Ladies and Gentlemen,
Dear Colleagues,

It is my honour and pleasure to invite you to acquaint yourselves with the abstracts of speeches presented at the 4th International Symposium on Otosclerosis and Stapes Surgery, which this year takes place in the beautiful city of Cracow.

After such big scientific events held in Poland and co-organized by the Institute of Physiology and Pathology of Hearing as IX European Symposium Paediatric Cochlear Implant (2009), X Congress of the European Federation of Audiology Society (2011), XXV International Evoked Response Audiometry Study Group Biennial Symposium (2017), XII International Tinnitus Seminar and I World Tinnitus Congress (2017) this is another unique meeting which, I hope, will result in reaching valuable conclusions that will be successfully applied in clinical practice.

The scientific program of the Symposium focuses on problems of the middle ear, seeking to exchange and propagate knowledge of the latest developments concerning otosclerosis and stapes surgery. Otosclerosis is one of the most common causes of progressive deafness in young adults. We know that hearing loss generally begins between the ages of 10 and 30, and, if untreated, may eventually result in total deafness. Surgical intervention is a common and highly effective treatment, and its success relies very much on the surgeon's skill and experience.

Organization of the 2018 Symposium in Cracow is an occasion to commemorate the works of Prof. Jan Miodoński (1902–63), a Polish otolaryngologist who was professor at Jagiellonian University, a pioneer of audiometry and ear surgery, creator of Polish neurootology, and inventor of the tympanoplasty procedure.

My hope is that the invited guests and speakers will bring valuable contributions to this meeting. I am sure that the 4th International Symposia on Otosclerosis and Stapes Surgery will become an important event in the history of otolaryngology and will bear fruit in terms of a growth in expertise which will in turn maximise our success rate and benefit our patients.

I look forward to an interesting and productive meeting.

Sincerely yours,

Prof. Henryk Skarżyński, MD, PhD, dr h.c. (multi)
President of the 4th International Symposium
on Otosclerosis and Stapes Surgery

Dear Guests, Ladies and Gentlemen,
Colleagues and Friends, Fellow Members,

It is a great pleasure and honor to welcome you to the 4th International Symposium on Otosclerosis and Stapes Surgery.

We are here, in this distinguished group of specialists, to spend these three upcoming days on a special scientific and cultural event. On the scientific program of the symposium you will find main lectures, round table discussions, and workshops covering a wide range of topics in the fields of middle ear problems, otosclerosis and stapes surgery. I am convinced that this meeting will be a perfect forum for exchanging our knowledge and experience in the area.

The Symposium is the fourth in a series begun in 2004 in Saas Fe, Switzerland, and followed by a Symposium in Biarritz, France, in 2008 and then in Siófok, Hungary, in 2014. The first two Symposia were organized under the auspices of the Politzer Society; since 2014, it has been an independent Symposium.

The symposium takes place in parallel with the conference Guidelines in the otology of the Society of Polish Otolaryngologists, Phoniatricians and Audiologists. I have the pleasure to co-organize such an important scientific event for the third time. This is a continuation of the meetings started two years ago, organized in Krynica Zdrój and in Zakopane under the patronage of the National Consultant in the Field of Otorhinolaryngology. Both, first and second edition gathered over 500 participants.

I believe that the 4th International Symposium on Otosclerosis and Stapes Surgery will meet your expectations, and prove to be a valuable scientific event as well as a very pleasant social and cultural experience.

Enjoy your stay in Cracow,

Ass. Prof. Piotr Henryk Skarżyński
M.D., Ph.D., MSc., ENT Specialist
Scientific Secretary
of the 4th International Symposium
on Otosclerosis and Stapes Surgery

4TH INTERNATIONAL SYMPOSIUM ON OTOSCLEROSIS & STAPES SURGERY

CRACOW, POLAND, 5-7 APRIL 2018

Opening Session

Counselling patient surgery or hearing aids.

Fraysse B., Molinier C.E.

Service ORL, Hôpital Pierre-Paul Riquet, Toulouse, France

The author will describe the information given to the patients between surgery or hearing aids based on a prospective longitudinal study of 30 patients comparing audiological outcomes with hearing aid then stapedotomy at 2 months.

Evaluation include pure tone audiogram speech discrimination sound localization binaural hearing GHSI and music perception.

Is There a Role for Active Middle Ear Prostheses in Management of Otosclerosis?

Jenkins H.A., Lupo E., Greene N., Tollin D.

University of Colorado School of Medicine, Aurora, Colorado, USA

Aim: Stapes surgery for conductive hearing loss secondary to otosclerotic fixation has been a mainstay in our treatment for many decades, since its rediscovery by John Shea Jr. in 1956. Refinements have led to less invasive fenestrations, self-crimping prostheses and introduction of lasers to minimize trauma to the inner ear. Even with these innovations, a significant number of patients have persistent or delayed conductive loss due to complications. Repeated attempts to improve this situation surgically with revision stapes procedures often are unproductive, depending on the middle ear changes with surgery. Much of the work over the last decade in our laboratory has been to study the potential for use of active middle ear prostheses. This presentation will discuss our laboratory experience in animal and human models of simulated otosclerosis and review the current state of surgical procedures.

Material and Methods: Artificial otosclerosis was simulated in the chinchilla with acrylic glue applied to the oval window area. Animals were stimulated acoustically, followed by mechanical stimulation of the round window in fixation and non-fixation states by an active middle ear prosthesis. Cochlear microphonics, compound action potential, and ABR were measured. Human cadaveric studies included laser Doppler measures of round window velocity in response to active stimulation of the stapes in fixation and following fenestration.

Results: Endocochlear potentials and ABR recordings reflected essentially identical responses to acoustical and round window mechanical stimulation. Fixation of the stapes showed similar pattern, though reduction of amplitude of response. Stapes fixation resulted in a 4-13 dB shift in cochlear microphonics and 2-24 dB in compound action potential, being frequency dependent. These losses could easily be compensated by the magnitude of the stimulator output. Similar results were seen in measuring the round window velocity in cadaveric specimen with response magnitude related to several factors that improved coupling of the prostheses. Direct drive of the vestibule via a stapedotomy produced excellent responses of the round window, equivalent to magnitudes associated with normal inner ear stimulation.

Conclusion: Direct mechanical stimulation of the round window with an active middle ear prosthesis is comparable to acoustic stimulation. Effective stimulation can be produced in a variety of stimulations and pathological states. In situations in which it is difficult to reconstitute the normal conductive mechanism, active middle ear prostheses offer an acceptable alternative.

Differential Diagnosis of Conductive and Mixed Hearing Loss. A question of Imaging.

Offeciers E.[1] , Casselman J.W.[2,3], De Foer B.[2], Zarowski A.[1], van Dinther J.[1], Somers Th.[1]

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The lecture will illustrate how state-of-the-art imaging improves the pre-operative diagnostic work-up in patients presenting with conductive or mixed hearing loss with an intact tympanic membrane, which is the typical presentation status of otosclerosis patients. The imaging work-up helps to establish the differential diagnosis with other causes, helps to avoid surgical complications or surprises as well as unwarranted interventions, and thus ultimately affects the therapeutic outcome.

The lower radiation burden of Conebeam CT and its improved spatial resolution (150 μm versus 500 μm) as compared to conventional multi-detector CT, has led the

authors to routinely apply it in the pre-operative work-up of patients presenting with conductive or mixed hearing loss with an intact tympanic membrane. CBCT plays an important role in pre-operatively confirming the presumed diagnosis, in identifying contra-indications to functional surgery and in informing the surgeon about potential difficulties and complications during surgery, thus allowing the clinician to counsel the patient more comprehensively and to better plan and prepare the operation.

Presenting illustrative clinical examples, the authors will treat in detail a list of ambiguities, generated by the traditional diagnostic methods at our disposal (personal and family history, otoscopy, audiometry) and solved by pre-operative diagnostic imaging. Some examples in point are: atypical history of hearing loss, suspected congenital hearing loss, suspect otoscopic image, asymmetric BC thresholds, profound mixed loss, suspect tympanometry and cases for revision stapes surgery. CBCT helps to document the diagnosis of the following pathologies: otosclerotic and other stapes fixations, tympanosclerosis, post-traumatic and post-inflammatory ossicular lesions, minor middle ear dysplasia's, labyrinth dysplasia's and SCCD. MRI can be used to exclude or confirm a schwannoma, a suspected congenital cholesteatoma or labyrinth dysplasia.

Pre-operative Conebeam CT imaging is very useful for the honest and realistic pre-operative counselling of the patient regarding the short and long term outcome of the surgical procedure. As such its use has medicolegal implications.

The aim of this lecture is to provide the clinician with the state-of-the-art information that allows him to ask his radiologist the pertinent questions, thus fostering a closer collaboration between the clinician and the radiologist.

Professor Jan Miodoński - an unappreciated Polish pioneer in the history of otolaryngology.

Szaleniec J.

Department of Otolaryngology, Jagiellonian University Medical College, Krakow, Poland

Professor Jan Miodoński is considered the greatest Polish otolaryngologist of the 20th century. His achievements and breakthrough discoveries are so numerous that it is almost impossible to believe that they were accomplished by a single person. He was the first otosurgeon in the world to perform tympanoplasty, the first otoneurologist to describe the four vestibular caloric tests, the first head and neck oncologist to introduce the subtotal partial laryngectomy. Why are these pioneer inventions attributed to other scientists who published their works years after Miodoński? Why is the name of Jan Miodoński not widely recognized by the international medical community?

To understand this paradox it is necessary to take a look at the historical context of his life and work.He started his medical career directly after the World War I. At this time Poland was struggling with many problems as it regained independence after 123 years of absence from the maps of Europe. Jan Miodoński worked with vigor and talent to create a modern laryngological clinic. Yet before the task was finished, another World War ruined his plans. Miodoński was arrested in a Nazi operation called the 'Intelligenzaktion' which was a plan to eradicate the Polish intellectual elite. He spent more than a year in a concentration camp. When the war was over, Poland fell under the Soviet influence. Behind the Iron Curtain the scientific ideas developed to some extent independently from the Western countries. It was not uncommon that scientists were forbidden to participate in international congresses and conferences or discouraged to publish in languages other than Polish or Russian. As a result Jan Miodoński lost a chance to present his discoveries abroad or claim his priority when the inventions were later introduced by other researchers.

Miodoński's areas of interest were very vast. He introduced novel ideas in the physiopathology of hearing, functional surgery of the middle ear, vestibular testing, treatment of otogenic cerebellar abscesses and larynx cancer surgery. He was both a brilliant scientist and an agile surgeon. The reader studying his works is impressed by the detailed descriptions of meticulous examination of the patient and thorough logical analysis of clinical observations. His educational texts were clear and comprehensive. But above all he was a man of high integrity. He claimed, that a good physician must be a good man - and to this principle he struggled to remain true in his whole life.

Thursday, 5th April

Round Table:

Otosclerosis and Active Hearing Implants

Introduction Active Middle Ear Implants (aMEI's) and otosclerosis.

Tange R.A.

Utrecht / Naarden, the Netherlands

Hearing impaired people can be treated with a multitude of different devices and surgical interventions. In otosclerosis surgical treatment is in most of the cases the treatment of choice. In a small number of otosclerostic patient this treatment is not sufficient. In severe profound mixed hearing loss due to advanced cochlear otosclerosis classic stapes surgery will not improve hearing without an extra device. Implantable hearing devices have been introduced to increase amplification options for those who have limited success or are unable to wear traditional hearing aids. Active implantable devices include bone conduction devices with percutaneous or subcutaneous components and middle ear implants providing auditory stimulation through manipulation of the existing or remnant ossicular chain. This paper introduces these aMEI's and the possible application in otosclerosis treatment.

Management of Advanced Otosclerosis: Option of Stapedotomy vs Cochlear Implantation.

Kameswaran M.

Department of Otolaryngology, Madras ENT Research Foundation, Chennai, India

Background: Advanced otosclerosis affects approximately 10% of patients with otosclerosis. Ossification of the cochlea may result in sensorineural or mixed hearing loss. Hearing aids, stapedotomy and cochlear implants are options for managing hearing loss associated with advanced otosclerosis.

Methods: A retrospective study of 153 patients with advanced otosclerosis was done in a tertiary ENT centre. 110 patients with advanced otosclerosis underwent stapedotomy and 43 patients with advanced otosclerosis underwent cochlear implantation (CI) from 1997 till date. The aim was to study the indications, selection criteria, as well as surgical issues of stapedotomy and cochlear implantation in patients with advanced otosclerosis.

Results: Stapedotomy followed by use of hearing aid was found to give good outcomes in advanced otosclerosis. Patients with advanced otosclerosis who had poor pre-operative speech discrimination underwent CI. The outcomes of cochlear implantation were found to be satisfactory. Facial nerve stimulation was seen in 5 patients who underwent CI; however this was successfully managed by reprogramming or switching off the concerned electrodes.

Conclusion: The management of advanced otosclerosis may prove to be a challenge to the otologist. The criteria for deciding on stapedotomy Vs CI has to be stringent for optimal outcomes. Several intra-operative issues, especially cochlear ossification may be encountered and the surgeon should anticipate these pre-operatively.

Hearing outcomes in cochlear implanted patients with far-advanced otosclerosis.

Mosnier I.[1,2], Cyna-Gorse F.[3], Tuset M-P.[2], Bernardeschi D.[1,2], Sterkers O.[1,2]

[1] Sorbonne Université, Inserm, Unité de Réhabilitation chirurgicale mini-invasive robotisée de l'audition, Paris, France
[2] AP-HP, GHU Pitié-Salpêtrière, Service ORL, Otologie, Implants auditifs et chirurgie de la base du crâne, Paris, France
[3] Radiologue, Paris, France

Far-advanced otosclerosis (FAO) refers to patients with a clinical diagnosis of otosclerosis, with an air-conduction threshold greater than or equal to 85 dB and a poor speech intelligibility with well-fitted hearing aids. Cochlear implanted patients with FAO may have various location and extension of otosclerosis that can lead to surgical difficulties and post-operative facial stimulation and influence the hearing outcomes.

We conducted a retrospective chart review of patients with FAO who underwent cochlear implantation in our department. Between 1991 and 2017, 100 adult patients with FAO underwent cochlear implantation (115 ears), which represent 5% of the cochlear implanted population during this period. The mean age at implantation was 61 ± 18 years. Pre-operative audiometric thresholds, auditory outcomes, surgical difficulties, complications and management in fitting strategies were compiled and analyzed as function of the otosclerosis extension on CT scan.

Pathogenesis and Diagnosis

Keynote Lecture

Prolonged hearing screening among workers in a Bulgarian shipbuilding plant.

Milkov M., Nedev P.

"Prof. Paraskev Stoyanov" Medical University of Varna, Bulgaria

Hearing reduction and hearing loss are common among the industrial workers exposed to continuous and intensive occupational noise, although they are completely preventable. Modern hearing protection devices are not efficient enough. Preventive otorhinolaryngologic examinations enable the early diagnosis and timely treatment of this severe pathology.

Our purpose is to analyze the dynamics of hearing reduction and loss of dockyard workers by comprehensive screening examinations as a first step in a broad prevention programme.

A total of 270 and 256 male workers from Varna dockyard are examined by otorhinolaryngologists in 2002 and 2009, respectively. Because of subsequent enterprise closure after 2009, the number of the patients followed-up until present fell down to 25 only. The workers belong to the following groups: helpingists (25%), pipe fitters (23%), ship-gear fitters and turners (22% each), moulders and woodworkers (3% each), and millers (2%). Otological screening consists in comprehensive clinical investigations such as otoscopy with microscopy, tympanometry, digital tonal threshold and over-threshold audiometry, otoacoustic emissions, videonystagmography, video head impulse test, routine blood tests, and, if necessary, consulting examinations by neurologists, x-ray, magnetic resonance imaging, and computer-assisted tomography.

In 2002, at 21-50 dB, the workers examined at 500 Hz are statistically significantly less than those at 1000 Hz and 4000 Hz ($p<0,001$) and those at 2000 Hz ($p<0,05$). At 61-70 dB, the workers examined at 500 Hz are statistically significantly less than those at 2000 Hz and 4000 Hz ($p<0,001$) while at 51-60 dB, the workers examined at 500 Hz and at 1000 Hz are statistically significantly less than those at 2000 Hz ($p<0,001$). In 2009, at 21-50 dB, the workers examined at 500 Hz are statistically significantly less than those at 4000 Hz ($p<0,001$) while at 61-70 dB, the workers examined at 500 Hz are statistically significantly less than those at 4000 Hz ($p<0,01$) and at 2000 Hz ($p<0,001$). Our recent examinations reveal otosclerosis in seven of these 25 patients. Acoustic neuritis is diagnosed in 18 patients and 22 workers complain of tinnitus. All the patients experience vestibular disorders during certain periods of time. Surgery is carried out in five otosclerosis patients.

There is a substantial improvement of the objective and subjective status in four of them. In three patients, additionally, ossicular prosthesis is implanted upon their request. It remains not clarified if otosclerosis has already occurred long ago, or not. Further research is needed to elucidate the relationship between labyrinth-related otosclerosis, on the one hand, and long-lasting exposure to noise and vibrations, on the other hand.

The conclusion is drawn that prolonged and regular otological screening could timely identify initial hearing disorders among the workers in unfavourable occupational environment.

An attempt to dissect the genetic background of familial otosclerosis.

Pollak A., Skarżyński H., Ołdak M.

World Hearing Center, Institute of Physiology and Pathology of Hearing, Warsaw/ Kajetany, Poland

The pathophysiology of otosclerosis is complex and contains both genetic and environmental factors. Genetic analysis conducted in patients with familial form of otosclerosis with high probability may bring new insight into the molecular mechanisms involved in the disease pathogenesis. Although there are about 10 loci reported to date linked with the otosclerosis, only two possibly causative genes (SERPINF1 and FOXL1) have been identified. Thus, in such cases the optimal solution is to conduct a wide research, which has become possible since the introduction of next generation sequencing (NGS). For this purpose we decided to perform the clinical exome sequencing, which is a universal and powerful discovery tool to reveal the genetic causes of diseases. The dedicated bioinformatics pathway combined with thorough experts analysis was applied to data analysis.

Genetic association analysis in Polish patients with otosclerosis.

Oziębło D., Adamiok A., Domagała S., Skarżyński H., Ołdak M.

World Hearing Center, Institute of Physiology and Pathology of Hearing, Warsaw/ Kajetany, Poland

Otosclerosis (OTSC) is one of the most common causes of adult-onset hearing loss in the Caucasian population. Development of OTSC is associated with abnormal bone remodeling in the region of otic capsule and stapes footplate. Etiology of OTSC is complex and there are a number of genetic variants reported to be associated with OTSC susceptibility. However, their frequencies have not been analyzed in the Polish population. The purpose of our study was to investigate the genetic variants most strongly associated with OTSC in Polish patients. DNA was isolated from blood samples and selected genetic variants in TGFB1 and RELN genes were genotyped in clinically diagnosed OTSC patients and control group using Custom TaqMan SNP Genotyping Assays and a real time PCR system. Allele and genotype frequencies were compared between the groups by Chi square test and odds ratio (OR) with 95% confidence intervals (95% CI) were calculated to estimate risk. Here we will present frequencies of the investigated variants and the strength of association between each of them and OTSC. Our data will verify the role of the selected genetic variants in OTSC susceptibility in Polish patients.

Genetic factors in the development of otosclerosis.

Ołdak M., Oziębło D., Pollak A., Lechowicz U., Adamiok A., Skarżyński H.

Department of Genetics, Institute of Physiology and Pathology of Hearing, Warsaw/Kajetany, Poland

Regardless of intensive research on the pathogenesis of otosclerosis, its background still remains poorly understood. The aim of the presentation is to give a comprehensive overview of the current state of knowledge in the field of genetics of otosclerosis. The intriguing correlation between geographic descent and prevalence of otosclerosis has been noted, indicating that this disease is significantly more frequent among Caucasians than Japanese and South Americans. Otosclerosis is extremely rare among Afro-Americans, American Indians and oriental populations. Taking into account interpopulation differences

in otosclerosis prevalence together with a positive family history of this diseased in almost 60% of patients, it is highly expected that genetic factors play a significant role in the pathogenesis of otosclerosis. Familial studies revealed several genetic loci linked with otosclerosis but up to now only one potentially causative gene (SERPINF1) has been identified. The etiology of otosclerosis seems complex involving both genetic and environmental components.

Otosclerosis or otosclerosis-like stapes fixation in patient with mixed hearing loss and COCH pathogenic variant?

Oziębło D.[1], Adamiok A.[1], Tacikowska G.[1], Furmanek M.[1], Skarżyński H.[1], Ołdak M.[1], Skarżyński P.H.[1,2,3]

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[3] *Institute of Sensory Organs, Kajetany, Poland*

The COCH gene encoding cochlin is highly expressed in the inner ear but the exact physiological function of the protein still remains unknown. Pathogenic variants in COCH cause autosomal dominant hearing loss with possible vestibular involvement. Interestingly, pathogenic variants in COCH gene have been also identified with superior semicircular canal dehiscence (SSCD) which introduces a third window into the inner ear and may mimic otosclerotic hearing loss. Using clinical exome sequencing we have identified a COCH pathogenic variant p.Ile374Thr causative for hearing loss in a patient diagnosed with bilateral sensorineural hearing loss. As the patient also suffered from unilateral otosclerosis we have verified whether the conductive hearing loss does not result from SSCD. Based on audiological and vestibular examination and reanalysis of the temporal bone imaging we have excluded SSCD as a possible cause of conductive hearing loss in a patient. Our results do not support the role of COCH p.Ile374Thr in SSCD development but considering different conditions that may mimic otosclerosis-like stapes fixation genetic testing is strongly recommended in diagnosing patients with hearing loss.

Stapes fixation in enlarged vestibular aqueduct due to SLC26A4 pathogenic variants.

Lechowicz U., Pollak A., Tacikowska G., Skarżyński H., Ołdak M.

World Hearing Center, Institute of Physiology and Pathology of Hearing, Warsaw/ Kajetany, Poland

Aim: Large vestibular aqueduct (LVAS) is diagnosed when diameter of the vestibular canal is wider than 1.5 mm at midpoint between the vestibule and its opening in the posterior fossa. LVAS is supposed to be the most prevalent reason of hearing loss in patients with congenital malformations of the temporal bone. Air-bone gap in patients with LVAS can be mistakenly taken as a disorder of aeration of the inner ear or stiffening of the ossicular chain, having an analogous clinical presentation to otosclerosis. However, diminished mobility of the stapes in such cases is induced by excessive pressure of the perilymph. Genetic and environmental factors might be involved in both conditions, therefore the genetic analysis might be a helpful tool in the differential diagnosis.

Material and Methods: An eight-year old female patient with mixed type hearing loss in both ears and repeated episodes of sudden deafness was consulted at the Institute of Physiology and Pathology of Hearing. Pure-tone audiometry, magnetic resonance imaging and a temporal bone CT-scan were performed. Additionally, whole exome sequencing (WES) was conducted to study the molecular background of hearing loss.

Results: Radiological examinations revealed inner ear malformation corresponding to incomplete partition type 2. Results of genetic testing showed two heterozygous pathogenic variants p.Glu29Gln and p.Leu117Phe in the SLC26A4 gene. **Conclusions:** Clinical picture of LVAS may cause diagnostic and therapeutic difficulties. In children with mixed hearing loss and repeated sudden worsening of hearing, we strongly recommend to look for pathogenic variants in the SLC26A4 gene and complete the diagnosis by radiological examinations.

This study was supported by National Science Centre grants: 2011/03/D/NZ5/05592 and 2012/05/N/NZ5/02629.

Stapes fixation in female carriers of POU3F4 pathogenic variants mimicking otosclerosis.

Pollak A., Lechowicz U., Tacikowska G., Niedziałek I., Furmanek M., Skarżyński H., Ołdak M.

World Hearing Center, Institute of Physiology and Pathology of Hearing, Warsaw/ Kajetany, Poland

Otosclerosis is defined as an abnormal process of bone resorption of the labyrinthine capsule followed by deposition of novel, immature sclerotic bone localized around the oval window. As a result of this process the stapes footplate fixation occurs causing conductive hearing loss. During the time the sclerotic bone has a tendency to increase in size and depth. In most cases patients suffering from otosclerosis undergo surgery consisting of replacing all or a part of the affected stapes with a prosthetic device which restores hearing abilities. The clinical diagnosis of otosclerosis is typically based on the (i) analysis of pure tone audiometry, in particular the presence of air-bone gap, (ii) no evidence of middle ear inflammation and (iii) regular picture of tympanic membrane. However, there are some rare cases when this common clinical picture is not linked with otoslerosis. Here, we present phenotypic features of female carriers of POU3F4 pathogenic variants, in whom based on molecular tests and radiological findings mixed type of hearing loss with inner ear malformations was established in opposite to the suspicion of otoslerosis.

This study was supported by National Science Centre grant 2011/03/D/NZ5/05592.

The pre – and postoperative assessment of tinnitus distress severity among patients qualified for stapedotomy – preliminary findings.

Skarżyński P.H.[1,2,3], Świerniak W.[1], Dziendziel B.[1], Bienkowska K.[1], Gos E.[1], Rajchel J. J.[1], Skarżyński H.[1]

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Aim: Stapes surgery is widely used in the treatment of otosclerosis in order to improve hearing. Furthermore, tinnitus is a frequent symptom related to hearing impairment. The aim of the study was to the pre– and postoperative assessment of severity tinnitus distress among patients qualified for stapedotomy.

Material: The prospective study included 161 patients with otosclerosis. The sample comprised 123 females and 38 males, with age on average M=49; SD=11.5 years. All patients underwent stapedotomy in the World Hearing Center, Institute of Physiology and Pathology of Hearing in Poland.

Methods: All patients were asked to complete the Tinnitus and Hearing Survey questionnaire (THS-POL) to differentiate the problems with hearing loss and tinnitus. The second tool was Tinnitus Functional Index (TFI-Pl) used for evaluation of changes in tinnitus severity after stapedotomy. This tool measures the impact on 8 domains such as: the intrusiveness of tinnitus, the degree of control the patient has over the disease, cognitive interference, sleep disturbance, auditory issues, relaxation issues, quality of life, and emotional distress.

Results: Before stapedotomy, 108 patients (67.1%) experienced unilateral chronic tinnitus in the ear qualified for surgery or bilaterally. Using guidelines created by Meikle et al. (2012) for group TFI scoring, before surgery the results of the TFI-Pl questionnaire amounted to an average of M=31.2; SD = 21.4 points, indicating a moderate problem with tinnitus. In early post-operative follow-up, the TFI-Pl total scores decreased to 22.1 points (SD=22.8).

Conclusion: Based on the results of the study, it was observed that more than half of patients with otosclerosis experience tinnitus, mostly of moderate problem. It is stated that after a stapedotomy, patients with otosclerosis can not only get better hearing but also experience reduced severity of tinnitus.

Evaluating of the prevalence and severity of tinnitus in otosclerosis patients.

Skarżyński P.H.[1,2,3], Dziendziel B.[1], Świerniak W.[1], Bienkowska K.[1], Gos E.[1], Kutyba J.[1], Skarżyński H.[1]

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Aim: Subjective tinnitus is, beside the progressive hearing loss, one of the primary symptoms of the development of otosclerosis. The aim of the study was to evaluate the prevalence and severity of preoperative tinnitus among a group of consecutive adult patients with otosclerosis using standardized research tools.

Material: The prospective study comprised 462 patients with clinically confirmed otosclerosis (326 women, 136 men), whose age was on average M=48; SD=11.5 years. All of them were qualified to stapedotomy in the World Hearing Center, Institute of Physiology and Pathology of Hearing in Poland between April – October 2017. The main inclusion criteria were: age ≥ 18 years, suspicion of otosclerosis and no previous stapes surgery in the ear eligible for surgery. We excluded patients based on intraoperative image excluding otosclerosis or with another surgical procedure than stapedotomy.

Methods: The Tinnitus and Hearing Survey questionnaire (THS-POL) was used to assess differentiating the problems with hearing loss and tinnitus. The Tinnitus Functional Index (TFI-Pl) questionnaire was used to assess the preoperative severity of tinnitus and impact on 8 domains such as: the intrusiveness of tinnitus, the degree of control the patient has over the disease, cognitive interference, sleep disturbance, auditory issues, relaxation issues, quality of life, and emotional distress. In addition, during the preoperative interview information on the age, sex of participants, the duration of tinnitus and hearing loss were collected.

Results: Among 462 patient diagnosed with otosclerosis, 298 (64.5%) reported chronic tinnitus. In 136 patients (45.6%) tinnitus occurred unilaterally, only in the ear qualified for surgery, in the other patients – on both sides. The results obtained in this group using a questionnaire THS – POL show that for nearly 22% patient, tinnitus was a greater or similar problem as hearing loss. The results of the TFI-Pl questionnaire amounted to an average of M=32.1; SD=21.4 points, indicating a moderate problem of tinnitus before surgery.

Conclusion: The number of women in the study was almost twice as high as that of men, which confirms the higher prevalence of otosclerosis in this group of patients. The results obtained by means of validated questionnaires indicate that over half of patients qualified for surgical treatment of otosclerosis experience tinnitus, mostly of moderate severity.

Genetic background of osteogenesis imperfecta in Polish patients.

Adamiok A., Osińska K., Oziębło D., Skarżyński P.H., Ołdak M., Skarżyński H.

World Hearing Center, Institute of Physiology and Pathology of Hearing, Warsaw/ Kajetany, Poland

Osteogenesis imperfecta (OI) caused by dominant autosomal mutations is a phenotypically and genetically heterogenous disease characterized by bone fragility, skeletal deformities and ossicles malformations. Clinical presentation and family history are two main diagnostic parameters in OI, although genetic test should be used to confirm the diagnosis. To date, only few genes were found to be causative for OI. In 85% of individuals OI results from mutations in genes encoding type I collagen (COL1A1, COL1A2). Collagen is known to be the main component of the extracellular matrix of bone and skin. Since our knowledge on OI genetic causes is still so obscure it is important to better understand the etiology of this disease. Our aim is to identify the genetic background of hearing loss in OI patients.

In the study genomic DNA of clinically diagnosed OI patients was used to perform clinical exome by next-generation sequencing. Segregation analysis of detected variants in OI families was performed by Sanger sequencing. We will present genetic data confirming the diagnosis of OI in the investigated families. We expect that in the future genetic testing for OI will be routinely used to identify OI patients and offer them appropriate treatment.

Lecture Session I

Keynote Lecture

Otosclerosis: from manual to robot-assisted surgery.

Sterkers O., Bernardeschi D., Ferrary E., Nguyen Y.

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Otosclerosis surgery is performed through a transcanal approach and requires long and thin instruments with sub-millimetric precision and precise amplitude of motion. The functional outcomes and complications of otosclerosis surgery are dependent on the experience of the surgeon. Thus, any technological assistance that enhances the surgeon's dexterity and reduces the learning curve could yield an even safer surgical procedure. One of the options is to use robotic assistance to achieve this goal. This lecture will first address the management of difficult surgical cases. The RobOtol system, a tele-operated device designed for middle and inner ear surgery will then be described and how we envision its use for otosclerosis surgery. At last, innovative methods for surgery teaching using modified artificial bones and a virtual simulator of the otosclerosis procedure will be presented.

Keynote Lecture

Same or one-day surgery for stapes surgery: clinico-economical issues.

Barbara M., Covelli E., Bianchi A., Talamonti R., Margani V., Monini S.

ENT Clinic, NESMOS Department, Sapienza, Rome, Italy

Aim: Otosclerosis is characterised by abnormal sponge-like bone growth in the middle ear causing progressive hearing loss. It mainly affects the ossicular chain and it can be treated surgically by removing (part of) the stapes and replacing it with a prosthesis—stapedotomy and stapedectomy, respectively. Increasingly, stapes surgery is performed as a One day-case procedure. The main reason of choice towards One day-case surgery is the result of economic considerations. Nevertheless, it is also an explicit patient request and leads to shorter waiting times for surgery, reduced risk of infection and most of all it positively influences the patient's quality of life as a result of rapid discharge and rehabilitation. According to the latest guidelines of day surgery procedures issued by National Healthcare System, stapedotomy needs a maximum of two days of hospitalisation. The primary objective of this study is to evaluate effectiveness of one day-case stapes surgery compared to in hospital stapes surgery in terms of days of hospital stay, complications and costs.

Material and Methods: 327 patients (116 male and 211 female; range 20-75 years, mean age 48.3) who underwent surgery for otosclerosis between 2008 and 2018 were retrospectively studied. The inclusion criterion was a diagnosis of otosclerosis based on a clinical history of progressive hearing loss, normal otoscopic findings, an audiogram showing a mean conductive hearing loss greater than 20 dB HL in the range of 0.5–4 kHz, the absence of cochleostapedial reflexes and surgery findings.

We have divided the patients into two separate groups. The first one includes 160 patients who underwent stapes surgery between 2008 and January 2012 under ordinary admission. The second group is composed by 167 patients who underwent stapes surgery under One day surgery between 2012 and 2018. Of these two groups, we considered mean days of hospitalisation, complications as vertigo, nystagmus, nausea, vomiting, hearing loss and hospitalisation costs according to the Regional Healthcare Service.

Results: In the ordinary inpatient group mean day of stay was 6 days, while in the one day-case patient group it was 2,2. Most patients of both groups did not show any complications.

Conclusions: According to our opinion, One day Surgery is the best choice for stapes surgery in terms of costs and improvement in quality of life. Indeed the impact of long

hospitalisation not influences the incidence of post-operative complications.

Keynote Lecture

Stapedotomy – How I Do It!

Vijayendra H.

Vijaya ENT Care Centre, Bangalore, India

Stapes surgery is one of the most delicate surgery in ear surgery. Utmost importance should be given to this surgery as middle ear is sterile in these cases and sterility of middle ear should be maintained. Most of my cases, I operate under local anaesthesia. Great advantage being, hearing can be tested on table. If piston length is more, then giddiness and nystagmus can also be checked.

Using proper and quality instruments is key in stapes surgery. One should always have good set of all instruments. Via transcanal approach, I create tympanomeatal flap. First secure tympanomeatal flap anteriorly using wet saline soaked gelfoam. Because as a beginner one will take more time to perform procedure. By the time procedure gets over, flap which has been raised in beginning will shrink because of dryness and then will be difficult to approximate. To obtain exposure, I curette posterosuperior canal wall. For adequate exposure and easy insertion of piston superiorly facial canal and posteriorly pyramidalis process should be visible. Chorda tympani nerve if coming in way then should be decompressed and secured away from working area so that it won't come in way during the procedure. I always first make controlled fenestra of 0.3mm in posterior half of the footplate. The advantage of making controlled fenestra is that it is very safe once contolled fenestra is made. If while widening fenestra something goes wrong with footplate then I have another instrument, 0.2mm Fisch pick with which i can easily manipulate footplate as it can be passed through 0.3mm diameter fenestra. Fenestra should be always made in posterior 1/3rd of the footplate in order to avoid injury to saccule and utricle which are placed near to anterior part of footplate as compared to posterior part. Then I drill posterior crus using 0.5mm diamond burr, disarticulate incudostapedial joint and cut stapedius tendon near pyramid. Then using 45 degree angles pick, keeping it at the neck of stapes, stapes suprastrucutre is fractured towards promontory. Fenestra gradually widened to 0.6mm using first 0.4mm and then to 0.6mm perforator. In my experience, I have seen that manual perforators are always better as surgeon can feel the pressure at the tip of his fingers and always have control over the procedure. Then using measuring rod, I measure the distance between the long process of incus and fenestra. I always use Teflon piston of 0.4mm diameter because sound conduction and ultimately hearing results will be better. I use lower magnification to insert piston as both fenestra and long process of incus should be visible in one view only. Teflon piston is inserted into fenestra, anchored to long process of incus and crimped. Surrounding of fenestra I always seal cicumferentially using post aural soft tissue harvested using non touch technique. In the end i place chorda in front of piston as additional support to the piston. After repositioning the flap, I check hearing on table.

Keynote Lecture

Nitibond - Systematic development of an optimized prosthesis.

Huber A.

University Hospital Zurich, Department of Otorhinolaryngology, Head and Neck Surgery, Zürich, Switzerland

The optimal prosthesis to use for otosclerosis surgery is still a matter of debate. It has been proposed that prostheses made of shape-memory alloy (SMA), produce better functional results with less variability and reduced risk because heat activation rather than manual crimping of the prosthesis loop forms a tighter fixation. However, risks in terms of mucoperiosteum lesions and incus necrosis are still discussed controversially. Therefore, the necessary properties for optimized stapes prosthesis were identified based on theoretical considerations, clinical experiences as well as mechanical tests. Then, a new generation of SMA stapes prosthesis was developed and tested in preclinical as well as clinical environments. A self-fixing coupling area with multiple contact points and recess spaces for the mucoperiosteum appear optimal for stapes prosthesis. A spring characteristic of the prosthesis loop allows a defined contact pressure on the incus and adaption to the geometry of different incus shapes. Optimizing the shape allows good intraoperative handling capabilities. Clinical testing revealed no adverse reactions and functional results were "non inferior" to conventional SMA prostheses. Therefore, the newly developed NiTiBOND stapes prosthesis overcomes limitations of existing prostheses. Our results suggest safety and reliability.

Keynote Lecture

Total stapedectomy. Technique and long term results.

Garcia-Ibañez E.

Clinica Garcia-Ibañez, Instituto de Otología, Barcelona, Spain

The use of total stapedectomy has been always the surgical option in our Institute for otosclerosis treatment. We present the technique, together with the short term and long term results. Also a small series of stapedotomies are compared with the total stapedectomy results.

These results and the results obtained in revisions of a big number of stapedotomies demonstrate that in our hands total stapedectomy is the technique of choice for us. However, we understand that for not experienced surgeons stapedotomy is safer for avoid inner ear damage and post-op complications.

Keynote Lecture

Surgical treatment of cochlear otosclerosis.

Diab Kh.M., Korvyakov V.S., Gamzatov K.N., Kaibov A., Kondratchikov D.S., Mikhalevich A.E., Pashchinina O.A.

Clinical Centre of Otorhinolaryngology of FMBA, Moscow, Russia

Aim: To increase effectiveness of speech and hearing rehabilitation for patients with cochlear otosclerosis accompanied by severe and profound hearing loss.

Materials and Methods: 28 patients with cochlea otosclerosis were diagnosed and underwent surgery: 16 with severe hearing loss (I group), 12 with profound hearing loss (II group). Patients from the 1st group underwent stapedoplasty according to methodology of using auto-cartilage on vein placement with the partial replacement of perilymph (antrum) to natural saline solution. This methodology allows to change electrolytic structure of inner ear's perilymph, its hydrodynamics, and also possible toxic effects of otosclerotic areas, that hypothetically could be the reason of sensorineural hearing loss occurence.

Patients from the 2nd group underwent cochlear implantation with insertion of straight electrode arrays. 8 patients from the 2nd group had undergone previous procedures of piston-type stapedoplasty, which caused profound hearing loss. During the pre-surgical examination of these patients were diagnosed enlargement of internal auditory canal and endolymphatic duct (according to CT-scan). Evaluation of audiological results was performed according to results of tone threshold audiometry (1st group) and free-field speech audiometry (2nd group) in 3 months after surgery.

Results: Patients from the 1st group showed decrease of air thresholds by 15-25 dB in average, that provided satisfactory socially-significant hearing. Full insertion of electrode array through round window was performed in all cases with the patients of 2nd group. 50% threshold of speech discrimination was reached in all patients' cases using the intensity of sound 31,5 dB in average.

Conclusions: Stapedoplasty performed in accordance with above mentioned methodology allows to obtain socially-significant hearing for patients with severe hearing loss. Cochlear implantation performed in cases of cochlear otosclerosis provides good results of hearing and speech rehabilitation under the condition of minimal trauma of inner ear.

Keynote Lecture

Cochlear Gene Therapy.

Lalwani A.K.

Department of Otolaryngology-Head & Neck Surgery, Columbia University College of Physicians and Surgeons, New York, USA

Cochlear gene therapy offers the potential of restoring hearing via either restoration of the damaged inner ear structures or protecting the delicate cells from harm. Protected by one of the hardest bones in body, the cochlea is nearly an impenetrable structure frustrating both bacteria and clinician trying to gain access to it. As a result, a means for reliable delivery of agents into the inner ear for therapeutic purposes, such as cochlear gene therapy, remains a formidable challenge. Another major issue includes the choice of delivery vector to carry the gene into the inner ear. Choices include viral and non-viral vectors that come with their own risks and benefits. The molecular machinery to control of gene expression has to be carefully chosen to restrict the expression of therapeutic genes to cells of interest (e.g. hair cells). Finally, there are safety concerns regarding gene therapy that must be addressed and overcome. With the availability of a reliable method of inner ear delivery, targeted delivery to hair cells, spiral ganglion neurons or other intra-cochlear structures, and managing safety, gene therapy could be accomplished for the treatment of variety of auditory and vestibular disorders such as sudden or progressive SNHL, Ménière's Disease, otosclerosis, and tinnitus.

Keynote Lecture

A tribute to John Shea.

Tange R.A.

Utrecht / Naarden, the Netherlands

This presentation devoted attention to the life of the discoverer of the stapedectomy. In 2015, John Joseph Shea (1924-2015) died at the age of 90 due to respiratory insufficiency. His legacy is invaluable for the current reconstructive ear surgery. Due to its progressively daring stapectomy operation (1956), the suffering of the millions of otosclerosis patients has since with substantially reduced hearing loss. Nowadays, the stapedectomy / stapedotomy with the interposition of a stapes prosthesis is the treatment of choice for otosclerosis and the procedure has become almost routine for the experienced ear surgeon. The procedure is now performed worldwide and taught to novice ear surgeons. All this thanks to the pioneering work of the pioneer John Joseph Shea. He was the founder of the current successful surgical treatment of otosclerosis.

Outcomes

Long-term functional results and Quality of Life 20 years after surgery for otosclerosis.

Sergi B., Lucidi D., Paludetti G.

Policlinico Gemelli, Catholic University of Rome, Italy

Aim: The aim of our paper is to analyze the hearing function and the quality of life in patients who underwent stapes surgery more than 20 years after treatment.

Material and Methods: From September 2016 to June 2017 we collected patients who underwent stapedotomy or stapedectomy between 1990 and 2010. We performed otomicroscopy, pure tone audiometry, speech audiometry, tympanometry, acoustic reflex testing. Pre-operative and post-operative PTA for air conduction (PTA AC), PTA for bone conduction (PTA BC) and air bone gap were collected according to Committee on Hearing and Equilibrium Guidelines. Moreover, we interviewed patients with two Italian language validated questionnaires: Glasgow Benefit Inventory and Adults Hearing Handicap Inventory. We added another set of seven questions about subjective symptoms, regarding: current hearing compared to preoperative and early post-operative hearing, the incidence of tinnitus, vertigo/dizziness, facial paralysis, "blocked ear" sensation, sound distortion/metallic sound perception, sensitivity to sound, hearing aid use.

Results: we reached 268 patients previously to submitted to a stapedotomy or to a stapedectomy at our Clinic. Among this series only 70 patients showed up to undergo to physical examination, audiometric evaluation and quality of life assessment. All surgeries were performed without intra-operative complications. None case of total sensorineural hearing loss, or "dead ear", was detected postoperatively. During follow-up (22 years on average), no patients had postoperative major complications needing revision surgery. Patients age at surgery procedure was 19-50 years. 37 patients were operated in both sides in 2.5 years (range: 1-5 years). 33 patients were operated only in one ear, with 10 patients diagnosed with monolateral otosclerosis and 23 patients with bilateral disease. Overall, we evaluated 107 ears. Mean follow-up was 22 years (ranging from 17 to 27 years). Patients age at last examination was 58 years. 23 patients were male and 47 were female. The median preoperative PTA AC was 57 dB, the PTA BC was 25 dB. Preoperative ABG was 32 dB. At early postoperative examination (1 year after surgery) PTA AC was 41 dB, PTA BC was 29 dB and early post-operative ABG was 12 dB, with a mean gain (ABG Gain) of 20 dB. At late postoperative evaluation median PTA AC was 49 dB, PTA BC was 37 dB and late ABG gain was 12 dB. The questionnaires showed that patients suffered for a slight handicap and patients, particularly at the GBI, still perceive a significantly positive impact on global well-being as many as 22 years after the intervention.

Conclusion: Patients undergoing otosclerosis surgery on average 22 years earlier showed a slight decrease in both the airway and the bone pathway, in the absence of late complications requiring surgical revision. These data confirmed the safety and effectiveness of the surgery for otosclerosis.

The pre – and postoperative assessment of quality of life and hearing benefits in group of patients with otosclerosis undergoing stapedotomy.

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Introduction: Otosclerosis leads to hearing disability and may impact on social and psychological function. Review of the literature showed that quality of life in group of patients with hearing loss is worsen and decreased compared with group with normal hearing. Problems with speech understanding impact on daily patients life and restrict their participation in social life.

The aim of this study was to assess pre- and postoperative quality of life and hearing benefits in group of patients with diagnosed otosclerosis who were undergone stapes surgery.

Material & Methods: This study included 242 patients (182 women and 60 men) who underwent stapedotomy in the World Hearing Center of the Institute of Physiology and Pathology of Hearing in Poland, in period from April to June 2017 and who signed the consent to participated in this study. According to the BIAP Classification, hearing loss degree in group of patients was: mild-29 (12%), moderate 155 (64%), severe 43 (17.80 %) and profound 15 (6.20%).

Pre and postoperatively, all patients were asked to fill in questionnaires: the Assessment of Quality of Life (AqoL-8d) and Abbreviated Profile of Hearing Aid Benefit (APHAB), Tinnitus Functional Index (TFI). The preoperative results of pure tone audiometry were considered.

Results: The results showed that the average of general quality of life in AQoL questionnaire before surgery was in group of women M= 73.27; SD= 9.58 and in group of men M= 76.74; SD=7.47. Significant differences of quality of life before and after stapedotomy were observed t= -3.16; p=0.002) and hearing benefits in all subscales of APHAB questionnaire: EC t= 14.97; p<0,001; BN t=13.61; p< 0.001; RV t= 13.61; p<0.001 and AV t= -2,65; p=0.009. Also, quality of life was correlated with subjective hearing benefits for EC: rho= -0.286; BN= rho=-0,392; RV rho=-0.335; AV rho= -0.246; p<0.001. The significant difference of quality of life after surgery was observed in outcome of TFI for subscale "quality of life" and was t=3.97; p<0.001. The relationship was observed between audiometric results and quality of life especially for subscale "relationships" (AC: rho= -0.146; p<0.001; BC: rho= -0.135; p<0.01; ABG rho= -0.141; p<0.01).

Conclusions: The above results indicate that postoperative quality of life is related to subjective hearing benefits after stapedotomy. Obtained results showed a positive change after of stapedotomy in aspect of social relationship which can significantly affect the general assessment of quality of life.

It needs to be highlighted that conception of general quality of life can be related with other factors such as family life, social status and other disorders not directly association with hearing problems. It is important to use questionnaires which measure general quality of life and also dedicated for group of patients with hearing problems. It is important to assess areas which are directly associated with hearing and changes after treatment of hearing for example after stapes surgery.

Measuring the postoperative change in quality of life and its significant correlates among patients with otosclerosis – the preliminary findings.

Skarżyński P.H.[1,2,3], Dziendziel B.[1], Świerniak W.[1], Bieńkowska K.[1], Rajchel J. J.[1], Gos E.[1], Skarżyński H.[1]

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Aim: Aside from measuring the audiometric results of the surgical interventions, there is a trend of assessing the broader aspects of postoperative benefits, such as health-related quality of life. Patients' perception of the postoperative outcomes can be influenced by many factors, including the subjectively perceived hearing benefits or the coexistence and severity of tinnitus. The aim of the current study was to evaluate the quality of life change after stapes surgery and its significant correlates in adult patients, using a self-report measures.

Material and Methods: The study group consisted of 110 subjects, including 82 women and 28 men with the mean age of M=47.68; SD=10.12. All patients signed an informed consent form to participate in the study. The eligibility criteria were age ≥ 18 years, lack of contraindications to take part in the questionnaire study, primary stapes surgery for otosclerosis and a minimum of 6 months postoperative observation period. To assess the postoperative health-related change in quality of life, Glasgow Benefit Inventory (GBI) was used. Tinnitus complaint was assessed using Tinnitus Functional Index (TFI) and hearing benefits in different acoustic situations were measured with Abbreviated Profile of Hearing Aid Benefit (APHAB).

Results: Statistically significant, moderate to strong negative correlations were observed between the GBI results and different domains evaluated by the TFI (r=-0.545-,0.652; p<0.001). Additionally, strong negative correlations were found between the APHAB Ease of Communication (r=-0.63; p<0.001), Background Noise (r=-0.68; p<0.001) and Reverberation subscales (r=-0.58; p<0.001). No relationship was found between APHAB Averseness subscale (p>0,05). Age and hearing loss duration were not related to the GBI results (p>0.05).

Conclusions: Stapes surgery conducted by an experienced surgeon is a beneficial procedure leading to the increased health-related quality of life. Lower tinnitus severity and improved hearing ability in different acoustic situations seem to contribute to the increased health-related quality of life reported by adult patients with otosclerosis.

Stapes surgery in patients with a small air-bone gap.

McClenaghan F., Lavy J.A.

The Royal National Throat, Nose and Ear Hospital, London, UK

Aim: To determine hearing outcomes in patients undergoing stapes surgery with a preoperative air-bone gap (ABG) <21.25dB.

Method: Patients with a unilateral or bilateral preoperative ABG <21.25dB undergoing primary stapes surgery were identified from a database of all stapes surgery performed by one surgeon in a tertiary centre. 254 ears met the inclusion criteria.

Intervention: All patients underwent stapes surgery under local anaesthetic. Ossicular reconstruction was achieved using a Smart 360 nitinol fluoroplastic piston (Gyrus) and complete posterior crurotomy performed using KTP laser. Hearing was assessed immediately postoperatively with clinical voice testing and with pure tone audiometry at 6 weeks postoperatively.

Main Outcome Measures: The primary outcome measure was the degree of closure of the preoperative ABG. Secondary outcome measure improvement in bone conduction thresholds at 4kHz.

Results: 248 ears (97.6%) demonstrated ABG closure to less than 10dB. Bone conduction thresholds showed an increase in 114 (44.8%), no change 75 (29.1%) and decrease in 66 (25.9%).

Conclusions: There is a slight increase in the risk of stapes mobilisation in ears with a small ABG when compared those with larger ABGs however this can be overcome by using a laser-assisted technique in combination with good surgical experience. The benefit in terms of hearing aid avoidance and the restoration of symmetrical hearing is both achievable and significant for the patient.

Intra-operative prognostic factors in stapes surgery.

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Aim: To assess the results obtained in a large cohort of patients undergoing to otosclerosis surgery and to determine

the impact of intra-operative variables on post-operative hearing function and complications.

Material and Methods: We enrolled 384 patients affected by otosclerosis who were subjected to stapes surgery between 2004 and 2013. Surgery was performed in all cases under local anaesthesia, using manual perforator and/or microdrill. Teflon-piston prosthesis were used in all patients. Audiological data obtained preoperatively and at last follow up examination (minimum 12 months) were compared. Statistical analysis was performed using the multiple regression model.

Results: peripheral rim otosclerosis and diffuse otosclerosis were associated with better functional results compared to the obliterative pattern ($p<0.05$). Mean post-operative ABG was significantly greater in the 0,4 mm, compared to 0,6 mm piston group at 0.5 kHz ($p<0.001$) and 1 kHz ($p<0.02$). Stratification according to the surgical technique shows that in the partial stapedectomy group mean post-operative ABG does not significantly vary between 0,4 and 0,6 mm piston groups (9.1 dB in both), while in the stapedotomy group a statistically significant difference was found between 0,4 and 0,6 mm piston groups, in favour of the latter ($p<0.05$).

Conclusions: our data might be helpful to predict the post-operative outcome and to address patients' expectations.

Does otosclerosis surgery change the spatial hearing?

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Aim: Otosclerosis seems to negatively influence the central auditory system, causing spatial hearing deficits.

Material and Methods: We tested several adult patients with otosclerosis together with 50 age and sex-matched healthy volunteers. Our patients were tested before surgery and 1 month after surgical treatment due to otosclerosis. We used pure tone audiometry (PTAud), stapedial reflex test (SRT) and the horizontal minimum audible angle test (HMAAT) for 8 azimuths with binaural stimulation.

Results: The chosen patients with otosclerosis had typical conductive unilateral and bilateral hearing loss and the controls had normal or near normal hearing in PTAud. All of patients with otosclerosis had abnormal HMAAT results. The biggest quantitative disturbances in HMAAT were present in the bilateral hearing loss. We presented results for 8 azimuths of spatial hearing. The worst sound localization ability in HMAAT was noted for azimuths 90° and 270° compared with the ones without the stapes involvement.

Conclusions: Our study confirms the strong relationship between otosclerosis and sound localization disability and sheds some light on the complexity of the relationship.

Impact of stapes surgery on the Carhart effect.

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Otosclerosis is associated with elevated air conduction thresholds and an air-bone gap. However, depressed bone conduction may also be observed in this condition. Often this may be attributed to the Carhart effect and has implications when assessing patient suitability for stapes surgery.

Aim: This study aims to examine the audiological impact of stapes surgery with particular examination of pre- and post-operative bone conduction thresholds and their relation to air conduction thresholds.

Materials and Methods: Retrospective study of 336 otosclerotic patients managed by stapedotomy and SMART 360° prosthesis placement (either primary surgery or revision). Audiometry was performed pre- and post-operatively. Air conduction (AC) and bone conduction (BC) thresholds were recorded at various frequencies (0.5kHz, 1kHz, 2kHz, 3kHz and 4kHz). Patients were subdivided by pre-op BC thresholds.

Results: AC thresholds improved by 22.98dB in patients with pre-op BC <30dB average across all frequencies ($p<0.05$). BC improved by 2.54dB in the same group. AC improved by 26.04dB in patients with pre-op BC > 30dB. BC improved by 4.41dB in this group. There was a marked decrease in the air bone gap (ABG) post-operatively at all frequencies. Carhart's notch remained visible post-operatively albeit there was improvement in the hearing loss as audiological frequencies increased.

Conclusion: Stapes surgery improves AC and BC thresholds in otosclerosis. In an era when surgeons are offering surgical intervention for smaller air-bone gaps than traditionally considered worthwhile, one must consider the possibility that the air conduction could be improved to beyond the pre-operative bone conduction threshold.

Effect of the stapedotomy on early post-operative bone conduction.

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Aim: To analyze changes in bone conduction (BC) thresholds before and after stapedotomy in patients with clinical otosclerosis.

Material and Methods: Retrospective case review of pure tone audiometry (PTA) in 116 patients who underwent a stapedotomy at a tertiary referral center between 2007 and 2017. The patients were grouped by age and degree of hearing loss. We measured a mean preoperative and postoperative hearing level for single frequencies 0.5, 1, 2, 3 and 4 kHz and groups of frequencies (0.5-1-2-3 kHz and 0.5-1-2-4 kHz).

Results: The mean preoperative hearing levels for single frequencies was 22.1dBHL, 24.4dBHL, 32.8dBHL, 30.3dBHL and 27.9dBHL and for group frequencies (0.5, 1, 2 and 3, or 4 kHz), was 26.8dBHL and 27.5 dBHL. The mean gain in BC, defined on PTA (0.5, 1, 2 and 3, or 4 kHz), was 3.4 dBHL and 2.6 dBHL after stapedotomy.

Conclusions: Reconstruction of the ossicular chain in patients did not produce significant changes in the average values of bone conduction.

The assessment of the auditory benefits after stapedotomy using audiometric tests and self-report questionnaire.

Skarżyński P.H.[1,2,3,4], Ratajczak A.[1], Dziendziel B.[1], Świerniak W.[1], Gos E.[1], Bieńkowska K.[1], Skarżyński H.[1]

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Aim: The evaluation of the success of stapes surgery in otosclerosis is measured by audiologists on the basis of an increase in air conduction threshold and a reduction in the size of air-bone gap. Important question is, whether the results of audiometric test correspond to the change in hearing perceived by patients. The aim of the study is to assess the correlation between auditory benefits based on audiometric tests and perceived improvement of hearing using self-report questionnaire in the group of patients who underwent stapedotomy.

Material: From over 235 stapes surgeries performed between April and June 2017 in the World Hearing Center, there was selected group of 66 patients with otosclerosis who underwent first-time stapedotomy and fulfil the study inclusion criteria. Presented group consists of patients aged from 25 to 70 years. The number of women predominated in the surveyed group - about 73%.

Methods: All patients underwent pure-tone audiometry before and after stapedotomy. In addition, patients were asked to complete the Abbreviated Profile of Hearing Aid Benefit (APHAB) questionnaire for the self-assessment of communication ability.

Results: The average hearing thresholds (for frequency from 0.5 to 4.0 kHz) for the air and bone conduction in the operated ear significantly increased after stapedotomy. There was also observed air-bone gap reduction by almost 16 dB after surgery. Statistically significant correlation between pure-tone audiometry results and particular APHAB scales was perceived.

Conclusion: The results of pure-tone audiometry in presented group of patients indicate improvement of hearing after stapedotomy, which confirms the success of surgery. The research also indicates that auditory benefits based on pure-tone audiometry results correspond with self-perceived improvement in APHAB questionnaire. Usage of this questionnaire is very important - it is a great complementary tool to audiometric tests, because it provides valuable information how big problem is hearing loss for the patient.

Assessment of tinnitus oppressiveness using the TFI questionnaire after the implantation of the CODACS system – a case study.

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Aim: Assessment of TFI (Tinnitus Functional Index) changes in patients with severe to profound mixed hearing loss before and after the implantation of the CODACS system – case studies.

Material: The analysis concerns four patients with diagnosed otosclerosis, severe to profound mixed hearing loss and tinnitus. The patients after numerous surgeries of both ears have undergone the implantation of the CODACS implant in the Institute of Physiology and Pathology of Hearing.

Method: The TFI questionnaire was performed as part of efficiency assessment of the applied solution. They were used to evaluate changes in oppressiveness of tinnitus. The tests and questionnaires were performed before the implantation and after the system had been activated.

Findings: The results of the questionnaire assessment indicate significant changes in the value of the TFI rate in

all the tested areas, confirming a reduction in the oppressiveness of tinnitus.

Conclusions: The gathered preliminary findings indicate that using the CODACS system may efficiently affect lowering oppressiveness of tinnitus.

Clinically important change in tinnitus sensation after stapedotomy.

Gos E., Raj-Koziak D., Dziendziel B., Skarżyński P.H., Skarżyński H.

World Hearing Center, Institute of Physiology and Pathology of Hearing, Warsaw/ Kajetany, Poland

Aim: Statistically significant difference in the outcome of treatment does not necessarily imply clinical importance, i.e. whether the observed improvement is important or meaningful to the patient. The aim of the study is to determine a change in tinnitus which is clinically important to patients with otosclerosis.

Material and Methods: Around 100 patients with otosclerosis reporting tinnitus complaints filled in tinnitus-related PROMs (Patients-Related Outcome Measures) including Tinnitus Functional Index. Tinnitus was measured before and 3 months after stapedotomy.

Results: Patients assessed the change in their tinnitus 3 months after stapedotomy using a 7-point scale with the following degrees: 1- very much worse; 2 – much worse; 3 – minimally worse; 4 – no change; 5 – minimally improved; 6 – much improved; 7 – very much improved. The mean change method and the receiver operating characteristic (ROC) method were used to determine minimal important change in tinnitus sensation.

Conclusions: The anchor-based approach using an external criterion (anchor) allows to determine change in tinnitus sensation which is meaningful to patients.

Audiometric results after stapedotomy with platinum prosthesis.

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Otosclerosis is disorder occurring with conductive or mixed hearing loss, often with coexisting tinnitus and vertigo. Crucial issue is stapes fixation. Purpose of our study was to assess hearing results and complications after stapedotomy when platinum wire prosthesis was used.

Material consisted of patients (228 women and 102 men, mean age 46,5 (range 6-76)), who underwent stapes surgery from September to December 2011 at World Hearing Center of the Institute of Physiology and Pathology of Hearing in Warsaw. Patents were analyzed retrospectively. Medical histories, surgery protocols and hearing examination outcomes were particularly reviewed.

Symptoms presented in our group were: progressive hearing loss, also tinnitus (>70%) and 20% vertigo. Comparing pure tone audiometry were mentioned frequencies: 0,25, 0,5, 1,2,4 kHz. After surgery the mean bone and air conduction decreased, and air bone gap also decreased. For frequencies range 0,25-2 kHz data were statistically significant ($p<0,05$). Mean gap reduction was from 22,7dB for 0,5kHz to 13,1dB for 2kHz.

Summing up this study and our previous experiences, stapedotomy is effective otosclerosis treatment method. Complications are rare, and hearing effect is satisfactory both for patients and surgeons. It's crucial due to frequent young age and professional activity of patients.

Friday, 6th April

Lecture Session II

Invited Lecture

The hidden history of stapes surgery.

McElveen J.T.

Carolina Ear & Hearing Clinic, Carolina Ear Research Institute / Camp Woodbine, Raleigh, USA

As otolaryngologists, we are well aware that stapedial otosclerosis is a disease process that involves the otic capsule of the temporal bone, resulting in a conductive hearing loss due to stapes fixation. In a classic study by Guild, histologic evidence of otosclerosis was found in 8.3% of 518 Caucasian temporal bones and 1% of 482 African-American temporal bones. More recently, Dr. Mike McKenna has implicated the measles virus in the development of the otosclerotic process. Regardless of the etiology, the universally accepted surgical treatment for stapedial otosclerosis is stapedotomy or stapedectomy.

However, this was not always the case. In this presentation, we will review the "Hidden History" of stapes surgery, beginning with the anatomical investigations of Valsalva, the histologic findings of Politizer and the surgical interventions of Kessel, Ménière, Holmgren, Sourdille, Lempert, Rosen and Shea.

As detailed in this presentation, the path to the current treatment of otosclerosis with stapedotomy has been a torturous one. Many of the pioneers were ridiculed, and some, such as Johannes Kessel, were forced to resign their positions. Other pioneers, such as Lempert, could not accept change and became obsolete. In the end, a young man, Dr. John Shea, who with the help of a politically savvy older man, Dr. Howard House, allowed stapes surgery to return to its rightful place in the treatment of otosclerosis.

Keynote Lecture

Do we need always a CT Scan in the diagnosis of otosclerosis.

Fraysse B., Escude B., Lagleyre S., Mary M.

Service ORL, Hôpital Pierre-Paul Riquet, Toulouse, France

The author will discuss the interest of imaging for the following reasons:

- To confirm a doubtful diagnosis.

Of 209 CT-scans of operated ears, 84% were classified positive, 9% doubtful and 7% negative. The sensitivity of CT-scan to otosclerosis was 95.1%. In the operated ears, the mean preoperative and postoperative bone conduction thresholds were significantly lower in cases of otosclerotic focus involving the endosteum ($p<0.005$ and $p<0.0001$ respectively).

- To define a surgical strategy in case of anatomical difficulties (small fenestra, obliteration, facial dehiscence) and he will describe all the causes of possible inner conductive hearing loss.

- To anticipate the evolution of postoperative bone conduction according to the extension of lesions by comparing on a series of 183 patients, two group with and without endosteal involvement.

- To analyze the cause of failure. The author will describe a new entities "the lateralization syndrome".

Keynote Lecture

Bone conduction in otosclerosis.

Röösli Ch., Chhan D., Rosowski J.

Department of ENT, Head and Neck Surgery, University Hospital Zürich, Switzerland

Department of Otology and Laryngology, Harvard Medical School, Boston, USA

Aim: To discuss the contribution of different pathways to bone conduction stimulation and its influence on hearing threshold in patients with otosclerosis.

Material and Methods: Measurements were performed in five anesthetized chinchilla. Acoustic stimuli were presented via an insert earphone, and bone conduction stimuli were transduced via a BAHA between 0.1 to 10 kHz. A pressure sensor to measure introcochlear pressure was placed is the scala tympani (Pst), and in the scala vestibuli (Psv). Additionally, sound pressure in the external auditory canal was recorded. Measurements were repeated with the ossicular chain intact, interrupted, and with the footplate fixed.

Results: With AC stimulation, both Psv and Pst drop by 30 to 40 dB after the interruption. In BC stimulation, Psv decreases after interruption by about 10 to 20 dB, but Pst is little affected. For fixation of the stapes, Pst changed less than Psv. The differential pressure changed significantly between 1 and 3 kHz.

Conclusions: The results suggest that the inner-ear mechanisms play a large role in bone-conduction stimulation in the chinchilla at all frequencies. However, the data also suggest the pathways that conduct vibration to the inner ear via ossicular-motion make a significant contribution to the response to BC stimulation in the 1 to 3 kHz range, such that interruption of these path leads to a 5 dB reduction in total stimulation in that frequency. This

phenomenon is similar to the Carhart's notch observed in patients with otosclerosis.

Keynote Lecture

A simplified analytical model of sound transmission in the human ear.

Gambin W.

Warsaw University of Technology, Warsaw, Poland

Aim: The subject of interest is behavior of the sound wave running from the sound source, through the outer, middle and inner ear, up to the round window membrane. Our goal is to find simple analytical rules for the level of sound intensity, amplitude and pressure of the going wave. It assumed that the intensity level of sound, the sound wave frequency, as well as, the dimensions of the ear and/or the parameters of ear prosthesis are given. Such the rules might be used for an analyses of ear activity, before and after the implantation of prosthesis. Among others, these rules show the effect of changes in the anatomy of the ear on the reception of sound.

Materials and Methods: At every stage of the sound propagation process, all looked rules yield from the energy conservation law. These rules can be determined on the basis of the following assumptions. (1) The outer ear is waveguide filled with air, in which propagates an elastic plane wave at a given frequency and sound level. The waveguide ends with a flexible tympanic membrane on which a partial reflection of the wave appears. (2) Three ossicles of the middle ear transmit oscillations of tympanic membrane to the inner ear. The whole is a vibrating mechanical system. This system converts the sound energy passed through the tympanic membrane on the work of the force moving the stapes footplate. (3) The inner ear is taken as a rectilinear waveguide filled with liquid, in which propagates an elastic plane wave forced by the vibration of the stapes footplate. Here propagates this part of the sound energy which has been passed through the tympanic membrane, transmitted by the mechanism of the ossicles and absorbed by the fluid of the cochlea. Along this waveguide extends a basilar membrane assumed to be a set of independent springs. The other end of the waveguide is closed by the elastic membrane of the round window, which completely absorbs the energy of the running wave. It is assumed that the sum of the energy of the running wave, the rippling basilar membrane and the deformed round window membrane is equal to the energy of the vibrating stapes footplate.

Results: For a set sound intensity level with a given frequency, the rules for sound intensity, amplitude and pressure of sound wave in outer and inner ear were obtained. In addition, associated displacements of the stapes footplate and round window membrane, were found.

Conclusions: The proposed approach neglect all viscous effects, a bone conduction and the real interaction of the basal membrane and the cochlea fluid. However, it gives some averaged results, which are close to the experimental ones. For example, experiments show that for intensity level equal to 90 dB we SF displacements drop from 16.4 nm to 4.63 nm, when frequency increases from 400 Hz to 2000Hz. SF displacements get from the analytical rule do not depend on the sound frequency. The rule gives the value 10.2 nm, which corresponds to the frequency 1000 Hz.

Keynote Lecture

Revisiting the learning curve for stapes surgery.

Barbara M., Covelli E., Monini S.

ENT Clinic, NESMOS Department, Sapienza, Rome, Italy

Stapes surgery is a challenging procedure that needs a long and intense specific activity by the otologists. Although residency programs include this procedure within the core of the course, very rarely a resident achieves to perform the entire procedure and is uniquely approaching single steps, usually the initial ones, such as the tympano-meatal flap. So, the real learning curve is actually starting after the residency and exclusively in those academic or hospital Centres where this type of surgery is performed with great numbers. Considering the reported worldwide decrease of otosclerotic patients, even in the major otologic Centres the operated cases just cover the maintenance of dexterity of the senior staff so that teaching the technique is left to temporal bone lab or occasional access to the operative theatre. After this premise, it is worth adopting the safest technique for allowing the beginner to get knowledge of the different steps of the procedure. In this regard, the reversal step Stapedotomy assisted by Laser could be considered an appropriate procedure.

Diagnostic Approaches

Keynote Lecture

Imaging of patients with suspected otosclerosis.

Szymański M.

Department of Otolaryngology Head and Neck Surgery, Medical University of Lublin, Poland

The main diagnosis of otosclerosis is based on audiological tests. However there are several entities resembling otosclerosis which may require different management or surgery carries higher risk of complication. The use of high resolution CT and/or Cone Beam CT enables visualisation and localisation of otosclerosis foci. Advanced imaging studies can differentiate patients with otosclerosis from other causes of conductive hearing loss. The lecture presents typical picture of fenestral otosclerosis, the use of imaging in preoperative and postoperative assessment and differential imaging diagnosis in patients with audiologicaly detected oosclerosis.

Comparison of high-resolution CT, pure tone audiometry and surgical findings in patients with otosclerosis.

Kunelskaya N.L., Garov E.V., Zelikovich E.I., Zagorskaya E.E., Kurilenkov G.V., Kiselyus V.E.

Sverzhevsky Otorhinolaryngology Clinical Research Institute, Moscow, Russia

Introduction: Nowadays high-resolution CT (HRCT) is widely used for planning stapes surgery. This method is helpful for localization of otosclerotic foci, anatomic abnormalities and determines risks of complications. However, the correlation between CT-signs of prevalence otosclerotic foci and audiological testing are various.

Aim: to evaluate correlation between results of HRCT, pure tone audiometry (PTA) and surgical findings in patients with otosclerosis.

Materials and Methods: 691 stapedoplasties were performed between January 2010 and October 2017 in Sverzjevsky Otorhinolaryngology Healthcare Research Institute (Moscow, Russia). 89 patients (102 ears) who underwent stapedoplasty by the same surgeon were selected for retrospective study; 67 were females (75.3%) and 22 males (24.7%), the mean age was 44.6 years (from 25 to 71 years). All patients with intra-operatively confirmed otosclerosis has preoperative temporal bone HRCT (GE Discovery 750 HD, 0.625-mm-thick axial imaging with multiplanar reconstruction, USA), performed by the same radiologist. Patients with any additional non-otosclerotic reason for hearing loss were excluded. PTA was performed by the same clinician.

HRCT data: fenestral localization of otosclerotic foci were identified in 68 ears (66,7%), fenestral and retrofenestral (mixed) – in 18 (17,7%) and retrofenestral – in 2 (1,9%). In 14 ears, there were no significant signs of otosclerosis. Unilateral lesions identified in 7 cases. Otosclerotic foci ranged from 0.5 to 10 mm, mean density of fenestral foci was 1074,3 HU and retrofenestral - 970,8 HU. Obliteration of oval window niche detected in 5 cases (4.9%), and facial nerve canal overhanging – in 3 (2.9%). Evaluation of PTA conducted at 0.5, 1, 2 and 4 kHz. Conductive hearing loss (CHL) was identified in 16 cases (15,6%) and mixed hearing loss (MHL) – in 86 (84,4%), 12 were unilateral. Mean PTA bone-conduction levels were 28.4 dB (from 8.3 to 53.8 dB), mean preoperative air-bone gap (ABG) was 31.6 dB (range 19.8 to 55 dB). Intra-operative prevalence of otosclerotic foci was: promontory - 77 cases (75,5%), footplate - 58 (56,9%), anterior part of oval window niche and footplate - 33 (32,4%). Narrow oval window niche ≤0.3 mm identified in 5 cases (4.9%).

Results: Sensivity of HRCT in diagnosis of otosclerosis was 86.3%. Comparison of HRCT and PTA revealed appropriation of fenestral form and CHL in 10 cases (9.8%), mixed form and MHL – in 19 (18.6%). Analysis of surgical findings and prevalence of otosclerotic foci on HRCT revealed appropriation in 28 cases (27.4%). During surgery were confirmed 5 cases of obliteration of oval window niche and 3 cases of facial nerve canal overhanging, that identified in HRCT.

Conclusion: HRCT is sensitive method for diagnose otosclerotic foci prevalence; however, it remains vulnerable for lesions less than 0.5 mm. HRCT is suitable for planning surgical treatment, allowing to predict difficulty and complication risk.

Wideband absorbance measures for assessment of otosclerotic ears: A preliminary study.

Kochanek K., Śliwa L., Skarżyński H.

World Hearing Center, Institute of Physiology and Pathology of Hearing, Warsaw/Kajetany, Poland

Background: Wide Band Tympanometry (WBT) is a reliable methods for assessment of middle-ear disorders. Literature reports indicate usefulness of WBT for qualification to reconstructive surgery and assessment of surgery results, especially in otosclerosis.

Material and Methods: Energy absorbance (EA) measurement was performed, along with other diagnostic tests, in a group of patients diagnosed for otosclerosis. In some of them the tests were repeated in postoperative period and the effects surgery were assessed.

Results: EA value shows significant changes in otoslerotic ears compared to normal-hearing ones. A meaningful decrease in EA value is observed especially in mid-frequency region, 500-1500 Hz. In many cases, 226Hz tympanograms in otoseritic ears are normal.

Conclusion: WBT is an effective method for detecting middle-ear disorders, like ossicular chain fixation, disarticulation, etc. It proves much more sensitive than standard low-frequency tympanometry. It may also be used for assessment of surgery results.

Audiological profile of patients with large vestibular aqueduct.

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Aim: Audiological characteristics and treatment of patients with large vestibular aqueduct (LVA).

Material: Retrospective analysis of 15 patients (10 women and 5 men), mean age 25,1 (SD 11,4) years with radiologically confirmed large vestibular aqueduct (LVA) has been carried out. Bilateral LVA was diagnosed in 9 patients, right ear localization was in 1 patients and left ear localization in 5 patients.

Method: All patients underwent audiological diagnostic procedure including pure tone threshold specified in tonal audiometry, impedance audiometry, otoacoustics emissions (DP-gram) and auditory brainstem responses (ABR). Possible deterioration of hearing was monitoring by tonal audiometry tests conducted at two-year intervals.

Results: LVA was confirmed by CT or MRI examination, secondary confirmed by CT. LVA was associated with Pendred syndrome in three and hypothyroidism in one patient. Mixed hearing loss as a main syndrome, extended from mild to profound, was either stable or progressive. Associated tinnitus was reported by 5 patients. Episodes of one sided or bilateral sudden deafness were diagnosed in 53% of patients, mostly with no results after standard treatment. Imbalance disorders were noticed in 5 patients. Eight patients were supplied with hearing aids. Six patients with progressive severe and profound hearing loss were qualified as candidates for cochlear implantation.

Conclusions: LVAS should be taken into account with differential diagnosis of otosclerosis. Cochlear implantation is a possible solution for LVAS patients with progressive hearing loss.

Lecture Session III

Keynote Lecture

Far advanced otosclerosis: Stapedotomy or cochlear implantation.

Fraysse B., Kabbara B.

CHU Purpan, Service ORL, Toulouse, Paris

Objectives: To describe the hearing outcomes in a group of patients with far-advanced otosclerosis managed with either stapedotomy, cochlear implantation or both, and to identify the best initial approach based on preoperative speech discrimination, pure tone average (PTA), and radiological classification.

Study design: Retrospective chart review.

Setting: Academic neurotology tertiary referral center.

Inclusion criteria: All patients within the cochlear implant range with a pure tone average below 85 dB (HL) and a preoperative dissyllabic word recognition score below 50% at 60 dB (HL) with maximal hearing aid, along with evidence of otosclerosis on CT Scan.

Results: Overall, 58 patients fit the inclusion criteria and were divided into 3 groups: A- stapedotomy group (n=32) B- Cochlear implant as a first option (n=11) C- Cochlear Implant with previous stapedotomy (n=23).

Otosclerotic lesions were classified radiologically based on a new CT Scan grading system:

I - isolated lesion involving the footplate and/or pericochlear lesion without endosteum involvement (7,8%);

II – pericochlear lesion with endosteum involvement (50%);

III – Complete obliteration of the round window and/or pericochlear lesion with ossification of the basal turn (42,2%).

Changes in word recognition scores (WRS) were compared between the three groups after a minimum of 12 months follow up. The mean post op WRS for groups A,B and C were 49% ,63% and 77% respectively. 60% of patients in the stapedotomy group had a postoperative WRS ≥ 50% requiring no further treatment, subsequent cochlear implantation due to deterioration of hearing. This is compared with 72% for group B (cochlear implant alone), and 95% for group C (both). The preoperative audiometrical first was correlated with the outcome of stapedotomy. However, the specificity was very low.

Previous stapedotomy has not impact on cochlear implant outcomes. The rate of surgical difficulties during cochlear Implantation (n=37) was 51%, these included round window and/or basal turn extra drilling, scala vestibuli insertion, incomplete insertion or perilymphatic gusher. These complications occurred in patients with grade III lesions 79% of the time. Non acoustic stimulation occurred in 29,5% of patients, and these two tended to occur more frequently in patients with grade III lesions (70%).

Conclusion: Although cochlear Implantation gives better overall results in terms of speech discrimination, we believe that stapedotomy with a well fitted hearing aid should be first attempted due to a relatively high success rate, its low cost and low complication rate.

Keynote Lecture

How to avoid complications in stapes surgery.

Lavy J.

Royal National Throat, Nose and Ear Hospital, University College London Hospitals, The London Ear Clinic, London, UK

Stapes surgery is one of the most successful surgical interventions for the restoration of hearing with excellent results being achievable in over 90% of patients by experienced surgeons. However at each step of the operation complications can occur. These include perforations of the tympanic membrane, damage to the chorda tympani, mobilisation of the stapes, subluxation of the incus, injury to the facial nerve and sensorineural hearing loss. The speaker has carried out around 2000 stapes operations and at present does 150-200 primary and revision procedures per year. This personal, practical experience and surgical tips acquired to reduce the risk of complications will be presented.

Keynote Lecture

How to overcome hurdles during Stapes Surgery.

Vijayendra H.

Vijaya ENT Care Centre, Bangalore,India

Stapes surgery is one of the most delicate surgery in ear surgery. In classical stapedotomy, first I create tympanomeatal flap via transcanal approach. After entering middle ear, I first confirm diagnosis by touching the ossicular chain. To obtain exposure I curette posterosuperior canal wall. Chorda tympani nerve secured away from working area so that it won't come in way during the procedure. I always first make controlled fenestra of 0.3mm in posterior half of the footplate and then remove suprastructure. To remove suprastructure, I first drill posterior crus using 0.5mm diamond burr of which speed I control with footpad. Then i disarticulate incudostaedial joint and cut stapedius tendon near pyramidalis process. Then using 450 angled pick I remove suprastructure. Then I widen fenestra to 0.6mm diameter using Fisch's perforators. I always prefer to use 0.4mm diameter Teflon piston with length of 4.25mm most of the time. If I feel incus is medialised then I cut short the lenth and if footplate is very thick I will add extra thickness of footplate to this length of piston. First I insert piston into fenestra and then anchored it over long process of incus and crimp it. I always seal surrounding of fenestra using post aural soft tissue. On occasions, I do come across difficult situations like otoslcerosis fixing stapes along with fixation of incudomalleal joint, bony ankylosis of malleus, perilymph gusher, inadequate exposure to stapes footplate due to prominent posterior crus and dehiscent facial nerve overlapping footplate, fixation of incus to facial canal with very thick footplate, fixation of incus in fossa incudes, thick dense fibrous tissue encasing stapes suprastructure, thick anterior crus, very thick footplate, necrosis of long process of incus, brittle footplate.

I will be showing in my presentation how I have tackeled all this situatlons.

Keynote Lecture

Revolix JR Laser Assisted Stadedotomy: a personal experience.

Barbara M., Covelli E., Talamonti R., Bianchi A., Marrone V., Tarentini S., Volpini L., Monini S.

ENT Clinic, NESMOS Department, Sapienza, Rome, Italy

Aim: Otosclerosis is one of the main causes of middle-ear conductive hearing loss and it is characterised by the spongization of the otic capsule and re-ossification that causes ear–bone fixation, resulting in progressive conductive hearing loss and deafness. The main challenge in stapes surgery has always been to recover the auditory function without damaging the inner ear structures, complication reported in about 1 % of cases. Several laser systems have been employed for stapes surgery in order to obtain higher functional results compared to common traditional devices such as manual perforator and/or ultralight microdrill. Over the years, standardised and reproducible outcomes have been achieved, together with progressive reduction of the rates of intraoperative labyrinthine trauma, due to footplate manipulation.

The introduction of lasers in otologic surgery has been particularly welcomed in stapes surgery for which handheld or micromanipulator-mounted systems are available. In fact, lasers can provide a precise, non-traumatic mechanical procedure, the heating effects on the labyrinthine fluids being eventually related to the wavelength of each specific laser system. The aim of this study is the evaluation of air bone gap (ABG) for 250-4000 Hz, before and after at least 30 days from stapes surgery, and intra and post-operative complications in two groups of patients undergoing Revolix JR laser stapes surgery with and without manual perforator. The frequencies 4000 and 8000 Hz were considered separately to investigate the impact of the laser Revolix JR on high frequencies, I.e. on the cochlear structure.

Material and Methods: We retrospectively reviewed medical records of 187 patients (125 female and 62 male, range 21-74 years, mean age 47,5), treated for otosclerosis, from January 2009 to July 2017, at the NESMOS Department, ENT Unit, «Sapienza» University, Rome, Italy. 86 patients underwent Revolix JR laser stapedotomy "Group 1" (46 classical stapedotomy and 40 reversal-steps stapedotomy) and 101 patients underwent stapes surgery through the association of one shot laser and manual perforator "Group 2" (35 classical stapedotomy and 66 reversal-steps stapedotomy). Surgery was performed mostly under general anaesthesia. Audiological evaluation was performed according to the guidelines of the Committee on Hearing and Equilibrium. Audiological data from preoperative examination (24 h before surgery), and postoperative follow-up examination, were compared for hearing outcome. Hearing outcome was calculated as PTA ABG closure < 10 dB, 11-20 dB PTA ABG closure, 21-30 dB PTA ABG closure, 31-40 dB PTA ABG closure and >40 dB PTA ABG closure. We have also analysed intra and post-operative complications. Intra-operative complications included those related to damage to the stapes footplate or to the instrumentation used (laser heating, drill noise) that could be in relation to the occurrence of nausea and/or vomiting with vertigo spells, during or soon after surgery; postoperative complications included balance disorders, hearing loss, fullness during the first month after surgery.

Results: In Group 1: 74% of patients closed ABG within 10 dB, 23% in a range between 11-20 dB and 3% of patients > 20 dB. In Group 2: 72% of patients closed ABG within 10 dB, 21% in a range between 11-20 dB and 7% of patients > 20 dB. Intra-operative complications occurred in 8% of patients. Most of them were represented by damage to the stapes footplate (cracking, flotation or involuntary total removal), oozing, accidental injury of chorda tympani or eardrum perforation. No statistical differences were observed between the two stapes surgery techniques regarding intra and post-operative complications. Bone

conduction evaluation for 4000 Hz and 8000 Hz before and after surgery does not show noteworthy differences. In the majority of the patients it was shown the closure of air-bone gap in the postoperative period. Bone-conduction hearing thresholds were stable in both group.

Conclusion: Our experience on a large series of patients confirms the absence of damages for the inner ear and optimal functional results that remained stable at the postoperative follow up. Our data suggest that laser Revolixx JR for stapedotomy is an effective and safe procedure for the treatment of otosclerosis.

Keynote Lecture

Experiences with laser-assisted stapes surgery using the NiTiFLEX stapes prosthesis.

Plontke S.K., Zirkler J., Gey A., Rahne T.

Department of Otorhinolaryngology, Head and Neck Surgery; Martin Luther University Halle-Wittenberg, Halle, Germany

Background: Coupling of the prosthesis to the long process of the incus is a critical step in stapes surgery. We here report our experiences with a titanium stapes prosthesis with a superelastic nitinol clip (NiTiFLEX®), a further development of the Soft CliP® stapes prosthesis (KURZ, Dusslingen, Germany). Instead of pure titanium, the CliP® now consists of nitinol, a superelastic nickel-titanium alloy. This further reduces the pressure exerted on the long process of the incus, aiming to improve coupling and minimize the risk of incus luxation.

Methods: In a monocentric, retrospective cohort study, we evaluated air-bone gap reduction (4PTA0.5-4kHz), understanding of monosyllables at 65 and 80 dB SPL stimulation levels, and adverse effects, such as changes in bone conduction (4PTA0.5-4kHz). Stapes surgery with the NiTiFLEX® titanium prosthesis with a superelastic nitinol clip was performed in 73 otosclerosis patients. The footplate was perforated using a CO2 laser (scanning mode; 0.7-0.8 mm; 20-21W) in nearly all cases. The diameter of the titan piston was 0.4 mm. Immersion at an angle of almost 90° thus results in a gap between the footplate perforation and the piston of 0.15-0.2 mm. Positioning onto the long incus process was performed with a 90°microhook (0.6 mm). Prosthesis length varied from 4,25 to 4,75 mm.

Results: In all cases, the NiTiFLEX® stapes prosthesis was inserted without complications. A preliminary evaluation of the audiological results showed no significant change in the mean air-bone gap (4PTA0.5-4 kHz) and an average improvement of the mean air-bone gap (4PTA0.5-4 kHz) comparable to results from other reports with different prostheses. A recent update of the audiological assessment is awaited. Handling of the prosthesis was rated as very good.

Conclusion: This study suggests that the NiTiFLEX® stapes prosthesis is a successful further development of the Soft CliP® technique with very good intraoperative handling qualities. While long-term results from larger studies are needed, this case series demonstrates that the hearing outcome is comparable to other prostheses.

Differential diagnosis of otosclerosis vs. superior semicircular canal dehiscence syndrome.

Hüttenbrink K.B.

Department of ORL, University Clinic Cologne, Germany

Since the first description of a dehiscence of the superior semicircular canal (SSCD), debates have emerged on its implication for a loss of conductive hearing. This idea was based on animal experiments with an artificial opening for the SSCD towards the air of the tympanic cavity. This, however, is not a correct approach, as in humans the canal is covered by dura and loaded by the mass of the brain and cerebrospinal fluid. Due to physiologic principles, a third window anywhere in the labyrinth will have only in a minuscule effect on sound transmission. This had been confirmed by the data of animal temporal bone experiments with an opening of the labyrinth towards the air. A 10-15 dB decrease of sound transmission could be recorded only in the low frequencies; above 1kHz, no effect was measurable. Further temporal bone experiments with covering the dehiscence with a piece of dura showed no change of sound transmission as compared to the preoperative intact canal.

Therefore, no significant change of sound transmission, nor a presumably otosclerotic air bone gap with a transmission loss higher than 20dB, perhaps with the negative rinne can be expected in case of a defect of the superior semicircular canal.

Furthermore, it must be respected, that even modern high resolution CTs cannot exclude a very thin bony closure of a semicircular canal, even if radiologist cannot demonstrate this bone. Many other symptoms, also related to SSCD, like Tullio and Hennebert vestibular symptoms, are also not concurrent to a presumed fistula, as these assumptions forget that the interposed ossicular joints, due to their gliding, will eliminate any significant pressure transfer from the external ear canal into the inner ear. A bulging of the labyrinth through a bony defect into the dura is unrealistic.

Arguments of unsuccessful stapes surgery with unresolved conductive loss must be attributed to other factors, for example not mentioned surgical failure, bony obliteration of the round window membrane, malleus fixation etc., to name only a few. Furthermore, experts in the older generation of ear surgeons, like Schuknecht etc., had already attributed these unspecific symptoms, which are nowadays correlated to SSCD, to connective tissues strands between the footplate and the membranous labyrinth.

Some dramatic improvements of uncharacteristic vertigo symptoms and even hearing disorders after surgery for SSCD should also consider the beneficial effect of drilling in the temporal bone, well known from "sham"-operations for Meniere`s disease.

Treatment Approaches

Stapes surgery for patients with mixed hearing loss.

Rapier J.J., Theodoraki G.E., Manjaly J.G., Nash R., Jain A., Lavy J.A.

Royal National Throat, Nose and Ear Hospital, University College London Hospitals, The London Ear Clinic, London, UK

Otosclerosis is associated with elevated air conduction thresholds and an air-bone gap. However, depressed bone conduction may also be observed in this condition. This may imply a cochlear component to the hearing loss but may also be attributed to the Carhart effect and has implications when assessing patient suitability for stapes surgery. Patients with bone conduction thresholds of <30dB are not traditionally considered good candidates for stapes surgery, if the aim of surgery is to avoid the need for hearing aid use.

Aim: This study aims to determine if there is benefit in performing stapes surgery for otosclerosis in patients with better ear bone conduction (BC)<30dB.

Methods: Retrospective review of 111 patients with otosclerosis and BC thresholds <30dB undergoing stapedotomy and SMART 360° prosthesis placement by the same surgeon. Assessment was made of pre-and post-operative audiometric data, pre- and post-operative hearing aid usage and change in patient quality of life using the Glasgow Benefit Inventory (GBI).

Results: 94 patients had complete audiometric data and were included. Average postoperative air conduction improved from 63.7dB to 36.9dB. Average air-bone gap was closed from 29.5dB to 7.85dB. Bone conduction improved from 34.1dB to 29.1dB. One patient experienced significant increase in air conduction of 49dB. Importantly 35% of patients in the study achieved postoperative air conduction thresholds of <30dB, even though their preoperative BC threshold was >30dB. This would have been an unexpected result, if the Carhart effect hadn't taken place and provides evidence that preoperative bone conduction thresholds in otosclerosis patients are not an accurate marker of cochlear function. Patients' GBI scores showed a significant increase in their quality of life postoperatively. There was also a significant decrease in daily hearing aid usage post-surgery, with a number of patients going from wearing a hearing aid at all hours of the day to never wearing one.

Conclusions: This study results suggest that patients with otosclerosis and BC thresholds of >30dB can achieve significant benefits from surgery, both in objective audiometry data, as well as in subjective increase in quality of life and decrease in hearing aid usage needs. When assessing suitability for surgery, surgeons should consider the fact that pre-operative bone conduction thresholds may be a poor indicator of the true cochlear reserve and therefore the potential for improvement in air conduction thresholds.

Stapes surgery in patients with profound hearing loss secondary to otosclerosis.

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Royal National Throat, Nose and Ear Hospital, University College London Hospitals, The London Ear Clinic, London, UK

Objective: To evaluate the efficacy of stapes surgery in patients with otosclerosis and profound hearing loss, who meet audiometric criteria for cochlear implantation.

Study design: Retrospective study and questionnaire.

Setting: Tertiary care hospital. Patients: Thirty-five adult patients (thirty-seven ears) with profound hearing loss. Ages ranged from forty to eighty-five, with a mean age of 63.4. **Intervention:** Stapedotomy with prosthetic implantation.

Main outcome measures: Benefit from stapes surgery in this patient population was measured audiometrically, using pure tone threshold averages (PTA) from 0.5kHz, 1kHz, 2kHZ and 4kHz, before and after surgery. In a subgroup of eight patients, the Glasgow Benefit Inventory (GBI) tool was used to assess impact on quality of life.

Results: Thirty-one of thirty-seven ears demonstrated improvement on PTA. The mean PTA from thirty-six ears was an improvement of 32.3dB (one patient was excluded because their thresholds were not recordable, and thus could not be included in numerical analysis). Of those that did not benefit, there was no PTA change in four patients, while two patients experienced elevated PTA. Only two patients in this cohort went on to have cochlear implantation. The mean GBI score was +19.8. Seventy-five percent of patients demonstrated functional improvement on GBI. One patient experienced very significant improvement with a score of +77.8. Twenty-five percent demonstrated a decrease in function, with GBI scores of -5.50 and -25.0.

Conclusion: Patients with otosclerosis and profound hearing loss who would meet audiometric criteria for cochlear implantation derive benefit from stapes surgery both quantitively and qualitatively. In most circumstances, stapes surgery improves hearing thresholds to a level where hearing aids become beneficial to the patient, thereby avoiding the need to change quality and tonality of sound and commit the patient to further rehabilitation through cochlear implantation.

Stapedoplasty as a method of hearing loss treatment in patients with tympanosclerosis.

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Sverzhevsky Otorhinolaryngology Clinical Research Institute, Moscow, Russia

Aim: To evaluate the efficacy of stapedoplasty as a method of hearing loss treatment in patients with stapes

tympanosclerotic fixation. Stapedoplasty in patients with chronic purulent otitis media (CPOM) is performed in 0.3% cases in the event of massive tympanosclerosis (TSC) and stapes tympanosclerotic fixation with its mobilization inefficiency.

Materials and Methods: The first group included 25 patients with CPOM and TSC who underwent a two-stage surgery (2nd stage – stapedoplasty). The second group included 25 patients with CPOM and TSC who underwent a multistage surgery with a typical stapedoplasty as a final step, but using a surgical CO2 laser. As a method of hearing loss treatment in patients with TSC 50 stapedoplasties were performed (27 vestibuloincudopexies, 19 vestibulomyringopexies, 4 vestibulomaleolopexies). To evaluate efficacy of stapedoplasty techniques, tonal threshold audiometry (TTA) in the frequency range of 0.125 to 8 kHz as a method of air conduction (AC) and bone conduction (BC) were performed before surgery, then – before discharge, and three more times – in 1, 6, and 12 months after surgery, also we calculate mean air-bone gap (ABG).

Results: Analysis data of ABG mean values (35.8±6.4 dB before surgery) in TSC1 group showed their decrease in 1 month up to 27.1±7.3 dB; in 6 months – up to 18.3±8.3 dB, and in 12 months – up to 13.3±9.2 dB. Hence, instrumental stapedoplasty was effective during whole follow-up period, but its best efficacy was determined in 12 months after surgery. ABG values < 20 dB were reached in 71.4% patients; from 20 to 30 dB – in 85.7%, and ≤ 30 dB – in 100% patients. Analysis data of auditory threshold by BC values (36.8±5.4 dB before surgery) showed their decrease in 1 month after surgery up to 44.1±5.5 dB; in 6 months – up to 17.0±8.0 dB, and in 12 months – up to 28.2±2.1 dB determined by inner ear reaction on surgery treatment. Analysis data of ABG mean values (35.3±8.6 dB before surgery) in TSC2 group showed their decrease in 1 month up to 22.1±9.5 dB; in 6 months – up to 17.0±8.0 dB, and in 12 months – up to 14.9±7.4 dB. Stapedoplasty with laser assistance was effective during whole follow-up period, but its best results were shown in 6-12 months after surgery. ABG values <20 dB were reached in 76% patients; from 20 to 30 dB – in 92%, and ≤ 30 dB – in 100% patients. Analysis data of auditory threshold by BC values (30.3±11.3 dB before surgery) showed their consistency in 1 month after surgery – 31.1±13.2 dB; in 6 months – 28.9±13.1 dB, and in 12 months – 28.3 ± 12.8 dB.

Conclusion: Stapedoplasty is effective with ABG decrease <20 dB in 73.7% TSC patients. Laser assistance during stapes crus crossing stage and stapedotomy in the early follow-up period results in absence of inner ear reaction for surgery; moreover, ABG mean values decrease more rapidly than these during instrumental surgery technique.

Non-surgical treatment of otosclerosis.

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Sverzhevsky Otorhinolaryngology Clinical Research Institute, Moscow, Russia

Many surgeons face with active forms of otosclerosis during stapes surgery. Frequency of cochlear complications increases and functional results of stapedoplasty decreases in case of active otosclerosis.

Aim: To reduce complications caused by active otosclerosis.

Materials and Methods: 959 patients with otosclerosis were examined in Sverzhevsky Otorhinolaryngology Research Institute from 2009 to 2017 year. Diagnosis was based on anamnesis, pure tone audiometry (PTA), tympanometry, measurement of ultrasound threshold and its lateralization, high resolution multispiral computer tomography with multi-planar reconstruction and densitometry (areas of otospongiosis with density less then +900 HU was marked as "active", normal density of bone labyrinth is +2000-+2200 HU). All patients with active otosclerosis receive special drug therapy (bisphosphonates, calcium, natrium ftoride, vitamin D). Treatment was regularly controlled by blood tests (Calcium free and bound, vitamin D, urea, creatinine), PTA and CT. Duration of treatment was selected invidually and was based on initial density of otosclerosis foci (1 standard course – 3 months).

Results: In 288 patients (30%) active otosclerosis was determined (28 - cochlear type, 221 – mixed type, 39 – tympanic type). Tinnitus was determined in 64 patients. In case of unilateral hearing impairtment by PTA data, signs of bilateral process were determined in 73,3% by CT data. Treatment was considered successful when the density of otosclerotic foci reached 1000 HU or higher. During the treatment air conduction (AC), bone conduction (BC) and air-bone gap (ABG) was regularly measured. In most cases this data was stable (5 dB). In 6 cases were matched improvement of BC and AC for 5-15 dB with the same ABG; in 14 cases – decreasing of AC with growth of ABG (BC were stable). 34% of patients reported reduction of tinnitus. Stapedoplasty were performed in 166 cases, after several courses of treatment, when the density of otosclerotic foci reached 1000 HU or higher and ABG was 30 dB or more. Functional results were good in all cases. Also, there were no signs of active otosclerosis during all 166 surgical procedures.

Conclusion: Special examination (including PTA, tympanometry, CT scan with densitometry) allows us to diagnose active forms of otosclerosis. Also, it helps us to form plan the drug treatment, its duration and and evaluate its effectiveness. Complex inactivating therapy stabilizes auditory function and creates condition for safe and effective stapedoplasty.

Laser stapedoplasty in case of otosclerosis: different types and results.

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Sverzhevsky Otorhinolaryngology Clinical Research Institute, Moscow, Russia

Objective: To compare the functional results of different types of Laser- assisted stapedoplasty.

Materials: 959 patients with otosclerosis were examined from 2009 to 2017 years. 837 patients were operated with different types of stapedoplasty, including 30 reoperations (the first operation was performed in various clinics of Russia). 365 were underwent stapedectomy (method of closure large window with autovenous graft and using cartilage graft to replace the stape, 173 - stapedotomy, 298 - modified stapedotomy (with large window), in which perforation hole in the footplate was closed with a autovenous grafts and a piston prosthesis.

Methods: To perform perforation in footplate we used CO2 laser (l-10,6). This instrument has a scanner system which works together with micromanipulator that connected to the microscope. It can generate laser pulses of extremely short duration and high peak powers. We used cartilage graft from the ear of a patient, titanium prostheses K-Piston (Heinz Kurz GmbH, Germany), titanium Piston-Ti (Audio Italy) and platinum-titanium The Big Easy™ Piston (Medtronic, USA). The length of cartilage graft was-3.5 mm, SD - 0.8-1.0 mm, piston prostheses - L - 4,25 mm, SD - 0,4-0,6 mm. Bone-conduction (BC) and air-bone gaps (ABG) were measured in all patient before surgery, 10-12 days, 1,3,6 and 12 months after surgery.

Results: After stapedotomy best outcomes (ABG ≤ 10 dB) being observed 6 months post-op in 78 % of patients. The best early functional results were received, using stapes prosthesis with diameter 0,6 mm after stapedotomy. After modified stapedotomy (ABG less 10 The dB) were observed after 6 month post-op in 92 %. In case of stapedectomy, with cartilage graft the best results (ABG less 10 The dB) were observed 3 month post-op in 96% patients with improvement of BC thresholds on 10-20dB. After surgery we deal with some complications. Immediate postooperative audiograms shows us sudden hearing loss in 7 patients (0.8%): 4 patients (0,5%) after modified stapedotomy, 2 (0,2%) – stapedotomy, 2 (0,2%) - stapedectomy with cartilage graft. Completely deaf in 2 (0,2%) cases. The most cases were identified as obliterating otosclerosis.

Conclusion: The study results shows us influence of size of footplate perforation and early isolation perilymph of vestibulum with an autovenous graft. The stapedectomy with cartilage graft to replace the stape and modified stapedotomy with piston on venous graft is more effective and safe, then stapedotomy.

Instructional Course: Otosclerosis: from standard to challenging cases.

Otosclerosis: from standard to challenging cases.

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The gradual decrease in primary stapes cases available to the otologic surgeon makes the possibility of unsuccessful procedures more frequent (especially in less experienced surgeons), thus revision stapes surgery may also become more frequent; the aim of this course is to show the surgical technique in detail from the standard to the more challenging cases.

The course will be divided in 2 parts: in the first one, after an introduction regarding etiology, physiopathology and clinical aspect of otosclerosis, cases of standard stapes surgery will be shown with the description of the surgical technique, results and complications as well as the use the laser, new stapes prosthesis and endoscope. In the second part, challenging cases (encompassing malleus head fixation, dehiscent facial nerve, narrow oval window, obliterative otosclerosis and round window ossification) will be described with videos and a case series of revision stapes surgery (erosion/absence of the incus, perilymphatic fistula) will be shown.

Saturday, 7th April

Lecture Session IV

Keynote Lecture

Classification and Surgical management of Tympanosclerosis.

Vijayendra H.

Vijaya ENT Care Centre, Bangalore, India

Introduction: Surgery for tympanosclerosis is most rewarding surgery as nothing can replace natural ear bones. It is more rewarding surgery than stapedotomy.

Objectives: As an otologist the aim is to solve the conductive type of hearing loss in tympanosclerosis by surgery.

Methods: With my vast experience of two decades after performing more than 250 cases of tympanosclerosis depending upon the surgical classification each case is tackled according to the pathology. At least in 85% of the cases, the footplate is made mobile (with documentation) and good hearing results are achieved.

Results: With my documented special surgical techniques in 85% of the patients servicable hearing results are achieved.

Conclusion: As per surgical classification i.e Myringosclerosis, attic tympanosclersosis, Stapes tympanosclerosis (Mild, Moderate, Massive), Mastoid tympanosclerosis, Tympanosclerosis associated with bony ankylosis and multiple combination of the above classification I have my own documented surgical techniques to make the ossicular chain mobile to achieve good hearing results, which is the goal of an Otologist.

Keynote Lecture

Congential isolated middle ear anomalies: surgical treatment tactics.

Diab Kh.M., Daykhes N.A., Korvyakov V.S., Gulyamov Sh.B., Kondratchikov D.S., Zagorskaya D.A.

Clinical Centre of Otorhinolaryngology of FMBA, Moscow, Russia

Aim: To increase the efficiency of surgical treatment of isolated middle ear malformations by developing and implementing optimal surgical tactics depending on the features of the middle ear structures.

Material and Methods: 21 patients with isolated middle ear malformations examined and operated at our clinic from January 2015 to October 2017. Indications for surgical treatment were: the presence of an isolated anomaly of the middle ear development with a hearing loss by conductive or mixed type II-III degree, indication for surgery were the presence of air-bone gap (ABG) more than 30 dB. In all patients, the main complaint was a non-progressive hearing loss from an early age, which is an important differential diagnostic feature for establishing the correct diagnosis. The general stage of surgical treatment for all patients was revision tympanotomy. The piston stapedoplasty performed to the patients with the stapes ankylosis. A fenestra nov-ovalis formed for the patients without OW and an ossiculoplasty with autocartilage performed. To assess the immediate and long-term functional results, the tone threshold audiometry repeated after removing swabs from the external auditory meatus and 4-12 months after the surgery.

Results: The absence of anterior limb of stapes, fixation of stapes were noticed in 7 patients; the absence of long limb of incus, stapes superstructure, fixation of its footplate were noticed in 1 patient; the absence of stapes head, the stiffness of stapes were recorded in 1 patient; the fusion of anterior and posterior limbs of stapes, the stiffness of stapes were recorded in 5 patients; the absence of stapes and OW was recorded in 5 patients; the absence of stapes, OW and long limb of incus was recorded in 1 patient; the absence of stapes and OW, long limb of incus and promontory mattered together were recorded in 1 patient. An anomaly of the location of the facial nerve canal, which consisted of its bifurcation, overhanging the OW or passing in the middle of the promontory was found in 7 patients. The ABG in patients undergoing stapedoplasty decreased to an average of 15.25 dB and remained at this level throughout the entire observation period. The formation of the fenestra nov-ovalis allowed to reduce the ABG to an average of 21.50 dB in the early post-surgery period. However, the bone-air interval of one patient increased to 35 dB in the late postoperative period, which may indicate the formed fenestra nov-ovalis ossification.

Conclusions: The patients with isolated middle ear anomalies are good candidates for hearing-enhancing surgery. Certain difficulties during surgery may occur in case of abnormal path of the facial nerve canal, but with an adequate choice of surgical tactics, it is possible to achieve good functional results. However, the cases of OW aplasia require modification of the applied surgical techniques.

Keynote Lecture

Revision stapes surgery.

Lavy J.

Royal National Throat, Nose and Ear Hospital, University College London Hospitals, The London Ear Clinic

In the assessment for revision stapes surgery the history is essential for predicting the potential for a good hearing

outcome. A wide variety of conditions can be encountered in the middle ear and the surgeon needs to be prepared for every eventuality. Advances in material sciences including piston design and non-ototoxic bone cements have boosted the success rate of this challenging surgery. A very practical approach to revision stapes surgery will be presented with emphasis on how to achieve the best results with minimal complications.

Keynote Lecture

Revision surgery in otosclerosis.

Szymański M.

Department of Otolaryngology Head and Neck Surgery, Medical University of Lublin, Poland

Stapedotomy is a highly successful and safe procedure, however, in case of complications or bad hearing result patients require revision surgery. Management of different clinical situations like erosion of the long process of incus, prosthesis dislocation, perilimphatic fistula is discussed. The use of KTP and CO2 laser in revision surgery is also described.

Keynote Lecture

Morphologic basis of stapes surgery.

Djeric D.

Academy of Medical Science of the Serbian Medical Society, Belgrade, Serbia

The goal of stapes surgery is to re-establish sound transmission through an ossicular chain stiffened because of otosclerosis. Various techniques have been used to accomplish this goal, including stapes mobilization, fragmentation, small fenestration, and partial or total stapes footplate removal. Even at the beginning of stapes surgery, authors emphasized the significance of anatomical factors. The success of stapedoplasty depends on the extension of otosclerotic process, surgical techniques, experience of the surgeon and morphological variations and relations in the operative filed. Stapes surgery is one of the most standardized and successful procedures in otology. Many discussions and publications have dealt with successful results obtained by numerous surgeons. A precise knowledge of anatomical characteristics has great influence , because of stapes surgery already carries some risk of complications (additional hearing loss, tinnitus, vertigo, facial nerve palsy and other).

Morphological variations and relations that are seen during the operation may predispose the occurrence of complications. Special attention should be given to the anatomical variations related to the stapes , the oval window and the facial nerve. The deeply located narrow oval window and various facial nerve anatomic variations are the problem that are met commonly during primary operations. Visualization of the entire oval window niche and identification of the facial canal is very important during stapedotomy. The facial canal and nerve may show a morphological variations and anomalies (abnormal course, dehiscence, prolapsed and bifurcations of the nerve and other).

The purpose of this paper is to review an important anatomical variations and relations in the middle ear that may occur on the human temporal bones. We presented findings of specimens that demonstrated a morphological variations involved: the facial nerve canal, area of the oval window niche, ossicular chain, and upper jugular bulb and point out of their importance in stapes surgery. The studies were performed on the large collection of the temporal bones (2000 temporal bone specimens) in our laboratory for anatomical and surgical examinations, for many years.

In conclusion, better surgical techniques are not always sufficient to receive good functional outcome from stapes surgeries, and the surgeon should also be aware of possible variations that affect the result of the surgery.

Implants in Treatment of Otosclerosis

Evaluation of the effectiveness and efficacy of the CoDACS system in patients with otosclerosis.

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World Hearing Center, Institute of Physiology and Pathology of Hearing, Warsaw/ Kajetany, Poland

Aim: The study aims at preliminary evaluation of the effectiveness and efficacy of the CODACS system in patients with otosclerosis.

Material: The material consists of 4 patients with diagnosed otosclerosis and severe to profound mixed hearing loss after many surgeries, who were implanted unilaterally with CODACS system. Procedures were performed in the Institute of Physiology and Pathology of Hearing.

Method: The free-field audiometric tests (free-field audiometry and speech audiometry) were performed as part of efficiency assessment of the applied solution. They were used to evaluate hearing improvement. The tests were performed before the implantation and in a few intervals after the system had been activated.

Results: The outcomes of audiometric evaluation in free field indicates lowering of the hearing threshold and enhancement of speech discrimination while using the CODACS system. Analysis of the APHAB questionnaire revealed subjective benefits in hearing and speech discrimination.

Conclusions: The gathered preliminary findings indicate that using the CODACS system may efficiently affect hearing improvement.

Power Stapedectomy with Vibrant SoundBridge SP coupler in Mixed Hearing Loss.

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The therapeutic options in otosclerosis are limited to medications (acute disease/active symptoms) as well as the rehabilitation of the hearing loss (HL). Rehabilitation can be with hearing aids (HA), stapedectomy, middle ear devices, osseointegrated implants and even with cochlear implants, depending on the HL degree. Conventional HA stimulates the hearing acoustically, with sound amplification, so there are feedback limitations, distortions, limited auditory gain, eczema of the external auditory canal and other disadvantages. HA advantages remain on aesthetic aspects, it does not require any surgical approach, with less morbidity.

In MHL, the ideal is to combine technologies to achieve a complete improvement of hearing. Therefore, the option of middle ear/osseointegrated implants is a great solution, already well established in the world medical literature as one of the gold standard treatments for the correction of mixed/moderate hearing loss. Such devices achieve great auditory benefits in cases of MHL of mild/moderate degree, because of the power of a mechanical auditory stimulation technology. This allows them to provide a great auditory gain without sound distortion and with great sound quality. Their disadvantages lie in not being as aesthetic compared to HA and the need for the patient to undergo surgery, which implies obvious risks. The therapeutic options in your health and health, as well as the hearing rehabilitation. The patient can be rehabilitated only with hearing aids, stapedectomy surgery, AMEI or OI and even CI, one dependent on the degree of deafness and involvement of his hearing. It is worth mentioning that conventional hearing aids stimulate hearing in an acoustic way, with an amplification of the children, on the other with limitations of feedback, distortions, difficulty in obtaining adequate auditory gain, eczema of the external auditory canal, within other limitations. Its advantages are more aesthetic and does not require any surgical surgery, which greatly reduces the risks for the patient. The therapeutic options in otosclerosis are limited to medications (when the disease is in its acute stage, with active symptoms) as well as the rehabilitation of its hearing.

Materials and Methods: To evaluate the surgical and clinical outcomes of the Treatment for MHL in Otosclerosis with Stapedectomy & VSB. All patients receive a SP (short process) coupler of VSB/Medel and a stepedectomy (teflon prostheses/0.4 mm).

Results: Five patients underwent to surgery (stapedectomy with VSB SP) for treating MHL in Otosclerosis. All patients get closed the air-bone gap with the stapedectomy and get normal pure tone thresholds after activation of the VSB. Excellent vocal and speech outcomes. There were no surgical or audiological complications until now. The follow up time is 16 months.

Conclusion: Stapedectomy & VSB is a good option for surgical rehabilitation for treating MHL in Otosclerosis.

Cochlear Implantation in Adult Patients with Far Advanced Otosclerosis - Systematic Review.

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Introduction: Otosclerosis is a chronic pathological process of the labyrinthine capsule, which leads to the conductive or mixed hearing loss. Typically it starts in the 4th or 5th decade of life, but pathology has a tendency to gradually worsen so it is possible that the first otosclerosis changes occur earlier. The pathomechanis of otosclerosis is multifactorial and incompletely understood, the most frequently mentioned causes of the disease include genetic predisposition, viral infections and various types of disorders: immunological, hormonal, mineral and enzymatic activity. The treatment (stapedotomy) enables restoring the normal functions of the ossicular chain and gives a chance of eliminating or reducing hearing loss. In patients with far advanced otoslerosis, when stapes surgery failed, cochlear implantation should be considered.

Aim: The aim of this study was to analyze of the benefits from cochlear implantation in patients with far advanced otosclerosis.

Material and Methods: This study was prepared as a result of literature review. The preliminary analysis covered 120 works. After the selection of the articles, some of the works were removed, including the duplicates, non-Polish and non-English works, articles about ossification of the cochlea due to meningitis as well as cochlear implantation in children. Ultimately, 40 pieces of literature were qualified.

Results: Cochlear implantation in patients with far advanced otoslerosis leads to statistically greater improvement in speech recognition scores than stapedotomy. However it is worth mentioning that outcomes after surgery are highly variable and speech recognition results were similar in patients with cochlear implants and group of successful stapetodomy.

Conclusion: Stapes surgery is a simple and effective method of otosclerosis treatment and should be considered before cochlear implantation. Cochlear implant seems to be a good option for patients with poor or none benefits after stapedotomy.

Basilar membrane excitation mechanism in bone conduction hearing pathway in ear with otosclerosis of the oval window.

Tudruj S., Kamieniecki K., Piechna J., Borkowski P.

Warsaw University of Technology, Warsaw, Poland

Aim: The aim of the work is to understand the mechanism of the basilar membrane (BM) excitation in the bone conduction hearing pathway in the ear with the otosclerosis of the oval window. The investigation was preformed using a numerical simulation.

Material and Methods: The cochlea is modeled as uncoiled. The model consists of an oval window (OW), a round window (RW), a BM, a scala vestibule (SV), a scala tympani (ST) and a helicotrema. Interaction between structural and fluid parts (the perilymph) is taken into account. The cochlear scalas are represented by a three dimensional canals. The BM is modeled as a set of resonators tuned to different frequencies. The model is excited by applying a harmonic velocity to the rigid cochlea's walls in the direction along the basilar membrane (X) and perpendicular to the BM (Z). Oscillating walls of cochlea transmit the boundary motion to fluid inside. The model was calculated for 10kHz excitation frequency.

Results: Results of the models are the pressure – time histories in the cochlea scalas and the displacement – time histories of the basilar membrane. Based on the analysis, it was shown that the vibration amplitude for the direction perpendicular to the BM is much smaller than the amplitude for the excitation direction along the BM. Referring to the time domain solution it was shown that this behavior is related to the geometrical and mechanical properties of the cochlea with otosclerosis of the OW.

Conclusions: In bone conduction hearing pathway the largest amplitude of the BM vibrations is for the excitation in the direction along the BM. This fact is related due to the geometrical and mechanical properties of the cochlea in the otosclerosis state, especially due to a stiff OW. Understanding a mechanism of such behavior can be helpful in design of hearing recovery devices in otosclerosis. It is expected that the behavior of the cochlea in the normal state (both windows has ability to move) will be different.

Time domain one and three dimensional models of the cochlea.

Kamieniecki K., Piechna J., Tudruj S., Borkowski P.

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Aim: To design good and efficient hearing recovery devices, for example middle ear prostheses, it is necessary to properly understand the cochlear physiology. The experiments are one of the ways to gain the knowledge but they are difficult to conduct due to difficult to access location of the inner ear, small size of the inner ear and due to the very dynamic character of the phenomena in the inner ear. A tool which can complement the experimental knowledge are the numerical analysis. The aim of the work is to present two numerical models of the cochlea, one dimensional (1D) and three dimensional (3D), developed in the time domain and to show the advantages of numerical modeling in understanding the cochlea's operation.

Material and Methods: 1D and 3D approach the cochlea is modeled as uncoiled. In 1D model the velocity profile and the pressure are assumed to be uniform across the cross section of the cochlear scalas. The basilar membrane (BM) is modeled as a set of resonators tuned to different frequencies. In 3D model the velocity profile and the pressure distribution at cross sections area are three and two dimensional fields respectively. The BM is modeled as a solid, thin membrane, fixed at the boundaries. The models were excited by applying a harmonic oscillations to the stapes footplate (SF) of the oval window (OW). A coupling between the fluid (perilymph) and the structural parts (BM, OW and round window) is taken into account.

Results: Results of the models are the pressure – time histories in the cochlea scalas and the displacement – time histories of the basilar membrane. As the unsteady process is simulated in the time domain, it is possible to observe a mechanism of the basilar membrane excitation by a pressure waves traveling in the perylimph.

Conclusions: The presented models are able to mimic the physiological cochlea operation according to the literature data [Bekesy 1960, Stenfelt 2003]. As the models, described the hearing physics in the time domain, they are helpful in better understanding the cochlea's operation, especially in the first moments after applying the excitation. The three dimensional model can be used to assess the effectiveness of the middle ear prostheses (piston or chamber prostheses) or investigate the cochlear macro – mechanics in the bone conduction hearing path way.

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Is the human cochlea and basilar membrane length a coincidence?

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Aim: The inner ear, especially the cochlea, is one of the most sophisticated human sense organs. It is able to precisely distinguish thousands of tones. The cochlea has a specific length which determines the length of the basilar membrane. It is interesting whether the dimensions of the cochlea scalas and then the basilar membrane are the coincidence or not? The aim of the work is to answer why the human's cochlea canal length and basilar membrane length equals to approximately 31.9mm and how does this affects hearing. Additionally it was checked if the inner ear

size is related to the body size or to the hearing frequency range. The proper understanding of the influence of the cochlea's geometry on the mechanics of hearing may be helpful in better treatment of the conductive hearing loses.

Material and Methods: To answer the above question, a one dimensional model of the cochlea was used. The model assumed that the scala vestibule and the scala tympani are a pipes of a constant cross section. A coupling between the perylimph and the basilar membrane and the pressure wave reflection in the helicotrema are taken into account. The model was excited by a harmonic vibration of the stapes footplate of the oval window.

Results: The results of the analysis showed that the human cochlea and basilar membrane lengths are not a coincidence. The length equal to 31.9mm enables a specific pattern of the pressure wave distribution along the cochlea. This leads to amplification of the basilar membrane vibrations at the high frequencies and to decreases vibrations at the low frequencies.

Conclusions: The human cochlea and the basilar membrane lengths are not the coincidence. The length of 31.9mm allows on amplification of the basilar membrane vibrations at high frequencies (10kHz) and to decrease the vibrations at low frequencies (0.4kHz). This is consistent with the basilar membrane geometrical and mechanical properties. In the basal end, where the high frequencies are sensed, the basilar membrane is narrow and thick, while in the apex it is wide and thin. Thus at the base the membrane is stiff and the amplification is needed, while at the apex it is flexible, thus the amplification is not necessary. Additionally the length of the human basilar membrane was compared with other mammalians. This also confirms that its length is not a coincidence but it is rather related to the hearing frequency range not to the body size.

Bone conduction stimulator position and direction investigation using numerical simulation.

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Aim: The goal of this work was to determine the amplitude of the harmonic force imposed by the bone conduction stimulator located in the temporal bone by using finite element analysis. The forces were sought that caused the same vibrations of the round window and basilar membrane as those obtained for the cochlea stimulated by the stapes. The stimulator was placed at the surface of the labyrinth capsule above the lateral semicircular canal and in the center of the anterior semicircular canal loop and at the outer surface of the squamous part.

Material and Methods: The geometrical model of the temporal bone was built using CT imaging. Over 12,000 surfaces were used to describe the bone and fluid spaces. The finite element analysis was performed in ANSYS system. The model with the excitation applied on the stapes was validated on the basis of the literature. Then a harmonic analysis for the frequency range from 0.4 to 10kHz was performed to determine the amplitude-frequency characteristics used as the reference to assess the efect of the stimulator. In the bone conduction analysis the stimulator was represented by a lumped mass. The two force amplitudes were considered which caused the amplitudes of vibration close to those obtained for the cochlea stimulated by the vibration of the stapes. The first force amplitude for bone conduction concerned the round window vibration and the second was related to the basilar membrane.

Results: For the bone conduction analysis the six directions of the harmonic force applied at the point located on the labyrinth capsule over the lateral semicircular canal were considered, assuming that the directions were available after the mastoidectomy. For the fixed stapes (otosclerosis) there was a relationship between the direction of the force and cochlear response. The force amplitudes were decreasing with the increase of frequency. The next simulation of bone conduction was performed for the stimulator placed in the center of the anterior semicircular canal loop. The four depths from 0 to 8 mm were considered assuming the fixed force direction. The force amplitudes that caused the same vibration amplitudes on the round window and basilar membrane as obtained for the cochlea stimulated by the stapes were approximately one order lover than those obtained for the stimulator situated at the reference point located at the outer surface of the squamous part. The force amplitudes decreased about twice when the point of excitation was moved to the labyrinth capsule over the lateral semicircular canal. The increase of the implantation depth caused the increase of the force amplitudes.

Conclusions: The results of FE analysis showed a significant influence of the force direction and the position and depth of the implantation on the effectiveness of the bone conduction stimulation.

Assessment of wideband absorbance in patients with middle-ear implants: A preliminary study.

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Background: Wide Band Tympanometry is a reliable methods for assessment of middle-ear mechanoacoustic properties. The presence of middle-ear implant, e.g. FMT, alters ear impedance and absorbance characteristics. This problem has not been investigated yet.

Material and Methods: Energy absorbance (EA) was measured, along with other characteristics, in a group of patients qualified for middle-ear implants. The tests were repeated in postoperative period and the changes in EA properties were assessed.

Results: The presence of FMT changes middle-ear impedance and influences energy absorbance in a specific frequency range. Low-frequency and high-frequency EA remains virtually unchanged. An influence of FMT fixation method on EA is also observed.

Conclusions: Middle-ear implant, of FMT type, influences acoustic impedance and absorbance characteristic due to the increase of mass of the vibrating ossicular system. The observed effects conform qualitatively well to those predicted with the middle-ear model.

Lecture Session V

Keynote Lecture

Age and otosclerosis.

Tange R.A.

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The onset of hearing problems due to otosclerosis is often difficult predict. In the 20's the majority the disease otosclerosis commenced between 21 and 25 with hardly any appreciable difference in the sexes. The beginning of deafness started in 50% of the cases in the period between the 16th and 30th year according to Nager(1927). Now, almost a century later the onset of the symptoms of otosclerosis seem to appear in a later fase of life. Recent studies have demonstrated that the disease otosclerosis commenced between 36 and 45 of age. The age onset of the symptoms of otosclerosis has been increased during the last 80 years. The reason for the age onset increaseof otosclerosis is still unclear.

Keynote Lecture

Flying and diving after stapes surgery.

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The ear works as a pressure receptor. A piston prosthesis, which significantly modifies the function of the normal ossicular chain, will be displaced at changes of the atmospheric air pressure quite differently as compared to the normal stapes. These pressures may a reach significant dimensions in daily life, as during swallowing, with tubal opening, wind gusts at the external ear. Extreme pressures can occur during flying and diving, a condition, which evolution had not foreseen for the development of our ears.

Fortunately, the middle ear is designed to compensate even elevated and rapid changing static pressures. This could be shown in temporal bone experiments with measurements of stapes displacement during tympanometry. These pressures, which are the maximum driving force also during physiological stresses, due to the stability of the collagen fibers in the tympanic membrane at higher pressures, can displace a piston up to 0,5mm in the oval window. These movements are transmitted from the pressure-induced displacement of the tympanic membrane, which can reach up to 1000 µm, and the lack of the attachment of the piston in the annular ligament, unlike the normal stapes. This movement can explain the risk of a short piston to be lifted out of the footplate perforation (for example after sneezing). In case of an underpressure in the middle ear, or a pressure rise in the external ear canal, an impalement of a piston with excessive length into the membranous labyrinth can occur. The inward movement of the piston can cause vertigo, as can be experienced also to during stapes surgery in local anesthesia.

In conclusion of the experiments, flying or diving can be allowed for patients after stapesplasty, if the piston had been placed in the posterior part of the oval window niche, where the distance towards the saccule and utricule is more than 1 mm. A further control can be obtained in surgery under local anesthesia: If the pressing down of the piston in its attachment to the lenticular process towards the inner ear and in its correct place does not provoke a nystagmus nor vertigo, no danger can be assumed in the postoperative life due to the stability of the ligaments of the ossicles.

Postoperatively, a test can confirm the lack of danger, if the patient wants to fly or dive after surgery: A tympanometry with fast changes between 400 mm positive and negative pressure, with continuous recording of the eye movements in ENG. If the recording is nystagmus free, and if the patient does not describe any vertigo, no postoperative risk must be anticipated for flying and diving after stapesplasty.

Keynote Lecture

Patient satisfaction after stapes surgery for otosclerosis.

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Department of Otorhinolaryngology, Iuliu Hatieganu University of Medicine and Pharmacy, Cluj-Napoca, Romania

Aim: Health-related quality of life after stapes surgery has become a major concern nowadays as in other areas of the otology field. Therefore, correct reporting of results requires more complex data both technically (depending on technique and surgeon) and functionally (depending on patient satisfaction). The objective of this study was to review the current methods of reporting results after stapes surgery, and to correlate the health related quality of life outcomes after surgery with technical audiometric parameters.

Materials and Methods: A prospective study on 230 patients who underwent stapes surgery for otosclerosis was conducted at Cluj-Napoca University Hospital between Jan 2015 and Jan 2016. The audiometric results, at 12 months after surgery, were analyzed according to the guidelines of the Committee on Hearing and Equilibrium of the Academy of Otolaryngology-Head and Neck Surgery (AAO-HNS), and Glasgow plot. Surgical trauma to the inner ear was also evaluated using the Amsterdam plot. The subjective hearing

disability was evaluated by the hearing handicap inventory for adults and the Tinnitus Handicap Inventory. The quality of sound after surgery was also evaluated.

Results: Technical hearing results after surgery were good after surgery (we achieved a closure of the ABG to 10 dB in 70 % of all cases and to 20 dB in 92 %, of all cases, with an AC gain of 24 ± 10.00 dB. The postoperative quality of life has improved in terms of tinnitus score, social and emotional subjective hearing disability and the quality of sound scores. No significant correlations were found between some hypothesized predictive audiometric outcomes and the quality of life improvement.

Conclusions: Uncomplicated stapes surgery represents an effective surgical procedure for the treatment of otosclerosis which leads to improvement in patients' quality of life.

Keynote Lecture

How to Learn Stapes Surgery in Cadaver.

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Stapes surgery is one of the most delicate surgery in ear surgery. As middle ear and tympanic membrane are absolutely normal, utmost importance should be given to this surgery. In cadaveric temporal bone, one should learn stapes surgery very meticulously as if it is being done in live patient. Using proper and quality instruments is key in learning stapes surgery. We should have good set of all instruments. In cadaver, reverse stapedotomy is done as most of the time footplate will be normal and if we try to remove suprastructure before inserting piston, footplate will get dislodged and procedure won't be possible. I create tympanomeatal flap via transcanal approach and secure it anteriorly using wet saline soaked gelfoam. Because as a beginner one will take more time to perform procedure. By the time procedure gets over, flap which has been raised in beginning will shrink because of dryness and then will be difficult to approximate. To obtain exposure, I curette posterosuperior canal wall. For adequate exposure and easy insertion of piston superiorly facial canal and posteriorly pyramidalis process should be visible. Chorda tympani nerve if coming in way then should be decompressed and secured away from working area so that it won't come in way during the procedure. I always first make controlled fenestra of 0.3mm in posterior half of the footplate and then gradually it is widened to 0.6mm. Fenestra should be always made in posterior 1/3rd of the footplate in order to avoid injury to saccule and utricle which are placed near to anterior part of footplate as compared to posterior part. Using measuring rod distance between the long process of incus and fenestra is measured. Use lower magnification to insert piston as both fenestra and long process of incus should be visible in one view only. Teflon piston of 0.4mm diameter inserted into fenestra, anchored to long process of incus and crimped. Bend test and Lift test is performed to ensure that Piston length is correct. In Bend test when shaft of piston is bent, piston should not come out of fenestra. In Lift test, while lifting incus, piston should not come out of fenestra.

Ossicular Prostheses

Keynote Lecture

The efficiency of self-crimping superelastic Nitinol/PTFE piston and cartilage prosthesis in otosclerosis surgery: our experience.

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Aim: To analyze the effectiveness of stapedoplasty with self-crimping superelastic NITINOL/PTFE (Nickel and Titanium alloy / polytetraflorethylene) piston-prosthesis and methodology of "auto-cartilage-on-vein" placement.

Materials and Methods: 165 patients (182 ears) with otosclerosis, who underwent stapedoplasty, were included in this study. We used self-crimping superelastic NITINOL/PTFE piston-prosthesis in 143 cases. Piston-type stapedoplasty was performed in cases of simple non-complicated forms of otosclerosis without incomplete dislocation of incus, facial nerve overhang, obliteration of oval window and cochlea. In cases when patient had certain anatomical features (dehiscence and overhand of facial nerve, obliteration of oval window, obvious perilymph leakage), in cases of revision surgeries (aseptic necrosis of process) or in cases of unintentional mobilization of stapes foot we practiced methodology created in the Russian Federation – that is total platynoctomy with stapedoplasty using "auto-cartilage on vein" placement (39 cases). Shaped oval window after platynoctomy is covered with auto-vein sized 3*5 mm and is pressed by auto-cartilage prosthesis, fixed under lenticular incus process, that helps to provide total hermitization of inner ear. Pure tone audiograms analyzed to evaluate the functional results.

Results: The average air conduction was 31,5±5,6 dB before surgery and significantly decreased to 11,5±3.2 dB at 6 months postoperatively in patients who underwent stapedoplasty using a NITINOL/PTFE piston-prosthesis (n=130). In 10 cases, the piston in Titanium was use due to thick long incus process. Analysis of the functional results showed a statistically significant (p <0.05) decrease air-bone gap to 10,2 ± 2.3 dB (before operation 34,2 dB) in patients who underwent stapedoplasty using methodology of "auto-cartilage-on-vein" placement. Displacement of piston prostheses were not observed during research study. In 4 cases (3 cases after piston type stapedoplasty and 1 case after stapedoplasty with cartilage) development of sensorineural hearing loss was observed.

Discussion and **Conclusion:** Usage of NITINOL/PTFE piston-prosthesis decrease time of surgery and consequently impact on inner ear's structures. The hook elasticity reduces the surgery steps and the difficulties that may occur during the closing phase. The hook closes softly, uniformly

wrapping the pressure along the total periphery of the long incus process, and minimize the risk of compressive necrosis. Positive aspect of stapedoplasty using methodology "auto-cartilage on vein" placement is possibility to shape auto-cartilage prosthesis (creation of auto-cartilage with a certain angle position) that helps to go round overhang of facial nerve, saving the pressure on it. Observation and analysis of surgeries showed high effectiveness of mentioned treatment methods, that allows ENT surgeon to have certain algorithm of his activities in situation of any difficulty.

Audiological results of the new Nitinol Stapes Prosthesis.

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Aim: Stapes plasty is a well-established method for improving hearing in case of otosclerosis. Continuous optimization leads to periodically new developed prosthesis. The aim of the current study was to evaluate the audiological results of Stapes plasty with new Nitinol stapes prostheses.

Material and Methods: The surgeries were performed between May and November 2017. Nitinol Prostheses were implanted in the patients. Preoperative and postoperative pure tone audiograms of patients were evaluated. Postoperative measurements were taken 3 weeks after surgery. Furthermore, data on the intraoperative surgical method and possible complications were recorded. As a control group, 10 patients were selected, who were treated by the same surgeon with a conventional prosthesis.

Results: To date, nine stapes plastic surgeries using the Nitinol Alloy prosthesis were evaluated. The control group consists of patients with conventional Stapes plasty. The postoperative average air-bone gap in the study group was 13.5 ± 3.9dB (MV ± SD) while that of the control group was 13.9 ± 7dB (MV ± SD). Moreover, the average reduction in the air-bone gap was 18.5 ± 9.4 dB (MV ± SD) in the study group and 12.7 ± 3.5 dB (MV ± SD) in the control group. Intraoperative and postoperative serious complications were not observed in both groups.

Conclusions: Stapes plasty using the new Nitinol prosthesis is a feasible procedure in otosclerosis surgery with less conductive hearing loss. As this prosthesis needs not to be crimped intraoperative, it represents a practicable alternative to conventional prosthesis.

Keynote Lecture

Principle of work of the chamber prosthesis and its realization.

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Aim: The principle of operation and construction of new chamber stapes prosthesis is shown. Its task is to transfer accurately the image of the sound wave from the external ear to the inner ear. Commonly used piston prostheses do not meet that requirement. Due to the small contact surface of the piston with the perilymph, these prostheses transform the plane wave in the ear canal into a dispersed spherical wave in the cochlea.

Materials and Methods: In order to generate in the cochlea an image of a sound wave the same as that entering to the ear canal, a device known in the optics as a 'camera obscura' was used in the chamber prosthesis. A diaphragm with a small opening, placed between an illuminated object and a screen, focuses light wave running from the object, creating an inverted image on the screen. This idea was applied to the sound wave. A design of the chamber prosthesis design, which focuses the sound wave at the entrance to the cochlea, has been prepared. It consists of five elements: a chamber (1) filled with physiological fluid, a disc (2) suspended on a membrane (3), a pendant (4) connecting the disc with the malleus and a protective cover (5). The basic conditions to be met prosthesis, have been formulated. It was assumed that the main elements of the prosthesis will be made by 3D printing.

Results: With help of the Autodesk Inventor program, few proposals of the chamber prosthesis have been prepared. The chamber, shield and cover of the prosthesis was planned be made by 3D printing from a biocompatible polymer. The pendant will be prepared from a platinum wire. Two ways of connecting the pendent with the disc were proposed. Three designs of the membrane made of a thin Teflon film or a flexible silicon hydrogel have been analyzed.

Conclusions: The proposed solution for the middle ear prosthesis has the following features:

– the prosthesis provides a tight seal to prevent infection after surgery;

– the proper size of the prosthesis enables a safe surgery, in particular it allows to avoid a damage of the facial nerve;

– the stiffness of the membrane is close to the stiffness of the stapes ligament;

– the contact surface of the disc with the cochlea fluid is a spherical surface, to avoid an absorption of the sound energy by the chamber walls;

– the plane of the chamber outlet opening is inclined to the plane of the basilar plane to ensure energy transfer deep into the cochlea interior; in a healthy ear, this transfer ensure the stapes footplate rotations. The main obstacle in the realization of the project are small dimensions of the prosthesis elements. We have hope that a new 3D printer built at our Institute and adapted to manufacture the chamber prosthesis, will solve that problem.

Prospective clinical study on the influence of stapes prosthesis diameter on hearing outcomes.

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Objectives/Hypothesis: To evaluate the influence of the diameter of stapes prosthesis on functional outcomes in stapes surgery.

Study design: Prospective cohort study.

Methods: Fifty consecutive small fenestra stapedotomies performed using a 0.4-mm-diameter prosthesis were compared with 50 consecutive small fenestra stapedotomies carried out using a 0.6-mm-diameter piston. Audiological assessment following the recommendations of the Committee on Hearing and Equilibrium was performed 1 month after surgery. Postoperative complications between the two groups were noted.

Results: There were no statistically significant differences in demographic data between the two groups, and no differences in preoperative bone-conduction (BC) or air-conduction (AC) hearing thresholds for all frequencies (analysis of variance [ANOVA] and $\chi 2$ tests). No differences were found in the mean preoperative BC and AC pure-tone average and air-bone gap (ABG). In the postoperative evaluation, a statistically significant difference was found for the mean AC gain (20 ± 8.7 vs. 24 ± 11.5, P = .042, ANOVA) as well as for the postoperative AC threshold at 0.125 and 0.25 kHz and the postoperative BC threshold at 0.25 kHz (P < .01, ANOVA). A postoperative ABG ≤10 dB was obtained in 90% and 94% of patients in the 0.4-mm- and 0.6-mm-diameter piston groups, respectively (difference not significant, $\chi 2$ test). No postoperative dead ear and/or sensorineural hearing loss was noted in either group.

Conclusions: The 0.6-mm piston allowed a statistically significant higher AC gain compared with the 0.4-mm diameter piston. A larger diameter piston may be preferable if there are no anatomical or technical reasons that would favor a smaller prosthesis.

Keynote Lecture

The efficiency of prosthesis with extensible (adjustable) stem in ossicular chain reconstruction: our experience.

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Aim: To analyze the effectiveness of surgical treatment with ossiculoplasty of patients with chronic suppurative otitis media operated in a single clinical center from 2015 to 2017.

Materials and Methods: 263 patients (296 ears) with chronic suppurative otitis media, who underwent tympanoplasty with ossiculoplasty, were included in this study. We used partial titanium ossicular prosthesis in 170 cases and total titanium ossicular prosthesis in 126 cases. We used prosthesis with extensible (adjustable) stem (the stem is extensible through the traction and shortable through the compression), that showed good sound wave conductivity by oscillometric tests (unchanged conductivity of the sound wave in all stem configurations). The effectiveness of surgical interventions were assessed as short-term (up to 3 months post-operatively) and long-term (6 to 12 months) anatomical and functional outcomes. The anatomical results considered satisfactory if there was a well-formed mobile neotympanic membrane, air tympanic cavity and dry postoperative cavity. Pure tone audiograms analyzed to evaluate the functional results.

Results: Satisfactory anatomical results obtained in 93.5% of patients with the safe type suppurative otitis, 88.9% of patients with the unsafe type and 91.2% of patients after a revision surgery. The average air conduction was 33,7±7,1 dB before surgery and significantly decreased to 17.5 ± 5.8 dB at 6 months postoperatively in patients who underwent ossiculoplasty using a partial ossicular prosthesis with satisfactory anatomic results. Analysis of the functional results showed a statistically significant (p<;0.05) decrease average air conduction to 35.5 ± 5.7 dB (before surgery 57.1 ± 5.3 dB) and air-bone gap to 18.5 ± 2.3 dB (before surgery 36.5 dB) in 91.4% of cases in patients who underwent ossiculoplasty with a total prosthesis with satisfactory anatomical results. Displacement of total ossicular prostheses was the main cause of poor functional results (9 cases). The most common causes of unsatisfactory anatomical results were perforation (15 cases), cholesteatoma recurrence (3 cases) and lateralization of the neotympanic membrane (3 cases).

Discussion and **Conclusion:** Formation of a reliable sound conducting system with ossicular prostheses allows for persistent improvement of hearing. Various surgical techniques, such as extended posterior tympanotomy, endoscopic assistance, provide a good effect with the removal of non-aggressive cholesteatoma while preserving the bone structures, which are not involved in the disease. A complete removal of an advanced aggressive cholesteatoma with the opening of the temporal bone cell system ensures good functional and anatomical outcomes, makes it possible to prevent the spread of the pathological process and development of intracranial complications.

Keynote Lecture

The nature of vibrations of disc suspended on flexible rings in the chamber prosthesis.

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Aim: Due to small dimensions of the chamber prosthesis, the basic difficulty is a way to hang the prosthesis disc. A suspension of the disc on a membrane is the most popular solution. Linear vibrations of the membrane enable

transfer of the sound wave from the outer ear to the inside of the chamber without any disturbances. However, to form the membrane a separate process is necessary. Our aim is to investigate the character of vibrations of the disc suspended on two flexible rings instead of on the membrane. Such rings, alike the other elements of the prosthesis, could be produced by 3D printing. In addition, due to an increasing rigidity of the rings with the sound amplitude, such a suspension can create a natural protection of the ear against an excessive noise.

Materials and Methods: Two rings between the disc and the cover and the prosthesis chamber are made of a soft hydrogel with viscoelastic properties. Slightly concave contact surfaces of the cover and the chamber with the rings fix the rings position. The fixed distances between the contact surfaces of the rings with the cover and the prosthesis chamber cause the rings to pre-squeeze. When the disc oscillates, its cyclic displacements w(t) are transferred to the both rings by additional forces P(t) on the contact surfaces. These forces should be much smaller than the forces P0 caused by their initial squeeze w0 of the discs. According to the Hertz contact theory, a force P(t) compressing a cylindrical ring is proportional to the square of the pressure points displacement w(t). Because the displacement w(t) is much smaller than the initial squeeze w0, one can assume a linear approximation of the relation between P(t) and w(t).

Results: A nonlinear equation for forced and dumped vibrations of the disk has been formulated. Its linear vibrations was found when the equation has been linearized. It was noted, that if the initial compression of the ring is too small, some nonlinear disk vibrations may appear. They may have a fractal character, a typical one for chaotic vibrations. For a disk suspended on the pre-compressed ring, vibrations are more stable, but they still may have an anharmonic character.

Conclusions: A disc suspended on a flexible ring is sensitive to vibrations caused by sounds of the low intensity and stops when sound of the high intensity appear. On the other hand, an influence of non-linear vibrations of the disc may disrupt the sound wave transmitted from the external ear to the cochlea. It should be examined whether the influence of these disturbances, in the range of amplitudes of the sound wave received by the human ear, is significant.

Audiometric results after stapedotomy with KURZ Skarżyński prosthesis.

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Otosclerosis is disease of the bones of the middle ear and inner ear. Vast majority of patients presents conductive or mixed hearing loss with or without tinnitus, sometimes with vertigo. Very often the first symptom is tinnitus. Purpose of our study was to asess hearing results and complications after stapedotomy performed using titanum prosthesis.

After 25 years of succesfull experiences with platinum prosthesis surgeons from World Hearing Center of Institute of Physiology and Pathology of Hearing in Warsaw started performing stapedotomies with KURZ Skarżyński Pistons. From October 2012 to October 2013 we performed 317 surgeries with new prosthesis. Data, as medical histories, surgery protocols and hearing examination outcomes were reviewed and audiological results are gradually collected.

All patients presented progressive, conductive or mixed, unilateral or bilateral hearing loss, ears had never been operated before. Comparing pre- and post-operative pure tone audiometry on frequencies: 0,5, 1,2,4 kHz, results are quite satisfactory. After surgery the mean bone and air conduction decreased, and air bone gap also decreased. Period of observation, due to initiation time of titanum prosthesis in our Center was 6 months.

Stapedotomy with KURZ Skarżyński Piston is good treatment method. Satisfiying hearing effects for patients and surgeons, frequent tinnitus reduction, and chance for normal social living is enough to risk this statement. To be sure of that, further studies are needed.

Difficult Situations

Decision making in far advanced otosclerosis and in otosclerosis with mixed hearing loss.

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Knowing what is the true bone conduction threshold in cases of mixed and far advanced otosclerosis can be difficult. We know from the surgical treatment of simple otosclerosis that the Carhart effect can be considerable. In addition in bilateral far advanced disease it can be problematic to ascertain which ear is providing the bone conduction thresholds. The author will present his results in a series of these cases and debate decision making in how best to treat this difficult group of patients.

Solutions in Complicated Stapedectomy and Revision Stapedectomy.

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Introduction: Stapedectomy is a surgical procedure that has undergone little variation since its inception and most variations are related to technological advances (microsurgical

drills, lasers, prosthesis). Still, anatomic variability makes modifications on the surgical technique necessary. Treatment options are also diverse in Revision Stapedectomy.

Material and Methods: We studied 250 cases of Stapedectomy and Revision Stapedectomy. The variations in the surgical techniques used to solve the complications encountered have been studied. The complications encountered are divided into preoperative (Osteomas, Exostosis, Trauma Reconstruction or other), intraoperative (Promontory Prolapse 1%, Facial Nerve Dehiscence 1%, complicated Persistent Stapedial Artery (PSA) 1/3000), narrow oval window niche 0.5%, obliterative Type IV and V Focus requiring drilling (6-1%), angled Incus 0.5%, an angle of the ear cannal with the Incus which requires a broad atticotomy recess 5%) and postoperative (Necrosis of the long process of the Incus 6%, displaced prosthesis, adhesions, cholesterol granuloma or iatrogenic cholesteatoma and tympanic perforation).The solutions employed are described, they range from drilling of the Promontory or Footplate to modifying the prosthesis, or fixation by Otomimix cement in Revision Stapedectomy.

Results: The functional results with intrasurgical complications are solved successfully in 96% of the cases a year after surgery. In revision surgery the functional results are directly related to the state of the oval window niche. The presence of scar tissue or grafts may condition the outcome since in several cases its resection may involve injury to the inner ear. Therefore, in Revision Stapedectomy, and depending on the findings, an optimal functional result will mean an average air bond gap of 10dB or less in 80% of the cases.

Discussion/Conclusion: We can provide an adequate functional surgical solution for each case except for extreme cases of anatomical variations or revision cases when there is a high risk of injury to the inner ear.

Hearing loss among Osteogenesis imperfecta patients- experience of World Hearing Center.

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Introduction: Osteogenesis imperfecta is a congenital disorder of connective tissue, which results in malformations of organs with high amount of collagene. The main symptoms are numerous fractures of bones, blue sclera, hypermobility of joints, flexible skin, dentinogenesis and in almost half of the patients - different types of hearing loss.

Aim: The aims of this study are to characterize group of patients with hearing loss treated in the World Hearing Center and to assess the hearing results of surgeries.

Material and Methods: In the World Hearing Center 21 patients were treated due to hearing loss with clinical diagnosis of Osteogenesis imperfecta, 19 underwent surgeries due to hearing loss. Stapedotomies, revisions after stapedotomy, restapedotomies and ossiculoplasties were performed. We excluded ears which underwent surgeries in other hospitals. We analyzed results of surgeries of 22 ears. We assessed pure tone audiometry results before and after surgeries in short -term and long-term follow up.

Results: Results of postop air bone gap were very good [ABG <=10dB] and good[ABG >=11dB and <20dB] in 95,5% of cases. Hearing gain >=10dB was obtained in 86% cases in short term observation, 14% of results were between >0dB and <10dB. Sensorineural hearing loss [BC threshold decrease >15dB] was observed in 1 case.

Conclusions: Hearing loss in Osteogenesis imperfecta in most cases is connected with disorders within middle ear. The most common is stapes pathology. Surgeries of stapes allow to obtain air bone closure in many cases of Osteogenesis imperfecta. However some cases need reoperations and hearing gain diminish. Due to high level of malformations of the structures of middle ear it is advised to perform surgery in this group of patients only by the most experienced surgeons.

Hearing loss treatment in patients with imperfect osteogenesis.

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Objective: To analyze the causes of hearing loss and the effectiveness of surgical treatment in patients with imperfect osteogenesis.

Materials: In the otosurgery department of the Institute, 13 people (25 ears) with IO (imperfect osteogenesis) were observed from 2009 to 2015, including 11 women (age from 26 to 48 years) and 2 men (21 and 27 years old). In 8 people, the diagnosis of HB was first established after the onset of symptoms of hearing loss and ear noise based on CT of temporal bones, despite the large number of bone fractures of limbs, scoliosis and kyphoscoliosis of 3 and 4 degrees. In 2 patients the diagnosis was established in early childhood. In all patients, ear noise and hearing loss appeared much later than other symptoms of the disease, from 4 to 8 years before resorting to a surdologist. According to the tone audiometry (TA), in 12 patients bilateral bradyacoustic hearing was noted and in 1 case it was one-sided. Conductive hearing loss was observed in 20 ears (1 and 3 degrees) and mixed - in 5 (2-3 degrees). The thresholds of bone conduction (CP) averaged 14.7 dB, and the bone-air interval (QVI) was 32.1 dB. According to tympanometry in some patients, the type Ad was noted, which indicates the hypermobility of the tympanic membrane and / or rupture of the auditory ossicles.

Methods: Surgical treatment was performed in 13 patients, 12 of them in both ears (25 ears). In 22 cases, stapedoplasty (partial stapedectomy) with laser assist and using an cartilage prosthesis (3.5 mm long) installed on the autovein

graft. The choice of cartilage prosthesis is due to its elasticity, which reduces the risk of necrosis of the long anvil leg in this category of patients.

Results: As a result of surgical treatment, all patients in the postoperative period experienced improvement in hearing. On average, 1 month after the operation, the thresholds of CP were 11.4 dB, and the QVIs - 6.2 dB. After 6 months, the KP thresholds improved by 4.3 dB with the complete closure of the KVI. With dynamic monitoring of patients from 1 to 5 years, successful results persisted.

Conclusion: The cause of hearing loss in patients with HO is local changes in the stapes and demineralization of the capsule of the labyrinth. The choice of the technique of the operation is determined by the state of the arch of the stirrup and the degree of fixation of its base. Taking into account the peculiarities of the disease course, prosthesis of the stapes is necessary to perform an autochthonous transplant, which allows reaching early and distant effective results in this category of patients. For the successful conduct of surgical treatment and the preservation of its result, patients with non-AD should be observed and, if necessary, receive treatment of endocrinologist.

Revision Stapedectomy: Cement Fixation.

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Objectives: In revision stapedectomy we find different variables which cause the relapsing, hearing loss and vestibular symptoms. The most habitual causes are included here such as: prosthesis displacement, necrosis of the long process of the incus, middle ear adhesions, etc. The surgical findings are evaluated as well as their solutions and results.

Material and Methods: Sixty five patients with a history of stapedectomy are studied. Both the variables due to gender and the surgical findings are analyzed (the type of prosthesis, including its placement, the condition the ossicular chain and the middle ear are in, etc.), as well as the solution adopted and the audiological results.

Results: The findings are: prosthesis displacement (82%), distal necrosis of the long branch of the incus (64%), middle ear adherences (33%), tympanic retraction due to extensive atticotomy (28%). Data are also provided for other findings; cholesteatoma, perforations, or absence of prosthesis and reobliterated footplates. There is an analysis of the TC findings and their comparison with the intraoperative findings. The solutions available vary from the placement of a new prosthesis, or cement placement and fixation in those cases where the long process of the incus allows for it. TORP is in some cases the only alternative.

Conclusions: The complete closure of the audiometric gap is achieved in 74% of patients, partially in 16%, unchanged in 6% and worsened in 4%. There is also an improvement in 95% of the vestibular symptomatology.

Taste disorders after stapedotomy.

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Aim: Irritation of chorda tympani nerve during middle ear surgery can result in taste disorders. The aim of this study was to assess pre- and postoperative taste ability in patients undergoing primary stapedotomy.

Material and Methods: Prospective study were performed. Fifty nine (14 men, 45 women) adult patients from Institute of Physiology and Pathology of Hearing were included. Patients were examined with taste test (Taste Strips, Burghart Messtechnik) at 1 day preoperatively (T0), and at 1 (T1) and 7 (T7) days postoperatively.

Results: Due to the taste test protocol 6 patients had preoperative taste disorders. Taste test results were lower in nearly 30 percent (20/59) of patients at T1 comparing to T0 but in most of them (17/20) remained within the normal range. At T7 twelve patients had lower taste test score than in T0 and only one of them was under the range.

Conclusions: Taste disorders may be one of the minior complications after stapedotomy but they are mostly temporary. Severe taste disorders after stapedotomy are very rare. Due to another factors having impact on taste test results in first week of recovery another long follow up is needed.

Malleus ankylosis. Case report.

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The aim of the study was to present a case of a 42 years old male, who was admitted to our Clinic due to bilateral conductive hearing loss. The hearing loss was found in a random test; before that the patient was not aware of the anomaly. In anamnesis no causative factors, such as inflammatory or trauma, were established. The patient was referred for exploratory tympanotomy. During the operation otosurgeon found malleus ankylosis in epitympanum and incudal concretion with lateral wall of the tympanic cavity. Bony block, which caused the malleus and incus immobilization, was removed restoring normal mobility of the ossicular chain. Ossiculoplasty with preservation of the ossicular chain may be successfully applied in imobilisation of the malleus and incus.

Surgical treatment of the 5 – year – old boy with otosclerosis.

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Introduction: Otosclerosis is a disease that reveals itself in the form of unilateral or bilateral hearing deterioration practically in every age. The results of surgical treatment of the youngest boy described in available literature are presented below.

Aim: The aim of this work is to present that progressive, bilateral moderate hearing loss accompanied by tinnitus is the indication for surgical treatment, even at the age of 5 years.

Material and methodology: The youngest boy described in literature with otosclerosis was submitted to surgical treatment at the age of 5. Frontal tympanotomy was performed through the external ear canal. After noting the immobility of crus of stapes and lodgment of otosclerosis on thickened plate, superstructure of the stapes was removed. Then a calibrated hole with a diameter of 0.5 mm was performed. Titanium piston of the KURZ Skarżyński Piston prosthesis with a diameter of 0.4 mm was placed in the hole.

Results: The results of surgical treatment indicate a complete closure of air–bone gap in the ear operated on and rise of bone conductive threshold on 3 frequencies from 5 to 10 dB. In the ear operated on, after the removal of dressing, the child did not complain anymore of tinnitus, which previously was persistent. In non–operative ear which is already planned for treatment tinnitus is variable and periodic.

Conclusion: Surgical treatment of otosclerosis is indicated regardless of the patient's age. The additional indication for surgical treatment to small children, besides hearing loss, are tinnitus and deterioration of bone conductive threshold.

Posters

Tinnitus in the patient with neuroendocrine adenoma of middle ear (NAME).

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Neuroendocrine adenomas are tumors with double differentiation. Middle ear location of those tumors is rare. NAME causes various clinical signs. One of them could be tinnitus. This paper presents a case study of patient with tinnitus related to NAME. A 23-years old patient was admitted to the World Hearing Center because of hearing loss and tinnitus in the right ear. Physical examination suggested tumor of the middle ear. Patient underwent surgical treatment (atticoantromastoidectomy) with excision of pathological tissues and myringoplasty. Tinnitus was evaluated using the battery of questionnaires (TFI, THI, THS). The postoperative period was uneventful. Patient reported hearing improvement and tinnitus reduction. Histological examination revealed neuroendocrine adenoma of the middle ear.

BOR syndrome – case report.

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Branchio-oto-renal syndrome (BOR syndrome) is a rare, autosomal dominant genetic disorder. BOR syndrome is characterized by an association of I and II branchial arch anomalies and renal malformations. Ear malformations can include outer ear, middle ear and inner ear as well.

Objectives: We report 7,5 year-old girl who presented to World Hearing Center with bilateral hearing loss. Past medical history included right-sided renal agenesis.

Materials and metods: Patient had bilateral mixed hearing loss. We present diagnostic evaluation and treatment of this patient.

Conclusion: I and II branchial arch anomalies present in patients with BOR syndrome require proper diagnosis. Treatment depends on the severity of symptoms and variety of ear malformations.

Stapedotomy instead of cochlear implantation in otosclerosis hearing rehabilitation.

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Aim: To evaluate hearing outcome and patient´s quality of life in patients with far-advanced-otosclerosis (FAO) undergoing stapedotomy.

Material and Methods: Data between 2013-2016 was collected from the Swedish Quality register of Otosclerosis Surgery (SQOS) including 1330 patients. Sixty-five patients (68 cases) with a preoperative PTA AC > 70 dB on the non-operated ear were evaluated to determine audiologic improvement and correlated to the results of the patient´s questionnaire one year after surgery.

Results: Sixty-eight cases, mean age 63 years (33-83) were included in the study. Hearing aid was used preoperatively in nearly all cases (98.5%) and surgery was previously performed on the other ear in 43% of the cases. In six patients hearing thresholds were not measurable and defined as deaf preoperatively but all improved with an air conduction gain. Successful surgery, defined as an ABG closure ≤ 10 dB, improvement in AC ≥ 20 dB and BC not worsens by > 5 dB was achieved in 31% of all cases. Two cases became deaf postoperatively necessitating a cochlear implantation. The patient questionnaire was responded by 66% of the patients and a majority were satisfied with the aided hearing postoperatively.

Conclusion: Stapedotomy can be beneficial even in otosclerotic patients with profound hearing loss fulfilling the inclusion criteria for cochlear implantation candidacy.

Optical pressure sensor for intracochlear measurement – design and making.

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Aim: The aim of this study was the analysis of the sensor making process suggested by Elizabeth S. Olson, PhD, and building intracochlear pressure sensor due to this instruction.

Methods and Results: The sensor operates as follows: light emerges from the optic fiber, disperses and is reflected from the membrane. The amount of light which returns to the fiber is correlated with pressure-induced membrane motion and distance to the membrane. This is the way how sensor detects pressure. Making the sensor requires making sensor tip, putting the membrane on the tip, etching optic fibers, making sensor rig and connecting the rig

and tip together. Then, the calibration process is needed. To make the sensor we used a diode with ST connector, multimode optic fiber, a glass capillary tube and a photodiode. Firstly, glass capillary tubing was cut into pieces about 1 cm long. Once, the tips have no rough edges, adding the membrane was performed (Fig.1). We have made the membrane from adhesive glue (NOA 68, Norland Products INC, USA). A drop of the NOA 68 was placed on surface of deionized water. After spreading, the membrane was pre-cured, placed on the tip and finally cured. All the presented images are self-made photographs. Fig. 1 Preparation the membrane from adhesive glue NOA 68. A: The amount of demineralised water, B: a drop of NOA, C: the drop spreading on the surface, D: experimental membranes on straws. Evaporating the tip (Fig. 2) is necessary to make the membrane as reflective. We performed the evaporation process in a special vacuum chamber. Fig. 2 Evaporation process. A: a laboratory chamber uncovered, B: the amount of Aluminium, C: the chamber during the process. Subsequently, we have etched (Fig. 3) the optic fiber, as its outer diameter (OD; 125 um) is too large to fit into the tip. We etched one end of the fiber by immersing it in a 6% hydrofluoric (HF) acid solution for several minutes. In order to quicken the process, the solution temperature was kept between 65-75 degrees. After etching, the OD was about 65-70 um. Then, the etched fiber was inserted into the tip. Fig. 3 Controlling of the OD during the etching process. A: after 5 min, B: after 10 min, C: after 15 min. Our team is during the process of constructing the rig with a Fiber Optic coupler. The system will be supplied in light with Laser Emitting Diode (LED). The light, reflected by the sensor tip will be detected by a Photo Diode (PD). The strength of the reflected light is correlated with the measured pressure fluctuations. Before the voltage signal from the PD will be recorded by an acquisition system, it should be augmented by an operational amplifier.

Conclusions: The process of making the sensor is demanding, however self-made sensors are needed as their sensitivity and specificity are high and their functioning is known.

How to measure pressure in the perilymph fluid?

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Aim: The most efficient parameter defining the effectiveness of middle ear prosthehis is pressure generated in perylimph. The aim of this study is to formulate the methodology of performing pressure measurements using optoelectronical sensor.

Material and Methods: In order to carry out the experiment several devices and tools presented in Figure 1 are needed. Temporal bones should be fresh, ear canal shall be shorten to 1 cm and extended facial recess approach carried out. 1) In order to carry out the experiment, a sound of particular intensity (90 dB SPL) will be calibrated, generated and controlled. The acoustic frequency will vary from 400 Hz up to 10 000 Hz. 2) Temporal bone preparation: requires extended facial recess approach and performing stapedotomy or cochleostomy.3) Placing measuring tools: firstly, insert earphone in external ear canal, place the probe microphone with a tube 2 mm and a speaker about 4-5 mm from tympanic membrane. 4) Insertion of pressure sensors consists in thinning the cochlear promontory and fenestration of 0.5 mm diameter in SV and ST. Each tip of the transducer is supposed to be inserted about 100-300 μm into the scalae and sealed with alginate material. 5) During experiment immersing the TB in saline helps prevent air presence. Figure 1 Measuring system. Figure 2 Temporal bone and measuring tools, on the right: an example of TB (1 – earphone, microphone probe with a tube, 3 – round window membrane). The differential intracochlear pressure, which correlates with inner ear excitation, is measured simultaneously in SV and ST.

Results: Different teams from all over the world have examined intracochlear pressure. In the study by Nakajima (2008) there is a major difference between sound pressure in scala vestibuli and scala tympani for middle frequencies, pressure in SV is much greater. As far as differential pressure is considered it was reduced by 30 to 50 dB for disarticulated ossicular chain. Olson (1998) presented results showing the SV pressure for upper frequency limit was approximately 30 dB higher than pressure in ear canal. In these measurements the SV input pressure has a range between 105 and 120 dB SPL. The phase of SV decreases linearly with frequency. The study by Nakajima (2010) resulted in a higher magnitude of pressure in SV between 0.6 to 8 kHz. The phase of differential pressure below 0.6 kHz was between 0-150 degrees and became negative with the frequency increase.

Conclusions: A sensible clinical methodology is proposed. It consists in measurements of intracochlear pressure of perilymph before and after stapedotomy. Results relying on the technique presented above are to be used to fulfil the preclinical data inevitable in the process of assessing the effectiveness of a new chamber stapes prosthesis.

Is the chamber stapes prosthesis safe for patients?

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Aim: New chamber stapes prosthesis (ChSP) is a medical device intended for use in the stapedotomy surgery. The ChSP has an innovative design and is composed of three elements: the chamber, the membrane, and the plate. Although the simulation and experimental studies have shown the effectiveness of the ChSP functioning, its safety of use has not yet been demonstrated. The aim of this work is to perform the preliminary risk analysis (RA) and to assess the risk-benefit ratio.

Material and Methods: The RA was performed according to ISO 14971. The hazards were determined and risk for each hazardous situation (HS) was estimated in qualitative

manner based on expert opinion. The risk was defined as the combination of the probability (P) of occurrence and the severity (S) for each harm. The P levels were: P1-high (likely to happen), P2-medium (can happen but not frequently), P3-low (unlikely to happen). The S levels were: S1-significant (death or loss of function or structure), S2-moderate (reversible or minor injury), S3-negligible (will not cause injury or will injure slightly). The benefit arising from the ChSP was estimated in comparison with the piston prosthesis.Results.Each of hazards can progress to the HS if foreseeable sequences of events (SE) occur. Relationship between hazards (Hz), SEs, HS and the harm (Hm) that can occur are as follows:Hz1: Acoustic or mechanical energy. SE:1. Sudden loud sound, noise exposure or violent head injury; 2.Prosthesis membrane damage. HS: ChSP cannot transmit sound. Hm: Conductive or sensori-neural hearing loss (CHL/SNHL) or mixed hearing loss (MHL), reoperation. HZ2: Biological. SE:1.Sudden loud sound or violent head injury, 2.Unsealing of chamber. HS:Bacteria or viruses enter to inner ear, fluid leakage. Hm:Bacterial or viral infection, SNHL, pain, reoperation. Hz3:Biocompatibility. SE:1.Incorrect chemical composition of fluid filled the chamber, 2.Fluid merges with perilymph. HS:Toxicological reaction. Hm: SHL, dizziness. Hz4:Function. SE:1.Incorrect fatigue strength of membrane or plate, 2. Fatigue failure. HS:Long-term reduced or impossible sound transfer. Hm: Long-term CHL. The estimated risks associated with the biological and biocompatibility hazards are on the unacceptable S1 level and risk reduction is required. The benefit arising from the ChSP in comparison with the piston includes: (1) the chamber cannot be lifted out of the vestibule nor move in too deep into the vestibulum, (2) the cochlear stimulation is on the physiological level, (3) masking effect for high-frequency tinnitus can occur.

Conclusions: There are any clinical data to confirm compliance with relevant essential requirements for safety (Directive 93/42/EEC) when using the ChSP. Therefore, to validate that the ChSP meets acceptable risk-benefit criteria, clinical testing is required.

Chamber stapes prosthesis – preclinical study of sound transmission.

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Aim: New chamber stapes prosthesis (ChSP) is intended for patients with stapes otosclerosis. Before being introduced to clinical practice, a number of preclinical and clinical tests are required. The ChSP functioning has been numerically simulated (Kwacz, Hear Res, 2014) and it was shown that the prosthesis effectively transmits vibration for frequencies 0.4-10 kHz. The aim of this study is to assess the sound transmission in ear implanted with a ChSP prototype.

Materials and Methods: The ChSP prototype (Fig.1A) consists of a conical chamber (1) filled with fluid, a flexible membrane (2, FM) that closes the wider base of the chamber, and a rigid plate (3) placed on the FM. The chamber and the rigid plate were made from acrylonitrile-butadiene-styrene (ABS-M30i) using ProJet 3600 Printer. The FM was self-made from UV-light curing adhesive NOA 68 (Norland Optics, USA) according to technique described by Olson (JASA, 1998). The FM had a thickness of ~25 um. The ChSP prototype (Fig.1B) was filled with deionized water, covered with the FM and implanted in a human cadaveric temporal bone specimen. Fig. 1. The ChSP prototype (A, B) and experimental setup (C, D). B – temporal bone, S – acoustic signal, M – microphoneA scanning laser-Doppler vibrometer (PSV400, Polytec, Germany) was used to measure vibrations of the round window membrane (RWM) before and after the ChSP-stapedotomy. An acoustic signal (90dB SPL, 0.8-8 kHz) was introduced to the external ear canal via an ER-2 loudspeaker (Etymotic Research, USA). The 90-dB SPL near the tympanic membrane was controlled by an ER-7C microphone (Fig.1C).The RWM vibration in a normal ear was measured at 155 scan points. Then, the stapes footplate (SF) was immobilized, the stapedial muscle was cut, the stapes suprastructure was removed and a 0.6-mm hole was drilled in the SF. Into this hole, the thin tube of the ChSP (4 in Fig.1A) was inserted and the chamber was fixed to the oval window niche. The long process of the incus was fixed between the sidewalls of the rigid plate (Fig.1D). Then, the RWM vibration in the post-stapedotomy ear was measured.Based on the measured displacement amplitudes, the magnitude of fluid volume displacement (VD) at the RWM was calculated. **Results:**In Figure 2, the VD magnitude at the RWM before and after the ChSP-stapedotomy are shown. Fig. 2. Fluid VD at the RWM in the normal ear and the ChSP-stapedotomy ear. 90 dB SPL in the external ear canal.For f > 1.0 kHz, the VD induced by the ChSP is significantly higher than the VD in the normal ear. The magnitude of the VD depends on both the stiffness of the FM (e.g. its thickness and material parameters) and the surface area of the chamber wider base.

Conclusions: The ChSP prototype works correctly and effectively transmits sound for frequencies 0.8-8 kHz. Further studies are necessary to design the biocompatible FM.

Coexistence of congenital stapes ankylosis with middle ear malformations in children.

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Aim: The aim of the study was the analysis of intraoperative findings in the middle ear and hearing results in patients after stapedotomy performed in treatment of congenital hearing loss.

Material and Methods: Study group comprised 36 patients operated before 18 years of age, with congenital conductive or mixed hearing loss. All underwent stapedotomy. Anomalies found in the tympanic cavity have been classified according to the Teunissen and Cremers classification. Preoperative diagnostics included laryngological, audiological

and radiological examination. Postoperative diagnostics included otological and audiological assessment.

Results: Conditions found intraoperatively in the middle ear have been described and classified. Hearing results have been assessed at 12 months after surgery.

Conclusions: Stapes ankylosis may be coexistent with different changes in the middle ear. Stapedotomy is a safe method of treatment of hearing loss in children with congenital middle ear defect and stapes ankylosis. CT imaging is an essential element of diagnostics before surgery in order to exclude hypertension in the inner ear, but not always conclusive.

Progressive otosclerosis with bilateral sensorineural hearing loss and vestibular impairment- case report.

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Hearing loss, tinnitus and dizziness are common complains among patients with otosclerosis. Usually it takes years from early onset to deafness. The aim of the study is to present a case of progressive otosclerosis with episodes of sudden hearing loss and loss of balance within 2 years.43-years old woman with bilateral sensorineural hearing loss was treated for episodes of sudden loss with steroids and after 2 years received cochlear implant because of non-functional residual hearing. Her otoneurological problems included imbalance in motion especially in darkness and oscillopsia. Magnetic resonance imaging revealed intensive otosclerotical process of the inner ear. Authors present audiological, otoneurological and radiological findings of the patient. In progressive otosclerosis with residual hearing and bilateral vestibular impairment, cochlear implantation and vestibular rehabilitation were the optimal treatment solution.

Stapes surgery for paediatric otosclerosis – a literature review.

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Aim: The objective of the study was to synthesize published data on stapes surgery for pediatric otosclerosis, regarding course of the diagnostic process, the usefulness of pre-operative imaging techniques, the characteristics of patients undergoing surgery, the course of the operation, its results and possible complications.

Material and Methods: A literature search was performed using the following databases: PubMed, Web of Science and Medline. Using the combination of keywords: "stapes surgery", "child", "juvenile" and "otosclerosis", 188 articles were found by PubMed, 31 by Web of Science and 46 by Medline. As the literature on the topic is scanty and one of the objectives was to analyse the development that occurred in the treatment of pediatric otosclerosis, we did not limit the timeframe of our search. Based on the eligibility criteria, 19 articles were found suitable for the final elaboration.

Results: About 400 cases of surgical treatment of pediatric otosclerosis have been described in the articles included in the review. The age of patients undergoing stapes surgery ranged from 5 to 21 years with the preponderance of girls in comparison to boys (in a ratio: 4-3/1). The authors of the analyzed papers emphasized rather low sensitivity of the computed tomography in the diagnosis of pediatric otosclerosis. Recent studies on stapedotomy audiological outcomes in children indicated that this procedure allows for the air-bone gap closure in more than 92% of operations. Due to the diversity of grafts and prostheses used, it was impossible to establish their relationship with postoperative results. Complications of stapes surgery were extremely rare and included in most cases transient dizziness. There was also no relationship between the age of pediatric patients and postoperative closure of the air-bone gap and the frequency of postoperative complications.

Conclusions: Stapes surgery for pediatric otosclerosis is a safe procedure resulting in closing air-bone gap in long- and short-term observation. The patient's age should not be a contraindication to the operation.

Stapedotomy in the only hearing ear with persistent tinnitus.

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The treatment of the only hearing ear is always a challenge for the otosurgeon and even more so for the patient, especially if previous surgery in the other ear had been with complications. However, many years of experience with such cases suggests that indications for stapedotomy in conductive hearing loss should be the same whether we making a decision about surgery in the only hearing ear or in both ears. The aim of this study was to present a case of a patient with only one hearing ear with additional atypical intraoperative finding and to demonstrate the such findings do not warrant resigning from performing a stapedotomy.

The patient was a 62 years-old woman admitted to the clinic due to hearing problems and tinnitus, who had undergone surgeries in both ears 19 and 22 years earlier. The audiometry showed total deafness in the left

ear and profound mixed hearing loss in the right ear. Hearing thresholds in the right ear in the past 22 years from the surgery were deteriorating gradually and presently the air conduction was at the level of 60 dB, and bone conduction at 25-30 dB with tenacious tinnitus. It has been decided to perform the stapedotomy in the right ear under general anesthesia. An intrameatal incision was performed in the narrow external auditory canal. The external ear canal was observed to be widened during a previous surgery that failed to improve patient's hearing. Intraoperatively surgeons had observed disconnection of the stapes suprastructure from the plate and very narrow oval window. The divulsion of the stapes occurred most probably during a previous operation which involved stapes mobilization. The stapes suprastructure was removed and a 0.5 mm diameter hole was drilled in the stapes plate. A titanium prosthesis KURZ Skarżyński Piston 0.4 mm was applied.

After removal of the ear canal packing full closure of the air-bone gap was achieved with improvement of the bone conduction curve by 5-10 dB. The patient reported immediate diminishment of the tinnitus and after 2 months tinnitus was completely resolved.

The only hearing ear cannot be nowadays a contraindication for surgical treatment. In each of such cases it is essential to assure that patient has full knowledge and understanding of possible complications. Surgeon must have adequate experience in such procedures. In our patient's case the surgery had been mandatory due to lowering of the bone conduction curve and burdensome tinnitus.

Tinnitus evaluation method in adult patients with otosclerosis.

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Aim: Subjective tinnitus is a common complaint among patients with otosclerosis. It can exacerbate the negative impact of hearing loss on people's daily life. Hence, distinguishing patients with bothersome and nonbothersome tinnitus is a diagnostic challenge with a possible impact on postoperative benefits. The aim of the study is to present a protocol for evaluating severity of tinnitus in adult patients with otosclerosis. The secondary objective is to assess the hearing benefit and quality of life in patients with and without tinnitus.

Material: Patients over 18 years of age are included in the study. The eligibility criteria are occurrence of tinnitus and first-time scheduled for the operation in the investigated ear.

Methods: The participants are asked to fill in the following questionnaires: Tinnitus Functional Index (TFI), Tinnitus and Hearing Survey (THS), Abbreviated Profile of Hearing Aid Benefit (ABHAB) and Assessment of Quality of Life (AQoL-8D). The protocol of the study includes completing the questionnaires before surgery and after 3 and 6 months. Additionally, pure-tone audiometry is conducted to evaluate the hearing benefit.

Results: There are a few scientific publications on the prevalence and severity of tinnitus in patient undergoing stapes surgery. Nowadays, there is still a tendency to report only postoperative hearing outcomes based on pure-tone audiometry. The preliminary study carried out in our department showed that tinnitus affects a substantial group of stapes surgery candidates. This finding prompted us to create the comprehensive protocol of assessing tinnitus in relation to both subjective and objective hearing benefit and quality of life.

Conclusion: The study allows the development of an original protocol to evaluate the original protocol to evaluate severity of tinnitus in the course of otosclerosis and their impact on the subjective assessment of hearing and quality of life.

Audiologic outcomes after stapedotomy in Ehlers – Danlos Syndrome – pediatric cases report.

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Aim: Ehlers – Danlos syndromes (EDS) is a rare inherited group of connective tissue disorder characterized by joint hypermobility and delicate, hyperelastic skin. The aim of the study was to assess audiologic outcomes in two adolescents with EDS after stapedotomy.

Material: The study involves two pediatric patients with history of increasing hearing loss in both ears. They both showed clinical features of the EDS syndrome. The 14-year-old boy was observed bilateral conductive hearing loss, progressing since the year 8. The patient underwent mobilization in the left ear and two years later - stapedotomy in the right ear. The second case concerns a 13-year-old girl who suffered from bilateral mixed hearing loss, progressing from early childhood. In addition, the girl had severe tinnitus and periodic dizziness. The patient underwent a bilateral, sequential stapedotomy.

Methods: Pure tone air- and bone – conduction audiometry were used to evaluate hearing outcomes. The pure – tone average was calculated for air and bone conduction using the frequencies: 0.5, 1.0, 2.0 and 4.0 kHz. For these frequencies was calculated also air – bone gape before and after surgery.

Results: In the boy, the average air conduction threshold decreased by 13 dB after mobilization surgery and 23.8 dB after stapedotomy. In the girl who underwent

bilateral stapedotomy, the avereage air conduction threshold decreased by 30.3 dB in the left ear and 27.5 dB in the right ear. In the 2-year observation, the stability of hearing thresholds was observed.

Conclusion: Few studies have been published regarding the outcomes surgery of patients with EDS with the majority of EDS literature focusing of surgical complication. To our knowledge, these are the first pediatric patients with EDS, who underwent stapedotomy. The obtained good results of hearing improvement, which are stable in the 2-year observation, confirmed the validity and effectiveness of performed stapes surgery.

Long-term hearing results after stapes surgery- observation of 5 and 10 years follow-up.

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Aim: Otosclerosis is an abnormal growth of bone near the middle ear which can lead to conductive, mixed or even sensorineural hearing loss. According to literature one of the most effective treatment of this disease is stapes surgery. The aim of this study was to evaluate the results of stapes surgery after 1, 5 and 10 years follow-up, according to pure-tone audiometry results.

Material: From over 21 000 stapes surgeries (stapedotomies, restapedotomies and stapedotomy revisions), which constitute the World Hearing Center material, 100 patients operated in 2004-2006 were randomly selected. This group of patient underwent stapedotomy in process of otosclerosis. The observed group is consisted on men and women aged 18-60.

Methods: Preoperative and postoperative results of puretone audiometry for air and bone conduction were analyzed to evaluate hearing outcomes. The mean hearing thresholds for air and bone conduction were calculated using the frequencies: 0.5, 1.0, 2.0 and 4.0 kHz. To evaluate improvement after stapes surgery mean preoperative and postoperative air-bone gap (ABG) was calculated.

Results: Basing on the long-term results after stapes surgery, it was found that in the majority of selected group of patients air-bone gap closed to within 10 dB. The extensive results of long-term observation after stapes surgery are going to be presented during the 4th International Symposium on Otosclerosis and Stapes Surgery in Cracow.

Conclusions: The stapes surgery in process of otosclerosis is a save and successful treatment for long-term hearing improvement. It is shown that many patients had air-bone gap reduction and in turn significant hearing gain. The results of surgery to obtain satisfactory postoperative hearing is also depended on the experience of a surgeon.

Ossiculoplasty in single sided conductive hearing loss with suspected otosclerosis.

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Introduction: Single sided conductive hearing loss, lack of stapedius muscle reflex in both ears, type A tympanometry, no changes observed in otoscopic examination suggest usually the diagnosis of otosclerosis. In such cases the preoperative evaluation should involve decision that intraoperative assessment is necessary for the surgeon to make a final decision about treatment.

Material and Methods: The analysis is based on the case study of 38-years old patient, who in the diagnostic process had been repeatedly informed that her hearing loss is caused by otosclerosis.

Results: Patient has been qualified for surgical treatment – exploratory tympanotomy with possible stapedotomy or other type of reconstruction of middle ear conductive apparatus. Intraoperatively was observed the destruction of the long process of incus, with its fragments in a strap of tendon tissue between the body of incus and the head of stapes. This is why the CT images made before surgery had shown that the long process is thinned but preserved. The long process of incus has been reconstructed with glass-ionomer cement. Very good hearing results have been achieved with closure of the air-bone gap.

Conclusions: Presented stages of the diagnostic process and intraoperative findings demonstrate then it is necessary to carefully formulate the inferences, even initial, with regard to the final diagnosis and implemented otosurgical treatment.

Certain causes of surgical treatment failure of otosclerosis - case report.

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The course of treatment of a 55 years old woman hospitalized due to progressive, bilateral, mixed hearing loss caused by otosclerosis is discussed. Prior to admission, the patient had undergone: stapes mobilization, then right ear stapedotomy and restapedotomy and finally left ear stapedotomy. Transitory improvement of hearing after stapes mobilization with following deterioration with lowering both air and bone conduction thresholds in the right

ear were the indication for stapedotomy. After the operation improvement of hearing was achieved but air bone gap from 35 to 15 dB persisted. Due to this fact after almost a year restapedotomy resulted in full closure of the air bone gap. Finally because of hearing loss in the left ear stapedotomy on this side was performed. Revision operations in otosclerosis demand sufficient experience due to a variety of possible causes of failure. It is especially difficult when the first operation was performed by another surgeon in another center. Stating significant air bone gap is an indication for reoperation by the most experienced otosurgeon in the team. The most common cause of failure of stapedotomy is occurrence of concretion after subsequent operations.

Promontorium drilling for better visualization of oval window - case report

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We present a case of a 62 years old woman with progressive, bilateral hearing loss lasting for 30 years due to otosclerosis. We describe one of the difficulties encountered during stapedotomy and remedial measures necessary for successful operation. During operation the otosurgeon found a narrow oval window niche with overhanging promontory. It was decided to drill a fragment of the promontory for better visualization of the niche and to perform successful stapedotomy. The operation enabled closure of the air-bone gap without sensorineural hearing deterioration. The patient is awaiting the operation of the other ear.Stapedotomy gives an outstanding opportunity to improve hearing in otosclerosis, however certain situations require nonstandard measures. In the described case drilling a fragment of promontory enabled successful accomplishment of the operation.

Stapedotomy in the only hearing ear.

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Over 25 year experience in surgical treatment of otosclerosis, during which the second author has performed more than 20,000 operations, allow authors formulating the opinion with regard to decisions about treatment of the only hearing ear. This subject has been presented for discussion for the first time at the national conference in Katowice in Poland in 1998. The author presented then a research based on a survey carried out among outstanding international surgeons. Author's opinion at that time was very careful and the only proposed indication for such intervention was sudden deterioration of hearing with a fast process of vestibule obliteration, confirmed by computed tomography and audiometric results indicating elevated hearing thresholds preventing effective amplification. In the survey responded: Prof. W. Hause, prof. E. Offeciers, prof. M. Tos., prof. U. Fisch, prof. J.M. Sterkers, prof. J. Helms, prof. T. McDonald, prof. T. Balkany, prof. E. Myers, prof. E. Lehnhard, who presented conservative approach. The proposed method of treatment involved conventional hearing aids for as long as possible. Only half of survey responders considered that exceptional situations may be an indication for surgery.

In the last 16 years, the author performed this procedure in 613 only hearing ears . The contralateral ears were deaf because of advanced, long-term otosclerotic lesions and damage of the inner ear, or damaged by the previous stapedotomy. All patients were informed on conditions of their hearing, possible chances and on rare but possible, not intended complications. Full cooperation of patients and patients' consent were the basis for decision on stapedotomy of the only hearing ear. Our results in this group of patients indicate hearing efficiency preserved in 99.2% in the only hearing ear.

These exceptionally good results obtained in a significant number of patients with otosclerosis and patients with the only hearing ear, that in treatment of such cases we should use the typical criteria. The surgeon should make a conscious and confident decision on the operation. Lack of decision will not, however, stop the destructive processes in the inner ear, but it may prevent future effective amplification or cochlear implantation.

Authors underline that the decision on surgery for otosclerosis in the only hearing ear should be taken with care and attention paid to fast elevating thresholds, increase of tinnitus and vestibular changes seen on CT images. Surgery in the only hearing ear is as risky as any other stapes surgery. Stable hearing after surgery in the only hearing ear confirms the effectiveness of proposed treatment, but does not exclude the possibility of complications.

Impact of stapes surgery on tinnitus in patient with osteogenesis imperfecta – a case study.

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Osteogenesis imperfecta (OI) is the most common hereditary disorder of connective tissue. It is associated with defects of the osteoarticular system, dentinogenesis imperfecta and progressive hearing loss. The aim of study was to retrospectively evaluate tinnitus and hearing of patient with Osteogenesis Imperfecta after stapes surgery. A 60 – year old woman has undergone bilateral, sequential stapes surgery. She was asked to complete the Tinnitus Functional Index (TFI) to assess the annoyance tinnitus and Abbreviated Profile of Hearing Aid Benefit (ABHAB) to subjective

assess hearing. Questionnaires consisted of two sets. The first set was about preoperative state, the second about postoperative state. The audiological findings of this patient have been described earlier. A patient reported progressive hearing loss and constantly tinnitus over 35 years before first operation. Preoperative TFI results showed, that tinnitus was the biggest problem in subscales: intrusive, sense of control, sleep and relaxation. She reported a complete disappearance of tinnitus after surgery. Preoperative audiometric tests showed bilateral mixed hearing loss. After surgery, closure of the air – bone – gap in left ear and significant reduction in right ear was observed. The APHAB results confirmed the reported improvement of hearing after stapes surgery. Osteogenesis imperfecta is characterized by disorders of bone metabolism, with hearing loss being a frequently reported factor. Patients with this disorder often have malformation of stapes, which may influence the postoperative hearing results. There are many publication which present exclusively postoperative hearing result. Little is known about annoyance of tinnitus in this group. This case shows that there is an opportunity to obtain improved hearing and also reduction of tinnitus. This effect was maintained in 5 year follow-up period. As a result of sequential stapedotomy in both ears, a patient reported subjectively improved hearing and total reduction of tinnitus.

Effectiveness of stapes surgery in patients over 60 years old.

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Aim: Otosclerosis is a disease that leads to progressive, unilateral or bilateral hearing loss, firstly conductive, and with time mixed or sensorineural hearing loss (depending on the location of otosclerotic foci and the severity of the disease). Otosclerosis is the most common in group of women aged 15-40. However, as live expectancy increases, there may be a tendency to observe more elderly patients who need stapes surgery.

The aim of this study was to analyze hearing results after stapes surgery in otosclerotic in group of patients older than 60 years.

Material and Methods: The object of these retrospective study were patients undergoing stapes surgery. From over 21 000 stapes surgeries (stapedotomies, resapedotomies and stapedotomy revisions), which constitute the World Hearing Center material, 50 patients operated on in 2004-2006 were randomly selected. In all cases otosclerosis were diagnosed preoperatively. Age was an inclusive criterion. All patients at the time of surgery were over 60 years old.

The evaluation of effectiveness of stapes surgery in elderly patients was made on the grounds of pure-tone audiometry (air conduction and bone conduction thresholds). The results were analyzed preoperatively, at 1, 5 and 10 years postoperatively. The mean preoperative and postoperative air-bone gap (ABG) was calculated as the difference between the air conduction (AC) and bone conduction (BC) thresholds. The average hearing gain was calculated as the difference between the preoperative and the postoperative ABG.

Results: The results will be presented during the 4th International Symposium on Otosclerosis and Stapes Surgery in Cracow.

Conclusion: According to literature stapes surgery should be offered to the elderly patients (especially when it is possible to recover social hearing) with the same indications as younger patients with otosclerosis even if audiometric results are better in younger groups. Stapedotomy or stapedectomy in the elderly is indicated to avoid hearing aid use or to facilitate hearing aid fitting, when the association of otosclerosis and presbycusis makes hearing aid unserviceable.

Effectiveness of stapes surgery in the elderly – review of literature.

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Aim: Otosclerosis is a complex and progressive disease of pathological bone remodeling that affects the otic capsule of the temporal bone, resulting in conductive and with time sensorineural or mixed hearing loss. Otosclerosis is most common in women aged 15-40. However, as live expectancy increases, there may be a tendency to observe more elderly patients.

The aim of this study is to measure effectiveness of stapes surgery in the elderly according to literature review.

Material and Methods: The review is based on electronic databases, containing scientific articles: PubMed, Web of Science and Google Scholar. The publications have been found by using consecutive key words: „otosclerosis in the elderly", „stapedotomy in the elderly", „age-related otosclerosis", age-related stapedotomy", „stapes surgery in the elderly". Eventually, 10 English-language publications, in which at least one of the presented groups of patients were patients over 60 years of age were selected and analyzed.

Results: According to the authors, decisive factor for successful surgery is closure of air-bone gap in 0-10 dB. This result was obtained in over 70% of older patients. Based on a literature review, it was observed that in groups of elderly patients preoperative bone conduction thresholds were elevated (> 30 dB HL), which may be dictated not only by otosclerosis, but also by presbycusis hearing loss.

Conclusion: Surgical methods of otosclerosis treatment are equally effective in the elderly as in other age groups. However, in three publications, a different position was postulated, paying attention to poorer postoperative results of people over 60 years of age compared to younger patients. At the same time, the value of other aspects demonstrating the benefits of otosclerosis treatment in the elderly, such as: inhibition of the disease process, but also older patients experience greater satisfaction especially in terms of daily activities and interpersonal relations.

Evaluation of auditory benefits in a patient with Treacher Collins Syndrome after stapedotomy - a case report.

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Aim: The Treacher Collins Syndrome is characterized by numerous deformities within the facial skeleton. Congenital malformations of the external and middle ear are also observed in this group of patients which results in conductive or mixed hearing loss. The aim of the study is to assess the auditory benefits of a patient with Treacher Collins Syndrome after a stapedotomy.

Material: The work concerns the 26-year-old woman with Treacher Collins Syndrome with mixed hearing loss caused by bilateral congenital defects of the middle ear. The patient did not accept sound amplification using bone anchored hearing aids and classic hearing aids for air conduction. At the age of 11 a stapedotomy in the ear with a larger hearing loss (right ear) was performed.

Methods: The results of pure-tone audiometry for air and bone conduction were analyzed to evaluate hearing outcomes. The mean hearing thresholds for air and bone conduction were calculated using the frequencies: 0.5, 1.0, 2.0 and 4.0 kHz. To assess the subjective benefits of hearing after the surgery, the Abbreviated Profile of Hearing Aid Benefit (APHAB) questionnaire was used. Its individual scales helped to estimate understanding of speech in different environmental conditions and reactions to surrounding sounds.

Results: Before the operation, the mean air conduction threshold was 60 dB HL, and the size of air-bone gap was 26 dB in the operated ear. After the stapedotomy, a significant improvement in hearing was achieved. Postoperative air-bone gap shows the improvement of air and bone thresholds obtained after stapedotomy versus preoperative air bone gap. In the long 15-years observation hearing in the operated ear has not exacerbated. The results of individual APHAB scales showed improvement in subjective hearing in different environmental conditions.

Conclusion: Basing on results of pure-tone audiometry and the APHAB questionnaire, significant and long-term auditory benefits after stapedotomy were observed. There are few scientific reports on the surgical treatment of congenital malformations in the course of concomitant rare diseases. In many medical centers, the standard procedure is compensation of hearing loss using classic hearing aids or bone anchored hearing aids BAHA. The decision to use a stapedotomy in case of congenital defects of the middle ear depends mainly on the degree of malformation. The otosurgeon's experience is another important factor determining the success of the operation.

Review of possible postoperative otoneurological complications after stapedotodomy.

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Aim of the study: Operation of otosclerosis, despite its effectiveness and popularity, may cause disturbance in vestibulum function. The aim of the study is to present possible postoperative vestibular complications and their mechanism.

Materials and Methods: Review of the literature and retrospective analysis of the history of the patients' after stapedotomy procedure, operated in Otorhinolaryngosurgery Clinic in Institute of Physiology and Pathology of Hearing.

Results: The postoperative vertigo after stapedotomy may be divided in: short-lasting and persistent. The first is observed in 3-7% of the patients, the latter in less than 0,5%. The most frequent causes for short lasting vertigo include: the contact of prosthesis or surgical instruments with membranaceous labirynthus of the sacculus and utricle, the change of the fluids pressure in vestibulum after drilling the opening in the footplate of the stapes, perylimph aspiration during the procedure, labirynthitis, enzymatic process conjoined with releasing the toxic substances by the otosclerotic plaques. The causes of persistent vertigo may be: replacement of the otolith with consequent benign paroxysmal positional vertigo, persistent perylimphatic fistula, too deep penetrating prothesis with pressure on the membranaceous labirynthus of utricle and sacculus, vestibular damage, dislocation of the prosthesis (too short prosthesis, necrosis of long process of the incus), adhesiones.

Conclusions: Vestibular disorders are not rare complications of stapedotomy procedure. In the majority of cases the symptoms are present only in postoperative period and are the consequence of disturbance in inner ear fluid homeostasis, resolves spontaneously and do not demand any further interventions. Sometimes, the persistent vestibular symptoms are the indication for reoperation and repositioning maneouvers. Every patient, before signing the informed consent, should be informed about the possible influence of stapedotomy on vestibular organ.

Simplified three-dimensional basilar membrane model in FSI analysis.

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Aim: Numerical simulations of FSI (fluid structure interaction) type are time-consuming. This is due to the need of using two solvers, which have to communicate one with each other during work. One of these solvers performs calculations in the domain of a solid body (FEM solver), while the other in the fluid domain (CFD solver). To speed up the calculation, it was proposed to use one solver working in the fluid domain, with the possibility of performing simple calculations for a solid body, that is, the basilar membrane.

Material and Methods: ANSYS FLUENT is a solver designed to perform fluid dynamics calculations. A user-defined function, or UDF, is a function that you program that can be dynamically loaded with the ANSYS FLUENT solver to enhance the standard features of the code. Using this tool, a basilar membrane model was created and added to the solver. The basilar membrane is modeled as a set of resonators. The movement of resonators is transferred directly to the fluid by means of appropriate boundary conditions. Thanks to this, the geometry of the model does not change and there is no need to modify its mesh, which is a time-consuming task.

Results: The proposed solution resulted in a significant acceleration of the calculation compared to the traditional FSI model.

Conclusions: The simplified FSI basilar membrane model, based on the ANSYS FLUENT solver and the UDF mechanism, seems to be a good alternative to traditional, time-consuming FSI models for simulating the inner ear operation.

High Resolution Computed Tomography of the Inner Ear: Effect of Otosclerosis on Cochlear Aqueduct Dimensions.

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Objectives: The aim of this study was to determine the radiographic detection of otosclerosis and compare to intraoperative findings, as well as to provide radiographic metric classification of the cochlear aqueduct.

Methods: A retrospective chart review of seventy-seven high resolution CT scans in patients with a diagnosis of otosclerosis based on clinical and operative findings. All scans were evaluated by neurotologists, neuroradiologists and general radiologists. The location and extent of otosclerotic foci were recorded. Unified methods were then used to determine the dimensions of the cochlear aqueduct in both affected and unaffected ears.

Results: Otosclerotic foci were visualized in 74% 68%, and 59% of the cases by neurotologists, neuroradiologists and radiologists, respectively. Radiographically, fenestral otosclerosis was noted in 97% of the above cases with 14% of ears also having cochlear involvement. In all patients with positive CT scan reads by neurotologists, otosclerosis was confirmed intraoperatively. Out of all reviewed ears, bilateral otosclerosis was noted in 67% of cases. The average cochlear aqueduct length, midline diameter and aperture dimensions in otosclerotic ears were 12.19, 0.68 and 2.60 mm, respectively. There was no noted statistical difference in these dimensions among otosclerotic and non-otosclerotic ears.

Conclusions: In our series, the rate of radiographic detection of otosclerosis widely differed based on the training background of the individual reviewing the scans. Hypodense otosclerotic foci were primarily situated along the anterior footplate. Metric evaluation of cochlear aqueduct dimensions revealed no statistically significant difference among otosclerotic and normal ears. The average dimensions were comparable to previously published data regarding healthy ears. Hence, we can conclude that the structure of cochlear aqueduct and potentially its patency are not effected by otosclerosis.

MicroRNA Expression Profiling in Perilymph Specimen of Otosclerotic Patients.

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Introduction: Otosclerosis is a relatively rare inflammatory bone remodeling disorder of the temporal bone with a complex and not fully understood etiology. A progression of inflammatory bone resorption, most commonly along the fissula ante fenestram, results in progressive conductive hearing loss due to fixation of the stapes. Ultimately, 70% of patients with conductive hearing loss undergo stapedectomy or stapedotomy. (1) At our institution, we have been serially collecting perilymph samples for microRNA (miRNA) analysis. MiRNA are 19-23 base pair single stranded RNA sequences that regulate mRNA translation and play a vital role in a myriad of cellular pathways and disease pathologies. (2) MiRNA regulate and define expression of genes through guided degradation through various mechanisms and have been implicated in several neurodegenerative disorders through modulation of stress and survival pathways. (3,4) Of clinical significance, miRNA have been successfully identified in numerous body fluids including blood serum, saliva, urine, and CSF and were found to serve as biomarkers for detecting and monitoring numerous pathophysiological conditions. (5)

Aim: The aim of this project is to describe the miRNA profile in the perilymph of patients with otosclerosis undergoing surgical interventions.

Materials and Methods: The perilymph of 4 patients undergoing stapedectomy with identified otosclerotic foci was collected. Perilymph was collected using a sterile glass capillary applied to the CO2 laser created oval window

opening. MiRNA expression profiling was carried out using the Affymetrix GeneChip miRNA 4.0 Arrays. Data sets were then analyzed using Ingenuity Pathway Analysis (IPA) software. IPA is an online software tool for identifying significant molecular interaction networks and biological functions associated with a set of genes (miRNAs) based on information gathered in the Ingenuity Pathway Knowledge Base (IPKB). Furthermore, miRNA profiles from above mentioned patients were compared to miRNA perilymph expression norms established in our lab to detect components specific to otosclerosis.

Results: Collected samples on average yielded 5-2 ng of total RNA per sample in a volume of 2-5 µl. Among all four specimen, miRNA specific to middle ear and cochlear otosclerosis and not noted in our comparative general population were found. Correlation between these specific miRNA and processed involved in osteoporosis were further noted.

Conclusion: Otosclerosis is a relatively rare disease making the access to perilymph samples limited. Preliminary data shows possible relationships which will need to be further investigated with a larger patient sample size. Ultimately, the goal is to identify a panel of molecules that serve as otosclerosis markers by defining miRNA that are unique to perilymph and networks of miRNAs that interact with molecules believed to be involved in the pathogenesis of otosclerosis.