Dear Colleagues,

Welcome to the **XIII Otorhinolaryngology International Academic Conference ORLIAC**!

The XIII ORLIAC in Warsaw proudly continues the tradition initiated by Professor Jan Veldman to facilitate the exchange of ideas and knowledge between the East and West to broaden the horizons of research and clinical work and create opportunities for expanding global collaboration and relationships.

We present here a comprehensive collection of abstracts from the XIII ORLIAC incorporated into a scientific program whose aim is to encourage discussion and new research ideas.

We wish you a productive and inspiring conference!

Prof. Henryk Skarzynski
President

Prof. Piotr H. Skarzynski
Scientific Secretary

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Workshops

Otosclerosis: from standard to challenging cases

Di Bari M.[1,2], Alciato L.[2], Delille H.[2], Sterkers O.[2], Colombo G.[1], Bernardeschi D.[2]

[1] *Otorhinolaryngology Department, Ospedale Nuovo di Legnano, Milan, Italy*
[2] *Otology, Auditory, Implants and Skull Base Surgery Department, Pitié-Salpetriere Hospital, Paris, France*

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, the proper patient and surgeon positioning, the step by step surgery with tips and tricks, results and complications as well as the use the laser and new stapes prosthesis. 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 will be shown. The objective of the course will be understand the classical technique of stapes surgery, recognize and manage difficult cases and identify prognostic factors when dealing with revision stapes surgery.

Study of visual-postural integration

Kaźmierczak W.

Department of Human Physiology, Nicolaus Copernicus University, Collegium Medicum in Bydgoszcz, Poland

In the formation of static and dynamic balance, a properly conducted process of integration of visual and postural stimuli is necessary. It determines the compatible reading of otolith and visual information. The maculae, vestibular nuclei, cerebellum, medullary reticular structures, thalamus, pol 8,18,19 cortex and thalamus play an important role in it. The integration study allows us to assess the function of otoliths and their central representation. It is used a dynamic posturograph with forward-backward and right-left mobility and a gonioscope or goggles giving visual stimuli of the type of saccadic, tracking movements in the horizontal plane and OPK in the sagittal plane in the up and down directions. Saccadic stimuli are administered in the test at a rate of 40/s with an amplitude of 35 degrees from zero position, tracking stimuli at a rate of 2–3 s in both directions, amplitude of 20–35 degrees from zero position, OPK stimulus at a rate of 40 or 60 degrees per second, resulting in 1 to 2 fixation points per second. According to the given test plan, course participants perform the listed registrations, for each test protocol, the wage and height of the test person should be given.

Keynote Lectures

Advanced head and neck carcinoma, surgical options and limitations

Stankovic M.

Medical Faculty, University Clinical Center Nis, Serbia

Locally advanced head and cancer represents significant challenge from surgical, and also from other therapeutical aspects. The data on the incidence, etiology, therapy, and survival rate are widely reported. However, the level I evidence is scant or lacking for the most of the affected areas. Different treatment options have evolved, and the decision process is mainly influenced by the tumor, patient, and physician factors. The dominant factors are nowadays centered on patohistology, TNM status and comorbidity. Different guidelines are regularly updated, NCCN being one of them, all showing treatment options, and their indications. Recurrent malignant disease, whether local relapse, or progression from related or independent field, is particularly difficult to treat. Reference data on treatment of recurrent or persistent malignant disease are particularly scant concerning surgical treatment. Treatment options for locally advanced head and neck cancer are: radical resection with adjuvant chemoradiotherapy, organ preservation, or chemoradiotherapy alone for unresectable or unfit patients. Author presents his surgical experience in the treatment of advanced head and neck cancer. Indications and contraindications for surgical treatment are explained. Examples of wrong surgical approach, neglected advanced tumors, or tumors with unknown primary are

presented. Extensive primary tumor is one of the most important challenges, especially for resectability and reconstruction. Extensive neck metastasis, especially bilateral must be adequate concerning TNM status, with preservation of important neurovascular structures. Secondary post irradiation tumor, affection of multiple locations, preservation of jugular vein, carotid artery, subclavian artery, as well as their reconstruction after resection are explained. Prevention and treatment of chylous fistula is presented. Pharyngocutaneous fistula is a relatively frequent complication of extensive resections, so prevention and proper reconstruction are mandatory. Other surgical topics are also discussed. Future treatment of locally advanced head and neck malignant tumors should be based on improvement of surgical options, but also on metastatic potential, chemoradiosensitivity, and growth speed.

Association of senile hearing loss with cognitive impairment in the Polish cohort of the PURE study

Połtyn-Zaradna K., Pazdro-Zastawny K., Szcześniak D., Basiak-Rasała A., Wołyniec M., Zatońska K., Zatoński T.

Otolaryngology-Head and Neck Surgery Department, Wroclaw Medical University, Wrocław, Poland

Dementia is a growing public health challenge, especially in the context of aging populations. The increase in the number of people with dementia, as predicted by the WHO, requires the identification of risk factors, including age-related hearing loss (ARHL). Studies indicate a significant association between ARHL and cognitive decline, possibly due to social isolation, reduced cognitive reserve, or common vascular lesions in the brain and inner ear, among other factors. The purpose of this study is to analyze this relationship in the PURE study population and to assess the risk and prevalence of ARHL concerning biological and sociodemographic factors of residents of Wroclaw and the surrounding area.

BCI and CI in patients after cholesteatoma surgery

Profant M., Kabátová Z., Varga L.

Comenius University, University Hospital Bratislava, Slovakia

Introduction: Cholesteatoma remains a controversial topic in otology. Optimal surgical technique to manage cholesteatoma is not yet defined. Recurrence rate of cholesteatoma varies significantly. Obliteration techniques nowadays offer improved results. Hearing results in cholesteatoma surgery are significantly worse than those in the non-cholesteatoma tympanoplasty. In this presentation the outcome of bone conduction implants and cochlear implants in cholesteatoma patients are evaluated.

Material and methods: *Bone conduction implants*: we evaluate 25 patients (10 patients with BAHA and 15 patients with Bonebridge). Pre- and postoperative audiograms are evaluated as well as the questionnaire evaluating the quality of life and patient's satisfaction are presented. *Cochlear implants*: in retrospective analysis we evaluated 560 patients with cochlear implantation. There were 7 patients with deafness due to cholesteatoma otitis with 9 cochlear implantations. Following parameters were evaluated: hearing loss, type of middle ear surgery before CI, surgical procedure for CI, complications, hearing results after CI.

Conclusions: BCI are powerful instruments to manage conductive and mixed hearing loss. Functional results and acceptance by patients confirm qualification for clinical use. Individual selection of devices based on audiological evaluation is utmost important. Device selection is also changing with time, new studies and new technology.

CI in patients with chronic otitis and severe hearing loss is an excellent method to restore hearing and communication skills. Different surgical procedures bring different outcomes regarding the problems and complication rate. Optimal procedure seems to be lateral petrosectomy with autologous fat obliteration and ear canal blind closure.

Carcinomas of nasal vestibule: setting new standards from classification to treatment

Bussu F.

Otorhinolaryngology Department, University of Sassari, Italy

Introduction: Nasal vestibule squamous cell carcinoma (NVSCC) is an ill-defined underestimated condition.

Aim: To define the current standard of care.

Material and methods: We review recent acquisitions concerning clinical features and therapeutic approaches.

Results: The current AJCC staging system, which attributes to nasal vestibule the same topographic code as nasal cavity proper and the same T-classification criteria as ethmoid, appears inadequate. As for treatment of primary lesions without bone invasion, current evidence suggests that brachytherapy is at least equivalent to surgery and superior to external beams in terms of oncological outcomes, and superior to both modalities in terms of cosmesis and function.

Conclusions: As for classification and staging, the nasal vestibule should be defined as a subsite of the nose and paranasal sinuses, distinct from the "nasal cavity proper and ethmoid", with specific topographic code and T-classification criteria. This will improve the assessment of prognosis and prevalence, underestimated also because of misdiagnosis with skin cancers.

Secondly, brachytherapy should become the new standard for the treatment of primary lesions without bone invasion. To optimize the advantages of brachytherapy, we propose novel anatomic criteria for the implantation. *Significance*: increasing evidence supports a paradigm shift in staging and treatment of NVSCC.

Cochlear implantation in children with congenital cytomegalovirus infection: what can we learn from the outcomes?

Goh B.-S.[1,2], Lokman F.L.[2], Cho K.-S.[3], Yusoff N.M.[4], Zaki F.M.[5]

[1] *Department of Otorhinolaryngology – Head and Neck Surgery, Faculty of Medicine, University Kebangsaan Malaysia, Kuala Lumpur, Malaysia*
[2] *Department of Otorhinolaryngology – Head and Neck Surgery, Hospital Canselor Tuanku Muhriz, University Kebangsaan Malaysia, Kuala Lumpur, Malaysia*
[3] *Speech and Language Therapy Unit, University Kebangsaan Malaysia Specialist Children's Hospital, Kuala Lumpur, Malaysia*
[4] *Audiology Program, Faculty of Allied Health Sciences, University Kebangsaan Malaysia, Kuala Lumpur, Malaysia*
[5] *Department of Radiology, Faculty of Medicine, University Kebangsaan Malaysia, Kuala Lumpur, Malaysia*

Introduction: Congenital cytomegalovirus (cCMV) infection is a one major cause of sensorineural hearing loss (SNHL) in children under TORCHES infection. This etiology was a relative contraindication for cochlear implantation due to the concerns regarding auditory and speech outcomes due to the co-morbidities and neurological impact of cCMV. This study aims to evaluate auditory performance and speech perception outcomes in children with cCMV infection following cochlear implantation.

Objective: To assess the auditory performance and speech recognition outcomes in children with cCMV infection.

Material and methods: A retrospective review was conducted on 34 children with cCMV infection who underwent cochlear implantation. Patients with either diagnosed with CMV serology, urine or MRI findings suggestive of CMV-related brain abnormalities was reviewed. Pre and post-implantation auditory performance was assessed using the Category of Auditory Performance (CAP-II) and Speech Intelligibility Rating (SIR) score.

Results: Majority of patients showed favourable outcomes following cochlear implantation with CAP-II score of at least 5 at 3 years. Similarly, SIR scores showed improvement in speech intelligibility of at least 3 at 3 years post CI, with many patients attaining intelligibility levels that allow for functional communication. The average CAP-II and SIR score post-implantation indicated improvement which is statistically significant in auditory and speech performance at 3 years and 5 years when compare to preoperative level. Despite the presence of CMV-related brain abnormalities seen on imaging in congenital hearing loss patients, the overall outcomes in terms of auditory performance and speech recognition were encouraging.

Conclusions: This retrospective review showed that cCMV should not be considered as a contraindication for CI. The outcome suggests that CI is an effective hearing rehabilitation for children with congenital CMV infection as they achieve positive auditory and speech outcomes after cochlear implantation. Therefore, all hearing loss children with CMV should have a thorough assessment performed by the CI team and paediatricians, so that the decision on cochlear implantation is made with appropriate expectations. Early intervention is still the key in paediatric hearing loss management.

Cochlear implant revision surgery: analysis of the 30 revision cases

Kalcioglu M.T.[1,2], Celik S.[1,2], Turan N.E.[1,2], Kokten N.[2], Kosaner J.[3]

[1] *Department of Otorhinolaryngology, Istanbul Medeniyet University, Istanbul, Turkey*
[2] *ENT Clinic, Goztepe Prof. Dr. Suleyman Yalcin City Hospital, Istanbul, Turkey*
[3] *Med-El Company, Istanbul, Turkey*

Today, as the frequency of cochlear implant surgery increases, so does the rate of revision surgery. After 722 cochlear implant surgeries, all performed by the same surgeon at our clinic, patients who require revision surgery have gained insight into the reasons and solutions. Since December 2008, the outcomes of patients who required revision after cochlear implant surgery at our clinic and patients who underwent cochlear implant surgery at different centers but were sent to our clinic for revision have been evaluated. Of the 30 patients who required revision, 13 had undergone primary surgery at other centers. Reimplantation was performed in 26 patients. In the remaining 4 cases, in one patient the problem was solved by replacing the magnet in the patient where the magnet was displaced after magnetic resonance imaging (MRI). In the second case of migration, the problem was solved by changing the position of the receiver in the scalp and reimplantation was not required. In the third case, the ground electrode became visible under the skin. The implant was functional and reimplantation was not possible due to financial reasons. The problem was then solved by a strong repair of the defect with a cartilage sliding flap. One patient with bilateral implants refused revision surgery in one ear. In one patient, where the magnet of the implant was displaced after MRI imaging, the problem was solved without surgery by changing the pole of the magnet in the transmitter. Revision surgery was performed in 2 of the 26 patients who underwent reimplantation due to receiver migration after recurrent infections. These patients were reimplanted. Four patients were found to have active electrode-related device failure and 9 patients were found to have trauma-related damage. Device damage assessments were not completed for the remaining patients. In all patients with traumatic damage, revision surgery showed that new bone formation had almost completely covered the mastoidectomy site and the lead had passed through a tunnel in this new bone and advanced into the mastoid region. The mean time from initial surgery to revision was 39.1 months (range 7–101 months). The revision rate was 2.35% (17/722) in patients who underwent primary surgery at our clinic. Trauma and device failure are the main reasons for reimplantation in our clinical series. In order to reduce the need for reimplantation due to trauma, it is predicted that research to reduce the possibility of traumatising the electrode by preventing new bone formation at the mastoidectomy site may help to solve the problem. Periosteal-preserving techniques may be useful to minimise local flap and infection problems.

COMMeND: a mentoring network within the otitis media research community

Santos-Cortez R.L.P., Ziniel S., Mayher L., Lee R., Preciado D., Ryan A.F.

[1] *Department of Otolaryngology-Head and Neck Surgery, School of Medicine, University of Colorado Anschutz Medical Campus, Aurora, CO, USA*
[2] *Department of Pediatrics, Children's Hospital Colorado, Aurora, CO, USA*
[3] *Department of Medicine, School of Medicine, University of Colorado Anschutz Medical Campus, Aurora, CO, USA*
[4] *Departments of Pediatrics and Surgery, George Washington University School of Medicine, Washington DC, USA*
[5] *Department of Otolaryngology, University of California, San Diego School of Medicine and Veterans Affairs Medical Center, La Jolla, CA, USA*

Introduction: Otitis media (OM) is the most common diagnosis among children and remains the main cause of the global burden of hearing loss in children < 5 years old, the age group when the development of speech, language and cognition is most important. OM in childhood also predisposes to sensorineural hearing loss and tinnitus in adults, and ongoing OM may cause permanent hearing loss at any age. OM, therefore, represents a significant health and economic burden, even in developed countries but especially in developing countries where access to advanced and adequate medical care is lacking. However, the false perception that OM is no longer important persists, resulting in less funding for OM research and attrition among OM researchers, with too many of those who completed training migrating to other fields of investigation.

Objective: To present the initiation and progress of the Cross-disciplinary Otitis Media Mentoring Network towards Diversity (COMMeND).

Material and methods: The initial survey assessing feasibility and interest in a mentoring network among otitis media researchers will be described. Additional surveys were also administered among recruited mentees ($n = 14$) and mentors ($n = 21$) during academic year 1.

Results: The development of an otitis media researcher network was funded by the R01 NIH-NIDCD based on preliminary data from the initial survey that demonstrated feasibility and interest in the network. We recruited a diverse network of mentees and mentors, with diversity based on self-reported ethnicity or being part of an under-represented minority in science, gender and career level. Majority of mentees indicated perceived benefit and usefulness of information obtained from bimonthly webinars, and overall satisfaction with mentoring sessions with matched mentors.

Conclusions: Taken together, the COMMeND network has achieved measurable progress towards its goals of enhancing the scientific, career and cultural training of early-career investigators and of strengthening the collaborative environment of the OM community.

Endoscopic DCR with and without silicone intubation

Dehgani-Mobaraki P.[1,2]

[1] *Naso Sano Association, Umbria, Italy*
[2] *Otorhinolaryngology and Head & Neck Surgery, Gubbio-Gualdo Tadino Hospital, Italy*

Introduction: Endoscopic dacryocystorhinostomy (DCR) is a minimally invasive surgical procedure used to treat nasolacrimal duct obstruction (NLDO). A key consideration in endoscopic DCR is whether to use silicone intubation, which may help maintain patency of the newly created lacrimal outflow pathway but also carries potential drawbacks, such as increased inflammation and granulation tissue formation.

Objective: This review aims to compare the outcomes of endoscopic DCR with and without silicone intubation in terms of surgical success rates, symptom resolution, complications, and long-term patency.

Material and methods: A systematic analysis of clinical studies, meta-analyses, and randomized controlled trials was conducted to evaluate the efficacy and safety of endoscopic DCR with and without silicone intubation. Key outcome measures included anatomical success (patency of the nasolacrimal duct), functional success (resolution of epiphora), operative time, and postoperative complications.

Results: Studies indicate that both techniques achieve high success rates, with anatomical and functional success ranging from 80% to 95%. Silicone intubation is particularly beneficial in cases with fibrotic stenosis, canalicular involvement, or revision surgery, as it helps maintain the ostium patency during healing. However, it may be associated with an increased risk of granulation tissue formation, synechiae, and foreign body reaction, which can compromise long-term outcomes. In contrast, primary endoscopic DCR without intubation demonstrates comparable success rates in uncomplicated cases while avoiding the potential drawbacks associated with stent placement.

Conclusions: Endoscopic DCR is an effective treatment for NLDO, with or without silicone intubation. The decision to use intubation should be individualized based on patient factors, the severity of obstruction, and surgical indications. Further high-quality, long-term studies are needed to establish standardized guidelines for the optimal use of silicone intubation in endoscopic DCR.

Epidemiology of HPV-related cancers

Mäkitie A.A.[1,2], Paavonen J.[2]

[1] *Department of Otorhinolaryngology-Head & Neck Surgery, Helsinki University Hospital, Finland*
[2] *University of Helsinki, Finland*

Introduction: The number of new cancers caused by human papillomavirus (HPV) continues to increase in the population that has not been vaccinated against HPV. There is no screening for other HPV-related cancers besides cervical cancer, so monitoring the incidence is important. We aimed to search

for any early decreasing trends in the age-standardized incidence ratios of HPV-related cancers in the Nordic countries.

Material and methods: We examined the age-standardized incidence of HPV-related cancers (cervix, vulva, vagina, anus, penis and oropharynx) in 2015–2023 from both the Finnish Cancer Registry and NORDCAN.

Results: Although the causative role of HPV in these cancers varies, an increase is observed. The age-standardized incidence ratio for cervical cancer showed a plateau or an increasing trend over the study period in Finland and Norway, but a clear decreasing trend in Sweden and Denmark. The annual incidence of oropharyngeal cancer in the Nordic countries is clearly increasing, more sharply in men than in women. This upward trend has been observed in several high-incidence countries in the 21st century. An increase in the incidence of both anal and penile cancer was observed in the Nordic countries, although the numbers are small. For penile cancer only in Denmark a decreasing trend was evident.

Conclusions: The ratios of HVP-related cancers in the Nordic countries are somewhat variable, and no remarkable decreasing incidence can be identified. The increasing cervical cancer rate in Finland was a surprise. Similarly, increasing trends for anal cancer in the Nordic countries were unexpected. The rates of oropharyngeal cancer continue to increase. Around two thirds of them have been reported to be HPV positive. This is in accordance with reports from other high-incidence areas.

Evaluation of the results of transcanal endoscopic ear surgery for treatment of attic retraction pockets

Thuy T.P.C.[1], Thai K.M.[2], Huy L.K.[2]

[1] *University of Health Sciences, Vietnam National University HCMC, Vietnam*
[2] *ENT Hospital, Ho Chi Minh City, Vietnam*

Objective: Assessment of the results of transcanal endoscopic ear surgery in patients with attic retraction pockets.

Material and methods: A descriptive study of 25 patients with attic retraction pockets were performed transcanal endoscopic ear surgery to take out the retractions pockets at Ear Nose and Throat Hospital of Ho Chi Minh City, Vietnam.

Result: In 25 cases, it was able to preserve the mastoid bone with transcanal endoscopic approach, the clinical symptoms improved a lot after surgery. After mean follow-up time 20.28 ± 8.57 months, our success rate is 96% with 1 recurrent case needed second operation. 11/25 cases with ossicular chain discontinuity have undergone ossicular chain reconstruction with mean PTA improved 10.4 dB.

Conclusions: Transcanal endoscopic ear surgery is an effective method for treatment of attic retraction pockets, improve patient's clinical symptoms. Endoscopic surgery helps preserving the mastoid bone and provide good view and control of the attic space. However, during surgery, checking the facial recess was important to prevent recurrence at this place after surgery.

Genetic background of hearing loss in patients with inner ear malformations

Baldyga N.[1,2], Gan N.[1,2], Ozieblo D.[1], Leja M.L.[1], Furmanek M.[3], Skarzynski H.[4], Oldak M.[1]

[1] *Department of Genetics, World Hearing Center, Institute of Physiology and Pathology of Hearing, Warsaw/Kajetany, Poland*
[2] *Doctoral School of Translational Medicine, Centre of Postgraduate Medical Education, Warsaw, Poland*
[3] *Bioimaging Research Center, World Hearing Center, Institute of Physiology and Pathology of Hearing, Kajetany/Warsaw, Poland*
[4] *Oto-Rhino-Laryngology Surgery Clinic, World Hearing Center, Institute of Physiology and Pathology of Hearing, Warsaw/Kajetany, Poland*

Introduction: Inner ear malformations (IEMs) can occur in isolation or as part of genetically determined syndromes. Enlarged vestibular aqueduct (EVA) is the most frequently observed IEM, often accompanied by incomplete partition type 2 of the cochlea and a dilated vestibule. SLC26A4 is the major gene associated with IEMs, with 50% of EVA/IP2 patients carrying either two pathogenic SLC26A4 variants or one SLC26A4 pathogenic variant together with the CEVA haplotype. The genetic background of other IEMs is more diverse and remains poorly understood.

Material and methods: DNA was isolated from blood samples of 40 hearing loss (HL) patients with bilateral IEMs. Sequencing was performed using a custom multigene panel (237 or 263 genes).

Results: Genetic testing identified the cause of IEMs in 67.5% (27/40) of individuals. Among EVA/IP2 patients, six carried two pathogenic variants in *SLC26A4*, while another six had one *SLC26A4* pathogenic variant together with the CEVA haplotype. The genetic cause still remains unknown in six EVA/IP2 patients. In patients with other IEMs, pathogenic variants were identified in *POU3F4* as well as genes associated with CHARGE (*CHD7*), BOR (*EYA1*, *SIX1*), Waardenburg (*SOX10*), and Kabuki (*KMT2D*) syndromes. Additionally, a novel pathogenic variant in *HOXA2* was identified in a single patient.

Conclusions: Mutations in *SLC26A4* and the CEVA haplotype account for more than half of EVA/IP2 cases. Patients with other IEMs are frequently diagnosed with syndromic forms of HL. Further research is needed to identify pathogenic variants in noncoding regions of known HL genes or in novel genes.

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Management of petrous bone cholesteatoma

Szymański M.

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

Petrous bone cholesteatoma is a life threatening disease defined as a presence of squamous epithelium mass in petrous

part of the temporal bone. It may be congenital or acquired. It is also classified as: apical, supralabyrinthine, infralabyrinthine, infralabyrinthine-apical or massive. The symptoms depend on the location and invasion of adjacent structures by the disease. Facial nerve palsy and /or meningitis may be the presenting symptom of the disease.

The treatment involves different surgical techniques adjust to the type of the cholesteatoma and possibility to preserve hearing and/ or facial nerve function. Main features in CT and MRI as well as surgical treatment methods are presented in this lecture.

Mastoid obliteration with bioactive glass: why, when and how?

Di Bari M.[1,2], Alciato L.[2], Delille H.[2], Sterkers O.[2], Colombo G.[1], Bernardeschi D.[2]

[1] *Otorhinolaryngology Department, Ospedale Nuovo di Legnano, Milan, Italy*
[2] *Otology, Auditory Implants And Skull Base Surgery Department, Pitié-Salpetriere Hospital, Paris, France*

This instructional course will delve into the principles and techniques of mastoid obliteration in both canal wall up (CWU) and canal wall down (CWD) mastoidectomy approaches. The course will also encompass the role and technique of obliteration in rehabilitation of an open cavity, subtotal petrosectomy with middle ear exclusion +\– cochlear implant insertion and meningoencephalocele and repair of a tegmen defect. The session will focus on the indications and surgical methods for obliteration, emphasizing the rationale behind these techniques and their impact on clinical outcomes. Participants will gain a comprehensive understanding of the decision-making process, technical nuances, and postoperative considerations essential for successful mastoid obliteration. This course is tailored for otologists aiming to refine their skills and optimize patient care in chronic ear surgery.

Mucin gene regulation by middle ear epithelial cells in otitis media

Ryan A.F.[1], Zemke N.[2,3], Lee E.J.[1], Kurabi A.[1]

[1] *Department of Otolaryngology, University of California San Diego, La Jolla, CA, USA*
[2] *Department of Cellular/Molecular Medicine, University of California San Diego, La Jolla CA, USA*
[3] *Center of Epigenomics, University of California San Diego, La Jolla, CA, USA*

Introduction: Mucus secreted by middle ear (ME) epithelial cells forms a first line of defense against infection. Transmembrane mucins shield the ME mucosa from pathogens. Secreted mucins trap pathogens and export them via the Eustachian tube. Which mucins are produced in the ME, and which epithelial cell types of the five traditional categories (secretory, non-secretory, intermediate, ciliated and basal) produce them before and during otitis media (OM), are not well understood.

Material and methods: We assessed single-cell mRNA expression throughout an episode of acute OM, induced in mice by ME inoculation with nontypeable *Haemophilus influenzae* (NTHi). We also assayed transposon-accessible chromatin (ATAC), which detects DNA accessible for binding by regulatory factors, at 24 hours after NTHi infection. This allowed us to compare altered mucin gene expression with changes in genomic DNA accessibility.

Results: Based on gene expression, we classified MER epithelial cells as high-secretory, low-secretory, intermediate, ciliated and basal. They expressed the transmembrane mucin genes *Muc1*, *Muc4*, *Muc16* and *Muc20*, and well as the secreted mucin genes *Muc5ac* and *Muc5b*. Only transmembrane mucin genes were expressed in the normal ME. This expression increased and secreted mucins were added after infection, peaking 24 hours after inoculation. High-secreting epithelial cells expressed the most mucin, including all six types, but all five epithelial cell types expressed mucins. Infection-related changes in mucin gene DNA accessibility were observed for all six mucin genes.

Conclusions: Strong upregulation of mucin genes throughout an episode of OM by all epithelial cell types indicates less specialization for mucus production than might be expected from their morphology. Changes in DNA accessibility during OM likely participate in up-regulated mucin gene expression. Regulatory proteins that bind to the identified accessible sites would also participate. This includes transcription factors such as NFκB and AP1, both of which were upregulated in epithelial cells during OM.

Perioperative care to reduce intraoperative bleeding in rhinosurgery

Dehgani-Mobaraki P.[1,2]

[1] *Naso Sano Association, Umbria, Italy*
[2] *Otorhinolaryngology and Head & Neck Surgery, Gubbio-Gualdo Tadino Hospital, Italy*

Introduction: Intraoperative bleeding is a significant concern in rhinosurgery, as it can obscure the surgical field, prolong operative time, and increase the risk of complications. Effective perioperative management strategies are essential to optimize surgical outcomes and enhance patient safety.

Objective: This review aims to summarize the current evidence on perioperative strategies to minimize intraoperative bleeding in rhinosurgery, focusing on preoperative preparation, intraoperative techniques, and postoperative considerations.

Material and methods: A comprehensive analysis of perioperative interventions, including patient optimization, pharmacological agents, anesthetic techniques, and surgical approaches, was conducted. Evidence from randomized controlled trials, meta-analyses, and expert guidelines was reviewed to identify best practices.

Results: Preoperative measures such as discontinuation of anticoagulants and antiplatelet therapy when appropriate, blood pressure control, and patient hydration play a crucial

role in reducing bleeding risk. Pharmacological agents like tranexamic acid (TXA), corticosteroids, and topical vasoconstrictors (e.g., epinephrine, oxymetazoline) have shown efficacy in minimizing intraoperative hemorrhage. Anesthetic techniques, including controlled hypotension and total intravenous anesthesia (TIVA), contribute to improved surgical visualization. Intraoperative strategies, such as meticulous hemostasis, bipolar cautery, and modern surgical instruments like microdebriders and powered instruments, further aid in reducing blood loss. Postoperative care, including nasal packing and proper patient monitoring, prevents rebleeding and enhances recovery.

Conclusions: A multimodal approach integrating preoperative, intraoperative, and postoperative strategies is essential for reducing intraoperative bleeding in rhinosurgery. Standardized protocols and individualized patient management can significantly improve surgical efficiency and patient outcomes. Further research is needed to refine best practices and establish evidence-based guidelines for perioperative care in rhinosurgical procedures.

Searching for molecular biomarkers of neuroplasticity in congenital deafness treatment in serum and in perilymph

Matusiak M.[1], Ozieblo D.[2], Oldak M.[2], Rejmak E.[3], Kaczmarek L.[3], Skarzynski H.[1]

[1] *Oto-Rhino-Laryngology Surgery Clinic, World Hearing Center, Institute of Physiology and Pathology of Hearing, Warsaw/Kajetany, Poland*
[2] *Department of Genetics, World Hearing Center, Institute of Physiology and Pathology of Hearing, Warsaw/Kajetany, Poland*
[3] *Laboratory of Neurobiology, Nencki Institute of Experimental Biology, Warsaw, Poland*

Introduction: Molecular and genetic biomarkers of neuroplasticity in congenitally deaf children treated with cochlear implantation (CI) would allow to implement better clinical management, taking into account individual, personalized needs, especially giving them better chances of spoken language rehabilitation.

Aim: The objective of the study was to verify the prognostic value of carrying a certain variant of *MMP-9* gene and plasma level of matrix metalloproteinase 9 (*MMP-9*), measured at cochlear implantation, to the outcome of speech and language rehabilitation after 18 months of CI use in long term follow-up.

Material and methods: We performed a prospective observational study analysis of serum activities of *MMP-9* at CI activation, 8, and 18 months after CI activation in the cohort of 61 children, diagnosed with bilateral profound sensory-neural non-syndromic hearing loss, aged below 2, treated with unilateral cochlear implantation. Language acquisition was assessed with Little Ears Questionnaire (LEAQ). We studied associations between serum activities of *MMP-9* and *BDNF* in the aforementioned intervals and LEAQ scores over follow-up intervals of the implanted children. In the other group of 100 deaf born children enrolled according to the same criteria association analysis of functional MMP9 rs3918242 variant and the child's auditory development measured at CI activation and 1, 5, 9, 14 and 24 months post CI activation with LittlEARS Questionnaire (LEAQ) was conducted.

Results: Correlation analysis shows that there is a significant relation between plasma level of MMP-9 measured at cochlear implantation and LEAQ score in 18 month follow up (rho = –0.25, $p < 0.05$). Statistical analysis in the subgroup implanted after 1 year of life ($n = 53$) showed significant association between MMP9 rs3918242 and LEAQ scores at 1 month ($p = 0.01$), at 5 months ($p = 0.01$), at 9 months ($p = 0.01$) and at 24 months ($p = 0.01$) after CI activation. No significant associations in the subgroup implanted before 1 year of life were observed. Multiple regression analysis ($R^2 = 0.73$) in the subgroup implanted after 1 year of life revealed that MMP9 rs3918242 was a significant predictor of treatment outcome.

Conclusions: *MMP-9* plasma level measured at cochlear implantation below 150 ng/ml predisposes deaf children to good response to cochlear implantation after 18 months follow-up. C/C rs3918242 MMP9 predisposes their deaf carriers to better CI outcomes, especially when implanted after the 1st birthday, than carriers of C/T rs3918242MMP9.

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Stapedotomy step by step: a surgical approach

Skarzynski P.H.[1,2]

[1] *Teleaudiology and Screening Department, World Hearing Center, Institute of Physiology and Pathology of Hearing, Warsaw/Kajetany, Poland*
[2] *Institute of Sensory Organs, Warsaw/Kajetany, Poland*

Introduction: Stapedotomy is a microsurgical procedure performed to treat conductive hearing loss caused by otosclerosis, a condition leading to abnormal bone growth in the middle ear. This procedure involves the creation of a small fenestra in the stapes footplate and placement of a prosthesis to restore sound transmission. Stapedotomy has been shown to significantly improve hearing outcomes, reduce tinnitus, and enhance quality of life in patients with otosclerosis.

Method: Surgical technique – the step-by-step approach to stapedotomy involves several key phases:

1. Patient Preparation and Anesthesia
   – The procedure is typically performed under local anesthesia with sedation or general anesthesia, depending on patient preference and surgical conditions.
   – The ear is prepared using an antiseptic solution, and an operative microscope is positioned for optimal visualization.
2. Tympanomeatal Flap Elevation
   – A postauricular or transcanal approach is used to access the middle ear.
   – The tympanomeatal flap is carefully elevated to expose the middle ear structures, particularly the stapes.
3. Stapes Visualization and Mobilization
   – The incudostapedial joint is identified and separated using otosurgical microinstruments.
   – The stapes suprastructure is removed to provide access to the footplate.

4. Fenestration of the Footplate
– A small hole (fenestra) is created in the stapes footplate using 0,6 mm diamond microdrill to allow sound transmission through the prosthesis.
5. Prosthesis Placement
– A suitable titanium prosthesis is inserted into the fenestra.
– The prosthesis is securely attached to the incus to facilitate effective sound conduction and appropriate sealing with blood cloth.
6. Tympanomeatal Flap Closure and Postoperative Care
– The tympanomeatal flap is repositioned with attachement with fibrin glue, and a small packing of material is placed in the ear canal to stabilize the flap.
– Patients are advised to avoid heavy lifting, straining, and sudden head movements postoperatively.
– Hearing improvement should be typically assessed at follow-up visits after 1, 6 to 12 weeks.

Results: Studies indicate that stapedotomy effectively reduces the air-bone gap, improves auditory thresholds, and enhances the overall quality of life in patients with otosclerosis. Postoperative complications are rare but may include transient dizziness, tinnitus changes, or in rare cases, sensorineural hearing loss. Patient selection, surgical expertise, and appropriate postoperative care play a crucial role in optimizing surgical outcomes.

Conclusions: Stapedotomy is a well-established and effective procedure for the treatment of otosclerosis-related hearing loss. A meticulous, stepwise surgical approach ensures optimal hearing restoration with minimal risks. Further studies on long-term outcomes and innovations in prosthesis design may contribute to enhancing surgical success rates and patient satisfaction.

The changing role of surgery in the multimodality treatment of head and neck cancer

Golusinski P.

Department of Otolaryngology and Maxillofacial Surgery, Collegium Medicum, University of Zielona Gora, Poland

Historically surgery has been a dominant and sole treatment modality of malignancies arising from head and neck anatomical subsites for decades. The advances in other therapeutic modalities has changed the paradigm of present diagnostic and therapeutic protocols worldwide. Apart from disease with very-low locoregional advancement, the multimodality treatment based on the diagnostic work-up and the recommendation of the multidisciplinary team should be offered to the patient. The development of non-surgical approaches based on modern radiotherapy, systemic treatment including immunotherapy has significantly limited the indications for primary surgical treatment. At the same time, the development of reconstructive options has made the surgery the treatment of choice for salvage of the recurrent disease after non-surgical primary management. On the other hand the development of the minimally invasive approaches with the use of the transoral laser and robotic technologies have become again a gold standard treatment alternative for oropharyngeal tumors particularly linked to the HPV infection. The talk is an overview of the evolving role of the use of surgical treatment as an important modality for head and neck cancer patients.

The clinical effect of steroids for hearing preservation in cochlear implantation: conclusions based on three cochlear implant systems and two administration regimes

Skarzynska M.B.[1,2,3], Kolodziejak A.[4], Gos E.[4], Walkowiak A.[5], Lorens A.[5], Pastuszak A.[6], Plichta L.[6], Skarzynski P.H.[2,4]

[1] *Department of Pharmacotherapy and Pharmaceutical Care, Medical University of Warsaw, Poland*
[2] *Institute of Sensory Organs, Kajetany, Poland*
[3] *Center of Hearing and Speech Medincus, Kajetany, Poland*
[4] *Teleaudiology and Screening Department, World Hearing Center, Institute of Physiology and Pathology of Hearing, Warsaw/Kajetany, Poland*
[5] *Implants and Auditory Perception Department, World Hearing Center, Institute of Physiology and Pathology of Hearing, Warsaw/Kajetany, Poland*
[6] *Oto-Rhino-Laryngology Surgery Clinic, World Hearing Center, Institute of Physiology and Pathology of Hearing, Warsaw/Kajetany, Poland*

The main aim of this study was to assess the clinical effect of steroids (dexamethasone and prednisone) on hearing preservation in patients who underwent cochlear implantation with different cochlear implant systems (Oticon®, Advanced Bionics®, Med-El®). 147 adult patients met the inclusion criteria and were enrolled to the study and divided into three groups depending on the brand of cochlear implant they received and participated in all follow-up visits regularly. They were also randomly divided into three subgroups depending on the steroid administration regime: (1) intravenous dexamethasone (0.1 mg/kg body weight twice a day for three days); (2) combined intravenous and oral steroids (dexamethasone 0.1 mg/kg body weight twice a day plus prednisone 1 mg/kg weight once a day); and (3) no steroids (control group). The results were measured by pure tone audiometry (PTA) at three time points: (i) before implantation, (ii) at processor activation, and (iii) 12 months after activation. A hearing preservation (HP) figure was also calculated by comparing the preoperative results and the results after 12 months. Further measures collected were electrode impedance and hearing threshold in the non-operated ear. The highest HP measures (partial and complete) were obtained in the subgroups who were given steroids. Of the 102 patients given steroids, HP was partial or complete in 63 of them (62%). In comparison, partial or complete HP was achieved in only 15 patients out of 45 (33%) who were not given steroids. There were differences between the three cochlear implant groups, with the Med-El and Advanced Bionics groups performing better than the Oticon group (45% and 43% of the former two groups achieved partial or complete HP compared to 20% in the latter). Hearing thresholds in the non-operated ear were stable over 12 months. Generally, impedance was slightly lower in the 12 month follow-up in comparison with the activation period, with the exception of the Oticon group. Conclusions: Pharmacological treatment with steroids in patients undergoing cochlear implantation helps to preserve residual hearing.

Update on middle ear implants

Barbara M.

Sapienza University, Rome, Italy

In selected cases with different severity of hearing loss, the surgical application of several devices could be considered as an alternative to conventional hearing aids (cHA) or even the only feasible rehabilitative procedure. Over the last 20 years, these devices have been included in the armamentarium for the otologist to recover the auditory disability close to normality. This latter can be achieved, for example, by the use of bone conduction devices (BCI) in conductive and mixed type of hearing loss in place of conventional middle ear surgery or ear malformations. Differently from BCI, the clinical introduction of active middle ear implants (AMEI) has been object of controversies since they were generally advised when hearing could not be rehabilitated by cHA, that is more and more unlike with the advent of the new generations of digital HA. Differently, when the deaf population is in search of a cosmetically-acceptable solution, fully-implantable devices can surely be helpful. Over last two decades, AMEI have risen and fallen, abandoning most of them from the clinical availability, also leaving in the otologist's responsibility the handling of the already implanted patients. Moreover, among the major concern is the economic burden that derives from the lack of insurance coverage of the AMEI which justifies their withdrawal by many Companies. Surgical training for their application has also to be taken into account, with differences among the AMEI and their clinical indication, that should be routinely part of any temporal bone training course.

Vestibular dysfunction in the sagittal, frontal and horizontal planes

Kaźmierczak W.

Department of Human Physiology, Nicolaus Copernicus University, Collegium Medicum in Bydgoszcz, Poland

Vestibular dysfunctions both vestibulocochlear and postural in the frontal or sagittal planes evidence damage at the level of the central part of the vestibular structures, this localization also applies to selected symptoms in the horizontal plane. The dysfunctions discussed in the sagittal plane are primarily vertical nystagmus, postural disorders associated with impaired motion perception and spatial orientation Among their etiologies are MS, spinal or cerebellar strokes, degenerative changes of the cerebellum, poisoning, CNS tumors, syringobulbia, head trauma, Arnold Chiari syndrome. It is important to take into account macular damage, here it is possible to meet otolithic vertigo, disorders of vertical OPK, small amplitude of vertical nystagmus in the rotational test. Vestibular disorders in the frontal plane are mainly torsion reaction of the eyeballs, oblique deviation of the eyes and rotational nystagmus. In addition, oblique head tilt, lateropulsions, and vertical reading disorders. Depending on the location of the lesion, these symptoms are either unilateral (bridge lesion below the junction of the graviceptive pathways) or contralateral (midbrain or interbrain lesions). Torsional eye reaction as the only symptom may be present in sternocleidomastoid lesions, in combination with others described above in spinal and midbrain dysfunctions. In the differential diagnosis of,functional vestibular lesions in these planes, unsupported free rotation testing, OVAR, VEMP, vertical and horizontal reading tests, electrootolithography are recommended. The vestibulo-oculomotor symptoms in the horizontal plane are characteristic of receptor-derived appointments. Central dysfunctions manifesting in this plane, in addition to conjunctival nystagmus, atactic nystagmus, positional nystagmus type I or III, include positional nystagmus – divergent and convergent, as well as canal paresis of medium degree according to Dix–Hallpike. In conclusion, it should be emphasized that the discussed functional tests are an essential component of otoneurological diagnosis in addition to structural tests.

Oral Presentations

APOBR is downregulated in EBV+ tonsils of children with obstructive sleep-disordered breathing

Santos R.L.P., Gomez H.Z., Elling C.L., Mayher L., Diala O.R., Gardner C., Willford K., Zamora V.C., Agyepong A., Lee N.K., Green K.K., Darr O.A., Wine T.M., Francom C.R., Larson4E.D., Gitomer S.A., Schell A.E., Frank D.N., Friedman N.R., Herrmann B.W.

[1] *Department of Otolaryngology-Head and Neck Surgery, School of Medicine, University of Colorado Anschutz Medical Campus, Aurora, CO, USA*
[2] *Department of Pediatric Otolaryngology, Children's Hospital Colorado, Aurora, CO, USA*
[3] *Pathways in Genomic Research for Undergraduate Experiences (PATH-GREU) Program, University of Colorado Denver/ Anschutz Medical Campus, Aurora, CO, USA*
[4] *Colorado Research Experiences (CORE) Program, University of Colorado Anschutz Medical Campus, Aurora, CO, USA*
[5] *Department of Basic and Translational Science, School of Dental Medicine, University of Pennsylvania, Philadelphia, PA, USA*
[6] *Division of Infectious Diseases, Department of Medicine, School of Medicine, University of Colorado Anschutz Medical Campus, Aurora, CO, USA*

Introduction: Obstructive sleep-disordered breathing (oSDB) is a heterogeneous phenotype that is increasing in prevalence worldwide and has many potential comorbidities that could severely affect quality of life. There is a need to identify

biomarkers for oSDB and its comorbidities to im-prove clinical management, particularly in children.

Objectives: To describe the tonsillar transcriptome of children with oSDB, according to (1) EBV in tonsil tissues and (2) recommendation for CPAP as treatment.

Material and methods: We performed bulk mRNA-sequencing, differential expression analysis, and qPCR replication of selected differentially expressed genes (DEGs) using RNA samples extracted from tonsils of children with oSDB. Two variables were used as classifier, namely, detection of Epstein-Barr virus (EBV) in tonsils and need for continuous positive airway pressure (CPAP) treatment. Standard statistical tests were used to determine associations across clinical, EBV, and DEG variables.

Results: Nineteen genes were dysregulated in tonsils that are EBV+ or from children needing CPAP. Of these genes, APOBR was downregulated in both EBV+ and CPAP+ tonsils, and this downregulation was replicated by qPCR in an independent set of pediatric samples. In the tonsils of adult patients with oSDB, APOBR was positively correlated with age, and potentially with diastolic blood pressure.

Conclusions: Taken together, APOBR and DEGs in tonsillar tissues may be useful as potential biomarkers of oSDB severity and comorbidity across the lifespan, with APOBR levels being dependent on latent EBV infection.

Cochlear biomechanics after cochlear implantation with hearing preservation

Walkowiak A.[1], Polak M.[2], Lorens A.[1], Skarzynski P.H.[3,4], Skarzynski H.[5]

[1] *Implants and Auditory Perception Department, World Hearing Center, Institute of Physiology and Pathology of Hearing, Warsaw/Kajetany, Poland*
[2] *Med-El, Innsbruck, Austria*
[3] *Teleaudiology and Screening Department, World Hearing Center, Institute of Physiology and Pathology of Hearing, Warsaw/Kajetany, Poland*
[4] *Institute of Sensory Organs, Kajetany, Poland*
[5] *Oto-Rhino-Laryngology Surgery Clinic, World Hearing Center, Institute of Physiology and Pathology of Hearing, Warsaw/Kajetany, Poland*

Introduction: The primary objective is to evaluate if impeded biomechanics affects the hearing preservation after cochlear implantation.

Material and methods: Seventeen adults were implanted with Flex 20 ($n = 1$); Flex 24 ($n = 8$); Flex 28 ($n = 1$); Flex Soft ($n = 5$); Medium ($n = 1$) and Standard electrode array ($n = 1$) with cochlear implants Pulsar, Concerto or Sonata (Med-El). Each of the subjects were implanted using the round window insertion technique. Intracochlear acoustically evoked potentials were recorded from the cochlear implant electrodes. Tone pip of frequency 500 Hz was presented from the inserts and generated by the Nicolet EDX system (Natus). Postoperative CT was performed and evaluated. The audiogram prior the implantation was compared with the audiogram performed at the testing.

Results: Nine patients had the highest amplitude response to 500 Hz tone pip maximum peak matching the 500 Hz excitation area evaluated by the postoperative CT. The impeded biomechanic of basilar membrane was observed in 8 patients. These eight patients had the highest amplitude to 500 Hz tone pip either apically (1 case) or basally shifted (7 cases) from the 500 Hz excitation area evaluated by the postoperative CT. Low frequency pure tone average drop for the tonotopy group of subjects was 12.4 dB, while for the group with impeded biomechanic was 8.4 dB. No difference in mean was found.

Conclusions: These preliminary data suggest that impeded biomechanics of basilar membrane does not necessary influence hearing preservation.

Cochlear implantation in children with congenital herpes virus

Kolodziejak A.[1], Skarzynski P.H.[1,2], Czajka N.[1], Skarzynski H.[3]

[1] *Teleaudiology and Screening Department, World Hearing Center, Institute of Physiology and Pathology of Hearing, Warsaw/Kajetany, Poland*
[2] *Institute of Sensory Organs, Kajetany, Poland*
[3] *Oto-Rhino-Laryngology Surgery Clinic, World Hearing Center, Institute of Physiology and Pathology of Hearing, Warsaw, Poland*

Introduction: The herpes simplex virus belongs to the *Herpesviridae* family (along with cytomegalovirus and varicella-zoster virus). Like cytomegalovirus, it is never completely eliminated from the host's body and has the ability to cause latent infections with periodic reactivation. It is one of the most common viruses, with an infection rate of up to 90% of the population. Depending on the patient's age, the type, and the location of the infection, it can cause a range of clinical symptoms, from mild local infections to life-threatening infections involving the central nervous system. Genital herpes in women is particularly dangerous for the fetus. Infection can occur during fetal life (5–8%), during delivery (85%), or after birth (8–10%). Congenital herpes infection typically results from exposure to the virus during vaginal delivery through contact with genital secretions containing the virus. Infection can also occur via droplet transmission or through lesions caused by the virus. Herpes virus infection can lead to eye and mucous membrane damage, disseminated diseases, intellectual impairment, and hearing loss. The hearing loss that occurs after the infection is sensorineural, and it can be congenital, delayed, or progressive. Therefore, children should undergo routine hearing screening at least once between the 24th and 30th months of life.

Aim: The aim of this study is to present the results of treatment in children who completely lost their hearing due to congenital herpes virus infection and underwent cochlear implantation.

Material and methods: The study group consisted of 10 patients (4 girls and 6 boys) aged from 7 to 56 months, with a mean age of 22.5 months ($SD = 15.222$), operated on

between 2010 and 2020. Five patients were bilaterally implanted (at appropriate time intervals). Eight right ears and seven left ears were operated on. All patients underwent auditory brainstem response (ABR) testing before the surgery. Postoperatively, patients underwent two tests: an adaptive speech understanding threshold test (AAST) and pure tone audiometry in free-field conditions. Nine patients used hearing aids before cochlear implantation, with the average age of hearing aid use initiation being 4.5 months.

Results: Preoperatively, in the ABR test, the hearing threshold for the operated ear was as follows: for 500 Hz, $M = 88.67$ dB; for 1000 Hz, $M = 98$ dB; for 2000 Hz, $M = 98.67$ dB; for 4000 Hz, $M = 99.29$ dB. In the AAST test, the average result in silence was 36.3 dB, and in noise, –10.8 dB SNR. The averaged hearing threshold obtained in pure tone audiometry in the free-field (for frequencies ranging from 250 Hz to 6000 Hz) was 36.5 dB.

Conclusions: Cochlear implantation is an effective method for treating hearing loss in children with congenital herpes virus infection. Patient outcomes may vary depending on the age of implantation, comorbidities, and the rehabilitation approach.

Continuous monitoring of temporal skills during long-term training by cochlear implant users

Szymański K.R., Gawryluk K., Brancewicz M.

Faculty of Physics, University of Bialystok, Poland

Objectives: To evaluate the efficacy of the EARPLANTED platform, a free tool developed for auditory rehabilitation, in monitoring temporal changes in musical perception among cochlear implant users and normally hearing individuals during long-term training.

Material and methods: Participants included 50 cochlear implant users and 45 normally hearing volunteers. The study utilized the EARPLANTED internet application, developed at the Faculty of Physics, University of Bialystok, which features a melodic contour identification test accessible on personal computers and mobile devices. Repeated testing sessions were conducted using the melodic contour identification test. Logistic regression with restricted cubic splines was applied to analyze temporal changes in scores, examining distribution and progression over time.

Results: Continuous temporal monitoring of skills was achieved for binary data (i.e., data representing correct or incorrect melodic contour identification, valued at 0 or 1). Nonlinear dependencies were analyzed, and uncertainties were included. Normally hearing volunteers significantly outperformed cochlear implant users, though many found the test challenging, indicating its inherent complexity. Temporal analysis demonstrated a general improvement in scores among cochlear implant users with extended use of the platform.

Conclusions: The platform has proven to be a valuable tool for monitoring auditory skill development, particularly for unilateral cochlear implant users. The findings emphasize the need for cautious interpretation of melodic contour identification test results due to the difficulty of the task for both cochlear implant users and normally hearing individuals. This study highlights the benefits of continuous monitoring and remote data collection, reinforcing the platform's potential to advance long-term auditory rehabilitation strategies.

COVID-19 – induced acute laryngitis: a case series

Stavridopoulos M., Oikonomou A., Nikitopoulos S., Tsiouma G.

ENT Department, General Hospital of Volos 'Achillopouleio,' Volos, Thessaly, Greece

Objectives: The objective of this study is to present a novel clinical manifestation of COVID-19 with characteristic endoscopic laryngeal findings. A group of patients who reported similar symptoms, displayed akin laryngoscopic features, and received appropriate treatment is analyzed. Endoscopic images are provided and the pattern of this entity is discussed.

Material and methods: This single-center descriptive analysis of a case series was performed in the General Hospital of Volos (Greece), during a 6-month period (from April 2022 to September 2022). Twenty-three patients who suffered from COVID-19 and were simultaneously diagnosed with acute laryngitis were enrolled.

Demographic data, clinical and endoscopic findings, laboratory results, and treatment courses were recorded. Descriptive statistics were performed with the statistical package SPSS (IBM Corp. Released 2017. IBM SPSS Statistics for Windows, Version 25.0. Armonk, NY: IBM Corp.).

Results: The majority of the patients were male and fully vaccinated, as defined by Greek legislation at the time. None of them was a smoker. All patients were infected with Severe Acute Respiratory Syndrome Coronavirus 2 for the first time and presented with acute odynophagia. The characteristic endoscopic finding was an erythematous larynx with white undetachable lesions mainly in the supraglottic area. Pooling of saliva in the pyriform fossae was an independent predicting factor for patients' hospitalization ($P < 0.001$). None of the patients required intubation or tracheostomy and all responded to the systemic treatment with corticosteroids and antibiotics.

Conclusions: COVID-19 – induced laryngitis should be considered in any patient with positive COVID-19 who complains of acute odynophagia. Fiberoptic laryngoscopy is necessary to confirm the diagnosis. In our series, timely initiation of treatment minimized the need to secure the airway and ensured a favorable prognosis.

Exploring the genetic background of autosomal dominant hearing loss

Ozieblo D.[1], Leja M.L.[1], Gan N.[1,2], Baldyga N.[1,2], Sarosiak A.[1], Skarzynski H.[3], Oldak M.[1]

[1] *Department of Genetics, World Hearing Center, Institute of Physiology and Pathology of Hearing, Warsaw/Kajetany, Poland*
[2] *Doctoral School of Translational Medicine, Centre of Postgraduate Medical Education, Warsaw, Poland*
[3] *Oto-Rhino-Laryngology Surgery Clinic, World Hearing Center, Institute of Physiology and Pathology of Hearing, Warsaw/Kajetany, Poland*

Introduction: Autosomal dominant hearing loss (ADHL) is the second most common form of hereditary HL, typically manifesting after the first decade of life. It primarily affects high frequencies and worsens over time. Autosomal-dominant genes account for about 20% of prelingual non-syndromic deafness cases, with 63 genes identified to date.

Material and methods: This study included 110 ADHL families. Targeted next-generation sequencing (NGS) of 237 HL-related genes was performed in all probands. In six large unsolved families, linkage analysis and whole genome sequencing (WGS) were conducted. Presence of the selected probably pathogenic variants and their segregation with HL within the family were confirmed by standard Sanger sequencing.

Results: A genetic cause was identified in 51% (56/110) of families. Among 56 HL variants, 27% (15/56) were previously reported, while 73% (41/56) were novel. Variants included missense (35/56, 62%), splice site (8/56, 14%), frameshift (6/56, 11%), nonsense (6/56, 11%), and one synonymous variant (1/56, 2%). The most frequently affected genes were *MYO6* ($n = 11$), *TBC1D24* ($n = 6$), *WFS1* ($n = 6$), *GSDME* ($n = 5$), *POU4F3* ($n = 5$), and *KCNQ4* ($n = 4$). Variants in *NLRP3*, *LMX1A*, *FGFR3*, *CD164*, *GRHL2*, *TMC1*, *COCH*, *ATP2B2*, and *CEACAM16* were found in single families. Linkage analysis and WGS identified non-coding variants in *EYA4* and *ATP11A* and novel candidate genes.

Conclusions: Our custom multigene panel demonstrated good diagnostic performance. Given the frequent discovery of novel variants, clinical assessment and segregation analysis are essential. Linkage analysis and WGS improve variant detection, particularly in non-coding regions, and help identify new HL genes.

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Functional tests as a means to elucidate the role of variants in hearing loss genes – minigene assay in verification of pathogenicity

Gan N.[1,2], Leja M.L.[1], Ozieblo D.[1], Baldyga N.[1,2], Skarzynski H.[3], Oldak M.[1]

[1] *Department of Genetics, World Hearing Center, Institute of Physiology and Pathology of Hearing, Warsaw/Kajetany, Poland*
[2] *Doctoral School of Translational Medicine, Centre of Postgraduate Medical Education, Warsaw, Poland*
[3] *Oto-Rhino-Laryngology Surgery Clinic, World Hearing Center, Institute of Physiology and Pathology of Hearing, Warsaw/Kajetany, Poland*

Introduction: The next generation sequencing (NGS) is becoming a common tool in diagnostic setting and its especially useful in diagnosing genetically heterogenous diseases, such as hereditary hearing loss (HL). This technique generates thousands of genetic variants for each patients, making it challenging to differentiate between those causative for disease and benign polymorphisms. Notably difficult to interpret are noncoding variants (i.e intronic variants), because their mode of pathogenicity is difficult to determine. Functional studies, such as the minigene assay, might help to define their role in pathogenesis of hereditary hearing loss.

Material and methods: 10 variants detected using dedicated panel of HL genes (237 genes) in HL patients of Genetic Outpatient Clinic of Institute of Physiology and Pathology of Hearing were selected for the minigene assay: *EYA1* c.1475+1G>T, *EYA4* c.1282-12T>A, *GSDME* c.991delT, *GSDME* c.1127A>G, *MYO6* c.816+1G>A, *MYO6* c.1984-1G>A, *MYO6* c.3281-13A>G, *MYO7A* c.2829G>A, *MYO15A* c.9230-4A>T, *SLC26A4* c.1001+1G>A. The fragments of genes of interests were amplified and cloned into expression vector pDEST-pCI-Neo-RHO using Gateway system. Obtained vectors were transfected transiently into HEK293T cell culture. After 48 h of incubation cells were lysed and RNA extraction was performed. Desired transcripts were analysed using RT-PCR and Sanger sequencing.

Results: 9 out of 10 studied variants did show their effect on splicing. Most commonly observed consequence was exon skipping and intron exonisation, resulting mostly in the creation of premature stop codon. Variant MYO15A c.9230-4A>T showed no changes in splicing.

Conclusions: Minigene assay allowed for the determination of the mode of pathogenicity for all studied variants – 9 of them did disrupt the correct splicing of the transcript. In case of variant MYO15A c.9230-4A>T performed assay suggests its benign nature. Splicing may be influenced not only by variants disrupting canonical splice sites but also coding variants and variants in deeper intron sequence. Both isolated HL (*EYA4*, *GSDME*, *MYO6*, *MYO7A*, *MYO15A*) and syndromic HL (*EYA1*, *SLC26A4*) can be caused by splicing affecting variants.

Local delivery of steroids to inner ear via medical device INCAT in partial deafness patients during cochlear implantation

Skarzynska M.B.[1,2,3], Skarzynski P.H.[2,4], Gos E.[4], Lorens A.[5], Walkowiak A.[5]

[1] *Department of Pharmacotherapy and Pharmaceutical Care, Medical University of Warsaw, Poland*
[2] *Institute of Sensory Organs, Kajetany, Poland*
[3] *Center of Hearing and Speech Medincus, Kajetany, Poland*
[4] *Teleaudiology and Screening Department, World Hearing Center, Institute of Physiology and Pathology of Hearing, Warsaw/Kajetany, Poland*
[5] *Implants and Auditory Perception Department, World Hearing Center, Institute of Physiology and Pathology of Hearing, Warsaw/Kajetany, Poland*

Introduction: The administration of steroids to preserve residual hearing during cochlear implantation has been described, although the results are mixed. Nevertheless, according to current knowledge, steroids may have an important role in reducing post implantation fibrosis and loss of hearing due to electrode insertion trauma and progressive effects of inflammation. The aim of the study was to assess separately the effectiveness and safety of three different algorithms of using steroids and INCAT (a medical device) Med-El® in partial deafness patients who underwent cochlear implantation and secondly – the assessment of the impact of the depth of the catheter (INCAT) on hearing preservation after cochlear implantation.

Material and methods: Ten patients underwent a cochlear implantation with an inner ear catheter Steroid administration followed three different algorithms: 1) methylprednisolone 62 5 mg/ml in solution – 3 patients; 2) methylprednisolone 40 mg/ml in suspension – 4 patients 3) dexamethasone 4 mg/ml in solution – 3 patients. Pure tone audiometry (0.125 – 8 kHz) was performed preoperatively and at the cochlear implant activation (one month after surgery). Hearing preservation was assessed according to the HEARRING group formula. Impedance measurements were taken at two days and one month after surgery.

Results: Patients treated with methylprednisolone 40 mg/ml in suspension showed the best hearing preservation, with 50% achieving complete preservation and 50% partial preservation. This group also had the lowest impedance changes (ranging from 1.06 to 2.11 kΩ). A shorter INCAT insertion depth appeared to be more favorable than a longer one. The smallest changes in the hearing thresholds were observed in the second group (methylprednisolone 40 mg/ml in suspension, Depo-Medrol). Hearing preservation (HP) in all patients at the CI activation was as follows: complete hearing preservation (HP) was observed in 2 patients (20%), partial HP in 5 patients (50%), and minimal HP in 3 patients (30%). No patients experienced total hearing loss at the time of CI activation.

Conclusions: All these considerations suggest that patients treated with methylprednisolone 40 mg/ml in suspension had better outcomes compared to others. The generalizability of the results is limited due to the small sample size and the inability to control for.

Otitis media in children with Down syndrome is associated with shifts in the nasopharyngeal and middle ear microbiotas

Elling C.L., Goff S.H., Hirsch S.D., Tholen K., Kofonow J.M., Curtis D., Robertson C.E., Prager J.D., Yoon P.J., Wine T.M., Chan K.H., Scholes M.A., Friedman N.R., Frank D.N., Herrmann B.W., Santos-Cortez R.L.P.

[1] *Department of Otolaryngology-Head and Neck Surgery, School of Medicine, University of Colorado Anschutz Medical Campus, Aurora, CO, USA*
[2] *Department of Pediatric Otolaryngology, Children's Hospital Colorado, Aurora, CO, USA*
[3] *Division of Infectious Diseases, Department of Medicine, School of Medicine, University of Colorado Anschutz Medical Campus, Aurora, CO, USA*
[4] *Center for Children's Surgery, Children's Hospital Colorado, Aurora, CO, USA*

Introduction: Otitis media (OM) is defined as middle ear (ME) inflammation that is usually due to infection. Globally, OM is a leading cause of hearing loss and is the most frequently diagnosed disease in young children. For OM, pediatric patients with Down syndrome (DS) demonstrate higher incidence rates, greater severity, and poorer outcomes. However, to date, no studies have investigated the bacterial profiles of children with DS and OM.

Objective: We aimed to determine if there are differences in composition of bacterial profiles or the relative abundance of individual taxa within the ME and nasopharyngeal (NP) microbiotas of pediatric OM patients with DS ($n = 11$) compared with those without DS ($n = 84$).

Material and methods: We sequenced the 16S rRNA genes and analyzed the sequence data for diversity indices and relative abundance of individual taxa.

Results: Individuals with DS demonstrated increased biodiversity in their ME and NP microbiotas. In children with OM, DS was associated with increased biodiversity and higher relative abundance of specific taxa in the ME.

Conclusions: Our findings indicate that dysbiosis in the ME of children with DS contributes to their increased susceptibility to OM compared with controls. These findings suggest that DS influences regulation of the mucosal microbiota and contributes to OM pathology.

Results of cochlear implantation in patients with congenital rubella

Kolodziejak A.[1], Skarzynski P.H.[1,2], Czajka N.[1], Zdanowicz R.[1], Skarzynski H.[3]

[1] *Teleaudiology and Screening Department, World Hearing Center, Institute of Physiology and Pathology of Hearing, Warsaw, Poland*
[2] *Institute of Sensory Organs, Kajetany, Poland;*
[3] *Oto-Rhino-Laryngology Surgery Clinic, World Hearing Center, Institute of Physiology and Pathology of Hearing, Warsaw, Poland*

Introduction: Congenital rubella syndrome (CRS) is an infection caused by rubella virus that transmitted to the fetus during pregnancy which can cause congenital hearing loss. Cochlear implant can be an effective therapy in patients with severe to profound bilateral hearing loss. The aim of this study was to evaluate the benefits of cochlear implantation in patients with profound hearing loss caused by congenital rubella syndrome.

Aim: The aim of this study was to evaluate the benefits of cochlear implantation in patients with profound hearing loss caused by congenital rubella syndrome.

Material and methods: 38 patients with profound hearing loss caused by intrauterine rubella virus infection were considered for cochlear implantation. Patients ranged in age from 8 to 72 years on the day of surgery, with a mean age of 27 years (*SD* = 13.2). Preoperatively, all patients underwent pure-tone audiometry and was free-field speech audiometry conducted in a quiet environment with the patient wearing a fitted hearing aid. Postoperatively patients underwent pure-tone audiometry, to assess residual hearing, and free-field speech audiometry conducted when the patient had an active implant.

Results: The average preoperative hearing threshold (averaged across the seven frequencies from 0.125 to 8 kHz was 99.2 dB HL (*SD* = 6.79), while the average postoperative hearing threshold was 103.4 dB HL (*SD* = 5.74). Twelve months after the operation patients achieved WRS scores ranging from 10% to 90%, with an average of 59.1% (*SD* = 25.8).

Conclusions: Rubella during pregnancy can lead to severe congenital defects, with sensorineural hearing loss being the most common. Cochlear implants appear to be an effective treatment for profound hearing loss caused by congenital rubella syndrome.

Revisiting musculoclavicular bone graft reconstruction for post-hemimandibulectomy ameloblastoma in a tertiary government hospital: a case series

Coronado J.L., Aragon E., Nonato R.M., Claudio E.R.

Department of Otorhinolaryngology-Head and Neck Surgery, Corazon Locsin Montelibano Memorial Regional Hospital, Bacolod City, Negros Occidental, Philippines

Ameloblastomas are uncommon odontogenic tumors originating from ectodermal epithelium. These tumors are benign in nature but may demonstrate aggressive behavior undergoing malignant transformation. Radical surgical intervention is considered the most effective treatment approach for ameloblastoma but can lead to functional impairments, cosmetic deficiencies, and psychological discomfort. The defect caused by hemimandibulectomy may result in impaired mastication, speech disability, and loss of mandibular contour. Vascularized free bone grafts from the fibula, ilium, scapula, or radius are the gold standard for mandibular repair. In some instances, free flaps may not be feasible within the local context due to lack of expertise, insufficient equipment, or nonviable donor sites among subjects. The paper discussed outcomes associated with musculoclavicular bone graft for the reconstruction of mandibular defect after hemimandibulectomy among mandibular ameloblastoma patients. This is a case series of mandibular ameloblastoma requiring hemimandibulectomy in a tertiary public hospital in the Philippines. All six patients underwent hemimandibulectomy and were subjected to musculoclavicular bone flap reconstruction. Successful integration of the clavicular flap to the mandible was monitored through Orthopantomography done 1 month post-surgery. Two out of six cases exhibited protrusion of the clavicle bone edge in its recipient site, repaired through debridement and rotational skin flaps. Four out of six cases had no post-operative complications. Mandibular contour was maintained with the use of the clavicular flap attached by mandibular titanium plates. Favorable cosmetic results were attained, and the potential for post-operative dental reconstruction remains an option. The musculoclavicular flap remains a viable alternative for reconstructing mandibular defects after hemimandibulectomy, especially in settings where microvascular free flaps are not applicable.

Subjective evaluation of the advantages of the new middle ear implant processor compared to the older generation processors

Ratuszniak A.[1], Lorens A.[1], Obrycka A.[1], Witkowska J.[1], Skarzynski P.H.[2,3], Skarzynski H.[4]

[1] *Implants and Auditory Perception Department, World Hearing Center, Institute of Physiology and Pathology of Hearing, Warsaw/Kajetany, Poland*
[2] *Institute of Sensory Organs, Kajetany, Poland*
[3] *Teleaudiology and Screening Department, World Hearing Center, Institute of Physiology and Pathology of Hearing, Warsaw/Kajetany, Poland*
[4] *Oto-Rhino-Laryngology Surgery Clinic, World Hearing Center, Institute of Physiology and Pathology of Hearing, Warsaw/Kajetany, Poland*

Objectives: Vibrant Soundbridge (VSB) middle ear implant is a partially implantable solution available in Poland for over 20 years for patients with various types of hearing loss. It consists of an internal part (implanted under the skin behind the ear) and an external part (audio processor). As technology advances, audio processors are continually improved to provide users with better auditory performance. The aim of the study is to compare the subjective benefits of using the Samba 2 processor and previous generation processors. The assessment was made using the SSQ12 and APSQ questionnaires.

Material and methods: Forty-five experienced VSB users (average VSB use time 9 years, *SD* = 2) who replaced their previously used processor (D404, Amadé©, Samba 1) with the model Samba 2. The average age is 56 years, *SD* = 20 years.

Results: The results of the questionnaire evaluation confirm the benefits of using the latest processor compared to the previous generation processors. On the SSQ questionnaire, both the total score and the subscale scores were statistically significantly better on the new processor than on the previous generation processor. In the APSQ questionnaire, statistically significant differences were observed for the Usability subscale.

Conclusions: The new audio processor is subjectively rated better than the previous generation processors. Access to modern technologies for patients with VSB implants brings measurable benefits.

Success of targeted sequencing in the search for genetic causes of Usher syndrome type 2

Baldyga N.[1,2], Ozieblo D.[1], Reurink J.[3], Skarzynski H.[4], Kremer4H.[3,5], Oldak M.[1]

[1] *Department of Genetics, World Hearing Center, Institute of Physiology and Pathology of Hearing, Warsaw/Kajetany, Poland*
[2] *Doctoral School of Translational Medicine, Centre of Postgraduate Medical Education, Warsaw, Poland*
[3] *Department of Human Genetics, Radboud University Medical Center, Nijmegen, The Netherlands*
[4] *Oto-Rhino-Laryngology Surgery Clinic, World Hearing Center, Institute of Physiology and Pathology of Hearing, Warsaw/Kajetany, Poland*
[5] *Department of Otorhinolaryngology, Radboud University Medical Center, Nijmegen, The Netherlands*

Objectives: Usher syndrome is one of the most common rare diseases in which both hearing impairment and retinitis pigmentosa coexist. Currently, four types of Usher syndrome are known and they are genetically and clinically heterogenous. The aim of the study was to characterize the genetic background of Usher syndrome type 2 (USH2) in a group of Polish patients.

Material and methods: A total of 55 patients with a clinical diagnosis of USH2 were recruited to the study. The DNA was isolated from blood and genetic testing was performed using three different methods: RT-PCR genotyping, targeted sequencing of the USH2A gene, and a multigene panel (237 genes). Segregation analysis was performed using Sanger sequencing and RT-qPCR.

Results: The cause of USH2 was identified in all patients. In 98% (54/55) of the individuals, causative variants were located in the *USH2A* gene. In one patient (2%; 1/55), a new homozygous terminating variant in the *ADGRV1* gene was identified. In *USH2A* gene, 42 different genetic variants were identified (28 known and 14 novel). A total of 74% (31/42) of the variants were deleterious. The most frequently identified genetic cause of *USH2* was c.11864G>A (p.Trp3955Ter) (29 alleles). Deletions of exons 22–24 (17 alleles) and 10–11 (8 alleles) of the *USH2A* gene also contributed frequently to *USH2* development.

Conclusions: Genetic testing of *USH2* patients should be based on high-throughput tests that enable simultaneous identification of SNVs and CNVs. The gathered data can serve as a starting point for further genotype-phenotype association analyses.

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Taste disorders after various otosurgical operations

Buksinska M.[1], Skarzynski P.H.[2,3]

[1] *Oto-Rhino-Laryngology Surgery Clinic, World Hearing Center, Institute of Physiology and Pathology of Hearing, Warsaw, Poland*
[2] *Teleaudiology and Screening Department, World Hearing Center, Institute of Physiology and Pathology of Hearing, Warsaw/Kajetany, Poland*
[3] *Institute of Sensory Organs, Kajetany, Poland*

Introduction: One of the nerves involved in the transmission of taste stimuli is the chorda tympani. This nerve runs through the tympanic cavity. During middle ear surgery, the chorda tympani can become irritated or damaged, which can be associated with taste disorders.

Aim of the study: To assess the incidence of taste disorders after different types of otosurgery.

Material and methods: Patients who required middle ear surgery were included in the study. Before surgery, a screening Taste Test was performed in patients. Those with normal results were given a retest one week after surgery (97 participants). The study participants were divided into three groups according to the type of surgery performed. Group 1 included patients who underwent surgery with access to the tympanic cavity via anterior tympanotomy, except for stapedotomy. Patients in group 2 underwent stapedotomy, while patients in group 3 required a posterior tympanotomy.

Results: One week after surgery, taste disturbances were detected in 17 patients (17.5%). The results in each group were as follows: group 1–4 patients (10.3%) with taste disorders after surgery, group 2–9 patients (27.3%), group 3–4 patients (16.0%). 3 months after surgery, the disorder persisted in 1 person (1.0%). None of the operated patients had damage to the chorda tympani during surgery.

Conclusions: Taste disorders can occur after any middle ear surgery. They are more common in patients undergoing stapedotomy than other surgeries, which may be related to the course of the chorda tympani and the surgical technique, requiring widening of the external auditory canal posteriorly.

The AMSA® manosonic nebulizer for ENT diseases among children in Poland

Skarzynska M.B.[1,2,3], Gos E.[4], Dominici Sanfins M.[4,5], Hartwich P.[6], Skarzynski P.H.[3,4], Szkielkowska A.[7]

[1] *Department of Pharmacotherapy and Pharmaceutical Care, Medical University of Warsaw, Poland*
[2] *Center of Hearing and Speech Medincus, Kajetany, Poland*
[3] *Institute of Sensory Organs, Kajetany, Poland*
[4] *Teleaudiology and Screening Department, World Hearing Center, Institute of Physiology and Pathology of Hearing, Warsaw/Kajetany, Poland*
[5] *Department of Speech-Hearing-Language, Federal University of São Paulo (UNIFESP), São Paulo, Brazil*
[6] *Department of Otolaryngology, Jagiellonian University, Collegium Medicum, Krakow, Poland*
[7] *Audiology and Phoniatric Clinic, World Hearing Centre, Institute of Physiology and Pathology of Hearing, Warsaw/Kajetany, Poland*

Introduction: The AMSA® manosonic nebulizer uses acoustic vibration and a flow of air to create an aerosol from a solution or suspension of a drug. The aerosol created this way is claimed to have enhanced penetration and drug delivery. It is administered under short-term overpressure, meaning that the aerosol is able to penetrate into the middle ear through the Eustachian tube (ET).

Aim: The aim of this study is to identify the active substances used in AMSA® manosonic nebulization for treating common ENT diseases in children aged 2–17 years and to evaluate the overall effectiveness of AMSA® manosonic nebulization in this context. Assessments were done by comparing conditions before and after nebulization using the following tests: (1) Eustachian tube function test, (2) tympanometry, and (3) otoscopy.

Material and methods: This study was a retrospective study with ethics committee consent. 129 children, comprising 56 girls and 73 boys. They were aged between 2 and 17 years, with a mean age of 6.9 years (*SD* = 3.0). There were 74 children up to 6 years and 55 children over 6 years of age. Children had the following conditions: (1) chronic otitis media with effusion, OME ($n = 86$), (2) Eustachian tube dysfunction, ETD ($n = 34$), (3) other conditions, e.g., cholesteatoma, retraction pocket ($n = 9$). Combination of medicines administered in this study was: Budesonide + Ambroxol (with or without NaCl), Budesonide (with or without NaCl), Budesonide + N-acetylcysteine (with or without NaCl), Budesonide + Hyaluronic acid, Budesonide + Ambroxol (with hyaluronic acid), Ambroxol (with or without NaCl).

Results: The number of nebulizations ordered was between 1 and 20 treatments, but most commonly, patients were given a nebulization series of 10 treatments. This was the case for 80.6% of the patients. Most patients with OME and ETD had 10 treatments ordered (79% and 79.5%, respectively), while all patients with other conditions had 10 treatments. Analysis of the tympanometry results was done in terms of the number of affected ears (not by individual). There were 210 ears with complete tympanometry (both pre and post), including 142 ears with OME, 54 with ETD, and 14 others. Statistically significant changes (improvements) after AMSA nebulizations were found for statistic compliance and middle ear pressure. Otoscopy assessments were done in all ears. The results were abnormal in 155 ears (73.8%) and normal in 55 ears (26.2%). After AMSA nebulizations, the number of abnormal results decreased to 117 ears (55.7%) and normal results were found in 93 ears (44.7%).

Conclusions: Use of the AMSA manosonic nebulizer appears to be an effective way of improving chronic medical conditions in children – such as chronic otitis media with effusion and Eustachian tube defect – but only if patient compliance can be achieved. The most frequently used active substance was budesonide, irrespective of whether additional secretolic/mucolytic agent was administered.

The Bonebridge active bone conduction hearing implant: safety, effectiveness and outcomes based on a largest cohort study – 355 patients

Skarzynski P.H.[1,2], Cywka K.B.[3], Czaplicka E.[1], Skarzynski H.[4]

[1] *Teleaudiology and Screening Department, World Hearing Center, Institute of Physiology and Pathology of Hearing, Warsaw/Kajetany, Poland*
[2] *Institute of Sensory Organs, Kajetany/Warsaw, Poland*
[3] *World Hearing Center, Institute of Physiology and Pathology of Hearing, Warsaw/Kajetany, Poland*
[4] *Oto-Rhino-Laryngology Surgery Clinic, World Hearing Center, Institute of Physiology and Pathology of Hearing, Warsaw/Kajetany, Poland*

Introduction: Hearing loss is often associated with a lower quality of life, leading to communication difficulties, social isolation and stigma. The scale of difficulties faced by people with hearing loss has, over the years, driven professionals to seek increasingly advanced treatments. Rapid technological advancements have enabled the development of implantable devices for patients who do not benefit from or cannot use traditional hearing aids. The Bonebridge implant is a major advancement in bone conduction technology, offering a safe and effective hearing loss solution. However, limited sample sizes in studies highlight the need for further research on its long-term efficacy and safety.

Aim: The aim of this study is to evaluate the safety, efficacy and audiological outcomes of the Bonebridge implant in a large cohort of patients with different types of hearing loss.

Material and methods: A total of 355 patients across a wide age range underwent Bonebridge implantation. Pre- and post-implantation evaluations included pure-tone audiometry, speech recognition tests and free-field audiometry. Word recognition was measured using the Polish Monosyllabic Word Test, while speech reception in noise was assessed using the Polish Sentence Matrix Test. Subjective benefit was assessed using the APHAB questionnaire. Follow-up tests were performed 3–6 months after activation.

Results: Revision surgery was required in 17 patients (4.8%) due to complications, including implant removal in five cases. Reimplantation was successful in four of these cases. The APHAB questionnaire showed improved hearing function and all hearing tests such as free field thresholds, speech discrimination and matrix tests showed significant improvement after implantation.

Conclusions: Active bone conduction implantation is an effective method for the rehabilitation of conductive hearing loss, mixed hearing loss and unilateral deafness. The large cohort study confirms significant hearing improvements and subjective benefits. The low complication rate supports the reliability of the Bonebridge system.

The role of the rhomboid lip and choroid plexus in microvascular decompression (MVD) for vestibulocochlear diseases

Tursun M.

Department of Neurosurgery, First Affiliated Hospital of Xinjiang Medical University, Ürümqi, China

Introduction: Microvascular decompression (MVD) is a surgical intervention for functional vestibulocochlear diseases, but its efficacy and the underlying mechanisms remain incompletely understood. Recent studies have focused primarily on neurovascular compression, while other potential factors, such as abnormalities in the rhomboid lip (RL) and choroid plexus (CP), have been largely overlooked. This study investigates the role of RL and CP in the development of vestibulocochlear symptoms and evaluates the surgical outcomes of MVD with and without addressing these structures.

Material and methods: We retrospectively reviewed 15 patients who underwent MVD for vestibulocochlear diseases between 2013 and 2022. Patients were categorized into four groups: vestibular paroxysmia (VP), benign positional paroxysmal vertigo (BPPV), Ménière disease (MD), and a "tinnitus" group. Preoperative imaging, intraoperative findings, symptom relief, recurrence rates, and patient satisfaction were analyzed. Additionally, we evaluated the impact of RL incision and CP excision on surgical outcomes.

Results: Following MVD, 6 of 7 patients with VP, 1 patient with BPPV, and 1 of 2 patients with MD achieved complete relief from vertigo. Notably, 4 patients who underwent RL incision and CP excision were also free of vertigo, despite the absence of confirmed vascular compression. In these cases, we hypothesize that abnormalities in the RL and CP, such as an elongated RL and overexpanded CP, may contribute to vertigo by obstructing cerebrospinal fluid (CSF) flow and stimulating the eighth nerve. Furthermore, some patients reported improved hearing postoperatively, suggesting that addressing these anatomical abnormalities may have additional benefits beyond vertigo relief. However, patients in the "tinnitus" group showed no significant improvement, indicating that tinnitus may have different underlying mechanisms and requires further investigation.

Conclusions: Our study highlights the potential role of RL and CP abnormalities in the pathogenesis of vestibulocochlear diseases. MVD is effective for treating VP and may also benefit patients with BPPV and MD, especially when RL and CP abnormalities are addressed. These findings suggest that preoperative imaging of the RL and CP should be considered in patients with refractory vestibulocochlear symptoms. Future research should focus on elucidating the anatomical and physiological interactions between these structures and the eighth nerve, as well as exploring targeted interventions to improve surgical outcomes.

Posters

Brown–Vialetto–Van Laere Syndrome – a case report of riboflavin treatment and cochlear implants in a 4-year-old girl with progressive hearing loss and delayed speech development

Piecuch A.K.[1], Skarzynski P.H.[2,3], Skarzynski H.[1]

[1] *Oto-Rhino-Laryngology Surgery Clinic, World Hearing Center, Institute of Physiology and Pathology of Hearing, Warsaw/Kajetany, Poland*
[2] *Teleaudiology and Screening Department, World Hearing Center, Institute of Physiology and Pathology of Hearing, Warsaw/Kajetany, Poland*
[3] *Institute of Sensory Organs, Kajetany, Poland*

Introduction: Brown–Vialetto–Van Laere (BVVL) syndrome is a rare autosomal recessive disorder caused by mutations in intestinal riboflavin transporter genes, resulting in a motor neuron disorder of childhood, which can be associated with sensorineural deafness. This report describes a 4-year-old Polish girl with progressive hearing loss and delayed speech development diagnosed with Brown–Vialetto–Van Laere syndrome who was treated with riboflavin (vitamin B2) and cochlear implants.

Case report: The case report concerns a girl from Poland who, at the age of 2 years 10 months, developed progressive atypical neurological symptoms of unknown etiology: ataxia of the upper and lower limbs, gait abnormalities, generalized muscle weakness, visual and hearing problems, and regression of speech development. A karyotype study (whole-exome sequencing) revealed alterations within SLC52A2, leading to the diagnosis of Brown–Vialetto–Van Laere syndrome and initiation of high-dose riboflavin treatment. As a 4-year-old child, she presented to the Institute of Physiology and Pathology of Hearing, Poland with progressive hearing loss and speech regression. Hearing tests revealed bilateral profound sensorineural hearing loss with auditory neuropathy. Surgical treatment was applied in the form of bilateral cochlear implantation.

Conclusions: This report shows the importance of genetic testing in infants who present with atypical symptoms or signs. In this case, the diagnosis of Brown–Vialetto–Van Laere syndrome resulted in timely correction of the genetic riboflavin (vitamin B2) deficiency and improved hearing following the use of cochlear implants.

Disrupted *GRHL2* transcriptional activity as a mechanism of autosomal dominant hearing loss development (DFNA28)

Baldyga N.[1,2], Ozieblo D.[1], Leja M.L.[1], Skarzynski H.[3], Oldak M.[1]

[1] *Department of Genetics, World Hearing Center, Institute of Physiology and Pathology of Hearing, Warsaw/Kajetany, Poland*
[2] *Doctoral School of Translational Medicine, Centre of Postgraduate Medical Education, Warsaw, Poland*
[3] *Oto-Rhino-Laryngology Surgery Clinic, World Hearing Center, Institute of Physiology and Pathology of Hearing, Warsaw/Kajetany, Poland*

Introduction: *GRHL2* is one of over 50 genes linked to autosomal dominant hearing loss (ADHL) and is also implicated in cancers. It encodes a transcription factor. Only few ADHL-related *GRHL2* pathogenic variants have been reported and their mode of action remains unclear. This study aimed to identify the genetic basis of ADHL in a multigeneration family with progressive hearing loss (HL) and explore the molecular mechanism of DFNA28 *GRHL2* mutations.

Material and methods: Genomic DNA from the proband and family members ($n = 8$) was analyzed using a 237-gene HL panel and Sanger sequencing. Expression vectors for four ADHL-related *GRHL2* variants were generated, and their transcriptional activity was assessed in HEK293T cells using a luciferase reporter assay.

Results: A novel heterozygous *GRHL2* variant (c.1061C>T; p.Ala354Val) segregating with HL was identified within the DNA-binding domain. Functional assays showed reduced transcriptional activity for this variant and two others (c.1258-1G>A, p.Gly420Glufs*111; c.1276C>T, p.Arg426*). In contrast, a variant affecting the dimerization domain (c.1609_1610insC, p.Arg537Profs*11) strongly activated the GRHL-responsive promoter.

Conclusions: The pathogenicity of novel missense variant was supported by functional assays. *GRHL2* mutations can suppress or activate transcription depending on variant location – DNA-binding domain variant causes haploinsufficiency, while dimerization domain variant shows gain-of-function effect. This study provides new insights into *GRHL2*-related hearing loss mechanisms.

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Duplication of internal auditory canal as a pathognomonic sign of severe cochlear nerve hypoplasia or aplasia and Bone Conductive Implant Bonebridge implantation in CROS configuration – a case report

Piecuch A.[1], Cywka K.B.[2], Skarzynski P.H.[3,4], Skarzynski H.[1]

[1] *Oto-Rhino-Laryngology Surgery Clinic, World Hearing Center, Institute of Physiology and Pathology of Hearing, Warsaw/Kajetany, Poland*
[2] *World Hearing Center, Institute of Physiology and Pathology of Hearing, Warsaw/Kajetany, Poland*
[3] *Teleaudiology and Screening Department, World Hearing Center, Institute of Physiology and Pathology of Hearing, Warsaw/Kajetany, Poland*
[4] *Institute of Sensory Organs, Kajetany, Poland*

Introduction: Duplication of the internal auditory canal, i.e. separate canals for the cochlear and facial nerves, is a very rare congenital anomaly arising during embryonic development. The aim of this study was to present the case of a patient with internal auditory canal duplication (separate canals for the cochlear and facial nerves), unilateral cochlear nerve aplasia, and a method of treatment for single-sided deafness (SSD) with a Bonebridge implant as CROS (contralateral routing of signal).

Case report: The Institute of Physiology and Pathology of Hearing, Poland was visited by a 14-year-old female patient who was diagnosed with unilateral hearing loss during a school balance sheet. Her hearing screening at birth was normal. Pure tone audiometry revealed unilateral deafness of the right ear. Computed tomography scan showed asymmetry of the internal auditory canals: the right canal was duplicated – divided into a separate canal for the facial nerve (2.2 mm) and the vestibulocochlear nerve (<1 mm). Magnetic resonance imaging confirmed a right bipartite canal with severe stenosis of the duct to VIII nerve, without identification of the VIII nerve on that side. Unilateral aplasia or severe hypoplasia of the right nerve VIII was suspected. In view of the test results obtained, the patient was referred for diagnosis of an implant using bone conduction as a CROS, taking advantage of the phenomenon of bone conduction of sound from the deaf ear to the well-functioning ear. During the simulation performed with a bone conduction sound device mounted on a soft band, the patient achieved MATRIX test results (in SSD configuration): with the device SRT threshold = –0.4 dB SNR, without the device SRT = 3.5 dB SNR. At the age of 14, the patient was implanted with a Bonebridge 602 implant in the right ear as a CROS. During implant activation in the Matrix test with the Bonebridge implant, the patient achieved SRT= –10.3 dB SNR.

Conclusions: Duplication of the internal auditory canal is pathognomonic for severe cochlear nerve hypoplasia or aplasia, which may have a significant impact on the choice of treatment and implant. It is essential to perform an imaging study before deciding on implantation. In the case of an isolated canal anomaly without a cochlear defect, hearing screening at birth may not detect a hearing loss. In this defect, it is possible to record otoacoustic emissions in an unaltered cochlea with impaired conduction through the hypo- or aplastic auditory nerve, as the embryogenesis of the inner ear and the auditory canal proceeds independently. In the case of a unilateral anomaly with no hearing impairment on the opposite side, bone conduction implantation should be considered as a CROS.

Effectiveness of bone conduction implants in patients with single-sided deafness (SSD)

Skarzynski P.H.[1,2], Cywka K.B.[3], Czaplicka E.[1], Skarzynski H.[4]

[1] *Teleaudiology and Screening Department, World Hearing Center, Institute of Physiology and Pathology of Hearing, Warsaw/Kajetany, Poland*
[2] *Institute of Sensory Organs, Kajetany, Poland*
[3] *World Hearing Center, Institute of Physiology and Pathology of Hearing, Warsaw/Kajetany, Poland*
[4] *Oto-Rhino-Laryngology Surgery Clinic, World Hearing Center, Institute of Physiology and Pathology of Hearing, Warsaw/Kajetany, Poland*

Introduction: Single-sided deafness (SSD) significantly affects speech understanding in noisy environments and spatial hearing, reducing overall quality of life. Traditional hearing solutions such as contralateral routing of sound (CROS) hearing aids or cochlear implants may not be suitable for some patients due to anatomical constraints or intolerance. Bone conduction implants offer an alternative rehabilitation approach by transmitting sound from the non-hearing side to the functioning cochlea. This study evaluates the audiological and subjective benefits of active transcutaneous bone conduction implants in SSD patients.

Material and methods: A prospective study was conducted at a tertiary referral center, including 40 participants aged 13 to 48 years (mean: 32.5 years) diagnosed with SSD. The leading causes of SSD were congenital deafness and mumps-related deafness. All participants were deemed unsuitable for CROS hearing aids or cochlear implants due to anatomical limitations or intolerance. Each patient underwent surgical implantation of a bone conduction device on the deaf side, allowing sound transmission to the functioning ear. Speech recognition in noise was assessed using the Polish Sentence Matrix Test. Patient satisfaction and quality of life were evaluated before and after implantation using the Abbreviated Profile of Hearing Aid Benefit (APHAB) questionnaire.

Results: Patients demonstrated a significant improvement in speech recognition in noisy environments. The mean Polish Sentence Matrix Test scores improved from +0.4 dB SNR at the initial assessment to –3.9 dB SNR six months after activation. The APHAB questionnaire results indicated enhanced communication abilities, with a mean benefit score of 20.5 points. The surgical procedures were well tolerated and no major complications were reported.

Conclusions: Bone conduction implants provide an effective and safe rehabilitation option for SSD patients, significantly enhancing speech understanding in noisy environments and overall quality of life. Early intervention with this technology may improve hearing outcomes in individuals with SSD who are not candidates for traditional hearing solutions. Further research is needed to assess long-term benefits and refine patient selection criteria.

First results of the new bone conduction implant SENTIO in patients with conductive hearing loss and patients with mixed hearing loss

Skarzynski P.H.[1,2], Cywka K.[3], Balwicka-Kowalczyk M.[1]

[1] *Teleaudiology and Screening Department, World Hearing Center, Institute of Physiology and Pathology of Hearing, Warsaw/Kajetany, Poland*
[2] *Institute of Sensory Organs, Kajetany, Poland*
[3] *World Hearing Center, Institute of Physiology and Pathology of Hearing, Warsaw/Kajetany, Poland*

Poland is the third country in the world and the second in Europe where the Sentio implant has been used. The paper describes the efficacy of the implant in a group of 20 patients diagnosed with conductive or mixed hearing loss. The Sentio device is the smallest transcutaneous implant available on the market and consists of an external Sentio 1 Mini sound processor and a Sentio Ti implant placed under the skin. The system works by transmitting vibrations through the skull bone to the inner ear, which is particularly helpful for patients with problems in the outer ear (microtia, atresia). Sentio is also used in patients with hearing loss due to chronic ear infections and after surgery. The size of the device opens up a wide field for a large group of people with hearing loss, including younger patients in the future. The system is comfortable to wear due to its size and convenience. Bone conduction overcomes the limitations of hearing loss in the outer and/or middle ear. Patients implanted with the Sentio implant experience a significant improvement in hearing in a variety of acoustic conditions. Audiological tests show improved hearing and speech understanding. Patients report improvements in sound localisation and reduced noise-induced fatigue. This is confirmed by the Abbreviated Profile of Hearing Aid Benefit (APHAB) questionnaire, which patients complete before and after implantation. Verbal audiometry using Demenko and Pruszewicz one-syllable words was used to assess speech in quiet. Speech understanding in noise was assessed using the Polish Sentence Matrix Test (SNR). The many advantages of the Sentio system make bone conduction technology particularly valuable for people with certain hearing problems who cannot use conventional hearing aids.

Long-term results of bilateral cochlear implantation in a patient with congenital inner ear malformation: incomplete partition type I – a case report

Piecuch A.[1], Skarzynski P.H.[2,3], Skarzynski H.[1]

[1] *Oto-Rhino-Laryngology Surgery Clinic, World Hearing Center, Institute of Physiology and Pathology of Hearing, Warsaw/Kajetany, Poland*
[2] *Teleaudiology and Screening Department, World Hearing Center, Institute of Physiology and Pathology of Hearing, Warsaw/Kajetany, Poland*
[3] *Institute of Sensory Organs, Kajetany, Poland*

Introduction: Incomplete partition type I (IP I), also known as cystic cochleovestibular anomaly (CCVA), accounts for approximately 20% of congenital defects of the inner ear and is characterised by: a cochlea lacking the modiolus and spiral lamina, giving it the appearance of an 'empty cyst', an enlarged cystic vestibule and a wide connection between them. The modiolus defect may be subtotal or total, with the latter – total modiolus aplasia – resulting in communication between the cochlea and the internal auditory canal, with associated hypertension of the inner ear fluids and an increased risk of recurrent meningitis. The pathomechanism of the defect is most likely due to the defective structure of the inner layer of the cochlea (endosteum), which is abraded or absent from the cochlea and vestibule, resulting in a defect in the stapes plate.

Case report: A 13-month-old female patient presented to the Institute of Physiology and Pathology of Hearing, Poland with bilateral profound hearing loss. During surgery, after opening the mastoid process, a defect in the area of the stapes plate was found to be leaking intense fluid – the fistula was successfully repaired. CT and MR imaging revealed a bilateral congenital inner ear defect of incomplete partition type I. Treatment with bilateral cochlear implants followed. During 12 years of follow-up, the patient achieved a speech understanding threshold of mild hearing loss with both implants.

Conclusions: Patients with congenital malformation of the inner ear (incomplete partition type I) may be suitable candidates for cochlear implantation. The results of implantation after long-term follow-up may be satisfactory and comparable to those of patients with other defects. Pre-operative CT and MRI scans are essential to better plan for possible intra-operative complications, fluid leak, stapes plate defect, selection of an appropriate implant electrode. Of particular note in intraoperative management are careful inspection of the stapes plate for a defect with CSF leakage, with possible removal of the cyst and provision of a fistula to prevent the development of recurrent meningitis, and careful sealing of the cochleostomy after insertion of the cochlear implant electrode.

Removal of a metallic foreign body from the Eustachian tube by anthromastoidectomy – posterior tympanotomy in a welder with a cerebellopontine angle tumour: a case report

Piecuch A.[1], Mrówka M.[1], Skarzynski P.H.[2,3], Skarzynski H.[1]

[1] *Oto-Rhino-Laryngology Surgery Clinic, World Hearing Center, Institute of Physiology and Pathology of Hearing, Warsaw/Kajetany, Poland*
[2] *Teleaudiology and Screening Department, World Hearing Center, Institute of Physiology and Pathology of Hearing, Warsaw/Kajetany, Poland*
[3] *Institute of Sensory Organs, Kajetany, Poland*

Introduction: Metallic spatters generated during welding that reach small sizes, high velocities and high temperatures can penetrate the middle ear and the Eustachian tube.

Case report: A case report of the surgical treatment of a welder treated at the Institute of Physiology and Pathology of Hearing, Poland who had to remove a ferromagnetic foreign body from the Eustachian tube due to the need to monitor a tumor of the cerebellopontine angle. This is the first such case report of removal of a metallic foreign body from the Eustachian tube by anthromastoidectomy – posterior tympanotomy.

Conclusions: Difficult to heal chronic otitis externa or otitis media in a welder should prompt a search for a retained foreign body in this location. If the foreign body has penetrated the Eustachian tube, there is a possibility of removal by anthromastoidectomy – posterior tympanotomy. Use personal protective equipment: ear protection with ear muffs or ear plugs during welding.

Sentio system used in CROS configuration in patient with unilateral deafness – case report

Skarzynski P.H.[1,2], Cywka K.[3], Balwicka-Kowalczyk M.[1]

[1] *Teleaudiology and Screening Department, World Hearing Center, Institute of Physiology and Pathology of Hearing, Warsaw/Kajetany, Poland*
[2] *Institute of Sensory Organs, Kajetany, Poland*
[3] *World Hearing Center, Institute of Physiology and Pathology of Hearing, Warsaw/Kajetany, Poland*

The paper describes the first case of a patient with unilateral deafness treated with the Sentio device, an implant that uses bone conduction of sound. The operation was performed at the World Hearing Centre at the Institute of Physiology and Pathology of Hearing in Kajetany.

Single-sided deafness (SSD) is described as a profound hearing loss in one ear with normal hearing in the other. For many years it was thought that patients with unilateral hearing loss did not need any treatment, hearing aids or implants. It was also believed that speech in SSD patients developed normally and that there were no problems with understanding speech. However, recent reports have shown that patients with unilateral deafness face many difficulties in everyday life, including sound localisation, understanding speech over noise and lack of binaural hearing. Due to various factors, it is not always possible to use a cochlear implant in patients with profound hearing loss. These difficulties were addressed by Prof. Henryk Skarzynski and Prof. Piotr H. Skarzynski and their team who implanted the Sentio system in the CROS configuration in a patient suffering from SSD. The implant is implanted on the side of the deaf ear and uses vibrations to stimulate the opposite ear. Normal hearing in the better ear is required for use of the implant in the CROS configuration. With this solution, it is possible to improve speech understanding in difficult acoustic conditions and reduce the "head shadow" effect. The benefits associated with the Sentio implant are confirmed by the APHAB (Abbreviated Profile of Hearing Aid Benefit) subjective hearing assessment questionnaire. Among the most important are: improvement in hearing ability, support in learning and working, and overall improvement in quality of life. The Polish Sentio Matrix Test was used to assess understanding in noise. The Sentio system is one of the latest developments in hearing technology. It is an effective form of treatment for SSD patients for whom a cochlear implant is not an option and classic hearing aids are not profitable. The Sentio implant in CROS configuration facilitates daily functioning and reduces the problems caused by hearing loss.

The A2ml1-knockout mouse as an animal model for non-syndromic otitis media

Elling C.L., Gomez H.Z., Lee N.K., Hirsch S.D., Santos-Cortez R.L.P.

Department of Otolaryngology-Head and Neck Surgery, School of Medicine, University of Colorado Anschutz Medical Campus, Aurora, CO, USA

Introduction: Inflammation and infection of the middle ear, known as otitis media (OM), is a leading cause of hearing loss and the most frequently diagnosed disease in children worldwide. Traditionally, mouse models for OM rely on inducing acute infection through inoculation of the middle ear, e.g. with the human otopathogen non-typeable Haemophilus influenzae, and with very few genetic models with spontaneous or chronic OM. A2ML1 variants, including loss-of-function variants, were associated with susceptibility to OM in humans, but no animal model has been reported for A2ml1-related OM. Here, we report our middle ear findings in a mouse line with a CRISPR-induced knockout (KO) of A2ml1.

Objectives: The objectives of this project were to determine the presence of spontaneous OM and any phenotypes consistent with a Noonan-like syndrome via body morphology measurements, cranial x-rays, and histologic examination of the ME mucosa of KO mice.

Material and methods: Mice were X-rayed prior to harvest to determine if there are craniofacial or skeletal abnormalities. Tissues from mouse middle ears, as well as other upper respiratory mucosal tissues, were harvested. The harvested middle ear bullae were examined under microscope and submitted for histologic preparation to study phenotypic indications of OM. RNA samples isolated from middle ear tissue were assayed for expression of genes correlated with A2ML1 expression in humans.

Results: Data from a total of 119 mice (35 wildtype, 40 heterozygous, 44 homozygous) will be presented, with each analysis being performed on subsets of these mice. There were no significant craniofacial differences by genotype ($n = 22$). Findings in mice with the A2ml1-KO indicated an increased incidence of OM ($n = 29$; Fisher exact two-sided $p = 0.02$) with tympanic membrane perforations or thickening, as well as cases of middle ear effusion, inflammatory cells, or fluid from histologic sections. Dsp was upregulated in the middle ear tissues of homozygous mice (Wilcoxon test $p = 0.001$).

Conclusions: Thus far, our results in this A2ml1-KO mouse line indicate spontaneous occurrence of OM and dysregulation of Dsp in the middle ear as a potential disease mechanism for A2ml1-related OM.

The Stimulation of Polymodal Sensory Perception by Skarzynski 2.0 (SPPS-S 2.0)

Skarzynski P.H.[1,2], Czajka N.[1], Skarzynski H.[3]

[1] *Teleaudiology and Screening Department, World Hearing Center, Institute of Physiology and Pathology of Hearing, Warsaw/Kajetany, Poland*
[2] *Institute of Sensory Organs, Kajetany/Warsaw, Poland*
5 Oto-Rhino-Laryngology Surgery Clinic, World Hearing Center, Institute of Physiology and Pathology of Hearing, Warsaw/Kajetany, Poland

Introduction: Central auditory processing disorders are a set of symptoms that can significantly affect an individual's daily functioning. According to current guidelines, the primary and most important pillar on which rehabilitation interventions for patients with central auditory processing disorders should be based is the use of auditory training. One such method is the Skarzynski Method of Polymodal Sensory Perception Stimulation.

Objective: The aim of this study is to present new therapeutic possibilities through the introduction of the new version of the Skarzynski Method of Polymodal Sensory Perception Stimulation 2.0.

Conclusions: The continuous progress in technological capabilities and ongoing research leads to the development of various diagnostic and therapeutic tools. The introduction of the Skarzynski Method of Polymodal Sensory Perception Stimulation 2.0 significantly expands therapeutic options for patients with central auditory processing disorders.

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The use of bone-conduction implants in patients with rare genetic syndromes associated with ear malformations

Skarzynski P.H.[1,2], Cywka K.B.[3], Czaplicka E.[1], Skarzynski H.[4]

[1] *Teleaudiology and Screening Department, World Hearing Center, Institute of Physiology and Pathology of Hearing, Warsaw/Kajetany, Poland*
[2] *Institute of Sensory Organs, Kajetany/Warsaw, Poland*
[3] *World Hearing Center, Institute of Physiology and Pathology of Hearing, Warsaw/Kajetany, Poland*
[4] *Oto-Rhino-Laryngology Surgery Clinic, World Hearing Center, Institute of Physiology and Pathology of Hearing, Warsaw/Kajetany, Poland*

Introduction: Bone-conduction implants have emerged as a viable solution for patients with conductive or mixed hearing loss who are unable to use conventional hearing aids due to anatomical abnormalities. Individuals with rare genetic syndromes, such as Treacher Collins, Goldenhar, Klippel–Feil, Charge syndromes, and mandibulofacial dysostosis with microcephaly, often experience significant hearing impairments due to external and middle ear malformations. This study evaluates the effectiveness and safety of bone-conduction implants in this patient population.

Material and methods: A retrospective cohort study was conducted at a tertiary referral center, including nine patients diagnosed with the aforementioned syndromes. All patients exhibited conductive or mixed hearing loss and were not candidates for conventional hearing aids. Bone-conduction implants were surgically placed, and hearing improvement was assessed through pure-tone audiometry and speech audiometry. Additionally, subjective satisfaction was measured using the Abbreviated Profile of Hearing Aid Benefit (APHAB) questionnaire.

Results: The results demonstrated significant hearing improvements following implantation. Audiometric assessments showed enhanced hearing thresholds and speech recognition in both quiet and noisy environments. Patients reported high satisfaction levels, noting improved daily communication abilities. The surgical procedures were performed safely, with a low incidence of minor complications.

Conclusions: These findings highlight the importance of bone-conduction implants as an effective intervention for managing hearing loss in patients with rare genetic syndromes associated with ear malformations. Early implantation supports auditory rehabilitation, facilitates speech development, and enhances overall quality of life. Further research and clinical application of this technology may improve outcomes for this unique patient population.

The use of the new OSIA OSI300 in adults and children with conductive hearing loss and mixed hearing loss

Skarzynski P.H.[1,2], Cywka K.[3], Balwicka-Kowalczyk M.[1]

[1] *Teleaudiology and Screening Department, World Hearing Center, Institute of Physiology and Pathology of Hearing, Warsaw/Kajetany, Poland*
[2] *Institute of Sensory Organs, Kajetany, Poland*
[3] *World Hearing Center, Institute of Physiology and Pathology of Hearing, Warsaw/Kajetany, Poland*

The study will evaluate the effects of the Osia OSI300 implant in a group of 20 patients with conductive or mixed hearing loss. The Osia device is a percutaneous active bone conduction implant. In 2024, the first operation with the Osia OSI300 in a child was performed at the Institute of Hearing Physiology and Pathology, representing a breakthrough in the treatment and rehabilitation of hearing loss in a younger group of patients. Thanks to the piezoelectric stimulation used, the implant has exceptional high frequency sensitivity compared to other bone anchored implants. In addition to hearing benefits, the Osia OSI300's advanced technology allows for full diagnostic imaging (MRI) without the need to remove the implant. A number of benefits in the treatment of hearing loss have been observed in patients who have received the implant. The first is an improvement in hearing quality in both quiet and noisy environments, with a consequent reduction in speech difficulties. Audiological studies show a significant improvement in speech understanding in both quiet and noise. The degree of speech discrimination was assessed using the Polish Demenko and Pruszewicz Verbal Test, while the Polish Matrix Sentence Test was used to assess understanding in noise. Patients' subjective impressions based on the APHAB (Abbreviated Profile of Hearing Aid Benefit) questionnaire include better discrimination of sounds in noise, improved sound quality and clearer perception of high frequency sounds. Patients appreciate the aesthetics and the lack of restrictions in daily use. The Osia OSI300 system opens up new possibilities for hearing impaired people, thanks to its advanced technology. This is especially true for those who have not been successful with traditional hearing aids.

Valsalva maneuver during computed tomography for the diagnosis of tracheal diverticulum: a case report

Tsiouma G.[1], Stavridopoulos M.[1], Oikonomou A.[1], Megagiannis L.[2], Malamouli I.[2]

[1] *ENT Department, General Hospital of Volos "Achillopouleio", Volos, Thessaly, Greece*
[2] *Radiology Department, General Hospital of Volos "Achillopouleio", Volos, Thessaly, Greece*

Objectives: Tracheal diverticula constitute a subtype of paratracheal air cysts (PACs) that are characterized by a connection with the trachea through a thin neck. Patients with tracheal diverticulum rarely develop symptoms and are usually diagnosed on computed tomography (CT) performed for an unrelated indication. However, identifying the communication with the trachea on imaging may be challenging.

Material and methods: This report presents the case of a 55-year-old male patient who was referred to the emergency department with a possible fracture of the scapula caused by a fall from a height of three meters and was diagnosed, incidentally, with a paratracheal air cyst on thoracic CT.

Results: The paratracheal air cyst was recognized as a tracheal diverticulum by having the patient perform the Valsalva maneuver during CT. The cyst's volume increased in all three dimensions and a communication with the lateral tracheal wall was revealed, confirming the suspicion of tracheal diverticulum.

Conclusions: Diagnosing and distinguishing tracheal diverticula from other PACs is challenging, even with advanced imaging techniques such as multidetector CT and multiplanar or 3D reconstruction. Accurate identification of tracheal diverticula is crucial for effective patient management, as early diagnosis may inform monitoring strategies and surgical considerations if symptoms arise. This case report proposes that differential diagnosis can be accomplished safely and effectively by utilizing the Valsalva maneuver during CT scans.