COL2A1 pathogenic variant (NM_001844.4: c.1833+1G>A) causative for Stickler syndrome and one pathogenic change in COL4A5 (NM_000495.4: p.Gly624Asp/c.1871G>A) causative for Alport syndrome. In SLC26A4 (NM_000441.1: p.Leu597Ser/c.1790T>C) only one known pathogenic variant was found and the CEVA haplotype was not detected. **Conclusions:** Simultaneous testing of many genes using next-generation sequencing followed by family analysis identified a de novo pathogenic variant in the COL2A1 gene and after clinical reanalysis, the patient was diagnosed with Stickler syndrome, which is inherited in an autosomal dominant manner. The patient was found to be a carrier of a COL4A5 variant for the X-linked Alport syndrome, which explains the occurrence of microhematuria and may account for some degree of her hearing loss. The cause of LVAS still remains unknown as only one SLC26A4 pathogenic variant and no CEVA haplotype were found, and the disorder is inherited in an autosomal recessive manner. Our study identifies the presence of three rare known hearing loss syndromes in one patient and emphasizes the important role of genetic testing in guiding clinical diagnosis.

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### Sound localization and patients' satisfaction in SSD-CI users

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**Introduction:** Since 2011 single sided deafness (SSD) is an indication for Cochlear Implantation (CI). Previous conventional, non-invasive treatment options, such as CROS hearing, never rehabilitated the affected, deaf ear. Those methods only tried to compensate the disabilities in difficult environments transferring sound to the normal hearing ear (NH). A CI works differently, by electrically stimulating the deaf ear, thus providing bilateral hearing. Most important after CI is the rehabilitation. In bilateral hearing loss, the exercises can be done easily by presenting the audio files via loudspeaker. In SSD, this setting is not useful because the NH compensates the speech understanding in quiet, but practise for the CI ear is necessary. Therefor a direct stimulation of the CI witch auditory accessories like a FM-cable or a neck loop is mandatory to bypass the NH and just exercise the CI side alone. The aim of this study was therefore to test SSD-CI users in different setting for hearing in noise and sound localisation abilities together with an subjective evaluation of their performance via questionnaires. **Material and methods:** All measurements were performed in the unaided SSD condition (SSD) and in the CI aided SSD condition (SSD-CI). Benefit of speech understanding in noise was tested with the adaptive OLSA (S0N0). For the sound localization test, a seven loudspeakers cemicircle setup was used. To demonstrate the individual failure a Root Mean Score Error (RMSE) and the bias was calculated. To evaluate the patients' quality of hearing and satisfaction two questionnaires were used. The first, was the "Abbreviated Profile of Hearing Aid Benefit questionnaire" called APHAB. The questionnaire is structured in four subscales: Ease of Communication (EC), Reverberation (RV), Background Noise (BN), and Aversiveness (AV). The second questionnaire used was the "Speech, Spatial and Qualities of Hearing Scale", (SSQ12). Inclusion criteria was based on manufacturers recommendation and additionally patients had to have stable hearing loss >1year), aged 18 years or older and required CI experience of at least one year. **Results:** Ten subjects (eight females and two males) with a mean age at implantation of 45.6 years were included. The mean PTA4-NH (normal hearing ear) was 11.25 dB and the mean PTA4-SSD (affected ear was) 93,875 dB. The unaided mean SNR ws at −3.35 dB and in SSD-CI aided condition −5.29 dB SNR. The difference of speech understanding in noise with the CI was 1.95 dB SNR which is an enhancement of 32.98% ($p$<0.05). The mean RMSE showed a significant improvement in sound localization ($p$<0.05). The results in the unaided condition showed that most of the signals were located on the normal hearing side. Subjects also reported that their individual quality of life has improved with the CI, which was also reflected in the questionnaire outcomes. The results of the APHAB questionnaire show that the biggest difference and improvement, is in the category BN. For all ten patients the individual, subjective assessment in situations of understanding when there is a background noise, is 28% better in the condition of SSD-CI. The overall APHAB score resulted in a significant improvement in the two different conditions ($p$<0.05; 41% vs 29%respectively). The overall mean SSQ12 score was 57% in unaided SSD and in SSD-CI 40% which means an improvement of 17% ($p$<0.05). **Conclusions:** The results for the ten SSD-CI patients showed significant improvements after Cochlear implantation in all tested categories: in speech understanding in noise with the OLSA in the condition S0N0, in sound localization with seven speakers as well as in the two different questionnairs, the APHAB and the SSQ12, ($p$<0.05).

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### Speech-in-noise perception through audio-tactile sensory substitution

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Understanding speech in background noise is challenging. Wearing face-masks, imposed by the COVID-19-pandemic, makes it even harder, by limiting access to visual speech cues. We developed an audio-tactile setup, including a sensory substitution device (SSD) that can deliver speech simultaneously through audition & as vibrations on the fingertips. We examined whether speech understanding in noise can improve through audio-tactile training. We also evaluated changes in resting-state networks following audio-tactile speech integration & learning. Twenty(20) participants with normal-hearing participated in the study. They performed three tests of repeating vocoded sentences in background noise: a)only through audition(A), b)with matching tactile vibrations (representing low-frequencies of speech, ATm) and c)with non-matching tactile vibrations(ATnm) on the two fingertips of the dominant hand, before and after short training of repeating sentences with visual feedback (<45min).The outcome measure in each test was mean Speech Reception Threshold (SRT).The data was analysed in SPSS 20 using paired $t$-tests. Seventeen (17) subjects participated in a resting-state fMRI exam (10 min), both before and after training. The data was analysed in CONN toolbox 20. We found that participants improved in A from 21.46±10.68 dB to 6.71±7.96 dB [$t(19)=8.48$, $p\leq 0.001$], in ATm from 14.66±8.68 dB to 1.89±6.28 dB [$t(19)=9.45$, $p<0.001$], and in ATnm from 14.60±8.96 dB to 10.44±8.62 dB [$t(19)=2.66$, $p$=0 016] Before training the SRT values in A and in ATm [$t(19)=3.52$, $p=0.002$] and between A and ATnm [t(19)=4.54, $p<0.001$] were significantly different, whereas scores in the two audio-tactile speech tests were not different ($p$=0 97).After training all three test scores were found statistically significantly different [A vs ATm, $t(19)=3.97$, $p=0.001$; A vs ATnm, $t(19)=3.3$, $p=0.004$; ATm vs ATnm, $t(19)=6.7$, $p<0.001$]. The rsfMRI analysis revealed increased functional connectivity between the ICA component encompassing multisensory temporo-parieto-occipital regions with the left insular cortex [peak in MNI (x,y,z)=–39–5 0], & the ICA component encompassing bilateral auditory cortices with the right occipital pole/lateral occipital cortex, for the PRE>POST comparison (FDR cluster corr at 0 05). We show that speech understanding in noise can improve through short multisensory training and generalize to both a unisensory and a multisensory test situation. The degree of improvement >10 dB indicates more than a doubling of the perceived loudness. The best score (lower SRT) after training for ATm indicates multisensory enhancement due to the inverse effectiveness rule. It seems that the training, by removing some difficulty from the auditory signal, facilitated use of the concurrent tactile inputs (participants learned to ignore the non-matching tactile stimulation).These effects might be related to the revealed changes in resting-state networks. After speech understanding has improved, we found lesser involvement of the insular cortex indicated in associative learning; as well as decreased communication between the auditory cortices and the visual system. Early visual cortex has been found involved in multisensory audio-visual speech contexts, whereas LOC has been indicated in the integration of multisensory inputs, including those delivered via touch. In general, our results show that long after the classical "critical periods" of development are over, a new pairing between a certain computation (here, speech processing) and an atypical sensory modality (here, touch) can be established & successfully trained.

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### Superior Canal Dehiscence (SCD) – case report

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**Objectives:** Vertigo, dizziness and imbalance are commonly correlated with hearing problems. We present case of patient with mixed hearing impairment and vestibular symptoms. **Material:** 36 years old woman was suffering from fluctuating hearing on her left side, dizziness after position changing and imbalance during jogging. She had plugging and tinnitus in her left ear as well as imbalance problems and unspecific vertigo in everyday activity. She was not suffering on any other medical conditions. She was not taking any medications. Betahistine was not helpful in her vestibular symptoms. **Methods:** We present audiological and otoneurological findings. Audiometry revealed mixed hearing impairment mostly on lower frequencies. Intensity of hearing impairment was fluctuating. In tympanometry there was type A in both ears but reflex from stapes muscle was also variable. In videonystagmography where was no vestibular loss of function. We present also VEMP findings, which suggested fistula in left inner ear.„CT scans confirmed SCD on her left side. **Results:** For patients with dizziness correlated with hearing problems differential diagnosis should include: otosclerosis, Meniere's disease, inner ear malformations, SCD. **Conclusions:** In case of dizziness and fluctuating mixed type hearing loss CT of temporal bone and VEMP should be obligated to confirm SCD.

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### Surgical treatment of patient with one-sided bamboo nodule of the vocal fold – case study

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**Objectives:** Bamboo nodule is one of benign lesions of the vocal folds. In videolaryngostroboscopy it is seen as transverse cream-yellow band located typically in the middle of vocal folds resembling bamboo joint. It is described as laryngeal manifestation of autoimmune diseases. **Material:** This study describes the case od 53-year old woman who presented with 6-month history of hoarseness and ten-year history of rheumatoid arthritis (RA), currently without symptoms. **Methods:** The patient underwent otolaryngological and phoniatric examination including videolaryngostroboscopy (VLS), perceptual evaluation (GRBAS scale), acoustic analysis of voice (MDVP software) and subjective voice evaluation with questionnaire VHI-30 (Voice Handicap Index). Microsurgery was performed using a CO2 laser. **Results:** VLS revealed subepithelial white mass in ½ of the superior surface of the right vocal fold referred as "bamboo nodule". MDVP revealed significant voice disturbances in amplitude parameters. The GRBAS and VHI-30 showed mild voice disturbance. The patient was referred for direct microlaryngoscopy and microsurgery. Inspection of the vocal folds during microlaryngoscopy showed white transversal stripe originated from the vocal ligament of the right vocal fold. The mass was removed. Evaluation 3 and 12 months after surgery revealed improved quality of voice and no recurrence of the bamboo nodule. **Conclusions:** Laser microsurgery is effective method of treatment of bamboo nodule. After appearance of bamboo nodule on the vocal fold, patient should undergo specialist diagnostics of autoimmune diseases, because changes in the larynx could be the first and sometimes only symptoms.

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### Suspicion of delayed speech development and auditory central conductivity of the brainstem in children with autism

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**Introduction:** A child's speech delay can be caused by various reasons. The causes causing a delay in speech development can be in various locations ranging from speech engines