Single-Sided Deafness: Diagnosis and Treatment

Presenter: Susan Arndt, MD; Iva Katharina Speck, MD

- It is my pleasure to introduce our two guest speakers today. They're gonna be presenting to us on the topic of single-sided deafness in regards to diagnosis and treatment. Today we have Dr. Susan Arndt and also Dr. Iva Speck. If you'd like to learn more about either professor, you can read about their course by, the bio on the course registration page, but at this time I'd like to hand the mic over to you. Dr. Arndt.

- Yeah, hello everybody. Dear Caroline, thank you for your kind introduction. And it's an honor for me to giving a course on... After having the honor of giving a course thing inside of deafness already in 2016, I'm delighted to have been asked about it again. And I think, I hope it's a good sign. That means it was educational for the audience in 2016. As you all know, the treatment of unilateral deafness started at the end of notice, which means between 2008 and 2010, which means that we can look now back on 15 years of experience with the treatment of patients with single-sided deafness. This allows us to learn from our mistakes and implement some approaches differently than at the beginning. I would like to say in advance, in my opinion, patient selection and particular counseling and informing patients about expectations of success, the advantage and disadvantages of any treatment and treatment options are the most important factors for satisfied patients in the end.

Furthermore, the necessity of daily practice on, and continuous usage should be explained even before the indication is given. But I don't want to anticipate any content of today's webinar. As you know, there are two of us today. I would like to introduce you to Iva Speck. With her, I have been working and doing research since 2017. She's a very dedicated and ambitious colleague, so I'm now looking forward to her presentation and of presentation of our joint work. We're expecting an exciting discussion in the end and I'm happy to answer your questions after the lecture. Iva, this is your stage now.

- Thank you very much for that kind introduction. It's a great honor for me to present today. I wanna talk about single-sided deafness, diagnosis and treatment, and present our work at the University Hospital in Freiburg. These are our disclosures. The learning outcomes of today are etiologies causing single-sided deafness and the correct way to diagnose these etiologies. We wanna talk interpretation of measurements to determine which subjects with single-sided deafness are candidates for cochlear implant. And also explain how to select the treatment options, from bone conducting hearing systems, CROS, and cochlear implant. And which treatment is best for which subject with single-sided deafness. We'll first talk about the impact of single-sided deafness on daily life.

Of those are that are affected with single-sided deafness. We wanna talk about epidemiology and etiologies of single-sided deafness. We wanna go over the diagnostics that we use at the University Hospital in Freiburg, especially the preoperative worker. We wanna talk about the various treatment options that are available to us, the state of the art, our long-term outcome. As Professor Arndt already told, we have 15 years of experience now and have some exciting data for the long-term outcome. We'll wanna give a short cleanse into the current research, especially on neuroplasticity. We wanna sum up and then discuss with you about these very exciting topics. Why is single-sided deafness a problem? For the longest time, it didn't seem like a problem, but we know that adults and children have problems in daily life because of single-sided deafness, especially speech recognition in background noise is reduced for these subjects and localization of sound sources is nearly impossible.

This is a big problem in traffic and also safety hazard there. Tinnitus can be a problem and can also cause low quality of life. All patient with single-sided deafness have increased listening effort. All of that can lead to low self-efficiency, low self-confident and social withdrawal due to disease of single-sided deafness. Especially for children,

we know that children with single-sided deafness require more speech therapy than their normal hearing peers. We also see a higher number of grade failures in children with single-sided deafness. This academic disadvantage keeps on into adult life. We see lower schooling and academic achievements in patients with single-sided deafness compared to normal hearing peers. So, yeah, single-sided deafness is a problem for everyday life for adults and children with single-sided deafness.

What is causing single-sided deafness? Here we have to differentiate between children and adults. In children, Rauch et al in 2021, could show that the leading cause of single-sided deafness in children is hypoplasia or aplasia of the acoustic nerve. Therefore, MRI is a crucial part of diagnosing these patients. The second leading cause is a infection with the cytomegalovirus of the mother during the pregnancy. From our clinical routine, we know that this is most likely in second pregnancies because the mothers infect themselves through the fluids of their first child, for example, spit or pee. So, we always have to counsel future mothers on the importance on keeping it distant to the first child's fluid during their pregnancies if they didn't had the CMV infection before.

In adults we see a different etiology cluster. Usmani et al in 2017 showed that the leading cause of single-sided deafness in adults is sudden hearing loss. It's followed by chronic otitis media, so the chronic middle ear infection. And vestibular schwannomas of the acoustic nerve. This is also important to perform memorize in adults to rule out vestibular schwannomas, so any malformation of the acoustic nerve. And we also see another leading disease in adults, the intracochlear schwannoma. Why is it increasing in number? There are two reasons for that. First, before we had the cochlear implant, most adults never underwent MRI because we had no treatment options for these patients, so an MRI was most necessary. Now that we have that options, almost every adult with a single-sided deafness will go into MRI and have that diagnosis done.

And the second reason why it seems to increase is that our MRI technology is improving. We have higher quality, so these really tiny tumors that are up to one centimeter can be diagnosed by now. So, that's why this disease seems to rise, but it's just that we can detect it now. What diagnostics do we need? I already told a little bit about imaging, but I will now give an overview about the preoperative workup that we applied our university hospital. This takes two days for the patients and includes medical personal history, ENT examination, audiological and vestibularis examination, imaging, rehabilitation capability, and important, the consultation of the patients. Medical and personal history is based on the duration of deafness, the onset of deafness, and possible causes of the single-sided deafness.

In ENT examinations, we focus on microscopic otoscopy, but we also diagnose the whole patient to see if we see any other diseases that might interfere with our treatment. I will now go a little bit more into detail on audiological examination. Here we perform subjective hearing tests and objective hearing tests. The objective hearing tests will be performed with and without hearing aids. For our insurance, it's really important to also prove that with a hearing aid, the deaf ear or the almost deaf ear cannot be rehabilitated properly. We'll perform therefore pure-tone audiometry. Speech recognition, we use numbers and monolithic words. And very specific for single-sided deaf patients, we will use sentence in background noise, and localization of sound sources.

Because as you remember from the beginning, these are the key problems from the audiological side that our patients with single-sided deafness face. For sentence recognition in background noise, we use the setup shown above. We either present both speech and noise from this, excuse me, from the sound speaker in front of the patient. Then we present the noise to the poorer ear and the speech to the better-hearing ear and vice versa. So, noise to the better ear and speech to the poorer

ear. For the localization of sound sources, we use the setup shown below. We have a semi-circle of seven loudspeakers in front of the patient and they will identify the loudspeaker where the stimulation is coming from, and so, we calculate the angle arrow that our patients display.

This is also important because it's something to monitor our treatment after, so we can see if the problems the patients face with single-sided deafness can be improved by the treatment we provide for them. Objective hearing tests include otoacoustic emissions, brainstem evoked response audiometry, or electrocochleography, or promontory test. If we use BERA, electrocochleography, or promotory test is based on the hearing capacity of the poorer hearing ear. If we still have some number or watch recognition, we will use brainstem evoked response audiometry. If we still have some pure-tone recognition on the poorer hearing ear, we can use the electrocochleography. If we have neither word nor tone recognition, we will apply the promontory test. Here we'll place a needle through the eardrum directly on the promontory and apply a small current and then we ask the patient if they hear or feel this current in their ear to test if the auditory nerve is still intact.

This is most important in patient that have their single-sided deafness due to a vestibular schwannoma. So, most of these patient has an operation on the auditory nerve and we have to see if the auditory nerve is still functional to see if maybe cochlear implant is an option for them. So, we ask them if they feel or hear the current. We ask if they can detect rhythm and if they have any hearing fatigue. So, we'll give them a tone for one minute and ask them if it gets less loud during that time. We also use the literature to determine, which diagnostics we use here. Here Van de Heyning et al in 2016, proposed a unified framework for single-sided deafness.

They recommended to measure pure-tone audiometry, speech perception and noise, sound localization, determine quality of life with a questionnaire in the mother tongue of

the patient, to measure tinnitus if applicable, and also, after treatment, the daily device use. Here, they propose to measure all these at baseline three weeks after treatment, one month, three months, six months, and up to 12 months. We usually measure all the time that our patients are in rehabilitation. In German that is around two to three years. And for the long-term follow up we invite the patients to participate in the study. Imaging is also very important in preoperative workout. I already talked a little bit about that during the etiologies of single-sided deafness.

Here the MRI is the key imaging technique that we use. Here we see a case of a aplasia of the auditory nerve. You see the inner ear canal on both sides. Here on the right side it is formed correctly. I'll explain what you see there. There you can see the vestibule nerve with the upper and lower part. Up here is the facial nerve going through the inner ear and the structure the arrow is pointing to is the auditory nerve. So, this is the normal picture we see from the inner ear canal. Here we see that there's something missing. We have the vestibular nerve with the upper and lower part. We have the facial nerve, but we have no auditory nerve.

So, that would be a typical picture for a child with single-sided deafness with an aplasia of the auditory nerve on the left hand side. The MRI is also used to detect intralabyrinthine schwannoma as I talked about before. Here you see the T1 with a contrast agent and you see this very small contrast enhancement in the vestibular. And therefore, it's really important to always perform the MRI with a contrast agent to be sure to detect these very small tumors. We can only see due to the contrast enhancement. Here we have the intraoperative view into the vestibulum. And we see this very small structure in the vestibulum, that is the intra leverage melanoma. Here you see that is under a half a centimeter diameter and this small tumor causes a single-sided deafness.

So, that's something that we can see in the new MRIs and then cure and put in a cochlear implant, for example for these patients. We use CT or diffusion volume tomography in children if we have any abnormalities in the MRI. So, as we see any abnormalities in the MRI, we'll perform a CT to rule out any malformations of the vestibulum or the cochlear. In adults we prefer the diffusion melanoma tomography, but also use CT if necessary based on the individual medical history. Coming to the next two very important parts of our preoperative workup that are less medical, but still really important for our patients. We have to evaluate if they can be rehabilitated with a cochlear implant.

As I mentioned before, it will take two to three years for them to come back to our clinic to do the rehabilitation stays. So, they have to be really motivated to do that and to relearn hearing with the cochlear implant. Especially important in patient with single-sided deafness because the cochlear implant has performed really well to be a match to the normal-hearing ear. And for the rehabilitation capability, we will look at that as doctors in patients above 65 years. We will always do a mock test to look into their cognitive abilities, maybe a beginning dementia. If we have any questions there, we're gonna involve neurologists or geriatric doctors into our team. The second pillar are the speech therapists.

They talk to the patients, explain which kind of therapy they need afterwards and also give their statement if they can be rehabilitated. And we also have a psychologist in our facility that is involved if necessary. We will also have a counseling of our technicians that will explain the various models that we have for the various treatment options and tell the patient the technical stuff they have to work with with the implants. For example, older patients sometimes have problems with small buttons to press, so they will also show that before, so that they know what they're getting into. And in the end, we ask the patient what they want and also give our medical opinion and the decision which treatment is correct for which patient is always individualized.

So, for every patient, the solution will be different. In the next slide, I will show you which treatment options we have and what are the advantages of each treatment option. So, we can do observance and rely on compensation mechanisms. That was the only treatment option for a long time to just observe our patients and rely on the compensation mechanisms. Then we have the possibility of the CROS hearing aid, so contralateral routing of signals. Here, the patient wears hearing aids in both ears and the information from the deaf ear will be transmitted wirelessly to the other side. So, all information will be transmitted to the better-hearing ear. Bone conductive devices work similarly. Also we have the conduction, so everything will be heard from the normal hearing to the better-hearing ear.

So we here have the possibility to overcome the head shadow effect with a CROS hearing aid or bone conductive device. The head shadow is a problem that if this is your deaf ear and someone's talking from that side to your ear, your head is in between the information and the ear that can perceive that information. So, if we put, for example, the CROS hearing aid here, the wireless connection will overcome that head shadow effect and give that information to the normal-hearing ear. So, everything will be transmitted to the normal-hearing ear. Then we have the possibility to use wireless microphone technology to the normal-hearing ear. Here we have the transmission of speech directly into the normal-hearing ear.

This is most important when we have a noisy background or a speaker that is far away. Usually it works like that, that the speaker will wear, for example, a Phonak Pen and talk into that, and then this information will be directly transmitted to your normal-hearing ear. Giving you the possibility, for example, a classroom setting, to understand the teacher. And the fifth option is a cochlear implant. The only option that allows us to rehabilitate binaural hearing because here we have the possibility to make the deaf ear have a hearing impression and speech recognition. So, you have two

independent speech recognitions from the normal-hearing ear and the cochlear implant. And when we have all these treatment options, we have to then find the optimal therapy for each patient.

We will always give them the possibility to test CROS hearing aids and bone conductive devices and wireless microphone technology before they ask for cochlear implant. So, during the preoperative workout they will be applied with the CROS, the bone conductive device on a headband, and the wireless microphone technology and test that out to see if these less invasive treatment options might be the correct option for that patient. And then, we talk to them and counsel them on which treatment option they would prefer, so that we can counter them to the individual needs and avoid non-users. Something that we don't want in any devices to implant something and then have a non-user in the end. Here you can see a meta-analysis of Kitterick loss system has the main advantage that reduces the head shadow, but we have interference with the normal-hearing ear.

So, a big problem is that all the information is put to the normal-hearing ear, putting stress on that normal-hearing ear. We have no summation effect. Summation effect is if you have the noise and the speeches coming directly from the front to you that the speech is perceived a little bit louder than the noise and you can only do that if you have two, so binaural hearing. We have no reduction of tinnitus and we have no improvement of localization in CROS systems. In cochlear implants, we can overcome the head shadow effect as well. We have no interference with the normal-hearing ear. We have reduction of tinnitus in 70% of the patients while they're wearing the cochlear implant.

And we have an improvement of localization. The literature on the summation effect is unclear. We have studies that show that the cochlear implant can provide the summation effect if noise and speech is coming from the front. But we have also

studies that do not show a summation effect in the cochlear implant. Next, I wanna look into the long-term outcomes. So, if we have our patient implanted for a long time, how do they perform with the cochlear implants? Here we'll look into a study from Arndt et al 2023, and a study from Speck et al 2021 on children and on adults. And here we look into speech recognition and background noise. One of the key problems patients with single-sided deafness face.

And we see a clear improvement in speech recognition in one situation. And that situation is when the speech is coming to the implanted, so the cochlear implant ear, and noise is coming to the normal-hearing ear. So, in this situation, children with postlingual deafness and adults improve significantly with the cochlear implant. In both studies we could not see the summation effect. This is this part here, so when speech and noise is coming from the front. And interestingly, when the noise is coming to the CI and the speech is coming to the normal-hearing ear, that CI noise recognition will not interfere with speech recognition for adults and for children. So, we have improvement, but in our long-term study we did not see a summation effect.

When we look into localization, we see that for postlingual children localization improves significantly after cochlear implantation, of course, 6 to 11 years. Here, the lower the angle arrow, the better the localization is. Here on the congenital and perilingual children we could only include four children because most of them were studied so many times that they could not come back to do that intervention, but we could receive some questionnaires from these children. For the adults, we saw that 70 showed 6 to 11 years after implantation a significant improvement of the angle arrow. And therefore, an improvement of the localization of sound sources with their cochlear implant. When we look into tinnitus, we see that, as we know, children suffer less from tinnitus than adults, only seven of our 19 children had tinnitus, so 12 showed no tinnitus.

And out of these seven, only one reported increase with the cochlear implant. So, six of these children either had a decrease or a neutral experience with the tinnitus through their CI. In our adults, we saw that 22 patients reported in tinnitus before implantation. The rank that on an numerical rating scale from 0 to a 10. And rank that once preoperatively. And then, 6 to 11 years with CI experience. And we saw that the intenseness of the tinnitus decreased significantly with the usage of the CI. Very important for us is also the CI usage. So, how long and if patients still use their CI after long term. And here we have very low numbers for non-users. For the children it was 11% non-users and for the adults with 15% non-users.

That is one part a good counseling for sure, but I also have to say that we have some bias here. Always in long-term follow-ups, you tend to have more motivated patient enroll and also patients that are more closely connected to the hospital. So, I think we might miss some non-users. Nevertheless, the better counseling will provide less non-users. In our... On the pediatric population, we saw that most non-users had a congenital onset of single-sided deafness and were implanted at the age of three. So, they had three years of no hearing experience from birth and that's why they never reached speech recognition and then stopped using their CI. In the adults, the reasons were more varied.

We had four non-users in our adult population. One told us he had a lot of dirt and splashes that worked, that's why he stopped using his cochlear implant. Professional and private stress prohibited one patient from doing the rehabilitation for the cochlear implant. One was to set aside with the hearing impressions or the quality. This patient reached good speech recognition, but the quality was off-putting. And then, we had one patient that did not reach speech understanding. There we had a infection of the cochlear and it took us a long time, one and a half years to fight with the insurance to get him implanted. And then, unfortunately, the outcome was not satisfying.

Fortunately, now the insurance will not fight that long anymore for the implantation of a cochlear implant in single-sided deafness.

So, here, again, one of our key messages, patient selections prevent non-users in adults cochlear implant in single-sided deafness. And as symmetry hearing loss patients is, from our point of view, the superior treatment because we have the restoration of the binaural hearing. A contraindication is always the absence of the cochlear nerve, so a pupil or aplasia of the cochlear nerve. It also depends on the duration of deafness in the adults, in children with acquired hearing loss. On that point, I will talk a little bit later our ongoing research. How we might quantify this rather difficult predictor. Duration of deafness is always difficult to get precise from our patients and we wanna look into, maybe the neuroplasticity telling us if an adult is a good or poor fit with a cochlear implant.

In children, we still don't know the optimal time we look for the fatal outcome for children with congenital single-sided deafness. We always compare to bilateral hearing loss, but we are not unsure if it's vital or narrower than for bilateral hearing loss. We, in our study saw that congenital children with single-sided deafness benefit from a cochlear implantation before the age of three. So, that's our cutoff that we use at the moment. For acquired unilateral deafness in children, we also don't have an age limit, we just look on the duration of deafness. In all cases, if we don't have the indication for CI or it's not wished by the patient, we will try to provide treatment with CROS, or also integrated bone conduction system, or other device systems like the Phonak to give them all the options.

But if they refuse or they don't feel a benefit, we can always observe these patients and monitor them closely. And now we have a small look into our current research because we wanna see if we can identify better markers to provide us information about the outcome with the cochlear implant, especially for subject with single-sided

deafness. Here we wanted to see if we can look above the function of the auditory nerve that we can monitor with the promotory test to see if we can look into the brain. So, the primary auditory cortex and the brainstem. And therefore, our first question was, is neural activity altered by single-sided deafness in a meaningful way? And here we worked together with the clinic for nuclear medicine and performed FDG PET, so positron emission tomography, using a radioactively labeled sugar.

So, there we can see where the sugar is used in the brain and therefore see where we have neural activity. Here we measure patients with single-sided deafness that not yet had any treatment. And here I will show you one exemplary picture that we get and explain you what you can see. The more yellow, the more activity we see. The blue circle parts are the auditory cortex, so the first region of the brain where tone recognition is performed. And here we have a single-sided deaf patient and we know that the auditory system crosses. So, on that side was the normal-hearing ear displayed on that primary auditory cortex. And here we had the deaf ear that displayed on that part.

And on the arrow part you can already see we have less yellow than on the other. So, less activity contralateral to the deaf ear compared to contralateral to the normal-hearing ear, which makes sense to us because we have a lot of information coming through the normal-hearing ear to the brain, but there would be no or very little information from the deaf ear. And we can also see that quantifiably, that the glucose metabolism, so the uptake of radioactive sugar is less contralateral to the more hearing impaired ear compared to contralateral to the normal-hearing ear. But what we could also see is that this asymmetry varies widely. In some patient it's reversed, it's almost nonexistent, then sometimes it's very pronounced.

So, that is something that is, has a great inter-individual range. So, maybe something that we could use for our prediction of outcome. If something varies between the

patients, maybe it's a factor that can tell us how good they'll perform with the cochlear implant. So, we can answer our first question. Yes, neural activity is altered by single-sided deafness. And of course the second question, is neural activity a predictor for success with cochlear implant? Here we'll show you a small first study of nine patients and here you can see the asymmetry of the auditory cortex. So, we would like to see a great asymmetry because we believe there's information coming from the normal-hearing ear, so a lot of activation of the auditory cortex.

And we have a deaf ear where there's no information coming up, so no activation on the other side. And then, when we have a small auditor asymmetry between the auditory cortex, we believe that this part of the brain that would usually perform auditory tasks from the deaf ear is overtaken by other sense. So, the visual or the sensor monitoring system is using that brain capacity to interpret visual and some other sensory information. And we can see that here. So, the more asymmetry we have, so the less overtake we have, the better our patients perform six months in with the cochlea implant. So, there, we have sentence recognition. Recognition can be better in higher symmetry than in lower symmetry.

Number recognition as well. And then work recognition, we see a trend level here. So, we see that the neuroplasticity, so our brain makeup, can tell us something about how good we can perform with our cochlear implant. So we can answer a small yes because we have just a small amount of patients, but we are working on a big follow up study with more patients and longer follow up. And hopefully, we can involve that in our clinical practice at one day to give our patients a better understanding how their brain is made up and if which treatment options would be best for them. Coming back to, from the research to day-to-day life. To sum up, cochlear implant has the possibility to eliminate the head shadow, improve localization and reduced tinnitus for patient with single-sided deafness.

Patient selection and education are essential to provide non-users. Secondhand choices after cochlear implantation are bone-anchored hearing aids and cross hearing aids. And we need to rely on the biomarkers and we are working on that. Thank you very much. And I also wanna thank everybody on our research and clinical team. When two people present something, there's always a lot of people behind us that did a lot of work and I wanna thank them at this point. I hope we have a good discussion and I'm happy to answer all your questions if possible.

- [Christy Huynh] Thank you Dr. Speck and Dr. Arndt. We're gonna go ahead and open up the floor. For any questions that you might have, feel free to type them in the queue in that chat box in the lower right hand corner.

- If we have no questions, maybe we can talk a little bit. Ah, now we have a question, perfect. What is the importance of doing a vestibular examination on individual with single-sided deafness? I think most importantly is to get an idea how the vestibular system works before our implantation, and to follow up after the implantation on the vestibular system. We have a lot of patients, for example is with MIA's disease that also receive other treatment here when we do the cochlear implantation surgery. So, decompression, for example, and something that we have to monitor. Also, cochlear implantations can have the effect of inducing vertigo, so we have to see if we changed something of that system before and after implantations.

Do you want to elaborate on that, Susan?

- No, it's okay. I think we have to rule out that the vestibular system is not infected on the other side. So, that's important for the cochlear implantation, especially in single-sided deafness. And we should do it and it's important to counsel the patients which treatment is best for them. The next question, how long do you recommend the patient try each treatment prior to cochlear implant? It's difficult, when we started with

a study in single-sided deafness, we have recommended to try each treatment options across device and the bone conduction device, each for three weeks. I think it's sometimes not possible to date that each patient should, or that has to try, or that has to use devices for six weeks and AWOL.

So, therefore, we recommend to treat, to try each treatment at least one week. And most of the patients are aware of an improvement after one week if it is okay, if they have an improvement with this treatment option after one week.

- The next question is what are, what age are you allowed to plant in Germany for children? So, to my knowledge there's no restriction. We are restricting ourselves to one year because also we have the anesthesia and we have to be sure that the moderation of the auditory system is up to the point that we can really see how high the hearing loss is on that ear. So, we usually opt for one year, at the age of one year to implant the patients. Am I correct, Susan?

- Yeah, congenital patients or the children with congenital SSD. I know that in the US they are restricted to the age of five. And as you heard from Iva before, we recommend the implantation to date of children with congenital deafness up to the age of three years. So, to have good results and they still don't know the perfect point, but therefore we wouldn't recommend an implantation now of these children with congenital deafness over the age of five. Because we have a lot of non-users over the age of five, there are varying results especially in this group of children and we can't, yeah, predict the outcome, especially in children with congenital deafness.

- And on the question on which data you might base yourself, there's data from our hospital and other hospitals where an implantation is possible before the age of five. Maybe that would be a good argument for the FDA to allow that we saw that we have these many non-users as was onset above the age of five. To the next questions, has

there been any evidence of COVID and, or vaccines causing single-sided deafness? Here we have some studies, some published, some in the working, on the COVID infection and the impact on the auditory system. And yes, we saw single-sided and bilateral deafness in COVID infection primarily higher. So, patient that needed long hospital stays, intensive care, that showed hearing loss on one or both sides.

Some of them are already implant for the cochlear implant because there was no restoration of the hearing after the COVID infection was done. So, we see that, but it's uncommon, but possible. I think in almost all virus infections there are some amount of patients experiencing hearing loss due to that infection. I have no data on vaccines and I just recently read a meta-analysis and summing up the research we have and there was no concrete data on vaccines to causing hearing loss, at least the COVID vaccines. Do you have more informations on that?

- No, it's okay. I just read the next question, sorry. The question is, for a patient who has a sudden single-sided deafness due to cochlear stroke, where would you recommend we start? I would... I don't know how is the overall situation of the patient. If we consider a single cochlear implant in those patients, we have to be sure that cochlear nerve is still working. And if the patient has no residual hearing as Iva already explained before, we would recommend to start with a pulmonary test to be sure that cochlear nerve is still working after a stroke.

- We are aware that pulmonary test is, has a low sensitivity, but a high sensitivity that, so it's not a perfect test of course, but in these situations where we are unsure about the function of the auditory nerve, it's the only possible measurement we have, so we always incorporate it to see if we maybe have a hearing impression and then we see way better outcomes. But we don't have a hearing impression, the counseling of the patient then is more crucial to tell them that we might try, but they might not perceive

speech recognition with the cochlear implant and then they have to agree to that setting. Am I correct?

- Yeah, but what did you mind, I just looked at the question due to cochlear stroke, I think you mean that it's a sudden hearing loss. And in case of sudden hearing loss and this residual hearing, you can start with a normal cochlear implant preexamination.

- So maybe we can, yes, I think it's resulting way more non-users for the patriate population. I think that a comment is based on our information on the time span. I think Susan told me about a new publication on that theme that did not show the three years mark we found. Can you maybe tell us something about that in the remaining time?

- Yeah. It's from Park et al . It was very interesting. They haven't found an implantation age, difficult implantation age of three years. More important for the usage of the CI is a daily usage. The daily usage in waking times for the children. When the children are awake and the duration of daily usage, date daily usage time and the practice with the CCI.

- I think.

- But I still think that, especially in congenital deafness, the three years are quite important because a lot of studies have shown that patients over the age of three years, implantation over the age of three years are quite difficult and we have varying results, especially from all have shown it and a lot of researchers. So, I would be very careful to recommend an implantation over the age of three.

- So, maybe constant individuals is difficult in certain population. How do we avoid individuals from becoming non-users? Yeah, it's very difficult and it's a task that we of course face every day. That's also why we are doing our research in a neuroplasticity

because sometimes you don't know how to counsel them when they have a long duration of deafness, but they're very motivated. And then, you also in doubt if they will perceive speech recognition with the CI or not. But this is a small part of the population. Most patients, the medical indication is quite clear like a cochlear implant or across our bone anchored device is better for them from the medical standpoint and then we just take time to talk to them.

And we also give out addresses of other patient that have single-sided deafness that received a cochlear implant, a bone conductive device will cross, so that they can also connect via the patient. Sometimes these informations are more, more, way more malleable than what we professionals can tell the patients.

- It is important to counsel the patients that they will not get back the auditory hearing, a normal hearing on the side of the cochlear implant side. That is really important to reduce the expectations to have real expectations of the cochlear implantation. They will have an improvement in hearing a noise, but it will not be getting normal. They will have experience better localization ability, but it's not normal in the end. And they have to practice with the cochlear implant. It's not enough to have the speech processor at the year. They have to practice every day at least 20 minutes with the Bluetooth over the auditory, over the with cochlea and the speech processor.

- For children, they don't want to turn on the bone production hearing aid in noise in classroom. Do you recommend turning it off in noisy situations for children? They have to refer to Susan, but I see that you're nodding your head.

- We normally don't recommend bone conduction hearing aid or CROS hearing aid in children, in small children. If they have normal hearing on the other side, we additionally give the normal hearing ear more strengths and more, more efforts for hearing. So, therefore, we don't recommend, normally in up to the age of 10 years, bone

conduction hearing aid or CROS hearing aid to preserve the normal-hearing ear. And if the children and the parents... If the children have still problems after the age of 10 or 12 years, then we can try a bone conduction hearing aid or a CROS hearing aid. It's sometimes better to give them digital Phonak device, Phonak Pen that is sometimes quite good. And especially in classrooms.

So, as Iva already explained before, when the teacher speaks into a microphone and it's transferred directly to the normal-hearing ear of the child.

- Yeah, we actually did a study into single-sided deaf children between the years of 5 and 10 and we made a noisy classroom environment and gave the teacher the pen and we saw that the speech recognition improved drastically when they had the Phonak device and the better-hearing ear. And we also put it in the conference mode, so that, for example, when there's a working group, so the other children sit around the table. It also improved so that it's something that you can recommend in the situation if it's too noisy otherwise. Maybe that would be a better fit for these patients or children. If there are... Thank you. Maybe we can look into the slides for the exam afterwards where the information's provided, so that we have a little refresher for all of you.

I will just go back to the beginning and show you the slides that the questions are answered on to sum up the seminar. So, one, to remind you the impact of single-sided deafness. We have speech recognition in background noise that is reduced. We have problems with localization of sound sources. We have tinnitus and increased listening effort. Problems with low self-efficiencies, self-confidence and social withdrawal. And in children, additionally increased speech therapy needs and grade failures for academic disadvantages. Then, looking at the etiology in children, the most common is hypo or plasia of the auditory nerve. And then, we have congenital infections with cytomegalovirus, rubella, and we also have more uncommon abnormalities like enlarged vestibular aqueduct that might cause single-sided deafness.

Maybe that's a good point to answer the next question. In congenital single-sided deafness, how is the auditory development different? So, for the neuroplasticity part, we have no information coming in from the deaf ear. So, for example, in EEG studies we see that the normal neural response of the auditory system doesn't develop like it does in normal hearing peers. You can see that the neural feedback even after stimulating with a cochlear implant is not normal. And we see that the brain region, so the auditory cortex and associated auditory cortexies will be overtaken by other sensibilities. So, for example, visual information will be processed in the part of the brain that would normally be served the deaf ear.

Susan, any other remarks to that?

- I'm not sure if the question was what is the difference between normal hearing peers and SSD children? Perhaps there's also the question, what's the difference between congenital SSD and acquired? And an acquired SSD that means children, this, they already, they heard before, before they becoming deaf on one side. The central region was already stimulated and they already stimulated with auditory input. And that's a different between congenital deafness and acquired deafness. And as we know from bilateral deaf children, the earlier they are implanted, the better as it results in the end. And we wouldn't implant also a bilateral deaf child over the age of five. So, and especially in single-sided deafness the asymmetry between both ears are way greater compared to bilateral deafness.

So, that's the reason we wouldn't recommend an implantation over the age of three because of the great variety of the results. I hope this answered your question.

- And we maybe look into the other slides concerning the questions. Here we see again the imaging and the importance of the MRI and what we can diagnose. We can

diagnose abnormalities. For example, we can diagnose a cochlear nerve deficiency and vestibular schwannoma. And also other abnormalities, like an enlarged vestibular aqueduct. Other problems like infections, we cannot see in the MRI there, we have to rely on medical history for our patients. And I see we have another question. Do you know of any studies discussed not using a BAHA for SSD because of amplification of noise for children? I cannot answer to that.

- No, there are not existing studies about that. I know it's a good point for discussion. Also, in Germany, a lot of surgeons in Germany also recommend the BAHA or a CROS hearing aids in children with single-sided deafness from the beginning. But in our opinion, the speech development of the normal-hearing ear is so important that it's, that we not want to disturb the speech development of the children to this noise, which is coming from the BAHA or the CROS hearing aid from the deaf side. And with the BAHA, we prevent the children for the development of compensation mechanism. And this is also important in patient, in children with congenital deafness where we can not implant the cochlear, or I recommend the cochlear implant that they have to develop compensation mechanism and a lot of children perform quite good with that.

You know that also. And the next question.

- Here we have the third question. So, recapping the selection of the patients. Really important is to look into the cochlea indications. For example, the absent of the cochlea nerve. It's really important to look into the duration of deafness. And the optimal window for children is unclear. We would recommend before the age of three. In congenital deafness, acquired unilateral deafness, we have not, do not have an age limit besides cochlear implant. If that is not an option or is not wished, we would recommend CROS hearing aids or bone conductive systems or assistive devices. For example, the Phonak wireless system. And we always have to cons on the limits of each of these treatments. And then, I will show you the summary for one more time.

So, to sum up, cochlear implant is a great option because it can eliminate the head shadow, it can improve localization and reduce tinnitus. Selection education is key. The second choice for us would be bone anchored hearing aids, CROS hearing aids or wireless systems. And we need more research to have more reliable predictors to consult even better. We have a question, thank you.

- [Christy Huynh] Thank you so much Dr. Speck and Dr. Arndt. We really appreciate and value your expertise and coming to share. Thank you everyone who joined us today on Audiology Online. We hope you enjoy today's presentation. Have a great day everyone.