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Single-Sided Deafness
– Diagnosis and Treatment

Iva Speck, MD & Susan Arndt, MD

Department of Otorhinolaryngology, Medical Center – University of Freiburg, Germany

Susan Arndt, MD

Since 2003 Susan Arndt has experience as PI and Co-investigator in numerous projects. Special fields of interests and research are cochlear implantation, especially single-sided deafness, maturation of auditory pathway (EEG, fMRI), bimodal fitting, development of new electrode designs, vertigo and QoL, preservation of residual hearing and hearing effort. Furthermore she has many years of experience in development, research and improvement of implantable hearing aids, especially middle ear and bone conduction systems (optimizing surgical techniques, indication, performance).

Especially, her research in treatment of patients with single-sided deafness has influenced the knowledge with regard to auditory rehabilitation worldwide. As one of the pioneers in this field, Susan and her research group facilitated that cochlear implants are an accepted treatment modality in single-sided deafness nowadays. Susan Arndt's scientific work was awarded with the Award of the Research Association of German Hearing Aid Acousticians and the Award of Eleonore-und-Fritz-Hodeige Foundation in 2012. Her scientific and educational activity is documented by over 80 journal publications and several book chapters, and over 100 official invitations to conferences and scientific seminars. Her h-index is 22 (February 2020).

Iva Katharina Speck, MD

Since 2017 Iva Speck works in the department of otorhinolaryngology of the university hospital Freiburg as a clinician scientist and resident. Her research focused on single-sided deafness and its effect on the central nervous system using fMRI, PET, EEG and fNIRS. Using the PET to evaluate the auditory pathway, she was able to visualize the inferior colliculus, an important part of the auditory pathway, for the first time. Besides the improvement of scientific knowledge of the central auditory pathway and its plasticity this project might be of clinical value regarding the prediction of the therapeutic outcome after CI.

As a PI Iva Speck received founding from the German Research Found and from the Else-Kröner-Fresenius-Stiftung to neural plasticity and its effect on treatment of single-sided deafness. Iva Speck received the ADNAO innovation award 2022 and the Alavi-Mandell Publication Award in 2021.

Disclosures

- **Presenter Disclosure**: Financial: Susan Arndt is employed by the University Medical Center Freiburg in Germany. She received an honorarium for this course. Non-financial: Susan Arndt has no relevant non-financial relationships to disclose. Financial: Iva Katharina Speck is employed by the department of otorhinolaryngology of the university hospital Freiburg. She received an honorarium for this course. Non-financial: Iva Katharina Speck has no relevant non-financial relationships to disclose.
- **Content Disclosure**: This learning event does not focus exclusively on any specific product or service.
- **Sponsor Disclosure**: There is no external sponsor for this course.

Learning Outcomes

After this course, participants will be able to:

- List etiologies causing single-sided deafness and the correct way to diagnose these etiologies.
- Name and discuss how to interpret measurements used to determine which subjects with single-sided deafness are candidates for a cochlear implant.
- Explain how to select the treatment option (BAHS, CROS, CI) best fit for subjects with single-sided deafness.

Table of Contents

Impact of Single-Sided Deafness

– What is the problem?

Adults and children

- Speech recognition in background noise
- Localisation of sound sources
- Tinnitus
- Listening effort
- Low self-efficiency, low self-confidence, social withdrawal

Impact of Single-Sided Deafness – What is the problem?

Adults and children

- Speech recognition in background noise
- Localisation of sound sources
- Tinnitus
- Listening effort
- Low self-efficiency, low self-confidence, social withdrawal

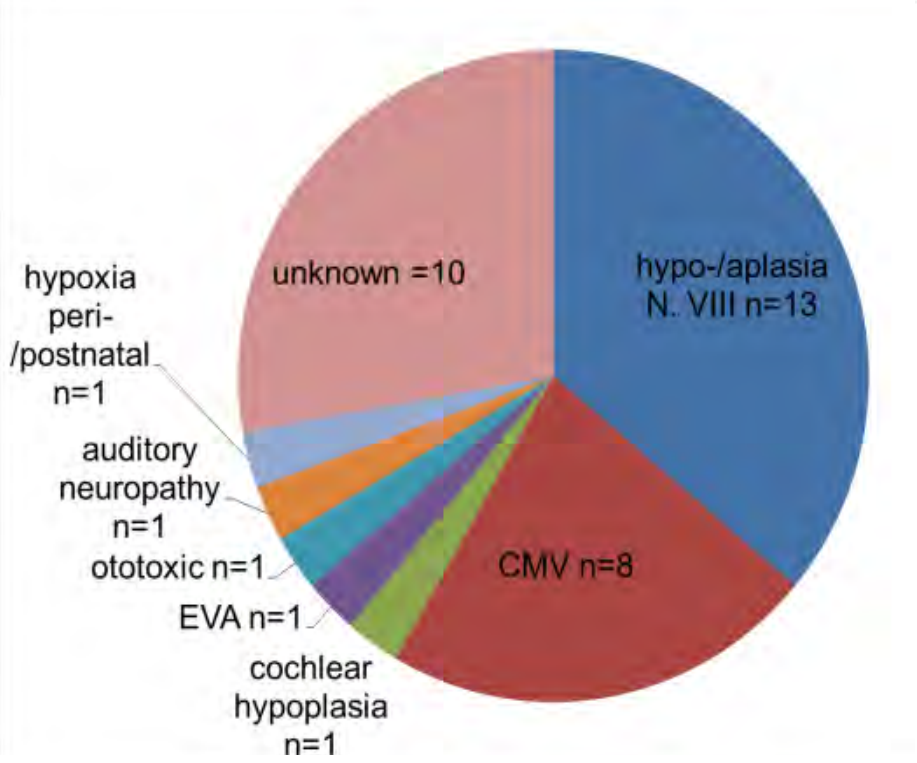
Children specific

- Speech therapy
- Grade failures

Etiology of Single-Sided Deafness

Children

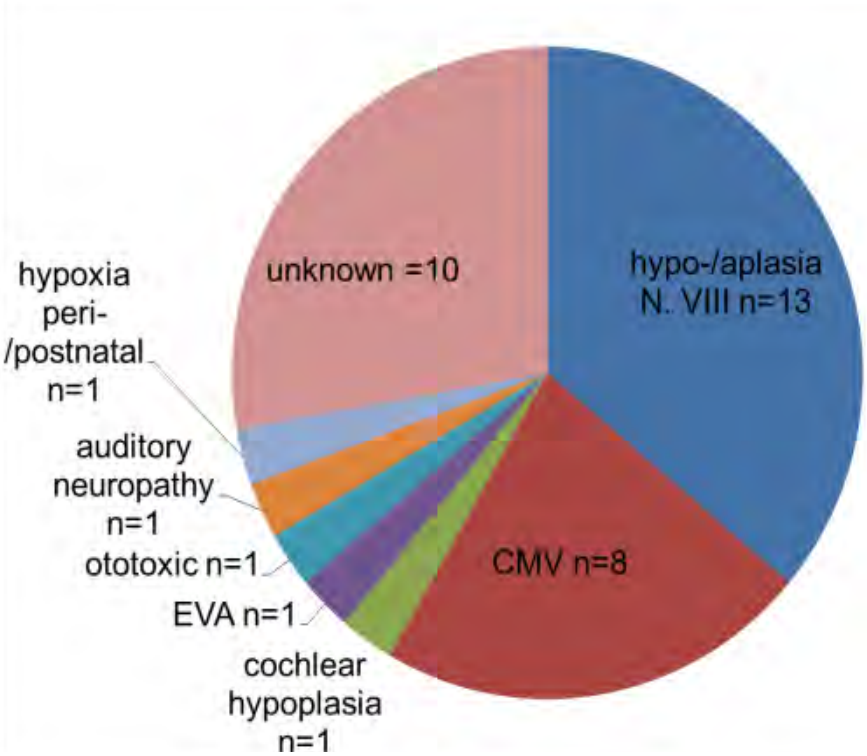
Rauch et al.
Eur Arch Otorhinolaryngol. 2021 Sep;278(9):3245-3255.



Etiology of Single-Sided Deafness

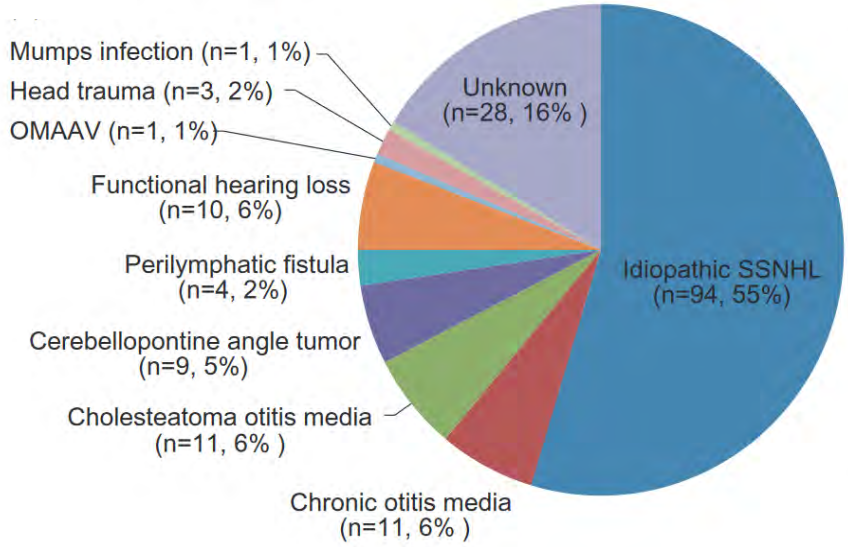
Children

Rauch et al.
Eur Arch Otorhinolaryngol. 2021 Sep;278(9):3245-3255.



Adults

Usmani et al.
Acta Otolaryngol. 2017 137(sup565):S2-S7.



Intrachochlear schwannoma 0.81 to 1.1 cases of 100,000 cases

Diagnostics

– Preoperative workup

1. Medical and personal history
2. ENT examination
3. Audiological examination
4. Vestibularis examination
5. Imaging
6. Rehabilitation capability
7. Consultation

Diagnostics

– Preoperative workup

1. Medical and personal history
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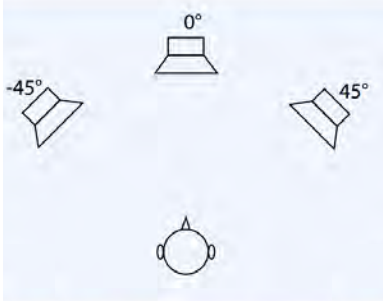
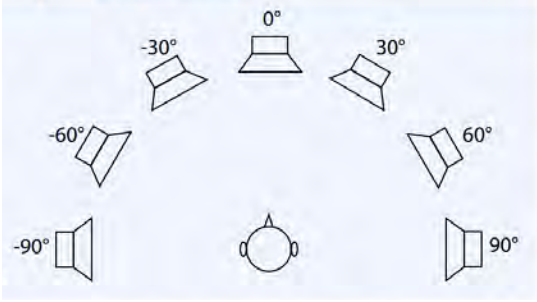
Diagnostics
– Preoperative workup

3. Audiological examination



- Subjective hearing tests with and without hearing aids
  - a. Pure-tone
  - b. Speech: numbers and word
  - c. **Sentence** in quiet and **background noise**
  - d. **Localisation of sound sources**



Arndt et al. HNO 2011 May;59(5):437-46.

Diagnostics
– Preoperative workup

3. Audiological examination

- Objective hearing tests
  a. OAEs
  b. BERA or
  c. Electrochochleography or
  d. Promontory test

Diagnostics
– Preoperative workup

Towards a Unified Testing Framework for Single-Sided Deafness Studies: A Consensus Paper

Paul Van de Heyning[a,b] Dayse Távora-Vieira[c,d] Griet Mertens[a,b] Vincent Van Rompaey[a,b] Gunesh P. Rajan[c,d] Joachim Müller[e] John Martin Hempel[e] Daniel Leander[e] Daniel Polterauer[e] Mathieu Marx[f] Shin-ichi Usami[g] Ryosuke Kitoh[g] Maiko Miyagawa[h] Hideaki Moteki[g] Kari Smilsky[i] Wolf-Dieter Baumgartner[j] Thomas Georg Keintzel[k] Georg Mathias Sprinzl[l] Astrid Wolf-Magele[l] Susan Arndt[m] Thomas Wesarg[m] Stefan Zirn[m,n] Uwe Baumann[o] Tobias Weissgerber[o] Tobias Rader[p] Rudolf Hagen[q] Anja Kurz[q] Kristen Rak[q] Robert Stokroos[r] Erwin George[r] Ruben Polo[s] María del Mar Medina[s] Yael Henkin[t] Ohad Hilly[u] David Ulanovski[u] Ranjith Rajeswaran[v] Mohan Kameswaran[v] Maria Fernanda Di Gregorio[w] Mario E. Zernotti[x]

- **Recommended outcome measurements over time**

Outcome measure	Baseline	After each 3-week trial	Months after device provision or nontreatment			
			1	3	6	12
PTA	both ears	better ear	better ear	better ear	better ear	better ear
Speech perception in noise	yes	yes	yes	yes	yes	yes
Sound localization	yes	yes	yes	yes	yes	yes
QoL	yes	yes	yes	yes	yes	yes
Tinnitus, if applicable	yes	yes	yes	yes	yes	yes
Daily device use	–	yes	yes	yes	yes	yes

Van de Heyning et al. Audiol Neurotol 2016;21:391-398

Diagnostics

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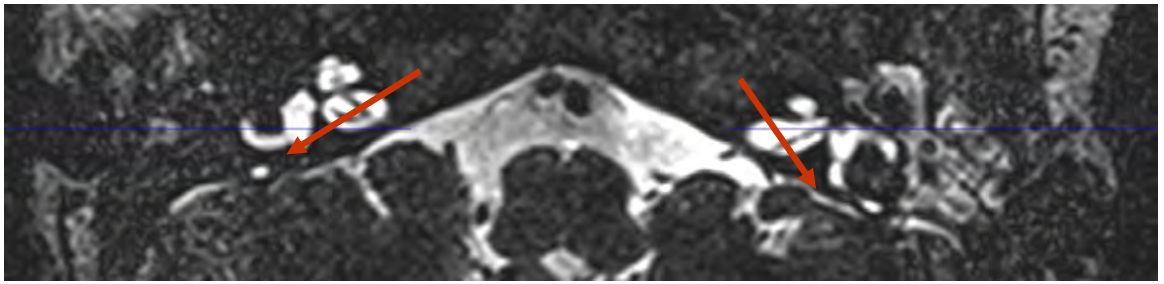
4. Vestibularis examination
5. **Imaging**
6. Rehabilitation capability
7. Consultation

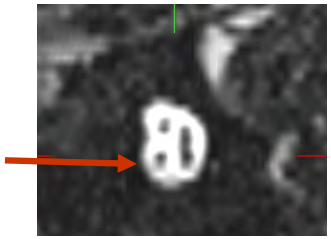
Diagnostics

– Preoperative workup

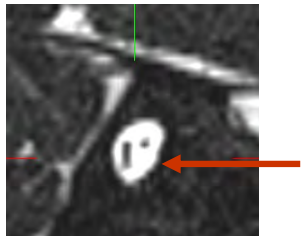
5. Imaging

- MRI





CND left

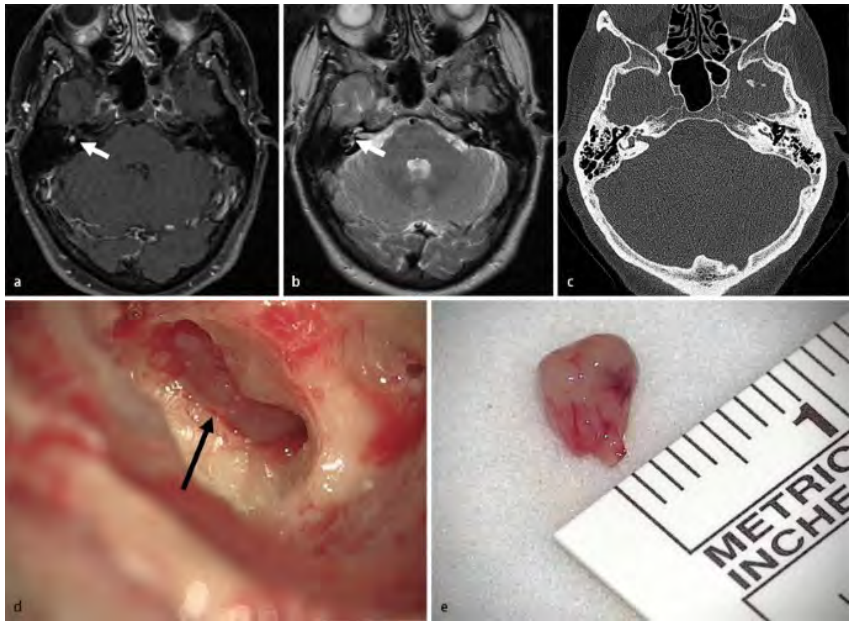


- CT or DVT
  - Children: CT only if necessary based on MRI
  - Adults: DVT preferred, CT only if necessary based on medical history

Tsukada et al. Acta Otolaryncol. 2021 Sep;141(9):835-840.

Diagnostics
– Preoperative workup

Intralabyrinthine schwannoma right



5. Imaging

- MRI
- CT or DVT
  - Children: CT only if necessary based on MRI
  - Adults: DVT preferred, CT only if necessary based on medical history

Aschendorff et al., HNO. 2017 Jan;65(Suppl 1):46-51.

Diagnostics
– Preoperative workup

1. Medical and personal history
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4. Vestibularis examination
5. Imaging
6. **Rehabilitation capability**
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Treatment Options

1. Observation and rely on compensation mechanisms
2. CROS hearing Aids (Contralateral Routing Of Signal)
3. Bone conduction devices (percutan/transcutan)
4. Wireless microphone technology (Phonak Roger Focus)
5. Cochlear Implant

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Overcoming the head shadow
Transmission of signals from the deaf side to the normal hearing ear

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Transmission of signals from the deaf side to the normal hearing ear

Transmission of speech directly into the normal hearing ear

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Re-Habilitationof binaural hearing

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Overcoming the head shadow
Transmission of signals from the deaf side to the normal hearing ear

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Transmission of speech directly into the normal hearing ear

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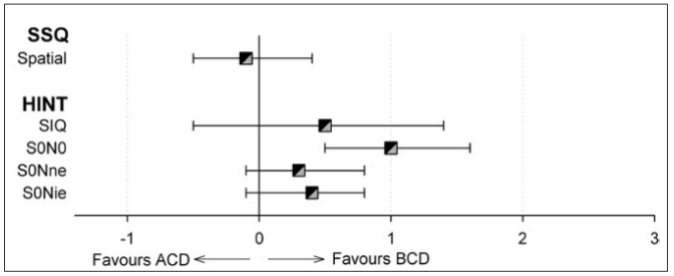
Re-Habilitationof binaural hearing

→ individual treatment concept for optimal therapy
Why? Individual needs; avoidance of non-users

Treatment Options

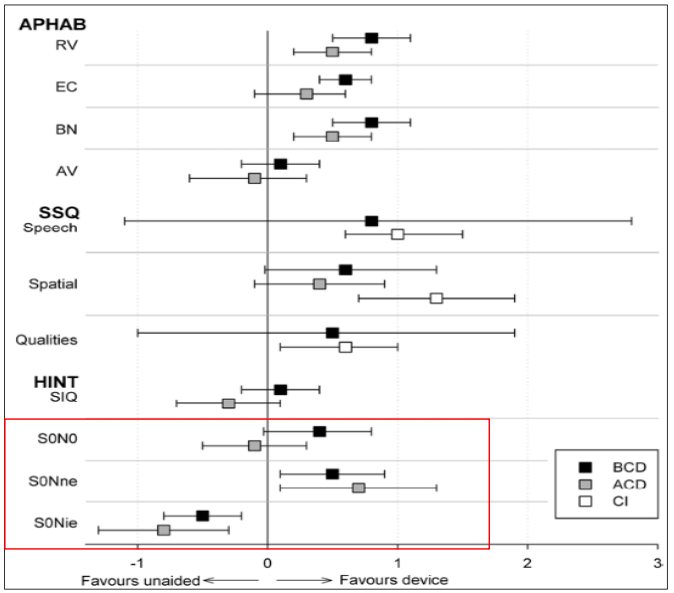
CROS systems (CROS HG / BAHS)

- **Reduces head shadow**
- Interference with normal-hearing ear
- No summation effect
- No reduction of tinnitus
- No improvement in localization



Cochlear Implant

- **Reduces head shadow**
- **No interference with normal-hearing ear**
- Summation effect unclear
- **Reduction of tinnitus in 70%**
- **Improvement of localization**

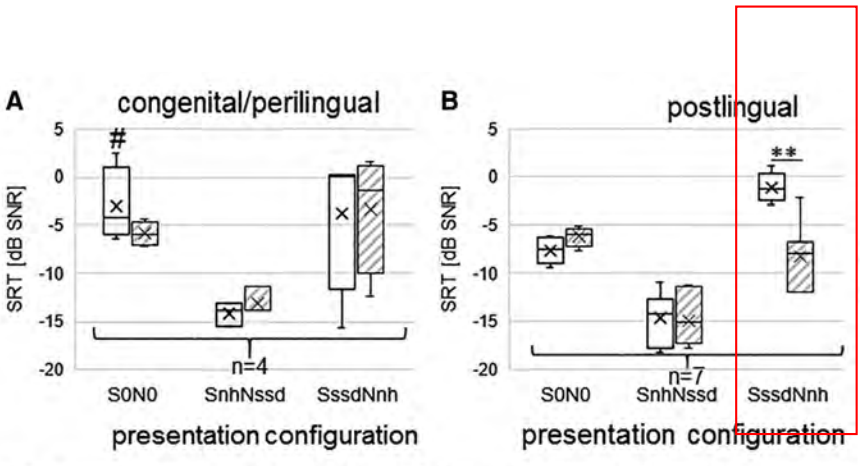


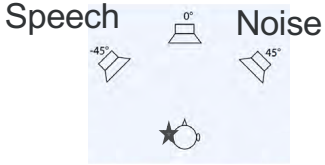
Kitterick et al. Ear & Hearing 2016;37;495–507

Speech Recognition in Background Noise
– Long-term outcome

Children

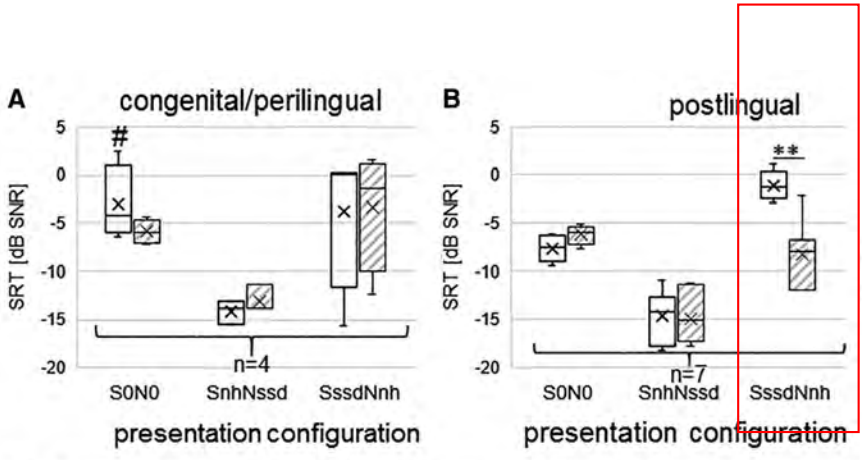
Arndt et al.
Ear & Hearing 2023 Sep 20.

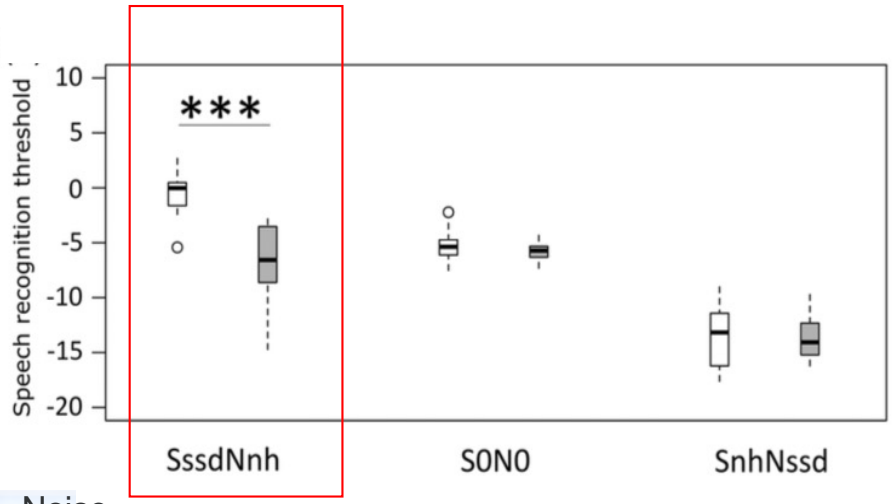


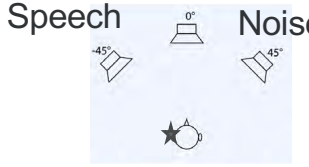


Speech Recognition in Background Noise
– Long-term outcome

Children	Adults
Arndt et al. Ear & Hearing 2023 Sep 20.	Speck et al. Eur Arch Otorhinolaryngol. 2021 Sep;278(9):3257-3265



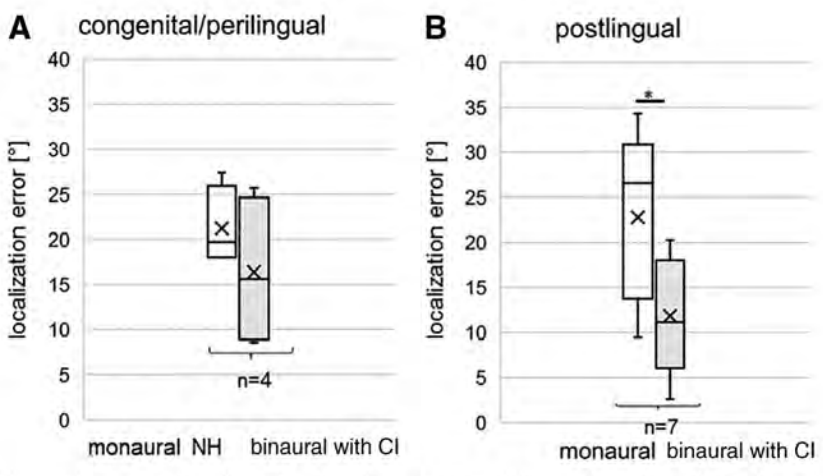




Localization of Sound Sources – Long-term outcome

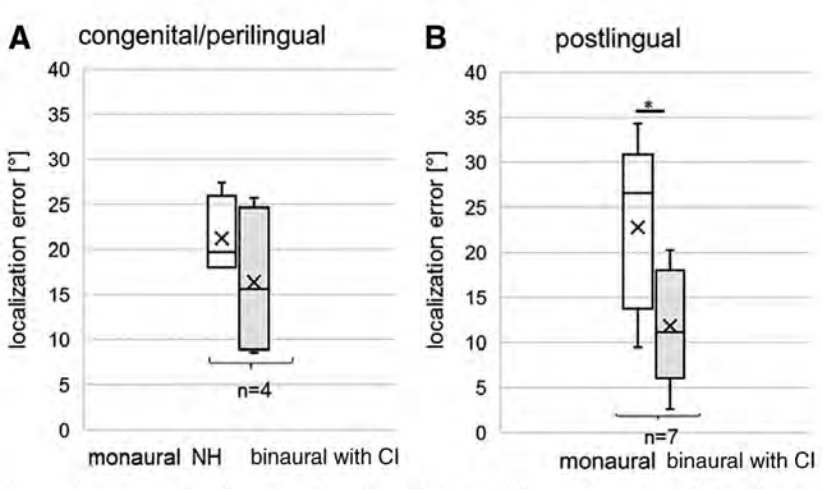
Children

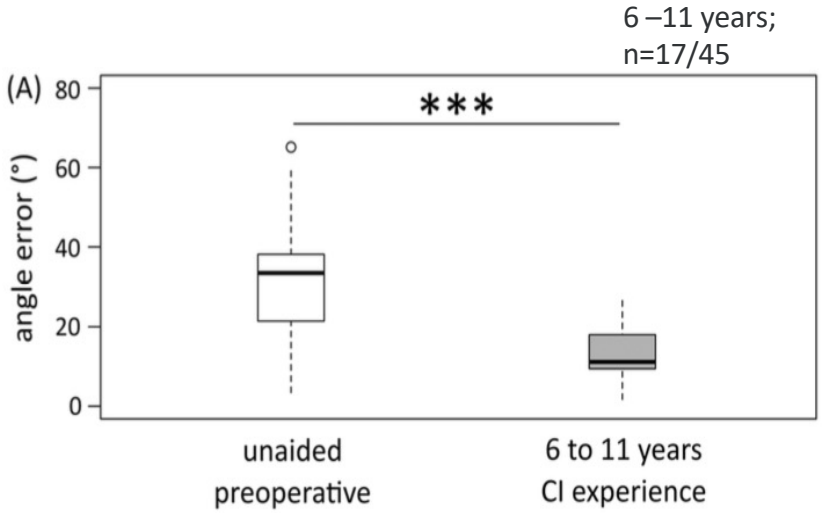
Arndt et al.
Ear & Hearing 2023 Sep 20.



Localization of Sound Sources – Long-term outcome

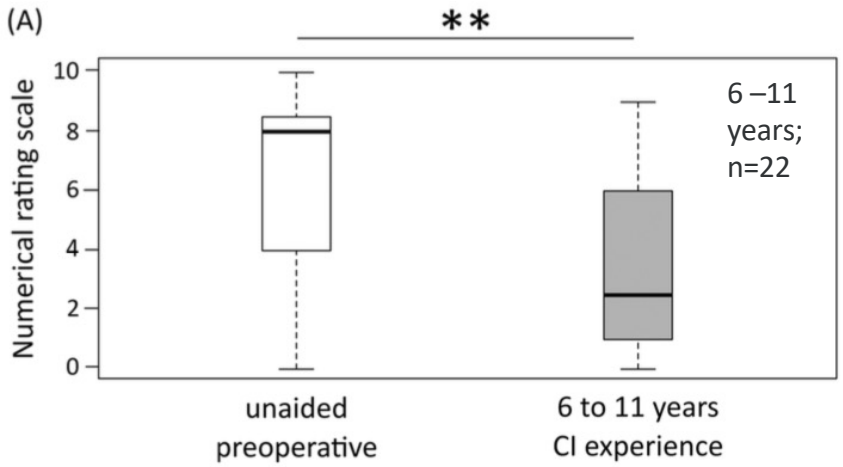
Children	Adults
Arndt et al. Ear & Hearing 2023 Sep 20.	Speck et al. Eur Arch Otorhinolaryngol. 2021 Sep;278(9):3257-3265





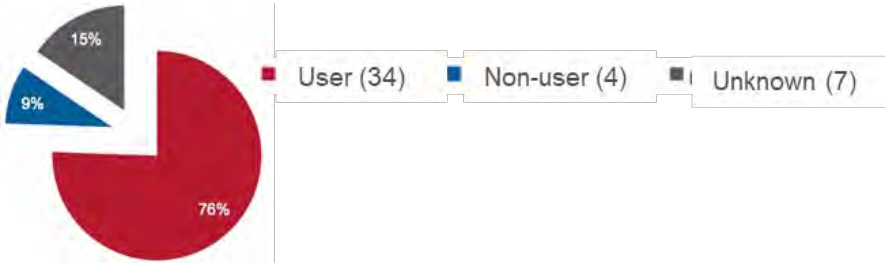
Tinnitus

– Long-term outcome

Children	Adults
Arndt et al. Ear & Hearing 2023 Sep 20.	Speck et al. Eur Arch Otorhinolaryngol. 2021 Sep;278(9):3257-3265
12 of 19: no tinnitus **1 of 7 with tinnitus reported an increase with CI**	

CI-Usage

– Long-term outcome

Children	Adults
Arndt et al. Ear & Hearing 2023 Sep 20.	Speck et al. Eur Arch Otorhinolaryngol. 2021 Sep;278(9):3257-3265
	
Non-user (n=4/36)= 11% All 4 nonuser: congenital onset of SSD and implanted after the age of 3 years	**Non-user (n=4/45)= 15%** 1. risk of dirt and splashes at work 2. professional and private stress 3. dissatisfied with the hearing impression 4. no speech understanding

Patient Selection to Prevent Non-users

- **Adults**
  - CI in SSD/ AHL patients is a superior treatment option after positive pre-therapeutic diagnostics (contraindication: absence of cochlear nerve), independent of the duration of deafness in adults and children with acquired hearing loss.
- **Children**
  - We still don't know the optimal time window for favorable outcomes in children with congenital SSD – is it wider or narrower than in bilateral hearing loss?
  - Congenital SSD children benefit from cochlear implantation before the age of 3 years.
  - Acquired unilateral deafness without age limit in children

- In all other cases (no CI-indication, refusal of CI-surgery) the Bi-/CROS and osseointegrated bone conduction systems or other assistive devices are still an option. Counselling of limited benefit and no prevention of unilateral auditory deprivation is necessary.
- Identification of markers that may provide information on outcome with CI

Is Neural Activity…

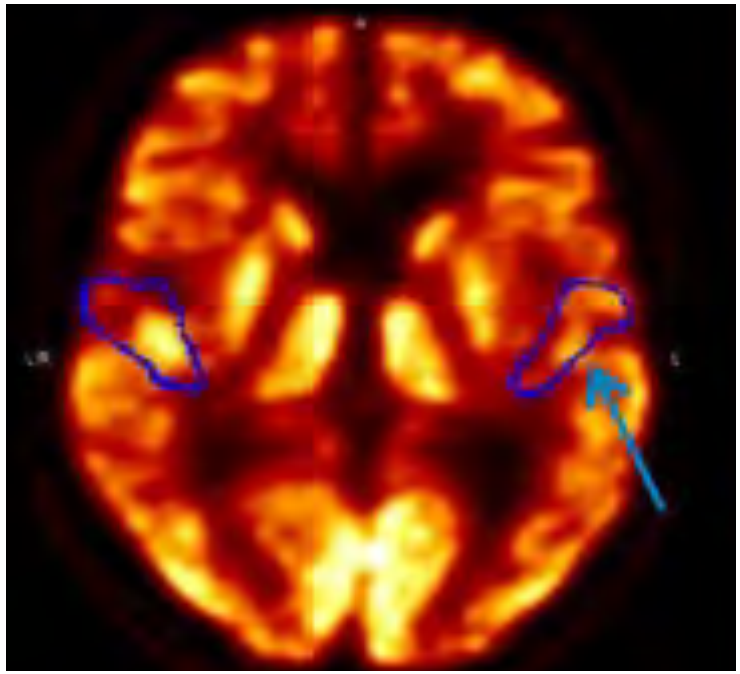
– Current research on neural plasticity

- …altered by single-sided deafness?

Is Neural Activity…

…altered by single-sided deafness?

Primary auditory cortex



^{18}F-FDG PET

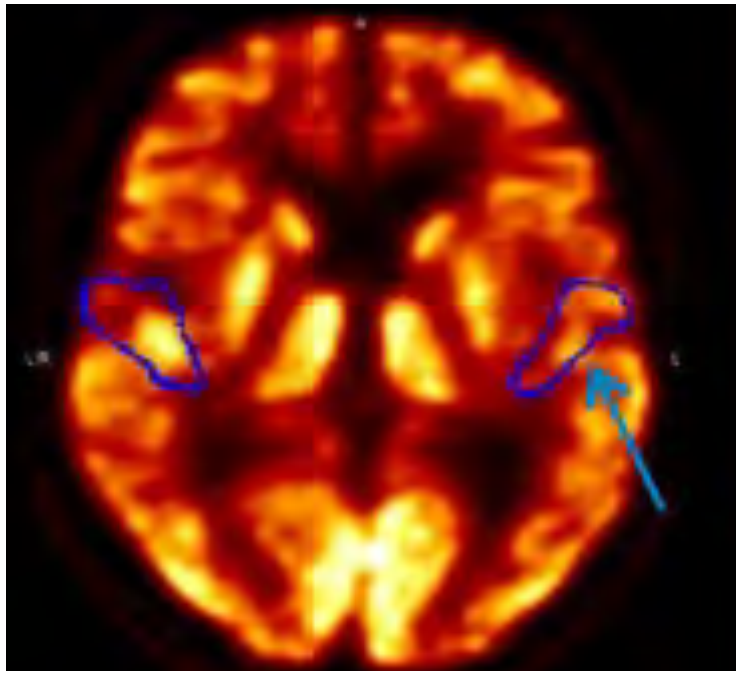
1.6
1.2
0.8
0.4
0.0

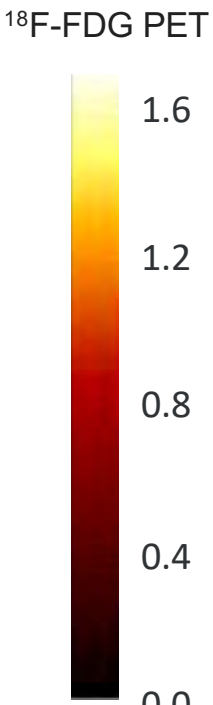
Speck I et al. J Nucl Med. 2020 Mar;61(3):418-422

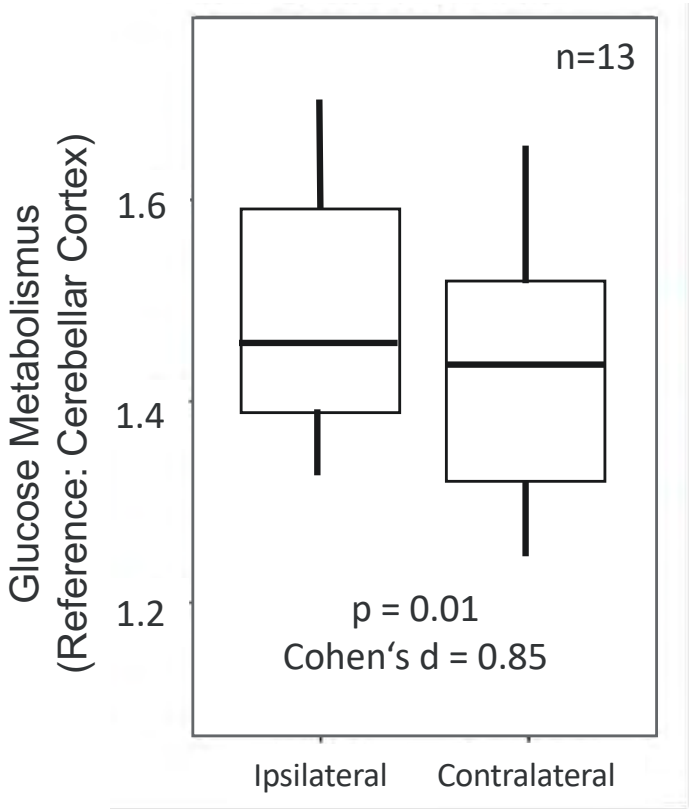
Is Neural Activity…

…altered by single-sided deafness?

Primary auditory cortex







Speck I et al. J Nucl Med. 2020 Mar;61(3):418-422

Is Neural Activity…

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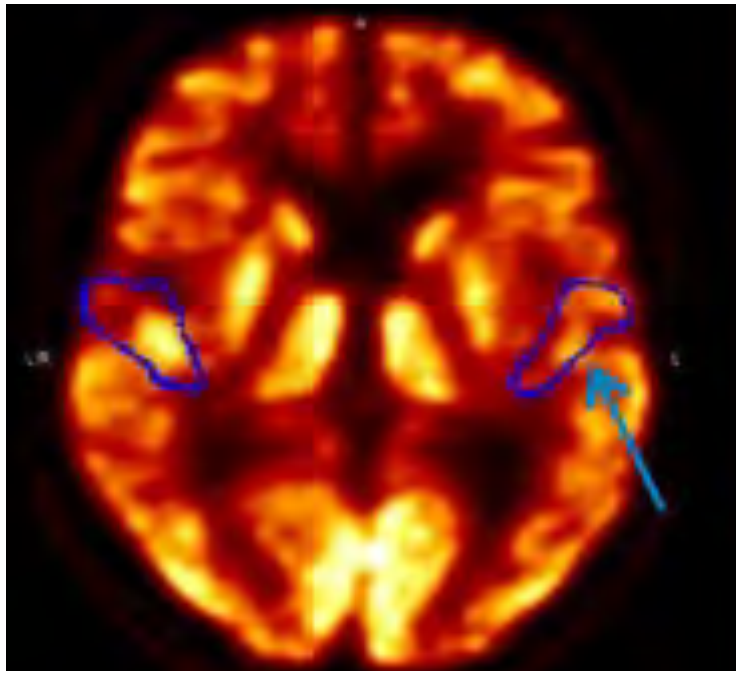
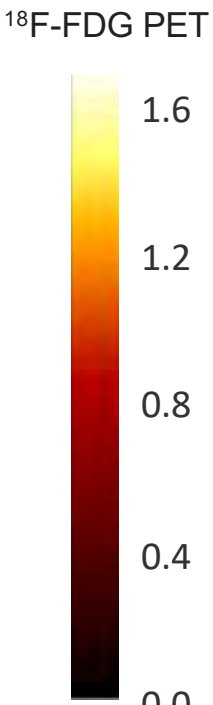
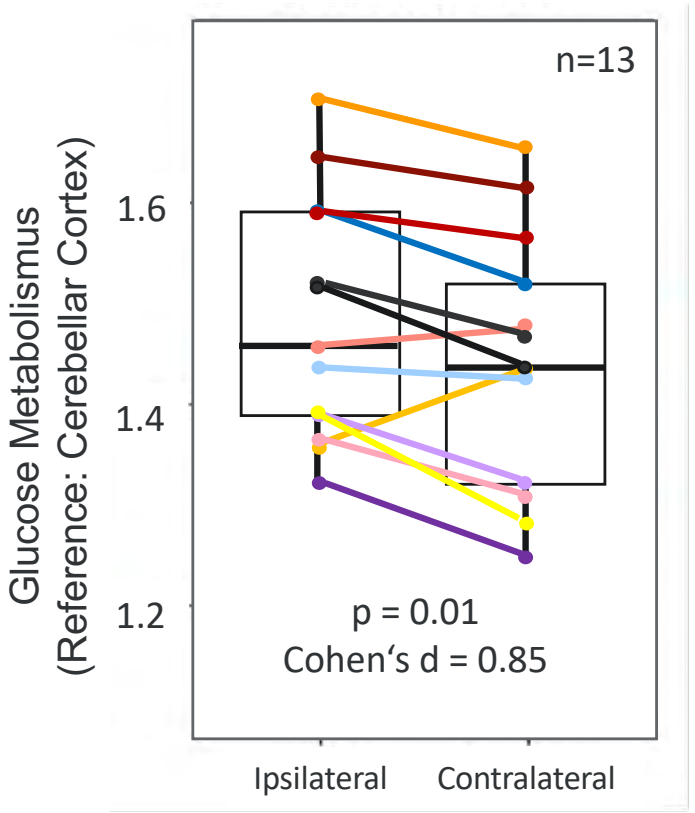
Primary auditory cortex







Speck I et al. J Nucl Med. 2020 Mar;61(3):418-422

Is Neural Activity…

– Current research on neural plasticity

1. …altered by single-sided deafness? **– YES**

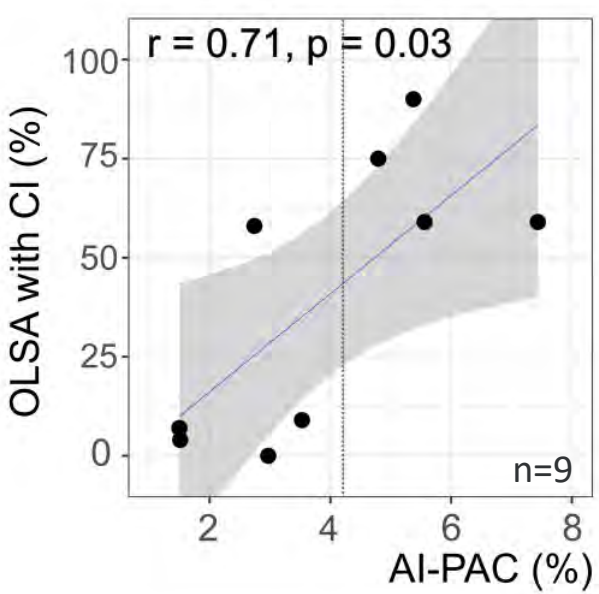
Is Neural Activity…

– Current research on neural plasticity

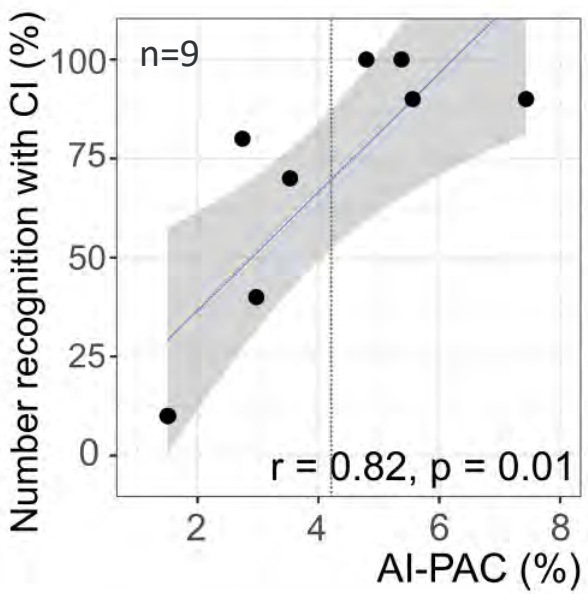
1. …altered by single-sided deafness? **– YES**
2. …a predictor for success with CI?

Is Neural Activity…
…a predictor for success with CI?

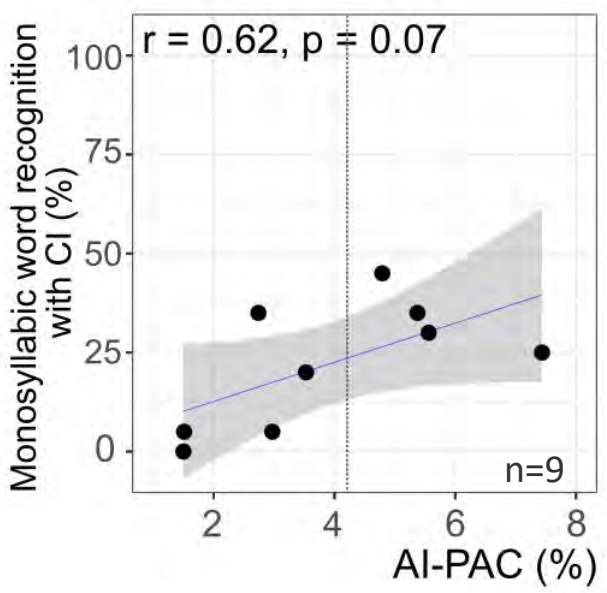
Sentence recognition with CI



Number recognition with CI



Word recognition with CI



Speck I et al. Sci Rep 2022 16;12(1):8068.

Is Neural Activity…

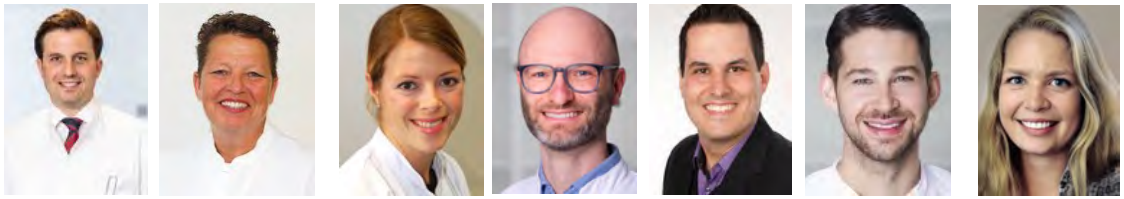
– Current research on neural plasticity

1. …altered by single-sided deafness? **– YES**
2. …a predictor for success with CI? **– YES: more pat. and longer follow**

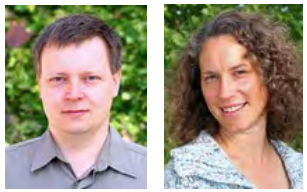
Summary

- Elimination of head shadow, better localization ability and tinnitus reduction prove that CI is a successful treatment option for patients with SSD
- Patient selection and education are essential to avoid non-users
- Second choice after CI is a bone anchored hearing aid
- Reliable biomarker as predictor for outcome after CI still missing – ongoing research

Many thanks to our research team



Iva Speck & Susan Arndt



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