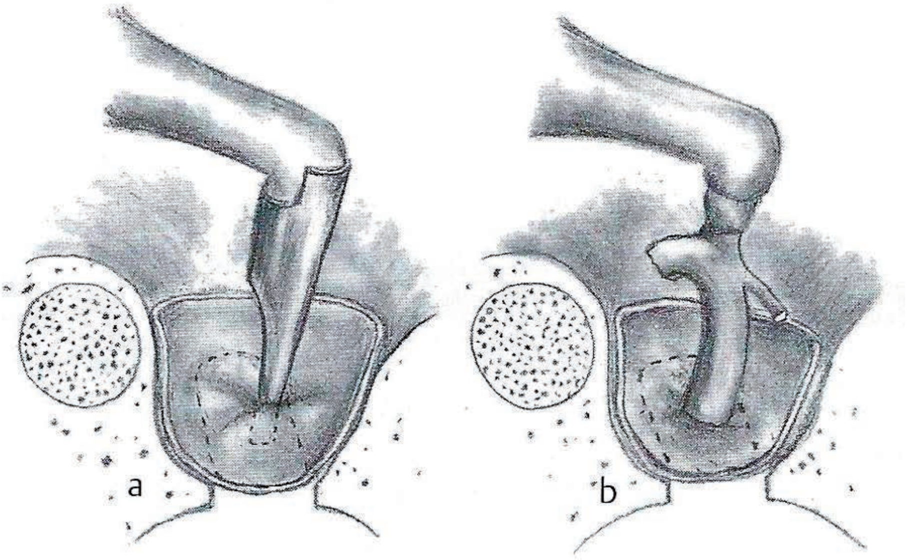

Figure 6. Various types of large fenestra stapedectomy and interposition technique: semi-schematic view into the posterior aspect of the oval window niche and the vestibule) a) Shea's vein-poly-ethylene-strut technique. b) Portmann's vein-posterior crus interposition. (With the courtesy of M. Tos, Surgical solutions for conductive hearing loss, Thieme Stuttgart 2000)

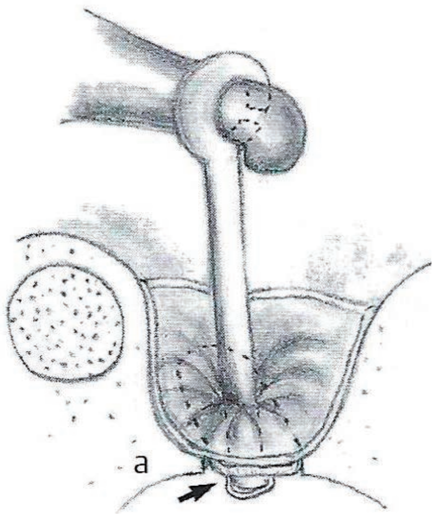

Figure 7. Stapedectomy technique. a) 0.8-mm stapedotomy and Causse's vein-Teflon interposition technique (arrow) (With the courtesy of M. Tos, Surgical solutions for conductive hearing loss, Thieme Stuttgart 2000)

CAUSSE'S SMALL FENESTRA STAPEDECTOMY

On page 108-110 Tos (ref Tos, 2000) has published a text with drawings when and how the Causse total stapedectomy technique with vein graft and Teflon interposition became refined into a small fenestra stapedotomy.

The figures 248a, 286 (Figure 8) and 287 (Figure 9) with their legends are reproduced in this text as illustrations (ref. Tos, 2000). This since the Causse stapes replacement surgical technique, with some later refinements, have been applied in the stapes surgery patients series presented in 5 of the first 6 original published manuscripts/ articles presented in this PhD thesis.

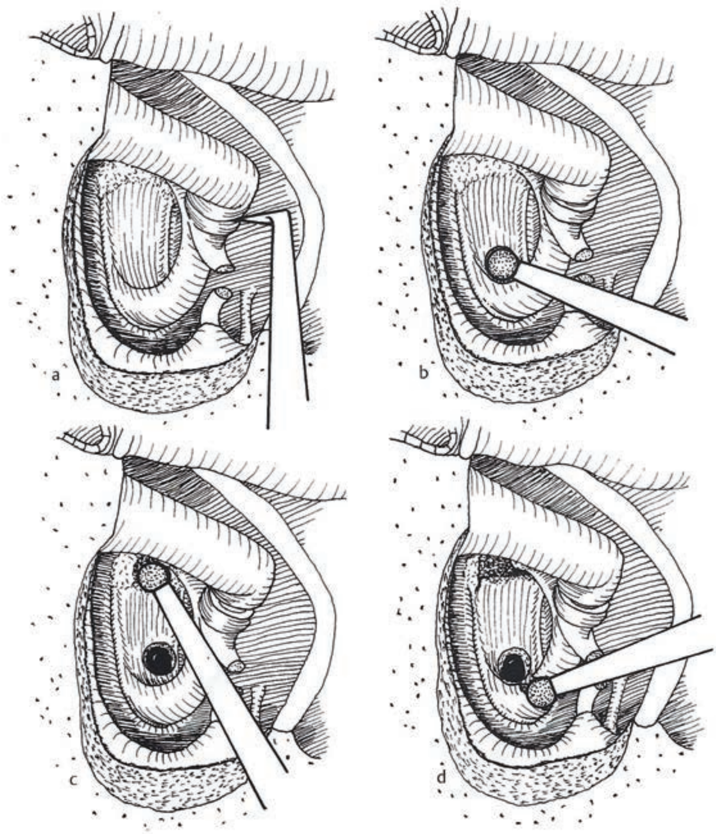

Figure 8. The Causse small fenestra stapedectomy and Teflon interposition technique. a) The stapes tendon is cut and the incudostapedial joint separated. b) A large safety hole is drilled in the posterior part of the footplate. The anterior crus (c) and the posterior crus (d) are separated from the footplate using a Treace-Causse microdrill. (With the courtesy of M. Tos, Surgical solutions for conductive hearing loss, Thieme Stuttgart 2000)

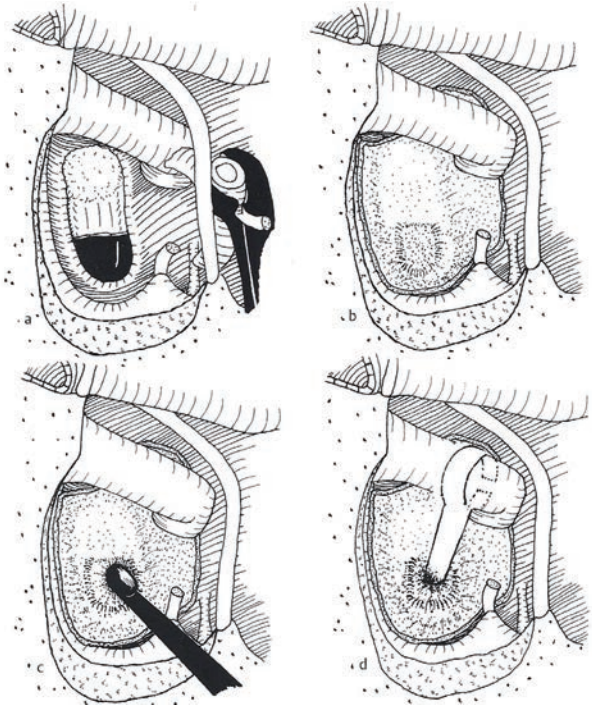

Figure 9. a) Using either a microdrill, picks, or hooks the posterior part of the footplate is removed, as are the crura. b) The vein is spread over the oval window niche. c) the vein is adapted to the oval window walls and carefully brought into contact with the oval window. d) A 0.8-mm Teflon piston is interposed between the vein and fixated to it. (With the courtesy of M. Tos, Surgical solutions for conductive hearing loss, Thieme Stuttgart 2000)

The combination of a 0.8 mm Teflon piston and vein graft interposition efficiently protects against a perilymph fistula and infection. The elasticity of the vein graft prevents damage to the membranous labyrinth by the Teflon piston (ref. Causse JR, 1960; 1961; 1964; 1965).

After relatively wide exposure of the stapes region, showing the facial nerve and the bony pyramidal eminence, the stapedial tendon was then cut, the mobility of the malleus handle and of the malleus/incus complex tested, and the incudo- stapedial joint disrupted (figure 8a). A hole was then drilled in the posterior part of the footplate (Figures 8b, 8c) and the crura drilled away using a microdrill (Figure 8d). The crura and the posterior part of the footplate are removed. The vein is spread over the oval window niche and a 0.8 mm all Teflon prosthesis interposed (Figure 9). (ref. Causse & Causse in 1985). In more recent times, with LASER assisted surgery to perform the 0.8 mm stapedotomy and with the application of a 0.4 mm all Teflon piston, this surgical technique became even more refined (Figure 10).

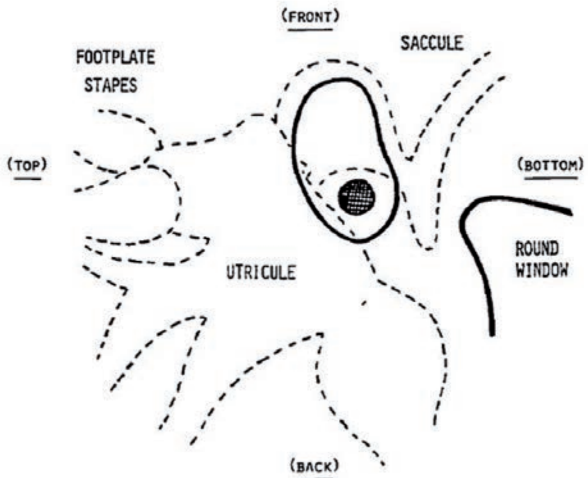

Figure 10. An 0.8 mm platinectomy in the posterior third of the footplate. (With the courtesy of M. Tos, Surgical solutions for conductive hearing loss, Thieme Stuttgart 2000)

Tos (ref. Tos, 2000 pp 126 etc.) spent additional pages 126-129 with text and illustrative surgical drawings (his figures 331, 332, 333, 334, 335, 336, 337- respectively figures 11, 12, 13, 14, 15, 16, 17) to present the "Jean Bernard Causse 0.8 mm stapedotomy with vein interposition. These figures 331-337 with their legends are reproduced to explain in more detail the outline of the more recent Jean Bernard Causse stapes surgery technique.

As Tos (ref Tos, 2000) comments: "the principle of a 0.8 mm calibrated hole, which is then covered with a vein graft; a 0.4 mm all Teflon piston is placed into the hole. The idea is to reconstruct optimally the function of the annular ligament (see the original figure 330 (Figure 18). The vein graft offers good protection for the inner ear and prevents any excessive movement and diving of the piston into the vestibule in contrast to stapedotomy without interposition and large fenestra stapedectomy".

Tos (ref. Tos, 2000) concludes in his textbook that "Jean Bernard Causse presents excellent results using the described method, but it is difficult not to argue that this is due in a large part to the greater experience (and so craftsmanship) of the surgeon with this particular method than the interposition itself." The author of this PhD thesis is working in the Causse Clinic. He too has performed this surgical stapes replacement technique during decades, as reported with its results in the first 6 original contributions for very specific and rare indications to improve the impaired hearing levels.

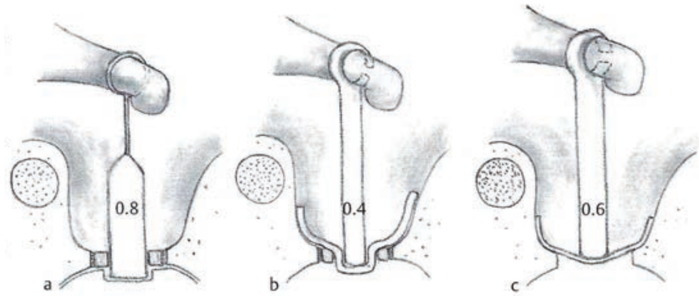

Figure 11. JB Causse's illustration of comparisons of movement of the footplate in various techniques. a) Stapedectomy with a 0.8-mm Teflon piston with endosteum covering the medial end of the piston. b) 0.8-mm stapedotomy with the interposed vein graft and 0.4-mm Teflon piston. c) Large fenestra stapedectomy with vein graft and interposition of a Teflon piston. (With the courtesy of M. Tos, Surgical solutions for conductive hearing loss, Thieme Stuttgart 2000)

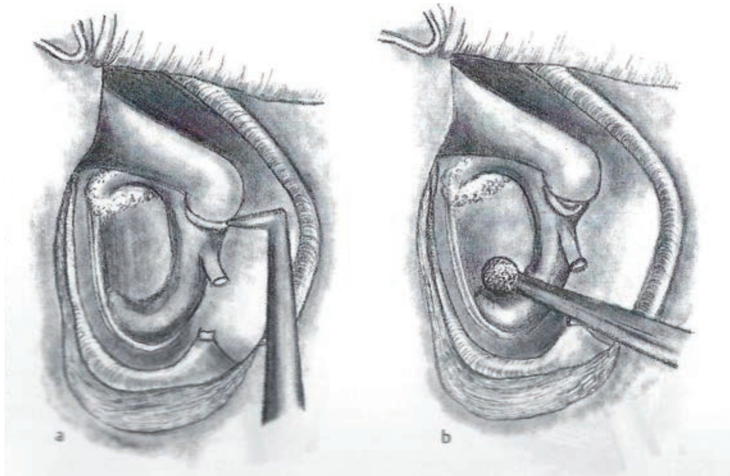

Figure 12. JB Causse's 0.8-mm stapedotomy technique. a) Relatively wide otosclerosis drilling is performed, exposing the posterior aspect of the stapes niche. The chorda tympani is pushed inferiorly, the stapedial tendon cut, and the incudostapedial joint separated. b) using a Treace-Causse microdrill, a 0.8-mm hole is formed, mainly in the posterior half of the footplate, by gently touching the footplate with the rotating head of the burr. (With the courtesy of M. Tos, Surgical solutions for conductive hearing loss, Thieme Stuttgart 2000)

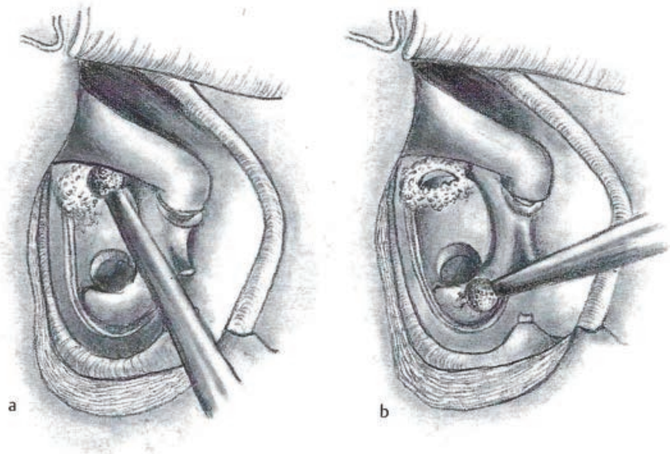

Figure 13. The anterior crus is drilled away close to the footplate; the drilling goes through the crura, without fracturing it. b) The posterior crus is drilled away. Drilling of the posterior crus can be performed before drilling of the anterior crus. (With the courtesy of M. Tos, Surgical solutions for conductive hearing loss, Thieme Stuttgart 2000)

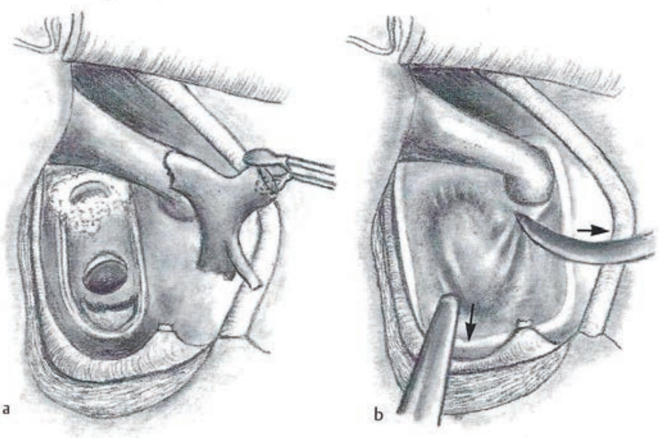

Figure 14. a) the stapes crura are removed. b) The vein is placed laterally over the oval window niche and pulled in various directions with the tip of the suction and the Rosen needle (arrows). (With the courtesy of M. Tos, Surgical solutions for conductive hearing loss, Thieme Stuttgart 2000)

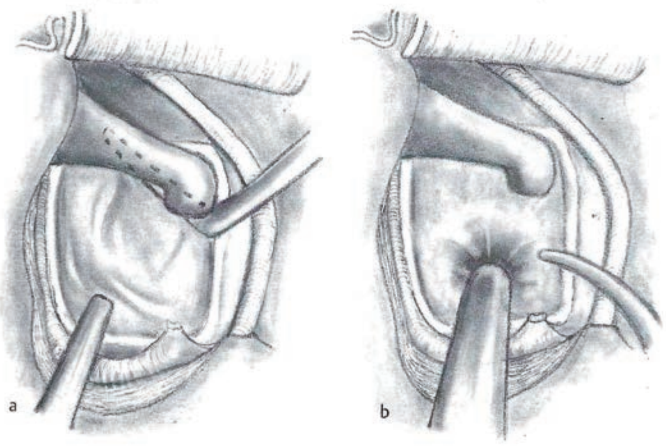

Figure 15. a) Further adaptation of the vein to the interior wall of the oval window niche, using a large hook. Movement of the vein is performed laterally in the niche, constantly elevating the vein from the footplate. b) Attempt to interpose the vein using a suction tip and Rosen needle. (With the courtesy of M. Tos, Surgical solutions for conductive hearing loss, Thieme Stuttgart 2000)

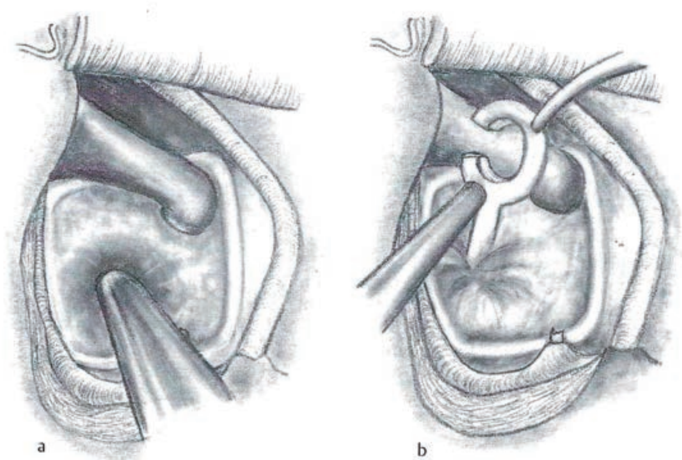

Figure 16. a) The suction tip pushes the vein into the 0.8-mm stapedotomy hole. The interposition is completed. b) A Teflon piston is placed into the stapedotomy hole with the interposed vein, the loop posterior to the long process of the incus. The limb of the loop is pushed with a suction tip and a Rosen needle over the incus. (With the courtesy of M. Tos, Surgical solutions for conductive hearing loss, Thieme Stuttgart 2000)

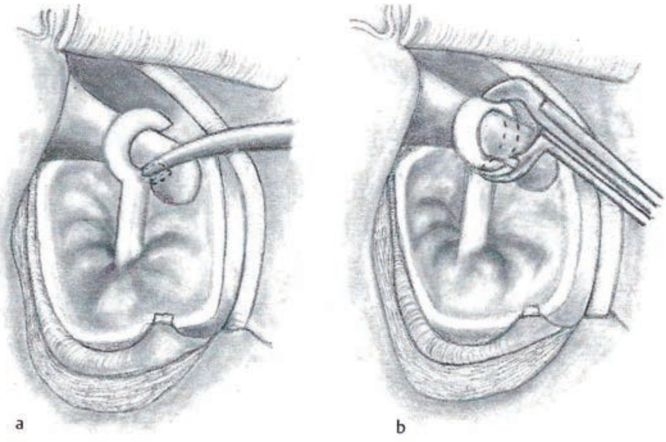

Figure 17. 1) The piston is slightly rotated and the loop is pushed in an anterior direction. b) The loop is gently crimped with curved cup forceps. (With the courtesy of M. Tos, Surgical solutions for conductive hearing loss, Thieme Stuttgart 2000)

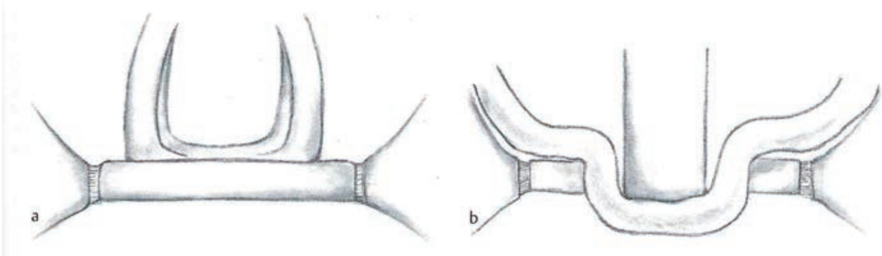

Figure 18. Schematic illustration of the JB Causse philosophy of function of 0.8 stapedotomy with interposition of a 2-mm thick piece of vein and a 0.4-mm piston. a) The normal 0.2-mm stapedial ligament. b) The vein, in Causse's explanation, replaces the ligament. (With the courtesy of M. Tos, Surgical solutions for conductive hearing loss, Thieme Stuttgart 2000)

Stapes revision surgery

Häusler (ref Häusler, 2004) published an overview of the hearing outcomes of primary stapes surgery for otosclerosis. Covering a period from 1978-2000, the number of surgeries in these series varied from N=49 to N=10.900. The mean air-bone gap closure for these series was < 10 dB in 70% and < 20 dB in 92%. These data show that in a low percentage of primary stapes surgery the hearing gain did not fulfill the expected and desired hearing gain. However, some cases needed revision surgery.

The main reasons for revision stapes surgery are a too large air-bone gap in the short or long term and/or persistent dizziness following surgery. Sometimes this can be due to a coexisting disease/anomaly like the presence of a third cochlear window as present in case of a bony dehiscence of the superior semicircular canal or the x-recessive stapes gusher syndrome.

A quick scanning of the revision surgery hearing outcomes over about 2 decades (ref. Häusler, 2004) did not show much improvement. However, over time improvements in surgical facilities and aids have become available to the surgeon, such as the use of preoperative high-resolution CT-scanning, LASER assisted micro- instruments, better micro-drills, and improved blood pressure control during general anesthesia. Also, the change from total stapedectomy to stapedotomy had a positive impact on the outcome of revision surgery.

In another table Häusler (ref. Häusler 2004) presents an overview of the hearing outcomes for stapes revision surgery for the period 1970-1995. Those hearing outcomes are rewarding but are nevertheless not very good.

Like Häusler (ref. Häusler ref. 2004), Vincent et al (2010) provides in his table 7 an overview of the hearing outcomes of his revision stapes surgery series published from 1997-2005. In table 2 he summarizes the different types of failure identified during revision surgery. Incus erosion (27.6%) and prosthesis displacement (18.2%) were the most common causes of failure. Since this original publication has become part of this PhD thesis, we will not provide in this "Chapter Introduction" an in-depth analysis of the data presented.

Ossiculoplasties: intact malleus and stapes

To evaluate the outcome of ossicular chain reconstruction in case of a missing or eroded incus, one must specify the presence, integrity and mobility of the malleus and the stapes.

It is also relevant whether the ossicular chain reconstruction is performed as an independent procedure or as part of a complicated canal wall up or canal down procedure. The aeration of the middle ear cleft is also relevant. Degrees of atelectasis of the tympanic membrane will influence the tension present in the tympanic membrane itself. Myringosclerosis, as a feature of tympanosclerosis, might affect the mobility of the tympanic membrane, especially when the myringosclerotic plaque is fixed to the bony annulus. (ref. Wielinga & Cremers, 1995) Most frequently, an eroded incus and the ensuing discontinuity of the ossicular chain is caused by a chronic otitis with or without cholesteatoma. It might also be the result of a skull base fracture with a dislocation at the level of the malleo-incudal joint and/or the incudostapedial joint. The fracture may displace the malleus from its natural site in the tympanic membrane to a more anteriorly position in relation to the position of the head of the stapes. Usually these ears are well aerated which is advantageous for the functional outcome.

For reconstruction of the ossicular chain in general the use of an unaffected autograft or allograft incus has been preferred over modelled bone grafts. As an alternative various biomaterials are available. In the metal group steel, tantalum, gold, platinum and titanium have been used, in the plastics group Vinyl acrylate, silicone, polyethylene and Teflon (polytetra fluoroethylene), and in the ceramics group aluminium oxide ceramic, glass ceramics, tricalcium phosphate ceramic and hydroxy apatite. (ref. Dost & Jahke, 2004).

Based on extrusion rates and new designs for Partial (PORP) and Total (TORP) Ossicular Replacement Prosthesis new surgical series with short- and long-term outcomes for hearing have been published. Although many confounding factors may probably bias the selection of the cases involved in these series, nevertheless a

measure of success identified related to the size of the airbone gap and the length of the follow up period. Therefore, in this PhD thesis an original contribution is enclosed, discussing the question whether the outcomes with a partial ossicular chain prosthesis (PORP) or a total ossicular chain prosthesis (TORP) are more favourable. (Vincent et al, 2011) The original malleus relocation and silastic banding techniques and their outcomes are described and reported as part of this series.

When performing ossiculoplasty it remains the goal of every otologist to work toward restoring the hearing mechanism to achieve the best possible hearing result and satisfy patient expectations. As the search for the perfect prosthesis continues, the importance of other factors, such as surgical techniquehave been highlighted as clear variables that require consideration when aiming for optimal results.

Ossiculoplasty: missing malleus and stapes

The more ossicles that are missing, the more complicated the reconstructive middle ear surgery will be to improve hearing. Earlier on we described that in case of a fixed stapes footplate a piston can be placed in a stapedotomy hole and fixed to the incus (incudo-stapedotomy) or to the malleus (malleostapedotomy). In situations with a fixed footplate and both a missing incus and malleus, a neomalleus can be placed in contact with the tympanic membrane in order to help fix/stabilise a stapes replacing piston. Such a technique has been described by Fisch as "Neomalleus and incus replacement with stapedotomy (NMRIS procedure) (ref. Fisch, 1994). Using a vein graft to cover the open footplate a type of columellar reconstruction can be used while reinforcing the tympanic membrane with a thinned cartilage graft. Smyth from Belfast mentioned such columellar reconstruction on a vein graft covering the oval window too, as illustrated in his figure 25 in 1976. (ref. Smith, 1976).

In case the footplate is mobile, various ways of columellar reconstruction towards the footplate may be possible and can be combined with cartilaginous reinforcement of the tympanic membrane. The “German Otologic School” started with the “Palisade technique” to repair and reinforce a tympanic membrane using thinned strips of autologous cartilage. Larger parts of the tympanic membrane can be reinforced with sliced tragal, cymbal or conchal cartilage. It helps to prevent extrusion of biomaterial ossicular replacement prostheses too. In this PhD thesis another new original clinical contribution is included (Vincent et al, 2012), viz. the evaluation of a new experimental surgical technique for ossiculoplasty to solve cases in which both the malleus and incus are missing.

Aim of this thesis

The main goal of this thesis is to analyze the outcomes of reconstructive middle ear surgery for specific groups of patients with a conductive hearing loss.

The first three original contributions concern the outcomes of exploratory tympanotomies for congenital minor ear anomalies Class I & II, III and IV following the Cremers classification. The next two original consecutive series present the outcomes of stapedotomy in Osteogenesis Imperfecta and the outcomes of stapedotomy in adolescents with otosclerosis. The sixth original contribution is to analyze the outcomes of 652 revision stapedotomy procedures.

The last two contributions relate to the outcomes of different types of ossiculoplasties. The first situation concerns the missing incus situation with still an intact malleus and an intact stapes in place. The last contribution is the evaluation of a new experimental surgical procedure to improve hearing in case of a missing malleus, incus and stapes superstructure with a mobile footplate.

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PART I

MALFORMATIONS

Chapter 2

Congenital Stapes Ankylosis in Children: Surgical Findings and Results in 35 Cases

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ABSTRACT

Objective: To evaluate surgical findings and hearing results in children undergoing middle ear surgery for congenital stapes ankylosis with or without other ossicular malformations (Teunissen and Cremers class I and class II malformations).

Study Design: A nonrandomized, nonblinded case series of prospectively collected data.

Setting: A tertiary referral center.

Patients: Twenty-eight consecutive pediatric patients who underwent 35 surgical procedures for congenital stapes ankylosis with or without other ossicular malformations and had available postoperative pure-tone audiometry.

Intervention: Primary stapedotomy with vein graft interposition and reconstruction with a Teflon piston, bucket handle prosthesis or total ossicular replacement prosthesis.

Main Outcome Measures: Pre- and postoperative audiometric evaluation using four-frequency (0.5, 1, 2, and 4 kHz) audiometry. Air-conduction thresholds, bone-conduction thresholds, and air-bone gaps (ABGs) were measured. Postoperative audiometry was performed at 3, 6, 9, 12, 18, and 24 months after surgery and at a yearly interval thereafter.

Results: Overall, a postoperative ABG closure of 10 dB or less was achieved in 73% of class I cases and in 50% of class II cases. A postoperative ABG closure of 20 dB or less was achieved in 77% of class I cases and 67% of class II cases. Postoperative sensorineural hearing loss occurred in one class I case (4%) and none of the class II cases.

Conclusion: Stapedotomy is a safe and feasible treatment option in children with congenital stapes ankylosis.

Conductive hearing loss is rarely caused by congenital middle ear malformations. Reported incidence varies between 0.5 and 3% in patients undergoing surgical treatment for conductive hearing loss (1,2). Congenital middle ear malformations are classified as major when they are accompanied by tympanic membrane or external ear malformations. Minor congenital middle ear malformations are limited to the middle ear and can be classified into four main groups according to the Teunissen and Cremers classification (3): (class I) isolated congenital stapes ankylosis, (class II) stapes ankylosis associated with other ossicular malformations, (class III) congenital malformations of the ossicular chain with a mobile stapes footplate, and (class IV) severe (IVa) aplasia or (IVb) dysplasia of the oval or round window. The majority of minor congenital middle ear malformations are classified as class I (31%) or class II (38%) malformations (3). In the past, stapes surgery in case of congenital middle ear malformations has been thought to be associated with increased risks of perilymphatic gusher and sensorineural hearing loss (SNHL) (4,5). Meanwhile, numerous authors have published the results of case series of patients undergoing stapes surgery for congenital middle ear malformations with satisfactory results (2,3,6–10). The objective of this study was to evaluate surgical findings and hearing results in children undergoing middle ear surgery for congenital stapes ankylosis with or without other ossicular malformations (class I and class II malformations).

METHODS

Between January 1991 and October 2014, 225 operations for congenital malformations of the middle ear were performed by the same surgeon (first author) in the same tertiary referral center. Of these 225 operations, 69 were performed in children and congenital stapes ankylosis was identified in 35 of these cases. The remaining 34 cases underwent surgery for class III and class IV congenital malformations and were excluded from further analyses.

Study Population

This is a nonrandomized, nonblinded study of prospectively collected data of 28 children, younger than 18 years of age, who underwent 35 consecutive operations for class I and class II congenital middle ear malformations from 1991 to 2014. Whenever we refer to cases in this article, we refer to operations and not patients. Of the 28 included children, seven had sequential bilateral middle ear surgery. Postoperative pure-tone audiometry was available for all of these patients. No patients were excluded because they lacked postoperative pure-tone audiometric data. None of the included children were diagnosed with a concomitant syndrome. The presence of concomitant syndromes was evaluated using computed tomography (CT) imaging only.

Children diagnosed with juvenile otosclerosis or Osteogenesis Imperfecta were excluded, because these disease entities have their own approach and outcome, the results of which have been published previously (11,12). Juvenile otosclerosis and osteogenesis imperfecta were diagnosed based on family history, a clinical history of progressive conductive hearing loss, intraoperative findings, and (otosclerotic foci on) preoperative CT imaging. CT imaging was also used to exclude other causes of conductive hearing loss, such as enlarged vestibular syndrome.

Audiometric Assessment

A serial assessment of hearing status was conducted before and at 3, 6, 9, 12, 18, and 24 months after surgery and at a yearly interval thereafter. Audiometric evaluation included pre- and postoperative air-bone gaps (ABGs), air-conduction (AC) thresholds, and bone-conduction (BC) thresholds. Only AC and BC thresholds that were obtained at the same time postoperatively were used for calculation of the mean postoperative ABG. Four-frequency (0.5, 1, 2, and 4 kHz) pure-tone averages for AC and BC thresholds, obtained at the last follow-up visit, were used in this study. Furthermore, closure of the ABG to within 10 dB or less was evaluated, as well as ABG closure to within 20 dB or less. Audiometry was reported according to American Academy of Otolaryngology-Head and Neck Surgery guidelines (13), except for thresholds at 3 kHz, which were substituted in all cases with those at 4 kHz. Audiometric data were collected prospectively using the Otology-Neurotology Database (ONDB) (AS Multimedia, Inc., Cassagne, France) (14).

Statistical Analyses

Means, standard deviations (SDs), and percentages were calculated. Differences in pre- and postoperative hearing outcomes were calculated as well as their corresponding 95% confidence intervals. Pre- and postoperative BC thresholds, AC thresholds, and ABGs were compared using a paired-samples t test. All analyses were performed using SPSS version 20.0 (SPSS Inc., Chicago, IL).

Surgery

A transcanal procedure with facial nerve monitoring using the NIM 2.0 (Medtronic, Inc., Jacksonville, FL) was performed in all cases. The anatomy and mobility of the ossicular chain were carefully checked. The facial nerve was identified and its relationship with the ossicular chain and/or stapes remnant was assessed (15). The Teunissen and Cremers classification system (3) was used to define the type of malformation and ossicular chain status. The surgical technique for the repair of the ossicular chain was dictated by the surgical findings at the time of surgery and were described according to the Teunissen and Cremers classification. Surgical data were collected prospectively using the ONDB (AS Multimedia, Inc.) (14).

Teunissen and Cremers

Class I Cases Of the total series of 35 cases, 23 cases (66%) were class I cases, defined as stapes fixation only without any other simultaneous malformation (Supplementary Digital Table 1, http://links.lww.com/MAO/A383). Of these 23 cases, 22 were primary operations and one was a revision operation (case 14).

Table 1. Overall pure-tone audiometric results for Teunissen and Cremers Class I cases (number of procedures = 23)

Outcome	Preoperative	Postoperative	Difference (95% CI)	P
AC (mean SD) dB)	46 (10)	32 (22)	14 (4-24)	0.008
BC (mean SD) dB)	17 (7)	20 (19)	-3 (-11 to 5)	0.436
ABG (mean SD) dB)[a]	29 (8)	12 (15)	17 (-24)	0.000
ABG closure ≤ 10 dB (n (%))[a]	-	16 (73)	-	-
ABG closure ≥ 20 dB (n (%))[a]	-	17 (77)	-	-
SNHL > 15 dB (n (%))	-	1 (4)	-	-

95% CI indicates 95% confidence interval; AC, air conduction; BC, bone conduction; ABG, air-bone gap; SD, standard deviation; SNHL sensorineural hearing loss. Means were calculated using AC and BC thresholds at 0.5, 1, 2, and 4 kHz.
[a]One case with SNHL was not included in the assessment of the postoperative ABG and ABG closure to within 10 and 20 dB (n=22).

Eleven procedures were performed on the right ear (48%) and 12 on the left ear (52%). Four patients were operated sequentially on both sides. Sex ratio was 44% women and 57% men. Mean age was 11.5 years (with a range of 7–17 yrs). Some authors have advocated waiting until age 8 to 10 years before surgical intervention and treating children with hearing aid amplification instead (3,5,8). In a few cases in this series, surgery was performed before the age of 8 years, because these cases suffered from severe hearing loss and a hearing aid was not well tolerated.

In two of 23 cases (9%) middle ear exploration revealed fixation of the stapes owing to a bony bar between the pyramidal process or the promontory and the stapes superstructure (cases 1 and 21) (Figure 1A). The stapes footplate was normal in both cases. The bony bridge was carefully drilled out with the Skeeter drill using a 0.7-mm diameter diamond dust burr. The stapes remained mobile and the entire ossicular chain was preserved. In one case (case 1), no ossiculoplasty was attempted whereas in another case (case 21) a 0.4-mm diameter Teflon piston was positioned between the incus and the mobile stapes footplate without removing the superstructure (incus-to-footplate assembly). The choice between these two techniques was arbitrary.

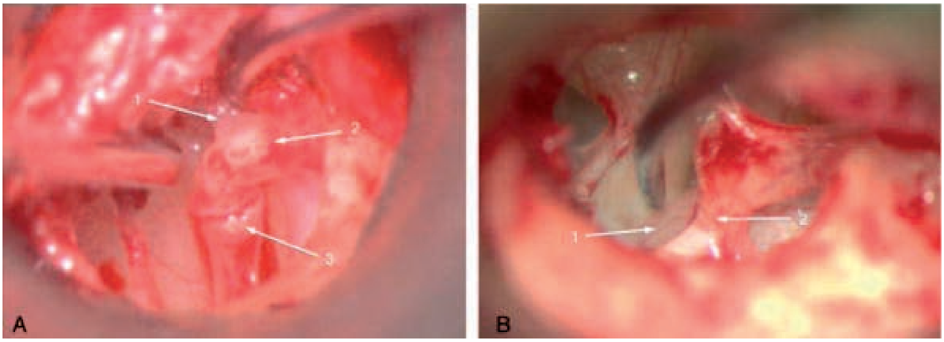

Figure 1A. Operative view of a left ear showing a stapes-pyramidal fixation by a bony bar. 1 = stapes-pyramidal (anterior) bony bar; 2 = incus; 3 = stapes. B, Operative view of a left ear. The facial nerve is coursing across the oval window, which is totally obstructed. 1 = dehiscent facial nerve; 2 = stapes.

An isolated congenital stapes fixation without the presence of a bony bridge was seen in 21 cases (91%). The technique was similar to our approach in primary stapedotomy: a regular 0.8-mm stapedotomy was performed using a combination of a laser and the Skeeter Microdrill (Medtronic Xomed, Inc., Jacksonville, FL), followed by vein graft interposition and incus-to-stapedotomy assembly (14). The KTP laser (Lumenis, Inc., Salt Lake City, UT) was used in 16 cases (70%), the CO_2 laser (OmniGuide, Inc., Cambridge, MA) in six cases (26%), and the Revolix Thulium laser (LISA Laser, Inc., Pleasanton, CA) in one case (4%). Ossiculoplasty was performed using a 0.4-mm-diameter Teflon piston of appropriate length when the incus was of normal length (20 cases [95%]). A bucket-handle (cup)–type prosthesis made of Teflon was used when the facial nerve was dehiscent (one case [5%]). Of these 23 cases, a perilymphatic gusher occurred bilaterally in one patient (cases 2 and 3) (two cases [9%]). Both cases were treated with stapedotomy with vein graft interposition. Gelfoam was used to fill the oval window and keep the vein graft in its proper position.

Teunissen and Cremers Class II Cases

Of the entire series of 35 cases, 12 cases (34%) were class II cases, defined as stapes fixation with other ossicular malformation (Supplementary Digital Table 2, http://links.lww.com/MAO/A384). Of these 12 cases, eight were primary operations and four were revision operations (cases 2, 4, 10, and 11). One patient was operated bilaterally. Eight procedures were performed on the right ear (67%) and four on the left ear (33%). Sex ratio was 75% women and 25% man. Mean age was 12.4 years (with a range of 6–18 yrs).

Table 2. Overall pure-tone audiometric results for Teunissen and Cremers class II cases (number of procedures¼12)

Outcome	Preoperative	Postoperative	Difference (95% CI)	P
AC (mean SD) dB)	49 (10	29 (15)	20 (8-32)	0.004
BC (mean SD) dB)	18 (6)	17 (3)	2 (-1 to 4)	0.221
ABG (mean SD) dB)[a]	31 (12)	13 (14)	18 (6-31)	0.008
ABG closure ≤ 10 dB (n (%))[a]	-	6 (50)	-	-
ABG closure ≥ 20 dB (n (%))[a]	-	8 (67)	-	-
SNHL > 15 dB (n (%))	-	0 (0)	-	-

95% CI indicates 95% confidence interval; AC, air conduction; BC, bone conduction; ABG, air-bone gap; SD, standard deviation.

A simultaneous malleus ankylosis was identified in one case (case 7) (8%) and a total ossicular replacement prosthesis, composed of a hydroxyapatite head and a 0.4-mm-diameter Teflon shaft, was positioned between the malleus and the stapedotomy (malleus-to-stapedotomy procedure). The malleus relocation technique was used to increase the stability of the prosthesis (14,16). The malleus relocation technique was introduced by the first author in 1997. The malleus is dissected free from the tympanic membrane and the tensor tympani is sectioned close to its insertion into the malleus. The incus is then removed. The anterior malleal ligament is stretched by placing a hook anterior to the neck of the malleus, which is then pulled posteriorly, repositioning it so that it lies directly over the footplate. The position of the malleus is maintained by its superior ligament, which is then preserved.

The incus was malformed in 10 cases (83%). In eight cases, the incus appeared hypoplastic and short, but it was preserved and a Teflon piston was inserted (incus-to-stapedotomy procedure). A total ossicular replacement prosthesis was positioned between the malleus and the stapedotomy (malleus-to-stapedotomy procedure) in two cases. In one of these cases, the long process was short, just reaching the area of the facial nerve. In the other case, the incus consisted of a fibrous band, which was connected to the malformed and fixed stapes. The malleus relocation technique was used in the two latter cases.

In one case (8%), stapedotomy was not performed at all, because the facial nerve coursed across the oval window, which itself was totally obstructed (case 12) (Figure 1B). The surgery was aborted for the high risk of facial nerve injury. This was the only surgery that was aborted out of all class I and class II cases.

Furthermore, one case (8%) with a persistent stapedial artery (case 2) and one case (8%) with a perilymphatic gusher (case 1) were observed in this class II group.

RESULTS

Teunissen and Cremers Class I Cases. Overall Results (Table 1 and Supplementary Digital Table 1, http://links.lww.com/MAO/A383)

Postoperative data were available for all cases. The average follow-up duration was 21 months (with a range of 3–83 mo). A significant postoperative SNHL, defined as a change in the average BC threshold of 15 dB or more, was observed in one case (4%) in this series. This case experienced a dead ear immediately after surgery. This case was not included in the assessment of the postoperative ABG and ABG closure to within 10 and 20 dB (n = 22 cases), but was included in the assessment of the postoperative AC and BC thresholds (n = 23 cases). Overall, the postoperative four-frequency ABG was closed to 10 dB or less in 16 of 22 cases (73%) and to 20 dB or less in 17 of 22 cases (77%). The postoperative four-frequency average ABG was 12 dB (SD 15) compared with 29 dB (SD 8) preoperatively. The postoperative four-frequency average AC threshold was 32 dB (SD 22) compared with 46 dB (SD 10) preoperatively. Both ABG and AC threshold improved significantly with mean gains of 17 dB (95% CI, 9–24, p = 0.000) and 14 dB (95% CI, 4–24, p = 0.008). Post-operatively, the four-frequency average BC threshold was 20 dB (SD 19) compared with 17 dB (SD 7) preoperatively, thus the BC threshold deteriorated with 3 dB (95% CI, -11 to 5, p = 0.437).

Hearing Results With at Least 1-year Follow-up (Supplementary Digital Table 1, http://links.lww.com/MAO/A383, and Supplementary Digital Table 3, http://links.lww.com/MAO/A385)

Postoperative data with at least 1-year follow-up were available for 14 of 23 cases (61%). The average follow-up duration was 28 months (with a range of 12–83 mo). The case presenting with a significant postoperative SNHL was not included in the assessment of the postoperative ABG and ABG closure to within 10 and 20 dB (n = 13 cases), but was included in the assessment of the postoperative AC and BC thresholds (n = 14 cases). The postoperative four-frequency ABG was closed to 10 dB or less in 8 of 13 cases (62%) and to 20 dB or less in 9 of 13 cases (69%). The postoperative four-frequency average ABG was 16 dB (SD 18) compared with 29 dB (SD 9) preoperatively.

Table 3. Reported pure-tone audiometric results following surgical treatment of Teunissen and Cremers class I and class II cases in literature

References	N	Class	Mean Follow-up Duration in Months (range)	% ABG Closure <10 dB	% ABG Closure <20 dB	Post ABG (mean [SD])	Post AC (mean [SD])	Gain AC (mean [SD])	% SNHL >10 dB
Kisilevsky et al. (2)[a]	22	Class I	NR	NR	NR	11	35	18	NR
	15	Class II		NR	NR	13	34	18	NR
Teunissen and Cremers (3)[b]	41	Class I	12	49	71	NR	NR	NR	NR
	51	Class II	12	39	72	NR	NR	NR	NR
Albert et al. (6)[c]	28	Class I	19 (6-82)	NR	NR	10 (3-28)[d]	15 (5-40)[d]	33 (5-52)[d]	0
Massey et al. (7)[a]	25	Class I	> 4	48	80	12	NR	NR	8
Thomeer et al. (8)[c]	39	Class I	12	46	74	14	25	21	0
Thomeer et al. (9)[c]	30	Class II	12	23	70	20 (14)	31 (18)	18 (21)	7
Welling et al. (10)[a]	21	Class I	6	24	67	16	22	21	0
Park and Choung (18)[a]	46	Class I	NR	NR	80	NR	NR	NR	NR
	10	Class II			30				Unclear (8 with severe SNHL
									4
									0
Raveh et al. (19)[a]	12	Class I	>12	17	25	NR	NR	NR	
This study[c]	23	Class I	21 (3-83)	73	77	12 (15)	32 (22)	14 (23)	
	12	Class II	26(3-119)	50	67	13(14)	29 (15)	20 (19)	

n indicates number of procedures; ABG, air-bone gap; AC, air conduction; SNHL, sensorineural hearing loss; SD, standard deviation; NR, not reported.

[a] 0.5, 1, 2, and 3 kHz.

[b] 0.5, 1, and 2 kHz.

[c] 0.5, 1, 2, and 4 kHz.

[d] Median with range instead of mean with SD.

The postoperative four-frequency average AC threshold was 39 dB (SD 24) compared with 48 dB (SD 10) preoperatively. Postoperatively, the four-frequency average BC threshold was 25 dB (SD 23) compared with 19 dB (SD 7) preoperatively. Only the average ABG threshold improved significantly with a mean gain of 13 dB (95% CI, 1–25, p = 0.037). The audiometric results of all individual cases are visualized using the Amsterdam Hearing Evaluation Plots in Figures 2 and 3(17). The gain in AC thresholds following surgery is dependent on preoperative ABG and both of these measures are plotted in Figure 2. Every dot below the solid diagonal line indicates a gain in AC threshold that is larger than the preoperative ABG. Every dot above the dotted diagonal line indicates an unsuccessful result, an ABG closure of more than 10 dB. The effect of surgery on BC thresholds is depicted in Figure 3 by plotting pre- and postoperative BC thresholds. The two dotted diagonal lines enclose those cases in which BC threshold did not change more than 10 dB. Every dot above the upper diagonal line indicates a case in which the BC threshold deteriorated by more than 10 dB, whereas every dot below the lower diagonal line indicates an improvement of the BC thresholds of more than 10 dB.

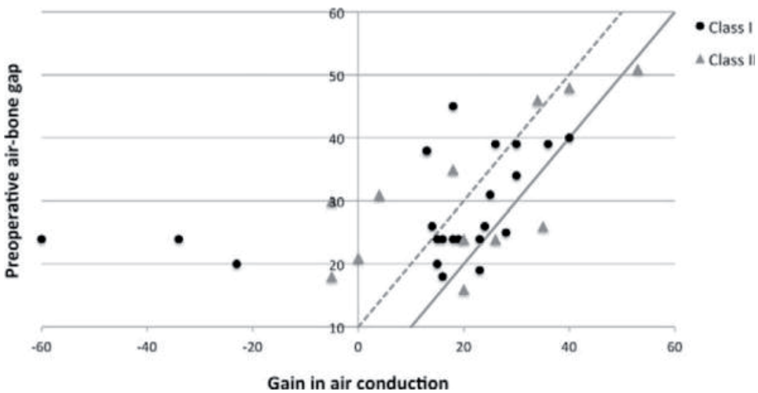

Figure 2. Amsterdam Hearing Evaluation Plot. Each black dot represents one case. The solid diagonal line indicates total air-bone gap closure. The area between the diagonal lines indicates successful surgery with an air-bone gap of 10 dB or less. Audiometric data at last follow-up were used (23 class I cases and 12 class II cases).

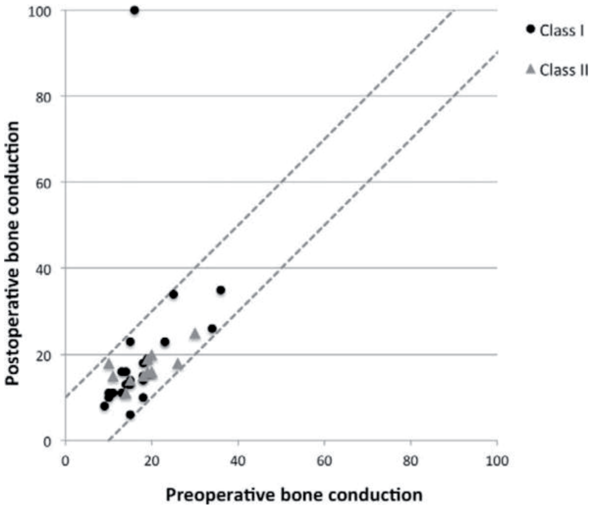

Figure 3 . Amsterdam Hearing Evaluation Plot. Each black dot represents one case. The two diagonal lines enclose the cases in which bone conduction did not change more than 10 dB. Audiometric data at last follow-up were used (23 class I cases and 12 class II cases).

Teunissen and Cremers Class II Cases

Overall results are available in (Table 2 and Supplementary Digital Table 2, http://links.lww.com/MAO/A384).

Postoperative data were available for all cases. The average follow-up duration was 26 months (with a range of 3–119 mo). No significant postoperative SNHL was observed in this series of cases. Overall, the postoperative four-frequency ABG was closed to 10 dB or less in six of 12 cases (50%) and to 20 dB or less in eight of 12 cases (67%). The postoperative four-frequency average ABG was 13 dB (SD 14) compared with 31 dB (SD 12) preoperatively. The postoperative four-frequency AC threshold was 29 dB (SD 15) compared with 49 dB (SD 10) preoperatively. Both ABG and AC threshold improved significantly with mean gains of 18 dB (95% CI, 6–31, p = 0.008) and 20 dB (95% CI, 8–32, p = 0.004). Postoperatively, the four-frequency average BC threshold was 17 dB (SD 3) compared with 18 dB (SD 6) preoperatively; thus, the BC threshold improved with 2 dB (95% CI, -1 to 4, p = 0.221).

Hearing Results With at Least 1-year Follow-up (Supplementary Digital Table 2, http://links.lww.com/MAO/A384, and Supplementary Digital Table 4.

Postoperative data with at least 1-year follow-up were available for eight of 12 cases (67%). The average follow-up duration was 37 months (with a range of 12–119 mo). The postoperative four-frequency ABG was closed to 10 dB or less in five cases (63%)

and to 20 dB or less in seven cases (88%). The postoperative four-frequency average ABG was 8 dB (SD 11) compared with 33 dB (SD 14) preoperatively. The postoperative four-frequency AC threshold was 25 dB (SD 14) compared with 52 dB (SD 12) preoperatively. Both ABG and AC threshold improved significantly with mean gains of 25 dB (95% CI, 9–41, p = 0.008) and 27 dB (95% CI, 12–41, p = 0.003). Postoperatively, the four-frequency average BC threshold was 17 dB (SD 3) compared with 19 dB (SD 7) preoperatively; thus, the BC threshold improved with 2 dB (95% CI, -3 to 6, p = 0.375).

Stability of Hearing Results

Short-term follow-up (3 mo) was compared with long-term follow-up (1 year) for those cases with available pure-tone audiometry at these two follow-up moments (Supplementary Digital Table 5, http://links.lww.com/MAO/A387). The differences in postoperative four-frequency average AC thresholds, BC thresholds, and ABG between these follow-up moments were not statistically significant. Nor were the differences in ABG closure to within 10 dB or less and ABG closure to within 20 dB.

DISCUSSION

In this study, we evaluated the hearing outcomes after DISCUSSION In this study, we evaluated the hearing outcomes after stapedotomy for congenital stapes ankylosis in 23 pediatric class I cases and 12 pediatric class II cases. Overall, a postoperative ABG closure to within 10 dB was achieved in 73% of class I cases and in 50% of class II cases. A postoperative ABG closure to within 20 dB was achieved in 77% of class I cases and 67% of class II cases. Postoperative SNHL occurred in one class I case (4%) and none of the class II cases. A rapid systematic literature search identified nine articles (2,3,6–10,18,19) reporting audiometric results following stapes surgery in children with class I and/or class II congenital stapes ankylosis. We only included studies that included at least 10 cases and performed a subanalysis for class I and/or class II cases. Overall, an ABG closure of 10 dB or less was reached in 17 to 49% in cases with class I congenital stapes ankylosis in five studies (Table 3). An ABG closure of 20 dB or less in class I cases was reported in six studies and was reached in 25 to 80%. Two studies reported the percentage of class II cases with ABG closure to within 10 dB: 39 and 23%. ABG closure to within 20 dB in class II cases was reported in three studies and was reached in 30 to 72%.

Both juvenile otosclerosis and congenital stapes fixation are indications for middle ear surgery in children. Juvenile otosclerosis is more likely in cases with progressive conductive hearing loss and a positive family history for otosclerosis (20). Furthermore, intraoperatively established white otosclerotic foci in the anterior oval window with a lack of vascularization are characteristic for juvenile otosclerosis (20). Although the family history may contribute to a definitive diagnosis, it needs to be noted that both diseases can be hereditary and may simultaneously run in one family. CT imaging can be helpful in ruling out large vestibular aqueduct syndrome or other inner ear abnormalities that are associated with a higher risk of perilymph gusher during surgery and consequent SNHL (21). Furthermore, incudostapedial discontinuity and increased thickness of the oval window may be witnessed on CT. However, it is not uncommon for the CT scan to be entirely normal in congenital stapes ankylosis and these abnormalities are not pathognomonic for congenital middle ear malformations (6,21). Although CT scans were performed routinely in all cases included in this study, the findings were not routinely collected in the ONDB and therefore the results of CT scanning were not described in this paper. It is not unlikely that congenital stapes ankylosis is sometimes falsely considered to be juvenile otosclerosis and the other way around. Both are characterized by fixation of the stapes and both can be successfully treated with stapes surgery. Several studies were identified that compared the surgical results of congenital stapes ankylosis with juvenile otosclerosis. Children with juvenile otosclerosis generally showed better hearing results following stapes surgery than children with congenital stapes fixation. Risk differences were calculated. A risk difference is the difference between the observed risks in two treatment groups (22). Risk differences for ABG closure to within 10 dB differed between 4 and 38% in favor of juvenile otosclerosis in eight studies (23–30). Risk differences for ABG closure to within 20 dB differed between 0 and 30% in favor of juvenile otosclerosis in six studies (23–25,27,29,30) It needs to be noted that these studies included a variety of congenital stapes malformations and did not report them according to the Teunissen and Cremers classification or did not perform subanalyses according to this classification.

Furthermore, all of the sample sizes were small and as a result none of these risk differences were statistically significant. In the past, stapes surgery in case of congenital middle ear malformations has been thought to be associated with an increased risk of SNHL. In our series, only one class I case and none of the class II cases showed a postoperative SNHL exceeding 15 dB. In the included studies, SNHL was usually defined as a change in BC thresholds exceeding 10 dB. In these studies, SNHL was reported in less than 10% of surgically treated class I and class II cases (6–10). Case series are vulnerable to selection bias. This case series was characterized by a clear study objective, explicit in- and exclusion criteria, consecutive patient enrollment, prospective outcome data collection, and a high follow-up rate. These strengths help to limit selection bias (22,31). Limitations of this case series include a relatively low sample size and the lack of a comparator group. Furthermore, follow-up duration was not standardized with a wide range of 3 to 119 months as a result.

CONCLUSION

Stapedotomy in children with congenital stapes ankylosis class I and class II was associated with satisfactory pure-tone audiometric results, with an ABG closure of 10 dB or less in 73% of class I cases and in 50% of class II cases. An ABG closure of 20 dB or less was achieved in 77% of class I cases and 67% of class II cases. Postoperative SNHL occurred in one class I case (4%) and none of the class II cases.

2

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Chapter 3

Congenital ossicular chain malformations with mobile stapes in children: Results in 17 cases

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ABSTRACT

Objectives/Hypothesis: To prospectively evaluate surgical findings and hearing results in children undergoing surgery for congenital malformations of the ossicular chain with a mobile stapes footplate (Teunissen and Cremers class III malformations).

Study Design: A nonrandomized, nonblinded, prospective case series.

Methods: Fifteen consecutive pediatric patients who underwent 17 surgical procedures for congenital ossicular malformations with a mobile stapes at a tertiary referral center were included. In 16 cases, postoperative pure-tone audiometry was available. The surgical technique for repair of the ossicular chain was dictated by the surgical findings at the time of surgery. The majority of the cases underwent ossiculoplasty. A Teflon piston, partial ossicular replacement prosthesis, or total ossicular replacement prosthesis was used in these cases. Associated surgical techniques included malleus relocation, Silastic banding, drilling out of a bony bridge, and a combination of these techniques. Hearing loss was evaluated using preoperative and postoperative four-frequency (0.5, 1, 2, and 4 kHz) audiometry. Air-conduction thresholds, bone-conduction thresholds, and air-bone gaps were measured. Postoperative audiometry was performed at 3, 6, 9, 12, 18, and 24 months after surgery and at a yearly interval thereafter.

Results: Overall, a postoperative air-bone gap closure to 10 dB or less was achieved in 63% of the included cases. A postoperative air-bone gap closure to 20 dB or less was achieved in 75%. Postoperative sensorineural hearing loss did not occur in any of the cases.

Conclusions: Ossicular reconstruction is a feasible treatment option in children with congenital malformations of the ossicular chain with a mobile stapes footplate.

INTRODUCTION

Conductive hearing loss due to congenital middle ear malformations occurs in approximately 1 per 11,000 to 15,000 patients.[1, 2] A division can be made between minor and major middle ear malformations; in case of the latter, malformations of the tympanic membrane and external ear are also present. Minor middle ear malformations are a small fraction of the total and can be classified according to the Teunissen and Cremers classification[3]: class I, isolated congenital stapes ankylosis; class II, stapes ankylosis associated with other ossicular malformations; class III, congenital malformations of the ossicular chain with a mobile stapes footplate; and class IV, severe aplasia or dysplasia of the oval or round window. The majority of minor congenital middle ear malformations are classified as class I (31%) or class II (38%) malformations.[3] Class III malformations consist of less than a third of all minor middle ear malformations. Class III malformations can be further subdivided in ossicular chain discontinuity (class IIIa) and epitympanic fixation (class IIIb).[3]

The classification system described above is used because of the implications for the different surgical reconstruction possibilities. Surgical treatment of class III malformations consists of reconstruction of the ossicular chain, whether or not in combination with mobilization. There are various surgical techniques for reconstruction of the ossicular chain; the method used depends on the type of malformation.

The objective of this prospective study was to evaluate surgical findings and hearing results in children undergoing middle ear surgery for congenital malformations of the ossicular chain with a mobile stapes footplate (class III malformations).

MATERIALS AND METHODS

Between January 1991 and October 2014, 225 operations for congenital malformations of the middle ear were performed by the same surgeon (first author) in the same tertiary referral center. Of these 225 operations, 69 were performed in children, and congenital malformations of the ossicular chain with a mobile stapes footplate were identified in 17 of these cases. The remaining 52 patients underwent surgery for class I, class II, and class IV congenital malformations and were excluded from further analyses.

Patients

This was a nonrandomized, nonblinded, prospective study of 15 children, under 18 years of age, who underwent 17 consecutive operations for class III congenital middle ear malformations from 1991 to 2011. Of the 15 included children, two had sequential bilateral surgery. Postoperative pure-tone audiometry was available for 16 cases (94%). Children diagnosed with juvenile otosclerosis or osteogenesis imperfecta were excluded, because these disease entities have their own approach and outcome.

Audiometric Assessment

A serial assessment of the hearing status was conducted before and at regular intervals after surgery. Audiometric evaluation included preoperative and postoperative air-bone gaps (ABGs), air-conduction (AC) thresholds, and bone-conduction (BC) thresholds. Only AC and BC thresholds that were obtained at the same time postoperatively were used for calculation of the mean postoperative ABG. Four-frequency (0.5, 1, 2, and 4 kHz) pure-tone averages for AC and BC thresholds, obtained at the last follow-up visit, were used for analysis of the outcome in this study. Furthermore, closure of the ABG to 10 dB or less was evaluated, as well as ABG closure to 20 dB or less. Audiometry was reported according to American Academy of Otolaryngology–Head and Neck Surgery guidelines,[4] with the exception of the thresholds at 3 kHz, which were substituted in all cases with those at 4 kHz. The audiometric results of all individual cases were visualized using the Amsterdam hearing evaluation plots.[5] Audiometric data were collected prospectively using the Otology- Neurotology Database (ONDB) (AS Multimedia, Inc., Cassagne, France).[6]

Surgery

A transcanal procedure with facial nerve monitoring using the NIM 2.0 (Medtronic Xomed, Inc., Jacksonville, FL) was performed in all cases in this series. The anatomy and mobility of the ossicular chain were carefully checked. The facial nerve was identified, and its relationship with the ossicular chain and/or stapes remnant assessed.[7] The Teunissen and Cremers classification system[3] was used to define the type of malformation and ossicular chain status. The surgical technique for repair of the ossicular chain was dictated by the surgical findings at the time of surgery and was described according to each Teunissen and Cremers class. Surgical data were collected prospectively using the ONDB.[6]

Statistical Analyses

Means, standard deviations (SDs), and percentages were calculated. Preoperative and postoperative AC thresholds, BC thresholds, and ABGs were compared using the nonparametric related-samples Wilcoxon signed rank test. All analyses were performed using SPSS version 20.0 (SPSS Inc., Chicago, IL).

Preoperative and Surgical Findings

Thirteen cases underwent primary surgery, three cases underwent second stage surgery, and one case underwent revision surgery (Table 1). Ten procedures were performed on the right ear (59%) and seven on the left ear (41%). Two patients were operated sequentially on both sides. Sex ratio was 41% female and 59% male. Mean age at the time of surgery was 12 years (range, 7–18 years).

Malleus ankylosis was observed in five out of 17 cases (29%) during middle ear exploration. One of these cases was also associated with a simultaneous stapes malformation (case 10). Ossiculoplasty was performed using a partial ossicular replacement prosthesis (PORP) with malleus-to-stapes head assembly in one case and a total ossicular replacement prosthesis (TORP) with malleus-to-footplate assembly in four cases, despite the evidence of a stapedial arch. A study performed in our center comparing the use of PORP with the use of TORP with Silastic banding showed better hearing outcomes when using the latter.[8] The one case in which a PORP was used was operated on before the introduction of the Silastic banding technique. The prosthesis shaft of the TORPs was inserted between the fallopian canal and the stapes superstructure, and the distal end of the prosthesis shaft was in direct contact with the mobile stapes footplate. The malleus relocation technique was used in two of the TORP cases.[9] A combination of malleus relocation and Silastic banding was used in the other two TORP cases.[8, 10] The Silastic banding technique was first introduced by the first author in 2000. A Silastic band is fashioned by punching a 1.2-mm disc from a thin sheet of Silastic with a specially designed punch (Medtronic Xomed, Inc., Jacksonville, FL). The center of the disc is then fenestrated using a 0.8-mm-diameter punch. The band should fit snugly around the stapes neck. The stapedius tendon is divided as closely as possible to the pyramid, as an attempt is then made to place the band beneath the stapedius tendon to avoid lateral displacement of the band. The shaft of the TORP is cut to the appropriate length and is introduced through the band. The shaft is introduced between the stapes and the fallopian canal, with the distal end lowered onto the center of the footplate. The prosthesis' head is then introduced under the relocated malleus handle. In its final position, the prosthesis' head should rest directly under the malleus handle without undue tension, and the malleus neck should stay away from the superior wall of the external auditory canal. The band is then gently pulled and positioned around the stapes head, beneath the stapedius tendon, which has been cut. By snugging the band down over the sectioned tendon, the prosthesis becomes firmly apposed to the stapes, allowing precise placement

Table I. Individual Surgical Findings and Audiometric Results for Teunissen and Cremers Class III Patients (Number of Procedures 517).

Patient	Case No.	Age,yr	Follow-up No.	Type of malformation	Previous Surgery for Microtia	Surgical technique	Ossiculoplasty	Pre ABG,dB	Post BG,dB	Pre AC,dB	Post AC,dB
A	01	8	24	Malformed malleus/ malformed incus	No	Malleus relocation	PORP (malleus to stapes head)	25	26	63	59
B	02	9	75	Malformed TM/bony bridge between malleus and EAC	No	Drilling out of the bony bridge	No	43	31	50	45
B	03	10	114	Bony bridge between malleus and EAC	No	Drilling out of the bony bridge	No	26	4	36	10
C	04	17	145	Malformed incus/ malformed stapes	No	—	TORP (malleus to stapes footplate)	38	0	53	8
D	05	18	169	Bony bridge between malleus and EAC	No	Drilling out of the bony bridge	No	23	0	30	5
E	06	10	23	Malformed stapes	No	—	Piston (incus to footplate)	35	5	46	19
F	07	12	121	Malformed stapes	No	—	Piston (incus to footplate)	23	30	35	44
G	08	12	129	Malformed TM (bony plate)/ malleus and incus absent/ malformed stapes	Yes	Drilling out of the bony plate underlying the TM and Silastic banding	TORP (TM to footplate)	49	11	68	29
H	09	9	29	Malleus ankylosis	No	Malleus relocation	TORP (malleus to footplate)	25	0	40	13
H	10	10	17	Malleus ankylosis/ malformed stapes	No	Malleus relocation	TORP (malleus to footplate)	21	3	36	16
I	11	10	100	Malleus ankylosis	No	—	PORP (malleus to stapes head)	28	16	41	30

Table I. Individual Surgical Findings and Audiometric Results for Teunissen and Cremers Class III Patients (Number of Procedures 517). (continued)

Patient	Case No.	Age,yr	Follow-up No.	Type of malformation	Previous Surgery for Microtia	Surgical technique	Ossiculoplasty	Pre ABG,dB	Post BG,dB	Pre AC,dB	Post AC,dB
J	12	7	12	Malformed incus/ malformed stapes	No	Malleus relocation	TORP (malleus to footplate)	46	0	58	14
K	13	11	82	Malformed TM (bony plate)/malformed malleus/ malformed incus	No	Drilling out of the bony bridge and Silastic banding	TORP (TM to footplate)	40	4	53	15
L	14	14	15	Malleus ankylosis No	No	Malleus relocation and Silastic banding	TORP (malleus to footplate)	20	0	25	10
M	15	18	—	Malformed incus/ malformed stapes (absence of superstructure)	No	Malleus relocation	TORP (malleus to footplate)	28	—	69	—
N	16	15	15	Malleus ankylosis	No	Malleus relocation and Silastic banding	TORP (malleus to footplate)	21	5	33	14
O	17	15	6	Malformed TM (bony plate)/ malformed incus	Yes	Drilling out of the bony bridge	TORP (malleus to footplate)	33	21	48	36

ABG = air-bone gap; AC = air conduction; EAC = external auditory canal; PORP = partial ossicular replacement prosthesis; Post = postoperative; Pre = preoperative; TM = tympanic membrane; TORP = total ossicular replacement prosthesis.

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on the stapes footplate with sufficient rigidity. This assembly facilitates an almost perpendicular position of the prosthesis from the footplate to the malleus handle and tympanic membrane. By keeping the distal end of the prosthesis in proper position using the Silastic band, the tendency of the prosthesis to tip or migrate is virtually eliminated. This combined technique of malleus relocation and Silastic band results in an increasingly stable and more functional ossicular arrangement.

A bony plate was present just underneath the tympanic membrane in three out of 17 cases (18%) (cases 8, 13, and 17). The bony plate was drilled out of the tympanic membrane using a Skeeter otologic drill with a 0.7-mm-diameter diamond dust burr. The remaining tympanic membrane was preserved. A simultaneous malformed incus was present in one of these cases (case 17) and ossiculoplasty was performed using a TORP (malleus-to-stapes footplate procedure). A simultaneous malformation of the malleus and incus was identified in a second case (case 13). Ossiculoplasty was performed in this case with a TORP with tympanic membrane-to-footplate assembly, in combination with the Silastic banding technique, as the malformed malleus could not be used. Simultaneous absence of the malleus and incus was present in a third case (case 8), and ossiculoplasty was performed with a TORP with tympanic membrane-to-footplate assembly in combination with the Silastic banding technique.

In one out of 17 cases (6%), a malleus malformation was identified in combination with an incus that was too short (case 1). The incus was removed and ossiculoplasty was performed using a PORP with malleus-to-stapes head assembly. The malleus relocation technique was used in this case. This case was operated before introduction of the silastic banding technique, and therefore a PORP was used instead of a TORP.

In three out of 17 cases (18%), middle ear exploration revealed fixation of the malleus due to a bony bridge between the posterior wall of the external auditory canal and the handle of the malleus (cases 2, 3, and 5). No other malformations were observed. The bony bridge was drilled out and the malleus remained mobile. The entire ossicular chain was preserved in all cases, and no ossiculoplasty was attempted.

There were two cases with a simultaneous incus and stapes malformation (cases 4 and 12). Ossiculoplasty was performed using a TORP with malleus-to-footplate assembly in both cases, and malleus relocation was performed in one case (case 12).

In one case, an incus malformation was associated with the absence of the stapes superstructure, and a TORP was positioned between the malleus and the mobile footplate (case 15). The malleus relocation technique was used in this case.

In two cases, the stapes was malformed and was not connected to the incus. Ossiculoplasty was performed using a 0.4-mm-diameter Teflon piston, which was positioned between the incus and the stapes footplate (cases 6 and 7).

No abnormal route of the facial nerve was observed in this series of class III patients.

Audiometric Assessment—Overall Results

Postoperative data were available for 16 cases (94%). The average follow-up duration was 67 months (range, 6–169 months). Significant postoperative sensorineural hearing loss (SNHL), defined as a change in the average BC threshold of 10 dB or more, was not observed in this series. Overall, the postoperative four-frequency ABG was closed to 10 dB or less in 10 out of 16 cases (63%) and closed to 20 dB or less in 12 out of 16 cases (75%) (Table 1, Table 2). The postoperative four-frequency average ABG was 10 dB (SD 11) compared with 31 dB (SD 10) preoperatively. The postoperative four-frequency average AC threshold was 23 dB (SD 16) compared with 45 dB (SD 12) preoperatively. Both ABG and AC threshold improved significantly, with mean gains of 21 dB (SD 14) and 22 dB (SD 15). Postoperatively, the four-frequency average BC threshold was 13 dB (SD 6) compared with 14 dB (SD 7) preoperatively; thus, the BC threshold deteriorated by 1 dB (SD 4).

The overall audiometric results of all individual cases were visualized using the Amsterdam hearing evaluation plots[5] shown in Figures 1 and 2.

Table 2. Overall Pure-Tone Audiometric Results for Teunissen and Cremers Class III Patients (Number of Procedures = 16).

Outcome	Preoperative	Postoperative	Difference
AC, mean (SD), dB	45 (12)	23 (16)	22 (15)a
BC, mean (SD), dB	14 (7)	13 (6)	1 (4)
ABG, mean (SD), dB	31 (10)	10 (11)	21 (14)a
ABG closure ≤10 dB, %	--	63	--
ABG closure ≤20 dB, %	--	75	--
SNHL >10 dB, %	--	0	--

Means were calculated using AC and BC thresholds at 0.5, 1, 2, and 4 kHz.
[a] Differences were statistically significant when using the related-samples Wilcoxon signed rank test with a significance level of .05.
ABG = air-bone gap; AC = air conduction; BC = bone conduction; SD = standard deviation; SNHL = sensorineural hearing loss.

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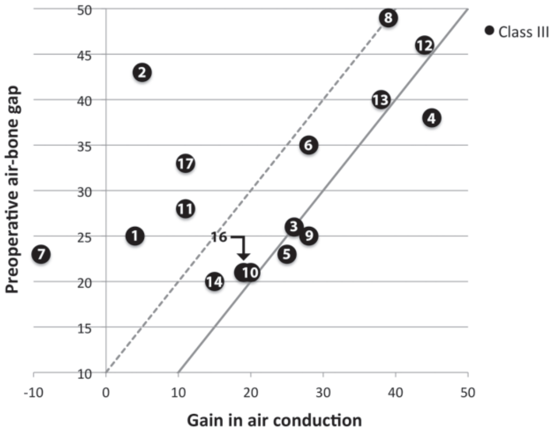

Figure 1. Amsterdam hearing evaluation plot. Each black dot represents one case. The solid diagonal line indicates total air-bone gap closure. The area between the diagonal lines indicates successful surgery with an air-bone gap of 10 dB or less. Audiometric data at last follow-up visit were used (16 cases).

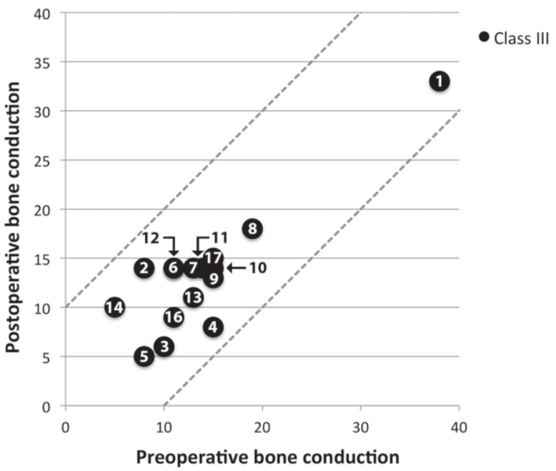

Figure 2. Amsterdam hearing evaluation plot. Each black dot represents one case. The two diagonal lines enclose the cases in which bone conduction did not change more than 10 dB. Audiometric data at last follow-up visit were used (16 cases).

Audiometric Assessment—At Least One-Year Follow-up

Postoperative data with at least one-year follow-up were available for 15 cases (88%) (Table 3). The average follow-up duration was 71 months (range, 12–169 months). The postoperative four-frequency ABG was closed to 10 dB or less in 10 out of 15 cases (67%) and closed to 20 dB or less in 12 out of 15 cases (80%).

Table 3. Pure-Tone Audiometric Results for Teunissen and Cremers Class III Patients With at Least 1-Year Follow-up (Number of Procedures = 15).

Outcome	Preoperative	Postoperative	Difference
AC, mean (SD), dB	44 (12)	22 (16)	22 (15)a
BC, mean (SD), dB	14 (7)	13 (6)	1 (4)
ABG, mean (SD), dB	31 (9)	10 (11)	22 (15)a
ABG closure ≤10 dB, %	--	67	--
ABG closure ≤20 dB, %	--	80	--
SNHL >10 dB, %	--	0	--

Means were calculated using AC and BC thresholds at 0.5, 1, 2, and 4 kHz.
[a] Differences were statistically significant when using the related-samples Wilcoxon signed rank test with a significance level of .05.
ABG = air-bone gap; AC = air conduction; BC = bone conduction; SD = standard deviation; SNHL = sensorineural hearing loss.

3

The postoperative four-frequency average ABG was 10 dB (SD 11) compared with 31 dB (SD 9) preoperatively. The postoperative four-frequency average AC threshold was 22 dB (SD 16) compared with 44 dB (SD 12) preoperatively. Both the ABG and the average AC threshold improved significantly, with a mean gain of 22 dB (SD 15) and 22 dB (SD 15), respectively. Postoperatively, the four-frequency average BC threshold was 13 dB (SD 6) compared with 14 dB (SD 7) preoperatively.

Failures

ABG closure to 20 dB or less was not achieved in four cases (25%). In all of these cases, revision surgery was carried out. Two of these patients (cases 1 and 7) underwent revision surgery when they were still under 18 years of age, whereas the other patients (cases 2 and 17) showed a delayed failure and were over 18 years when they underwent their revision surgery. In the first case (case 1), the previous prosthesis (PORP) was found to be dislocated during revision surgery. A TORP was placed during the revision surgery and the Silastic banding technique was performed. In one case (case 2), the malleus was dislocated. A TORP was used to reconstruct the ossicular chain in combination with the Silastic banding technique. In one case (case 7), the incus was eroded. Ossiculoplasty using a TORP and malleus relocation were performed. In the last case (case 17), the tympanic membrane was perforated. The previously placed prosthesis was left in place, and a transcanal myringoplasty type I was performed using a tragal perichondrium graft. The ABG was closed to within 20 dB in all four of these cases at the time of the last follow-up (mean, 39 months; range, 23–54 months).

Table 4. Reported Pure-Tone Audiometric Results Following Surgical Treatment of Teunissen and Cremers Class III Patients in the Literature.

First author (year)	No. of cases	Classification	Mean follow-up duration, mo (range)	%ABG closure <10 dB	%ABG closure <20 dB	Post ABG, Mean (SD)	Post AC, Mean (SD)	Gain AC, Mean (SD)	% SNHL > 10 dB
Funasaka (11)a	11	None, mobile stapes	>18	NR	NR	NR	29 (11)	30 (15)	NR
Hashimoto (2002) 12b	21	Own classification, group I	> 24	NR	95‡	7 (NR)	18 (NR)	37 (NR)	NR
	4	Own classification, group III		NR	75‡	6 (NR)	18 (NR)	37 (NR)	NR
Park (2009)13d63	13	PC type II	>3	NR	85	NR	NR	NR	NR
Philippon (2013)14e	16	None, mobile stapes	>6	25	63	19 (11)	29 (13)	19 (13)	6
Raveh (2002)15d	22	None, mobile stapes	>12	14	28	NR	NR	NR	0
Sakamoto (2011)16e	41	PC type I and II	>12	NR	56	NR	40 (24)	19 (14)	NR
Teunissen (1993)3, 17b	31	TC class III	12	29	74	NR	29 (17)f	19 (16)f	3
Thomeer (2012)18e	23	TC class III	12	NR	65	19 (12)	30 (17)	16 (17)	9
	22		48 (11–175)	NR	65	20 (9)	30 (15)	16 (16)	9
This studye	16	TC class III	67 (6–169)	63	75	10 (11)	22 (16)	22 (15)	0

DISCUSSION

In this study, we evaluated the hearing outcomes after middle ear surgery for congenital ossicular malformations with a mobile stapes in 16 pediatric cases. Overall, a postoperative ABG closure to within 10 dB or less was achieved in 63%. A postoperative ABG closure to within 20 dB or less was achieved in 75%. Postoperative sensorineural hearing loss did not occur in any of the cases.

A rapid systematic literature search identified nine articles[3,11-18] reporting audiometric results following middle ear surgery in children with class III congenital middle ear malformations with a mobile stapes (Table 4). In two of these,[3,17] a large portion of the included cases overlapped. The results of these articles were combined and were considered to be part of one study. We included only those studies that analyzed at least 10 cases and were written in English, Dutch, French, or German. ABG closure to 10 dB or less was reached in 14% to 29% of the cases in three studies.[3,14,15,17] ABG closure to 20 dB or less was reported in seven studies[3, 12-18] and was reached in 28% to 95% of the cases. The percentages of ABG closure to within 20 dB found in this study accord well to those described in the literature. However, when comparing the percentage of cases with an ABG closure to 10 dB or less, our series shows a substantially higher success rate.

Several studies have compared the hearing outcome of middle ear surgery for congenital ossicular malformations with a mobile stapes (Teunissen and Cremers class III) with the outcome of surgical treatment for other congenital middle ear malformations (Teunissen and Cremers class I and class II). The reported ABG closure following surgical treatment in patients with Teunissen and Cremers class I and II to 10 dB or less and to 20 dB or less range from 17% to 73% and 25% to 80%, respectively.[3,13,15,19-22] One study[13] showed a comparable outcome in patients with a mobile stapes footplate (84.6%) and patients with an isolated stapes footplate fixation (80.4%). However, if the stapes footplate fixation was associated with other middle ear malformations, the hearing outcome was significantly lower in this study (30%). Another study[15] found a better outcome for patients with a mobile stapes compared to those with a fixed stapes.

In an earlier study, the results of which have not been published yet, conducted in our center, the hearing outcome of 45 cases with Teunissen and Cremers class I and II congenital malformations were analyzed. Success rates were comparable to that of class III malformations, showing an ABG closure of 10 dB or less in 73% (class I) and 50% (class II) of all cases and a closure to 20 dB or less in 77% (class I) and 67% (class II) of all cases. Overall, the success rate seems to be highest in class I and III malformations.

The occurrence of postoperative SNHL was reported in four studies,[3,14,15,17,18] and varied from 0% to 9%. SNHL was defined as deterioration in mean BC threshold of more than 10 dB. In our series, none of the patients showed postoperative SNHL exceeding 10 dB. The incidence of SNHL in patients with class III congenital malformation is comparable to that of patients with class I and II congenital malformations, with an incidence reported of less than 10% in the literature.[19-23]

CONCLUSION

The results of stapedotomy in children with congenital ossicular malformation and a mobile stapes are satisfactory, with ABG closure to within 10 dB or less in 63% to within 20 dB or less in 75% of cases. Postoperative sensorineural hearing loss did not occur in any of the cases

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Chapter 4

Congenital oval or round window malformations in children: Surgical findings and results in 17 cases

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ABSTRACT

Objectives/Hypothesis: To prospectively evaluate surgical findings and hearing results in children undergoing surgery for congenital oval or round window malformations (class IV malformations).

Study Design: A nonrandomized, nonblinded, case series of prospectively collected data.

Methods: Fourteen consecutive pediatric patients who underwent 17 surgical procedures for congenital oval or round window malformations in a tertiary referral center were included. Postoperative pure-tone audiometry was available in 15 cases. The surgical technique for repair of the ossicular chain was dictated by the surgical findings at the time of surgery. The majority of the cases underwent ossiculoplasty using a Teflon piston, bucket-handle prosthesis, or total ossicular replacement prosthesis. Associated surgical techniques included malleus relocation and oval window drill-out procedure. The main outcome measures were preoperative and postoperative hearing status using four-frequency (0.5, 1, 2, and 4 kHz) audiometry. Air-conduction thresholds, bone-conduction thresholds, and air-bone gap were measured. Postoperative audiometry was performed at 3, 6, 9, 12, 18, and 24 months after surgery and at a yearly interval thereafter.

Results: Postoperative air-bone gap closure to 10 dB or less was achieved in 47%. A postoperative air-bone gap closure to within 20 dB or less was achieved in 60%. Postoperative sensorineural hearing loss did not occur in this series.

Conclusions: Middle ear surgery for class IV abnormalities is feasible, but success percentages are much lower compared to other types of congenital ossicular malformations. Surgeons should be particularly careful in case of facial nerve abnormalities on computed tomography or during middle ear exploration.

INTRODUCTION

Congenital middle ear malformations account for approximately 0.5% to 3% of all cases with conductive hearing loss.[1,2] These malformations can be divided in major and minor anomalies. In case of involvement of the tympanic membrane and external ear, this is referred to as a major anomaly. Minor middle ear malformations can be classified according to the Teunissen and Cremers classification,[3] based on the different surgical reconstruction possibilities: (class I) isolated congenital stapes ankylosis, (class II) stapes ankylosis associated with other ossicular malformations, (class III) congenital malformations of the ossicular chain with a mobile stapes footplate, and (class IV) severe 1) aplasia or 2) dysplasia of the oval or round window. Among these minor congenital middle ear malformations, class IV malformations are most uncommon (<10%).[3]

The facial nerve, stapes, and otic capsule all develop from the second branchial arch derivates. One widely accepted theory suggests that the developing facial nerve is displaced anteriorly, preventing the stapes and otic capsule to make contact, resulting in malformation of both the oval window and the stapes.[4,5] Because of the association with facial nerve anomalies or an aberrant course of the facial nerve, surgical intervention can be challenging. Surgical intervention consists of the performance of a stapedotomy, oval window drill-out procedure, or fenestration of the promontory or lateral semicircular canal fenestration, if possible followed by ossicular reconstruction.[6-8] Alternatives to surgical treatment are conventional hearing aids or osseointegrated implants.[6-8]

The objective of this study of prospectively collected data was to evaluate surgical findings and hearing results in children undergoing middle ear surgery for congenital oval or round window malformations (class IV malformations).

MATERIALS AND METHODS

Between January 1991 and October 2014, 225 operations for congenital malformations of the middle ear were performed by the same surgeon (first author) in the same tertiary referral center. Of these 225 operations, 69 were performed in children, and congenital oval or round window malformations were identified in 17 of these cases. The remaining 52 cases underwent surgery for class I, class II, and class III congenital malformations. The outcomes of surgery in these patients were described elsewhere.[9,10]

Study Population

This is a nonrandomized, nonblinded, study of prospectively collected data of 14 children, under 18 years of age, who underwent 17 consecutive operations for class IV congenital middle ear malformations from 1993 to 2014. An international board review was not required for this study. Whenever we refer to cases in this article, we refer to operations and not patients. Of the 14 included children, three had sequential bilateral surgery. Postoperative pure-tone audiometry was available for 15 of these cases (88%). Children diagnosed with juvenile otosclerosis or osteogenesis imperfecta were excluded, as they have their own approach and outcome, the results of which are described elsewhere.[11,12]

Surgery

A transcanal procedure with facial nerve monitoring using the NIM 2.0 (Medtronic, Inc., Jacksonville, FL) was performed in all cases in this series. The anatomy and mobility of the ossicular chain were carefully checked. The facial nerve was identified and its relationship with the ossicular chain and/or stapes remnant assessed.[13] The surgical technique for repair of the ossicular chain was dictated by the surgical findings at the time of surgery and was described according to each Teunissen and Cremers class.[3] Surgical data were collected prospectively using the Otology–Neurotology Database (ONDB) (AS Multimedia, Inc., Cassagne, France).[13]

Audiometric Assessment

Hearing status was assessed before and at 3, 6, 9, 12, 18, and 24 months after surgery and at a yearly interval thereafter. Audiometric evaluation included pre- and postoperative bone-conduction (BC) thresholds, air-conduction (AC) thresholds, and air-bone gaps (ABGs). Furthermore, closure of the ABG to 10 dB or less was evaluated, as well as ABG closure to 20 dB or less. BC and AC thresholds that were obtained at the same time postoperatively were used for calculation of the mean postoperative ABG. Four-frequency (0.5, 1, 2, and 4 kHz) pure-tone averages for BC and AC thresholds, obtained at the last follow-up visit, were used in this study. Audiometry was reported according to American Academy of Otolaryngology–Head and Neck Surgery guidelines,[14] with the exception of thresholds at 3 kHz, which were substituted with those at 4 kHz in all cases. The audiometric results of all individual cases were visualized using the Amsterdam Hearing Evaluation Plots.[15] Audiometric data were collected prospectively using the ONDB.[13]

Statistical Analyses

Means, standard deviations (SDs), and percentages were calculated. Preoperative and postoperative BC thresholds, AC thresholds, and ABGs were compared using the nonparametric related-samples Wilcoxon signed rank test. All analyses were performed using SPSS version 20.0 (SPSS Inc., Chicago, IL).

RESULTS

Preoperative and Surgical Findings

Twelve cases underwent primary surgery, three cases revision surgery following primary surgery elsewhere, and two cases underwent second stage surgery (see Supporting Table 1 in the online version of this article). Both second-stage procedures were preceded by reconstructive surgery for microtia performed by another surgeon. Six procedures were performed on the right ear (35%) and 11 on the left ear (65%). Sex ratio was 41% female and 59% male. Mean age at the time of surgery was 12 years, with a range of 5 to 17 years.

Table 1. Overall Pure-Tone Audiometric Results for Teunissen and Cremers Class IV Patients (Number of Procedures = 15).

Outcome	Pre-operative	Post-operative	Difference	P Value (Related-Samples Wilcoxon Signed Rank Test)
AC, mean (SD), dB	57 (12)	37 (22)	21 (17)a	0.003a
BC, mean (SD), dB	16 (11)	17 (12)	−1 (6)	0.575
ABG, mean (SD), dB	41 (13)	20 (21)	21 (17)a	0.002a
ABG closure ≤10 dB, n [%]	—	7 [47]	—	—
ABG closure ≤20 dB, n [%]	—	9 [60]	—	—
SNHL >15 dB, n [%]	—	0 [0]	—	—

Means were calculated using AC and BC thresholds at 0.5, 1, 2, and 4 kHz.
[a] Differences were statistically significant when using the related-samples Wilcoxon signed rank test with a significance level of .05.
ABG = air-bone gap; AC = air conduction; BC = bone conduction; SD = standard deviation; SNHL = sensorineural hearing loss.

Oval window aplasia (class IVa) was observed in one out of 17 cases (6%) (case 6). An oval window depression could not be identified, in accordance with a previous report[16] on patients with congenital absence of the oval window. There was a concomitant absence of the round window, and all three ossicles were missing. Fenestration was not attempted in this case, and the surgery was aborted without performing ossicular reconstruction.

Oval window dysplasia (class IVb) was observed in 16 out of 17 cases (94%), and was defined as the presence of a depression clearly demarcating an oval window area,4 without an identifiable footplate or annular ligament[17] (Figure 1). A complete osseous obliteration of the oval window was identified in 12 out of 17 cases (71%) and a malformed oval window niche without osseous obliteration in four cases (24%). In one out of 12 cases with osseous obliteration of the oval window, surgery was aborted without performing an ossicular reconstruction (case 15). The facial nerve coursed across the oval window, obstructing it completely (Figure 2). An oval window drill-out procedure was performed to fenestrate the vestibule in the other 11 cases. The Skeeter microdrill was used to thin the bone of the oval window area and create a 0.8-mm-diameter fenestration followed by a vein graft interposition over the fenestration. A Teflon piston, with incus-to-stapedotomy assembly, was placed in the presence of a normal malleus and incus (six cases). In one case, the incus was too short and a bucket-type Teflon prosthesis was used. A total ossicular replacement prosthesis (TORP), with malleus-tostapedotomy assembly, was used in case of a simultaneous malleus and incus malformation (four cases). In one patient (case 2 and 3) the malleus and incus were fused in both ears. In another case (case 4), the malleus was malformed, and both the incus and stapes were absent. In the last case (case 7), both the malleus and incus were malformed.

In four out of 17 cases (24%), the oval window could be identified, but the footplate was malformed. The footplate was abnormally small and fixed in the first case (case 5). The malleus and incus were fused, and a malformed, monopodal stapes was found in this case. A stapedotomy and ossiculoplasty were performed using a TORP with malleus-to-stapedotomy assembly. In the second case (case 8), a rudimentary, monopodal stapes was present in combination with a dysplastic footplate, which was small but mobile. The footplate was preserved, and ossiculoplasty was performed using a 0.4-mm-diameter Teflon piston, which was positioned in between the incus and the mobile footplate. In the third case (case 9), the posterior half of the malformed footplate was fixed, whereas the anterior half was mobile. The stapes consisted of an amorphous piece of bone attached to the fixed posterior half of the footplate. The malformed superstructure was drilled out, and ossiculoplasty was performed using a 0.4-mm-diameter Teflon piston, which was positioned in between the incus and the mobile part of the footplate. In the fourth case (case 14), the footplate was dysplastic and fixed, and surrounded by a membrane (Figure 3). A rudimentary stapes was connected to the fixed footplate. A regular 0.8-mm stapedotomy with vein graft interposition was performed, and ossicular reconstruction was performed using a bucket-type Teflon prosthesis with incus-to-stapedotomy assembly.

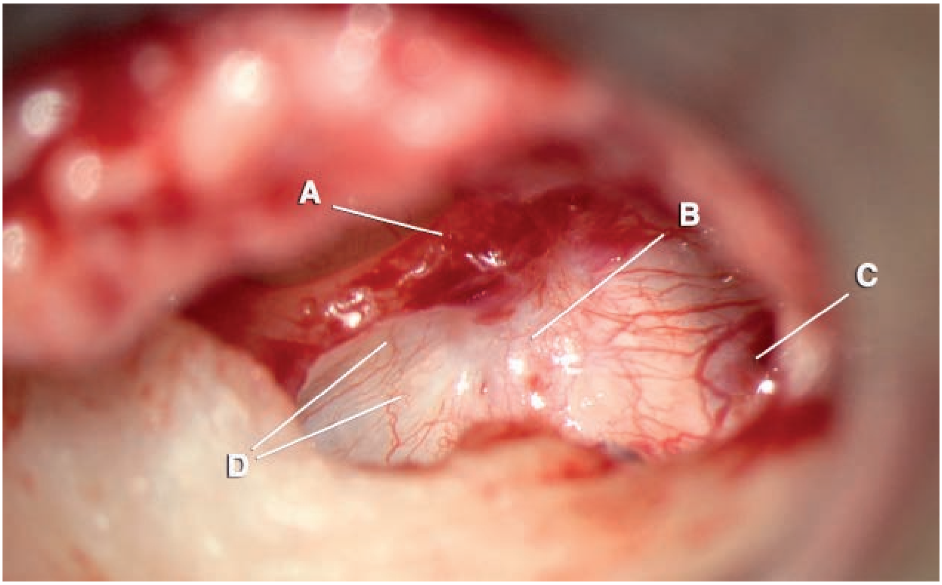

Figure 1. Operative view of a right oval window dysplasia (class IVb) showing a depression clearly demarcating an oval window area without an identifiable footplate or annular ligament. The stapes is absent. A = incus; B = oval window area; C = round window; D = facial nerve. [Color figure can be viewed in the online issue, which is available at www.laryngoscope.com.]

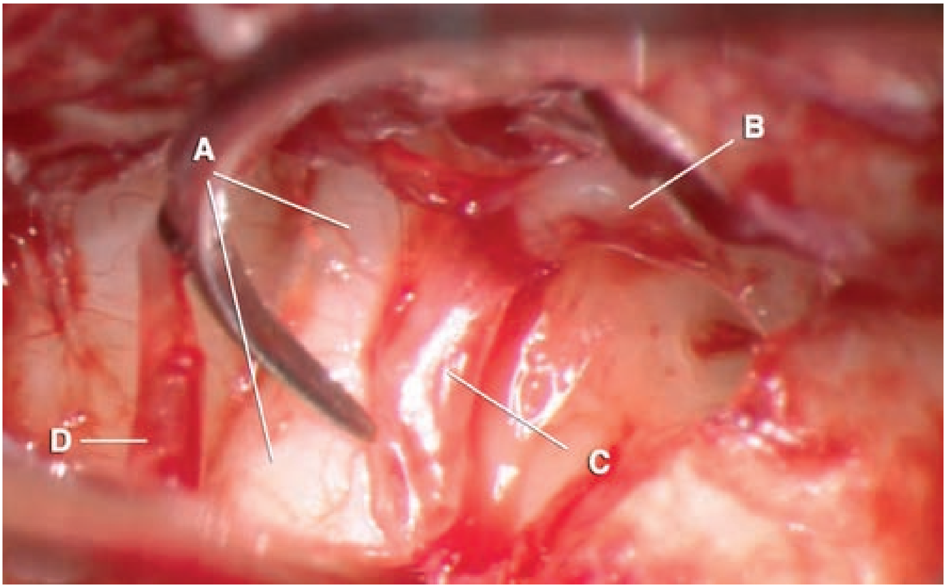

Figure 2. Operative view of a left oval window dysplasia. The facial nerve courses across the oval window, obstructing it completely. A = facial nerve; B = incus; C = stapes superstructure; D = chorda tympani. [Color figure can be viewed in the online issue, which is available at www.laryngoscope.com.]

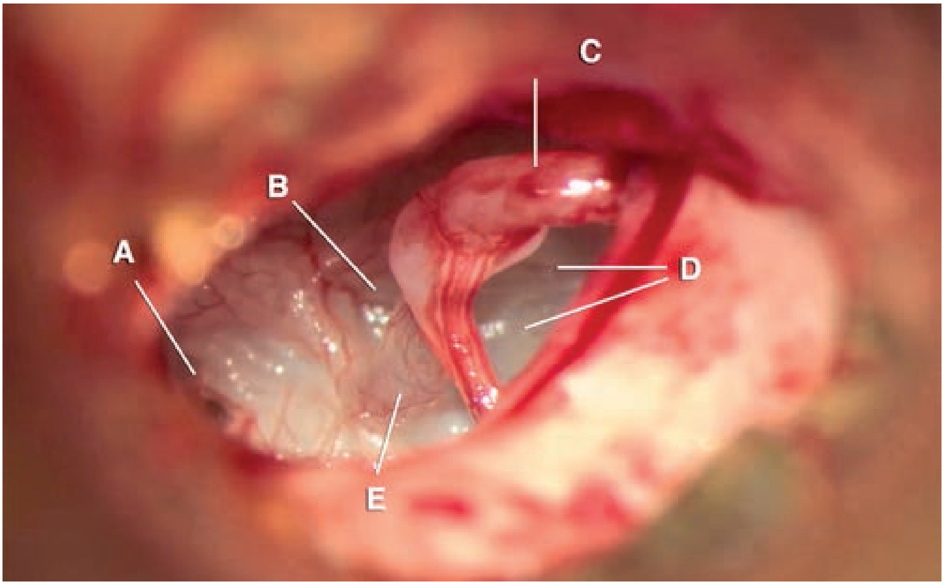

Figure 3. Operative view of a left ear showing a small, dysplastic, and fixed footplate surrounded by a membrane. A = round window; B = dysplastic and fixed footplate; C = incus; D = membrane surrounding the footplate; E = stapes superstructure (posterior crus). [Color figure can be viewed in the online issue, which is available at www.laryngoscope.com.]

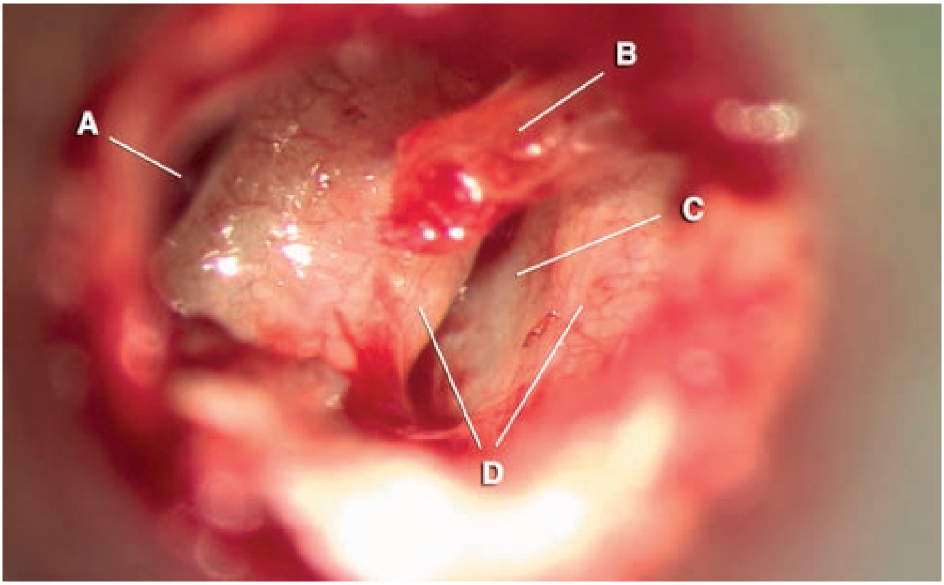

Figure 4. Operative view of a left ear showing a congenital duplication of the facial nerve. One segment courses over the promontory. The second segment appears to be in the normal facial nerve position. The stapes superstructure was not connected to the oval window and was removed. A = round window; B = incus; C = osseous obliteration of the oval window; D = congenital duplication of the facial nerve. [Color figure can be viewed in the online issue, which is available at www.laryngoscope.com.]

An abnormal route of the facial nerve was observed in eight out of 17 cases (47%). Seven of these cases (cases 7, 10–13, 15, 17) presented with an osseous obliteration of the oval window in combination with a discontinuity of the stapes superstructure and oval window. Both the malleus and incus were normal in all seven cases. The facial nerve coursed over the oval window in four cases and over the promontory in one case. A congenital duplication of the facial nerve, dividing the nerve in two segments, was present in three cases. One segment coursed over the promontory, whereas the second segment was in the normal facial nerve position (Figure 4). In one of the eight cases (case 16), the oval window was obliterated and the stapes was absent (Figure 1). The facial nerve was dehiscent in all cases. It was possible to fenestrate the footplate superior to the facial nerve in seven cases. In one case (case 15), fenestration was not attempted, and the surgery was aborted because the facial nerve obstructed the oval window totally.

One case of perilymph gusher was observed in this class IV group (case 7). This case was treated with stapedotomy with vein graft interposition. Gelfoam was used to fill the oval window and keep the vein graft in its proper position.

A syndrome was diagnosed in two patients. Both patients were operated bilaterally. Branchio-oto-renal (BOR) syndrome was diagnosed in one patient (case 12 and 13). Similar malformations were identified in both ears: osseous obliteration of the oval window and a discontinuity of the stapes superstructure and footplate. The facial nerve followed an abnormal route, coursing over the promontory in between the round and oval window. Both the malleus and incus were normal in both ears. Treacher Collins syndrome was diagnosed in the other patient (case 4 and 5). The types of malformations were not the same for both ears. The incus was malformed and the stapes was missing in one ear. A combination of malleus ankylosis and a malformed incus, which was not connected to a monopodal stapes, was present in the contralateral ear.

Audiometric Assessment—Overall Results

Postoperative data were available for 15 cases (88%) (Table 1) (see also Supporting Table 1 in the online version of this article). The average follow-up duration was 46 months with a range of 3 to 131 months. The postoperative mean ABG was 20 dB (SD 21) compared with 41 dB (SD 13) preoperatively. The ABGs improved significantly with a mean gain of 21 dB (SD 17, P = .002 using the related-samples Wilcoxon signed rank test). The postoperative mean AC threshold was 37 dB (SD 22) compared with 57 dB (SD 12) preoperatively. The AC threshold improved significantly with a mean gain of 21 dB (SD 17, *P* = .003). The postoperative mean BC threshold was

17 dB (SD 12) compared with 16 dB (SD 11) preoperatively; thus, the BC threshold deteriorated with 1 dB (SD 6, *P* = .575). This difference was not statistically significant, nor was it clinically relevant. Overall, the postoperative four-frequency ABG was closed to 10 dB or less in 47% and to 20 dB or less in 60% of the 15 cases with postoperative pure-tone audiometry. Significant postoperative sensorineural hearing loss (SNHL), defined as a change in the average BC threshold of 15 dB or more, was not observed in this series.

The audiometric results of all individual cases are visualized using the Amsterdam Hearing Evaluation Plots[15] in Figure 5 and Figure 6.

Audiometric Assessment—At Least 1-Year Follow-up

Postoperative data with at least 1-year follow-up were available for 12 cases (71%) (see Supporting Table 2 in the online version of this article). The average follow-up duration was 57 months, with a range of 24 to 131 months. The postoperative mean ABG was 18 dB (SD 20) compared with 42 dB (SD 11) preoperatively. The postoperative mean AC threshold was 35 dB (SD 22) compared with 59 dB (SD 11) preoperatively. Both the ABG and the average AC threshold improved significantly, with a mean gain of 24 dB (SD 18, *P* = .006) and 24 dB (SD 16, *P* = .004), respectively. Postoperatively, the mean BC threshold was 17 dB (SD 13) compared with 17 dB (SD 12) preoperatively. The postoperative mean ABG was closed to 10 dB or less in 50% and to 20 dB or less in 67% of the included 12 cases with at least 1-year pure-tone audiometric follow-up.

Failures

ABG closure to 20 dB or less was not achieved in six cases (40%). In two of these cases (cases 5 and 11), revision surgery was carried out. In the first case (case 5), the previously placed TORP was too short and was replaced by a TORP of the appropriate size. In the second case (case 11), the previously placed Teflon piston was displaced. During revision stapedotomy, a TORP was placed instead. (Re)obliteration of the oval window was not observed in these two cases. The ABG was closed to within 20 dB in both cases at the time of last follow-up (87 and 78 months, respectively).

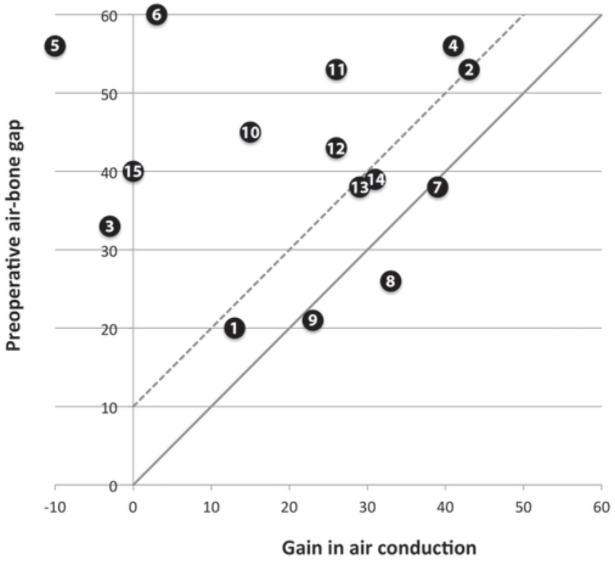

Figure 5. Amsterdam Hearing Evaluation Plot. Each black dot represents one case. The solid diagonal line indicates total air-bone gap closure. The area between the diagonal lines indicates successful surgery with an air-bone gap of 10 dB or less. Audiometric data at last follow-up were used (15 cases).

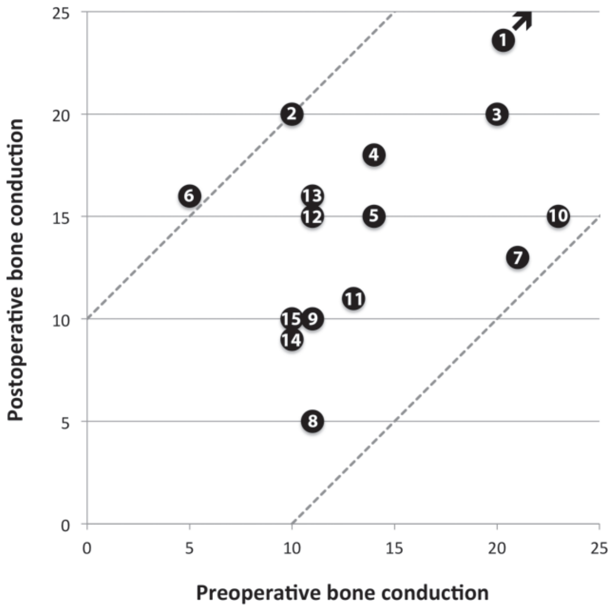

Figure 6. Amsterdam Hearing Evaluation Plot. Each black dot represents one case. The two diagonal lines enclose the cases in which bone conduction did not change more than 10 dB. Audiometric data at last follow-up were used (15 cases).

Table 2. Reported pure-tone audiometric results following surgical treatment of Teunissen and Cremers class IV patients in literature

Author (year)	Classification	Mean follow-up duration, mo (range)	No. of cases	%ABG closure <10 dB	%ABG closure <20 dB	Post ABG, Mean (SD)	Post AC, Mean (SD)	Gain AC, Mean (SD)	% SNHL > 10 dB
de Alarcon (2008)[6]	None (CAOW)	1 (NR)a	13	NR	NR	23 (NR)	31 (NR)	30 (NR)	0
		20 (6–47)a	10	NR	NR	31 (NR)	43 (NR)	18 (NR)	0b
Ashtiani (2010)[18]	None (CAOW or abnormal facial nerve course)	62 (12–125) a	22	NR	NR	19 (6)	25 (9)	33 (NR)	0c
Jahrsdoerfer (1980)[19]	None (CAOW)	NRd	6	17	67	NR	NR	NR	NR
Su (2014)[7]	None (CAOW)	NRa	56	NR	NR	31 (NR)	49 (NR)	18 (NR)	0
Thomeer (2012)[8]	TC class IV	1 (NR)a	8	0	25	30 (14)	55 (35)	5 (37)	25
		115 (12–226)a	8	0	13	43 (11)	68 (28)	−9 (30)	38
This study	TC class IV	67 (6–169)a	15	47	60	20 (21)	37 (22)	21 (17)	0

[a] 0.5, 1, 2, and 4 kHz.
[b] Bone conduction remained normal in all patients.
[c] None of the included patients suffered from postoperative SNHL, not otherwise specified.
[d] Frequencies unknown.
ABG = air-bone gap; AC = air conduction; CAOW = congenital absence of the oval window; NR = not reported; SD = standard deviation; SNHL = sensorineural hearing loss; TC = Teunissen and Cremers classification.

DISCUSSION

In this study, we evaluated the surgical findings (17 cases) and hearing outcomes following middle ear surgery (15 cases) for congenital oval or round window malformations (class IV malformations) in pediatric cases. Overall, a postoperative ABG closure to 10 dB or less was achieved in 47%. A postoperative ABG closure to within 20 dB or less was achieved in 60%. Postoperative SNHL did not occur in this series.

The outcomes of surgery in patients with class I, class II, and class III congenital malformations were described previously.[9,10] A postoperative ABG closure to 10 dB or less was achieved in 73% of class I cases, 50% of class II cases, and in 63% of class III cases. A postoperative ABG closure to 20 dB or less was achieved in 77% of class I cases, 67% of class II cases, and in 75% of class III cases. Even though middle ear surgery for class IV abnormalities is feasible, success percentages are lower compared to other types of congenital ossicular malformations. Given the lower success percentages, one might argue for treating class IV malformations with hearing aids instead of surgery. The decision-making process in this series involved case-by-case discussions with the patients and their families. The surgeon's experience has led him to believe that many children in France do not like wearing hearing aids over time. Knowing that hearing loss has negative effects on school performance, especially in case of bilateral hearing loss, the patients that are discussed in this article and their families opted for surgery.

We conducted a rapid systematic literature search, identifying five articles[6-8,18,19] reporting audiometric results after middle ear surgery in children with congenital oval or round window malformations (Table 2). Studies were included if at least five cases were analyzed, and if they were written in English, Dutch, French, or German. Only two of the analyzed studies reported the percentage of cases with postoperative ABG closure. One study[20] reported an ABG closure to 10 dB or less in 17% and an ABG closure to 20 dB or less in 67%. In the other study,[8] ABG closure to 10 dB or less was achieved in none of the included cases, and ABG closure to 20 dB or less was achieved in 25% at 1-month follow-up. At long-term follow-up (mean, 67 months; range, 6–169 months), again no cases achieved ABG closure to 10 dB or less, and the percentage of ABG closure to 20 dB or less declined to 13%. Postoperative mean gain in AC threshold was described by all but one article[18] and varied from −9 dB to 33 dB.[6-8,18] Two of these studies[6,8] compared short- and long-term mean gain in AC threshold, showing a deterioration of the AC threshold of 12 dB and 14 dB over time, respectively. Several authors found bone regrowth in the oval window area during revision surgery, which may explain deterioration of the mean AC threshold over time.[6,8] These findings suggest that the otic capsule is capable of regenerating new bone matrix, possibly as a reaction to traumatic insult, such as the oval window drill-out procedure.6,20 However, Lambert[14] revised three cases with postoperative AC threshold deterioration, and found that the vestibulotomies were patent, with no occurrence of bone regrowth. Another possible explanation for the postoperative hearing loss is limited prosthesis mobility in cases with an overlying facial nerve, placed against the prosthesis.[8,19] A dislocated piston causing AC threshold deterioration after initial postoperative improvement has been described in two studies.[6,8]

Approximately 25% of all types of congenital ossicular malformations are associated with a genetic syndrome.[21] Goldenhar syndrome, Möbius syndrome, Saethre Chotzen syndrome, BOR syndrome, Treacher Collins syndrome, and hemifacial microsomia have been previously associated with congenital oval or round window malformations.[6,8,19] Thomeer et al.[8] reported a prevalence of 40% of syndrome-associated class IV malformations. It has been suggested that malformations are more severe when they are associated with a branchiogenic syndrome.[21] Nonetheless, the only case in which ABG closure to within 20 dB was reached in the study by Thomeer et al.[8] suffered from BOR syndrome. In our study, two cases were associated with BOR syndrome and two cases with Treacher Collins syndrome. Both cases with BOR syndrome and one case with Treacher Collins syndrome reached ABG closure to within 20 dB. The second case with Treacher Collins syndrome was less successful, with a postoperative mean ABG of 65 dB.

Facial nerve anomalies are found in 59% to 76% of patients with congenital round or oval window malformations.[6-8,19] These include partial or complete overlay over the oval window or promontory, dehiscence or partial absence, and bifid facial nerves.[7,22,23] In this series, eight cases presented with an abnormal course of the facial nerve, all of which were accompanied by an absent or malformed stapes and a dysplastic oval window. This triad of abnormalities may be explained from an embryologic point of view. As stated in the introduction, a displacement of the facial nerve in the early stages of development may prevent normal stapes and oval window formation.[4,5] It can be difficult to adequately assess the course of the facial nerve during middle ear exploration. Malformation of the stapes, a lack of contact between the stapes and oval window, and oval window dysplasia are more easily identified. Any surgeon should be aware of a risk of abnormal facial nerve routing if a simultaneous stapes to oval window discontinuity and oval window dysplasia is identified during middle ear exploration.

CONCLUSION

Middle ear surgery for class IV abnormalities is feasible, but success percentages are much lower compared to other types of congenital ossicular malformations. Surgeons should be particularly careful in case of facial nerve abnormalities on computed tomography or during middle ear exploration.

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Part II

Stapedotomy in Osteogenesis Imperfecta

Chapter 5

Stapedotomy in osteogenesis imperfecta: a prospective study of 32 consecutive cases

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ABSTRACT

Objective: To prospectively evaluate hearing outcomes in patients with Osteogenesis Imperfecta undergoing primary stapes surgery and to isolate prognostic factors for success.

Study Design: A nonrandomized, open, prospective case series.

Setting: A tertiary referral center.

Patients: Twenty-five consecutive patients who underwent 32 primary stapedotomies for Osteogenesis Imperfecta with evidence of stapes fixation and available postoperative pure-tone audiometry.

Intervention: Primary stapedotomy with vein graft interposition and reconstruction with a regular Teflon piston or bucket handle-type piston.

Main Outcome Measures: Preoperative and postoperative audiometric evaluation using conventional 4-frequency (0.5, 1, 2, and 4 kHz) audiometry. Air-conduction thresholds, bone-conduction thresholds, and air-bone gap were measured. The overall audiometric results as well as the results of audiometric evaluation at 3 months and at least 1 year after surgery were used.

Results: Overall, postoperative air-bone gap closure to within 10 dB was achieved in 88% of cases. Mean (standard deviation) gain in air-conduction threshold was 22 (9.4) dB for the entire case series, and mean (standard deviation) air-bone gap closure was 22 (9.0) dB. Backward multivariate logistic regression showed that a model with preoperative air-bone gap closure and intraoperatively established incus length accurately predicts success after primary stapes surgery.

Conclusion: Stapes surgery is a feasible and safe treatment option in patients with Osteogenesis Imperfecta. Success is associated with preoperative air-bone gap and intraoperatively established incus length.

Osteogenesis Imperfecta is a generalized disorder of the connective tissue, resulting in bone fragility and susceptibility to fractures. Other nonosseous abnormalities include blue sclera and hearing loss. Four major types of Osteogenesis Imperfecta have been recognized[1]. The classic triad of spontaneous fractures, blue sclera, and hearing loss is known as the Van der Hoeve–de Kleyn syndrome[2]. Of all patients with osteogenesis imperfecta, 45% to 80% experience hearing loss[3-5]. Although, historically, hearing loss has been reported to be most common in type 1 osteogenesis imperfecta and rare in type 4 osteogenesis imperfecta[6], a recent study suggests that hearing loss develops independently of the underlying genotype[7]. Hearing loss may be conductive, sensorineural, or mixed. Treatment options include the use of hearing aids or stapes surgery for conductive hearing loss and cochlear implants or bone-anchored hearing aids for sensorineural hearing loss[8]. Stapes surgery has been considered to be more risky in osteogenesis imperfecta until the 1970s. Since then, several authors have published the results of relatively large case series of patients with osteogenesis imperfecta undergoing stapes surgery. Results of stapes surgery in patients with osteogenesis imperfecta show closure of the air-bone gap to within 10 dB in 75% to 85% of patients[9-13] compared with the results of up to 90% to 95% of patients with otosclerosis[14]. The objectives of this study were to prospectively evaluate hearing outcomes in patients with osteogenesis imperfecta undergoing primary stapes surgery and to isolate prognostic factors for success.

MATERIALS AND METHODS

Patients

This is a nonrandomized, open, prospective case series of 25 consecutive patients who underwent 32 primary stapedotomies from 1994 to 2013 for Osteogenesis Imperfecta with evidence of stapes fixation. A total of 46 procedures for stapes fixation in patients with Osteogenesis Imperfecta have been carried out in a tertiary referral center, 10 of which (21.7%) were revision stapedotomies. No postoperative pure-tone audiometry was available for another 4 (8.7%) of these 46 cases. Revision procedures and patients lacking postoperative pure-tone audiometry were excluded. All patients included in this analysis suffered from Osteogenesis Imperfecta type 1.

Surgery

All procedures were performed by the same senior ENT surgeon (R. V.) in a tertiary referral center. In all cases, a transcanal procedure was undertaken. Ossicular mobility was assessed by gentle palpation of the stapes head with a curved needle after incudostapedial joint separation. Stapedotomy was performed using the Argon laser

(HGM Medical Laser Systems, Inc., Salt Lake City, UT, USA) in 27 cases, CO_2 laser (OmniGuide, Inc., Cambridge, MA, USA) in 4 cases, and Revolix Thulium laser (LISA Laser, Inc., Pleasanton, CA, USA) in 1 case, followed by vein graft interposition in all cases. In 27 cases, a 0.4-mm-diameter Teflon piston was used. In 5 cases, a 0.4-mm-diameter Teflon bucket handle-type piston was used because the incus was too short to use a regular Teflon piston (3 cases) or the facial nerve was dehiscent (2 cases). Intraoperative findings were prospectively reported by the surgeon (R. V.) and stored using the Otology–Neurotology Database[14].

Audiometric Assessment

Audiometric evaluation included preoperative and postoperative air-conduction (AC) thresholds, bone-conduction (BC) thresholds, and air-bone gap. Four-frequency (0.5, 1, 2, and 4 kHz) pure-tone audiograms were used for the calculation of AC and BC thresholds. Furthermore, closure of the air-bone gap to within 10 dB or less was evaluated, which is generally considered a successful outcome of stapes surgery in literature[14-17]. The AC and BC thresholds obtained at the same time before and after surgery were used for the calculation of air-bone gap closure. The overall results as well as the results of pure-tone audiometry obtained at 3 months and at least 1 year after surgery were presented. Audiometry was reported according to the American Academy of Otolaryngology – Head and Neck Surgery guidelines[18]. However, thresholds at 3 kHz were substituted in all cases with those at 4 kHz. Audiometric data were stored using the Otology–Neurotology Database[14].

Statistical Analyses

Means, standard deviations (SDs), and percentages were calculated. Preoperative and postoperative BC thresholds, AC thresholds, and air-bone gap were compared using a paired-samples t test. A predictive model for success was created[17,19]. Prognostic factors for success, defined as air-bone gap closure within 10 dB, were identified using univariate analyses. Predictors that were associated with success ($p < 0.10$) were included in multivariate logistic regression analyses. Predictors with $p > 0.10$ were included in the multivariate model when they were clinically relevant. The multivariate model was reduced through exclusion of predictors with $p > 0.10$, using backward elimination. The predictive accuracy of the multivariate model was estimated based on their goodness-of-fit using the Hosmer-Lemeshow test[20]. The model's ability to discriminate between patients was estimated as the area under the receiver operating characteristic (ROC) curve of the model[21]. The area under the ROC curve suitably summarizes the discriminative or predictive value of a test or predictive factor. It ranges between 0.5 (no discrimination, like flipping a coin) and 1.0 (perfect discrimination). All analyses were performed using SPSS Version 21.0 (SPSS, Inc., Chicago, IL, USA).

RESULTS

Patients

Mean age was 36 years (range, 18–59 yr). Of the included 25 patients, 14 (56%) were female. In 20 patients, both ears were affected (80%). All patients suffered from Osteogenesis Imperfecta type 1.

Surgery

Of the included 32 procedures, 17 (53%) were performed on the right ear and 15 (47%) were performed on the left ear. The stapes footplate was found to be fixed in all cases.

Fixation of the anterior one third of the footplate was identified in 8 cases (25.0%), diffuse involvement of the footplate was identified in 11 cases (34.4%), and obliteration of the oval window niche was identified in 13 cases (40.6%). Malleus, incus, and stapes crura were not abnormally slender, brittle, or fractured in any of the cases.

Audiometric Assessment

Preoperative and postoperative audiometric data are summarized in Tables 1 and 2. At the first follow-up, the air-bone gap was closed to 10 dB or less in 87.5% of all cases. Both AC thresholds and air-bone gap improved significantly, with mean (SD) gains of 21.5 (11.7) and 22.3 (9.0) dB, respectively. Sensorineural hearing loss, defined as changes in postoperative BC thresholds exceeding 15 dB, occurred in 1 case (3.2%). In another case, temporary sensorineural hearing loss of 16 dB was witnessed at first follow-up. At 9 months of follow-up, the difference in preoperative and postoperative BC thresholds

5

Table 1. Overall audiometric results

Outcome	Overall results (n=32)
Follow-up duration, mean (range), mo	7 (2Y41)
Mean AC, mean (SD), dB	30.7 (15.2)[a]
Mean BC, mean (SD), dB	26.5 (12.9)
Mean ABG, mean (SD), dB	4.3 (6.2)[a]
Gain in AC, mean (SD), dB	21.5 (11.7)
Gain in BC, mean (SD), dB	j0.8 (8.2)
Gain in ABG, mean (SD), dB	22.3 (9.0)
ABG closure e10 dB,	% 87.5
Sensorineural hearing loss (BC deterioration 915 dB), %	3.2

[a]p value G 0.001 using paired-samples t test.
ABG indicates air-bone gap; AC, air-conduction; BC, bone-conduction; N, number of procedures; SD, standard deviation.

Table 2. Short-term and long-term audiometric results

Outcome	Short-term follow-up (n = 18)	Follow-up at least 1 yr (n = 18)
Follow-up duration, mean (range), mo	4 (3Y6)	26 (12Y47)
Mean AC, mean (SD), dB	30.8 (11.9)[a]	32.4 (16.4)[a]
Mean BC, mean (SD), dB	26.1 (11.0)	26.5 (13.1)
Mean ABG, mean (SD), dB	4.7 (6.2)[a]	5.9 (9.5)[a]
Gain in AC, mean (SD), dB	19.7 (11.8)	17.9 (15.5)
Gain in BC, mean (SD), dB	-1.9 (8.1)	-2.1 (10.5)
Gain in ABG, mean (SD), dB	21.7 (9.4)	19.9 (10.5)
ABG closure e10 dB, %	94.4	72.2

[a]*p* value <0.001 using paired-samples *t* test.
ABG indicates air-bone gap; AC, air-conduction; BC, bone-conduction; N, number of procedures; SD, standard deviation.

was only 11 dB. The mean (SD) loss in BC threshold was 0.8 (8.2) dB for the entire series. Eighteen patients underwent pure-tone audiometry at 3 to 6 months. The air-bone gap was closed to 10 dB or less in 94.4% of these cases. Both mean (SD) gains in AC and air-bone gap (19.7 [11.8] and 21.7 [9.4] dB, respectively) were comparable to the mean values for the entire series of cases. The success rate declined to 72.2% in 18 patients who were followed up for at least 1 year. Moreover, the mean (SD) gains in AC and air-bone gap were slightly less as well (17.9 [15.5] and 19.9 [10.5] dB, respectively). Figure 1 shows the audiometric results over time for the entire series of cases. Mean postoperative air-bone gap, mean improvement in AC, and success percentages slightly deteriorated over the years. The audiometric results of all individual cases are visualized using the (modified) Amsterdam Hearing Evaluation Plots in Figures 2 and 3.[15]

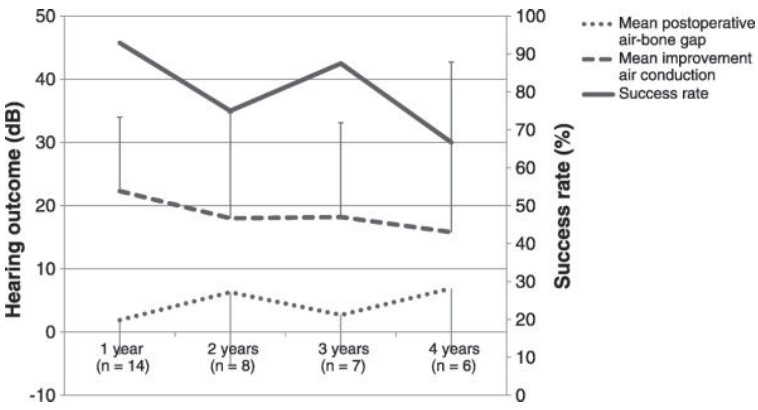

Figure 1. Audiometric results over time for the entire series of cases. N = number of procedures. The error bars indicate standard deviations.

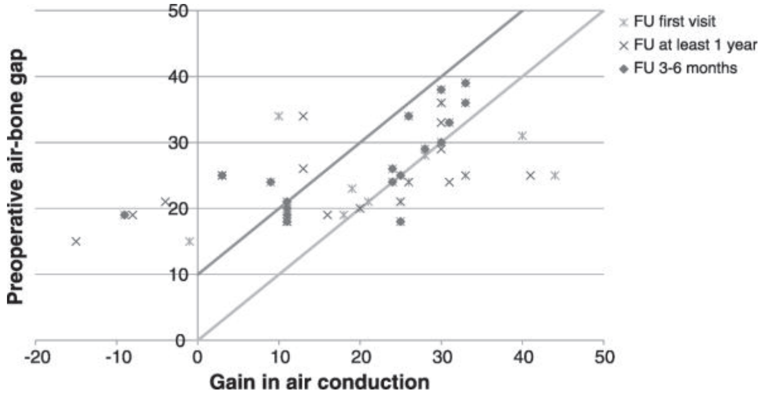

Figure 2. Modified Amsterdam Hearing Evaluation Plot. The solid diagonal line indicates total air-bone gap closure. Any point below this line is defined as overclosure. The dotted diagonal line indicates a gap between the preoperative bone-conduction and postoperative air-conduction of 10 dB. Any point above this line is representative of a patient in whom air-bone gap closure to within 10 dB was not achieved.

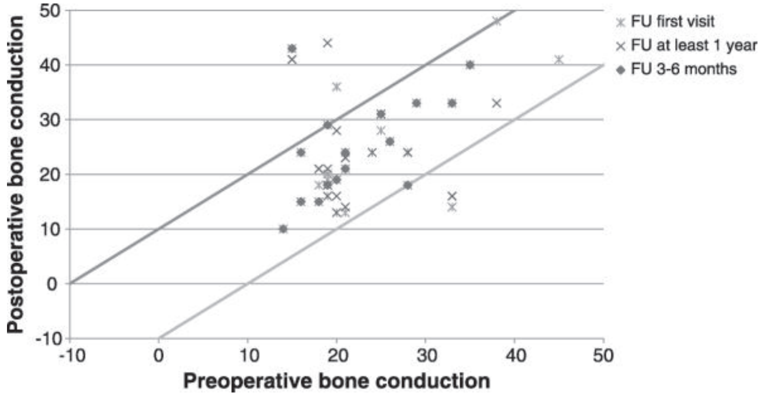

Figure 3. Amsterdam Hearing Evaluation Plot. The 2 diagonal lines enclose the cases in which bone-conduction did not change more than 10 dB.

5

Failures

Adequate air-bone gap closure to within 10 dB was not achieved in 4 cases (12.5%) at the first follow-up and in an additional 2 cases (6.3%) at last the follow-up. In 4 of these 6 cases (12.5% of all 32 procedures), revision surgery was carried out. Re-exploration revealed 1 eroded incus, 1 dislocated incus, 1 case of oval window obliteration, and 1 piston that was too short. No audiological follow-up was available after revision surgery for one of these patients. In 1 patient, a second revision was necessary because the placed prosthesis was too long. The second revision surgery in this patient was successful. In the other 2 cases, the first and only revision surgery was successful with air-bone gap closure to within 10 dB.

Risk Model

We identified 1 predictor that was associated with success ($p < 0.10$) using univariate analysis, namely, the presence of a normal incus (see Table 3 for odds ratios and their 95% confidence intervals for all variables after univariate analysis). Preoperative air-bone gap was not a statistically significant predictor but was included in the multivariate model because it is a clinically relevant predictor. Multivariate logistic regression analysis showed that both incus length and preoperative air-bone gap were associated with success, $p < 0.10$ (see Table 3 for odds ratios and their 95% confidence intervals for the variables in the final multivariate risk model).

Table 3. Predictive model for success (postoperative air-bone gap closure 910 dB) for the entire study population

	First postoperative pure-tone audiometry (n=32)	
Variable	**OR (95% CI) univariate**	**OR (95% CI) multivariate**
Age	1.066 (0.961-1.182)	-
Sex	1.545 (0.189-12.640)	-
Mean preoperative BC[a]	1.006 (0.912-1.110)	-
Mean preoperative AC[a]	0.977 (0.914-1.045	-
Mean preoperative ABG[a]	0.926 (0.822-1.042)	**0.820 (0.668-1.007)**
Type of laser	0.315 (0.055-1.791)	-
Narrow oval window	>2 (0-0)	-
Dislocated stapes	0.231 (0.016-3.371)	-
Short incus	**0.120 (0.012-1.193)**	**0.011 (0.000-0.596)**
Facial nerve dehiscence	0.231 (0.016-3.371)	-
Obliteration	0.647 (0.079-5.2920	-
Hemorrhagic mucosa	>2.0 (0-0)	-
Footplate involvement	0.499 (0.109-2.277)	-

Predictors with p G 0.10 are printed in bold.
[a]Odds ratios and their corresponding 95% CI for pure-tone audiometric outcomes per decibel.
95% CI indicates 95% confidence interval; ABG, air-bone gap; AC, air-conduction; BC, bone-conduction; N, number of procedures; OR, odds ratio.

Figure 4 shows the distribution of the calculated probabilities in the 32 included cases. The mean probability is 0.88, which corresponds with previously reported success rates. The probabilities ranged from 0.202 to 0.996. The model showed a good fit, using the goodness-of-fit Hosmer-Lemeshow test ($p = 0.82$). This means that there is a high level of agreement between the calculated risk using our multivariate risk model and the observed outcomes. The model distinguishes well between success and failure, with an area under the ROC curve of 0.90.

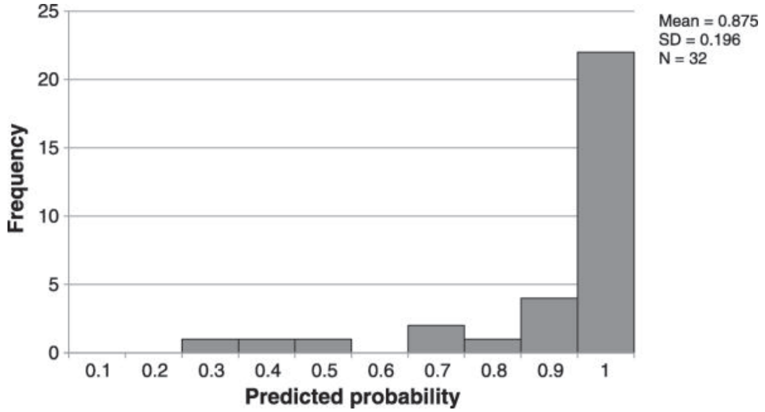

Figure 4. Distribution of calculated probability.

DISCUSSION

In this study, we evaluated the hearing outcomes after primary stapes surgery in 32 consecutive Osteogenesis Imperfecta cases. Furthermore, we developed a risk model that calculated the probability of success for patients undergoing primary stapes surgery for Osteogenesis Imperfecta. Success was defined as air-bone gap closure to within 10 dB. Our study shows a success rate of 88%. Backward multivariate logistic regression showed that a model with preoperative air-bone gap closure and intraoperatively established incus length accurately predicts success after primary stapes surgery. Other authors have reported success rates of 75% to 85% and sensorineural hearing loss in 0% to 8% of patients with osteogenesis imperfecta treated with primary stapes surgery[9-13]. Postoperative sensorineural hearing loss occurred in 3.2% of the cases included in this study. In one additional case, temporary sensorineural hearing loss of 16 dB was witnessed at first follow-up. Various intraoperative findings have been reported in Osteogenesis Imperfecta. Fixed footplates and thickening of the footplate were found in all cases in this study and have been reported by several others[10]. Obliteration of the footplate was found in 41% of cases in this study. Incidences between 27% and 80% have been previously reported. Brittleness, atrophy, and fractures of the stapes crura have been reported previously[10,22,23] but were not observed in this study. Although footplate obliteration, hemorrhagic mucosa, excessive bleeding, and brittle, atrophic, or fractured stapes crura have been thought to be associated with a higher incidence of failure, no prognostic studies have been published up to now. All of these predictors were not associated with success or failure in our case series, using univariate analyses. In interpreting our findings, the following considerations need to be taken into account. First, in accordance with the Committee of Hearing and Equilibrium Guideline[18], a follow-up duration of at least 1 year is to be preferred when reporting AC threshold (improvement) and air-bone gap closure. Many patients treated in this tertiary referral center have to travel long distances to reach the clinic, and therefore, long-term follow-up is not achieved in a large number of cases. For 12 cases, no long-term follow-up, exceeding 1 year, was available, and therefore, we decided to include the results of the first postoperative pure-tone audiometry in our analyses. Second, the sample size of 32 cases is rather small. Given that sensorineural hearing loss occurred in 0% up to 8% of stapes surgeries for Osteogenesis Imperfecta, this study and most of the case series that have been reported are not large enough to make reliable statements about this outcome measure. Small sample sizes can lead to overestimation or underestimation of intervention effects.

CONCLUSION

Although stapes surgery in Osteogenesis Imperfecta is less successful than in otosclerosis, the results of this prospective study show that stapes surgery is a feasible and safe treatment option in patients with Osteogenesis Imperfecta. Hearing thresholds improve significantly. Success, defined by air-bone gap closure to within 10 dB, seems to be associated with preoperative air-bone gap and intraoperatively established incus length. The case series presented here and the case series described by other authors are not large enough to make reliable statements about the risk of sensorineural hearing loss.

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Part III

Stapedotomy in childhood otosclerosis

Chapter 6

Primary stapedotomy in children with otosclerosis: A prospective study of 41 consecutive cases

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ABSTRACT

Objectives/Hypothesis: To prospectively evaluate hearing outcomes in children with otosclerosis undergoing primary stapes surgery.

Study Design: A nonrandomized, nonblinded, prospective case series.

Methods: Thirty-four consecutive pediatric patients who underwent 41 primary stapedotomies for otosclerosis in a tertiary referral center were included. Patients were included when there was evidence of otosclerotic stapes fixation and they had available postoperative pure-tone audiometry. Patients underwent primary stapedotomy with vein graft interposition and reconstruction with a regular piston, bucket handle prosthesis, or total ossicular replacement prosthesis. Hearing results were evaluated using pre- and postoperative four-frequency (0.5, 1, 2, and 4 kHz) audiometry. Air-conduction thresholds, bone-conduction thresholds, and air-bone gaps were measured. Postoperative audiometry was performed at 3, 6, 9, 12, 18, and 24 months after surgery and at a yearly interval thereafter.

Results: Overall, a postoperative air-bone gap closure of 10 dB or less was achieved in 93% of cases and to within 20 dB in 98% of cases. Mean gain in air-conduction threshold was 23 dB for the entire case series, and mean air-bone gap closure was 23 dB. Postoperative sensorineural hearing loss, defined as changes in bone-conduction thresholds exceeding 10 dB, occurred in one case at last follow-up. The bone-conduction threshold deteriorated 13 dB in this case.

Conclusion: Primary stapedotomy is a safe and feasible treatment option in children with juvenile otosclerosis.

INTRODUCTION

Otosclerosis is characterized by an abnormal growth and remodeling of bone predominantly in the middle ear around the otic capsule, which can result in progressive conductive hearing loss. Stapes surgery has proven to be a safe and effective treatment option in adult otosclerosis patients, with success percentages ranging between 75% and 95%.[1,2]

Although highly prevalent in adults with conductive hearing loss, the incidence of otosclerosis in children is low. One histologic study shows an incidence of less than 1% in children aged under 5 years (one case out of 161 temporal bones) and 4% in children aged between 5 and 18 years.[3] Treatment options include hearing aids or stapes surgery. In the past, stapes surgery has been thought to be more risky in children. Meanwhile, numerous authors have published the results of case series of children undergoing stapes surgery, all of which are characterized by small sample sizes.[4-18] Nonetheless, caution is advocated when considering surgery in children for otosclerosis.[19]

The objective of this study was to prospectively evaluate hearing outcomes in children with otosclerosis undergoing primary stapes surgery.

MATERIALS AND METHODS

Patients

This is a nonrandomized, nonblinded, prospective case series of 34 consecutive children under 18 years of age who underwent 41 primary stapedotomies from 1991 to 2014 for otosclerosis with evidence of stapes fixation. A total of 49 procedures for stapes fixation in pediatric otosclerosis patients have been carried out in a tertiary referral center, five of which (10%) were revision stapedotomies. No postoperative pure-tone audiometry was available for another three (6%) of these 49 cases. Revision procedures and patients lacking postoperative pure-tone audiometry were excluded.

Surgery

All procedures were performed by the same senior surgeon (r.v.) in a tertiary referral center. A transcanal procedure was undertaken in all cases. Stapes mobility was assessed by gentle palpation of the stapes head with a curved needle after incudostapedial joint separation. Juvenile otosclerosis was diagnosed intraoperatively by establishing the presence of white otosclerotic foci. Stapedotomy was performed using the KTP laser (Lumenis, Inc., Salt Lake City, UT) in 39 cases (95%) and the CO_2 laser (OmniGuide, Inc., Cambridge, MA) in two cases (5%). A vein graft interposition followed in all cases. In 37 cases (90%), a 0.4-mm-diameter Teflon piston was used. In two cases (5%), a 0.4-mm-diameter Teflon bucket-handle type piston was used because the incus was too short to allow the use of a regular Teflon piston given that the facial nerve was not bridged. In two cases (5%), a total ossicular replacement prosthesis (TORP), composed of a hydroxyapatite head and a 0.4-mm-diameter Teflon shaft, was used because the stapes fixation was associated with a malleus-incus ankylosis. Intraoperative findings were prospectively reported by the surgeon and stored using the Otology-Neurotology Database (ONDB).[2]

Audiometric Assessment

Audiometric evaluation included pre- and postoperative air-conduction (AC) thresholds, bone-conduction (BC) thresholds, and air-bone gaps. Four-frequency (0.5, 1, 2, and 4 kHz) pure-tone audiograms were used to evaluate the hearing results using AC and BC thresholds. Furthermore, closure of the air-bone gap to 10 dB or less was calculated, as well as air-bone gap closure to 20 dB or less, which are both generally considered successful outcomes of stapes surgery in literature.[2,20,21] We believe that closure of the air-bone gap to 10 dB or less should be considered a successful outcome of otosclerosis surgery in both adults and children from the surgeon's perspective. AC and BC thresholds obtained at the same time postoperatively were used for calculation of the air-bone gap closure. Audiometry was reported according to the American Academy of Otolaryngology–Head and Neck Surgery guidelines.[22] However, thresholds at 3 kHz were substituted in all cases with those at 4 kHz. Audiometric data were stored using the ONDB.[2] Audiometric results at last follow-up and at least 1-year follow-up were evaluated.

Statistical Analyses

Means, standard deviations (SDs), and percentages were calculated. Pre- and postoperative BC thresholds, AC thresholds, and air-bone gaps were compared using a paired-samples *t* test.

All analyses were performed using SPSS version 20.0 (IBM Corp., Armonk, NY).

RESULTS

Patients

Mean age at the time of surgery was 14.4 years (range 8 to 18 years). Of the included 34 patients, 27 were female (79%). In 14 patients, both ears were affected (41%).

Surgery

Of the included 41 procedures, 22 were performed on the right ear (54%) and 19 on the left ear (46%). The stapes footplate was found to be fixed in all cases. Fixation of the anterior one-third of the footplate was identified in 29 cases (71%) and involvement of the entire footplate in three cases (7%). Obliterative otosclerosis, defined as hard new bone filling the fossa that requires an oval window drill-out for an excessively thick footplate, was seen in nine cases (22%). A simultaneous malleus ankylosis was found in two cases (5%). No other ossicular chain abnormalities were identified; children with congenital malformations of the ossicular chain were not included.

Audiometric Assessment

Pre- and postoperative audiometric data were summarized in Tables 1 and 2. The average follow-up duration was 81 months (range 3 to 196 months) for the entire case series of 41 cases. The average follow-up duration for 38 cases, with at least 1-year follow-up, was 90 months (range 12 to 196 months).

Table 1. Overall Pure-Tone Audiometric Results (n = 41).

Outcome	Preoperative	Postoperative	Difference (95% CI)
AC (mean [SD]) dB	42.7 (8.7)	19.6 (9.0)	23.0 (20.3; 25.8)
BC (mean [SD]) dB	17.0 (6.6)	16.7 (7.0)	0.3 (–1.4; 2.0)
ABG (mean [SD]) dB	25.7 (5.7)	3.0 (5.3)	22.7 (20.6; 24.9)
ABG closure ≤ 10 dB (%)	–	92.7	–
ABG closure ≤ 20 dB (%)	–	97.6	–
SNHL > 10 dB (%)	–	2.4	–

Means were calculated using AC and BC thresholds at 0.5, 1, 2 and 4 kHz.
ABG = air-bone gap; AC = air conduction; BC = bone conduction; CI = confidence interval; n = number of procedures; SD = standard deviation; SNHL = sensorineural hearing loss.

Overall, at last follow-up for the entire case series of 41 cases, the air-bone gap was closed to 10 dB or less in 93% of all cases and to 20 dB or less in 98% of all cases. Both AC thresholds and air-bone gaps improved significantly with mean gains of 23 dB (95% CI, 20; 26) and 23 dB (95% CI, 21; 25) respectively. Sensorineural hearing loss, defined as changes in postoperative BC thresholds exceeding 10 dB, occurred

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Table 2. Pure-Tone Audiometric Results With at Least One-Year Follow-Up (n = 38).

Outcome	Preoperative	Postoperative	Difference (95% CI)
AC (mean [SD]) dB	42.9 (9.0)	19.7 (9.2)	23.2 (20.3; 26.1)
BC (mean [SD]) dB	17.0 (6.7)	16.8 (7.0)	0.2 (–1.6; 2.0)
ABG (mean [SD]) dB	25.8 (5.8)	2.8 (5.5)	23.0 (20.7; 25.3)
ABG closure ≤ 10 dB (%)	–	92.1	–
ABG closure ≤ 20 dB (%)	–	97.4	–
SNHL > 10 dB (%)	–	2.6	–

Means were calculated using AC and BC thresholds at 0.5, 1, 2 and 4 kHz.
ABG = air-bone gap; AC = air conduction; BC = bone conduction; CI = confidence interval; n = number of procedures; SD = standard deviation; SNHL = sensorineural hearing loss.

in one case at last follow-up. The BC threshold deteriorated 13 dB in this patient. The average gain in BC threshold was 0.3 dB (95% CI, –1; 2) for the entire series.

The success rate was 92% in 38 cases that were followed for at least 1 year. The mean gain in AC and improvement in air-bone gap was comparable to the results at 3-month follow-up (23 dB [95% CI, 20; 26] and 23 dB [95% CI, 21; 25], respectively). The audiometric results of all individual cases are visualized using the Amsterdam Hearing Evaluation Plots in Figures 1 and 2[20] Figure 3 shows the audiometric results over time for the cases with available long-term pure-tone audiometry. Mean postoperative air-bone gap, gain in AC, and the success rate remained stable over the course of 5 years.

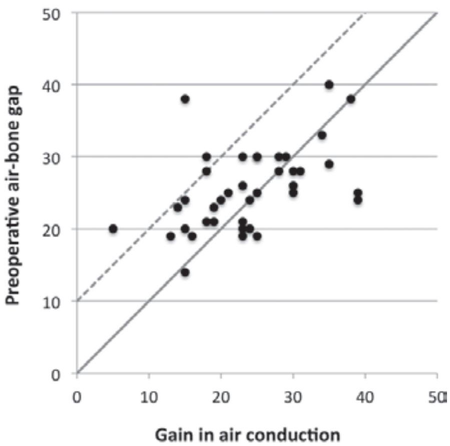

Figure 1. Amsterdam Hearing Evaluation Plot. Each black dot represents one case. The solid diagonal line indicates total air-bone gap closure. The area between the diagonal lines indicates successful surgery with an air-bone gap of 10 dB or less.

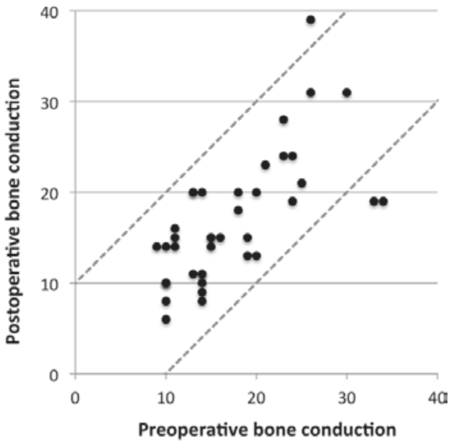

Figure 2. Amsterdam Hearing Evaluation Plot. Each black dot represents one case. The two diagonal lines enclose the cases in which bone conduction did not change more than 10 dB.

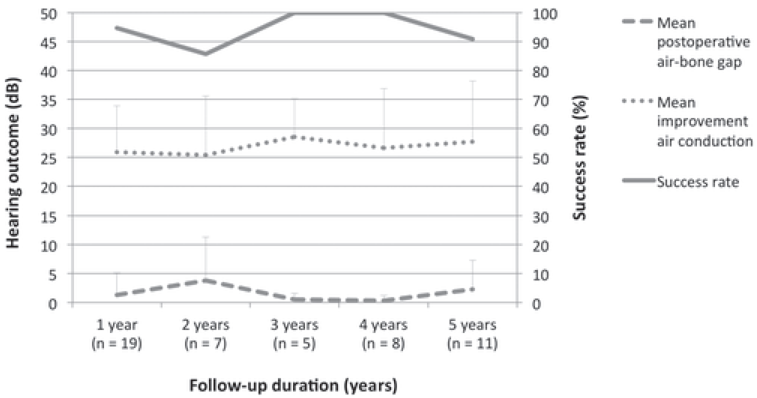

Figure 3. Audiometric results over time. n = number of cases. Error bars indicate standard deviations.

Failures

Adequate air-bone gap closure to within 10 dB was not achieved in four cases (19%) at 3-month follow-up. None of the cases were characterized by a simultaneous malleus ankylosis, nor were they all treated early in the case series. These four children were treated between 1999 and 2010.

All of these cases were initially treated using a 0.4-mm Teflon piston. Their preoperative AC thresholds and mean air-bone gaps, however, were above average. Their preoperative AC thresholds ranged between 43 and 54 dB compared to a mean of 43 dB for the entire population, and their preoperative mean air-bone gap ranged between 24 and 38 dB compared to a mean of 26 dB for the entire population. In one of these four cases, revision surgery was carried out 2.5 years later. Re-exploration revealed oval window reobliteration. During revision surgery, a stapedotomy with vein graft interposition was performed with placement of a 0.4-mm Teflon piston. The air-bone gap was closed to 11 dB at 3 months postoperatively. At 2 years postoperatively, the air-bone gap was closed to 6 dB in this case.

DISCUSSION

In this study, we evaluated the hearing outcomes after primary stapes surgery in 41 pediatric cases. Postoperative air-bone gap closure to within 10 dB was achieved in 93% of cases and to within 20 dB in 98% of cases at last follow-up. Mean gain in AC threshold was 23 dB (95% CI, 20; 26) for the entire case series, and mean air-bone gap closure was 23 dB (95% CI, 21; 25). Furthermore, in cases where long-term follow-up was available, the mean postoperative air-bone gap, gain in AC, and success rate remained stable over the course of 5 years. Postoperative sensorineural hearing loss occurred in one case at last follow-up. The bone-conduction threshold deteriorated 13 dB in this case.

Surgeons may choose to postpone surgical treatment of juvenile otosclerosis until children are old enough to participate in the decision-making process. In the past, it has been suggested that stapes surgery in children is associated with an increased risk of postoperative sensorineural hearing loss.[4,6,10] On the other hand, inadequate treatment of both unilateral and bilateral (conductive) hearing loss in children may lead to impaired speech and language development.[23,24] This current series of juvenile otosclerosis is associated with a higher incidence of simultaneous malleus fixation. In our previously published series,[2] simultaneous malleus fixation was found in 1% of adults compared to 6% in children. In this series, we also report a high incidence of obliterative otosclerosis (22%). In our previously published series,[2] 15% of the children had obliterative otosclerosis compared to 3% in adults. Several authors have also reported higher rates of obliterative otosclerosis in children compared to adults, and as a result, a greater need for drilling the stapes footplate: 42% in Cole,[7] 27% in Lippy et al.,[12] and 28% in Robinson.[16] A worsening of footplate pathology later on in life was described by Robinson[16]; therefore, the higher incidence of obliterative otosclerosis should not be a contraindication for stapes surgery in childhood. Furthermore, the majority of the studies on pediatric stapes surgery reported sensorineural hearing loss in less than 10% of surgically treated children (see Table 3)[4,6,9,10,13,15,18] Sensorineural hearing loss was defined as changes in BC thresholds exceeding 10 or 15 dB in these studies.

Table 3. Reported Pure-Tone Audiometric Results for Stapedotomy in Juvenile Otosclerosis in Literature.

Author/ year	Mean age in years (range)	No.	Mean follow-up duration in months (range)	% ABG closure < 10 dB^	% ABG closure < 20 dB^	% SNH > 10 dB
An (2014)[4]	NR	6	2 (SD 1)	67	100	NR
Bachor (2005)[5]	11 (?)	5	NR (3-12)	40	80	20
Carlson (2013)[6]	16 (5-18)	NR	NR	71	94	NR
Cole (1982)[7]	17 (6-20)	62	82 (2-264)	77	NR	13
de la Cruz (1999)[8]	15 (SD 3)	NR	NR	82	NR	NR
Denoyelle (2010)[9]	NR	5	2	80	100	NR
House (1980)[10]	14 (10-18)	24	NR	92	NR	4[b]
Lescanne (2008)[11]	14 (0-18)	10	80 (24-180)	90	100	NR
Lippy (1998)[12]	13 (7-17)	60	6	92[a]	95[a]	NR
Millman (1996)[13]	17 (7–21)	38	NR (2–3)	58	89	3
		40	305 (204–408)	50	90	NR
Murphy (1996)[14]	? (5–19)	9	NR	56	100	NR
Neilan (2013)[15]	11 (6–18)	7	NR (1–2)	57	86	14[c]
Robinson (1983)[16]	14 (5–18)	35	5	100	100	0[b]
Vick (2004)[17]	12 (10–13)	5	NR (1–8)	60	80	20
Vincent (2006)[2]	15 (8–18)	31	80 (4–160)	94	100	0[b]
von Haacke (1985)[18]	16 (6–21)	19	27 (3–180)	74	89	0[b]

In all studies, pure-tone averages were calculated using thresholds at 0.5, 1, 2, and 3 or 4 kHz, except for the studies by Cole[7] (0.5, 1, and 2 kHz), House et al.[10] (NR), Millman et al.[13] (0.5, 1, and 2 kHz), Robinson[16] (unclear), and von Haacke[18] (unclear).

[a] Preoperative bone conduction used for calculation of the postoperative ABG.

[b] SNHL defined as > 15 dB.

[c] Unclear what definition of SNHL was; cutoff value not mentioned.

ABG = air-bone gap; n = number of procedures; NR = not reported; SD = standard deviation; SNHL = sensorineural hearing loss.

A rapid systematic literature search identified 16 articles[2,4-18] reporting audiometric results following stapes surgery in children with otosclerosis. Overall, air-bone gap closure to within 10 dB was reached in 40% to 100% in these studies (Table 3). Air-bone gap closure to within 20 dB was reached in 80% to 100% of children. The studies that only included patients with juvenile otosclerosis seemed to report better results than the studies including a combined population of children with juvenile otosclerosis and children with congenital stapes fixation. Both juvenile otosclerosis and congenital stapes fixation are indications for stapes surgery in children. Juvenile otosclerosis is more likely in case of progressive conductive hearing loss and a positive family history. Both are characterized by fixation of the stapes and can be successfully treated with stapes surgery. Children with juvenile otosclerosis generally show better hearing results following stapes surgery than children with congenital stapes fixation. Risk differences for air-bone gap closure within 10 dB differ between 4% and 38% in favor of juvenile otosclerosis in eight studies.[4-6,8-10,15,17] Risk differences for air-bone gap closure within 20 dB differ between 0% and 30% in favor of juvenile otosclerosis in six studies.[4-6,9,15,17] It needs to be noted that all of the sample sizes are small; as a result, none of these risk differences is statistically significant.

Vincent et al.[2] previously reported a success percentage, defined as air-bone gap closure within 10 dB of 94% out of 3,050 stapedotomies, in a mixed population of children, adults, and elderly. The success percentage for children alone was 94% as well. Air-bone gap closure within 20 dB was achieved in 98% in the entire population and in 100% of children. The children described in this article were included in this overview as well. Although the results and conclusions were similar, this additional report is valuable for several reasons. We were able to report the audiometric details of these children in more detail in this article and included longitudinal data showing stability of the audiometric results over time. Furthermore, we were able to add more cases, which adds to the statistical power of the results described here. Lastly, we offer a valuable and detailed comparison to other published results of pediatric stapes surgery for otosclerosis.

CONCLUSION

The results of primary stapedotomy in children with otosclerosis are comparable to the results of primary stapedotomy in adult otosclerosis patients. Success percentages are high, with an air-bone gap closure to within 10 dB in 93% of stapedotomies and air-bone gap closure within 20 dB in 98% of stapedotomies. Sensorineural hearing loss occurred in one case.

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Part IV

Revision stapedotomy in otosclerosis

Chapter 7

Revision stapedotomy: operative findings and hearing results. A prospective study of 652 cases from the Otology-Neurotology Database

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ABSTRACT

Objective: To identify the causes of failure of primary stapes surgery and to evaluate the hearing results of revision stapes surgery in a consecutive series of 652 cases.

Study Design: Prospective nonrandomized clinical study.

Setting: Tertiary referral center. Patients: Six hundred thirty-four patients who underwent 652 consecutive revision stapes operations from April 1992 to December 2007 were enrolled in this study.

Main Outcome Measures: Preoperative and postoperative audiometric evaluation using conventional audiometry, namely, air-bone gap (ABG), bone-conduction thresholds, and air-conduction thresholds, were assessed. Postoperative audiometry was performed at 3, 6, 9, 12, 18, and 24 months and then annually for 15 years.

Results: The most frequently identified reason for primary surgery failure was incus erosion (27.6%) and prosthesis displacement (18.2%). The postoperative ABG was closed to 10 dB or less and 20 dB or less in 63.4 and 74.6% of cases, respectively. The mean 4-frequency postoperative ABG was 11.5 dB as compared with 28 dB preoperatively (mean difference, 16.5 dB; 95% confidence interval [CI], 15.1-17.9 dB, $p < 0.0001$). The mean 4-frequency postoperative air-conduction thresholds were 45.7 dB compared with 58.7 dB preoperatively (mean difference, 13 dB; 95% CI, 11.4-14.6 dB, $p < 0.0001$). The mean 4-frequency postoperative bone-conduction thresholds were 34 dB compared with 30.6 dB preoperatively (mean difference, -3.5 dB; 95% CI, -4.4 to -2.5 dB, $p < 0.0001$). A significant postoperative sensorineural hearing loss (>15 dB) was observed in 2.9% of cases in this series.

Conclusion: Improvement of a conductive hearing loss after initial unsuccessful primary or revision stapes surgery can be accomplished with further revision but is occasionally modest.

Surgical outcome after revision stapes surgery has consistently been reported as less favorable when compared with that after primary surgery. Revision surgery is also associated with a higher risk of postoperative sensorineural hearing loss (SNHL).[1-3] Hearing results seem to be determined by the primary surgical techniques used, the cause of failure identified at revision surgery, and the revision technique.[1] During the period of April 1992 to December 2007, 4,508 operations for otosclerosis were performed by the same surgeon (first author) in the same tertiary referral center. Of these 4,508 operations, 652 were revisions. The first aim of this prospective study was to evaluate the causes that led to the failure of the previous surgery. The second aim was to report hearing results of the revision surgery in a series of 652 consecutive surgical revisions performed by the same surgeon.

MATERIALS AND METHODS

Patients

This is a prospective study of 634 patients who underwent 652 revision stapes surgeries. All patients treated between April 1992 and December 2007 were included in this study and were operated on by the same surgeon. A serial assessment of hearing status was conducted before and at a regular interval after surgery. All patients had at least a 3-month audiology follow-up after the surgery. All data were tabulated prospectively using the Otology-Neurotology Database (ONDB) (AS Multimedia, Inc., Cassagne, France).[4]

Surgery

The technique of revision was similar to our approach in primary stapedotomy.[4] A transcanal procedure was used in all cases in this series. The malleus and incus were visually inspected, and then mobility was assessed by gentle palpation. The connection of the prosthesis to the incus or malleus and its position in the oval window niche were checked. In all cases, an argon laser was used to lyse adhesions in the middle ear and/or to dissect in the area of the oval window. The laser was used to progressively vaporize the fibrous and connective tissues surrounding the prosthesis's shaft until the position of the distal tip and penetration or not into the labyrinth can be ascertained. The original stapedectomy (partial or total) or stapedotomy fenestra was inspected. The original stapedectomy (partial or total) or stapedotomy fenestrum was inspected. In all cases a regular laser stapedotomy was created through the remnant of the stapes footplate if at the previous surgery the surgeon had not fenestrated the footplate. If a previous stapedotomy had been created the perilymph membrane was re-opened. This was followed by vein graft interposition.

In case of reobliteration with bony closure of the oval window, a regular 0.8-mm stapedotomy was performed using a combination of laser and Skeeter Microdrill (Medtronic Xomed Inc, Jacksonville, FL, USA) and then vein graft interposition.[4] The surgical technique used for the repair of the ossicular chain was dictated by the surgical findings at the time of revision. When more than one possible cause of failure was identified, all causes were recorded, but the one that was suspected to be the major cause was selected as the main cause of failure.

Prostheses that were fixed or displaced from the oval window or from the incus and prostheses that were too long or too short were removed, and a new prosthesis was introduced. Prosthesis fixation was defined as fixing the prosthesis to the promontory or fallopian canal.

A long prosthesis was defined by the prosthesis' length, which was greater than the distance measured intraoperatively between the incus and the stapes footplate.

When the incus and malleus had no abnormalities, a 0.4-mm-diameter Teflon piston of appropriate length was used in 234 cases, whereas a bucket-handle (cup-type) Teflon prosthesis was used in 48 cases when the incus was short or when the facial nerve was dehiscent. The choice of the diameter of the piston varied according to each surgeon's preference. We have always used 0.4-mm-diameter piston for our stapedotomy procedures at our center. When the incus was eroded, dislocated, or absent or when a malleus ankylosis was identified, a total ossicular replacement prosthesis (TORP), composed of an hydroxylapatite head and a 0.4-mm-diameter Teflon shaft, was used in 278 cases and a total 0.4-mm-diameter Teflon malleus piston was used in 6 cases. In 212 of the 278 cases where a TORP was positioned, the malleus relocation technique was used to increase the stability of the prosthesis.[4-5] Malleus relocation is a new technique introduced by this institution in 1997. The malleus is dissected free from the tympanic membrane and the tensor tympani sectioned close to its insertion into the malleus. The incus is then removed. The anterior malleal ligament is stretched by placing a hook anterior to the neck of the malleus, which is then pulled posteriorly, repositioning it so that it lies directly over the footplate. The position of the malleus is maintained by its superior ligament, which is then preserved.

The perilymph fistula was identified as the cause of failure in 36 cases. In these cases, fibrous adhesions were removed from the oval window with the argon laser, and the fistula was closed using a large vein graft. Prosthesis selection was again made based on the position and condition of the incus. In 18 cases, a 0.4-mm-diameter Teflon piston was used; in 4 cases, a bucket-handle (cup-type) Teflon prosthesis was used; and in 2 cases, a TORP was used. In case of preoperative dead ear (12 cases), the fistula was repaired, but no prosthesis was positioned.

In 49 cases, it was not possible to clearly identify the cause of failure at the time of revision, and the patients were therefore classified as having "negative findings".[6] (malleus and incus had no abnormalities, and the position of the previous prosthesis was correct with the presence of a clear round window reflex). In 5 of these 49 cases, the previous prosthesis was left in place, and no further procedure was attempted. In 34 of the 49 cases, the previous prosthesis was replaced by a 0.4-mm-diameter Teflon piston. The choice between removing the previous prosthesis or not was arbitrary. In 8 of the 49 cases, a bucket-handle (cup-type) Teflon prosthesis was placed. In 2 of the 49 cases, a TORP was positioned. In 1 case, a round window obliteration was identified as the suspected cause, the previous prosthesis was left in place, and no further procedure was attempted.

Audiometric Assessment

Audiometric evaluation included the preoperative and postoperative air-bone gap (ABG), air-conduction (AC) thresholds, and bone-conduction (BC) thresholds. Only AC and BC results that were obtained at the same time after surgery were used for the calculation of ABG and pure-tone averages (PTAs). We used a 4-frequency PTA for AC and BC thresholds (0.5, 1, 2, and 4 kHz) obtained at the last follow-up visit. The preoperative and postoperative BC and AC levels at 4 kHz were also assessed. All patients had at least a 3-month audiology follow-up. Audiometry was reported according to the American Academy of Otolaryngology-Head and Neck Surgery guidelines.[7] except for thresholds at 3 kHz, which were substituted in all cases with those at 4 kHz. This was necessary because the 3-kHz measurements were not routinely undertaken at the beginning of this study in 1991.

Statistical Analyses

Mean differences and percentage differences and their 95% confidence intervals (CIs) were calculated regarding the differences before and after surgery as well as between different groups, that is, according to type of surgery, follow-up time, and number of previous operations. Paired t tests were used to evaluate differences before and after surgery, whereas Student's t and $[chi]^2$ tests were used to evaluate possible differences in mean values and percentages between the groups. All analyses were performed using SPSS (version 15.0; SPSS, Inc., Chicago, IL, USA).

RESULTS

Patients

Of all patients, 67% were females, and the mean age was 51 years (range, 12-86 yr). There were 3 children (aged <=18 yr) who underwent 3 operations and 83 senior patients (aged >=65 yr) who underwent 85 operations. Of 652 operations, 583 (89.4%) were primary revision operations, 53 (8.2%) were secondary revision operations, 8 (1.2%) were tertiary revision operations, and 8 (1.2%) were quaternary revision operations. Of the 652 cases, 90 cases (13.8%) had previously been operated on by the author ("personal revision") and 561 cases (86.2%) had previously been operated on by another surgeon ("other surgeon revision"). All the 90 personal revision cases (13.8% of 652 cases) were operated for a previous failed primary surgery. Of the 562 other surgeon revision cases, 493 cases (75.6% of 652 cases) were operated on for a previous failed primary surgery and 69 cases (10.6% of 652 cases) were operated on for a previous failed revision surgery. The indications for revision surgery were a recurrent or persistent 4-frequency average ABG greater than 10 dB in 616 cases (94.4% of the 652 cases) or the suspicion of a perilymph fistula with dizziness, disequilibrium, and the presence of a fistula sign in 36 cases (5.6% of the 652 cases). In all cases where the preoperative ABG was the only criterion for revision surgery (616 cases), all preoperative ABGs were more than 20 dB. The residual 36 cases were operated on for the presence of fistula with or without ABG.

Operative Findings

To accurately assess the cause of failure after previous primary surgery, the causes of failure were specifically studied during revision after previous primary surgery (583 cases). Intraoperative findings in 583 primary revision cases were analyzed to define the main cause of failure, and these are listed in Table 1. The most frequently reported reasons for failure were incus erosion (27.6%), prosthesis displacement without incus erosion (18.2%), and short prosthesis (12.7%). In 8.7% of cases, failure was related to a previous operation that had not been entirely completed because the footplate was found intact and fixed in all cases. The patients were classified as having "incomplete previous operation." Less frequent were perilymph fistula (5.6%), long prosthesis (4.6%), and stapes refixation (4%). Stapes refixation was defined by the presence of an intact and refixed stapes in patients who had mobilization as the primary operation as described by the operative note (stapes refixation should not be confused with incomplete previous operation because the ossicular chain was found to be entirely intact in the group with stapes refixation, whereas the stapes superstructure was absent in the group with incomplete previous operation).

Other identified causes included malleus ankylosis (3.4%), incus dislocation (3.3%), and otosclerotic regrowth with obliteration of the oval window (2.7%), prosthesis fixation by adhesions (0.8%), stapes footplate subluxation into the vestibule (0.2%), and ossified round window (0.2%). In 8% of cases, no pathologic findings that could explain the failure were identified, and the patients were classified as having negative findings[6]. Surgical findings were also studied according to the previous surgeon (personal revision and other surgeon revision).

Table 1. Surgical findings in revision surgery after previous primary surgery (583 cases): main cause of failure

Main cause of failure	Entire series (n = 583)	Personal revision previous surgery: primary (n = 90)	Other surgeon revision previous surgery: primary (n = 493)
Eroded incus	161 (27.6%)	28 (31%)	133 (27%)
Dislocated incus	19 (3.3%)	1 (1.2%)	18 (3.7%)
Malleus ankylosis	20 (3.4%)	1 (1.2%)	19 (4%)
Dislocated stapes footplate	1 (0.2%)	-	1 (0.2%)
Dislocated prosthesis	106 (18.2%)	19 (21%)	87 (17.7%)
Short prosthesis	74 (12.7%)	12 (13.4%)	62 (12.6%)
Long prosthesis	27 (4.6%)	2 (2.2%)	25 (5%)
Prosthesis fixation	4 (0.8%)	-	4 (0.8%)
Incomplete previous operation	51 (8.7%)	-	51 (10.3%)
Stapes refixation	23 (4%)	-	23 (4.7%)
Perilymphatic fistula	33 (5.6%)	8 (8.8%)	25 (5%)
Oval window obliteration	16 (2.7%)	1 (1.2%)	15 (3%)
Ossified round window	1 (0.2%)	-	1 (0.2%)
Negative findings	47 (8%)	18 (20%)	29 (5.8%)

Table 2. Time of failure related to the cause of failure in personal revision series (n = 90)

	Time of failure			
Main cause of failure	Immediate (n = 22)	=12 mo (n = 18)	12-60 mo (n = 36)	960 mo (n = 14)
Eroded incus	-	5 (27.8%)	16 (44.4%)	7 (50%)
Dislocated prosthesis	2 (9%)	4 (22.2%)	10 (27.8%)	3 (21.4%)
Short prosthesis	9 (41%)	-	3 (8.3%)	-
Long prosthesis	2 (9%)	-	-	-
Perilymph fistula	3 (13.7%)	3 (16.6%)	2 (5.7%)	-
Oval window reobliteration	-	1 (5.6%)	-	-
Malleus ankylosis	-	-	-	1 (7.2%)
Dislocated incus	-	-	-	1 (7.2%)
Negative findings	6 (27.3%)	5 (27.8%)	5 (13.8%)	2 (14.2%)

Table 2 shows the main cause of failure correlated to the time of failure after the previous operation in the 90 cases of the personal revision series. Immediate failure and failures that occurred within the first year after the primary operation were mainly related to an inappropriate prosthesis' length (11 cases, 27.5%), whereas failures that occurred after one or several years were mainly related to incus erosion (23 cases, 46%) and prosthesis dislocation (13 cases, 26%). Perilymph fistula was mainly observed during the first year after the primary operation (6 of the 8 cases occurred within the first 12 mo).

Audiometric assessment

Hearing Results in 538 Cases

Of the 652 cases in which surgical data were compiled, 549 had audiologic data available (84%). Of these 549 cases, 11 ears that had total SNHL (dead ear) before the revision operations. These 11 ears were operated on for severe vertigo with the presence of a positive fistula sign. These 11 cases were excluded from the analysis of the hearing results (n = 538). Mean follow-up was 25.2 months (range, 3-164.9 mo; Table 3)

Table 3. Hearing results in 538 cases.

Variables	Preoperatively	Postoperatively	Mean difference (95% CI)	P
SNHL, %		2.9		
ABG ≤ 10 dB, %		63.4		
ABG 11-20 dB, %		11.2		
ABG ≤20 dB, %		25.4		
Mean ABG, dB	28	11.5	-16.5 (-17.9 to -15.5)	<0.0001
Mean BC, dB	30.6	34	3.5 (2.5 to 4.4)	<0.0001
Mean AC, dB	58.7	45.7	-13.0 (-14.6 to -11.4)	<0.0001
BC at 4 kHz, dB	40.8	44.9	4.1 (2.9 to 5.3)	<0.0001
AC at 4 kHz, dB	66.3	57.9	-8.5 (-10.1 to -6.8)	<0.0001

Of the 538 evaluated cases, a residual ABG of 10 dB or less was achieved in 63.4% and 20 dB or less was achieved in 74.6%. The postoperative incidence of SNHL was 2.9%. The mean ABG was 11.5 dB postoperatively compared with 28 dB, preoperatively. The mean BC was 34 dB postoperatively compared with 30.6 dB preoperatively, and the mean AC was 45.7 dB postoperatively compared with 58.7 dB preoperatively.

There were 16 cases of postoperative SNHL in the entire series (2.9%), defined as a change in the BC PTA of 15 dB or higher. These 16 cases were not included in the assessment of the postoperative ABG (n = 522) but were included in the assessment of the postoperative AC and BC thresholds (n = 538 cases). The 16 SNHL cases were not included in the assessment of the postoperative ABG because ABG may be perfectly closed after surgery despite SNHL. Instead of being too optimistic, we filtered results to exclude these cases from the analysis.

The postoperative 4-frequency ABG was closed to 10 dB or less in 331 cases (63.4%) and to 20 dB or less in 389 cases (74.6%). The postoperative 4-frequency average ABG was 11.5 dB compared with 28 dB preoperatively (mean difference, -16.5 dB; 95% CI, -17.9 to -15.5 dB, $p < 0.0001$). The postoperative 4-frequency AC threshold was 45.7 dB compared with 58.7 dB preoperatively (mean difference, -13 dB; 95% CI, -14.6 to -11.4 dB, $p < 0.0001$). The postoperative 4-frequency BC threshold was 34 dB compared with 30.6 dB preoperatively (mean difference, 3.5 dB; 95% CI, 2.5 to 4.4 dB, $p < 0.0001$), and overclosure (postoperative improvement of BC > 10 dB) occurred in 12 cases (2.2%). At 4 kHz, the mean postoperative AC threshold was 57.9 dB compared with 66.3 dB preoperatively (mean difference, -8.5 dB; 95% CI, -10.1 to -6.8 dB, $p < 0.0001$), and the mean postoperative BC threshold was 44.9 dB compared with 40.8 dB preoperatively (mean difference, 4.1 dB; 95% CI, 2.9-5.3 dB, $p < 0.0001$).

Table 4. Hearing results in 284 cases with at least a 1-year follow-up

Variables	Preoperatively	Postoperatively	Mean difference (95% CI)	P
ABG ≤ 10 dB, %		65		
ABG 11-20 dB, %		9		
ABG ≤20 dB, %		26		
Mean ABG, dB	27.5	11	-16.5 (-18.5 to -14.6)	<0.0001
Mean BC, dB	31.8	35.3	3.5 (2.4 to 4.5)	<0.0001
Mean AC, dB	59.3	46.2	-13.1 (-15.3 to -10.9)	<0.0001
BC at 4 kHz, dB	42.1	46.8	4.7 (3.1 to 6.3)	<0.0001
AC at 4 kHz, dB	67.2	59.1	-8.1 (-10.4 to -5.8)	<0.0001

Hearing Results in 284 Cases With at Least 1-Year Results

As outline previously, there were 16 cases of postoperative SNHL in the series (Table 4). One year or later after surgery, 6 of these 16 cases were still being followed up. These 6 cases were not included in the assessment of the postoperative ABG (n = 278 cases) but were included in the assessment of the postoperative AC and BC thresholds (n = 284 cases). The mean follow-up for this group was 44.2 months (range, 12-164.9 mo). The postoperative 4-frequency ABG was closed to 10 dB or less in 181 cases (65%) and to 20 dB or less in 206 cases (74%). The postoperative 4-frequency average ABG was 11 dB compared with 27.5 dB preoperatively (mean difference, -16.5 dB; 95% CI, -18.5 to -14.6 dB, $p < 0.0001$). The postoperative 4-frequency AC threshold was 46.2 dB compared with 59.3 dB preoperatively (mean difference, -13.1 dB; 95% CI, -15.3 to -10.9 dB, $p < 0.0001$). The postoperative 4-frequency BC threshold was 35.3 dB compared with 31.8 dB preoperatively (mean difference, 3.5 dB; 95% CI, 2.4-4.5 dB, $p < 0.0001$), and overclosure (postoperative improvement of BC > 10 dB) occurred in 7 cases (2.4%). At 4 kHz, the mean postoperative AC threshold was

59.1 dB compared with 67.2 dB preoperatively (mean difference, -8.1 dB; 95% CI, -10.4 to -5.8 dB, $p < 0.0001$), and the mean postoperative BC threshold was 46.8 dB compared with 42.1 dB preoperatively (mean difference, 4.7 dB; 95% CI, 3.1-6.3 dB, $p < 0.0001$).

Table 5. Long-term hearing results in the entire series of 538 cases.

Variables		Results at 1 yr (n = 197)	Results at 10 yr (n = 39)	Mean difference (95% CI)	P
Mean ABG, dB	Preoperatively	27.2	27.6	0.075 (-3.6 to 3.8)	0.97
	Postoperatively	11	11.7	-0.7 (-5.4 to 4.0)	0.77
Postoperative ABG, %	≤10 dB	67.5	65.8	1.8	0.89
	11-20 dB	15.5	7.8	7.7	
	>20 dB	17	26.4	-9.4	
Mean BC, dB	Preoperatively	31.3	32.8	-1.5 (j6.0 to 3.0)	0.52
	Postoperatively	33.9	40.6	-6.8 (-12.1 to -1.5)	0.01
Mean AC, dB	Preoperatively	59	60.4	-1.5 (-6.9 to 4.1)	0.61
	Postoperatively	45	52.4	-7.4 (-14.1 to -0.82)	0.03
BC at 4 kHz, dB	Preoperatively	41.8	46.3	j4.5 (j10.9 to 2.0)	0.18
	Postoperatively	45.5	53.3	0.075 (-3.6 to 3.8)	0.04
AC at 4 kHz, dB	Preoperatively	67.3	69.7	-2.5 (-9.0 to 4.1)	0.46
	Postoperatively	57.5	67.3	-9.8 (-17.6 to -2.0)	0.02

Long-Term Hearing Results in the Entire Series of Available Data (538 Cases)

Table 5 shows the audiometric results of the entire series from 1 to 10 years of follow-up. Not all data were available for every case. One hundred ninety-seven cases were tested at 1 year postoperatively, and 39 cases had follow-up data for 10 years. As outlined previously, there were 16 cases of postoperative SNHL in our series. One year after surgery, 3 of these 16 cases were still being followed up and were not included in the assessment of the postoperative ABG (n = 194) but were included in the assessment of the postoperative AC and BC thresholds (n = 197). Ten years after surgery, 1 of these 16 cases was still being followed up and was not included in the assessment of the postoperative ABG (n = 38) but was included in the assessment of the postoperative AC and BC thresholds (n = 39). Mean values of the postoperative ABG show a slight decrease of hearing improvement with time. At 10 years, successful closure of the ABG to 10 dB was present in 65.8% of cases compared with 67.5% of cases at 1 year. A slight progression of hearing loss with time was observed because the average 4-frequency postoperative gain in AC was 8 dB after 10 years of follow-up compared with 14 dB after 1 year. The gain in AC at 4 kHz was 2.4 dB after 10 years compared with 9.8 dB after 1 year. A continuous progression of sensorineural component was also observed for years because the mean 4-frequency BC threshold was 40.6 dB at 10 years compared with 33.9 dB at 1 year. The postoperative BC threshold at 4 kHz was 53.3 dB after 10 years of follow-up compared with 45.5 dB after 1 year, and the BC deterioration was 7 dB after 10 years of follow-up compared with 3.7 dB after 1 year.

Table 6. Long-term hearing results of a series of 32 cases during 10 years: preoperative and/or postoperative values of SNHL, ABG, AC, and BC

Variables		Results at 1 yr	Results at 10 yr	Mean difference (95% CI)	P
Mean ABG, dB	Preoperatively	28.5	28.5	0.0 (-5.3 to 5.3)	0.97
	Postoperatively	5	11.2	-6.2 (-12.2 to -0.14)	0.77
Postoperative ABG, %	≤10 dB	84.3	65	19.3	0.89
	11-20 dB	3.2	10	-6.8	
	>20 dB	12.5	25	-12.5	
Mean BC, dB	Preoperatively	31.6	31.6	-1.5 (j6.0 to 3.0)	0.52
	Postoperatively	31.8	39.5	-6.8 (-12.1 to -1.5)	0.01
Mean AC, dB	Preoperatively	60	60	-1.5 (-6.9 to 4.1)	0.61
	Postoperatively	36.8	50.7	-7.4 (-14.1 to -0.82)	0.03
BC at 4 kHz, dB	Preoperatively	44.3	44.3	j4.5 (j10.9 to 2.0)	0.18
	Postoperatively	46.5	51.8	0.075 (-3.6 to 3.8)	0.04
AC at 4 kHz, dB	Preoperatively	69	69	-2.5 (-9.0 to 4.1)	0.46
	Postoperatively	52.5	65.4	-9.8 (-17.6 to -2.0)	0.02

Long-Term Hearing Results of a Series of 32 Cases With Data Available at 1 Year and at 10 Years of Follow-Up

To assess deterioration of early results, we compared the results of a cohort of patients at 1 year with the results of the same cohort at 10 years of follow-up. Thirty-two cases had audiologic data available at the 1- and 10-year marks. Table 6 presents the long-term results of these 32 cases during 10 years of follow-up. The power of this analysis is low regarding the number of cases, but mean values of the postoperative ABG show a slight degradation of hearing improvement with time. At 10 years, successful closure of the mean ABG to 10 dB was present in 65% of cases compared with 84.3% of cases at 1 year. A progressive hearing loss with time was observed because the average postoperative gain in AC was 9.3 dB after 10 years of follow-up compared with 23.2 dB after 1 year. A continuous progression of sensorineural component was also observed during the years because the mean BC thresholds was 39.5 dB at 10 years compared with 31.8 dB at 1 year. The average annual deterioration was higher for AC (1.54 dB/yr) than the one observed for BC (0.85 dB/yr).

7

Oval Window Fistula

Of the entire series, of 652 cases, perilymph fistula was identified in 36 (5.5%). In all cases, a fistula was suspected preoperatively as evidenced by the presence of severe dizziness associated with a positive fistula sign. Postoperative fistula may cause SNHL and more severe problems as observed in this series. Of the 36 cases explored for perilymph fistula, 12 cases had total preoperative SNHL (33.4%), 22 cases (61%) had no ABG preoperatively, and 2 cases (5.6%) had a preoperative ABG 20 dB or less. The surgical technique used during the previous surgery was identified in 35 cases as total stapedectomy in 21 cases (60%) and stapedotomy in 14 cases (40%). Of these 36 cases, postoperative data were available in 29 cases (80.5%). Mean follow-up was 32.8 months (range, 3-129.1 mo). Of the 29 cases, symptoms of dizziness were alleviated in 28 cases (96.5%). The remaining case of persistent postoperative dizziness underwent another revision operation 1 year after, and a recurrent oval window fistula was identified. This was closed with another vein graft. After surgery, symptoms of dizziness were alleviated in this case during a follow-up period of 18 months. In the group of revision surgery after initial primary surgery, fistula was identified in 23 cases, and postoperative data were available in 18 cases. Of the 18 cases, the postoperative ABG was closed to 10 dB in 83.4% of cases (15 cases), and no severe postoperative SNHL was observed with a mean follow-up of 29 months (range, 3-95.7 mo). The postoperative mean ABG was 5.1 dB compared with 1.6 dB preoperatively. The mean postoperative BC thresholds was 40 dB compared with 35.5 dB preoperatively, and the postoperative AC thresholds was 45 dB compared with 37 dB preoperatively. In our personal revision series after primary surgery (90 cases), postoperative fistula occurred in 8 cases. These 8 cases were all revised by the first author, and fistula were identified in all cases at the anterior pole of the oval window niche and were all related to a vein graft which was not perfectly adherent to the anterior pole of the footplate.

DISCUSSION

Our results showed that improvement of a significant conductive hearing loss after unsuccessful primary or revision stapes surgery can be accomplished in most cases. Although revision surgery may be clinically required when perilymph fistula is suspected, on the basis of the presence of dizziness with fistula sign, several factors may dissuade surgeons from performing another procedure. First, it is well recognized that postoperative hearing results in revision surgery are poorer than those in primary procedures. Second, revision is often technically far more challenging than primary stapedectomy. The surgical experience of the surgeon plays an important role because the technique of repair is usually determined by the surgical findings at the time of revision. Third, revision is associated with a higher risk of potential cochlear trauma with subsequent postoperative SNHL. Surgical findings in our study are consistent with the previously published series of revision surgery[1-3,6,8-14], in that the most common causes of failure of the primary surgery were incus erosion (27.6%) and prosthesis displacement (18.2%). Short and long prosthesis were identified in 12.7% and 4.6% of cases respectively. Most probably, many of these prosthesis displacements and improper length prostheses could have been avoided if the prosthesis has been firmly attached to the incus or if the proper length of the prosthesis has been selected on the basis of an accurate measurement of the distance between the incus and the stapes footplate. In 12.7% of cases, the cause of failure was related to a previous operation that was not entirely completed (8.7%) or to a stapes refixation (4%) in patients who had mobilization as the primary operation. Again, these failures could have been avoided during the primary procedure. Perilymph fistula was identified in 36 cases. As previously described[11], passage of time does not protect against the occurrence of fistula because in the series of Somers et al.[11], the mean delay was 5 years and the longest delay found was 13 years. In our personal revision series after primary surgery (90 cases), perilymph fistula was observed in 8 cases. Of the 8 cases, 6 cases occurred within the first 6 months after initial primary surgery and the remaining 2 cases were observed after 1 year and 3 years. These observations also show that postoperative perilymph fistula may also occur after vein graft interposition, which was used in all cases during the primary surgery by the first author. Revision surgery was performed without great difficulty for most cases in this series. Like others, we recommend the use of a fiber-optic handheld laser to enable easier and safer revision surgery[1,2,6,12] by vaporizing adhesions, dissecting the previous prosthesis free atraumatically, and reopening the oval window membrane. A significant proportion of the patients experienced hearing improvement because the postoperative ABG was closed to within 10 dB in 63.4% of cases. However, this is significantly less than in our primary stapedotomy patients where an ABG closure to within 10 dB was achieved 94.2% of cases.[4] The mean 4-frequency ABG, the AC, and the BC thresholds improved significantly after surgery but certainly not to reference levels. Air-bone gap closure does not universally improve the hearing thresholds enough to avoid the use of conventional hearing aids in addition to surgery. However, by improving the hearing thresholds, even partially, revision surgery may significantly facilitate the use of an appropriate hearing aid. A significant post-operative SNHL (>15 dB) was observed in 2.9% of cases in this series compared with 0.5% in our primary otosclerosis series.[4] The average annual AC and BC deterioration rates for 10 years were 1.54 and 0.85 dB/yr, respectively, which are higher than in our previously published series of primary otosclerosis, which were 0.4 dB/yr for AC and 0.36 dB/yr for BC.[4] This is consistent with other authors who reported a long-term mild deterioration of hearing results in revision surgery.[2] Our results are comparable with previously published series of revision stapes surgeries[1-3,6,8,9,11-14], reporting a success rate of 24% to 80% for ABG closure to within 10 dB and a postoperative incidence of SNHL varying from 0% to 7.7% of cases (Table 7).

Table 7. Hearing results of previously published series of revision stapes surgery

Series	n	Postoperative ABG e 10 dB, %	Postoperative SNHL, %
Gros et al[1].	63	52.4	1
De La Cruz and Fayad[2]	356	59.8	7.7
Han et al.[3]	74	45.6	5.4
Lippy et al.[6]	483	71	2
Hammerschlag et al. (8)	250	80	2
Berenholz et al. (9)	78	67.6	1.3
Somers et al. (11)	226	40	1.3
Wiet et al. (12)	23	52	0
Magliulo et al. (13)	63	24	3.1
Nissen (14)	21	43	1
Current series	652	63.4	2.9

These disparities in hearing results may be dependent not only on the cause of failure[1,2,6,11] but also on preoperative factors such as the timing of failure after initial surgery or the number of previous operations.[1,2,6,9] These preoperative factors should be studied accordingly to identify possible predictive factors. There were 49 cases of unidentified cause for ABG of greater than 10 dB in this series. These cases should have been examined by computed tomography to exclude superior semicircular canal dehiscence. Since the initial description by Minor et al.[15] in 1998, refinements in vestibular and radiologic testing have identified an increasing number of patients with superior semicircular canal dehiscence. As their current prospective study started in 1992, computed tomography was therefore not routinely performed in the first 6 years of this study. However, this is relatively rare entity, and there would still be a lot of cases with no demonstrable reason for an ABG.

In selected cases, the indication for revision surgery is not always hearing improvement but may include dizziness and a suspected perilymph fistula. This was the case for 36 patients in our series. In all cases, a fistula was suspected preoperatively as evidenced by the presence of severe dizziness associated with a positive fistula sign. However, because a fistula may exist without a positive fistula sign, the related preoperative assessments are not regarded as being reliable. Postoperatively, symptoms of dizziness were alleviated in all cases, and these patients remained clinically stable, thus improving their quality of life. The major strength of the study is that we report a large number of consecutive patients undergoing revision surgery for otosclerosis, who were followed subsequently, allowing us to study long-term (10 yr) results. Possible limitations should, however, also be discussed. First, all cases were operated on by the same surgeon in the same institution, so data are not, per definition, generalizable. Second, this study is descriptive and gives no clues, as yet, which patients might benefit to a greater or lesser degree from revision surgery. Further studies should focus on determining predictive factors of hearing outcome. This can then assist the surgeon to better select the patients, anticipate surgical findings, and estimate realistic expectations for better patient counseling. It would be worthwhile to develop a prognostic model predicting which patients benefit more or less from revision otosclerosis surgery.

CONCLUSION

Revision stapes surgery, although not as satisfactory as a primary procedure, seems to significantly increase hearing. However, the mean hearing improvement in AC was only 10 dB in the long-term assessment. Although this type of surgery remains challenging, there is a low risk of SNHL and a two-thirds chance of hearing improvement. For patients with insufficient hearing improvement, conventional hearing aids may provide additional auditory amplification.

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Part V

Expert surgical technique

Chapter 8

Ossiculoplasty in intact stapes and malleus patients: a comparison of PORPs versus TORPs with malleus relocation and Silastic banding techniques.

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ABSTRACT

Objectives: To compare hearing results in patients undergoing ossiculoplasty using either partial ossicular replacement prosthesis (PORP) or total ossicular replacement prosthesis (TORP) with Silastic banding and malleus relocation techniques in cases with malleus and stapes both present and mobile.

Study Design: Prospective nonrandomized clinical study.

Setting: Tertiary referral center.

Methods: Five hundred eighty-five patients undergoing ossiculoplasty were enrolled in this study from April 1991 to May 2010. Comparative analyses were made between a group of 304 patients who underwent ossiculoplasty with partial prosthesis positioned from the malleus to the stapes head and 281 patients who underwent ossiculoplasty with total prosthesis positioned from the malleus to the stapes footplate. Preoperative and postoperative audiometric evaluation using conventional audiometry, that is, air-bone gap (ABG), bone-conduction thresholds, and air-conduction thresholds were assessed.

Results: In the PORP group, the mean postoperative ABG was 13.1 dB compared with 8.9 dB in the TORP group, (95% confidence interval [CI], 2.2-6.2 dB; $p <= 0.001$). Fifty-four percent of patients from the PORP group had a postoperative ABG of 10 dB or less, compared with 68.9% in the TORP group (mean difference, 14.6%; 95% CI, 6%-23%; $p < 0.001$). The postoperative ABG was closed to within 20 dB in 70.4% of cases in the PORP group compared with 86.9% in the TORP group (mean difference, 14.5%; 95% CI, 10%-23%; $p < 0.001$).

Conclusion: In patients with an absent incus and intact stapes and malleus, ossicular reconstruction with TORP combined with our malleus relocation and Silastic banding technique results in significantly better hearing outcomes compared with reconstructions with PORP.

Reconstruction of the ossicular chain when the stapes and malleus are both present typically involves the use of a partial reconstruction prosthesis between the stapes head and the malleus handle or tympanic membrane (TM) with or without supportive tissue to strengthen the tympanic membrane. Few authors advocate the use of a total reconstruction prosthesis from the footplate of a normal stapes to the malleus (1-5).

During the period from April 1991 to May 2010, approximately 1,756 consecutive ossiculoplasty procedures were performed by the same surgeon (R. V.) in the same tertiary referral center. Of these, 832 (47%) were performed with both an intact stapes and intact malleus handle. The aim of this prospective study was to compare hearing results in patients undergoing ossicular reconstruction in this specific ossicular chain condition using either a partial ossicular replacement prosthesis (PORP) or a total ossicular replacement prosthesis (TORP).

MATERIALS AND METHODS

Patients

This is a prospective study of 700 patients who underwent 832 ossicular reconstructions for ossicular chain defects with both malleus and stapes intact. All patients were treated between April 1991 and May 2010 and were operated on by the same surgeon. Austin's (6) classification of ossicular defects as modified by Kartush (7) was used to define the ossicular status encountered. In this analysis, those patients allocated to Austin-Kartush Group A (stapes and malleus present) and patients having PORP and TORP implantation from the malleus and stapes head/footplate were selected. Patients having other types of prostheses than PORP or TORP and patients who had procedures in which the prosthesis' head was applied directly to the TM and not to the malleus handle (TM to stapes head or mobile footplate procedure) were excluded (204 cases).

Six hundred seventeen patients who underwent 628 ossicular reconstructions for ossicular chain defects using PORP or TORP were included in this study. Patients were divided in 2 groups according to the type of prosthesis used. For the PORP group, 304 patients were included with a total of 310 operated ears. Of these 310 cases, there were 237 primary operations and 73 revision operations. All PORPs were interposed between the malleus handle and the stapes head. In the TORP group, there were 313 patients with a total of 318 operated ears. Of these 318 cases, there were 190 primary operations and 128 revision operations. All TORPs were interposed between the malleus handle and the stapes footplate.

All data were tabulated using the Otology-Neurotology Database (AS Multimedia, Inc., Cassagne, France) (8). This is a commercially available software package developed at our center, designed to comply with the American Academy of Otolaryngology guidelines for reporting clinical and audiometric results (9).

Surgery

All procedures in both groups were performed by the same surgeon (R. V.) via a transcanal approach. The choice of the prosthesis (PORP or TORP) used for ossicular reconstruction was dictated by the progressive experience and surgical technique of the surgeon, and subsequently, the introduction of 2 original, previously published, techniques: malleus relocation (10) and Silastic stabilizing properties that were shown with the Silastic banding technique (11). Before the introduction of malleus relocation and Silastic banding techniques, the author preferred to use a partial prosthesis. After introducing these 2 techniques, the use of a TORP became the preferred method of ossicular reconstruction, despite the presence of a stapedial arch. All PORPs and TORPs were positioned underneath the malleus, which was used in all cases in both groups.

In the PORP group, a standard technique was used using a Flex/HA PORP (Medtronic-Xomed, Inc., Jacksonville, FL, USA) in all cases (310 cases). The prosthesis' shaft was cut to the appropriate length, and the PORP was placed from the malleus handle to the stapes head. These patients were enrolled in the study between April 1991 and August 2000. In 1999, the malleus relocation technique was introduced by the author to achieve an improved vertical position of the middle ear prosthesis (10). The malleus relocation technique was used in 131 cases of the PORP group and was not used in 179 cases. However, the malleus was used in all cases. Within the group overall, 137 ears (44%) required TM grafting. The main reasons for grafting were TM perforation, atrophic TM, and TM retraction. In all cases, tragal perichondrium was inserted as an underlay graft to repair the perforation.

In the TORP group, ossiculoplasty was performed with a Flex/HA TORP (Medtronic-Xomed, Inc.) in all cases (318 cases) despite the presence of a stapedial arch. The prosthesis shaft was cut to the appropriate length, and the TORP was positioned from the malleus handle to the stapes footplate. The prosthesis shaft was inserted between the fallopian canal and the stapes superstructure with the distal end of the prosthesis shaft in direct contact with the stapes footplate. When there was not enough room between the horizontal segment of the facial nerve and the stapes superstructure to place the shaft of the prosthesis, the TORP was inserted between the promontory and the suprastructure. These patients were enrolled in the study between January 2000 and May 2010. In 2000, the Silastic banding technique was introduced by the author to further stabilize the prosthesis (11). A combination of both techniques of malleus relocation and Silastic banding was used in all cases in this group (318 cases). Within the group overall, 153 ears (48%) required TM grafting for atrophic TM, perforation, or retraction of the TM. In all cases, tragal perichondrium was inserted as an underlay graft.

Malleus Relocation Technique

As previously described (10), after dissecting the malleus free from the TM, the tensor tympani was sectioned as close as possible to its insertion into the malleus handle. Entire separation of the malleus from the TM was required (Figure 1). Any remnant of the incus (even within the epitympanum) was removed. Relocation was done using a strong 90-degree hook placed anterior to the neck of the malleus. Progressive posterior retraction of the malleus was applied until it came to lie directly above the stapes capitulum or footplate. Subsequent anterior retraction of the malleus was avoided by overstretching the anterior malleal ligament, the position thereby being maintained by the superior ligament of the malleus, which was preserved. In the relocated position, the malleus should lie immediately over the stapes footplate (Figure 2). Precise measurement from the relocated malleus to the stapes capitulum or footplate can then be undertaken using a modified stapes-measuring rod. Placement of the shaft of the prosthesis was performed first, followed by easy positioning of the malleus handle

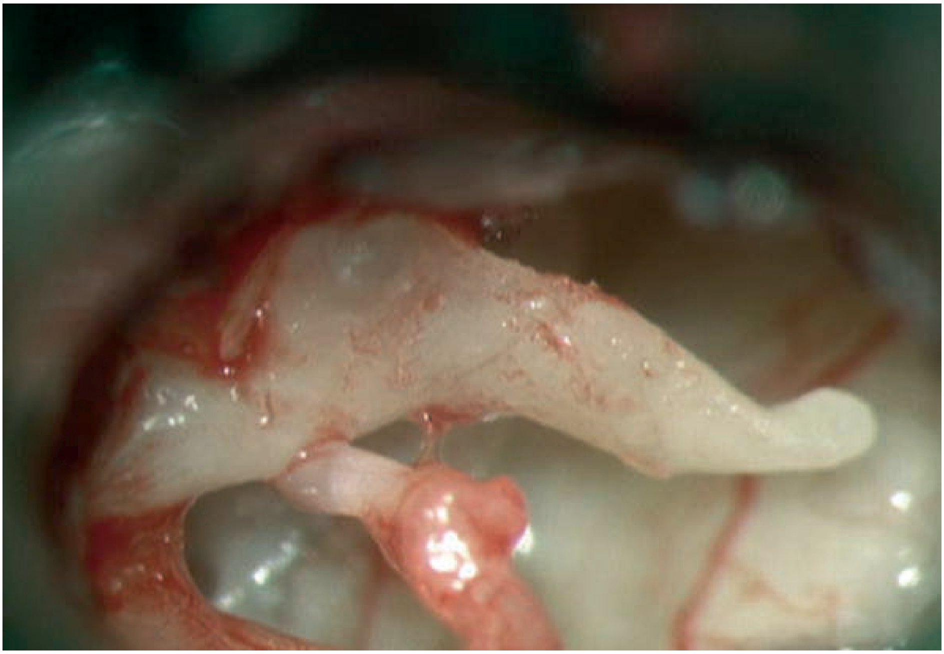

Figure 1. Operative view of a right ear. The malleus is dissected free from the TM. Entire separation of the malleus from the TM is required.

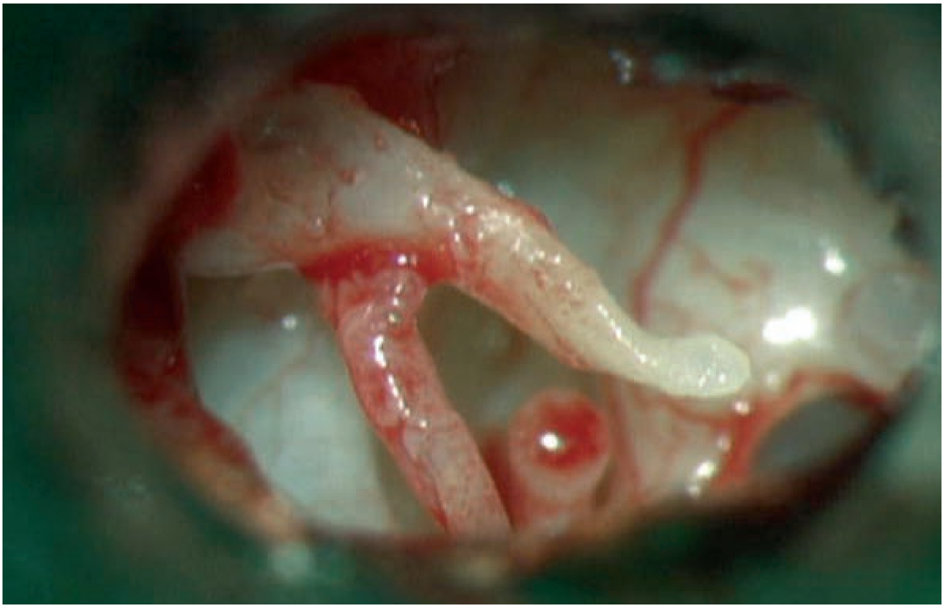

Figure 2. The relocated malleus should lie immediately above the stapes capitulum or footplate.

into the groove of the prosthesis head. In its final position, the prosthesis' head should rest directly under the malleus handle without undue tension, and the malleus neck should stay away from the superior wall of the external auditory canal.

Silastic Banding Technique

The technique for Silastic banding stabilization has previously been described (11) and was used in 287 cases within this study. A Silastic band was fashioned by creating a 1.2-mm disc with a specially designed punch (Medtronic-Xomed, Inc.) from a thin sheet of Silastic. The center of the disc was then fenestrated with a 0.8-mm diameter punch (Figure 3A). The band should fit snugly around the stapes neck. Because the band needs to be placed beneath the stapedius tendon to avoid lateral displacement, the tendon is sectioned as close to the pyramidal eminence as possible. The prosthesis shaft was cut to the appropriate length and was introduced through the band (Figure 3B). The shaft was introduced between the stapes and the fallopian canal with the distal end being lowered onto the center of the footplate (Figure 4). The prosthesis' head was then introduced under the relocated malleus handle (Figure 5). The band is then gently pulled and positioned around the stapes capitulum, beneath the stapes tendon which had been cut. By snugging the band down over the sectioned tendon, the prosthesis became firmly apposed to the stapes, allowing precise placement on the stapes footplate with sufficient rigidity (Figure 6). This assembly facilitates an almost perpendicular position of the prosthesis from the footplate to the malleus handle and TM.

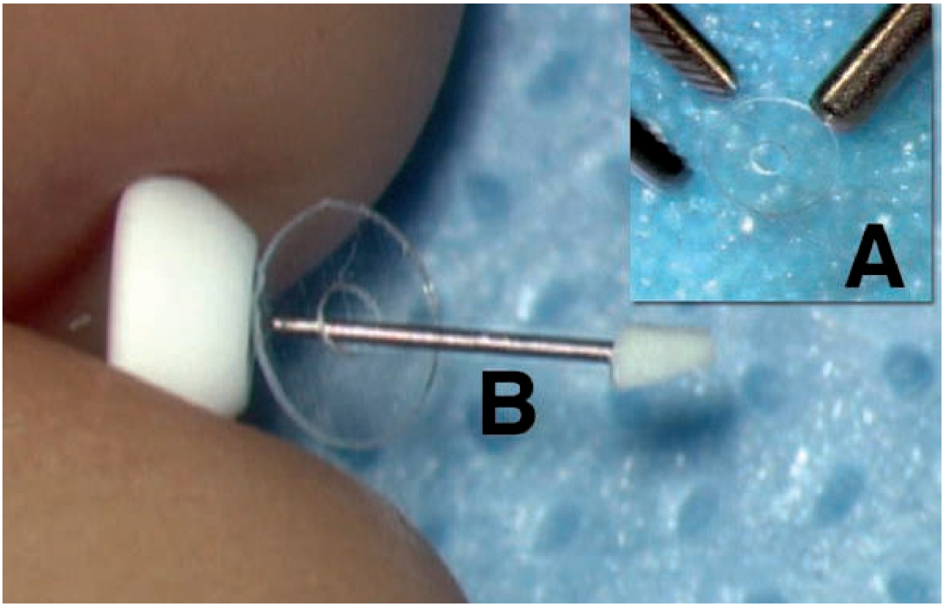

Figure 3A+B. A Silastic band is fashioned by creating a 1.2-mm disc with a specially designed punch from a thin sheet of Silastic (A). The prosthesis shaft was cut to the appropriate length and was introduced through the band (B).

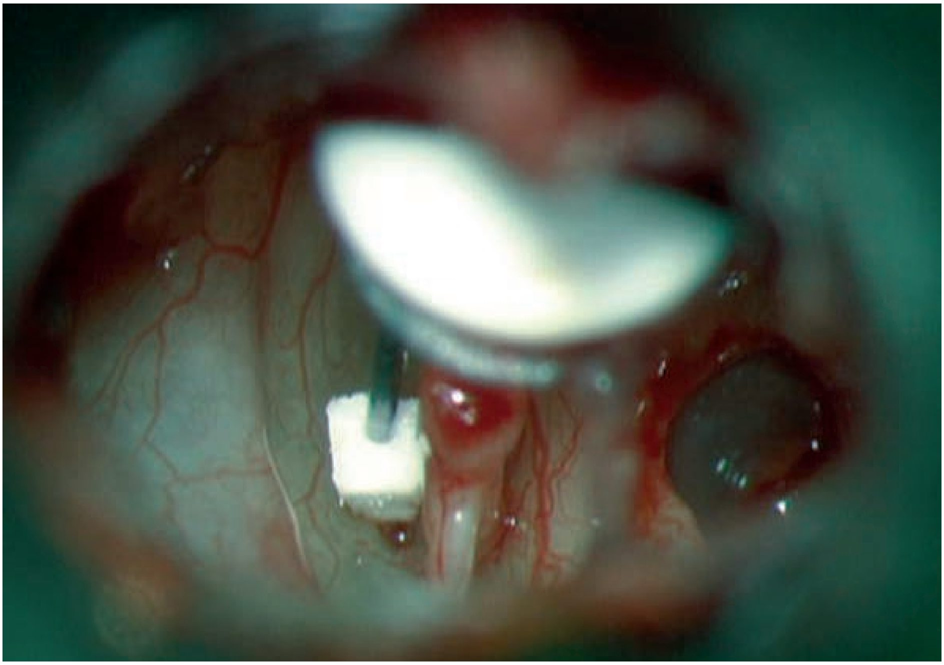

Figure 4. The prosthesis' shaft is introduced between the stapes and the fallopian canal, with the distal end being lowered onto the center of the footplate.

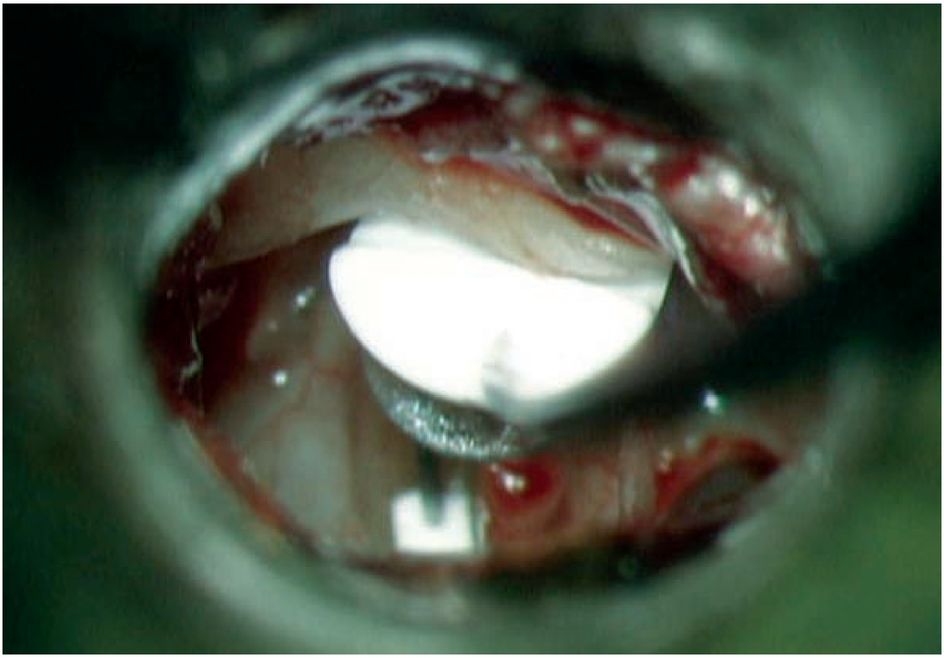

Figure 5. The prosthesis' head is introduced under the relocated malleus handle.

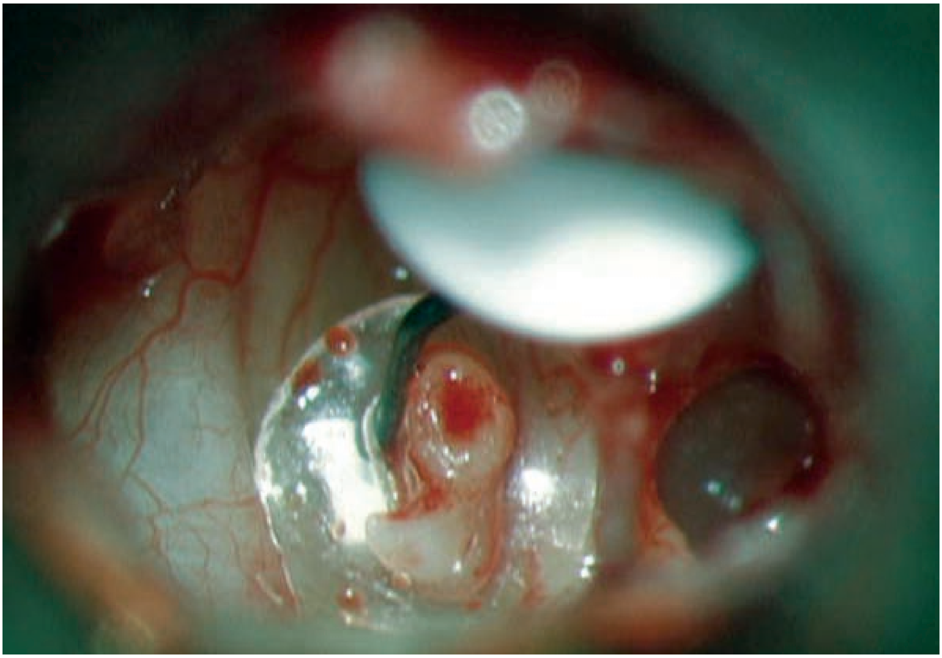

Figure 6. By snugging the band down over the sectioned stapes tendon, the prosthesis is firmly opposed to the stapes.

By keeping the distal end of the prosthesis in proper position using the Silastic band, the tendency of the prosthesis to tip or migrate is virtually eliminated. We think this is the most significant advantage of this technique.

Audiometric Assessment

Audiometric evaluation included preoperative and postoperative air-bone gap (ABG), air-conduction (AC) thresholds, and bone-conduction (BC) thresholds. Only AC and BC results that were obtained at the same time postoperatively were used for calculation of ABG and air-bone pure-tone average. Four-frequency pure-tone averages for bone and air thresholds, 0.5, 1, 2, and 4 kHz, obtained at the last follow-up visit, were used. Pure-tone averages for bone and air thresholds at 4 kHz alone also were analyzed separately. Audiometric data also were discussed for cases with a minimum follow-up of 12 months and at 5 years, where available. Audiometry was reported according to American Academy of Otolaryngology-Head and Neck Surgery guidelines (9), except for thresholds at 3 kHz, which were substituted in all cases with those at 4 kHz.

Statistical Analyses

Mean differences and percentage differences and their 90% confidence intervals (CIs) were calculated. Furthermore, Student's *t* tests or Wilcoxon tests and $[chi]^2$ tests were used to evaluate potential differences in mean values and percentages between the 2 groups. To adjust for potential confounders (e.g., age and sex), multivariate linear regression analyses were performed. All analyses were performed using SPSS (version 15.0; SPSS, Inc., Chicago IL, USA).

RESULTS

Patient Characteristics

Table 1 shows the preoperative patient characteristics for the 2 groups. In the PORP and TORP groups, 57% and 62% were female subjects, and the mean ages were 48.1 and 52.1 years, respectively. There were 51 senior patients (age, ≥65 yr) within the PORP group and 62 in the TORP group. There were 11 children (age, ≤18 yr) within the PORP group and 14 in the TORP group. Preoperative BC, AC, and ABG were similar in both groups. The mean preoperative BC threshold within the PORP group was 26 and 25.6 dB in the TORP group. The mean preoperative AC thresholds were 50.7 dB in the PORP group and 50.4 dB in the TORP group. The preoperative ABG was 24.6 dB in the PORP group and 24.9 dB in the TORP group. There was no statistical difference between the groups.

Figure 7 shows the pathology and ossicular chain status at the time of surgery for each group. Of the 310 cases of the PORP group, there were 285 cases of chronic suppurative otitis media (CSOM; 92%) and 25 cases of ossicular chain problems secondary to head trauma (8%).

Table 1. Preoperative patient characteristics in partial ossicular replacement prosthesis and total ossicular replacement prosthesis groups

Variable	Partial ossicular replacement prosthesis group (n = 310)	Total ossicular replacement prosthesis group (n = 318)	p
Age (yr), (min-max)	48.2 (5-95)	52.1 (7-80)	0.02
Sex (% female)	57	62	0.19
Mean BC (dB)	26	25.6	0.63
Mean AC (dB)	50.7	50.4	0.85
Mean ABG (dB)	24.6	24.9	0.71
BC at 0.5 kHz (dB) 1	7.2	17.5	0.72
BC at 1 kHz (dB)	20.2	21.4	0.21
BC at 2 kHz (dB)	29.1	29.3	0.87
BC at 4 kHz (dB)	37.7	34.1	0.02
AC at 0.5 kHz (dB)	44.7	43.9	0.52
AC at 1 kHz (dB)	48.4	48.8	0.76
AC at 2 kHz (dB)	51.3	50.8	0.71
AC at 4 kHz (dB)	58.3	58.4	0.98
ABG at 0.5 kHz (dB)	27.4	26.3	0.25
ABG at 1 kHz (dB)	28.2	27.3	0.36
ABG at 2 kHz (dB)	22.2	21.5	0.41
ABG at 4 kHz (dB)	20.6	24.2	<0.001

ABG indicates air-bone gap; AC, air conduction; BC, bone conduction.

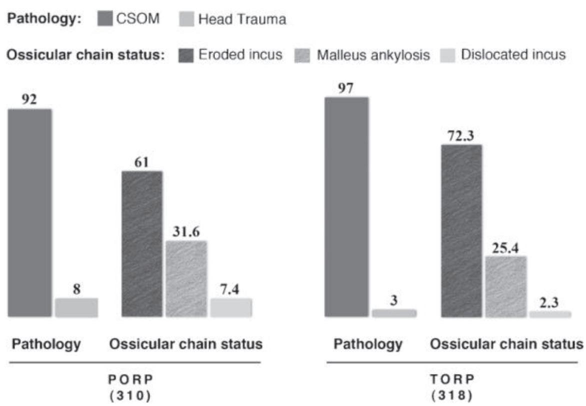

Figure 7. Pathology and ossicular chain status in PORP and TORP groups (%).

Incus erosion was identified in 189 cases (61%), malleus ankylosis in 98 cases (31.6%), and incus dislocation in 23 cases (7.4%). Of the 285 cases of CSOM, incus erosion was identified in 189 cases (66.3%) at the time of surgery, and malleus ankylosis was observed in 96 cases (33.7%). Of the 25 cases of post-head injury hearing loss, incus dislocation was seen in 23 cases (92%), and malleus ankylosis occurred in 2 cases (8%). Of the 310 operated ears within the PORP group, 73 were revision cases.

Of the 318 cases of the TORP group, there were 308 patients with history of CSOM (97%) and 10 cases with history of post-head trauma hearing loss (3%). Incus erosion was identified in 230 cases (72.3%), malleus ankylosis in 81 cases (25.4%), and incus dislocation in 7 cases (2.3%). Of the 308 cases of CSOM, incus erosion was seen in 230 cases (74.7%) at the time of surgery, and malleus ankylosis was identified in 78 cases (25.3%). Of the 10 cases of post-head injury hearing loss, incus dislocation was observed in 7 cases (70%) and malleus ankylosis in 3 cases (30%). One hundred twenty-eight operated ears within the TORP group were revision cases.

Postoperative Audiometric Assessment

Overall Results

The postoperative hearing results for both groups are shown in Table 2. Of the 310 cases of the PORP group, postoperative data were available for 288 cases (93%). Fifty-eight percent within this group were female subjects with an average time to follow-up of 43.6 months (range, 3-208 mo). The mean age was 48.7 years (range, 5-95 yr). Of the 318 cases of the TORP group, postoperative data were available for 283 cases (89%). Sixty-four percent of patients within this group were female subjects, and the average time to follow-up was 21.7 months (range, 3-121.8 mo). The mean age was 52.5 years (range, 7-80 yr).

There was only 1 case of postoperative sensorineural hearing loss (SNHL), defined as a worsening of the BC PTA of 15 dB or more. This occurred in the PORP group (1 [0.3%] of 288 cases). No SNHL was observed in the TORP group. The incidence of prosthesis extrusion was 3.4% in the PORP group (10 cases) compared with 4.2% in the TORP group (12 cases). In the PORP group, the postoperative 4-frequency average ABG was 13.1 dB compared with 8.9 dB in the TORP group (mean difference, 4.2 dB: 95% CI, 2.2-6.2; $p < 0.001$). The postoperative ABG was closed to 10 dB or less in 156 cases (54.3%) and to 20 dB or less in 202 cases (70.4%) in the PORP group compared with 195 cases (68.9%) and 246 cases (86.9%), respectively, in the TORP group (mean difference, 14.6%; 95% CI, 6-23; $p < 0.001$; and mean difference 14.5%; 95% CI, 10-23; $p < 0.001$, respectively). The mean postoperative gain in AC threshold within the PORP group was 9.8 dB compared with 16.3 dB in the TORP group (mean difference,

Table 2. Postoperative overall hearing results in partial ossicular replacement prosthesis and total ossicular replacement prosthesis groups

Variable	Partial ossicular replacement prosthesis (n = 288)	Total ossicular replacement prosthesis (n = 283)	Mean or percentage difference (95% confidence interval)	p
ABG <10 dB (%)	54.3	68.9	14.6 (6-23%)	<0.001
ABG <20 dB (%)	70.4	86.9	14.5 (10-23)	<0.001
Mean BC (dB)	28	25.5	2.5 (0.3-4.8)	0.03
Mean AC (dB)	41.1	34.4	6.7 (3.7-9.8)	<0.001
Mean ABG (dB)	13.1	8.9	4.2 (2.2-6.2)	<0.001
Mean change in BC (dB)	-1.7	+0.3	-2.0 (-3.1 to -0.9)	<0.001
Mean change in AC (dB)	+9.8	+16.3	-6.4 (-9.0 to -3.9)	<0.001
Mean BC at 4 kHz (dB)	42.2	35.7	6.5 (3.1-9.9)	<0.001
Mean AC at 4 kHz (dB)	56.6	45	11.5 (7.8-15.3)	<0.001
Mean ABG at 4 kHz (dB)	14.3	9.3	5.0 (2.8-7.3)	<0.001
Change in BC at 4 kHz (dB)	-4.3	-1.6	-2.7 (-4.5 to 0.9)	0.003
Change in AC at 4 kHz (dB)	+2	+13.4	11.4 (8.3Y14.4)	<0.001

ABG indicates air-bone gap; AC, air conduction; BC, bone conduction

-6.4 dB; 95% CI, -9 to -3.9; $p < 0.001$). The postoperative change in BC threshold was negligible in both groups (-1.7 dB in the PORP group and +0.3 dB in the TORP group). At 4 kHz, the postoperative ABG was 14.3 dB in the PORP group compared with 9.3 dB in the TORP group (mean difference, 5 dB; 95% CI, 2.8-7.3; $p < 0.001$).

Short-Term Hearing Results

Short-term hearing results in the PORP and TORP groups were defined as audiologic data available at 1 to 2 years of follow-up and are shown in Table 3. Not all data were available for every case. Many of our patients travel long distances for care at our center, and follow-up is sometimes not possible. Of the 310 cases of the PORP group, short-term postoperative data were available for 172 cases (55.4%). The average time to follow-up was 15.5 months (range, 12-24.3 mo). The mean age was 49.3 year (range, 5-95 yr). Of the 318 cases of the TORP group, short-term postoperative data were available for 155 cases (48.7%). The average time to follow-up was 14.8 months (range, 12-23.8 mo). The mean age was 51.3 year (range, 7-80 yr). The postoperative mean ABG was 11.2 dB in the PORP group compared with 7.2 dB in the TORP group (mean difference, 4 dB; 95% CI, 1.6-6.5; $p < 0.001$). The postoperative ABG was closed to within 10 dB in 104 cases (60.8%) in the PORP group compared with 113 cases (73.5%) in the TORP group (mean difference, 12.7 dB; 95% CI, 5-21; $p = 0.02$). It was closed to within 20 dB in 134 cases (78.4%) in the PORP group compared with 142 cases (91.6%) in the TORP group (mean difference, 13.2 dB; 95% CI, 7-19; $p <$

Table 3. Short-term postoperative hearing results in partial ossicular replacement prosthesis and total ossicular replacement prosthesis groups

Variable	Partial ossicular replacement prosthesis (n = 172)	Total ossicular replacement prosthesis (n = 155)	Mean or percentage difference (95% confidence interval)	p
ABG <10 dB (%)	60.8	73.5	12.7 (5-21)	0.02
ABG <20 dB (%)	78.4	91.6	13.2 (7-19)	0.001
Mean BC (dB)	26	24.8	1.2 (-1.6 to 4.1)	0.39
Mean AC (dB)	37.3	32	5.3 (1.5-9.2)	0.007
Mean ABG (dB)	11.2	7.2	4.0 (1.6-6.5)	0.001
Mean change in BC (dB)	0.1	+0.6	-0.5 (-1.8 to 0.8)	0.47
Mean change in AC (dB)	+13.6	+18.6	-5.0 (-8.6 to 1.6)	0.003
Mean BC at 4 kHz (dB)	40.5	34.6	5.9 (1.5-10.2)	0.008
Mean AC at 4 kHz (dB)	52.7	43	9.7 (4.7-14.6)	<0.001
Mean ABG at 4 kHz (dB)	12.1	8.4	3.7 (0.8-6.6)	0.01
Change in BC at 4 kHz (dB)	-3.3	-1.3	-2.0 (-4.4 to 0.4)	0.11
Change in AC at 4 kHz (dB)	+5	+15.4	10.4 (6.3-14.4)	<0.001

ABG indicates air-bone gap; AC, air conduction; BC, bone conduction.

0.001). The mean postoperative gain in AC threshold within the PORP group was 13.6 dB compared with 18.6 dB in the TORP group (mean difference, -5 dB; 95% CI, -8.6 to 1.6; p = 0.003). The postoperative change in BC threshold was negligible in both groups. At 4 kHz, the postoperative ABG was 12.1 dB in the PORP group compared with 8.4 dB in the TORP group (mean difference, 3.7 dB; 95% CI, 0.8-6.6; p = 0.01).

Long-Term Hearing Results

Long-term hearing results in the PORP and TORP groups were defined as audiologic data available with at least 4 years of follow-up and are shown in Table 4. Of the 310 cases of the PORP group, long-term postoperative data were available for 98 cases (31.6%). The average time to follow-up was 81.1 months (range, 48.3-119.8 mo). The mean age was 48.1 year (range, 5-95 yr). Of the 318 cases of the TORP group, long-term postoperative data were available for 42 cases (13.2%). The average time to follow-up was 65.8 months (range, 48.3-121.8 mo). The mean age was 47.5 years (range, 7-71 yr). The postoperative mean ABG was 11.7 dB in the PORP group compared with 9.2 dB in the TORP group (mean difference, 2.4 dB; 95% CI, -2.5 to 7.4; p = 0.33). The postoperative ABG was closed to within 10 dB in 60 cases (61.2%) in the PORP group compared with 27 cases (64.3%) in the TORP group (mean difference, 3.2 dB; 95% CI, -5 to 11; p = 0.73). It was closed to within 20 dB in 72 cases (73.5%) in the PORP group compared with 36 cases (85.7%) in the TORP group (mean difference, 12.2 dB; 95% CI, 5-19; p = 0.13). The mean postoperative gain in

AC threshold within the PORP group was 9.3 dB compared with 12.2 dB in the TORP group (mean difference, -2.9 dB; 95% CI, -9 to 3.2; $p = 0.35$). The postoperative BC deterioration was 3.2 dB in the PORP group compared with 2.9 dB in the TORP group (mean difference, -0.3 dB; 95% CI, -2.8 to 2.3; $p = 0.84$). At 4 kHz, the postoperative ABG was 14.4 dB in the PORP group compared with 8.6 dB in the TORP group (mean difference, 5.8 dB; 95% CI, 0.15-11.5; $p = 0.04$).

Table 4. Long-term postoperative hearing results in partial ossicular replacement prosthesis and total ossicular replacement prosthesis groups

Variable	Partial ossicular replacement prosthesis (n = 98)	Total ossicular replacement prosthesis (n = 42)	Mean or percentage difference (95% confidence interval)	p
ABG <10 dB (%)	61.2	64.3	3.2 (-5 to 11)	0.73
ABG <20 dB (%)	73.5	85.7	12.2 (5-19)	0.13
Mean BC (dB)	28.9	29.5	-0.5 (-6.1 to 5.1)	0.86
Mean AC (dB)	40.7	38.7	1.9 (-5.3 to 9.2)	0.60
Mean ABG (dB)	11.7	9.2	2.4 (-2.5-7.4)	0.33
Mean change in BC (dB)	-3.2	-2.9	-0.3 (-2.8 to 2.3)	0.84
Mean change in AC (dB)	+9.3	+12.2	-2.9 (-9.0-3.2)	0.35
Mean BC at 4 kHz (dB)	42.2	40.9	1.2 (-7.1 to 9.6)	0.77
Mean AC at 4 kHz (dB)	56.6	49.5	7.1 (-1.6 to 15.7)	0.11
Mean ABG at 4 kHz (dB)	14.4	8.6	5.8 (0.15-11.5)	0.04
Change in BC at 4 kHz (dB)	-4.2	-3.7	8.8 (2.6-14.9)	0.02
Change in AC at 4 kHz (dB)	+0.9	+9.6	0.5 (-4.3 to 5.4)	0.82

ABG indicates air-bone gap; AC, air conduction; BC, bone conduction.

Failures

For all of the cases within this series, failures were reported as cases falling into any of the following categories: severe SNHL, defined as a worsening of BC PTA by 15 dB or more; prosthesis extrusion, postoperative ABG greater than 20 dB; and dizziness in combination with a positive fistula sign. When more than 1 cause of failure was identified, all causes were recorded, but the one which was suspected as to be the major cause was selected as the main cause of failure.

The overall failure rate in PORP group cases was 31.2% (90 cases) compared with 15.2% (43 cases) in TORP group cases. Postoperative SNHL occurred in 0.3% of cases (1 case) in the PORP group and was not observed in the TORP group. There were 86 cases of postoperative ABG greater than 20 dB in the PORP group (29.7%) compared with 37 cases in the TORP group (13%). Prosthesis extrusion occurred in 10 cases of the PORP group (3.4%). Of the 10 cases of extrusion, 8 cases occurred within the first 12 months of follow-up, and there were 2 cases of late extrusion, which were observed after 5 years after surgery. Prosthesis extrusion was seen in 12 cases of the TORP group (4.2%). Of these 12 extrusions, 6 cases occurred within the first 12 months of follow-up, 3 cases at 2 years and 3 cases after 4 years. There was 1 case of dizziness with the presence of a positive fistula sign in the PORP group (0.3%). This was not observed in the TORP group.

First Personal Revisions

PORP Group

Within the PORP group, 75 of 90 failures underwent revision surgery during the follow-up period. Causes of failure are presented in Figure 8. Prosthesis dislocation was the most common cause of failure in this group and was observed in 50 cases (66.6%), and short prosthesis was the second cause of failure (13 cases [17.3%]). Other causes of failure were prosthesis extrusion (5 cases [6.7%]), stapes fixation (3 cases [2.7%]), and malleus ankylosis (2 cases [2.7%]). One patient (1.3%) was reoperated for a TM perforation, and the position of the previous PORP was found to be correct. In 2 cases (2.7%), it was not possible to clearly identify the cause of failure at the time of revision, and the patients were therefore classified as having "negative findings" (malleus and stapes were normal, and the position of the previous prosthesis was correct with the presence of a clear round window reflex).

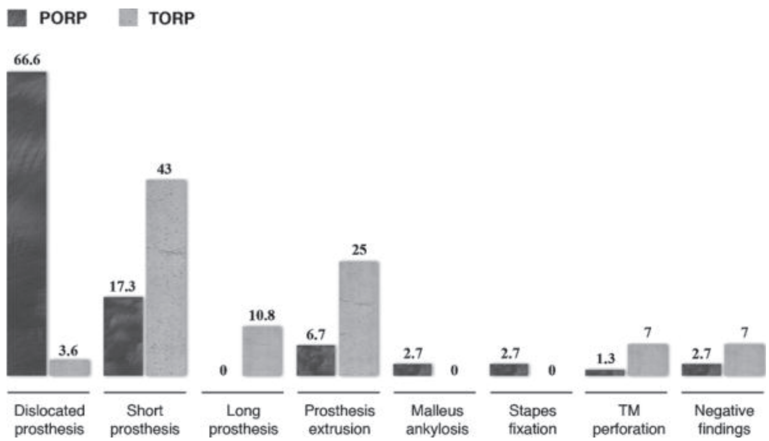

Figure 8. Cause of failure in PORP and TORP groups (%).

Further ossicular chain erosion was identified at the time of the revision. Erosion of the stapes superstructure was observed in 10 cases and malleus erosion in 2 cases.

In 27 cases, the previous PORPs were removed and replaced with a new PORP positioned from malleus to stapes head. In 15 cases, the previous PORPs were replaced with a TORP positioned from malleus to stapes footplate, and the Silastic banding technique was used. Of the 27 cases that underwent revision ossiculoplasty with PORP, postoperative data were available for 26 cases, with a mean follow-up of 48.4 months (range, 3-169.4 mo). Of the 15 cases who underwent revision ossiculoplasty with TORP, postoperative data were available for 12 cases with a mean follow-up of 24 months (range, 3-96.2 mo). Neither postoperative SNHL nor prosthesis extrusion was observed in either group. The postoperative mean ABG was 17 dB in the PORP group compared with 13 dB in the TORP group. The postoperative ABG was closed to within 20 dB in 16 cases (61.5%) in the PORP group compared with 9 cases (75%) in the TORP group.

TORP Group

Within the TORP group, 28 of 43 failures underwent revision surgery during the follow-up period. Causes of failures are presented in Figure 8. Short prosthesis was the most common cause of failure (12 case [43%]), and prosthesis extrusion was the second cause (7 cases [25%]). Long prosthesis was seen in 3 cases (10.8%), and prosthesis dislocation occurred only in 1 case (3.6%). Two patients (7%) were reoperated for a TM perforation, and the position of the previous TORP was found to be correct in both cases. In 2 cases (7%), it was not possible to clearly identify the cause of failure at the time of revision, and the patients were therefore classified as having “negative findings” (malleus and stapes were normal, and the position of the previous prosthesis was correct with the presence of a clear round window reflex).

Further ossicular chain erosion was identified at the time of the revision. Erosion of the stapes superstructure was observed in 7 cases and malleus erosion in 4 cases.

In 13 cases, the previous TORP was removed and replaced with a new TORP positioned from malleus to stapes footplate, and the Silastic banding was used. Postoperative data were available for all cases (13 cases), with a mean follow-up of 11.3 months (range, 3-22 mo). No postoperative SNHL or prosthesis extrusion was observed in this group. The postoperative mean ABG was 15 dB. The postoperative ABG was closed to within 20 dB in 10 cases (77%).

DISCUSSION

When performing ossiculoplasty, it remains the goal of every otologist to work toward restoring the hearing mechanism to achieve the best possible hearing results and satisfy patient expectations. As the search for the perfect prosthesis continues, the importance of other factors, such as surgical technique, also has been highlighted as clear variables that require consideration when aiming for optimal results. During surgery, a number of ossicular defects may be faced. Four groups were described by Austin (6) in the absence the incus; this was later modified by Kartush (7). The most common ossicular defect described the absence of the long process of the incus, with intact malleus handle and stapes, classified as Austin-Kartush group A. This group has formed the basis for the current study.

Overall hearing outcomes for the 2 groups in the present study showed significantly superior results for the TORP group for ABG closure (68.9% and 86.9% closure of ABG to <10 and <20 dB, respectively) compared with the PORP group (54.3% and 70.4% closure of ABG to <10 and <20 dB, respectively) and postoperative mean ABG (8.9 dB for TORP group and 13.1 dB for PORP group). The mean gain in AC also was significantly higher in the TORP group (16.2 dB versus 9.8 dB). We postulate that the reason for better hearing outcomes within the TORP group may be due to the favorable combination of malleus relocation, use of Silastic banding to secure the prosthesis, together with footplate placement. This amalgamation results in an increasingly stable and more functional ossicular arrangement. However, as previously stated, all the malleus relocation and banding cases were done later in the series when we had more experience as opposed to a random allocation of cases. This can be considered as a possible selection bias.

Mounting evidence in the literature confirms the presence of the malleus handle as an important prognostic factor in ossiculoplasty, leading to better hearing results and lower rates of extrusion due to its role in preventing TM lateralization (10). De Vos et al. (12) recently reiterated the absence of the malleus handle an adverse prognostic factor because its presence not only gives stability to the reconstruction but also acts as catenary lever, thereby contributing to acoustic gain. However, it is not only the presence of the malleus but also its position in relation to the stapes that affects hearing outcomes. Vlaming and Feenstra (13) demonstrated that the ideal reconstructive situation was when the malleus was positioned directly over the stapes. In practice, this anatomic configuration is very rarely seen but can be easily achieved with the malleus relocation technique as demonstrated in this study. The theory is that all of the force of vibration through a strut prosthesis would be converted into an efficient pistonlike motion at the footplate. When the malleus is malpositioned, ossiculoplasty is often unstable and mechanically inefficient, particularly if angulated more than 45 degrees from the axis of the stapes superstructure or footplate (14). Malleus relocation acts to reduce the angle to zero as the relocated malleus is positioned over the stapes head or footplate, allowing ideal placement of the prosthesis. In addition, the use of Silastic banding to secure the prosthesis at the distal end ensures that the prosthesis is centered properly on the stapes footplate, without great risk of displacement. This configuration further allows an almost perpendicular direction of forces to the footplate, with optimal transfer of function and minimal dissipation of energy (11).

Although many would advocate the use of PORP in Austin-Kartush group A patients, some authors are recognizing the benefits of TORP reconstruction. Moretz (1) commented that, although the Austin-Kartush classification describes ossicular chain defects, it does not take into account the distance and spatial arrangements between structures in the middle ear. Hence, technique selection can be varied to suit the anatomy, with several authors now favoring the use of TORP in the presence of an intact and normal stapes structure (1,3-5,15,16). Colleti and Fiorino (16) chose a TORP to perform a malleus-to-footplate reconstruction with good outcomes, resulting in an ABG closure to within 20 dB in 73.5% and 70.8% of subjects at 1 and 5 years, respectively. They argued that the superior results compared with other reconstructions, such as TM-to-footplate and the use of PORPs, were due to the stability and orientation of the reconstruction, producing optimal sound transfer.

Moretz (1) showed that the use of TORP from the footplate in patients with an intact stapes superstructure could be just as effective as the more traditional techniques of partial reconstruction, advocating their use. Conversely, other authors such as Rondini-Gilli et al. (17) found that the residual ABG was significantly lower in PORP cases compared with TORP reconstructions (16 ± 9.3 versus 23 ± 13.7 dB, $p < 0.05$). A comparison of ossicular reconstruction in Austin-Kartush group A patients from the literature is shown in Table 5.

The few articles cited in the table reflect the reluctance to use a total prosthesis in the presence of an intact stapes superstructure. A search of the literature revealed that the majority of studies comparing PORP and TORP used the more traditional approach of PORP in Austin-Kartush Group A patients, with TORP reserved for Groups B and D. These studies were therefore not included in the comparison analysis.

In our series, the overall failure rate in PORP group cases was 31.2% (90 cases) compared with 15.2% (43 cases) in TORP group cases. Postoperative SNHL occurred in

Table 5. Comparison of partial ossicular replacement prosthesis and total ossicular replacement prosthesis reconstructions in Austin-Kartush Group A patients in literature

			Partial ossicular replacement prostheses					Total ossicular replacement prostheses					
Study	**No. of cases**	**Minimum follow-up**	**Mean pre-operative ABG (dB) (standard deviation)**	**Mean post-operative ABG (dB)**	**Mean post-operative ABG < 10 dB (% cases)**	**Mean post-operative ABG < 20 dB (% cases)**	**Mean post-operative ABG < 30 dB (% cases)**	**Mean pre-operative ABG (dB)**	**Mean post-operative ABG (dB)**	**Mean post-operative ABG < 10 dB (% cases)**	**Mean post-operative ABG < 20 dB (% cases)**	**Mean post-operative ABG < 30 dB (% cases)**	**Comments**
Present study	571; partial, 2=288; total n=283	12	24.6	11.3	60.8	78.3	--	24.8	7.2	73	91.6	--	
Moretz (1)	70; partial; n = 41; total, n = 29	12	25 (11)	16	27	81	96	24 (10)	17	14	79	16	No significant difference found between groups
Colleti and Fiorino (16)	779; partial, n = 588; total, n = 191	12	30 (8)	15	--	--	--	28 (8)	9	38.3	73.6	90.7	Total ossicular replacement prostheses showed significantly better results at both 1 and 5 yr ($p < 0.01$) compared with partial ossicular replacement prostheses
Brackmann et al. (2)	1,042; partial, n = 438; total, n = 604a	6	--	--	50	73	85	--	--	27	55	74	Higher success rates in partial ossicular replacement prostheses but no statistical analysis.
Gersdorff et al. (18)	165; partial, n = 96; total, n = 69	9	33	16.7	31.3	64.6	88.5	36.7	22.7	24.6	52.2	79.7	

aIncludes total ossicular replacement prosthesis to oval window after stapedectomy (n = 125). (The authors did not give breakdown of results; therefore, these patients could not be excluded from the results analysis.)

0.3% (1 case) in the PORP group and was not observed in the TORP group. Prosthesis extrusion occurred in 10 cases of the PORP group (3.4%) and was seen in 12 cases of the TORP group (4.2%). In his series, Moretz (1) found no cases of SNHL or prosthesis extrusion. Overall failure rates secondary to disease state or prosthetic causes were quoted as 15% (115) in the PORP group and 9% (73) in the TORP group.

The causes of failure identified during revision surgery for the "personal" cases in which the original surgery had been performed by the senior author were different after the use of PORP and TORP. In the PORP group, the main cause of failure was prosthesis dislocation/displacement. This was observed in 66.6% compared to 3.6% in the TORP group. This observation supports the idea that the Silastic banding technique leads to enhanced stability with a TORP as compared with conventional PORP positioning onto the stapes head. In the TORP group, the main cause of failure was a short prosthesis, observed in 43% of cases. This is related to the malleus relocation technique. After dissecting the malleus free from the TM and cutting the tensor tympani and or stretching or cutting the anterior tympanomallear ligament, the malleus is left attached by the superior tympanomallear ligament alone. It has a tendency toward slight medialization. This artificially decreases the distance from the stapes footplate, and as a result, the prosthesis may be trimmed to short. This problem has been identified by the authors as part of their learning curve for this technique. As a result, the technique has been modified, and the malleus is now placed in contact with the TM in a horizontal plane before measuring the distance to the stapes footplate, giving better results.

The hearing results for the TORP group were better on all measures when compared with the PORP group. Only in the long-term analysis, this did not reach statistical significance. It is unclear to us why a significant difference was not demonstrated for long-term hearing outcomes because this does not correlate with our clinical observation. Because long-term data were available for only a small segment of each group, (31.6% of PORP group and 13% of the TORP group), it is possible that this may not be truly representative. Future reports will address this as more data are obtained.

CONCLUSION

Our study demonstrates that, in Austin-Kartush Group A, the use of TORP with malleus relocation and Silastic banding gives superior results over the more traditional partial prosthesis with or without malleus relocation. The addition of malleus relocation and Silastic banding seems to provide a more favorable anatomic vertical configuration for effective transfer of vibrations to the footplate and better hearing outcomes.

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Chapter 9

Ossiculoplasty in missing malleus and stapes patients: experimental and preliminary clinical results with a new malleus replacement prosthesis with the otology-neurotology database

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ABSTRACT

Objective: To present the preliminary results of new malleus replacement prosthesis combined with a total ossicular prosthesis in middle ear reconstruction in patients missing the malleus and stapes.

Study Design: Prospective experimental and nonrandomized clinical study.

Setting: Tertiary referral center.

Methods: An original titanium malleus replacement prosthesis (MRP) was designed to be inserted into the external auditory canal and to replace a missing malleus for various middle ear pathologies. The MRP was tested experimentally and clinically. The vibratory properties of the new prosthesis were measured using laser Doppler vibrometry. Ninety patients with missing malleus and stapes, undergoing 92 ossicular reconstructions were enrolled in this study from September 1994 to March 2012. Comparative analyses were made between a group of 34 cases of ossicular reconstructions with total prosthesis (TORP) positioned from the tympanic membrane to the stapes footplate (TM-to-footplate assembly) and a group of 58 cases of ossicular reconstructions with TORP positioned from a newly designed malleus replacement prosthesis (MRP) to the stapes footplate (MRP-to-footplate assembly). Preoperative and postoperative audiometric evaluation using conventional audiometry, that is, air-bone gap (ABG), bone-conduction thresholds (BC), and air-conduction thresholds (AC) were assessed.

Results: Experimentally, the vibratory properties of the MRP are promising and remain very good even when the MRP is cemented into the bony canal wall mimicking its complete osseous-integration, if this were to occur. This finding supports the short-term clinical results as in the TM-to-footplate group; the 3-month postoperative mean ABG was 23.3 dB compared with 12.5 dB in the MRP-to-footplate group (difference, 10.8; 95% confidence interval, 4.0–17.6); 37.0% of patients from the TM-to-footplate group had a postoperative ABG of 10 dB or less, and 48.1% of patients had a postoperative ABG of 20 dB or less, as compared with 58.1% and 79.1%, respectively, in the MRP-to-footplate group. The average gain in AC was 11.0 dB in the TM-to-footplate group as compared with 21.3 dB in the MRP-to-footplate group (difference, –10.3; 95% confidence interval, –18.2 to –2.4).

Conclusion: The results of this study indicate that superior postoperative hearing thresholds could be achieved using a MRP-to-footplate assembly, compared with a

TM-to-footplate assembly in patients with an absent malleus undergoing ossiculoplasty. The postoperative AC thresholds, after 3 months and 1 year, are significantly lower in patients treated with the MRP-to-footplate assembly.

The presence or absence of a malleus handle affects the hearing results in ossiculoplasty, particularly in the absence of a stapes superstructure. Numerous authors have emphasized the importance of the malleus in successful ossiculoplasty (1–4). Recreation of the malleus has been used by several authors to enhance middle ear prosthesis stability (5–7).

The present study evaluated the effectiveness of a newly designed titanium malleus replacement prosthesis (MRP), which was created to replace a missing malleus as an attempt to reduce the risk of prosthesis dislocation. The MRP is designed to be affixed to the bony ear canal to provide superior stabilization of a total or partial prosthesis coupled to it. During the period of January 1991 to March 2012, 1952 consecutive ossiculoplasty procedures were performed by the first author in the same tertiary referral center. Of these, 92 cases (4.7%) were performed with an absent malleus and stapes. Of these 92 cases, 58 were operated with a MRP implantation from January 2010 to March 2012. A total ossicular replacement prosthesis (TORP) was positioned from the MRP to the stapes footplate (MRP-to-footplate assembly) in all 58 cases. In 34 cases, operated from September 1994 to April 2009, a MRP was not used, and a TORP was positioned from the tympanic membrane to the stapes footplate (TM-to-footplate assembly).

The aim of this prospective study is to compare the hearing results in patients undergoing total ossicular reconstruction using either a TM-to-footplate assembly or a MRP-to-footplate assembly.

MATERIALS AND METHODS

The Malleus Replacement Prosthesis

The MRP (Figure 1) was designed in 2009 together with the Kurz company (Heinz Kurz Inc, Dusslingen, Germany). It is a titanium neo-malleus, which is implanted underneath the tympanic membrane at any position in the bony rim of the external auditory canal. The 0.8-mm-diameter handle is attached via a Y-shaped titanium wire with 2 posts (0.3-mm diameter), which are intended to fix to the bony canal wall of the EAC. These posts are inserted into 2 holes, which are drilled with a 0.6-mm burr. The surgeon introduces the MRP and can connect almost any partial or total replacement prosthesis because of the malleable MRP.

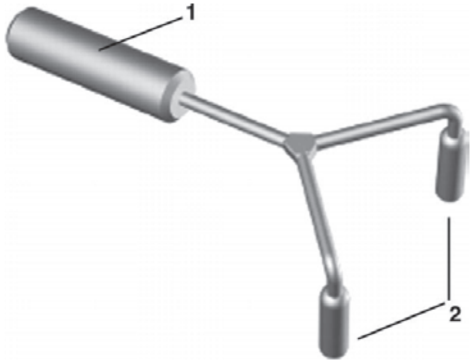

Figure 1. MRP: 1 = 0.8-mm-diameter handle; 2 = 0.3-mm-diameter posts, which will be inserted in the EAC.

Experimental Study

We tested the vibratory properties of the MRP experimentally using laser Doppler vibrometry, with and without TORP insertion and with and without MRP and tympanic membrane fixation. The MRP was inserted in human temporal bones, which were donated for scientific research. Fibrin glue was used to fix the eardrum to the external auditory canal in such way that would be stretched over the MRP handle. Because of the possibility that titanium might osseointegrate in living tissue, and this cannot be reproduced in cadaveric bone, dental cement (Carboxylat cement, *Durelon*; ESPE Dental AG, Seefeld) was used to fix the MRP into the bone. Stimulation parameters were 94 dB in the outer ear canal at 0.5, 1, 2, and 4 kHz.

The measurements were performed using laser Doppler vibrometer (LDV) (Polytec, OFV 551, Germany) focused onto the center of the handle of the MRP. Figure 2, A and B, represents a view through the microscope, showing the MRP coupled to a Flex/HA TORP (Medtronic-Xomed Inc, Jacksonville, FL, USA). All measurements were made in 4 different conditions: 1) Nonnon (tympanic membrane and MRP without being fixed in place); 2) Fixnon (MRP cemented into the bone, ear drum not fixed); 3) Fixfix (eardrum and MRP both fixed in place without TORP insertion); 4) TORP (TORP inserted between the stapes footplate and the handle MRP, eardrum, and MRP are fixed in place).

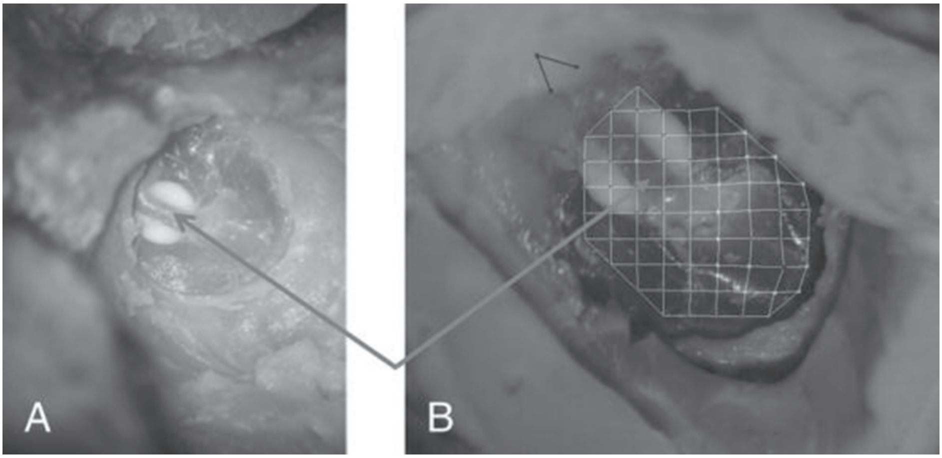

Figure 2. View of the tympanic membrane as seen through the microscope (A) and through the LDV videocamera (B) of a right temporal bone, showing the MRP, which is coupled to a Flex/HA TORP. 1 = TORP's head. 2 = Handle of the MRP. 3 = Tympanic membrane. The arrows point to the LDV measuring position.

Patients

This is a prospective study of 90 patients with absent malleus and stapes who underwent 92 ossicular reconstructions with a TORP implantation. All patients were operated by the same surgeon from September 1994 to March 2012.

All cases were revision tympanoplasty cases for chronic suppurative otitis media (CSOM) without active cholesteatoma. Austin's classification of ossicular defects as modified by Kartush was used to define the ossicular status encountered (8,9). In this analysis, only those patients allocated to Austin–Kartush Group D (stapes and malleus absent) were included. Patients were divided in 2 groups according to the type of prosthesis' assembly. Thirty-four consecutive cases of TORP repair occurring from September 1994 to April 2009 were compared with 58 consecutive cases using MRP occurring from January 2010 to March 2012. In TORP-only cases, the prosthesis was interposed between the tympanic membrane and the stapes footplate (TM-to-footplate assembly). In all MRP cases, the MRP was inserted followed by a TORP that was positioned from the MRP to the stapes footplate (MRP-to-footplate assembly). This was completed at the same operation or in a staged fashion.

All data were prospectively tabulated using the Otology-Neurotology Database (ONDB) (AS Multimedia Inc., Cassagne, France) (10). This is a commercially available software package developed at our center, designed to comply with the American Academy of Otolaryngology guidelines for reporting clinical and audiometric results (11).

Surgery

All procedures in both groups were performed by the same surgeon (first author) via a permeatal approach. In all cases, the ossiculoplasty was performed with the same Flex/HA TORP as the one which was used for the experimental study. The prosthesis' shaft was cut to the appropriate length, and the distal end of the prosthesis shaft was positioned to the stapes footplate. The choice of prosthesis' head placement was dictated by the progressive experience and surgical technique of the surgeon and subsequently the introduction of a new original malleus replacement prosthesis (MRP). The MRP was designed in 2009 to recreate a new malleus handle to enhance prosthesis' stability. The prosthesis head was universally placed in direct contact with the tympanic membrane before the introduction of the MRP. Thus, all encountered cases after January 2010 were done with the MRP.

In the TM-to-footplate group, the TORP's head was positioned in direct contact to the tympanic membrane in all cases (34 cases). These patients were enrolled in the study between September 1994 and April 2009. In the MRP-to-footplate group, a MRP was inserted, and the TORP's head was positioned underneath the handle of the MRP (58 cases). These patients were enrolled in the study from January 2000 to March 2012. Of these 58 cases, 49 cases (84.4%) were operated with a simultaneous implantation of a MRP and a TORP during the same operation (1-stage procedure). The remaining 9 cases (15.6%) were implanted with a MRP only (first-stage procedure), and the TORP was positioned during the second stage, which was performed 6 months later. The decision to stage the procedure was dictated by the status of the middle ear mucosa at the time of surgery.

Surgical Technique for MRP Implantation

Two tunnels were drilled through the external auditory canal wall (EAC) (from side to side) with a 0.6-mm diamond dust burr for the 2 posts of the MRP (Figure 3, A and B). The distance between the 2 EAC tunnels was equal to the distance between the 2 posts of the MRP. The drilling of the 2 tunnels required highly regulated speed with constant irrigation to prevent burning or pressure necrosis of the bone. The choice of the position of these 2 tunnels was dictated by the status of the EAC. In 52 cases (89.6%), they were drilled approximately at 11 and 9 o'clock for a right ear and 1 and 3 o'clock for a left ear; where 12 o'clock is positioned at the notch of Rivinus. Thus, the position of the titanium neo-handle of the MRP was slightly more posterior to a normal malleus handle. In 6 cases (10%), there was not enough residual posterior-superior bony canal wall to allow for drilling out the tunnels, which were then drilled at approximately 5 and 6 o'clock. In these cases, the position of the titanium neo-handle was exactly opposite but parallel to the position of a normal malleus handle.

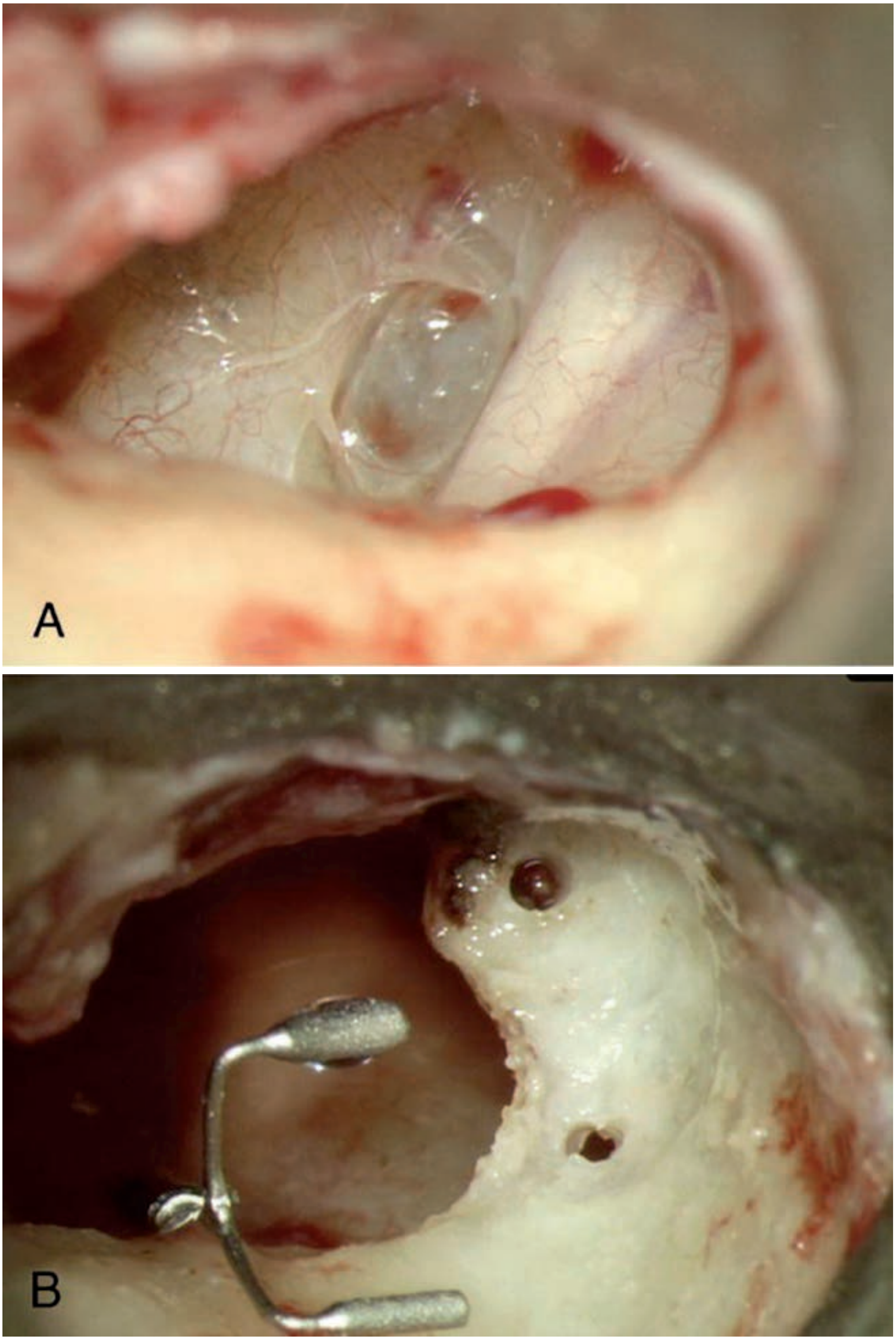

Figure 3 . A, Intraoperative view of a right ear showing the stapes footplate, which is mobile with a missing malleus and incus. B, Two tunnels were drilled through the EAC from side to side with a 0.6-mm diamond dust burr to leave room for the 2 posts of the MRP. The tunnels were drilled at 9 and 11 o'clock

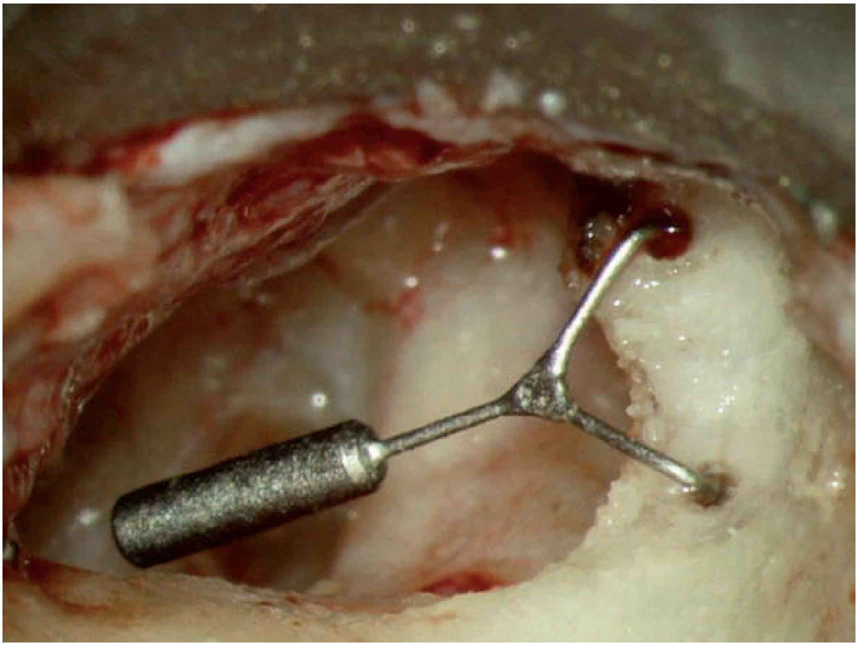

Figure 4. The 2 posts of the MRP were inserted in the 2 tunnels, and the titanium handle was positioned over the stapes footplate.

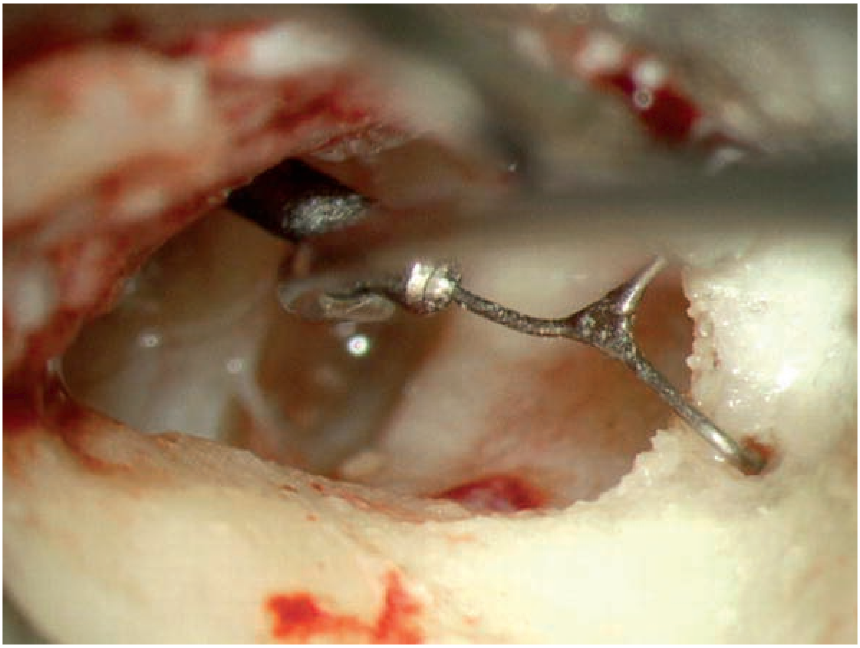

Figure 5. While the MRP was kept in position by the 2 posts, the thin titanium link between the handle and the posts enabled the surgeon to easily accommodate the position of the neo-handle to all anatomic conditions. The handle of the MRP was kept in proper position overlying the stapes footplate.

The 2 posts of the MRP were inserted in the 2 tunnels, and the titanium handle was positioned over the stapes footplate (Figure 4). This configuration prevents MRP displacement and avoids contact between the neo-handle and the canal wall. Although the MRP was kept in position by the 2 posts, the malleable titanium link between the handle and the posts enabled the surgeon to easily accommodate the position of the neo-handle to all anatomic conditions (Figure 5). It was possible to slightly move the handle laterally, inferiorly, or superiorly. The foremost advantage of this prosthesis is to keep the neo-handle in proper position overlying the stapes footplate in all cases.

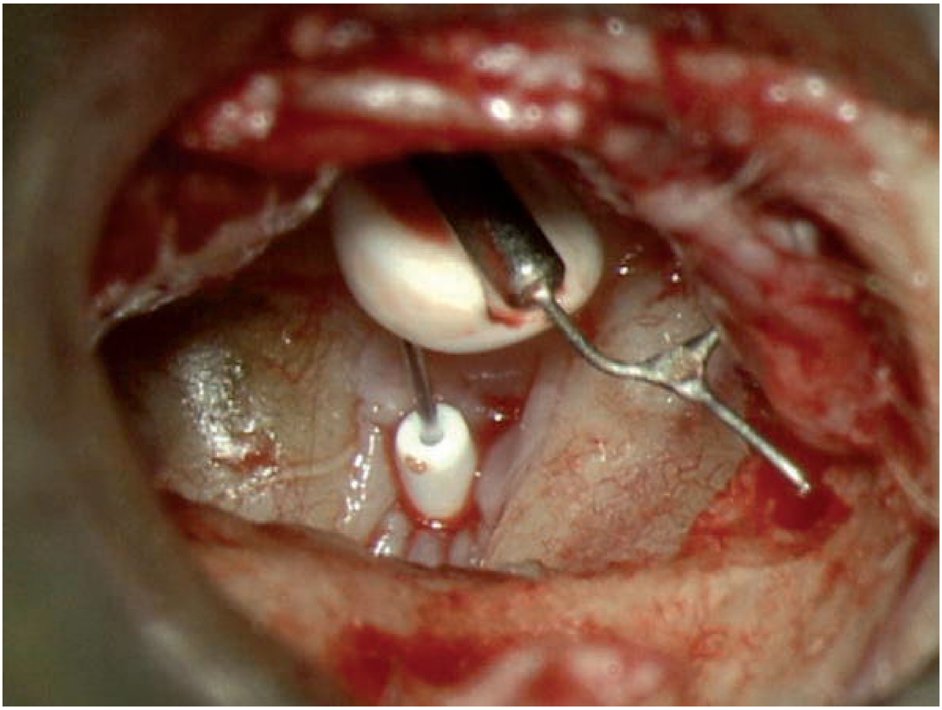

Figure 6. Final view showing the MRP's handle, which was introduced within the groove of the TORP's head.

The distance between the neo-malleus of the MRP and the stapes footplate was determined with an elongated stapes measuring rod. The TORP shaft was cut at the appropriate length, and the TORP was positioned from the handle of the MRP to the stapes footplate. The distal tip of the TORP's shaft is centered on the stapes footplate, and the MRP's neo-handle is easily introduced within the groove of the TORP's head (Figure 6). A thin layer of tragal cartilage was interposed over the MRP in all cases covering both the 2 posts and the neo-handle. When TM grafting was performed in combination with the MRP implantation, all fascia or perichondrium grafts were placed over the cartilage plate (lateral graft).

Audiometric Assessment

Audiometric evaluation included preoperative and postoperative air-bone gap (ABG), air-conduction (AC) thresholds, and bone-conduction (BC) thresholds. Only AC and BC results that were obtained at the same time postoperatively were used for calculation of ABG and pure-tone averages (PTAs). We used a 4-frequency PTA for AC and BC thresholds (0.5, 1, 2, and 4 kHz) obtained at 3-month and 1-year follow-up. The preoperative and postoperative BC and AC levels at 4 kHz were also assessed. Audiometry was reported according to American Academy of Otolaryngology–Head and Neck Surgery Guidelines (7) except for thresholds at 3 kHz, which were substituted in all cases with those at 4 kHz.

Statistical Analyses

To evaluate the effect of an ossicular reconstruction using either a TM-to-footplate assembly or a MRP-to-footplate assembly, preoperative and postoperative audiometric results were compared, using an independent samples *T* test. Mean differences and percentage differences and their 95% confidence intervals (CIs) were calculated. All analyses were performed using SPSS (version 15.0; SPSS Inc., Chicago IL, USA).

RESULTS

Experimental Results

Using the above-mentioned setting (Figure 2), the vibratory properties of the MRP were analyzed in 4 different fixation conditions (Figure 7, A and B). Our preliminary data demonstrated that the MRP does vibrate when stimulated from 0.5 to 4 kHz,

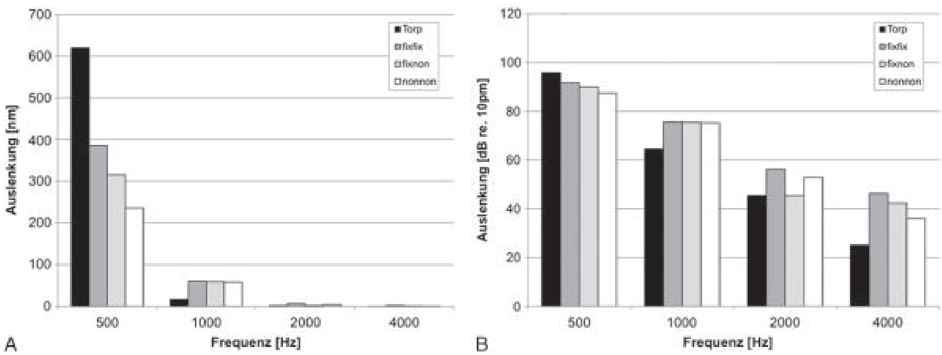

Figure 7. A and B, LDV analysis results plotted on a linear scale (A) and on a logarithmic scale (B) related to 10 pm demonstrating vibrations of the MRP in all 4 tested conditions 1. Nonnon = ear drum and malleus prostheses without being fixed in place; 2. Fixnon = malleus-prostheses cemented into the bone, ear drum not fixed; 3. Fixfix = ear drum and malleus prostheses fixed in place, no TORP; 4. Torp = TORP between the stapes footplate and the malleus-prostheses, ear drum, and malleus prostheses fixed in place.

having the motion amplitude decreasing with increasing frequency of the applied sound at the same sound pressure level (94 dB).

The vibrations are transmitted through the direct contact of the tympanic membrane to the MRP and are not diminished when the MRP posts have been fixed in place. This supports the expected long-term functionality of the MRP, even if ossification occurs around the posts. Additionally, the tympanic membrane vibrations were transmitted to the MRP by contact alone without the use of adhesive substances. This suggests that fibrous fixation of the MRP handle to the tympanic membrane is not essential for transmission. However, further studies are underway to optimize the mechanical force transfer between the tympanic membrane and the MRP.

Patient Characteristics

Of the 34 patients in the TM-to-footplate group, 27 had audiologic data available at 3-month follow-up. Of the 58 cases in the MRP-to-footplate group, 43 had audiologic

Table 1. Preoperative patient characteristics

Variable	TM-to-footplate TORP placement (n = 27)	MRP-to-footplate TORP placement (n = 43)	Differences (95% CI)	p
Age, yr (SD)	51.6 (15.3)	52.8 (14.7)	-1.25 (-8.6 to 6.1)	0.733
Sex (%female)	55.6	72.1	-16.5 (-39.6 to 6.5)	0.159
Mean BC (dB)	30.8 (14.8)	25.9 (12.5)	5.0 (-1.6 to 11.5)	0.137
Mean AC (dB)	65.4 (16.0)	59.0 (13.9)	6.5 (-0.7 to 13.7)	0.078
Mean ABG (dB)	34.6 (10.4)	33.1 (9.5)	1.5 (-3.3 to 6.3)	0.535
BC at 0.5 kHz (dB) 1	20.9 (10.7)	14.8 (8.4)	6.2 (1.6 to 10.7)	0.009
BC at 1 kHz (dB)	24.3 (13.3)	19.2 (11.5)	5.1 (-0.9 to 11.1)	0.096
BC at 2 kHz (dB)	35.6 (19.2)	30.6 (18.5)	5.0 (-4.2 to 14.2)	0.284
BC at 4 kHz (dB)	42.6 (23.5)	39.0 (18.7)	3.6 (-6.5 to 13.8)	0.477
AC at 0.5 kHz (dB)	59.1 (17.2)	52.6 (18.7)	6.5 (-2.4 to 15.4)	0.148
AC at 1 kHz (dB)	63.0 (17.4)	57.6 (16.8)	5.4 (-3.0 to 13.8)	0.201
AC at 2 kHz (dB)	66.1 (19.5)	57.9 (15.8)	8.2 (-0.3 to16.7)	0.058
AC at 4 kHz (dB)	73.5 (17.6)	67.8 (16.2)	5.7 (-2.5 to 14.0)	0.169
ABG at 0.5 kHz (dB)	38.2 (10.6)	37.8 (16.0)	0.4 (-6.6 to 7.3)	0.919
ABG at 1 kHz (dB)	38.7 (13.1)	38.4 (14.0)	0.3 (-6.4 to 7.0)	0.921
ABG at 2 kHz (dB)	30.6 (14.2)	27.3 (10.4)	3.2 (-2.6 to 9.1)	0.277
ABG at 4 kHz (dB)	30.9 (16.5)	28.8 (11.6)	2.1 (-4.6 to 8.8)	0.536

Means were calculated with 0.5, 1, 2, and 4 kHz.
ABG indicates air-bone gap; AC, air conduction; BC, bone conduction; MRP, malleus replacement prosthesis; SD, standard deviation; TORP, total ossicular replacement prosthesis.

data available at 3-month follow-up. Because of the fact that many of our patients travel long distances for care at our center, onsite follow-up care is sometimes not possible. Table 1 shows the preoperative patient characteristics for the 2 groups. In the TM-to-footplate and MRP-to-footplate groups, 55.6% and 72.1% were female subjects, and the mean age was 51.6 and 52.8 years, respectively. There were 4 senior patients (age, >=65 yr) in the TM-to-footplate group and 8 in the MRP-to-footplate group. There were 2 children (age, <=18 yr) in the TM-to-footplate group, and there were no children in the MRP-to-footplate group. The mean preoperative BC threshold was 30.8 dB in the TM-to-footplate group and 25.9 dB in the MRP-to-footplate group. The mean preoperative AC threshold was 65.4 dB in the TM-to-footplate group and 59.0 dB in the MRP-to-footplate group. The preoperative ABG was 34.6 dB in the TM-to-footplate group and 33.1 dB in the MRP-to-footplate group. The only statistically significant difference noted was the BC at 0.5 Hz.

Postoperative Audiometric Assessment at 3-Month Follow-Up

None of the patients had postoperative sensorineural hearing loss (SNHL), which was defined as change in the BC PTA of 15 dB or more. Postoperative hearing results are shown in Table 2. The postoperative ABG was closed to 10 dB or less in 37.0% of the cases and closed to 20 dB or less in 48.1% of the cases in the TM-to-footplate group compared with 58.1% and 79.1%, respectively in the MRP-to-footplate group (difference, -21.1; 95% CI, -44.5 to 2.3; and difference, -30.9; 95% CI, -53.4 to -8.5). The postoperative gain in AC threshold was 11.0 dB in the TM-to-footplate group compared with 21.3 dB in the MRP-to-footplate group (difference, -10.3; 95% CI, -18.2 to -2.4). The postoperative change in BC threshold was negligible in both groups (-0.3 dB in the TM-to-footplate group and 0.7 dB in the MRP-to-footplate

Table 2. Hearing results at 3-month follow-up according to the total ossicular replacement prosthesis placement

Variable	TM-to-footplate TORP placement (n = 27)	MRP-to-footplate TORP placement (n = 43)	Mean or percentage difference (95% CI)	p
ABG, ≤10 dB (%)	37.0	58.1	-21.1 (-44.5 to 2.3)	0.088
ABG, ≤20 dB (%)	48.1	79.1	-30.9 (-53.4 to -8.5)	0.008
Mean BC, dB (SD)	31.1 (16.7)	25.2 (13.1)	6.0 (-1.2 to 13.1)	0.101
Mean AC, dB (SD)	54.4 (22.7)	37.7 (18.3)	16.8 (6.9 to 26.6)	0.001
Mean ABG, dB (SD)	23.3 (16.7)	12.5 (11.9)	10.8 (4.0 to 17.6)	0.002
Mean change in BC, dB (SD)	-0.3 (7.0)	+0.7 (6.2)	-1.0 (-4.2 to 2.2)	0.533
Mean change in AC, dB (SD)	+11.0 (15.5)	+21.3 (16.4)	-10.3 (-18.2 to -2.4)	0.011

Means were calculated with 0.5, 1, 2, and 4 kHz.

group). In the TM-to-footplate group, the postoperative 4-frequency average ABG was 23.3 dB compared with 12.5 dB in the MRP-to-footplate group (difference, 10.8; 95% CI, 4.0–17.6).

Postoperative Audiometric Assessment at 1-Year Follow-Up

Postoperative hearing results are shown in Table 3. The postoperative ABG was closed to 10 dB or less in 15.4% of the cases and to 20 dB or less in 53.8% of the cases in the TM-to-footplate group compared with 52.6% and 84.2%, respectively, in the MRP-to-footplate group (difference, -37.3; 95% CI, -67.1 to -7.4; and difference, -30.4; 95% CI, -62.0 to 1.3).

Table 3. Postoperative hearing results at 1 year according to the total ossicular replacement prosthesis placement

Variable	TM-to-footplate TORP placement (n = 13)	MRP-to-footplate TORP placement (n = 19)	Mean or percentage difference (95% CI)	p
ABG, ≤10 dB (%)	15.4	52.6	-37.3 (-67.1 to -7.4)	0.035
ABG, ≤20 dB (%)	53.8	84.2	-30.4 (-62.0 to 1.3)	0.065
Mean BC, dB (SD)	37.9 (17.5)	26.8 (14.6)	11.1 (-0.5 to 22.8)	0.061
Mean AC, dB (SD)	60.4 (20.1)	39.7 (17.1)	20.7 (7.2 to 34.2)	0.004
Mean ABG, dB (SD)	22.5 (13.9)	12.9 (12.6)	9.6 (-0.1 to 19.3)	0.052
Mean change in BC, dB (SD)	-0.5 (5.2)	-1.5 (5.7)	1.0 (-3.0 to 5.1)	0.606
Mean change in AC, dB (SD)	+9.2 (14.6)	+19.9 (14.9)	-10.6 (-21.5 to 0.2)	0.055

Means were calculated with 0.5, 1, 2, and 4 kHz.

The postoperative gain in AC threshold was 9.2 dB in the TM-to-footplate group compared with 19.9 dB in the MRP-to-footplate group (difference, -10.6; 95% CI, -21.5 to 0.2). The postoperative change in BC threshold was negligible in both groups (-0.5 dB in the TM-to-footplate group and -1.5 dB in the MRP-to-footplate group). In the TM-to-footplate group, the postoperative 4-frequency average ABG was 22.5 dB compared with 12.9 dB in the MRP-to-footplate group (difference, 9.6; 95% CI, -0.1 to 19.3).

Failures

For all of the cases within both groups, failures were reported as cases falling into any of the following categoris: severe SNHL, defined as as worsening of BC PTA by 15 dB or more; prosthesis extrusion; postoperative ABG greater than 20 dB; and dizziness in combination with a positive fistula sign. When more than one cause of failure was identified, all causes were recorded, but the one which was suspected as the major cause was selected as the main cause of failure.

The overall failure rate in the TM-to-footplate group cases was 51.9% (14 of 27 cases) compared with 23.2% (10 of 43 cases) in the MRP-to-footplate group cases. Postoperative SNHL or dizziness was not observed in either group. There were 14 cases of postoperative ABG greater than 20 dB in the TM-to-footplate group (51.9%), compared with 9 cases in the MRP-to-footplate group (20.9%). Postoperative extrusion occurred in 2 cases in both groups (7.4% in the TM-to-footplate group compared with 4.6% in the and MRP-to-footplate group).

First Personal Revisions—Operative Findings

TM-to-Footplate Group

Within the TM-to-footplate group, 7 of 14 failures underwent revision surgery during the follow-up period. Prosthesis dislocation was the most common cause of failure in this group and was present in 5 cases (71.4% of revision surgeries). Prosthesis extrusion was the second ranked cause of failure (2 cases, 28.6%). In all cases, a MRP was inserted, and a TORP was positioned underneath the MRP (MRP-to-footplate assembly).

MRP-to-Footplate Group

Within the MRP-to-footplate group, 3 of 9 failures underwent revision surgery during the follow-up period. The causes of failure were related to the TORP and not to the MRP. Too long and too short prosthesis (TORP) were observed in 2 cases (66.6% of the 3 cases), and in 1 case (33.4% of the 3 cases), it was not possible to clearly identify the cause of failure at the time of revision (the MRP was stable and mobile, and the position of the previous TORP was correct with the presence of a clear round window reflex), and the patient was therefore classified as having " negative findings." No dislocation or ankylosis of the MRP was observed. In 2 cases, a new TORP was positioned underneath the MRP, and in the "negative finding" case, the previous TORP was left in place, and no further procedure was attempted.

DISCUSSION

The MRP was developed to improve the postoperative hearing results in patients with an absent malleus, undergoing ossiculoplasty that had been previously managed with columellae or neo-malleus technique. Recreation of the malleus has been used in the past (5–7). Wehrs (6) suggested the use of a homologous drum and malleus, which usually requires staging for a stable reconstruction, in patients without an intact malleus. Black (7) introduced a technique of neo-malleus ossiculoplasty by using an autograft neo-malleus strut and an assembly rather than columella in cases in which the malleus was unavailable for assembly techniques. More recently, we described an original technique of silastic banding in patients with missing malleus and intact stapes superstructure (Austin-Kartush group C) (12). The ossiculoplasty was performed with a TORP, positioned from the stapes footplate to the under-surface of the tympanic membrane, using a silastic band to stabilize the prosthesis.

With the design of the MRP, a new malleus handle is created and can be used with any type of PORP or TORP, which can be positioned under the neo-handle in a single or 2-stage procedure. This technique decreases the risk of displacement of the PORP or TORP. The causes of failure identified during revision surgery for the "personal" cases in which the original surgery had been performed by the senior author were different between the 2 groups. In the TM-to-footplate group, the main cause of failure was prosthesis dislocation. This was observed in 71.4% and was not observed in the MRP-to-footplate group. This observation supports the idea that the MRP leads to enhanced stability with a TORP when the malleus is missing as compared with conventional TORP positioning from TM to footplate. However, as previously stated, all MRP-to-footplates cases were done later in the series when we had more experience as opposed to a random allocation of cases. This can be considered as a possible selection bias as the results may be explained by a learning curve. One way to explore this possibility was to show ABG closure over time in the TM-to-footplate group (27 cases). We separated the results into 3 periods of 5 years' duration: first period (September 1994 to August 1999), second period (September 1999 to August 2004), and third period (September 2004 to August 2009). This gives roughly equally spaced 5-year intervals. Figure 8 shows no evidence of a learning curve as the results of the first period were better than those of the last period.

The experimental and preliminary clinical results of the present study demonstrated the general usability and safety of the MRP, which was associated with higher success rate than when the MRP was not used. At 3-month follow-up, the postoperative mean AC and ABG were significantly smaller, and the mean postoperative gain in AC was significantly higher in patients treated with the MRP-to-footplate technique. At 1-year follow-up, the mean AC remained significantly lower in the MRP-to-footplate group.

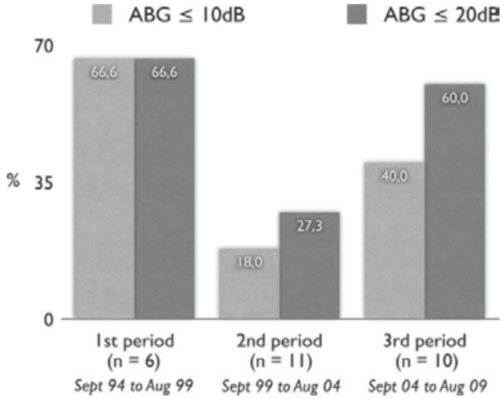

Figure 8. M-to-footplate assembly (27 cases). Postoperative ABG closure over 5-year periods.

Table 4. Hearing results in case of missing malleus in literature review.

Series	Austin-Kartush group	Material	n	Postoperative ABG G10 dB (%)	Postoperative mean ABG
Moretz (1)	C	PORP	6	0	24
	C	TORP	4	0	24
Vincent (10)	C	TORP	96	66	12
Austin (13)	C	Partial autograft	23	12	27
Black (14)	C	PORP	13	15	--
Goldenberg (15)	C	PORP	7	14	23
Current series at 3 months	D	TORP+MRP	43	58.1	12.5
Current series at 1 year	D	TORP+MRP	19	52.6	12.9

Table 4 shows the postoperative hearing results in comparable articles presenting ossiculoplasty cases with a missing malleus. This table shows that the results of the present study compare favorably with the results reported by other authors.

There were 34 cases of conventional TORPs positioning (TM-to-footplate) between 1994 and 2009. In contrast, there were 58 cases of MRP-to-footplate TORPs positioning between 2010 and present. There has been a dramatic increase in the TORP volume recently. Although this could also be considered as due to a possible selection bias, in fact, this was not the case. With the earlier group, the author had become increasingly disillusioned with the results of reconstruction in cases of missing malleus and stapes. Indeed, it was as a result of this disillusionment that he developed the malleus replacement prosthesis. Once this prosthesis was available, it became inevitable that there would be an increased usage when the preliminary results were very promising. Moreover, 7 of the 58 cases of the MRP-to-footplate group were revision surgeries after previous failed TM-to-footplate assembly.

Although these results seem to show an improvement in the hearing results with the MRP, which is included in the reconstruction, they are based on a restricted number of patients and a relatively short follow-up period, and there is the risk of deterioration of results with time. Possible long-term risks include extrusion of the MRP, which must be covered by a large cartilage plate. There were 2 cases of extrusion of the MRP in the MRP-to-footplate group. In both cases, the MRP seemed to be only partially covered by the cartilage plate, which was too small. This may increase the risk of extrusion. Originally, the cartilage was trimmed to a “T-shape” to cover only the 2 posts and the neo handle. This problem has been identified by the authors as part of their learning curve for this technique. As a result, the technique has been modified, and a larger cartilage plate is now interposed between the tympanic membrane and the MRP. Other risks include osseointegration and fixation of the MRP, with decline in hearing results. Although our laser Doppler results do not show this, it remains a possibility. Were this to happen, the MRP has been designed so that the titanium link can be cut at a second stage, freeing the neomalleus handle from any connection with the surrounding bone. Of course, atelectasis and scarification of the TORP could occur, but this is not a specific risk for the MRP.

According to McCulloch et al. (12), discussing the different stages among surgical innovation, the present study covered the “exploration phase,” which means prospective database research focusing on the development of the new surgical technique. Considering the safety and the superior postoperative hearing results of the MRP-to-footplate technique in the present study, the next phase should be a randomized trial to assess the effectiveness against the current standards.

CONCLUSION

The results of this study indicate that superior postoperative hearing thresholds can be achieved using a MRP-to-footplate assembly, compared with a TM-to-footplate assembly in patients with an absent malleus undergoing ossiculoplasty. The postoperative AC thresholds, after 3 months and 1 year, are significantly lower in patients treated with the MRP-to-footplate assembly.

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DISCUSSION

This PhD thesis contains eight original publications focussed on elective reconstructive ear surgery. The first five papers report on the outcomes of *primary* stapes replacing surgery. Three of those concern congenital minor ear anomalies. The fourth is on Osteogenesis Imperfecta and the fifth one is on the outcome of otosclerosis surgery during adolescence. The sixth paper discusses the outcome of a prospective series of 652 *revision* stapedotomies. Two factors make these papers special, viz. the systematic use of the Causse stapedotomy technique with vein graft interposition, and the relatively large size of these series of patients with a rare condition such as Osteogenesis Imperfecta and minor congenital ear anomalies.

The Causse Clinic, situated near Béziers in France, is recognized around the world as a referral centre for the specialised ear surgery discussed in this PhD thesis. This might cause a selection bias. However, if it does, it would rather be a negative selection bias, given the complexity of the cases that are usually sent to an international referral centre. Because of the distances involved, the usual follow-up period of one year could not be completed for all series included in this PhD Thesis.

The presentation of the audiometric data is in accordance with the Guide- lines of the American Academy (ref. American etc.) of Otolaryngology-Head and Neck Surgery. This facilitates comparison with the literature regarding hearing outcomes both now and in the future.

The following Chapter discusses in sequence the surgical findings and the hearing outcomes of the original publications presented in this PhD thesis.

Congenital Minor Ear Anomalies

Chapters 2-4 consist of three original publications on the outcomes of *primary* stapes replacing surgery in congenital minor ear anomalies, following the Cremers classification (ref. Tos, 2000; Teunissen & Cremers, 1993). The first paper presents the outcome of a consecutive series of patients with an isolated congenital stapes (footplate) ankylosis, treated by primary Causse stapedotomy with vein interposition. (ref. Vincent R., Wegner I., Kamalski D.M.A. et al., Otol Neurotol 2016;37:367-373)

It is a non-randomized, non-blinded study of prospectively collected data of 28 children, younger than 18 years of age, who underwent 35 consecutive operations for Class I and Class II congenital middle ear malformations (following the Cremers classification) from 1991 to 2014. None of the children were diagnosed with a concomitant syndrome. This might be the result of a selection bias, but might also be the result of missed diagnoses. Preoperative high-resolution CT-scanning of the petrous bones was performed. Those patients could thus be screened preoperatively for juvenile otosclerosis (ref. Vincent, Wegner, Vonck 2016) or Osteogenesis Imperfecta (ref. Vincent, Wegner Stegeman, 2014). Positive cases were excluded from this surgical report. Table 3 of Chapter 2 shows a review of the literature from 1993-2011, separately listing the hearing outcomes for surgery on congenital stapes ankylosis *without* (Class I) and *with* (Class II) associated ossicular anomalies.

The mean hearing gain for both classes I and II was reported as ranging from 18 dB to 21 dB. The percentage of air-bone gap closure < 10 dB and < 20 dB is presented for Class I and II separately for each series, when available. In general air-bone gap closure rates of < 10 dB are about 50%, while air-bone gap closure rates of < 20 dB are about 70%.

The air-bone gap closure rates for Class II were only rarely reported. For air-bone gap < 10 dB they vary from 23% (ref. Thomeer); 39% (ref. Teunissen), to 50% (ref. this PhD thesis Vincent). For air-bone gap < 20 dB they vary from 30% (ref. Park), 67% (ref. this study. Vincent), 70% (ref. Thomeer) to 72% (ref. Teunissen). These percentages have been placed in brackets since their number is less than 100 in total. The success percentages for Class II are somewhat worse when compared to Class I.

The reason for this might be the more complex anatomical ossicular chain anomalies, with possibly an epitympanic ossicular chain fixation or a missing long process of the incus. A malleostapedotomy procedure is usually a more difficult and sensitive procedure (Burggraaf et al, 2017).

A selection bias might influence the outcome of surgery in congenital stapes ankylosis Class I and II. This might complicate comparison between the published series. Nevertheless, detailed documentation of hearing outcomes and comparison with the litterature provides valuable data in these nowadays well defined pathologies.

The annual incidence of congenital minor ear anomalies (Chapter 2, 3 and 4) is fairly low, with wide individual variation in the specific ossicular chain anomalies. This makes strict application of evidence-based medicine principles not feasible.

The incidence of added postoperative sensorineural hearing loss is also shown, (Figure 2) by applying the Amsterdam Hearing Plots. This provides relevant information for the purpose of preoperative counseling. (Thomeer et al. Ann Otol Rhinol Laryngol, 2010; Thomeer et al. Arch Otolaryngol, 2011; Thomeer et al. Ann Otol Rhinol Laryngol, 2012)

In Table 1 and 2 the mean preoperative and postoperative sensorineural hearing components are mentioned separately for Class I and Class II. For Class I it is preoperatively 17 dB and postoperatively 20 dB, for Class II respectively 18 dB and 17 dB. Such variable sensorineural components in the total hearing impairment have also been presented and individualized for all Classes I to IV by Teunissen and Cremers (1993 Ann Otol Rhinol Laryngol; Thomeer et al., 2010 Ann Otol).

Thomeer provided for this aspect a review of the literature in his table 1, showing mean preoperative and postoperative sensorineural components varying in those series from 9 to 23 dB preoperative and from 6 to 24 dB postoperative. Such sensorineural components might be considered preoperatively as not really existing and being the outcome of audiometric variables, especially when coexisting with large conductive components. The outcomes from several series mentioned here are a warning not to do so.

Another frequent finding in the outcome is a remaining postoperative air-bone gap of 10-20 dB. When combined with a preoperatively existing sensorineural component, the remaining postoperative hearing impairment might necessitate fitting with a conventional air conduction hearing aid, even after successful surgery with an improved postoperative hearing level. An open ear canal fitting is recommended, if feasible.

As early as a hearing impairment has been diagnosed in bilateral and unilateral cases, hearing support must be organised, either by providing air conduction hearing aids or transcutaneous non-implantable bone conduction hearing aids, possibly attached to a softband or by an adhesive method. (Verhagen et al., 2008; Gawliczek et al, 2018) This must be done as early as possible, to stimulate auditory maturation at a cerebral level. It has great impact on the child's cognitive development, with repercussions on its lifelong educational and academic performance There is a large body of scientific evidence supporting early auditory rehabilitation. (Ref. Vogt et al. 2018)

Surgery for congenital minor ear anomalies in bilaterally and unilaterally affected children and adolescents aims at improving binaural hearing, if possible without the further need for conventional air conduction hearing aids or any type of partially implantable bone conduction hearing device. However, even after successful surgery, hearing aids may still be indicated. The outcome will in general be much better than it was preoperatively.

In **chapter 3** the surgical findings and outcomes for hearing are reported for 17 ears of children/adolescents under 18 years of age (mean age at time of this surgery was 12 years with a range 7-12 years) with congenital ossicular chain malformations with a mobile stapes footplate. (ref. Vincent et al. Laryngoscope 2015;126:682-688) These cases belong to Class III following the Cremers classification. (Tos, 2000; Teunissen & Cremers, 1993).

Table I presents the clinical and the preoperative and postoperative audiometric data, as well as the peroperative findings and the type of executed ossiculoplasty for each of 17 ears. One case has been lost to postoperative audiometric follow up. Ten cases (4, 8, 9, 10, 12, 13, 14, 15, 16, 17) had a columellar TORP reconstruction to the (neo) malleus/tympanic membrane and two cases (1 & 11) a partial ossicular chain (PORP) reconstruction. In two cases (6 & 7) a piston was placed between the long process of the incus and the mobile footplate.

In 3 cases (2, 3, 5) only a bony bridge needed to be removed to mobilise the intact ossicular chain. In two cases out of three (3 & 5) this successfully improved hearing. The postoperative air-bone gap was respectively 45, 10 and 5 dB.

A residual bony plate in the tympanic membrane was observed in 3 cases (8, 13, 17), fixing a somewhat malformed handle of the malleus. A malformed incus is reported to be present in all three. For one case (8) even a malformed stapes was mentioned. In all three cases a TORP reconstruction bypassing the stapes superstructure and the incus was executed.

The outcomes of the ossiculoplasty study on patients with an intact stapes and malleus, comparing the results of ossicular chain reconstruction using PORPs versus TORPs explain the preference given for a TORP ossicular chain reconstruction in those 3 cases. (Vincent, Rovers, Mistry, Oates, Sperling, Grolman, 2015).

The postoperative air-bone gap was respectively 11, 4, and 21 dB, resulting in postoperative hearing levels of 29, 15, and 36 dB. The hearing levels were influenced by the preoperative presence of a sensorineural component.

The presence of a residual partial bony plate in the tympanic membrane, usually in the anterior-superior part, is embryologically a classification transition marker in the distinction between major and minor congenital ear anomalies. According to the Tos and the Hear Subcommittee subclassification – (the extended Cremers classification) those 3 cases get the classification 2C1. To the best of our knowledge this is the first time that the surgical findings and hearing outcomes for each separate case of this specific anomaly have been reported. (Tos, 2000)

The final air conduction hearing levels are specified for each individual case. For case 1, 2, 7, & 17 the postoperative hearing impairment was still over 35 dB. So no or only a small improvement in postoperative hearing could be achieved. Four cases (25%) showed a postoperative air-bone gap (ABG) of >20 dB. Subsequent revision surgery performed before or after the 18th birthday closed the ABG to within 20 dB in those 4 ears. Thomeer et al (ref. 2012) scored 65% (15/23 cases) with an ABG ≤ 20 dB.

The overall audiometric outcomes of individual cases were visualized in two figures using the Amsterdam hearing evaluation plots (ref. de Bruijn et al). Table 3 shows a mean improvement in hearing level of 22 dB for 15 surgical procedures.

In comparison, Thomeer et al. (2012 ref) mentioned a mean improvement of 17 dB. (ref. Thomeer et al. 2012) Teunissen and Cremers reported for this class a mean improvement of 19 dB. (ref. Teunissen & Cremers, 1993)

Teunissen and Cremers reported for 5 of their 27 ears as syndromal diagnosis Treacher Collins, Klippel Feil and Frontometaphyseal Dysplasia. Thomeer et al. (ref. 2012 Ann Otol Rhinol Laryngol 2012;33:779-784) reported in 6 out of the 23 ears the following syndromal diagnosis: Treacher Collins, Branchio-Oto-Renal syndrome, maxillofacial dysostosis, Noonan syndrome, Turner syndrome and Crouzon syndrome. The Béziers study of Class III cases does not report syndromal diagnoses.

These three series from two centres differ in the applied surgical techniques for ossicular reconstruction and in the syndromic impact.

Nevertheless, the mean outcomes for hearing improvement do not vary that much, viz. 19 dB (Teunissen & Cremers 1993), 17 dB (Thomeer et al. 2012) and 22 dB (this study). For the reviewed other series in the literature, not clearly following the Cremers classification III as mentioned in table 4, the mean improvement varied from 16 to 37 dB.

In **Chapter 4** (ref. Vincent, Wegner, Derks & Grolman, 2016) the surgical findings and hearing results are reported for 17 ears in 14 children/adolescents under the age of 18 years with congenital oval or round window malformations (Class IV malformations). (Teunissen & Cremers Ann Otol Rhinol Laryngol, 1993).

The surgical findings and hearing outcomes are reported with a 1-year follow up for 12 out of 17 ears (71%). Hearing outcomes are only presented for 15 out of 17 ears. No details about those two missing ear surgeries were mentioned. The postoperative ABG was $<$ 10 dB in 6 out of 17 ears (40%), and $<$ 20 dB in 9 out of 17 ears. Table 2 summarises the outcomes from 4 other papers on Class IV malformation surgery, in comparison with the Béziers study.

As can be expected, our Class IV results are less good when compared to the results of Class I-II-III, but they are remarkably better than the results in the few other available publications.

Additional information on this topic was published by Sterkers and Sterkers (1988). They reported their surgical outcomes in 8 ears from two families with a congenital absence of the oval window with a malposition of the facial nerve (ref. Sterkers & Sterkers 1988). Included in this series were their first two cases published earlier on in 1980 (ref. Sterkers J.M, 1980). In 6 out of 8 ears the hearing improved dramatically, with an ABG $<$ 10 dB. Those patients were operated upon between 4 and 8 years of age.

Also Yang & Liu (2018 Ann Otol Rhinol Laryngol) reported on Class IV results, with an oval window atresia with or without round window atresia. However, they (mis) classified them as Class III. They reported on 12 cases and mentioned ossicular chain reconstruction by a vestibulum drill out and scala tympani fenestration. No specified audiometric outcomes were presented.

Tang et al. (2018 Eur Arch Oto Rhino Laryngol) reported on their series of 64 unilateral congenital minor middle ear anomalies applying the Cremers classification. They analyzed the outcomes only for 47 of 64 patients. In a table 3 they report their surgical findings for 7 ears with a Class IV anomaly (aplasia/dysplasia oval window/round window). The audiological outcomes are only presented in general for their total series of 47 ears. In the contralateral ear the hearing was normal in all 64 patients.

High resolution CT-scanning of the inner ear has now become available as a routine preoperative screening method. This allows timely identification of malformation cases.

Cochlear implant surgery opened up another new development. Creating an atraumatic cochleostomy has now become a regular surgical procedure. The availability of better diagnostics by preoperative HR CT-scanning of the inner ear, experience with the performance of atraumatic cochleostomies and the encouraging hearing outcomes presented in our study might generate the possibility for good and consistent hearing results, even for Class IV congenital minor ear anomalies. Moreover, the availability of percutaneous and transcutaneous, passive and active, partially implantable hearing devices have made optimal hearing revalidation possible for all. By applying a softband or adhesive application this option has already become available, even bilaterally, soon after birth.

The application of the Amsterdam Hearing Plot for reporting Class IV series hearing outcomes is useful to identify and visualise smaller or larger individual inner ear deteriorations, like it was done by Thomeer et al. (ref. 2012) and by Vincent. (ref. 2016) Thomeer (ref. 2012) reported for one ear (case 7) total inner ear deafness as result of a concomitant acute otitis media. The application of the vein interposition technique might possibly have prevented that complication. In the large consecutive series of Thomeer et al. (2012) with 106 cases and in the previous series by Teunissen and Cremers (ref. 1993) of 144 congenital minor ear anomalies, that was the only case with a postoperative total inner ear deafness. This indicates that, in general, the risk for inner ear deafness as the outcome of surgery for congenital minor ear anomalies is minimal. The data reported in this PhD thesis support this belief .

Pedersen and Felding (1991) (ref. Pedersen and Felding Ann Otol Rhinol Laryngol 1991;100:607-611) convincingly demonstrated that infection with the influenza virus is responsible for unexpected complications following stapes surgery. In an analysis of 1111 patients who had a stapedectomy or stapedotomy performed within a 10-year period and in a thorough investigation of eleven patients who suffered total hearing loss and eight patients who developed partial sensorineural hearing loss after surgery, a close relation was found between the appearance of the complication and a period of influenza epidemics. In fifteen of those nineteen ears no explanation was found for postoperative deafness. The primary operation was mainly a House wire large fenestra technique. In practically all anacusis cases hearing was good during and after operation. Pedersen and Felding (1991) showed a relation between the incidence of influenza in Denmark and the occurrence of anacusis and partial sensorineural hearing loss following stapedectomy in their series (ref. Tos, 2000; ref Pedersen & Felding, 1991).

As stated earlier in chapter 4 for this Class IV anomaly “the decision-making process” for the patients in this series was based on case-by-case discussion with the patients and their families. The surgeon’s experience has led him to believe that many children in France do not like wearing hearing aids. Knowing that hearing loss has negative effects on school performance, especially in case of bilateral hearing loss, the patients and their families opted for surgery.

Chapter 5 presents the surgical findings and hearing outcomes of primary stapes surgery in 32 ears of 25 patients with Osteogenesis Imperfecta type 1. In total 46 consecutive ears were operated from 1994 to 2013. (ref. Vincent 2014). However, in 4 cases the required postoperative hearing data were not available, and 10 cases concerned revision cases. These cases (N=14) were excluded for the report.

The mean age of these 25 patients at surgery was 36 years (Range 18-59 yr). In the literature this varied from 30.6 yr. to 32.3 yr. (Garretsen & Cremers, 1990;1991). Fourteen of these 25 patients (56%) were female. The male: female ratio was more to the female side in other larger series (Garretsen 1980; 1991).

The stapes footplate was found to be fixed in all cases. Obliteration of the oval window niche was identified in 13/32 cases. The mean audiometric follow-up time was 7 months, varying from 2 to 41 months, which is relatively short for a part of this series. In 28 out of 32 (88%) primary stapedotomies the postoperative air-bone gap was < 10 dB in the short term. This success rate subsequently declined to 72.2%. Swinnen et al (ref. 2012) had already reported this decline in the size of the air- bone gaps at the short time for the patients having a minimal follow up of 1 year. Mean postoperative air-bone gap, mean improvement in air conduction and success percentages slightly deteriorated over the years. A postoperative sensorineural loss exceeding 15 dB occurred once. For 6 out of these 32 cases the air-bone gap was > 10 dB. In 4 of those revision surgery was carried out. The mean postoperative bone conduction level was mentioned to be 26.1 to 26.5 dB for 18 ears having a 1 yr. follow up. The mean postoperative hearing gain was noted to be 22.3 dB (for 32 ears).

Other authors reported success rates of 75-85% and additionally some sensorineural hearing loss in 0 to 8%. We agree that in accordance with the Committee of Hearing and Equilibrium Guideline (ref. American Academy, 1995) a follow-up duration of at least 1 year is to be preferred when reporting AC thresholds (improvement) and air- bone gap closure. This report failed partly to fulfill that requirement because, as previously mentioned, many patients involved in this study had to travel long distances to reach the Béziers Causse Clinic.

Reporting on their surgical series for Osteogenesis Imperfecta, Garretsen and Cremers (ref. Ann Otol Rhinol Laryngol 1991;100:120-130) suggested a possible explanation for the slowly progressive postoperative sensorineural component in 12 out of 58 stapes surgeries. They showed that in the contralateral (non-operated) ear such a progressive sensorineural component was also found.

Garretsen and Cremers also analyzed the literature on cases with disappointing results, trying to identify possible predictive features, both in the preoperative work-up and at surgery, correlating with those disappointments. They named a quite progressive sensorineural component in the hearing loss as a factor. They also stated that the group with disappointing results directly after surgery was relatively young and had high preoperative bone conduction thresholds. All these patients had complication-sensitive changes in the footplate and oval window (Garretsen & Cremers, 1991). In our more recent study (ref. Vincent, 2014) we could not confirm these correlations. Swinnen et al (2012 Laryngoscope mineral density) reported on an association between bone mineral density and hearing loss in Osteogenesis Imperfecta.

The annual incidence of Osteogenesis Imperfecta especially with large conductive hearing losses is so low that strict application of evidence- based medicine principles is not feasible.

Chapter 6 presents the surgical findings and audiometric outcomes of a prospective study of 41 consecutive primary stapedotomies in 34 children/adolescents with otosclerosis, performed from 1991 to 2014. (ref. Vincent et al., 2015)

The cases described in this paper were also partially included in a previous series published in 2006 on the surgical findings and hearing results in 3.050 stapedotomies for primary otosclerosis. (ref. Vincent et al., 266;27:S25-S47)

This additional publication is valuable because it extends the series with additional cases and reports the audiometric data in more detail. This allows evaluation of the stability of the audiometric outcomes over time.

In the 2006 publication the success rate for children with a closure of the air-bone gap < 10 dB was 94%. In the 2015 publication the ABG closure of < 10 dB is 93%, and the ABG closure < 20 dB was 98%. An audiometric follow up of one year could only be realized for 38 out of 41 primary stapedotomies. The mean follow up was 9 months (range 12 to 196 months).

The mean hearing gain was 23 dB. A sensorineural additional hearing loss of 13 dB occurred in one case at last follow up. Since the natural course of otosclerosis most often shows a progressive sensorineural component over time, this could explain that slight deterioration.

The annual incidence of otosclerosis in adolescents is so low that strict application of evidence- based medicine principles is not feasible.

Otosclerosis in children

Häusler from Bern reported in his publication on advances made in stapes surgery (Häusler 2004 Thieme) also on the topic of otosclerosis in childhood. He referred to Lippy et al. (1998) stating that although a first slight hearing loss had already been noted before the age of 20 years in 15% of patients, the diagnosis of otosclerosis was first made when they were adults. In Häusler's series of otosclerosis cases six patients (1%) had surgery before the 18th year of life (11-17 years). A marked obliterative otosclerosis was present in three out of these six patients. A slight inner ear component was already present in two of them. Correct and even spectacular hearing improvements were obtained in all six children.

A review of the literature of the audiometric results for juvenile otosclerosis was given by Vincent et al. (ref. Vincent et al. 2016) over a period spanning from 1982-2013.

Air-Bone Gap closure < 10 dB was reached in 40% to 100% in these studies. Air-Bone Gap closure < 20 dB was reached in 80% to 100% of children.

Over time opinions have differed as to when and whether a stapes operation should be carried out in children. It was and is considered to be more appropriate for a hearing aid to be fitted as a first treatment option in children. It is also thought that in children with bilateral otosclerosis a stapedotomy should only be carried out on one ear (Fisch, 1994). In recent studies the postoperative results are reported as being as good as in adults. Earlier studies, however, reported less good hearing gains. It is recognized that before embarking on stapes surgery in general, and especially in children, there needs to be personal and delicate deliberation, coupled with careful family counselling, with the input of the child if appropriate.

In **chapter 7** the causes of failure of primary stapes surgery and the hearing results of revision stapes surgery in a consecutive series of 652 cases in 634 patients are presented. These 652 consecutive revision stapes operations were performed between April 1992 and December 2007. The first aim of this prospective study was to evaluate the causes that had led to the failure of the previous surgery. The second aim was to report hearing results of revision surgery in a series of 652 consecutive surgical revisions by the same surgeon.

Of 652 operations, 583 (89,4%) were primary revision operations, 53 (8.2%) were secondary revision operations, 8 (1.2%) were tertiary revision operations and 8 (1.2%) were quaternary revision operations. Ninety (13.8%) out of 652 cases had previously been operated on by the author ("personal revision") and 561 cases (86.2%) had previously been operated upon by another surgeon ("other surgeon revision").

The indications for revision surgery were a recurrent or persistent 4-frequency average Air-Bone Gap (ABG) greater than 10 dB in 616 cases (94.4% of the 652 cases) or the suspicion of a perilymph fistula with dizziness, disequilibrium and the presence of a fistula sign in 36 cases (5-6% of the 652 cases). So, the size of the ABG was the only criterion for revision surgery in 616 cases. All preoperative ABG's were more than 20 dB.

In table 1 the surgical findings in revision surgery after previous primary surgery (583 cases) showed as the main causes of failure: eroded incus (27.6%), dislocated prosthesis (18.2%), too short prosthesis (12.7%), too long prosthesis (4.6%).

In the personal revisions of previous surgery those causes of failures were equally ranked in order.

A perilymphatic fistula was present in 25/493 (5%) of the "other surgeon revision" cases and in 8/90 (9%) of the personal revision cases. A perilymph fistula was mainly observed during the first year after the primary operation (6 out of the 8 cases occurred within the first 12 months). In all 8 cases the fistula was identified at the anterior pole of the oval window niche and was in all cases related to a vein graft which was not perfectly adherent to the anterior pole of the footplate. Of the 36 cases explored for perilymph fistula, 12 cases (33%) had total preoperative sensorineural hearing loss (SNHL), anacusis, (33.4%); 22 cases (61%) had no ABG preoperatively and 2 cases (5.6%) had a preoperative ABG of 20 dB or less. A total stapedectomy was performed in 21 out of 35 cases (60%) while a stapedotomy was only performed in 14 out of 35 cases (40%).

As previously described the passage of time does not protect against the occurrence of fistula, because in the series of Somers et al. (ref JLO, 1997) the mean delay was 5 years and the longest delay found was 13 years. In the Bern series of 114 stapes revision series (R. Häusler, Bern, 1992-1998) perilymph fistulae were never found despite a meticulous search for these (Ref. Häusler 2004).

Table 2 shows the main causes of failure correlated to the time of failure after the previous operation in the 90 cases of the personal revision series. Immediate failures and failures that occurred within the first year after the primary operation were mainly related to an inappropriate prosthesis length (11 cases, 27.5%), whereas failures that occurred after one or several years were mainly related to incus erosion (23 cases, 46%) and prosthesis dislocation (13 cases, 26%).

The surgical findings in our study are consistent with the previously published series of revision surgery, in that the most common causes of failure of the primary surgery were incus erosion and prosthesis displacement. (Gros et al., 2005; De la Cruz & Fayad, 2000; Han et al., 1997; Lippy et al., 2003; Hammerschlag et al., 1998; Berenholtz et al., 2002; Lesinski, 2002)

However, the analysis of causes of 114 stapes revision operations by Häusler (Bern, 1992-1998) (Ref. Häusler 2004) was different. High revision scores were related to the stapedectomy with the Schuknecht wire adipose tissue prosthesis or House wire-loop with vein/perichondrium graft (29 out of 114 ears). Causes related to the stapedotomy with piston insertion (36 out of 114 ears) were the main causes in his series.

Our results show that improvement of a significant conductive hearing loss after unsuccessful primary or revision stapes surgery can be accomplished in most cases. The postoperative ABG was closed to within 10 dB in 63.4% of cases. However, this is significantly less than in our primary stapedotomy patients where an ABG closure to within 10 dB was achieved in 94.2% of cases. (ref. Vincent- Sperling-Oates-Jindal O&N, 2006)

In **Chapter 8** the hearing results of two different types for ossiculoplasty in patients with an absent incus and an intact stapes and malleus are compared to each other. (Ref Vincent et al. O&N 2011;32:616-625). Five hundred eighty-five patients undergoing ossiculoplasty were enrolled in this study from April 1991 to May 2010.

Comparative analyses were made between a group of 304 patients with 310 operated ears who underwent ossiculoplasty with a partial ossiculoplasty replacement prosthesis (PORP) positioned from the malleus to the stapes head and 281 patients with 318 operated ears who underwent ossiculoplasty with a total ossiculoplasty replacement prosthesis (TORP) positioned from the malleus to the footplate. The impact of the application of new surgical developments, such as the Silastic banding and malleus relocation technique, were also analysed.

The surgeon based his choice of the prosthesis (PORP and TORP) used for ossicular reconstruction on his progressive experience. During the period when he preferred using the PORP prosthesis, he introduced two original techniques: the malleus relocation technique (ref. Vincent et al., O&N 2006;25:223-230) and the Silastic banding technique, used to stabilize the PORP prosthesis. Later on, the use of a TORP became the preferred method of ossicular reconstruction despite the presence of an intact stapedial arch.

Later on, the use of a TORP became the preferred method of ossicular reconstruction despite the presence of an intact stapedial arch.

In this study Austin's classification (ref) of ossicular defects as modified by Kartush (ref.) was used to define the ossicular status encountered.

The malleus relocation technique was used only in 131 ears in the PORP group (in total 310 ears).

In 137 ears (44%) of the PORP group 1 (N=310), tympanic membrane grafting with a tragal perichondrium underlay technique was applied to repair the perforation.

In the TORP group, ossiculoplasty was performed with the same TORP prosthesis in all cases (N=318), despite the presence of a stapedial arch. The TORP was positioned from the malleus handle to the stapes footplate. The prosthesis shaft was inserted between the Fallopian canal and the stapes superstructure where possible. Where this was not feasible it was inserted between the stapes superstructure and the promontory.

In 153 ears (48%) out of 318 ears tympanic membrane grafting with a tragal perichondrium underlay technique was applied to repair the perforation.

Table 2 presents the hearing outcomes separately for the PORP and for the TORP procedures. Audiometric data were available for 93% of the PORP group and for 89% of the TORP group. The postoperative 4-frequency average ABG was 13.1 dB in the PORP group compared with 8.9 dB in the TORP group. The average follow-up time was 21.7 months (range 3-121.8 months). The mean age was 52.5 years (range 7-80 yr.).

The Austin-Kartush classification does not take into account the distance and spatial relationship between the structures in the middle ear. Hence, technique selection can vary to suit the anatomy. Vlaming and Feenstra (ref. Vlaming & Feenstra, 1986) demonstrated that a malleus positioned directly over the stapes provides the ideal situation for reconstruction. They have also explained that in the stapedo-malleopexy ossiculoplasty the transmittance of the otoacoustic energy to the footplate was the less effective the more the malleus handle was positioned anteriorly. Over time otologists have started to apply titanium PORP prostheses fixed with a clip fixation to the stapes head, acting as a columella directly to the medio-posterior part of the tympanic membrane and covered by sliced autologous cartilage underneath the tympanic membrane to prevent extrusion of the titanic PORP.

The aim of the malleus relocation technique is to ideally place the prosthesis in order to maximize transmittance of the otoacoustic energy from the malleus to the stapes head or footplate.

Table 5 presents an overview from the literature comparing hearing outcomes of PORP and TORP reconstructions. When compared to 4 other published series, our ossiculoplasty results in the Austin-Kartush group A still figure as the best. However, the literature shows that the outcomes of ossiculoplasties for the Austin-Kartush group A deteriorate remarkably at 1- year and 5-year follow-up. Therefore, it is relevant to have long-term results available to establish the stability of the TORP results and to compare them with the PORP results.

Also, when evaluating and comparing the hearing outcomes for ossiculoplasties, it is important to acknowledge the effect of many confounding factors on the outcome, even for ears within the same Austin-Kartush groups. These factors are often not clearly outlined in the published series. For our series, the following effects on outcome are under investigation: selection bias, the possible variance in measuring and interpreting individually audiometric outcomes between the presented ossiculoplasty series, the audiometric follow-up time, the aeration status of the middle ear cleft, the etiology of the ossicular damage (chronic otitis media or trauma), the definition of whether the ossiculoplasty was an isolated surgical act or part of a more comprehensive tympanoplasty and whether the follow-up has been completed for the whole consecutive series. The malleus relocation technique and the Silastic banding technique presented here are new surgical variables, gradually applied in this ossiculoplasty series for Austin-Kartush group A ossicular defects.

Evaluating and publishing the clinical and hearing outcomes of such new surgical procedures is needed to keep improving the outcomes of such elective surgical procedures. Evidence-based medicine principles might be applied in these studies.

Chapter 9 presents the preliminary results of a new malleus replacement prosthesis combined with a total ossicular prosthesis, used for reconstruction of the ossicular chain in 92 ears of 90 patients with a missing malleus and stapes superstructure. The patients for the study and surgery were enrolled from September 1994 to March 2012.

The results of 2 groups were analysed and compared. The first group comprises 34 ears in which the reconstruction was made with a TORP positioned from the tympanic membrane to the (mobile) footplate (TM-to-footplate assembly). Surgery took place between September 1994 and April 2009. The second group comprises 58 ears in which the reconstruction was made with a TORP positioned from a newly designed malleus replacement prosthesis (MRP) to the mobile footplate (MRP-to-footplate assembly). Surgery took place between January 2010 to March 2012. The surgical technique and the positioning of the malleus-replacement-prosthesis (MRP) connecting to the groove of the TORP's head is well illustrated in this paper.

All cases were revision tympanoplasty cases in ears with chronic suppurative otitis media (CSOM) without active cholesteatoma.

In this analysis only cases allocated to Austin-Kartush group D (stapes and malleus absent) were included. (ref. Austin, 1972; Kartush, 1994).

Table 1 presents the preoperative patient characteristics for both study groups.

Table 2 presents the hearing results at 3-months follow-up for each group separately.

The pre- and postoperative bone conduction levels were respectively 30.8 dB and 31.1 dB for group 1 (TM-to- footplate TORP) and respectively 25.9 dB and 25.2 dB for group 2 (MRP-to- footplate TORP).

The pre- and postoperative ABG was respectively 34.6 dB and 23.3 dB for group 1. For group 2 the pre- and postoperative ABG was respectively 33.1 dB and 12.5 dB (3 months in follow-up) and 12.9 dB (1-year follow-up).

In group one 14 out of 27 ears (52%) had a postoperative ABG > 20 dB, while in group 2 this was found in 9 out of 43 ears (21%).

None of the patients in the two surgical series experienced dizziness or increased sensorineural hearing loss (SNHL) during the postoperative follow-up.

Extrusion of the prosthesis occurred at short-term follow-up for each group twice. The extrusion rate was thus 7.2% (2 out of 27 ears) in group 1 and 5% (2 out of 43 ears) in group 2.

Comparing the results of both groups, the MRP group seems to show better hearing results. However, it must be acknowledged that the comparison is based on a restricted number of patients and on a relatively short follow-up period, which poses a risk for deterioration of the hearing results over time. The authors have indeed experienced the need to cover the MRP with a larger plate of cartilage interposed between the tympanic membrane and the MRP in order to keep the results stable.

Also osseointegration of the MRP in the bone of the tympanic frame is a potential risk, because it leads to fixation of the MRP and a decline of the hearing results. The present MRP has been designed so that the titanium link can be cut at a second stage, freeing the neomalleus handle from any connection with the surrounding bone.

Looking at the different stages of a surgical innovation process, the present study covered the "exploration phase", i.e. prospective database research focusing on the development of the new surgical technique. (Ref. McCulloch et al. 2009) Considering the safety and the superior postoperative hearing results of the MRP-to-footplate technique in the present study, the next phase should be a randomized trial to assess the effectiveness against the current standards.

The mean postoperative bone conduction levels in these 2 groups were around 31 dB (group 1) and 25 dB (group2). In combination with a mean air-bone gap of 13 dB (group 2 with 1 year follow-up) this means that many of the involved patients will additionally need a conventional air conduction hearing aid for an optimal hearing result, possibly with an open ear canal fitting.

Hüttenbrink stated in 2004 (ref. Hüttenbrink, 2004) that due to the piston-like mode of ossicular vibration, sound transmission becomes less efficient when the axis of the prosthesis is tilted away from the direction of the vibration. The decrease is proportional to the square of the cosine of the angle of the connection that is placed between the tympanic membrane and the footplate. Thus, a prosthesis mounted between the footplate and the malleus handle, whose long axis is approximately in line with the vibrational axis, will transmit sound better than a prosthesis place more obliquely between the stapes head and the umbo. Clinically this was already shown to be the case in our previous study of ossiculoplasty in patients with in intact stapes and malleus. (Vincent et al, 2011 O&N 32;616-625).

In this study the same biomechanical mechanism is applied, by positioning theneo-malleus straight above the oval window and the mobile footplate. An additional advantage of this position is that it improves the long term stability of the columella.

Also the Silastic banding technique adds to the stability, by securing the columella to the head of the stapes in ears with an intact malleus and stapes superstructure.

So, in those two new ossiculoplasty studies included in this PhD thesis the laws of biomechanics for the optimal functioning of an ossiculoplasty have been followed and have been applied. This results in better hearing outcomes, compared to previous studies from the literature in which the principles of biomechanics for a reconstructed ossicular chain were not yet applied.

Those outcomes now need to be confirmed in randomised prospective trials, to assess the effectiveness against the current standards, preferably by other otologists in tertiary referral centres worldwide. Implementation of evidence-based medicine principles might become part of those new studies.

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SUMMARY

This PhD thesis presents eight original publications on elective reconstructive ear surgery.

Five of the first six concern stapes surgery.

Three of those are series of primary surgery for congenital minor ear anomalies. Two of them also concern stapes replacement surgery. They present unique data, because of the systematic application of the Causse stapedotomy technique with vein interposition. They are also relevant because they provide the surgical outcomes of relatively large consecutive series of quite rare pathological conditions, such as minor congenital minor ear anomalies, Osteogenesis Imperfecta and primary stapedotomies for otosclerosis in adolescence. The sixth publication reports on the outcome of a prospective series of 652 revision stapedotomies.

In the seventh and eighth publication the outcomes of two different types of ossiculoplasties are presented and discussed.

Chapters 2, 3 and 4 present the results of three original surgical series with congenital minor ear anomalies, following the Cremers classification.

Chapter 2 presents the surgical findings and outcomes in consecutive series of ears with an isolated congenital stapes footplate ankylosis with or without associated ossicular anomalies (respectively Class I and II). In all cases the Causse stapedotomy technique with vein interposition was used. Twenty-eight children, younger than 18 years, underwent 35 consecutive primary stapedotomies from 1991 to 2014.

The mean hearing gain for both classes I and II ranged from 18 dB to 21 dB. A postoperative air-bone gap (ABG) closure of 10 dB or less was achieved in 73% of Class I cases and in 50% of Class II cases.

A postoperative ABG closure of 20 dB or less was achieved in 77% of Class I cases and 67 % of Class II cases.

We concluded that stapedotomy is a safe and feasible treatment option in children with congenital stapes ankylosis.

Chapter 3 reports on the surgical findings and hearing outcomes of 17 ear interventions in children / adolescents under 18 years of age (range 7-12 years), presenting with congenital ossicular chain anomalies with a mobile stapes footplate (Class III according to the Cremers classification). Various surgical techniques were needed and performed in this group. Both Partial Ossicular Reconstruction Prostheses (PORPs) and Total Ossicular chain Reconstruction Prostheses (TORPs) have been applied.

Sometimes only the removal of a bony bridge, fixating some part of the ossicular chain, was needed to mobilise the ossicular chain. In three ears we found an atretic bony plate in the tympanic membrane. This could be associated with a malformation of the incus or stapes. These 3 ears belong to the 2C1 Class, according to the classification presented by TOS and the HEAR group (the extended Cremers classification). The overall audiometric outcomes of individual cases are visualized in two figures using the Amsterdam hearing evaluation plots.

Chapter 4 reports on the surgical findings and hearing results of 17 ear interventions in 14 children / adolescents under the age of 18 years, presenting with congenital oval or round window malformations (Class IV Cremers classification). Hearing outcomes are presented for 15 out of 17 ears. Only 12 ears have a 1-year audiometric follow-up. For 6 ears a postoperative ABG < 10 dB was obtained, and for 9 ears an ABG < 20 dB.

Chapter 5 presents the surgical findings and hearing results of primary stapes surgery in 32 ears of 25 patients with Osteogenesis Imperfecta type 1. The required postoperative hearing data were not available for 4/32 ears.

The mean age of these patients at surgery was 36 years (range 18-59 yr.). The stapes footplate was found to be fixed in all cases. Obliteration of the oval window niche was identified in 13/32 cases. The mean audiometric follow-up time was 7 months (range 2 to 41 months), which is relatively short for a part of this series.

The short-term audiometric follow-up showed an ABG <10 dB in 28/32 ears.

Chapter 6 reports on the surgical findings and audiometric results of 41 consecutive primary stapedotomies in 34 children with otosclerosis. A part of this series was already previously published in 2006. Those cases are also included in this paper. The paper is relevant because it provides more detailed audiometric data. In addition, the series has a higher number of cases.

In 93% of the cases the ABG was < 10 dB, and in 98% the ABG was < 20 dB.

An audiometric follow-up of at least 1 year was available for 38/41 cases. The mean follow-up duration was 90 months.

Chapter 7 presents a large series of revision cases after failure of previously executed primary stapes surgery. The paper describes the causes of failure of the primary surgery and the hearing results of the revisions in a consecutive series of 652 cases in 634 patients. The 652 consecutive operations were performed between April 1992 and December 2007.

The most frequent causes of failure in this series were: eroded incus (27.6%), dislocated prosthesis (18.2%), prosthesis too short (12.7%) and prosthesis too long (4.6%).

A perilymph fistula was found in 25/393 (5%) cases previously operated by other surgeons and in 8/90 (9%) cases previously operated by the author.

The postoperative ABG was closed to within 10 dB in 63.4% of cases. The success rate is significantly less than in our primary stapedotomy patients where an ABG closure to within 10 dB was achieved in 94.2% of cases.

Chapter 8 compares the hearing results of two different types of ossiculoplasty in ears with an absent incus but an intact stapes and malleus.

The first group consisted of 304 patients with 310 operated ears. The ossiculoplasty was realised with a Partial Ossiculoplasty Replacement Prosthesis (PORP) positioned from the malleus to the stapes head.

The second group consisted of 281 patients with 318 operated ears. The ossiculoplasty was realised with a Total Ossiculoplasty Replacement Prosthesis (TORP) positioned from the malleus to the footplate. During the course of the series some innovative technical improvements were applied, viz. the Silastic banding technique and the malleus relocation technique. The postoperative 4-frequency average ABG was 13.1 dB in the PORP group and 8.9 dB in the TORP group. The average follow-up time was 21.7 months (range 3-121.8).

Chapter 9 presents the preliminary results of a new malleus replacement prosthesis combined with a total ossicular prosthesis, for ears in which both the malleus and the stapes superstructure are missing. The series consists of 92 ears of 90 patients, who were operated from September 1994 to March 2012.

Comparative analyses were made of the results of 2 subgroups.

The first subgroup consists of 34 cases. The operations were performed between September 1992 and April 2009. The ossiculoplasty was realised with a TORP positioned from the tympanic membrane to the mobile footplate (TM-to-footplate assembly).

The second subgroup consists of 58 cases. The operations were performed between January 2000 and March 2012. The ossiculoplasty was realised with a TORP positioned from a newly designed malleus replacement prosthesis (MRP) to the mobile footplate (MRP-to-footplate assembly).

All cases were revision tympanoplasty cases with a history of chronic supurative otitis media (CSOM) without active cholesteatoma.

In group 1 a postoperative ABG > 20 dB was still present in 14 out of 27 ears (52%), while in group 2 this was only the case in 9 out of 43 ears (21%). The hearing results at 3 months follow-up are presented for each group separately in a table. These outcomes need now to be confirmed in randomised prospective trials to assess their validity against the current standard techniques. The trials should preferably be done in other otosurgical tertiary referral centres.

SAMENVATTING

Dit proefschrift bevat acht originele publicaties over electieve oorchirurgie om het gehoor te verbeteren.

Vijf gaan over stijgbeugelvervangende chirurgie. Drie publicaties betreffen de bevindingen en resultaten van oorchirurgie voor verschillende klassen van aangeboren anomalieën van de gehoorbeenketen. In twee daarvan is ook sprake van stijgbeugelvervangende chirurgie. Deze vijf publicaties zijn relevant omdat systematisch “de Causse stapedotomie techniek met vene interpositie” werd toegepast als methode voor het vervangen van de stijgbeugel. Bovendien gaat het om tamelijk grote opeenvolgende series van ingrepen en werden ze uitgevoerd omwille van verschillende zeldzame oorziekten, met name aangeboren anomalieën van het middenoor, Osteogenesis Imperfecta en otosclerose op tiener leeftijd.

De zesde publicatie is een prospectieve studie die de peroperatieve bevindingen en gehoorresultaten rapporteert van een serie van 652 revisie ingrepen met stijgbeugelvervangende chirurgie.

In de laatste twee publicaties worden de resultaten van twee verschillende types van reconstructie van de gehoorbeenketen gepresenteerd en besproken.

In de **hoofdstukken 2, 3 en 4** worden drie originele series van ooringrepen omwille van aangeboren anomalieën van het middenoor gepresenteerd, waarbij de Cremers classificatie gevolgd wordt.

In **hoofdstuk 2** worden de chirurgische bevindingen en de resultaten van gehoorverbeterende reconstructieve chirurgie beschreven voor aaneengesloten series van oren met een geïsoleerde congenitale stapes voetplaat ankylose zonder of met een geassocieerde ketenanomalie (respectievelijk Klasse I en Klasse II). Hierbij werd de Causse stapedotomie met vene interpositie gebruikt. Tussen 1991-2014 werden bij 28 kinderen jonger dan 18 jaar in totaal 35 eerste stapedotomieën verricht. De postoperatieve gehoorwinst voor beide Klassen I en II samen lag tussen de 18 dB en 21 dB. Een luchtbeengeleidings component ≤ 10 dB werd voor Klasse I in 73% en voor Klasse II in 50% van die oren bereikt. Geconcludeerd wordt dat het verrichten van een stapedotomie bij kinderen met een aangeboren stijgbeugelfixatie een veilige en goede behandeloptie is.

In **hoofdstuk 3** worden de chirurgische bevindingen en de resultaten van de in opzet gehoorverbeterende chirurgische ingrepen beschreven voor 17 oren bij kinderen / adolescenten jonger dan 18 jaar (spreiding 7-12 jaar) met een aangeboren ketenanomalie met tevens een beweeglijke stijgbeugel voetplaat, dus Klasse III volgens de Cremers classificatie. Onderling nogal verschillende chirurgische procedures waren nodig en werden uitgevoerd. Er werd gebruik gemaakt van zowel partiële gehoorbeenketenprothesen (PORP) als totale gehoorbeenketenprothesen (TORP). Soms was alleen de verwijdering nodig van een benig bruggetje, dat de gehoorbeenketen ergens deels fixeerde, om die te mobiliseren.

Een gedeeltelijke nog in het trommelvlies aanwezige atretische plaat werd in 3 van deze 17 oren aangetroffen, al of niet geassocieerd met een tevens misvormd aambeeld of stijgbeugel. Deze 3 oren behoren tot de 2C1 klasse volgens de door Tos en de HEAR-groep gebruikte subclassificatie (de uitgebreide Cremers classificatie). De gehoorresultaten van deze chirurgie zijn voor deze groep als geheel en voor elk geval individueel in 2 figuren weergegeven met behulp van de Amsterdam Hearing plots.

Hoofdstuk 4 beschrijft de chirurgische bevindingen en de gehoorresultaten van 17 gehoorverbeterende operaties bij 14 kinderen / adolescenten jonger dan 18 jaar met een aangeboren anomalie van het ovale of ronde venster (Klasse IV Cremers classificatie). De audiometrische opvolging was beschikbaar voor 15 van de 17 oren, en slechts voor 12 hiervan gedurende een termijn van één jaar.

Bij 6 oren kon een lucht-beengeleidings component ≤ 10 dB bereikt worden en bij 9 oren een luchtbeengeleidings component ≤ 20 dB.

In **hoofdstuk 5** worden de chirurgische bevindingen en de resultaten van een primaire stapedotomie gepresenteerd voor 32 oren in 25 patiënten met Osteogenesis Imperfecta type 1. Voor 4 van deze 32 geopereerde oren ontbreken de benodigde audiometrische resultaten.

De gemiddelde leeftijd bij heelkunde was 36 jaar (spreiding 18 - 59 jaar). In alle gevallen bleek de stijgbeugelvoetplaat gefixeerd te zijn. Een obliteratie van de ovale vensternis werd in 13/32 gevallen gevonden. De gemiddelde audiometrische opvolging was 7 maanden (spreiding 2 van 41 maanden) en dus tamelijk kort voor een deel van deze serie.

Voor 28 van deze 32 eerste stapedotomieën (88%) werd op de genoemde korte termijn een lucht-beengeleidings component ≤ 10dB bereikt.

In **hoofdstuk 6** worden de chirurgische bevindingen en de resultaten van een primaire stapedotomie in 41 oren van 34 kinderen met otosclerose gepresenteerd. Een deel van deze chirurgische serie werd in 2006 reeds eerder beschreven. Deze aanvullende publicatie is van betekenis omdat nu de gehoorresultaten gedetailleerder worden beschreven. Bovendien is de huidige serie uitgebreider.

Een audiometrische opvolging van minstens 1 jaar kon slechts gerealiseerd worden voor 38 oren, met een gemiddelde opvolgingsduur van 90 maanden (spreiding 12 tot 196 maanden). In 93% werd een lucht-beengeleidings component ≤ 10 dB bereikt. In 98% werd een lucht-beengeleidings component ≤ 20 dB bereikt.

In **hoofdstuk 7** wordt gerapporteerd over een grote reeks revisie operaties na faling van eerder uitgevoerde primaire stijgbeugelvervangende heelkunde. Zowel de oorzaken van het mislukken van de primaire stapedotomie als de gehoorresultaten van de revisie chirurgie worden beschreven voor 652 gevallen bij 634 patiënten. Deze 652 elkaar opvolgende revisie stapedotomieën werden uitgevoerd tussen april 1992 en december 2007.

De belangrijkste oorzaken voor het falen van de primaire stapedotomie bleken te zijn: een gearrodeerd lange been van het aambeeld (27,6%), een gedisloceerde prothese (18,2 %) een te korte prothese (12,7%) of een te lange prothese (4,6%).

Een perilymfe fistula bleek aanwezig bij 25/393 (5%) gevallen van deze revisie serie wanneer ze afkomstig waren van andere chirurgen, en bij 8/90 (9%) van de persoonlijke revisie serie van de auteur.

Een postoperatieve lucht-beengeleidings component van ≤ 10 dB werd bereikt voor 63,4% van deze operaties. Dit percentage is opvallend veel lager dan de 94,2% die we behalen bij onze primaire stapedotomieën voor otosclerose.

In **hoofdstuk 8** worden de gehoorresultaten met elkaar vergeleken voor twee verschillende vormen van reconstructie van de gehoorbeenketen in oren met een ontbrekend aambeeld maar een intacte stijgbeugel en hamer.

De eerste groep van 310 oren van 304 patiënten kreeg een gehoorbeenketen reconstructie met een “Partial Ossiculoplasty Replacement Prosthesis” (PORP) welke geplaatst werd tussen de hamer en het kopje van de stijgbeugel.

De tweede groep van 318 oren van 318 patiënten onderging een reconstructie van de gehoorbeenketen met een “Total Ossiculoplasty Replacement Prosthesis” (TORP), die geplaatst werd tussen de hamer en de stijgbeugelvoetplaat. In de loop van deze serie werden nog technische details toegevoegd aan de techniek, met name het plaatsen van een Silastic bandje rondom het kopje van de stijgbeugel alsook een techniek om de hamer te reloceren naar het achterboven kwadrant van het trommelvlies. De postoperatieve lucht-beengeleidings component, berekend voor vier frequenties, bleek 13.1 dB te zijn in de PORP groep, terwijl dit 8.9 dB was voor de TORP groep. De gemiddelde duur van opvolging was 21,7 maanden met een spreiding van 3 tot 121,8 maanden.

In **hoofdstuk 9** worden de eerste resultaten beschreven zoals die bereikt werden met een nieuwe hamervervangende prothese in combinatie met een TORP voor oren waarin zowel de hamer als de stijgbeugel bovenbouw ontbraken. Het betreft een serie van 92 oren bij 90 patiënten geopereerd tussen september 1994 en maart 2012.

De serie bestaat uit 2 subgroepen waarvan de resultaten worden vergeleken. De eerste subgroep bestaat uit 34 oren, geopereerd tussen september 1992 en april 2009. Hierbij werd een TORP geplaatst tussen het trommelvlies en de beweeglijke stijgbeugelvoetplaat (TM-to-footplate assembly).

De tweede subgroep telt 58 oren, geopereerd tussen januari 2000 en maart 2012. Bij deze oren werd de nieuw ontworpen hamervervangende prothese (MRP-to-footplate assembly) gebruikt, waarbij een TORP de hamerprothese verbindt met de mobiele voetplaat.

Al deze ooroperaties betroffen revisie tympanoplastieken met als voorgeschiedenis een chronische otitis media (CSOM) zonder een actief cholesteatoom.

Voor groep 1 bleek er postoperatief nog een lucht-beengeleidings component te bestaan ≥ 20 dB bij 14 van de 27 oren (52%), terwijl dit voor groep 2 slechts het geval was bij 9 van de 43 oren (21%). Deze gehoorresultaten met een 3 maanden termijn opvolging worden voor beide groepen in een tabel getoond. De resultaten moeten nu bevestigd worden in gerandomiseerde prospectieve trials om de effectiviteit van deze nieuwe chirurgische techniek voor een totale reconstructie van de gehoorbeenketen te kunnen beoordelen en ze te kunnen vergelijken met de huidig toegepaste standaard technieken voor deze indicatie. Daarbij verdient het de voorkeur dat deze chirurgische trials worden uitgevoerd door oorchirurgen in andere tertiaire oorheelkundige centra.

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CURRICULUM VITEA

Robert Vincent was born on October 28, 1960 in Chateaubriant, France.

He passed the french baccalaureate in 1978.

From 1978 until 1986 he studied medicine at Montpellier University, France. He completed his internship and residency training program on ear, nose, and throat (ENT) surgery at the University Hospital in Nancy (France) under the supervision of Professor Michel Wayoff.

He passed his medical thesis on October 24, 1990 and became ENT specialist on October 31, 1990.

He joined the Otology group of the Causse Ear Clinic in Colombiers, France on January 1991. Since then he is otologist surgeon with special interest on stapes surgery and ossicular reconstruction of the middle ear. He has performed more than 7300 operations for otosclerosis, more than 4000 tympanoplasties and more than 260 operations for congenital malformation of the ossicular chain.

He became Honorary Fellow of the Royal College of Surgeon of Edinburgh (UK) on September 2005 and Honorary Member of the Indian Society of Otology on November 2006.

He is member of several ENT societies such as the American Academy of Otolaryngology-Head and Neck Surgery (AAO-HNS), the European Academy of Otology & Neuro-Otology (EAONO), the Otosclerosis Study Group, the Prosper Meniere Society, the Mediterraneen Society of Otology and the Politzer Society. Since 2005 he is the General Secretary and co-founder of the Live International Otolaryngology Network (LION). On October 4 2009 he has been awarded by the Board of Directors of the AAO-HNS a certificate of honor for exceptional services in the scientific program continuing education courses and instructional courses of this Society.

He is on the editorial board of several scientific journals such as the Otology & Neurotology Journal, the Journal of the Mediterraneen Society of Otology, and also serves as a reviewer for a number of professional publications.

Since 21 years he runs a annual advanced otology course in June at the Causse Ear Clinic where delegates from all parts of the world are participating every year. He is a yearly invited guest speaker and guest surgeon in many countries worldwide and has designed several middle ear prostheses and published new techniques in ossicular reconstruction.

He is the father of 3 children, Julie, Marion and Romain.

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