Cochlear Implantation: Concept, Results Outcomes and Quality of Life

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ABSTRACT

Cochlear implants today are an essential method of auditory rehabilitation in patients with severe to profound hearing loss. Due to the rapid development of implant technology the results have been markedly improved. Today about 80 % of patients can use the telephone and children achieve near to normal hearing and speech development. In consequence, more patients are candidates for a cochlear implant today including those with high frequency deafness and single sided deafness. However, today only 60,000 out of 1 Million CI-candidates in Germany have been implanted so far. In future multi modal universal auditory implants will provide combined electric-mechanical stimulation to make best use of the residual auditory hearing and the electrical stimulation of the auditory nerve. They allow a continuous adaptation of the stimulation strategy onto the given functional status of haircells and auditory nerve fibers especially in cases of progressive hearing loss. Brain computer interfaces will allow the automated fitting and adaptation to the acoustic scene by optimizing the signal processing for best possible auditory performance. Binaural hearing systems will improve directional hearing and speech perception in noise. Advanced implants are composed of individualized electrodes by additive manufacturing which can be inserted atraumaticly by computer and robot assisted surgery. After insertion they automatically adapt to the anatomy of the individual cochlea. These advanced implants are composed with additional integrated biological components for the preservation of residual hearing and regeneration of neural elements to improve the electrode nerve interface. This will allow to increase the number of electrical contacts as a major step towards the bionic ear. This will overcome the principal limits of today's cochlear implant technology. Advanced care models will allow an easy way for the patient towards hearing preservation cochlear implantation under local anesthesia using minimal invasive high precision cochlear implant surgery. These implant systems will become a personal communicator with improved connectivity. Remote care and self-fitting will empower the patient to optimize his own hearing.

Contents

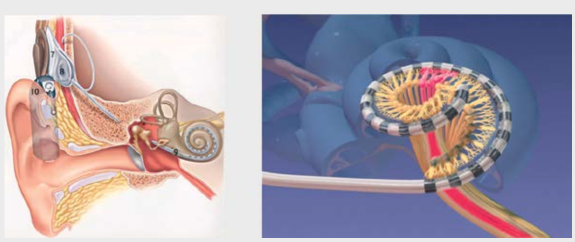
1. Cochlear implantation today

Today, cochlear implants (CI) are the therapy of choice for hearing rehabilitation in severe and profound hearing loss as well as deafness with sensory or perisynaptic origin in pediatric and adult patients [1, 2]. They replace the function of the deficient inner hair cells by direct electrical stimulation of the auditory nerve via intracochlear multichannel electrodes (▶ **Fig. 1**). Due to the rapid technological development, especially of microprocessors since the 1980s the hearing results could be significantly improved so that the indications for implantation were consecutively extended. With more than one million recipients, CI may be considered as success story of neuroprosthetics (▶ **Fig. 2**). While at the beginning of the 1980s only patients with bilateral deafness were expected to be candidates for cochlear implantation, nowadays more and more patients with residual hearing and speech comprehension are implanted (▶ **Fig. 3**).

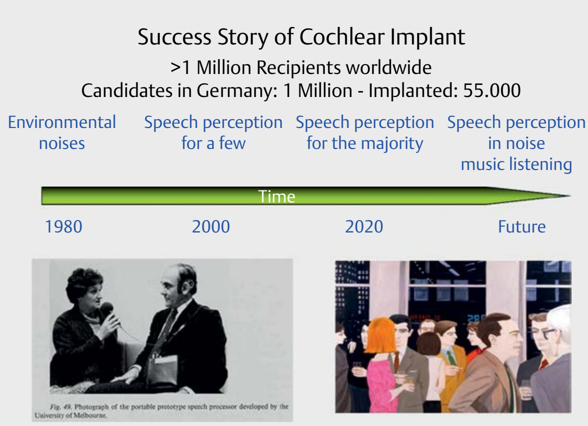
A CI is indicated when better speech comprehension may be expected compared to alternative methods like hearing aids, bone-anchored hearing systems, acoustic implants, or hearing-improving surgeries (▶ **Fig. 4**). Thus, one precondition is performing adequate, age-related multidimensional diagnostics as well as a reliable prediction of the expected hearing outcome. Numerous factors have to be included in the decision-making process in order to assure that the patient´s hearing and the quality of life improve.

The large spectrum of etiology and pathophysiology of hearing loss requires an individualized treatment concept. Today, hearing implants are available for all grades and types of hearing loss (▶ **Fig. 5**) to achieve the best possible hearing rehabilitation. Individualized cochlear implantation is an outstanding example for precision medicine using advanced technology to achieve the best possible auditory performance for patients suffering from severe or profound hearing loss. Important paramteres are the anatomy of the cochlea, residual hearing, the function of the auditory nerve as well as the information processing along the auditory pathways, cognitive factors and additional disabilities. These factors must be taken into consideration to select stimulation mode (electrical stimulation [ES] versus electro-acoustic stimulation [EAS]), electrode array, atraumatic surgical technique, appropriate speech processing strategies, and concept of auditory training.. Rehabilitation and lifelong follow-up are further components of an integrated care model of cochlear implantation.

Nowadays, objective measures are an indispensable tool for all phases of cochlear implantation. This includes preoperative objective audiometry for assessment of the severity and type of hearing loss, intraoperative monitoring of residual hearing and testing of function of the auditory nerve and pathways, postoperative fitting as well as follow-ups and longterm care with functional check-ups of the implant and the management of complications.

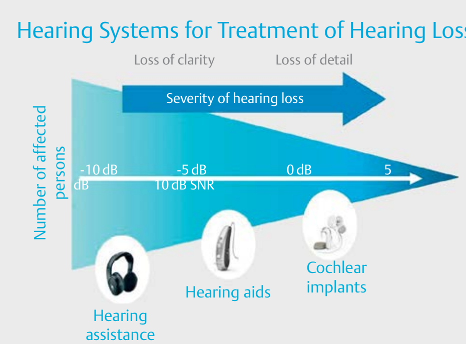


▶ **Fig. 1** Principle of current Cochlear Implant Systems (permit by Cochlear Pty, already published in T. Lenarz, 2017).

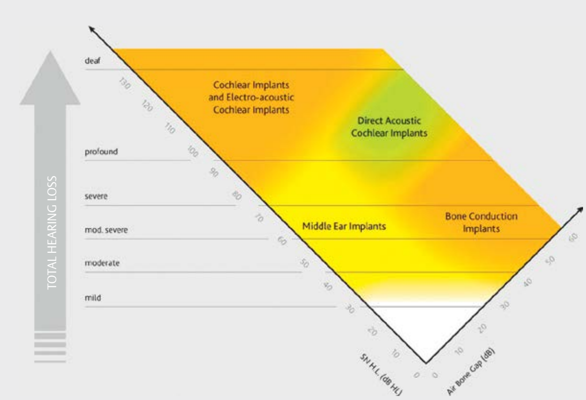


▶ **Fig. 2** Development of Cochlear Implant.

▶ **Fig. 3** Indications for CI since 1990.



▶ **Fig. 4** Indications for different hearing systems depending on speech in noise perception(permit by Cluster of Excellence Hearing-4All).



▶ **Fig. 5** Indications for different auditory implants in relation to air and bone conduction threshold (permit by Cochlear Pty).

Modern CI systems allow the wide use of audiotechnology by means of specific transmission protocols such as Bluetooth. This so-called connectivity enables, among other features, the easy use of cell phones and assistive listening devices. Telemedicine applications include implant check, remote fitting, and the transmission of technical and audiological data. Remote care allows a significant simplification of postoperative rehabilitation and aftercare with direct access to the expertise of the implant center. Above that, integrating regional cooperation partners allows establishing networks for integrated care models (hub and spoke).

The implant system allows patients to perform technical control tests of their implant, self fitting and self assessment of auditory performance (patient empowerment). In this way, fine adjustments for specific hearing situations can be done contributing to further improvements of the individual hearing performance.

The remote transmission of these data allows for continuous monitoring of implant function and electrode impedances leading to an automated detection of technical defects and medical complications.

Collecting the data of numerous patients allows to establish large databases including CI registries. The application of artificial intelligence on these data facilitate comparisons of different implant types, detection of technical defects and error patterns, and medical complications. This big-data approach even permits the calculation of models to predict the individual hearing outcome.

Telemedicine, remote care, and self-care are essential components of a far advanced implant technology which serves as a role model for an effective functional rehabilitation of chronic diseases.

2. CI Technology

2.1 CI System

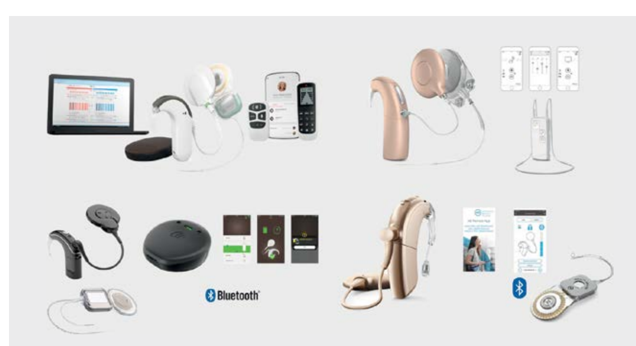
Today, CI systems are partially implantable and consist of an external speech processor and the actual implant, also called receiver-stimulator (▶ **Fig. 6**) [3]. The power transmission to operate the implant is inductive, the signal transmission occurs via a bidirectional radiofrequency transmission link. Fully implantable systems are currently being developed since basic experiences regarding the advantages and disadvantages could be collected with the first device generation in few pilot patients. The advantages of invisibility (invisible hearing) and situation-independent applicability are countered by acoustic disadvantages due to the retroauricular position of the subcutaneous microphone, the limited service life of the rechargeable battery of probably a maximum of 10 years with consecutively required reimplantation, and the technological upgrade limited to just software.

2.1.1 Implant (receiver-stimulator)

The implants are active systems for controlled electrical stimulation of the auditory nerve. They contain high-performance circuits able to create several ten thousand pulses per second. Based on the speech processing algorithm, these are distributed to the single contacts of the electrode array according to the transmitted input signal. The power is supplied inductively from the externally worn sound processor. The transmission pathway is transcutaneous via two aligned electrical coils (one located in the implant and the other one connected to the sound processor) that are kept centered in position by means of integrated magnets.

The implants also have a backward telemetry allowing the recording, amplification and reverse transmission of functional data of the implant as well as electrophysiological parameters. These objective measures are an important diagnostic tool that can be applied intra- and postoperatively at any time. In detail, they provide:

- Measurement of the electrode impedances for characterization of the function of the single electrode contact, the electrode-nerve interface and changes over time. By using different electrical stimulation frequencies the correct position of the electrode array in the cochlea and malposition like tip foldover (▶ **Fig. 42**) can be estimated.
- Measurement of evoked potentials of the inner ear and the auditory nerve
  - Cochlear microphonics (CMs) from the outer and inner hair cells for assessment and monitoring of residual hearing in hearing preserving cochlear implantation (so-called cochlear monitoring) during and after surgery (▶ **Fig. 28** and ▶ **29**). Acoustic stimulation of different frequencies are delivered through an insert ear phone in the external auditory canal.
  - The evoked hair cell responses are measured through the apical electrode contacts of the electrode array. The amplitude grows with increasing insertion depth, reductions reveal possible mechanical interactions with intracochlear structures, most probably with the basilar membrane. If the reduction can be reversed by correcting the electrode position, a low impact on residual hearing may be expected, otherwise substantial damage must be assumed [4, 5].
  - Electrical Compound Action Potentials (ECAP) of the auditory nerve evoked by intracochlear electrical stimulation to assess the neural responses of the auditory nerve. Depending on the cochlear implant manufacturer, the method is named neural response telemetry (NRT), imaging (NRI), or auditory response telemetry (ART). Hereby, action potentials are measured by means of the contacts neighboring the stimulation electrode. To some



▶ **Fig. 6** Current CI-Systems (Selection) with Receiver/Stimulator and externally worn speech processor.

extent they allow fitting of the CI to the individual properties of the auditory nerve (NRT based fitting in children) and the assessment (of changes) of the electrical stimulating properties of the auditory nerve. In a similar way the spread of excitation for assessing channel interaction inside the cochlea and the electric field distribution can be measured [6]. To some extent, the functional status of the auditory nerve can be checked [7] using this technique. Summating potentials (SP) generated by the (inner) hair cells can be also recorded to assess residual hearing during hearing preservation cochlear implantation and to determine the electrode position in the cochlea (rarely applied) [8].

Electrically elicited stapedius reflex (ESRT). This is an indirect measure of stimulation of the auditory nerve with suprathreshold stimuli. The threshold correlates with the so-called C level, i. e. the comfortable loudness level. If the stapedius reflex is elicited, an intracochlear position of the electrode can be assumed. Similar to the compound action potential, the threshold value can be used to support the fitting of the speech processor.

Objective measures play an increasing role in remote care, e. g. monitoring the implant function including failure detection or early detection of medical complications like inflammatory processes in the cochlea that can be identified due to increased electrode impedances.

2.1.2 Electrodes

Nowadays, electrodes are designed nearly exclusively as intracochlear electrode arrays to be placed in the scala tympani (▶ **Fig. 7**). Extracochlear electrodes are used as reference electrodes and are either case electrodes, part of the electrode array, or separate electrode arrays . The electrical pulses are delivered to the surrounding tissue via electrical contacts of the intracochlear electrode array. Depending on the manufacturer, their number varies between 12 and 22 active contacts. According to the design of the electrode array, there are different distances to the neuronal elements of the auditory nerve. Straight electrodes are available with different lengths of 16 to 31 mm and are placed at the lateral wall of the cochlea while preformed electrode arrays are designed for a perimodiolar position. Due to the high variability of the cochlear anatomy, in particular with regard to length, resulting individual differences of the so-called cochlear coverage, i. e. the part of the cochlea covered by the electrode (▶ **Fig. 18**) as well as the insertion angle using the same electrode have to be taken into consideration for individualized cochlear implantation.

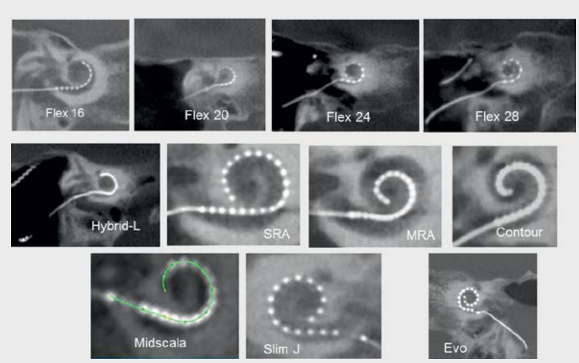
A selective stimulation of auditory nerve fibers shall be achieved through a high number of electrode contacts with sufficient channel separation. However, overlapping electric fields due to spread of excitation lead to non controllable channel interactions. Therefore a significant increase of the number of electrical contacts does not seem to be reasonable to improve the selectivity of stimulation with the current type of electrode-nerve interface. In this way, tonal hearing with presentation of single frequencies is not possible; instead of single frequencies, frequency ranges are allocated to the single electrode contacts. This distribution is based on critical bands and sufficient for speech comprehension while there are clear limits for music listening. Significant improvement of channel separation might be achieved by intraneural electrodes of the auditory nerve implant (ANI) (▶ Fig. 61) [9].

It is still unclear which parts of the auditory nerve are stimulated by the electrical pulse. In cases of complete deafness with degeneration of dendrites, mainly the spiral ganglion cells in Rosenthal's canal and the axons in the modiolus will be stimulated while in cases with residual hearing, i. e. predominantly in the apical region, also dendrites and hair cells (electrophonic hearing) may be additionally stimulated.

2.1.3 Speech processor and speech processing

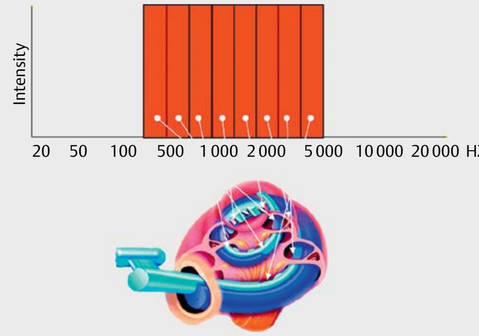
The external speech processor (▶ **Fig. 8**) is realized either as an ear level processor or as a button processor. It consists of a microphone system for sound registration, several stages for sound pre-processing, a processing stage for translating the sound signal into an electrical stimulation code and a device for transcutaneous transmission of the coded signal to the implant via a high frequency transmission link. The processor is powered by either single use or rechargeable batteries. The processor also powers the implant itself through an inductive link between the external coil of the processor and the internal coil of the implant. The implemented magnet inside the external coil as well as inside the implant keep the transmission coils centered to each other ensuring optimal positioning of the transmitter and receiver antennas. Further components encompass a backward telemetry to register device feedback regarding the functional status of the implant and the objective measures (see chapter 2.1.1). In addition, the speech processor incorporates features for connectivity like Bluetooth receivers and streaming of data or even broadband audio-signals to hearing systems worn on the contralateral ear.

Coding of the acoustic input signal into electrical stimulation patterns of the auditory nerve also requires an adaptation of the acoustic dynamic range. This is done by compressing the acoustic input signal of, for example, 60 dB to a generally much smaller electrical dynamic range of the individual patient of usually less than 15 dB. This is achieved by determining the stimulation levels of comfortable loudness (C level) as well as of the hearing threshold (T level) for every single electrode contact. Furthermore, an adap-



▶ **Fig. 7** Selection of currently available electrode systems from different manufacturers. Postop. Cone Beam CT in Stenvers projection.

▶ **Fig. 8** CI-System with integrated connectivity for contact to audiotechnology and internet. Key element for Telemedicine and Remote Care.



▶ **Fig. 9** Principle of speech coding. Tonotopic allocation of frequency bands to electrode contacts (permission by Advanced Bionics).

tation of the loudness growth function is possible to better adapt to individual needs of the patient.

The acoustic input signal is prepared for optimal speech processing by enhancing the relevant parts for speech comprehension. At the same time, reduction of negligible signal components and noise suppression are applied. Among others multi-microphone systems are applied as a beamformer in order to achieve better signal to noise ratios. The input signal processed in the above mentioned fashion (▶ **Fig. 9**). In this way, the limited information transmission capacity of the electrode-nerve interface is optimally used and not overburdened with signal components not necessary for speech perception.

To further reduce perceptually unnecessary stimulation, many speech processing algorithms apply the concept of the n-of-m strategy selecting the frequency bands with the highest amplitudes for each stimulation cycle.

A more advanced version of the n-of-m approach is based on psychoacoustic masking procedures where signal components, which are not perceivable by the auditory system, are simply omitted. This principle of psychoacoustic masking, where – simply speaking – softer tones are not perceivable in the presence of louder tones, has been studied for decades and has been used in audio compression for more than 20 years with great success (mp3 players). This principle has been transferred to the field of cochlear implants with some success [10].

Most algorithms use sequential stimulation so that the selected electrodes are addressed consecutively with the prepared stimuli. Due to the concept, an overlapping stimulation of the same nerve population with uncontrolled loudness effects is avoided.

Simultaneous stimulation strategies can be applied when a sufficient channel separation for neighboring electrode contacts with regard to the electrical field distribution may be achieved by an electrode position extremely close to the modiolus. That concept has the advantage of increased information throughput compared to sequential stimulation procedures, however, the power consumption is significantly increased.

This advantage of simultaneous stimulation is partially outweighed by today`s high rates of sequential stimulation, at least to some extent. Because of the refractory time, other neural elements are stimulated by the subsequent stimulus so that a more exact representation of the acoustic input signal becomes possible. An overview of the technical developments is provided by Büchner and Gärtner [3].

In cases of hybrid configuration for electroacoustic stimulation, the speech processor comprises an additional acoustic component similar to the components of a conventional hearing device for the low and middle frequency ranges, usually up to 2000 Hz.

2.1.4 Hybrid systems for electro-acoustic stimulation

In cases of patients with sufficient residual hearing in the low frequency range, the residual hearing may be used by natural acoustic stimulation (ENS) or by means of an additional acoustic component of the speech processor, i. e. an acoustic amplification of low frequency sounds presented into the outer ear canal (EAS). A postoperative hearing threshold at 500 Hz of 65 dB or better may be considered as the minimum for a successful application of the acoustic amplification. At 250 Hz, the minimum amounts to 50 dB. In general, stimulation strategies are applied that use a non-overlapping frequency separation between the acoustic and the electrical stimulation area. The acoustically presented part of the sound signal is below the so called cross-over frequency. The cross-over frequency is generally set at the frequency, above which an acoustic amplification is not being considered sensible, i. e. where any additional benefit for the speech understanding is not being expected. As a rule of thumb, the cross-over frequency is placed where the (usually sloping) hearing threshold of a patient dives below 70 dB HL. A certain overlapping of the electrical and acoustic frequency ranges in the cross-over area is intrinsic due to the limited edge steepness of the bandpass filters (▶ **Fig. 3** and ▶ **21**).

Due to the traveling time of the acoustic signal on the basilar membrane, the electrical stimulus is provided with delay (running time correction) in order to achieve a temporally adjusted presentation of the entire input signal.

2.1.5 Diagnostic components

Cochlear implant systems dispose of implemented diagnostic elements for technical check of the CI system, for characterization and monitoring of the electrode-nerve interface by means of electrode impedances and the neural response to electrical stimulation as well as the hair cell responses with acoustic stimulation. In this way, residual hearing and a neural response profile of the hearing nerve can be repeatedly measured objectively and compared over time. Variations of the measurement results may indicate technical or medical complications. Inflammations in the area of the inner ear (labyrinthitis) are characterized by increased electrode impedances, electrode migration by shifts of the NRT profile [11]. Intraoperatively, also a possible misplacement of the electrode array may be identified by impedance spectroscopy and corrected accordingly.

2.1.6 Telemedicine (▶ **Fig. 10**)

The connection of the cochlear implant system with the smartphone via e. g. Bluetooth does not only allow controlling and programming the implant but also to transmit data that are assessed by the implant itself (bidirectional communication). In this way, parameters like functional data of the implant and the speech processor may be transmitted wirelessly to the mobile phone and sent to an implant center. Reversely, this connection allows fine tuning of the implant as well as remote checkups regarding the integrity of the implant and the auditory performance of the patient. These so-called remote-care options play an increasing role for patients requiring close-to-home follow-up.

This opens up the possibility to monitor the integrity of the cochlear implants by means of automated algorithms. The implants can conduct continuous self-checks and objective measurements of the bioparameters and the connected smartphone sends these data to a secure central database or the database of the implanting center. In cases of critical deviations in the longitudinal data, technical or medical complications may be detected at very early stages and interventions may be scheduled (see chapter 2.1.5).

2.1.7 Self-fitting (▶ **Fig. 10**)

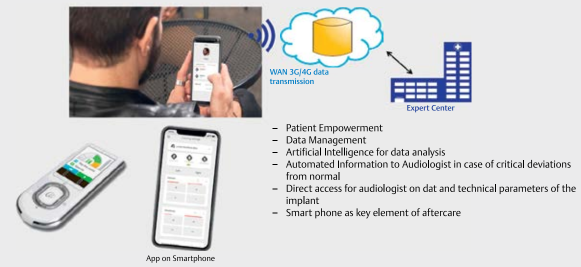
By means of computer devices like smartphones, the settings of the speech processors may be changed within predefined limits and adjusted to the respective hearing situation by the patients themselves. This leads to patient empowerment and subsequentl improvements of the settings for specific hearing situations. This approach also follows the general trend of individualized therapies.

2.1.8 Data logging

The user data of the implant system are automatically registered and give information about the intensity of use and the hearing-habits of the CI patients. In this way, valuable data may be gained for postoperative fitting and rehabilitation, such as variations of the hearing habits like increasingly avoiding noisy environments.

2.2 Bilateral stimulation

In cases of simultaneous bilateral cochlear implantation, synchronization of both implant systems is necessary. This refers to frequency allocation and thus equal pitch perception on both ears as well as on the loudness growth and balancing. Both are requisites



▶ **Fig. 10** Self Fitting and Remote Care with CI.

for use of the advantages of bilateral cochlear implantation, namely the improvement of directional hearing and speech perception in noise. Up to now, the CI systems do not use interaural time differences (ITD), phase differences, or spectral differences in their speech processing strategy, but rely mainly on interaural loudness differences (ILD).

2.3 Bimodal implantation

Patients with asymmetrical hearing may require bimodal stimulation with cochlear implant on one side and hearing aid on the other side. Also in this context, the coordination between both hearing systems is necessary in order to facilitate directional hearing and speech perception in noise. Both devices are connected to each other either wireless or via Bluetooth.

2.4 Impact of technical development on quality of life of implanted patients

In summary, the achieved technical development of CIs did lead to a significant improvement in auditory performance. At the same time, the handling became much easier. The connectivity allows the use of mobile phones, numerous audio-technological devices as well as the direct use of the internet. Improved diagnostic options facilitate troubleshooting as well as early detection of medical complications.

3. Preoperative diagnostics and indications

The process of cochlear implantation is divided into subsequent different phases that are described in various documents on quality management (AWMF guideline CI; White Paper on cochlear implantation of the German Society of Oto-Rhino-Laryngology, Head & Neck Surgery, QUINCI Quality Management of Cochlear Implantation of the health insurances companies):

- Candidacy evaluation
- CI surgery
- (Early) first fitting
- Follow-up and rehabilitation
- Aftercare

The different phases will be described in the following chapters.

3.1 Candidacy evaluation

The preoperative diagnostics are necessary to assess grade and type of hearing loss as well as to predict its further course and out-

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come with a cochlear implant. Further objectives are the identification of etiology and pathophysiology of hearing loss which can influence the therapy decision as well as the hearing outcome.

The diagnostic procedures must check the basic preconditions for cochlear implantation 1) the presence of a cochlea, 2) a functional auditory nerve, and 3) functional auditory pathways (▶ **Table 1**).

Residual hearing must be determined in relation to its impact on speech perception. This includes the air conduction threshold audiometry and speech audiometry in quiet and noise. Both methods give the essential information for the type of stimulation to be recommended, electric stimulation (ES) only or electro-acoustic stimulation (EAS) (▶ **Fig. 11**).

Optimum preoperative speech perception can be measured using the so-called Master Hearing Aid [12] (see chapter 3.3.1). The result has a high predictive value for the postoperative hearing outcome with a CI (▶ **Fig.12**).

Cognitive performance should be examined in older patients because it significantly influences the outcome of CI [13].

In children, objective audiometry plays a crucial role, especially in newborns and young children. The combination of different methods including transitory evoked otoacoustic emissions, electrocochleography with measurement of the compound action potential, the summating potential and the cochlear microphonics as well as auditory brainstem responses with the use of frequency-specific stimuli allows to determine hearing threshold as well as the type of hearing loss as conductive, sensory or neural [14]. It is also possible to detect perisynaptic hearing loss with an impaired transmission between the hair cells and the spiral ganglion cells of the hearing nerve. They have to be differentiated from cases of true neuropathy with damage of neural elements, e. g. spiral ganglion cells or axons of the auditory nerve [15].

Audiometry and vestibular examinations are completed by imaging and lab diagnostics.

High-resolution computed tomography (CT) or cone beam computed tomography (CBCT) [16] provide the anatomical information that is essential for cochlear implantation concerning size and shape of the cochlea, the internal auditory canal and the mastoid. Magnet resonance imaging (MRI) of the temporal bone and the brain allow to determine the content of the cochlea (fluid, tissue, tumor), the presence and thickness of the auditory nerve and ain structures with special reference to the auditory system and brain development. The anatomical integrity of the central auditory pathways can be visualized with diffusion tensor imaging (DTI). Functional imaging of the auditory system and associated areas of the brain can be assessed by means of functional MRI (fMRI) [17], near-infrared spectroscopy (NIRS) [18] and positron emission tomography (PET) .

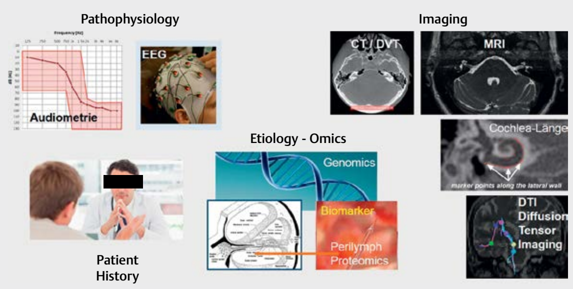
Laboratory diagnostics comprise genomics to assess genetic origin of hearing loss [19, 20] as well as proteomics of the perilymph for identification of disease-specific biomarkers [21].

3.2 Indications

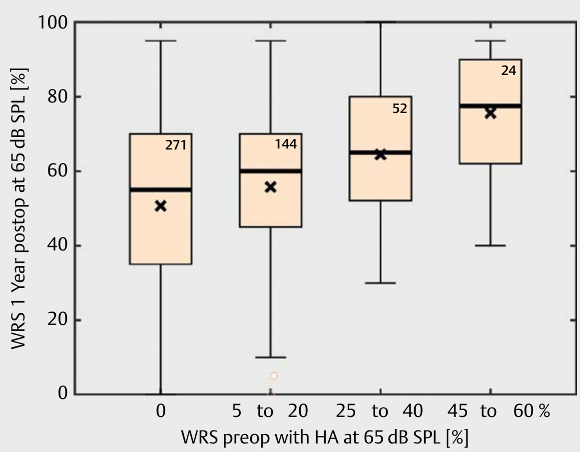
Cochlear implants are generally indicated when other therapies cannot restore hearing sufficiently for audio-verbal communication [22] (▶ **Fig. 4**). Thus, the indication requires the measurement of speech perception as well as its potential improvement by means of appropriate methods like hearing aids. Furthermore, the potential of ear surgery or implantable hearing aids for hearing improvement must be elucidated. The indication also depends on the current technological development of hearing implants (▶ **Fig. 5**). It must be mentioned that the hearing results with artificial electrical stimulation do not reach the level of natural hearing so that these general limits have to be taken into account. Therefore, the wide variability of performance must be considered (▶ **Fig. 12**) caused by different factors such as the functional status of the auditory nerve, cognitive abilities , duration and onset of deafness as well as anatomical factors like malformation of the cochlea. According to the current version of the guideline on cochlear implants

▶ **Table 1** Preoperative diagnostics for CI candidacy evaluation

1. ENT-examination including otoscopy
2. Pure tone audiometry for measurement of hearing threshold and uncomfortable loudness level as well as the dynamic range
3. Speech audiometry in quiet and in noise with and without fitted hearing aid
a) Freiburg speech perception test for numbers and monosyllables b) Sentence test without and with noise, e. g. HSM sentence test c) Matrix sentence test (OLSA test) in noise allowing the comparative assessment of the speech comprehension between different languages
4. Objective audiometry
a) Impedance audiometry with tympanometry and stapedius reflex measurement b) Transitory evoked otoacoustic emissions TEOAE c) Brainstem audiometry (BERA) d) Electrocochleography e) Promontory Test
5. Imaging of the temporal bone, auditory nerve, and auditory pathways
a) High-resolution CT scan or CBCT of the temporal bone b) High-resolution MRI of the temporal bone with imaging of the inner ear and auditory nerve c) MRI of the head for imaging of the auditory pathway d) DTI (Diffusion Tensor Imaging) e) Functional imaging by means of functional MRI (fMRI), positron emission tomography (PET), and near-infrared spectroscopy (NIRS)
6. Genomics for assessment of genetically associated hearing loss
7. Proteomics and metabolomics of the perilymph for differentiation of etiology of hearing loss
8. In pediatric patients additionally:
a) psychoacoustic measurement of the hearing threshold by means of age-related methods (behavioral and play audiometry) b) Frequency-specific BERA c) ASSR (Auditory Steady-State Responses) d) Hearing aid test e) Diagnostics of speech development and communication skills f) Developmental physiological status with psychomotor function, cognition g) Emotional development
9. Vestibular diagnostics for assessment of balance disorders
a) Caloric test b) Head impulse test c) c) VEMP d) Dynamic posturography



▶ **Fig. 11** Diagnostic areas for check of CI candidacy.



▶ **Fig. 12** Correlation between optimum speech perception preop with Master Hearing Aid and CI postop.

▶ **Table 2** Indications and contraindications of cochlear implantation.

Indications:
1. Severe to profound sensory-neural hearing loss or deafness
Postlingual bilateral hearing loss:
Comprehension of monosyllables under best-aided conditions at 65 dB SPL ≤ 60 %
or < 50 % without hearing aid at 80 dB
Prelingual hearing loss in children up to the age of 6 years:
Objectively measured hearing threshold > 70 dB
Missing or insufficient speech development
Perilingual hearing loss (onset of profound hearing loss after birth but before permanent speech acquisition at about 10 years)
Hearing thresholds of > 70 dB
Delayed, stagnating, or regressive speech development
1. Unilateral deafness or asymmetric hearing loss (single-sided deafness, SSD)
1. High-frequency deafness with hearing loss of > 80 dB above 1 KHz and hearing threshold better than 50 dB at 500 Hz and below
1. Auditory synaptopathy and neuropathy:
Missing auditory brainstem responses but present otoacoustic emissions and cochlear microphonics in electrocochleography and present auditory nerve shown by imaging.
Contraindications:
1. Missing cochlea or missing auditory nerve 2. Missing capacity to participate in the overall treatment process, e. g. cognitive impairment 3. Missing infrastructure for CI treatment 4. Negative subjective promontory test 5. Severe comorbidities significantly impairing the rehabilitation process

of the AWMF, the following indications and contraindications are given (▶ **Table 2**):

3.3 Prediction of outcome and individualized cochlear implantation

Depending on the diagnosic results, individualized cochlear implantation aims at providing the best possible hearing rehabilitation for the individual patient. In particular residual hearing, functional condition of the auditory nerve, cochlear anatomy, etiology and pathophysiology of hearing loss are important as well as the potential impact of future therapies and the patients' preference (▶ **Fig. 13**).

3.3.1 Preoperative speech perception under best-aided conditions with Master Hearing Aid

The evaluation of preoperative residual hearing does not only rely on pure tone threshold for air conduction but mainly on speech perception under best aided conditions. A high correlation with the postoperative speech comprehension with CI is found (▶ **Fig. 12**). Most significant in this context is the verification of the preoperative speech perception by means of a so-called master hearing aid, a PC-based hearing aid with optimized sound presentation [12]. The measured value is the optimum speech perception that cannot be reached under real world conditions using conventional hearing aids. In addition, the score is an indirect measure of the cochlear reserve, functionality of the auditory nerve and cognitive capacity of the patient to use the residual hearing for speech perception. The speech comprehension is tested by means of the Oldenburg sentence test (OLSA) or the so-called matrix test with speech simulating noise. The predictive value for postoperative outcome is $R^2 = 0.389$, the most important single prediction parameter identified so far.

3.3.2 Vocabulary Test

The vocabulary test according to Schmidt and Metzler (1992) allows testing the patients' ability to identify the meaningful word out of a series of otherwise five nonsense words. The vocabulary test has a predictive value of $R^2 = 0.158$.

3.3.3 Usable residual hearing for electro-acoustic stimulation

Generally, the decision must be made if electrical stimulation (ES) alone will provide better hearing results for the patient or electro-

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acoustic stimulation (EAS or ENS) (see chapter 3.4). Patients with EAS and ENS achieve significantly better speech perception, particular in noise, compared to patients with electrical stimulation only [23]. The postoperative speech perception score with EAS should be better than the median with ES (median HSM sentence test of 10 dB S/N = 65 percent) (▶ **Fig. 15**).

Precondition for EAS use is preserved postoperative residual hearing at 500 and 250 Hz. Postoperatively, it should be 65 dB or 50 dB, respectively, or better. Based on statistical data of implanted patients the preoperative hearing loss at 500 Hz must not exceed 55 dB and at 250 Hz not more than 40 dB (▶ **Fig. 14**). Furthermore, the patient should have sufficient experience with hearing aids and be motivated to use a hybrid system with a hearing aid . If the patient suffers from chronic external otitis, this treatment option should be not recommended.

With low frequency hearing threshold of 20 dB and better, residual hearing can be effectively used without hearing aid (so-called electro-natural stimulation, ENS).

Children should be also tested for residual hearing especially in low frequencies in order to use it for EAS or ENS. About 20 percent of the children are suitable for this kind of treatment.

3.4 Selection of the electrode system

For optimal stimulation of the auditory nerve, the selection of the most suitable electrode system for ES and EAS is essential and depends on several aspects (▶ **Fig. 16**).

3.4.1 Electrical stimulation

In patients with planned ES, the so-called cochlear coverage (CC) is the important parameter. It is defined as the part of the total cochlear length that is covered by an electrode. Based on preoperative CT/CBCT imaging the total cochlear length can be measured. Different procedures using for example the A and B diameters of the basal turn allow an estimation, more exact procedures use the so-called multiplanar regression, i. e. the cochlea is "unwound" and the total length of the lateral wall is measured (▶ **Fig. 17**). According to the length, the most suitable electrode can be chosen from the electrode portfolio stimulate a high number spiral ganglion cells (SGC) as well as the still present dendrites electrically. Due to the structure of the cochlea, the SGC are mostly located in Rosenthal's canal in the basal and second turn while the dendrites are preserved in areas with residual hearing. In the apical part of the cochlea, they run perpendicular to the respective SGC so that the electrical stimulation might not lead to additional benefit. On the other hand, an electrode that is too short will not stimulate all present SGC and dendrites.

By using straight lateral wall electrodes of different lengths, cochlear coverage (CC) depends mainly on the total length of the cochlea (▶ **Fig. 18**). The cochlear length at the outer wall varies significantly between 31 and 46 mm (▶ **Fig. 16**, [24]). The best speech perception is achieved for a CC between 0.72 and 0.80 (▶ **Fig. 19**), [25]. For lower as well as higher CC, the results are significantly poorer. Therefore electrode length should be selected to reach a coverage between 0,72 and 0,80.. Electrodes of different lengths are available (▶ **Fig. 20**).

3.4.2 Electro-acoustic stimulation

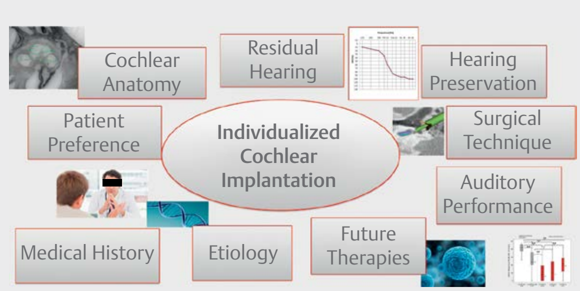
The objective of electro-acoustic stimulation is the combined use of residual acoustic and electrical hearing which restores the high and middle frequencies [26]. The so-called hybrid systems combine the cochlear implant speech processor with an additional hearing aid for the low frequencies (▶ **Fig. 21**).

In general, it is possible to achieve hearing preservation by atraumatic electrode insertion and additional inner ear protection by means of systemic or local cortisone application. Special short electrode systems have been developed with a maximum insertion depth limited to 16 mm. The insertion angle is about 270 ° with an average CC of 0.44. In this way, high frequencies can be functionally restored. Prospective trials could demonstrate high rates of hearing preservation [27, 28]. Hearing preservation in the low frequencies may be classified based on the postoperative threshold changes between 125 and 1,000 Hz:

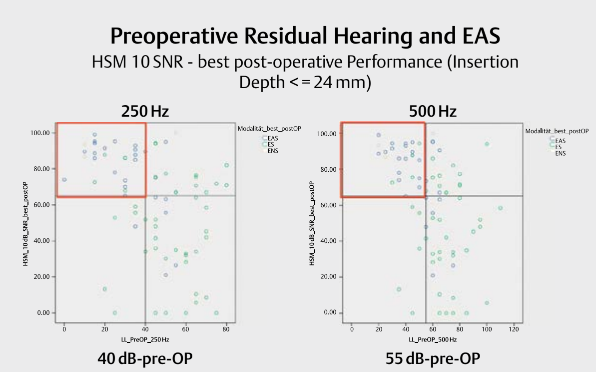
- Good hearing preservation: < 15 dB
- Moderate hearing preservation: 15 to 30 dB
- Complete functional hearing loss: > 30 dB

Using this scale, the following hearing preservation scores were found for short electrodes:

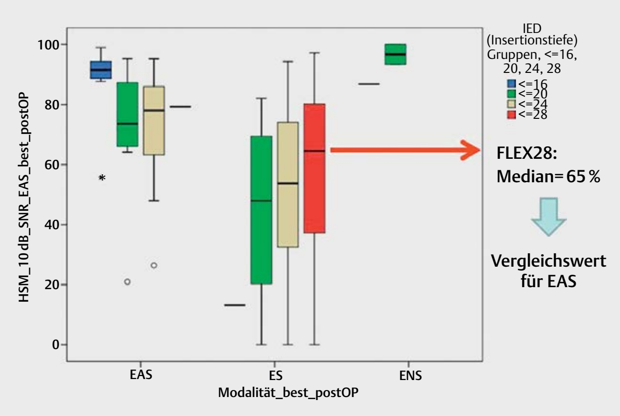
- Good hearing preservation: about 55 %
- Moderate hearing preservation: about 38 %
- Complete hearing loss: 7 %



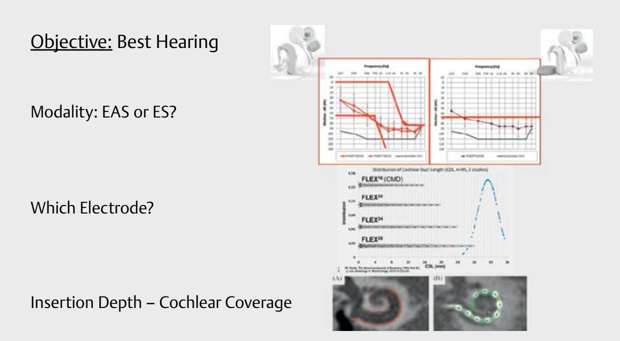
▶ **Fig. 13** Important parameters relevant for individualized cochlear implantation.



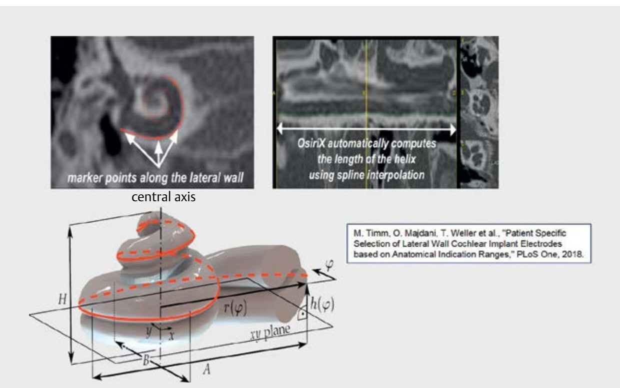
▶ **Fig. 14** EAS-candidacy using preoperative AC threshold at 250 und 500 Hz. Most CI-recipients with postop speech in noise perception of 65 percent and better are in the left upper rectangular with preop AC threshold of 40 dB or better at 250 Hz and 55 dB or better at 500 Hz.



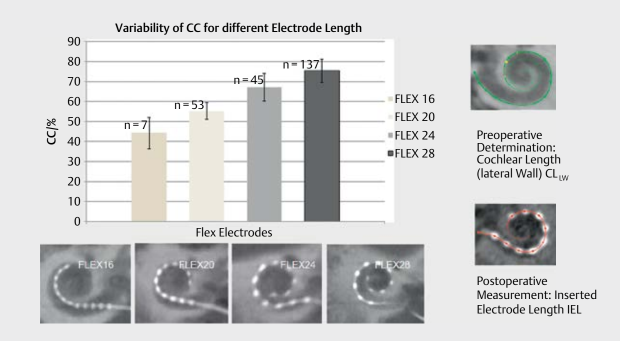
▶ **Fig. 15** Comparison of median speech in noise perception for patients with different stimulation modes. Patients with Electric-acoustic stimulation with additional hearing aid (EAS) perform or natural low frequency hearing (ENS) than those with best possible electric stimulation only ES (65 Prozent).



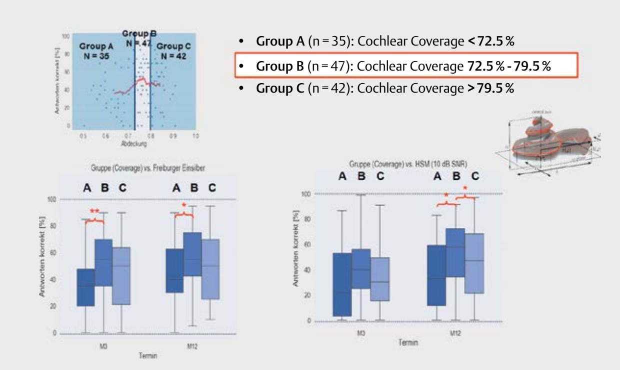
▶ **Fig. 16** Concept of individualized cochlear implantation with type of stimulation ES or EAS, electrode selection with respect to residual hearing and cochlear length/cochlear cocverage.



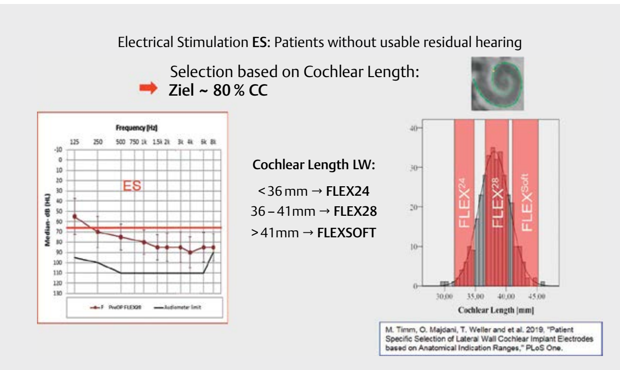
▶ **Fig. 17** Measurement of cochlear length at the lateral wall using multiplanar regression. Cochlear model derived from preop CT or CBCT for virtual surgery to calculate the achievable cochlear coverage.



▶ **Fig. 18** Effect of cochlear length on Cochlear Coverage, using electrodes of different length fully inserted.



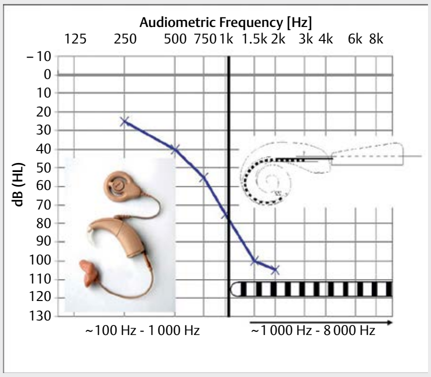
▶ **Fig. 19** Effect of Cochlear Coverage on postoperative speech perception with CI. Best results in group B with a CC between 72 and 80 percent.



▶ **Fig. 20** Individualized electrode selection for best Cochlear Coverage and electric stimulation only ES. Stratification according to electrode length (Timm et al., 2018).

The risk of deafness clearly increases when longer or pre-shaped electrodes are used (▶ **Table 3**) [29, 30].

This aspect may be explained by the anatomy of the cochlea. Along the cochlea, the height of the scala tympani, especially at the lateral wall, decreases continuously beyond an insertion depth



▶ **Fig. 21** Hybrid-System with CI component for electrical stimulation in the high frequency range and hearing aid for acoustic stimulation in the low frequency range (EAS). Crossing point at 75 dB.

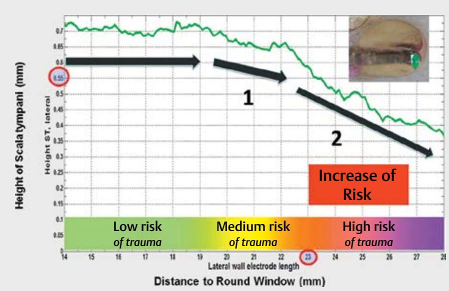
of about 18 mm so that the probability of a mechanical contact of the electrode array with the basilar membrane increases above this point leading to a mismatch between the electrode diameter and the height of the scala tympani in cases of a very flat scala tympani. The individual position of this high-risk zone varies with total cochlear length as well as height of the cochlea. With the imaging methods clinically available today, direct measurement of the height is not possible (▶ **Fig. 22**) [31]. This means that generally a higher risk of damage must be expected beyond an insertion depth of more than 18 mm and substantial damage of cochlear structures may occur with high probability when using longer electrodes. For good hearing preservation, electrode insertion depth should be adapted to this risk zone [32] (▶ **Table. 4**).

However, if the residual hearing decreases or is lost completely during or after cochlear implantation, patients with short insertions achieve clearly poorer hearing results due to the low cochlear coverage compared to patient with longer electrodes and larger cochlear coverage. This results in a trade off between good hearing preservation on one hand and sufficient cochlear coverage on the other hand (▶ **Fig. 23**), [23]. So if a patient experiences postoperative hearing loss after insertion of a short electrode, reimplantation would be necessary with a long electrode of appropriate length in order to achieve the best possible hearing with ES only.

To overcome this trade-off, the concept of the so-called partial insertion has been developed. Hereby, the electrode that was selected according to the total length of the cochlea is only partially inserted in order to have the highest possible chance for hearing preservation at the time of implantation. Some electrode contacts remain intentionally extracochlearly. If hearing loss increases, the electrode may be further advanced into the cochlea during a small intervention (so-called afterloading). This procedure was successfully used in some cases. It was possible to advance the electrode completely into the scala tympani without resistance. By selecting the length appropriate electrode at time of first surgery, sufficient cochlear coverage is achieved after complete electrode insertion to achieve the best possible hearing outcome with electrical stimulation only [33].

▶ **Table 3** Hearing preservation with CI electrodes of different lengths.

Electrode Patients Insertion length/mm	**Ø PTA loss (125..1 kHz)**	**Hearing loss pre – post surgery (125 Hz- 1 kHz) n/percent of patients**		
		≤ 15 dB	≤ 30 dB	Total hearing loss (> 30 dB or exceeds audiometer limit)
Hybrid-L (n = 97) 16 mm	10.0 dB	53 (**54,6 %**)	90 (**92,8 %**)	7 (**7,2 %**)
Nucleus 422 (n = 100) 20 mm	14.2 dB	36 (**36 %**)	83 (**83 %**)	17 (**17 %**)
FLEX20 (n = 46) 20 mm	17.5 dB	21 (**45.6 %**)	12 (**75.7 %**)	8 (**24.3 %**)
FLEX24 (n = 34) 24 mm	20.0 dB	10 (**29.4 %**)	18 (**79.3 %**)	6 (**20.7 %**)
FLEX28 (n = 40) 28 mm	24.0 dB	6 (**15.0 %**)	20 (**65 %**)	14 (**35 %**)
Nucleus 532 (n = 25) 16 mm	24.5 dB	8 (**32 %**)	9 (**68 %**)	8 (**32 %**)



▶ **Fig. 22** Risk zones for cochlear trauma during electrode insertion. Height of scala tympani decreases beyond 18 mm insertion depth with significant increase of risk of damage with electrode insertion (from Afci et al., 2016).

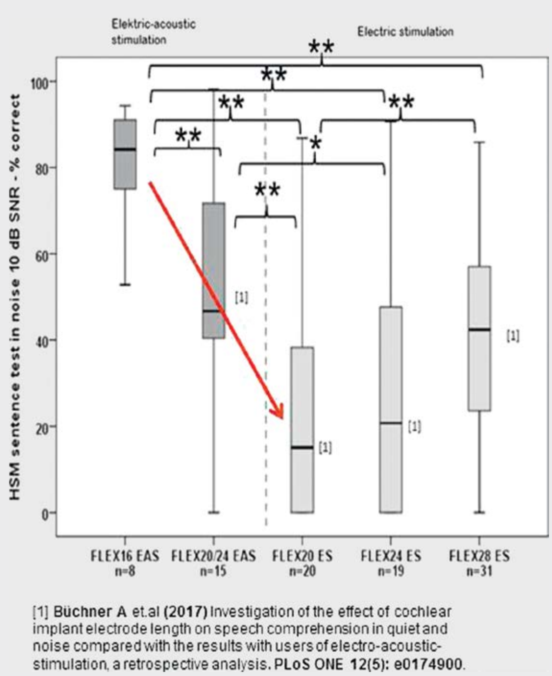
Previous investigations revealed that a low number of intracochlear electrodes is sufficient to restore high-frequency hearing with comparable speech perception to patients with a completely inserted short electrode and a higher number of intra-cochlear electrode contacts.

Performance data after partial insertion and EAS are above average of ES, especially for speech perception in noise (▶ **Fig. 49**).

The required insertion depth for restoration of high and middle frequencies by ES in the area of more than 70 dB hearing loss can be calculated preoperatively based on the following three parameters:

▶ **Table 4** Steps of standardized surgical technique.

1.	Retroauricular incision with creation of a periostal flap and soft tissue pouch
2.	Partial mastoidectomy with exposition of the posterior wall of the auditory canal, the antrum with incus, the facial nerve canal in the mastoid, the sinus-dura angle, and the labyrinthine block
3.	Creation of a bone bed for retromastoid fixation of the implant body
4.	Creation of a connecting tunnel or canal to the mastoid
5.	Posterior tympanotomy for exposition of the middle ear with promontory, incudo-stapedial joint, stapedius tendon, and round window niche
6.	Preparation of the round window membrane with removal of the bone overhang, if needed, to completely visualize the membrane
7.	Insertion of the implant in the prepared bone bed and positioning of the electrode array in the mastoid
8.	Incision of the round window membrane, if needed enlarged round window approach by drilling in anterior-inferior direction
9.	Atraumatic and slow insertion of the electrode into the scala tympani by using electrode-specific insertion techniques, if needed with application of cochlear monitoring for hearing preservation
10.	Cochlea-near fixation of the electrode to avoid electrode migration
11.	Intraoperative electrophysiology for control of the implant function and measurement of the neural responses
12.	Wound closure in several layers to securely cover the implant
13.	Intraoperative imaging to verify the electrode position



▶ **Fig. 23** Comparison of speech in noise understanding for EAS patients using a short electrode versus ES with longer electrodes. In case of loss of residual hearing patients with short EAS electrodes have significantly poorer scores for ES than patients with longer electrodes (from Büchner et al., 2017).

1. usable residual hearing
2. total length of the cochlea
3. predicted postoperative hearing threshold in low frequencies

The location of a single frequency along the basilar membrane is given by the so-called Greenwood function and respects the individual length of the cochlea. (▶ **Fig. 24**). The electrode is inserted virtually into the cochlea, the electrode tip is placed at the site of the transition frequency between electrical and acoustic hearing (70 dB hearing loss, the limit for useful acoustic amplification) using the predicted postoperative hearing threshold (▶ **Fig. 25**).

4. CI implantation

4.1 Standard surgical technique

During the last decades, a standard surgical technique could be established that includes the following steps [34], (▶ **Table 4**).

This standardized surgical technique can be safely applied with low complication rates for all implants and in all patients of different ages as well as all anatomical situations. It may be easily modified for specific situations. A standardized surgical technique is also a precondition for an effective quality management. Complications can be detected early, their origin may be identified, and they can be corrected by means of appropriate measures. Based on the available experience and with regard to minimizing the complication rate, the single steps are based on long term clinical experience. Alternative procedures like the suprameatal or transmeatal approach did not prevail [35].

1. Retroauricular incision and preparation of soft tissue

The retroauricular incision is performed in the length of the auricle one centimeter behind the retroauricular fold to provide an overview of the mastoid and the temporalis muscle. After exposure of the external auditory canal, a periostal flap is created that is pediciled at the external meatus and serves for safe covering of the mastoid and the implant. The objective is a two-layer coverage with soft tissue in order to avoid infection of the implant in cases of wound healing problems. The incision line does not cross the implant. A soft tissue pouch is created in occipital direction by further lifting the periosteum and the temporalis muscle. In this way, a safe soft tissue coverage of the implant with good blood supply especially over the receiver-stimulator is achieved.

2. Mastoidectomy (▶ **Fig. 26**)

Partial mastoidectomy is performed if mucosa is not diseased. In cases of chronic inflammation, the entire inflammatory tissue has to be removed. It is important to expose the anatomical landmarks. Those are the posterior wall of the auditory canal, the antrum with the incus for safe exposure of the fossa incudis, the mastoid canal of the facial nerve, and the canal of the chorda tympani for precise definition of the area for posterior tympanotomy, the sigmoid sinus, the labyrinthine block with the 3 semicircular canals, the cortical bone to the middle and posterior cranial fossa as well as the sinus-dura angle. Hereby, a cortical bone overhang remains superior, posterior, and inferior for safe positioning of the electrode array in the mastoid avoiding the direct contact with the covering soft tissue.

3. Creation of a bone bed (▶ **Fig. 26**)

The bone bed has to be sufficiently deep and even. It serves for safe fixation of the implant to avoid migration and protrusion with con-

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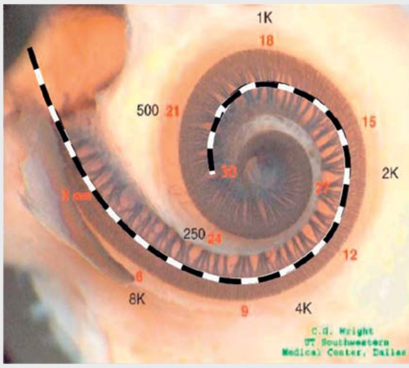
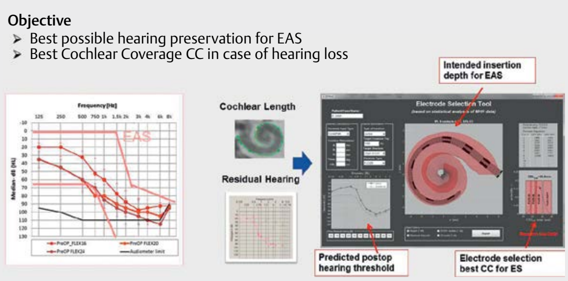


▶ **Fig. 24** Greenwood-function to localize frequency representation along the basilar membrane of the human cochlea (Greenwood 1961, 1990). (permission by MEDEL).



▶ **Fig. 25** Individualized Cochlear Implantation with partial insertion for EAS. Electrode selection and calculation of intended insertion depth according to preop AC threshold and cochlear length.

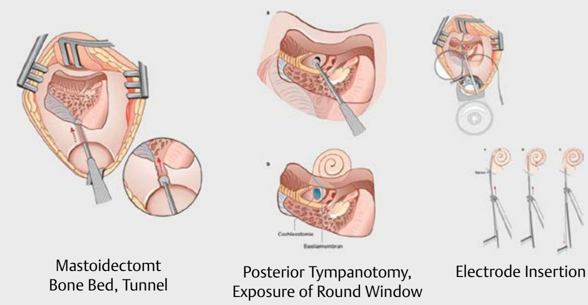
secutive problems of the covering skin. It should be created at 1 cm behind and above the sinus-dura angle. In this way, a sufficient distance to the auricle is kept for the transmission coil and conflict with eyeglass temples are avoided. In cases of implants with integrated magnet, the implant bed has to be positioned more posteriorly.

4. Tunnel or channel to the mastoid (▶ **Fig. 26**)

From the bone bed, a connecting canal or tunnel is created towards the sinus-dura angle. Here, the bone is thick enough, also in infants and small children. Exposure of the dura might be necessary. This connection is crucial for the protection of the electrode array. The tunnel provides additional fixation to avoid migration of the implant inferiorly and anteriorly. If performed correctly the implant body does neither protrude nor rock, which could otherwise cause significant problems of the skin including severe pain.

5. Posterior tympanotomy (▶ **Fig. 26**)

It is important to clearly expose the canal of the facial nerve in the mastoid from the level of the fossa incudis down to the branching of the chorda tympani which has to be identified in its bony canal. Posterior tympanotomy can be securely performed in the triangle between facial nerve, chorda tympani and the fossa incudis with



▶ **Fig. 26** Standardized surgical technique for Cochlear Implantation. Essential steps (permission by Endo-Press, Tuttlingen).

preservation of the bridge as cranial border. This allows a safe and sufficient approach to the middle ear with exposure of the incudostapedial joint, the stapedius tendon, and the promontory with the round window niche. At the inferior edge of the facial recess, the bone should be preserved to create a bone slit for secure fixation of the electrode array. The bone slit avoids migration especially of straight electrodes out of the cochlea. Alternatively metal clips may be used.For this step of the implantation, facial nerve monitoring is recommended.

6. Preparation of the round window membrane (▶ **Fig. 26**)

It is necessary to remove the variable bone overhang for complete exposure of the entire round window membrane. Contact of the drill with the membrane must be avoided. An anterior-inferior extension of the round window is indicated when the round window is too narrow for safe insertion of thicker electrode systems or when obliteration is found. Newly built tissue and bone that can be differentiated from labyrinthine bone due to its white color must be removed from the scala tympani with preservation of the cochlear structures of the basal turn, especially the modiolar wall and the basilar membrane.

7. Positioning of the implant (▶ **Fig. 26**)

The implant is securely inserted in the pre-shaped bone bed, the electrode is advanced into the mastoid and the posterior part positioned in the occipital pouch with an even position to avoid protrusion.

8. Incision of the round window membrane

Opening of the inner ear is performed by a sufficiently wide incision of the round window membrane so that the electrode array may be inserted in an atraumatic way and perilymph drains to avoid intracochlear pressure increase.

9. Insertion of the electrode (▶ **Fig. 26** and ▶ **27**)

The insertion of the electrode is performed slowly by means of special insertion forceps or pincers. An insertion duration of about 1–3 minutes should be kept, if needed with intermitted stops for cochlear monitoring. The insertion may be supported by robotic systems like Iota Motion [36], Robotol [37], or RoboJig [38].

10. Electrode fixation (▶ **Fig. 27**)

At the end of the insertion procedure, the electrode may be securely fixed in the created bone slit in order to avoid postoperative electrode migration. Also clips and tubes may be used. Afterwards, the electrode cable is positioned in the mastoid cavity so that contact with the overlying skin is avoided. Closure of the inner ear is performed with fresh venous blood, muscle, or connective tissue.

11. Intraoperative electrophysiology

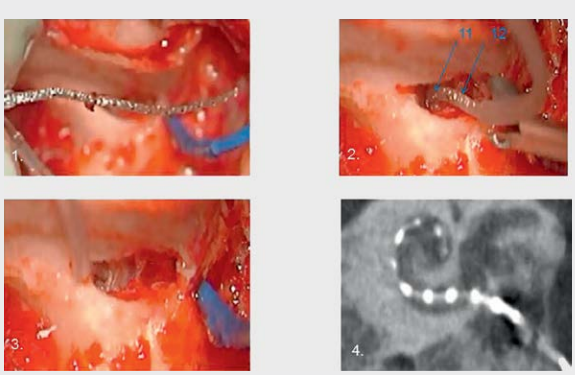
The function of the implant is controlled intraoperatively in order to detect possible damage of the implant already during insertion. The measurement of neural responses allows indirect control of the position of the electrode and allow preliminary estimation of the stimuli for fitting of the speech processor especially in young children. Essentially, the electrically elicited stapedius reflex with direct observation and determination of the reflex threshold as well as electrically evoked compound action potential of the hearing nerve (so-called NRT, NRI, or ART measurements) are used. They also give information about the correct position of the electrode, especially when using multifrequency impedance measurement (TIM Trans impedance measurement) (s. 2.1.1).

12. Wound closure

The closure should be performed in several tissue layers in order to avoid propagation of infections to the implant due to wound healing disorders. Periosteum and muscle form the inner layer and serve for covering the implant and the mastoid. Subcutaneous tissue and skin are two further separate closure layers. In this way, the rate of postoperative infections has been reduced significantly.

13. Intraoperative imaging for position control of the electrode (▶ **Fig. 27**)

The intraoperative imaging is essential in order to detect malposition of the electrode and then to correct it immediately during surgery. This is especially important in cases of difficult electrode insertion like malformations, obliteration, and reimplantation.



▶ **Fig. 27** Surgical technique for partial insertion. 1. Connective tissue marker for precalculated insertion depth on electrode 2. Atraumatic slow electrode insertion. Stop of insertion, when marker is positioned at the round window membrane. 3. Fixation of electrode in bone slit at the facial recess. 4. Control of electrode position with intraoperative CBCT

4.2 Cochlear implant surgery with hearing preservation

Preservation of hearing can generally be achieved by using an atraumatic surgical technique. In addition, the so-called cochlear monitoring can be applied (see chapter 2.1.1). The responses of the hair cells of the inner ear evoked by acoustic stimulation (cochlear microphonics CM) allow an online monitoring of residual hearing (▶ **Fig. 28** and ▶ **29**) [39, 40]. If the amplitudes decrease during advancement of the electrode, in particular in the area of the risk zone beyond 18 mm insertion depth, an interaction with cochlear mechanics by the inserted electrode may be expected. Further advancing the electrode may lead to structural damage of the basilar membrane or other cochlear structures which means a significant hearing loss up to deafness. If the insertion is stopped at the onset of amplitude decrease and the electrode is pulled back or the direction of insertion is changed, the response amplitude may recover. In this way, the maximum insertion depth for hearing preservation can be determined [5, 41, 42]. There is a good correlation between intraoperative changes and postoperative hearing (▶ **Fig. 30**). Beside amplitude variations, other parameters such as phase shifts or multi-tone measurements are applied in order to differentiate between damage-specific changes of the amplitude and physiological changes by phase shifts of the response while passing the frequency specific generator site along the basilar membrane [42]. In addition, the insertion may be performed under simultaneous fluoroscopy. The surgeons retrieves information about the position and the behavior of the electrode during insertion and can react and correct very early for example a so-called tip fold-over (▶ **Fig. 42**). This aspect is particularly important with the use of pre-shaped electrodes.

4.3 Cochlear implantation under local anesthesia

Cochlear implantation may be performed under local anesthesia, if needed with support of anesthesiologists, as an analgosedation procedure. It follows the proven principles of ear surgery under local anesthesia [43]. The patients have the following advantages:

- No risk of general anesthesia
- Rapid recovery
- Intraoperative control of the residual hearing by immediate response of the patient about changes of simultaneously presented acoustic stimuli, e. g. a low-frequency permanent tone for testing the residual low-frequency hearing

▶ **Fig. 28** Intraoperative Cochlear Monitoring. ECochG through CI. Insert ear phone for acoustic stimulation in external auditory canal . Real-time monitoring of residual hearing through intracochlear recording of Cochlear Microphonics CM.

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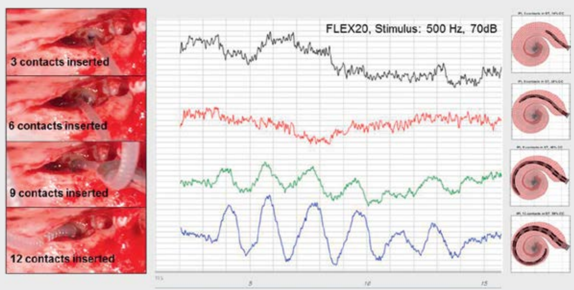
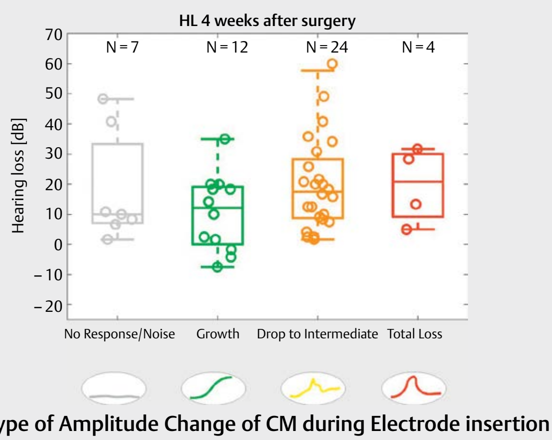


▶ **Fig. 29** CM recording during electrode insertion using a 500 Hz tone as acoustic stimulus. Increase of CM amplitude measured with the most apical electrode contact during electrode insertion. Probable hearing preservation.



▶ **Fig. 30** Correlation of type of intraoperative CM changes and postoperative hearing. Total loss of CM is a sign of severe cochlear damage with high probability of hearing loss. .

- Immediate information about vertigo as sign for an insertion trauma with the option to modify the electrode insertion
- Control of electrode position by intraoperative activation of the implant and stimulation of different electrode contacts to elicit different pitches and determine T and C levels for immediate fitting of the speech processor

Surgery under local anesthesia becomes more important, especially for older and old patients and simplifies the surgical procedure [44].

4.4 Computer-assisted and robotic surgery

The limitations of conventional cochlear implant surgery result from the fact that the surgeon has no direct visual control over the electrode insertion beyond the round window, respectively cochleostomy. The insertion trajectory of the electrode cannot be adjusted to the course of the scala tympani at the beginning of the basal turn. Even the rotation of the electrode cannot be controlled within the cochlea. Further limitations of the manual electrode insertion result from the minimally possible speed for an even and steady insertion of the electrode [45]. Experimental investigations show that insertion forces may be significantly reduced by (ultra) low insertion speed below the limit of manual insertion which leads to a reduction of the risk for damages of cochlear structures (▶ **Fig. 31**) [46]. Therefore, slow and steady insertion procedures should be developed that cannot be performed manually.

In order to achieve a higher surgical precision, computer- and robot-assisted surgery procedures have been or are currently being developed [38, 47, 48]. The basic process is as follows (▶ **Fig. 32**):

A preoperative CT or CBCT dataset is used for segmentation of the relevant anatomical structures of the temporal bone. Taking into account the residual hearing that may be preserved, the insertion depth of the electrode is defined. A cochlear model is computed for virtual cochlear implantation and anatomically feasible trajectories for optimal electrode insertion can be defined. These trajectories can be transmitted to the surgery site by means of a navigation system and be used for drilling and electrode insertion (▶ **Fig. 33**).

However, navigation-based procedures lack the necessary accuracy of planning if no bone-anchored markers are used. Furthermore, the application of trajectories causes problems with electrode insertion because the insertion tools cannot be sufficiently referenced and manually guided.

Robotic systems have been developed for precise implementation of the planned trajectories into a minimally invasive surgical procedure for an accurate electrode insertion. A pre-calculated drilling canal can be created from the mastoid surface into the cochlea with simultaneous temperature control and facial nerve monitoring. The accuracy requested with an overall deviation of < 0.3 mm may be achieved by applying high-resolution CT scans, bone-anchored marker systems for navigation-based procedures and use of rigid fixation systems (Mayfield clamp) for connecting the patient site with the robotic system. Furthermore, drilling jigs are applied that are individually manufactured intraoperatively. The drill as well as the insertion tool can be guided exactly in the pre-calculated trajectory. By means of a mini-stereotactic frame that is rigidly fixed at the patient’s head already before data acquisition, the jig with drilled trajectory is hold in the predefined position. The drill path is created from the mastoid directly to the cochlea (▶ **Fig. 34**).

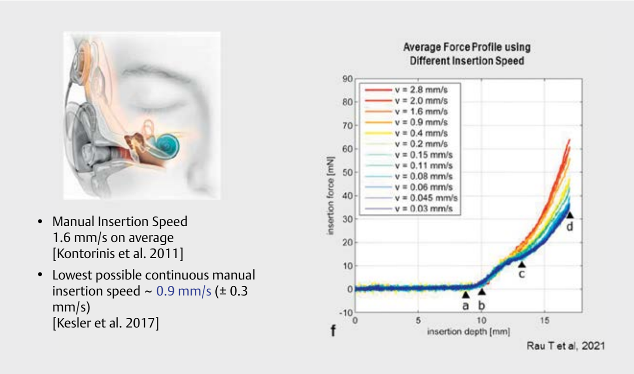
After drilling and opening of the cochlea, the electrode may be inserted manually or motorized by an insertion robot (Robotol) as slowly as necessary through the drilling canal into the cochlea (▶ **Fig. 35**), [37]. Integrated force sensors allow a haptic control. A simplified system for very slow electrode insertion has been described by Rau et al. (▶ **Fig. 35**), [49].

4.5 Surgery – special cases

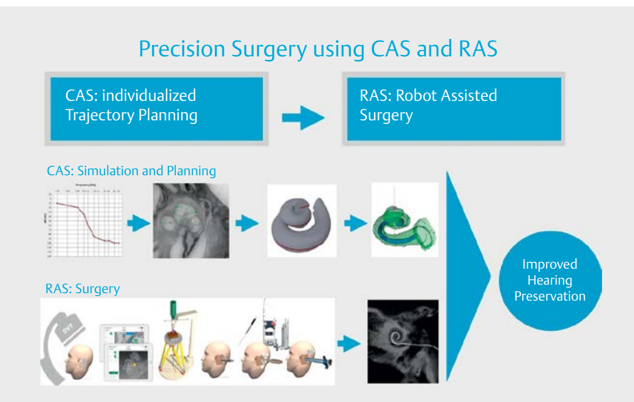
4.5.1 Chronic otitis media

The different types of chronic otitis media require procedures that are adapted to the disease process. The following types must be differentiated:

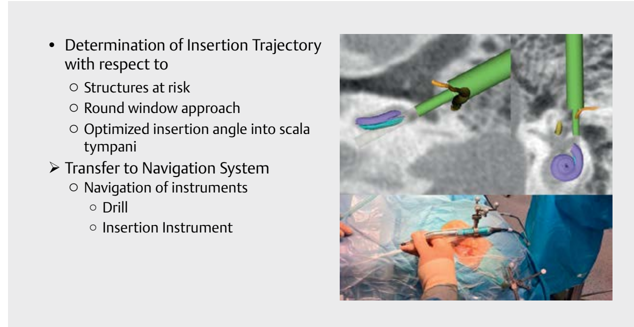
- serous or mucous otitis media; so-called sero- or mucotympanum
- chronic otitis media without cholesteatoma
- chronic otitis media with cholesteatoma
- Radical cavity



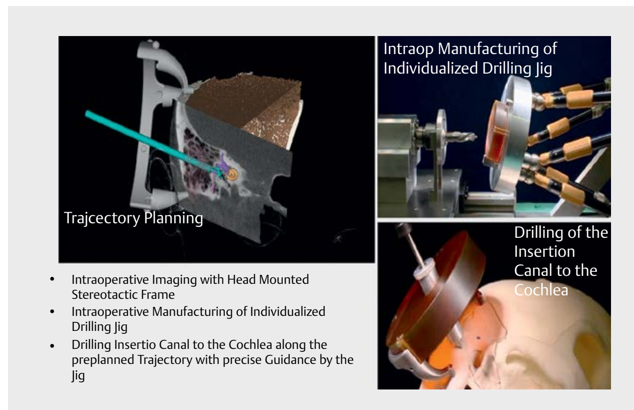
▶ **Fig. 31** Insertion forces during manual and robot assisted slow electrode insertion. Slow steady insertion reduces insertion forces significantly and avoids intermittend force peaks (Rau et al., 2021).



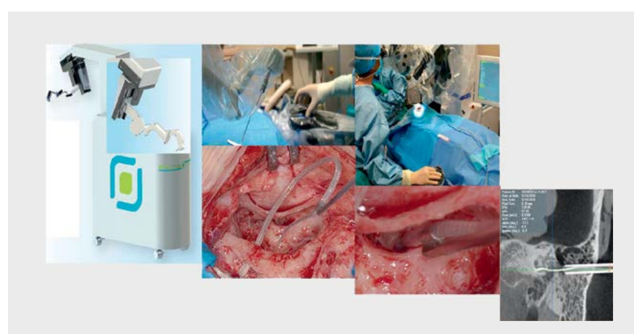
▶ **Fig. 32** Principle of Computer and roboter assisted precision surgery to improve hearing preservation



▶ **Fig. 33** Navigation based Cochlear Implantation using precalculated trajectory for optimized anatomy based electrode insertion. Navigation helps to visualize the insertion for the surgeon.



▶ **Fig. 34** Robot assisted Cochlear Implantation.



▶ **Fig. 35** Otosurgery robot for motorized electrode insertion (Robotol®, Fa. Collin).

In general, serous and mucous otitis media are cases of bacteria contaminated middle ear effusions that should be treated before cochlear implantation also from an audiological point of view in the context of CI diagnostics [50].

CI surgery may be performed in cases without infection even with recurrent OME in order to avoid a delay in the onset of hearing rehabilitation in children.

In cases of chronic otitis media tympanoplasty should be performed prior to implantation to eradicate the chronic inflammation. The tympanic membrane must be enforced by underlying cartilage to avoid retraction pouches with risk of cholesteatoma formation.

Depending on the severity of the findings, cochlear implantation may be performed simultaneously or as a staged procedure.

In cases with a radical cavity or if preservation of the posterior wall of the auditory canal is not possible a subtotal petrosectomy with blind sac closure of the auditory canal and Eustachian tube and fat obliteration of the cavity should be performed. In general, this leads to eradication of the inflammatory process and reduces the risk of implant loss due to infection or inflammatory reactions. Cochlear implantation can be safely done 6 months later. A single stage procedure is not recommended [51, 52].

4.5.2 Malformations

Malformations of the cochlea represent a special challenge for preoperative diagnostics, the surgical concept, the intraoperative management, and the postoperative fitting. According to Sennaroglu [53, 54] they may be classified into the following subtypes with increasing deviation from normal anatomy (▶ **Table 5**).

High-resolution imaging using CT or CBCT and MRI of the temporal bone, the auditory nerve and the auditory pathways with the use of surface coils is important. Surgery should only be performed by experienced surgeons, facial nerve monitoring is imperative.

The surgical concept depends on the type of the malformation with special respect to the position of the auditory nerve fibers as well as the possible access to the cochlea. Management of gusher and CSF leak must be known [55].

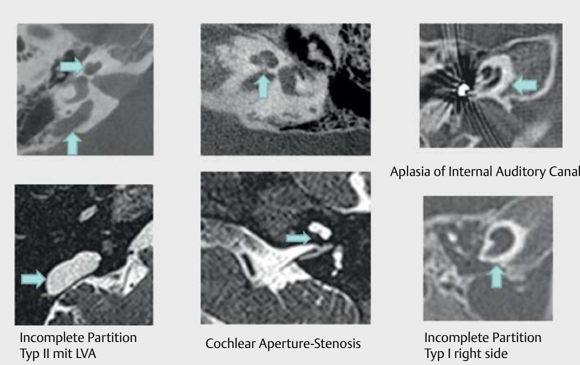
1. Incomplete partition type II (▶ **Fig. 36**). The cochlea is shorter than normal with 1.5–2 turns. The selection of the electrode follows the criteria for normal cochlea In general, gusher (uncontrolled release of perilymph due to unnatural connection to the CSF space) does not occur.
2. Incomplete partition type III. The cochlea has an abnormal wide cochlear aperture due to a missing bone floor of the basal turn. In this context, the use of a pre-shaped electrode is recommended to avoid malposition in the internal auditory meatus. Electrode insertion should be done using intraoperative fluoroscopy for a safe intracochlear positioning of the electrode. It may be inserted through the round window or in case of its absence through a cochleostomy adapted to the diameter of the electrode. Generally, gusher occurs that can be stopped by inserting small pieces of muscle or connective tissue into the cochleostomy. Only in very rare cases CSF drainage is required.
3. Incomplete partition type I (▶ **Fig. 36**). The cochlea has no internal structure and misses a modiolus. Since the position of the hearing nerve fibers is not known and/or a modiolus is not present, straight electrodes with ring contact should be inserted, e. g. Nucleus Straight Electrode, because they lie at the outer wall of the cochlea and stimulate the auditory nerve fibers located either there or in the cochlea. The position is also controlled by means of intraoperative fluoroscopy in order to detect malpositioning in the internal auditory meatus with need for repositioning (▶ **Fig. 36 and 37**). In general, a cochleostomy adapted to the diameter of the electrode is necessary. The treatment of gusher is performed as described above.
4. Common cavity (▶ **Fig. 37**). There is only one common structure for the cochlea and the vestibular organ. Since the modiolus is absent, a straight electrode with ring electrodes should be inserted. The opening of the cochlea should be kept as small as possible in order to safely manage the gusher that usually occurs by closing the opening with small pieces of connective tissue around the electrode. The electrode insertion should be performed under fluoroscopy. If malposition of the electrode in the internal auditory canal is found, a special loop technique can be used. A slit is drilled in the cochlea and the electrode is inserted as loop while the electrode tip is held outside the cochlea (▶ **Fig. 37**).
5. Cochlear aplasia with present vestibular structure (▶ **Fig. 37**). The procedure is performed as described for common cavity.
6. Complete absence of the inner ear. This is a contraindication for cochlear implantation, an auditory brainstem implant may be considered.
7. Vestibular malformations. Various types with involvement of the semicircular canals or the vestibulum are found. The severity of hearing loss is very different. Cochlear implantation follows the principles for normal anatomy of the cochlea.
8. Cochlear aperture stenosis (▶ **Fig. 36**). The bone canal from the modiolus into the internal auditory meatus is narrowed or missing with either a hypoplastic or aplastic auditory nerve. In general, these findings require the intraoperative thorough measurement of auditory nerve responses, for example by using a test electrode (ANTS electrode) with measurement of ECAPs and EABR before cochlear implantation.
9. Narrow internal auditory meatus (▶ **Fig. 37**) Procedure as for cochlear aperture stenosis.
10. Missing internal auditory canal.

This is a contraindication for cochlear implantation, an auditory brainstem implant may be considered.

Intraoperatively, different methods for identification of auditory nerve responses (ESRT, ECAPs, EABR, see above) must be applied,

▶ **Table 5** Inner ear malformations with relevance for cochlear implantation**. Malformations with predominantly cochlear involvement**

1.	Short cochlea, so-called incomplete partition type II or Mondini dysplasia (▶ **Fig. 36**), often associated with large-vestibular-aqueduct (LVA)
2.	Cochlea with missing limitation to the internal auditory canal, so-called incomplete partition type III or x-linked deafness
3.	Cochlear malformation without development of the modiolus and intracochlear structures, so-called incomplete partition type I (▶ **Fig. 36**)
4.	Common Cavity (▶ **Fig. 37**) without differentiation of cochlear and vestibular parts
5.	Cochlear aplasia (▶ **Fig. 37**) with development of only the vestibular part
6.	Cochlear aplasia with completely missing inner ear
Malformations with predominantly vestibular involvement	
1.	Vestibular malformations of different degree
Malformations of the auditory nerve	
1.	Cochlear aperture stenosis with narrow bone canal from the modiolus to the internal auditory meatus (▶ **Fig. 36**)
2.	Narrow internal auditory canal (▶ **Fig. 37**)
3.	Missing internal auditory canal (▶ **Fig. 36**)



▶ **Fig. 36** CI and malformations oft he inner ear 1.

including the use of test electrodes. The results are essential for the postoperative fitting especially in children. If responses cannot be measured, the decision must be made for every single case if cochlear implantation shall be tried. If no postoperative hearing reactions can be observed and hearing/speech development are missing, the only alternative is the auditory brainstem implant (ABI) [56].

Outcomes of cochlear implantation in cases of malformations

Auditory performance and development of speech and language depend on the type of malformation and of the associated additional handicaps (syndromic hearing loss). In cases of short cochlea, normal results may be achieved. Sometimes, the short cochlea is also associated with a large-vestibular-aqueduct syndrome (LVAS) (▶ **Fig. 36**) often characterized by a progressive hearing loss with sudden deterioration especially after head trauma. The perilingual hearing loss provides a favorable precondition for good auditory performance.

In cases of severe types of malformation, e. g. common cavity, the results are usually poorer than the average of children with normal anatomy [57].

4.5.3 Obliteration/ossification (▶ **Fig. 38**)

Obliteration and ossification may be induced by different underlying diseases, e. g. after trauma, meningitis, otosclerosis, chronic inflammatory diseases with involvement of the inner ear like Wegener's disease (granulomatosis with polyangiitis), Cogan syndrome or chronic otitis media. The staged inflammatory process is initiated by granulation tissue formation followed by connective tissue and ossification. Therefore, preoperative imaging with high-resolution CT scan or CBCT as well as magnet resonance imaging with contrast enhancement are crucial for the planning and the surgical procedure in order to identify the severity, degree, and stage of obliteration. A strong contrast enhancement is found in initial stages of the inflammatory process which is reduced in later stages. The bone remodeling processes of the cochlear capsule is pathognomonic for otosclerosis [58].

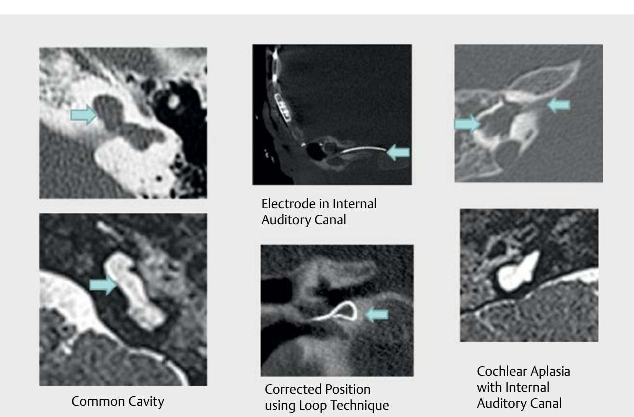
Obliteration mostly starts in the area of the round window and propagates towards apical direction, however, also different other obliteration sites may be observed, e. g. in cases of post-meningitic hearing loss.

In partial obliteration (▶ **Fig. 38**), the surgical concept aims to remove the new built tissue until the open lumen of the scala tympani or the scala vestibuli apically to the site of obliteration is reached. Prior to electrode insertion, a more rigid electrode dummy can be inserted, so-called stiff probe, which allows passing smaller obliteration sites and widens the cochlea. If obliteration reaches beyond the straight part of the basal turn, a second cochleostomy in the area of the second turn is required. It is performed anterior to the stapes, inferior of the lenticular process of the tensor tympani muscle. Generally, the lumen of the second turn is reached and the first turn can be approached in a retrograde direction [59]. Both drilled canals can be joined and the electrode inserted along the basal to the second turn. If the connection of both drilled canals is not possible, a split array with two electrode arrays can be implanted with one positioned in the basal turn and the other in the second turn (▶ **Fig. 38**).

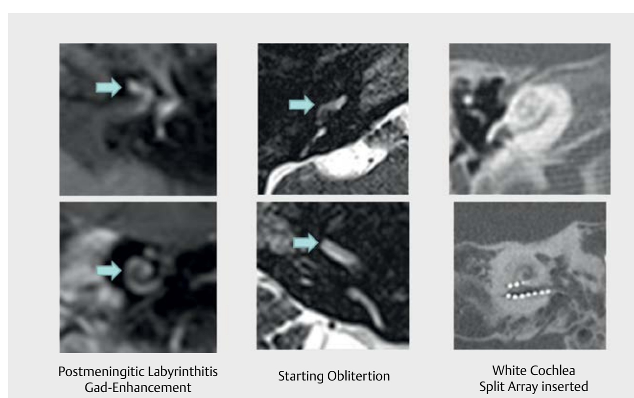
Overall, the hearing results are significantly poorer compared to cases with regularly inserted electrodes because the auditory nerve is only partially stimulated, the electrical field spread is changed due to ossification and in addition a damage or loss of spiral ganglion cells can be present due to the underlying disease. In cases of partial obliteration and completely inserted electrode there may be no differences to normal anatomy cochlear implant cases.

4.5.4 Cochlear implant and facial nerve stimulation

Sometimes an increased risk of facial nerve stimulation after cochlear implantation is found with certain diseases. The incidence amounts to 1–14 % [60]. These diseases include for example advanced otosclerosis, temporal bone fractures, malformations, or other types of bone remodeling processes. Severe facial nerve stimulation may end up in a non-use of the implant. In general, switching off single electrodes, reduction of the stimulation level, or modification of the stimulus paradigm, for example tripolar stimulation may solve the problem. If these measures fail or even lead to a significant decrement in performance, reimplantation with another cochlear implant system may be discussed. A shift from a lateral wall to a perimodiolar electrode may be successful [61]. It could be shown recently that use of a different stimulation strategy could eliminate the facial nerve stimulation also in severe cases and thus lead to clearly



▶ **Fig. 37** CI and malformations of the inner ear 2.



▶ **Fig. 38** Obliteration of the Cochlea. Different phases. Split Array-Electrode in case of total obliteration inserted.

improved hearing. The different stimulation mode uses so-called pseudo-monophasic stimulation and a combined common ground electrode. Both factors lead to a reduced spread of excitation. The amplitude of the cathodic phase of the biphasic electrical stimulus which is responsible for facial nerve stimulation is significantly reduced [62].

4.5.5 CI in cases of vestibular schwannoma (▶ **Fig. 39**)

If the hearing nerve is functionally intact and the cochlea suitable for implantation, a CI can generally be implanted for hearing rehabilitation.

The following constellations must be differentiated:

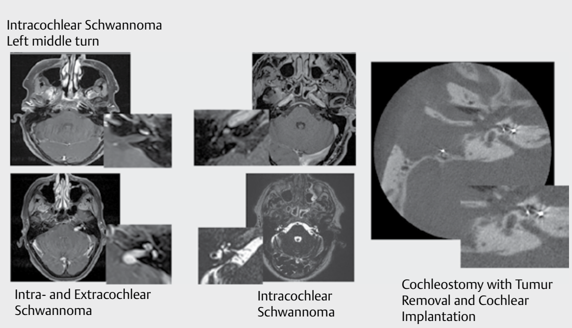
- Intracochlear schwannoma with progressive severe hearing loss
- Extracochlear schwannoma without previous treatment
- Extracochlear schwannoma after radiotherapy
- Condition after surgical removal of a vestibular schwannoma with consecutive hearing loss
- Bilateral schwannomas in neurofibromatosis type 2 (NF 2)

In the first case, the schwannoma leads to hearing loss without impairment of the function of the auditory nerve. The tumor may be removed through the round window or cochleostomy and the CI electrode may be inserted [63].

An untreated extracochlear vestibular schwannoma leads to hearing loss by impairment of the cochlear blood supply and to damage of the nerve fibers which increases with tumor growth. Depending on these two mechanisms, the function of the auditory nerve can be more or less impaired, even in a progressive way. Thus, the results vary in the inter- as well as intraindividual course. An initially good hearing outcome may significantly worsen over time. Preconditions for cochlear implantation are a positive promontory test, completed by intracochlear test stimulation, if possible.

The same applies for cases after radiotherapy.

Cases after microsurgical tumor resection with an anatomically preserved auditory nerve require verification of its functional integrity by promontory test and intracochlear electrical stimulation using test electrodes. If the nerve is intact, CI surgery may be performed with permanently stable hearing results (▶ **Table 6**). Typically, the results after microsurgery are better than after radiotherapy. The poorest outcome is observed in patients with neurofibromatosis type 2, probably due to the aggressive infiltrative tumor growth into the auditory nerve. In these cases, auditory rehabilitation is only possible with central auditory implants (auditory brainstem implant [ABI], auditory midbrain implant [AMI]) [56, 64].



▶ **Fig. 39** Vestibular Schwannoma and CI

4.6 Cochlear implantation in children

Specific quality requirements must be fullfilled [65, 66]. In general, organs like the lung and the cardiovascular system are immature, they are more prone to hypothermia and the children often have additional impairments. The main focus is on the safety of the child in order to avoid severe complications. The elective intervention should be planned and performed in collaboration with experienced pediatric anesthesiologists. In general, the intervention may be performed from the 7th month of life without increased risk regarding anesthesia or surgery. In cases of postmeningitic deafness, surgery may be performed at an earlier age if signs of beginning obliteration are found, taking into consideration the general status of the patient. Intraoperatively, thorough hemostasis must be performed in order to minimize the blood loss regarding the low total blood quantity in children. If necessary, sequential implantation must be performed if the blood loss is relevant for circulation, instead of planned simultaneous bilateral surgery.

Factors relevant for cochlear implantation are the size of the head, acute and chronic otitis media in its different types, thin calvaria as well as thin soft tissue. The objective is a long-term stable implant position with a low complication rate. In the context of age-related frequent middle ear diseases including otitis media, sealing of the inner ear is important to avoid labyrinthitis. Furthermore, it is important to completely drill out the mastoid to avoid potential mastoiditis. Due to its small size, it may be difficult to position the electrode array cable in the drilled-out mastoid. A tension-free position is crucial to avoid postoperative migration of the electrode caused by skull growth. It is also important to safely position the implant in a carefully created bone bed even with exposure of dura as well as the protection of the electrode by a connection tunnel or canal to the mastoid. The electrode fixation near the cochlear is also decisive to avoid electrode migration.

The complication rate amounts to about 10 % with 5 % of severe complications, often requiring surgical intervention. Most of them occur as late complications [66, 67].

4.7 Reimplantation

With a growing number of implanted patients, the number of reimplantations will also increase. They are indicated mainly for 3 reasons:

1. Technical defect of the implant

▶ **Table 6** Hearing results with CI in cases of vestibular schwannoma.

	n	Monosyllables %	HSM in quiet %	HSM S/N 10 dB %
Wait and Scan	44	55 (0–65)	59 (0–75)	25 (0–45)
Radiotherapy	19	33 (0–55)	30 (0–55)	12 (0–35)
Microsurgery	53	67 (0–85)	74 (5–85)	27 (0–55)
NF 2	11	21 (0–55)	27 (0–35)	5 (0–10)

2. Medical complications
3. Upgrade for older implants with no longer available spare parts or in cases of bad performance

Reimplantations are subject to the same surgical principles as first implantations, however, they are associated with an increased risk for complications. Depending on the reasons for reimplantation, attention should be paid to use the same electrode and implant in the newest version because in general a connective tissue sleave has developed around the electrode. Reimplantation with another electrode array may lead to difficult insertion. The application of suitable measures like the stiff probe or the removal of newly developed connective tissue is helpful. Furthermore, the insertion should be performed under fluoroscopy. After removal of the implant from the surrounding soft tissue, the electrode array in the mastoid is dissected to the posterior tympanotomy. There, the canal of the facial nerve, the chorda tympani, and the promontory with the entry of the electrode into the cochlea are identified. The implant is separated from the electrode. After shaping of the bone bed and the tunnel to the mastoid, the new implant is inserted. The reimplantation requires specific experience of the surgeon. The electrode replacement should only be performed after those preparatory steps in order to keep the intracochlear connective tissue around the electrode array open to facilitate electrode reinsertion.

Difficulties with electrode exchange may occur especially in cases of obliteration or ossification around the electrode (▶ **Fig. 40**). It is then required to remove the new built tissue around the electrode array for extraction. If this is not possible in a suitable measure, the electrode might break and parts of it remain in the cochlea. Afterwards, the new electrode can be inserted only partially (▶ **Fig. 40**) which leads to poorer performance. The use of a split array may be considered.

If electrode systems are no longer available, a similar system must be used. In patients with very deep electrode insertion, a long electrode should be used in order to stimulate also the apical parts of the cochlea. With short electrodes auditory performance may deteriorate, especially in cases of early implanted patients with prelingual hearing loss due to the missing auditory input from previously stimulated parts of the cochlea (▶ **Fig. 40**).

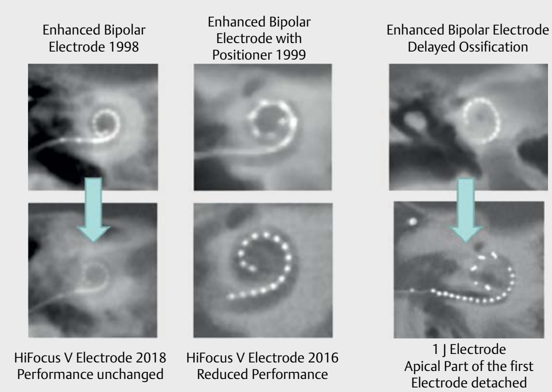
Revision rates between 7 and 8 percent are reported. [68, 69]. On the average, comparable hearing results are achieved with the use of the same electrode and the same implant if the electrode position is nearly the same [70]. This situation cannot always be achieved. Sometimes significant differences are found regarding the insertion depth before and after reimplantation. These and other factors may lead to significant differences in speech comprehension (▶ **Fig. 41**). In the context of technological upgrades, i. e. technologically advanced cochlear implant system, better hearing results may be achieved under these circumstances [68]. In cases of prelingual deafness, also poorer hearing results may be found, especially significant differences in electrode position, electrode type, and speech processing strategy [71]. If the electrode position is different after reimplantation, for example with smaller cochlear coverage or insertion angle, the performance may be poorer. Apparently, the hearing system in cases of prelingual deafness is dependent on a given electrical stimulation pattern for the auditory nerve.

4.8 Complications

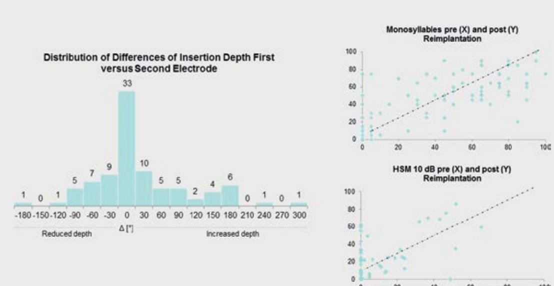
Technical and medical complications must be differentiated.

4.8.1 Implant failure (technical complications) = device failure

Implant failures occur in 2–4 % of the cases. They appear more frequently in children than in adults due to a higher incidence of external physical impact. Continuous improvement of the implant reliability could significantly reduce this rate. The incidence depends on the technological standard of the implant as well as its proper use. The results of postoperative failure analysis required by law and the experiences of users contribute to a continuous improvement of implant reliability and thus to a reduction of the failure rate. Prerequisite is a consequent data management for complete assessment of all potentially occurring device failures through implant registries. The latter might be established by the single manufacturers or independently from the manufacturers as for example a national CI registry that is currently being established by the German Society of Otolaryngology, Head and Neck Surgery. These registries require the cooperation of possibly all cochlear-implanting institutions in order to obtain complete datasets.



▶ **Fig. 40** Reimplantation using different electrodes. Impact on auditory performance. Secondary ossification can cause difficulties during electrode explantation which can lead to incomplete insertion.



▶ **Fig. 41** Reimplantation. Pre- postop differences in insertion depth and speech perception

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At the same time, standardized criteria for definition, classification, and reporting must be applied [72]. Different international standards have been established for a multitude of active implant systems that are generally applied also for cochlear implant systems. In this context, the long-term assessment of all implant types and implant generations that have ever been on the market is important to identify cumulative failure rates. This cumulative failure rate (CFR) defines how many implants of a certain type do no longer function at a certain point in time after implantation. On the other hand, the cumulative survival rate (CSR) states how many implants function impeccably at a certain time after implantation.

Besides complete failures, also partial failures may occur like the defect of a few electrode contacts [73]. For the patient, the decrement of performance caused by the failure is the important issue and should be the common basis for failure reporting. In most cases, decrement of performance is associated with a technical out-of-specification, which means that the implant does no longer meet all predefined technical functions. Not every technical defect has an impact on auditory performance such as the failure of single electrode contacts in an otherwise fully functional implant.

The technical specification (functioning) may be verified by suitable functionality tests of the implant, so-called integrity tests performed by the manufacturers on the patient. However, it is a limiting factor that not all functionalities of the implant may be checked so that it is possible that patients report about a decrement of performance as measured by hearing tests without identification of a specific device failure (so-called soft failure). If a certain failure type has never occurred and thus is not covered by the test configuration, it may be not detected. An according database query can identify a new device failure once several patients are affected. The manufacturer must perform a thorough analysis of explanted devices in order to specify the type of failure and report to the result to the implantiung surgeon as well as the competent authorities, in Germany to the BfArM [73].

Based on the occurred failures, the manufacturers have performed numerous so-called corrective actions and continuously improved the implant reliability over time which lead to a reduction of the cumulative failure rate. A prominent example of such corrective action is the change from ceramic to titanium cases that are clearly more robust and less error-prone in daily use, especially with regard to shock resistance and leakage.

Re-implantation is always indicated when the device failure significantly impairs auditory performance. This also applies for intermittent failures that may be difficult to assess because probably they do not occur during the test. Typical examples of intermittent failures are broken electrode wires at the entry site into the implant case.

Most frequent technical failures are case damages caused by external impact, e. g. hits or accidents, hermeticity problems at the feed troughs in the area of the electrode exit, failure of electronic components, or broken wires of the electrode or the antenna [74].

It is important to intraoperatively control the functionality of the implant so that already existing implant defects or those that occur during implantation are identified and re-implantation may be performed immediately during surgery.

In addition , data logging with self-check of the implant is an important tool to detect especially intermittent failures (see chapter 2.1.6–8).

Re-implantation is indicated when auditory performance is significantly impaired. Soft failures are difficult to verify when the patients credibly report about deteriorated hearing, but the available integrity tests cannot identify a deviation in the technical specifications of the implant.

In cases of confirmed implant failure, re-implantation should be performed as soon as possible in order not to jeopardize the auditory performance achieved until then. This is particularly important for children whose further hearing and speech development depends on a functioning implant. This aspect applies especially for children with only unilateral implantation [75, 76].

4.8.2 Medical complications (▶ **Fig. 42**)

Medical complications can usually be avoided by applying adequate surgical techniques. A low complication rate reflects a high quality standard of cochlear implantation and sufficient training due to an adequate minimum number of surgeries performed per year and surgeon [67, 77–79].

Similar factors are important as for technical complications. A systematic data management system is key to identify and reduce complications by corrective clinical actions. [67]. An improved surgical technique and the postoperative follow-up of patients in large centers contributed significantly to reduced complication rates. Nowadays, trans- or suprameatal approaches to the inner ear are only rarely performed because of the higher extrusion rates of the electrodes and skin break down. Furthermore, long incisions with wide exposition of the bone belong to the past.

Intraoperative complications Intraoperative complications mainly occur as damages to crucial anatomical structures, for example facial nerve, the tympanic membrane, the external auditory meatus, the chorda tympani, the sigmoid sinus, the dura, or the inner ear with opening of the modiolus, gusher, or electrode malposition (▶ **Fig. 42**) as well as damage of the labyrinth with postoperative vertigo and tinnitus. Damage of the internal carotid artery has also been described. Due to exact analysis of the preoperative imaging as well as application of intraoperative measures like monitoring or navigation, many of these complications may be avoided even in cases of difficult anatomical situations. An important precondition is sufficient training and experience of the surgeon performing the implantation. In order to secure the process quality a number of implantations per year should be performed by each CI surgeon. This is particularly important for CI surgeries in pediatric patients with additional age-related risks which must be handled by means of suitable conservative and surgical measures (see chapter on CI surgery in children). If intraoperative complications occur, they generally require immediate measures. Those are hemostasis, duraplasty, management of gusher, reconstruction of the facial nerve, the posterior wall of the outer auditory canal and the tympanic membrane as well as correction of an electrode malposition. The rate of intraoperative complications amounts to 1–5 percent [67, 77–79].

Postoperative complications Severe and mild complications must be differentiated. Mild complications like otitis media, sometimes with involvement of the mastoid, may generally be controlled with conservative measures such as antibiotics. In addition, transient hearing loss with increasing electrode impedances may occur that responds well on corticosteroids.

Severe complications such as breakdown of soft tissue with implant extrusion, facial nerve stimulation, electrode migration as well as infections with labyrinthitis or meningitis, cholesteatoma formation or brain abscess require appropriate surgical and conservative treatment.

One example of more frequent complication is the skin breakdown over the implant caused by too strong magnet pressure especially in combination with a protruding implant so that skin perforation and extrusion of the implant may occur and requires appropriate surgical measures (▶ **Fig. 43**). In cases of insufficient fixation of the implant, migration into inferior and anterior direction may lead to extrusion. In cases of insufficient soft tissue covering the electrode array for example in a radical cavity or defect of the posterior wall of the external auditory canal, electrode extrusion may occur (▶ **Fig. 42**). Subtotal petrosectomy with obliteration of the cavity is required in these cases. [51, 52]. The implant may be safed, or in cases of infection be explanted and reimplanted as a second stage procedure.

Postoperative facial nerve paresis can be primarly occur with direct injury of the nerve for example caused by the drilling or secondary caused by edema or reactivation of a latent virus infection. Postoperative high-resolution CT scan or CBCT shows potential injury of the bony nerve canal as well as the position of the electrode array in relation to the exposed nerve. If secondary paresis is found, high-dose cortisone is applied to see if the nerve function recovers rapidly, otherwise, revision surgery with inspection, decompression, and reconstruction is performed as in primary paresis [80].

If the electrode array cable is in contact with the covering soft tissue over the open mastoid, movement of the electrode induced by pressure from outside can be transmitted into the inner ear and lead to vertigo, tinnitus, and impairment of the residual hearing. This requires revision surgery with relocation of the electrode cable distant to the covering soft tissue.

Postoperative vertigo requires accurate vestibular diagnostics. Vertigo with and without activation of the implant must be differentiated [81]. Potential mechanisms are damage of the utriculus and sacculus during surgery, closure of the cochlear aqueduct by the electrode as well as (newly developed) tissue or opening of a semicircular canal. If vertigo is only observed during switch on of the implant, reprogramming is advised. If vertigo persists , revision surgery or re-implantation might be required. In cases of Menière's disease sac decompression may be adviced.

Postoperative tinnitus is often transient. Persistent tinnitus can be treated by cortisone. Chronic tinnitus may require additional measures like behavioral or cognitive therapy.

If complications occur, appropriate diagnostics and treatment should be started immediately.

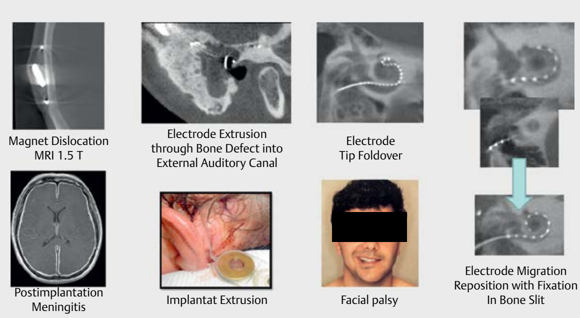
In cases of migration and extrusion, the implant should be anchored in the bone, for example by means of creating a bone bed, fixation of the implant with crossing threads, and protected by sufficient soft tissue coverage, for example by means of a muscle rotation flap from the temporalis muscle. In cases of migration, the electrode can generally be completely reinserted (▶ **Fig. 42**). Secure fixation near the cochlea with suitable methods must be done. In general, these measures may be performed without implant damage.

Infections on the implant can spread to the inner ear with consecutive labyrinthitis and post-implantation meningitis [82]. Due to the biofilm that has developed on the surfcae, explantation is usually necessary [83]. In cases without involvement of the inner ear (no cochleovestibular symptoms, see above), the electrode should remain in the cochlea until the extracochlear infection has been cured. In this way, re-insertion of the electrode is significantly easier.

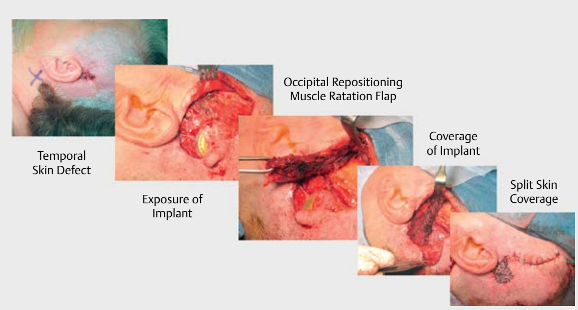
Sufficient infection prophylaxis has to be applied. Generally, CI recipients have a higher risk to acquire meningitis [84]. Thereforeall patients should undergo vaccination against *Haemophilus influenzae* and in particular *Streptococcus pneumonia* as the main bacterial specimens causing meningitis.

The overall rate of postoperative complications amounts to 2–10 percent [67].

The rate of inflammatory complications in children (6,9 percent) is clearly higher than in adults. The same is true for electrode migration, which occurs mostly with lateral wall electrodes (▶ **Fig. 42**) and can be diagnosed by CT or CBCT scan. Indirect signs are decrement of performance and missing NRT responses on the basal part of the electrode.In general, revision surgery with reinsertion of the electrode and adequate fixation [42] is necessary.



▶ **Fig. 42** Complications after Cochlear Implantation.



▶ **Fig. 43** Skin necrosis through implant migration. Repositioning of implant and muscle to cover th eimpalnt and close the skin defect.

Complications require an adequate management that a cochlear implant surgeon has to master. Continuous improvement of the surgical technique allows achieving a significant reduction of the complication rate.

An overview of complications can be found in ▶ **Table 7**.

▶ **Table 7** Long-term complications after cochlear implantation.

Long-term complications/n = 1150
– Skin necrosis: 4 – Chronic otitis media with/without cholesteatoma: 12 – Mastoiditis: 4 – Conservative therapy: 3 – Surgical therapy: 15 – Labyrinthitis: 5 – Meningitis (4–40 months postop): 6 – Implant failure: 35

5. Postoperative fitting and training of speech and hearing

5.1 Principles and contents

Postoperative fitting of the implant system to the individual stimulation conditions of the auditory nerve may be performed as early or as first fitting. Early fitting is done immediately after surgery, first fitting five to six weeks after surgery. With early fitting, the patient has the chance to get familiar with the cochlear implant hearing until the first fitting. However, fine tuning is not possible at this early stage of wound healing and patients might be disappointed with their new hearing.

During the fitting process, first the so-called T and C values are determined for each electrode contact, e. g. the minimal current needed for auditory sensation (T value) and that for comfortable loudness level (C value). The difference between T and C levels is the so-called dynamic range into which the acoustic signal has to be fitted. It is important to achieve a possibly equal loudness perception over all electrode contacts of the electrode array [85].

The entire frequency contents of the transmitted sound signal are distributed to frequency bands that are allocated to the single electrode contacts or channels of the implant. The allocation of frequency bands follows a tonotopic order so that high frequencies are coded near the round window and low frequencies near the apical part of the cochlea (▶ **Fig. 9**).

After first fitting, targeted hearing training may be performed for discrimination of simple sounds, rhythmical-prosodic elements and even words, followed by vowel and consonant differentiation exercises. In general, patients can already recognize single elements of speech during first fitting so that the complexity of the training tasks may be rapidly increased. In an iterative concept, fitting can be optimized and the auditory performance improves with extended hearing experiences.

The training aims to develop speed comprehension from vowels and consonants, numbers, then monosyllables toward sentences and open speech understanding. Further elements comprise even the use of the telephone and other communication devices, speech comprehension in noise, and directional hearing.

Due to the high technology standard of current cochlear implant systems as well as the significantly preconditions of current cochlear implant candidates with still present residual hearing in many cases, short duration of deafness, and sufficient hearing experience with hearing aids, a major success may be achieved rapidly so that patients usually start to acquire open speech understanding already after a few days.

Daily use of the implant as well as the conscious exposition to different listening situations may enlarge the hearing spectrum. Targeted exercises are the use of audiobooks, sometimes even with direct coupling to the speech processor which is particularly important for patients with single-sided deafness after cochlear implantation for targeted training of the deaf ear.

For EAS systems, the requirements of system settings are higher because two different types of hearing have to be combined effectively. This aspect concerns the cross over frequency for both types of stimulation as well as the same loudness of the electrically and acoustically supplied channels and the time alignment of the electrical stimulus to the time delay of the travelling wave on the basilar membrane.

Regular checkups of the implant as well as of the stimulus response of the auditory nerve, fine tuning of the settings, the use of auxillary devices such as an additional microphone or wireless transmission increasingly enlarge the patients' hearing experiences.

5.2 First fitting and training in children

In cases of congenital deafness, children do not have own hearing experience. Hereby, careful approaching the implanted child to the hearing world is important. Conscious combination of environmental events may connect them to the hearing space. The systematic use of the newly opened auditory sense finally allows the initiation of speech and language and based on consequent early support, spoken language may be started.

Longer periods of time have to be taken into consideration that require intensive care in specialized institutions as well as the continuous support by early support institutions as well as daily training by the parents with their hearing-impaired child.

In children, fitting can also be performed based on objective like intraoperatively measured parameters. The measured thresholds for the stapedius reflex and the electrically evoked compound action potentials (ECAPs) give hints for primary settings. Even completely ECAP-based maps may be created for which correction of the stimulation level is performed based on observation of the child's behavior. Gradually, the settings are improved according to the child's reaction.

In addition, electrically evoked brainstem potentials (EABR) and EEG signals [86] can be used to follow the development of hearing and speech and language over time. Deviations from the normative range can be detected early and appropriate measures for correction eg of speech coding or the training setting can be taken.. For control of the using habits by so-called data logging, the implant records several parameters like the daily duration of use. This information serves as support of rehabilitation [87].

In the future, the adaptation of the systems will be completed by automated elements. Further objectively measured parameters such as cortical, electrically evoked potentials play an increasing role. These EEG parts allow an assessment of conscious hearing, attention, and discrimination of the electrically coded acoustic stimulus. Iterative procedures allow optimizing the speech processing strategy for the single patient and different hearing situations [87–89]. This aspect can lead to different settings for different hearing situations.

Therefore, so-called closed-loop systems will have additional EEG measurement electrodes that will be placed epi- or subdurally at predefined sites of the temporal region during cochlear implantation. In this way, relevant EEG components like the N1P1 potential, mismatch-negativity responses, or even P300 can be measured quasi online [88]. In addition, certain frequency parts of the EEG, the so-called gamma activity, may be measured allowing to check the attention to an acoustic stimulus. Targeted variations of the speech processing strategy to increase these EEG components probably allow to optimize the speech coding algorithm for the single patient [89].

5.3 Hearing and speech therapy

Generally, cochlear implantation is divided into three phases of basic and subsequent therapy as well as lifelong aftercare. Basic and following therapy have a high significance and are an integral part of CI treatment. Already during fitting of the speech processor, basic therapy starts. In postlingually hearing impaired patients, hearing training is sufficient. The hearing training takes up present hearing experiences and the individual speech level and aims at making comprehensive acoustic experiences available for the contact with the environment and at fostering the binaural integration as cognitive process. Based on long-term practice, different learning contents turned out to be suitable for hearing therapy that are already started in basic therapy and expanded and consolidated in the follow-up therapy [90]. These contents can be assigned to different areas of central-auditive perception and processing. Nowadays, hearing training apps or online materials complete the process allowing CI users to train individually and on their own at home [91].

However, in the context of therapeutic measures for prelingually hearing impaired children aiming at using hearing impressions with the technical hearing devices, allowing optimal orientation in the acoustic environment, and creating basics for comprehensive speech and language development, the term of hearing education is used [91]. In the initial phase of hearing education, it is important that the child regularly wears the well-set hearing system, preferably during the whole day so that hearing may increasingly develop as everyday ability. Then the attention may be directed on perception and identification of certain sounds. The child learns that hearing, like vision, is associated with information and symbols. The higher the attention is for auditive signals, the better the child learns giving acoustic feedback. Therefore, hearing education always includes exercises for auditive attention and memory. These abilities together with cognitive intelligence are the basis for language and speech acquisition. The support of hearing-oriented language and speech development is performed simultaneously and is based on current speech therapeutic theories and methods.

The success of hearing training and education can be measured by means of current speech reception test methods.

5.4 Telemedicine/remote care

Patients increasingly benefit from technical progress with regard to patient empowerment. The objective is patient self-care for individual hearing situations. Associated aftercare has to be performed by the implant center as well as by selected cooperation partners in a connected network. Mostly, these partners are hearing aid acousticians and other hearing care professionals as well as cooperating otolaryngologists. The cooperation in a network and a standardized data management allow continuous patient data collection and exchange. This leads to multiple options of decentralized aftercare with permanent access to the expertise of the implant center, so-called hub and spoke system. The provision of spare parts, implant checkups, implant fitting and technological upgrade can be offered near the patient's residence.

Telemedicine allows remote fitting [92]. The patient is connected with the implant center via a remote data transmission line. The specialist in the cochlear implant center can observe the patient and communicate directly with him. Direct access to the implant is possible by means of a specialist on site or an interface that is controlled by the patient himself. In this way, implant fitting especially in the home environment, technology checkups, and software upgrades may be performed [93].

Telemedicine allows also daily control of implant function as well as the electrode-nerve-interface, e. g. increased impedances as hint for labyrinthitis onset can be identified early. Remote care is important especially for lifelong aftercare [94].

5.5 Self fitting

The patients themselves can use and optimize different maps for different listening situations. A controlled procedure is necessary in order to avoid either overstimulation or inappropriate loudness. The automated function control of the implants as well as the implemented options of performance testing allow an increasing contribution of the patients and thus also self-treatment for example by using suitable training apps. Apps also allow connection with the implant center at any time. The evaluation of the measurement data can be automated. The assessment by learning systems (AI) allows the continuous success control and comparison with the predicted auditory performance. In this way, variations of performance as well as technical and medical complications (increased electrode impedances) can be identified early.

5.6 Adjustments, rehabilitation, and aftercare

In the meantime, different treatment concepts have been developed that all aim at a possibly lifelong, individual aftercare for different listening situations of the patients. The patients' capacity in their social and professional environment shall be optimally used and individual support shall be provided. Different aspects have to be addressed.

1. Regular checkup of the implant function, upgrades of hard- and software, provision with spare parts and additional devices

2. Adaptation of the aftercare according to the circumstances by a local partner network (see above) or by the implant center
3. Permanent access to the expertise of the implant center and other partners by remote care

5.6.1 Rehabilitation

In order to support hearing and speech development, especially in children, specific rehabilitation measures are necessary. This aspect also applies for adults who experience slow progresses or who need a more intensive therapeutic approach due to unfavorable prognostic factors like long duration of hearing loss. In this way, significantly better hearing results may be achieved at least temporarily [95].

5.6.2 Aftercare

After implantation, the surgeon is responsible for the organization and quality assurance of lifelong aftercare. This refers to the technical checkups as well as the setting and fitting of the implants. Furthermore, regular updates of soft- and hardware are required. Thus, advances in implant technology are made available for the patient. Medical complications and device failure can be identified and managed .

5.7 Methods of technical check up and assessment of hearing outcome

The tasks of aftercare and rehabilitation described above require a systematic and standardized approach. Several test procedures are available (some of them are listed in ▶ **Table 1** and ▶ **8**) to check the following parameters:

1. Implant function
2. Electrode impedances
3. Objective audiometric parameters like ESRT, ECAP, and EABR
4. Psycho-acoustic procedures for measurement of T and C levels, loudness growth function, dynamic range, and frequency allocation
5. Speech comprehension in free field or direct coupling mode (▶ **Table 8**)

The already described procedures are applied for adults and children like the Freiburg speech perception test, HSM sentence test in quiet and noise as well as the OLSA test (matrix test).

For pediatric patients, standardized questionnaires for parents and teachers are used (e. g. LittleEars, FRAKIS) for documentation and assessment of beginning development of speech and hearing. As from the age of about 2 years on, the application of age-based standardized test procedures is possible to assess the speech and language development. These test procedures also allow the comparison of test results with normally hearing peers and at the same time an evaluation of the progress in hearing and speech and language development.

From 4 years on pediatric speech comprehension tests can be used.

Comparative evaluations with the preoperative hearing status as well as the development of speech comprehension in quiet and in noise can be documented over time by means of the CAP scale [96].

Bilateral hearing may be assessed by free field testing. The patients are often provided with either bimodal (cochlear implant and hearing aid) or bilateral (2 cochlear implants) systems or with a hybrid system for electroacoustic hearing in one ear and a hearing aid in the contralateral ear (so-called combined mode). The various hearing situations have to be assessed separately and the percentage of the different hearing modalities (acoustic, electrical, electroacoustic) regarding the whole hearing situation must be evaluated.

The amplified (aided) hearing threshold should be in the range of 20–30 dB over the entire electrode spectrum or in cases of EAS systems over the entire frequency range and be balanced between the two ears.

6. Results

The results of cochlear implantation could be significantly improved during the last decades. This is mainly due to the technological development and progress as well as the changed indications of patients with clearly better prognostic factors.

6.1 Outcome in postlingually deafened patients

The postoperative hearing results are usually associated with a rapid onset of open speech comprehension. The results improve continuously over a period of generally 6–12 months. In addition, a further increase especially of speech comprehension in noise and in specific hearing situations is observed [97, 98]. About 80 % of the patients reach open set speech comprehension with a wide variability (▶ **Fig. 44**). Based on a median value of about 65 % monosyllabic word understanding, the patients may be classified into good, average, and poor performers according to the percentiles of 30 and 70 (▶ **Fig. 45**). Only few patients have poorer speech comprehension compared to the preoperative situation under best aided conditions with hearing aid. In most cases, special conditions could be identified like incomplete electrode insertion, impaired cognitive function or stimulation of the facial nerve (▶ **Fig. 46**).

On the average, new implant generations achieve better auditory performance than previous ones. This is mainly due to the progress of processor technology, especially the increase in stimulation rate [99]. However, despite improved electrodes and innovative

▶ **Table 8** Test procedures for the assessment of auditory performance.

- Speech perception in free field or by means of direct coupling
- Adults: Freiburg monosyllables test, HSM sentence test in quiet and in noise, Oldenburg sentence test (OLSA)
- Children: LittlEars hearing questionnaire, Göttingen speech comprehension test for children, Oldenburg sentence test for children (OLKISA), age-related tests for evaluation of the Categories of Auditory Performance [55]
- Binaural treatment: testing of each modality as well as the overall situation
- Bilateral = CI in both sides
- Bimodal = CI + hearing aid in the contralateral ear
- Combined = EAS + contralateral hearing aid
- Directional hearing
- Children: language development tests
- Questionnaires for early childhood language development (FRAKIS)
- Language development tests for children (SETK 2–5;11)
- Marburg speech comprehension test for children (MSVK)

speech processing strategies, the results have reached a plateau that cannot be surpassed effectively with the current implant concept (▶ **Fig. 45**). The basic limitations of current cochlear implant systems become obvious, especially regarding the existing bottleneck of the electrode-nerve interface.

Important patient specific factors influencing auditory performance are the duration and etiology of deafness, cognitive skills, the age at implantation as well as the residual hearing.

Further individual factors are the position of the electrode, the cochlear coverage, the functional condition of the auditory nerve that is still very difficult to assess for example by the so-called cochlear analyzer [100].

6.1.1 Duration of deafness

The longer the duration of deafness, the longer the restoration of hearing, especially of speech comprehension takes [90]. Datasets of 1,002 postlingually deafened patients assessed at the occasion of the follow-up appointment after 5 years have been analyzed retrospectively. For more exact evaluation, the data were assigned to defined durations of deafness. All patients underwent the Freiburg monosyllables test, HSM sentence test in quiet and in noise (10 dB SNR) in the free field at 65 dB SPL.

Patients with a duration of deafness of less than one year achieved 60 percent speech comprehension in the monosyllables test. Patients with a duration of deafness between one and ten years had 65 %, patients with a duration of deafness of 10–20 years 63 %, patients with a duration of deafness of 20–30 years 45 %, and patients with even longer durations of deafness reached 28 %. The statistical analysis showed a reduction of the speech comprehension in dependence of the duration of deafness (Jonckheere-Terpstra test, $p < 0.05$) (▶ **Fig. 47**). Even after longer use of a CI, the duration of deafness has a decisive impact on the speech comprehension.

6.1.2 Influence of cognitive skills and age

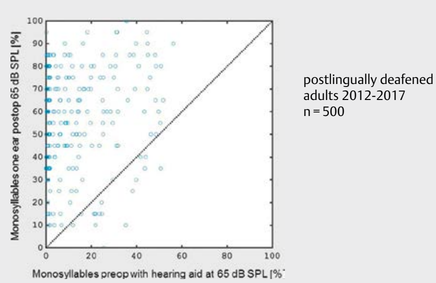
For quite a long time, the correlation between hearing loss and the reduction of cognitive skills is well-known [101]. There is a negative correlation with the degree of hearing loss [102]. The provision of a cochlear implant clearly improves hearing which also contributes to increased cognitive performance. Hereby, various skills can be improved to a particular measure like global cognition [103]. Non-inferiority tests about cognitive performances of the study group after cochlear implantation show that the global cognition, the figural episodic memory, and the attention control reached a similar level than the normally hearing control group after 12 months of CI use. The improvement of the global cognition is significantly correlated with the speech recognition after three months of cochlear implantation.

The influence of the patients' age on the hearing performance cannot be considered separately from the cognitive skills. In the age decades over 60 years, the median performance shows a clear reduction (▶ **Fig. 48**).

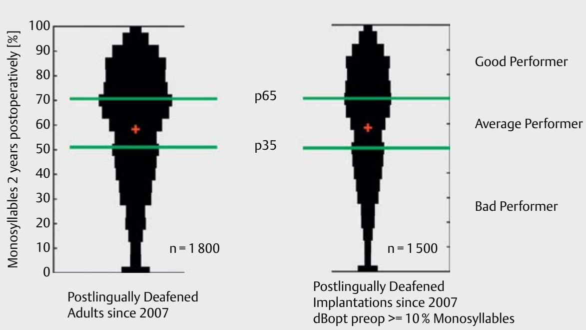
6.1.3 Influence of residual hearing

In general, hybrid systems lead to better speech comprehension, especially in noise and directional hearing, and increase the perception of music due to the preserved tonal hearing in the low frequency range. Hereby, suitable patients with sufficient residual hearing in low frequencies must be selected who may benefit from the acoustic component of the EAS system [26–28]. Pure tone thresholds should be better or equal 55 dB at 500 Hz and 40 dB at 250 Hz before surgery and 65 dB and 50 dB postoperatively.

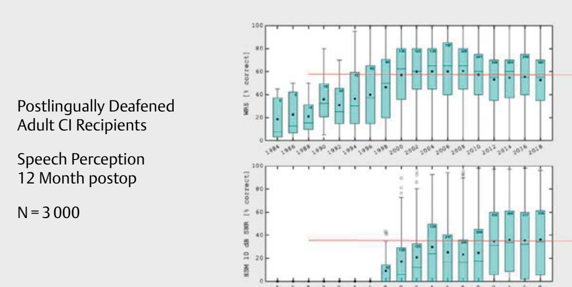
Hearing results achieved with electro-acoustic stimulation using short electrodes are significantly better, especially in noise compared to those with long electrodes and electric stimulation only. Electrode length together with the cochlear coverage are important for speech comprehension. Büchner et al. [23] could show that patients using electric stimulation and long electrodes have better



▶ **Fig. 44** Speech perception in adult postlingually deafened CI recipients. Scores 12 month postoperative versus preoperative. Significant improvement with large variability of auditory performance.



▶ **Fig. 45** Distribution of results and grades of performance. Postlingually deafened adult CI recipients, scores 12 month postoperative.



▶ **Fig. 46** Auditory performance in postlingually deafened adult CI recipients at Medizinische Hochschule Hannover 1984 until 2019.

speech in noise understanding and results than patients with shorter electrodes without acoustic. This also reveals the importance of apical (acoustic better than electrical) stimulation for good speech comprehension.

Especially patients with preserved residual hearing using EAS can benefit from very good speech understanding in noise. (▶ **Fig. 49**). Patients with high frequency deafness, e. g. due to presbyacusis and near to normal low and mid frequency hearing can achive better speech in noise understanding compared to hearing aid use through high frequency hearing restoration using a short electrode insertion for ENS (electro-natural hearing stimulation) (▶ **Fig. 50**). This gives a high potential for future development of hearing restoration strategies and subsequent extension of indication criteria (see chapter 8.1).

6.1.4 Bimodal hearing

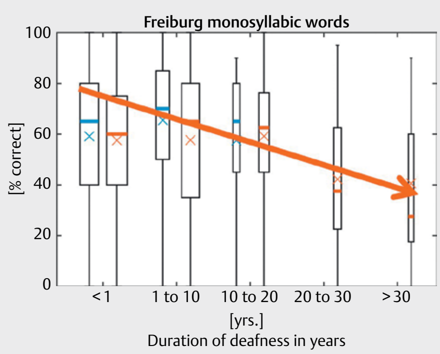
In bimodal hearing, the residual hearing of the contralateral side is used in combination with electrical hearing of the implanted ear. Significantly better hearing performance is achieved compared to the use of the cochlear implant only or the hearing aid only [104–107]. The additional use of the hearing aid leads to an improvement of low frequency hearing and better speech comprehension in noise (▶ **Fig. 51**). Cochlear implants cover the cochlea with electrical stimulation down to around 1000 Hz with a wide individual variability [108, 109]. An additional support with hearing aid may thus increase the range in direction towards low frequencies. Illg et al. [110] revealed that the hearing threshold must be 80 dB or better in the frequency range between 125 and 250 Hz in order to achieve improved speech comprehension through the bimodal effect.

However, a small patient population did not have any benefit from bimodal hearing or even poorer speech comprehension under bimodal conditions although this group did not have a significantly larger hearing loss of the frequencies below 1000 Hz compared to the group with bimodal benefit. It is assumed that other parameters such as cognitive skills have an impact on the speech comprehension and should be considered in bimodal treatment.

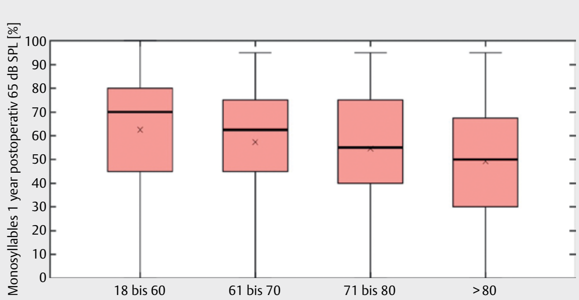
If the acoustic hearing of the contralateral side deteriorates below the above-mentioned values, the patient might be a candidate for bilateral CI with implantation of the contralateral side. In general, this leads to better outcomes regarding speech comprehension in noise as well as directional hearing. This aspect also applies for children.

6.1.5 Influence of the etiology of deafness, in particular malformations and hypoplasia of the auditory nerve

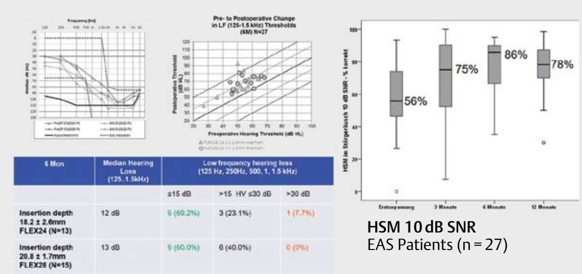
The influence of the etiology of deafness on auditory performance could not be definitely clarified up to now taking into account the



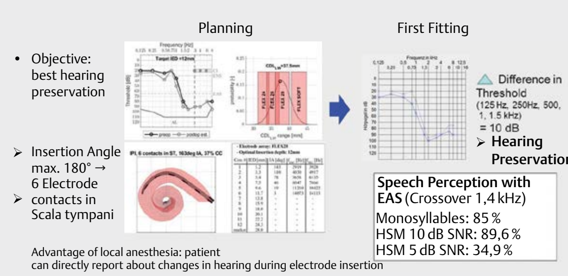
▶ **Fig. 47** Impact of duration of deafness on auditory performance. Signifcantly poorer performance in patients with long duration of deafness, but large interindividual variability reduces the possibility of individual outcome prediction.



▶ **Fig. 48** Impact of age at implantation on auditory performance in postlingually deafened adult CI recipients. Scores 12 month posteratively.



▶ **Fig. 49** Results in patients with individualized partial cochlear implantation. High rate of good hearing preservation and excellent speech perception in noise.



▶ **Fig. 50** Extended CI indication for patients with high frequency deafness due to presbycusis and no benefit with hearing aids. Complete hearing preservation. Electro-natural stimulation without hearing aid very good speech perception score in noise. Surgery under local anesthesia allows for direct feedback on residual hearing and potential changes during electrode insertion .

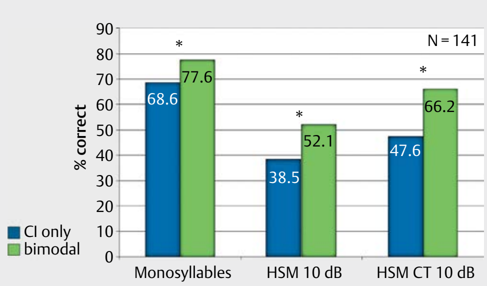
large population of CI users with unclear etiology. The impact is very clear in cases of malformations, obliterations, or perisynaptic audiopathies. Furthermore, additional disabilities and syndromes as well as genetic factors play a crucial role [111]. In the following, more detailed explanations will be given based on the example of malformations.

The high variability of different malformations is also reflected in the difference of the functional performance of the auditory nerve. The presence of a hearing nerve cannot always be identified by currently available imaging techniques. Therefore, children with suspected aplasia of the auditory nerve should always receive a CI within the first year of life unless aplasia of the cochlea or the internal auditory canal is found.

For children who receive CI within the first two years of life and who have a malformation of the cochlea and/or suspected dysplasia of the auditory nerve, the interdisciplinary cooperation of ENT specialists, neuroradiologists, audiologists, CI engineers, and therapists is extremely important to decide if the quantity and quality of the electrical stimulation with the CI is sufficient for auditory based speech and language acquisition. CI fitting in these children is different from that in children with normal anatomy and auditory nerve function. If no sufficient hearing and speech development is observed, a dysfunction of the auditory nerve can be assumed and an auditory brainstem implant (ABI) should be considered. EEG and NIRS measurements give information about the activation of the auditory cortex. In parallel, the progress of the audioverbal development is assessed by age appropriate methods including parental questionnaires.

6.2 Single-sided deafness (SSD)

Meanwhile, cochlear implant is a widely used option of auditive rehabilitation in cases of single-sided deafness. Generally, the second ear improves directional hearing as well as speech comprehension in noise and very often suppresses the accompanying tinnitus [112–116]. However, it must be stated that the hearing performance is always lower compared to the one of the normally hearing contralateral ear. In cases of asymmetric hearing loss, the hearing performance of the cochlear implant may become comparable with the one of the hearing impaired contralateral ear or even be superior. To ensure the achieved results, repeated isolated hearing training for the implanted ear is required, for example in direct coupling.



▶ **Fig. 51** Bimodal hearing improves speech perception through the acoustic stimulation of the contralateral ear (Illg et al., 2014).

Finke et al. [117] investigated the speech comprehension over a period of 12 months and the subjective benefit in CI users with single-sided deafness and correlated the results with speech comprehension of bilateral CI users who had been implanted sequentially.

Significantly lower values are achieved in unilaterally deaf patients with CI in all three speech comprehension tests applied. The success of cochlear implantation based on the speech comprehension is clearly different between both patient populations even if the patients of both groups had received comparable hearing training. In the first year after implantation the performance increase is comparable in both groups. The dominant normal hearing of the contralateral side that is always present in daily life seems to limit the quality of the speech comprehension with electrical stimulation in unilaterally deaf patients. Specific advantages of bilateral hearing, however, are maintained such as improved speech comprehension in noise and the localization of sound (▶ **Fig. 52**). Also in comparison with patients with bone conduction hearing aids like BAHA (bone-anchored hearing aid) or CROSS devices (contralateral routing of signal) [118], the rating in the "Bern Benefit in Single-Sided Deafness Questionnaire" is descriptively higher, i. e. the benefit due to the cochlear implant is obvious.

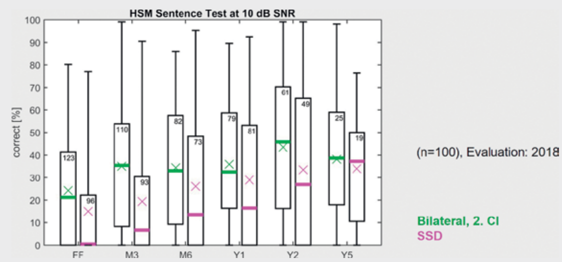
6.3 Outcome of prelingually deafned children

Corresponding to normal hearing children, the speech and language development generally takes two to six years. A comparison with the development of normal hearing children can only be made when no additional disabilities or particularities are present. In general, early implanted children reach very good speech development scores in quiet that are very similar to normally hearing children [119]. However, the hearing performance in noise is clearly poorer which supports the statement that the cochlear implant does not provide normal hearing but turns deafness to the status hearing impairment (▶ **Table 9**). Taking into account the overall development, it must be emphasized that deficits due to hearing impairment remain also in other fields of development, especially the entire cognitive development. This fact can also be explained by the close interrelation of the auditory system with other brain areas and their functions, the so-called connectome [120].

Results show a large variability in performance which is only partially explainable by some parameters. In this context, the most important prognostic factors are the time at implantation and the onset of the auditively supported speech acquisition. Hereby, bilateral auditory input is particularly important in order to develop directional hearing and speech comprehension in noise. Critical phases of brain development must be respected. In children, even the development of a true binaural hearing system is possible [121].

In cases of congenital hearing loss, hearing results and speech development depend strongly on the age at time of implantation (▶ **Fig. 53**). Children with congenital hearing loss who are bilaterally implanted within the first two years of life develop a significantly larger passive vocabulary than children who are implanted later (▶ **Fig. 54**). In acquired hearing loss, the age at onset of hearing loss and the duration until implantation are crucial for the achieved results.

This fact is also reflected with respect to schooling. About 70 percent of early implanted children are able to visit regular schools.



▶ **Fig. 52** CI and SSD (Single Sided Deafness). Open speech understanding on the implanted ear. But scores are poorer than those of the second CI in bilaterally implanted patients .

This percentage decreases significantly in patients who are implanted later [122].

Also in professional training and education, generally a higher level may be achieved in early implanted patients compared to late implantation. Cochlear-implanted children are meanwhile able to take every profession later in life. However, on average a lower school level and professional qualification are found compared to normal hearing peers [123].

In summary, cochlear implants may turn severe to profound hearing loss or deafness to the level of moderate hearing impairment. The increased hearing effort compared to normally hearing people remains. This means that a higher percentage of the cognitive capacity is bound by hearing which leads to increasing cognitive stress. The remaining cognitive capacity is limited for further tasks like learning ability or adaptation to particular tasks in the job compared to normal hearing individuals. At the same time, these facts show the significance of cognitive capacities for the success of auditory rehabilitation. The higher the cognitive capacity, the better is also the hearing success, especially in difficult listening situations.

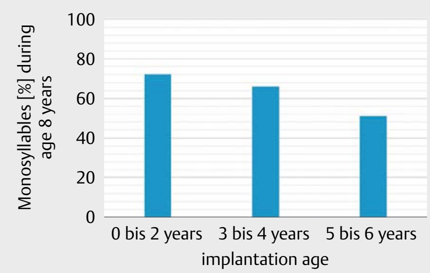
Fortunately, the introduction of the nationwide newborn hearing screening significantly improved the early identification of severely hearing-impaired children [124, 125]. However, the continuation of the screening for older children as nationwide screening is still pending. Thus, children with progressive hearing loss might be identified too late which leads to non-optimal benefit in cases of perilingual deafness.

Other factors significantly influencing the hearing and language development are malformations, obliterations, additional disabilities with impact on the rehabilitation capacities, syndromes, and genetic origins of deafness [111]. Therefore, children should always undergo diagnostics for the identification of further disabilities. Their spectrum is large. Most crucial are deficits of cognitive performance, for example disorders of the autism spectrum (ASS). 80 percent of the patients with ASS accept the speech processor well, some of them even use spoken language; however, only half of them are able to successfully communicate.

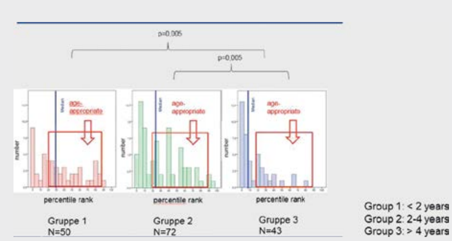
▶ **Table 9** Categories of Auditory Performance (Archbold et al., 1995) **[**167].

CAP category	Criterium
8 7	Conversation in noise without lip-reading Phone calles with known speakers
6	Conversation without lip-reading
5	**Sentence comprehension without lip-reading**
4	**Discrimination of parts of speech without lip-reading**
3	Recognition of environmental noise
2	Reaction on speech
1	Perception of environmental noise
0	No perception of noise or voices

CAP mean value: 4.63 (min – max 0–8).



▶ **Fig. 53** Impact of age at implantation on speech perception in congenitally deaf children.



▶ **Fig. 54** Impact of age at implantation on vocabulary in congenitally deaf children.

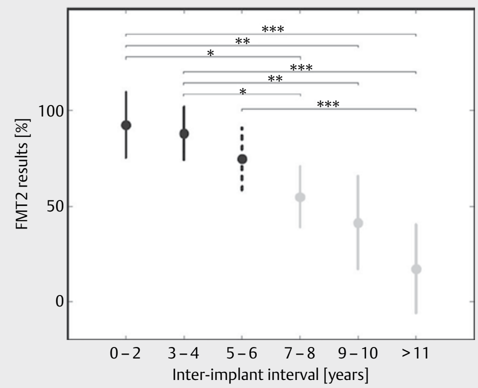
6.3.1 Influence of the inter-implant interval in sequential bilateral implantation

If unilateral cochlear implantation is performed in children with bilateral congenital deafness within the first two years of life, speech comprehension may develop that is highly beneficial in quiet environment. However, after unilateral cochlear implantation and an average duration of use of 12.86 ± 1.99 years speech in noise can only be understood in up to 22 % [126]. Similarly, directional hearing does not develop [127–129] because the separate perception of two or more sound sources is only possible with bilateral hearing which significantly supports speech comprehension in noise [130].

In order to optimize the speech comprehension in noise for bilaterally deaf and unilaterally CI implanted children, many of these children received a second cochlear implant. Despite audio-verbal training, it became obvious that the time gap between the implantations of the two ears and thus the older age of the children at the time of implantation of the second ear is the most crucial parameter for speech comprehension after CI implantation on the second side. The inter-implant interval should not exceed 4 years in cases of sequential bilateral cochlear implantation of congenitally deaf children because with a longer interval the speech comprehension of the second ear is significantly poorer than on the first side [131] (▶ **Fig. 55**).

As long as a hearing aid is used on the contralateral side in addition to the unilateral CI, a useable residual hearing seems to contribute to hearing pathway maturation. In children and adolescents with longer durations of use (3–16 years), the comprehension of sentences in quiet and in noise was significantly higher than in children with short durations of hearing aid use [126].

Furthermore, the duration of bilateral use of both cochlear implants finally has a positive impact on speech comprehension. Longer bilateral use of CIs leads to better speech comprehension. Significant improvements could be shown in monosyllables and HSM sentence tests in quiet [126].



▶ **Fig. 55** Impact of inter implant interval on speech perception of the second implanted ear in congenitally deaf children (Illg et al., 2017).

7. Quality of life

7.1 Assessment of quality of life of CI users

According to the World Health Organization (WHO), health is defined as "a state of complete physical, mental and social well-being and not merely the absence of disease or infirmity". Health assessment and the impact of healthcare must include changes of the severity of diseases as well as the improvement of the quality of life (QoL). Generic QoL measurements allow a general comparison of the effectiveness of a treatment and the quality-adjusted life years for different diseases (e. g. the effect of a pacemaker compared to a cochlear implant), while disease-specific QoL measurements focus on certain patient populations (e. g. cochlear implant users, possibly for comparison of indications and subgroups) or a certain indication (e. g. people with unilateral hearing loss, possibly for comparison of different treatment options).

Another perspective for the measurement of the (disease-specific) quality of life is provided by the International Classification of Functioning, Disability and Health, well-known as ICF [132]. The ICF is an addition to the ICD-11 (International Classification of Diseases). It is a classification model describing health status functions (body functions, activities, participation) and the disability (impairments, impaired activities, impaired participation) of a person.

During the last years, patient-reported outcome measures (PROM) of the subjective quality of life have gained importance and attention [133, 134] and the acceptance of patient-reported QoL results has increased during the last decades [135–138].

Several disease-specific parameters have been developed and applied in order to assess the effects of hearing loss and the use of hearing aids or implants for different indications [139–145].

Many disease-specific measures in the treatment of hearing loss focus on hearing in everyday life, i. e. on what a person is able to hear or not [146], the quality of hearing [147, 148], or the sound quality [149]. Only few questionnaires focus on how hearing loss influences a person's quality of life [150, 151] or have been developed specifically for hearing aid users but not for CI users [152, 153]. The questionnaire on the abbreviated profile of hearing aid benefit (APHAB) [151] was defined as gold standard for addressing communication problems with and without hearing aids, however, the criticism was that the scoring system is rather complex, the wording of the items is too complicated, and when the questionnaire is not completed, the score is not calculated based on the number of answered questions and the statement is issued that no conclusion can be drawn about the effect of hearing loss on a patient's quality of life [139].

The most frequently applied questionnaire in the context of CI is the Nijmegen Cochlear Implant Questionnaire (NCIQ). It has been developed specifically for the assessment of the quality of life of CI users. It also has some disadvantages: Many items focus rather on the hearing abilities or problems in different hearing situations than on the quality of life. With 60 items it is very long which complicates the implementation in follow-up practice. Further it seems that some sections do not provide reliable information [139], and finally the items were formulated more than 20 years ago and it might be stated that they do not correspond well to current expectations and results of CI users. According to our knowledge, there is only one questionnaire on the assessment of hearing loss that has been developed recently and that refers to the quality of life, which is the ERSA questionnaire (Évaluation du Retentissement de la Surdité chez l'Adulte) [139]. The ERSA comprises four sub-scores: quality of life, private life, professional life, and social life. The authors provided the questionnaire for hearing impaired adults and explained that it had been developed to assess the effect of hearing loss (independent from the severity of hearing loss or the rehabilitation status by means of cochlear implant or traditional hearing aid). This questionnaire includes the quality of life in four different areas of everyday life.

7.2 Quality of life of adult CI users

During the last years, several trials have been published about the effects of cochlear implantation on everyday life and psychological well-being of affected people [154–157]. The results show that cochlear implantation significantly improves the quality of life. Studies assessing the health-related quality of life of CI users of different age groups partly reveal heterogenic results. Olze et al. [158] described that the benefit of older participants (70–84 years) was higher than in younger ones (19–67 years), while Djalilian et al. [159] could not find any differences related to age. These differing results in the literature show that the hearing-related quality of life is influenced by additional factors like speech comprehension, psychological, cognitive, and audiological aspects.

The results of studies with older CI users ([158, 160, 161] show that the positive effect of cochlear implantation is not limited to the hearing capacity but also includes a reduction of tinnitus, depression, cognitive decline, somatization disorders, and isolation of CI users which leads to an increased quality of life. All studies report an increase of the quality of life within the first six months after cochlear implantation without further changes after 12 months. Current data of the Department of Otolaryngology at Hannover Medical School show that a significantly higher quality of life is observed after three months, measured by means of the NCIQ, in older patients which remains stable until 12 months after surgery. The data reveal that the benefit for younger patients (group 1: 60–70 years) is higher compared to older patients (group 2: 71–90), indicating that also patients older than 60 years benefit from cochlear implantation by increasing the quality of life and avoiding deterioration of the general health status (▶ **Fig. 56**).

Even in older hearing impaired people, the CI use does not only lead to a better speech comprehension in quiet and in noise, but it also has a positive impact on physiological and social deficits independent from age. Furthermore, both groups achieved the same level 12 months after surgery despite the presence of physical comorbidities (25 % in group 1 and 40 % in group 2). This fact confirms that also older patients with additional diseases may benefit from improved hearing with CI.

7.3 Quality of life in children

Tools for measurement of the health-related quality of life are also available for pediatric patients with chronic diseases. In most cases, they may be applied as self- or third-party assessments [162]. Measurement tools regarding the hearing-related quality of life in children, however, have not been explicitly developed. Morettin et al. [163] concluded that the measurement of pediatric quality of life comprises different concepts and methods. Regarding children using CI, the results show that it is difficult to develop a comprehensive concept which identifies the quality of life important for children and how these areas may develop further during childhood taking into account the multitude of the evaluated instruments and aspects.

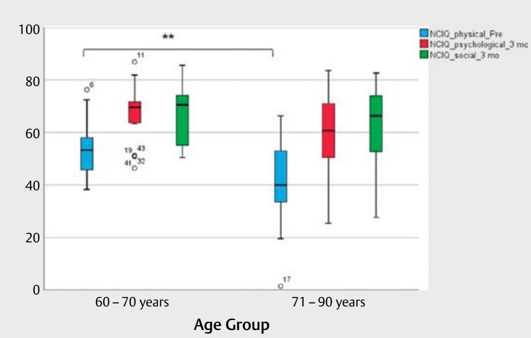
Well-known measurement tools in German speaking countries are for example “Disabkids” and “Kidscreen”. Both tools are interculturally developed questionnaires to assess the health-related quality of life of children and adolescents [164]. While the questionnaire entitled “Disabkids” has been provided for children and adolescents with chronic diseases, the “Kidscreen” questionnaire may be applied for sick as well as healthy children.

Haukedal et al. [165] measured the quality of life of children with CI aged 5.6 to 13.1 years by means of the Pediatric Quality of Life Inventory and compared the results with the ones of normal hearing peers. Most children with CI have quality of life very close to the one of the peers of the same age and sex. However, the children reported social and schooling related issues indicates that these areas require more attention during rehabilitation for a good quality of life. The improvement of spoken language may contribute to an improved QoL.

7.4 Education and occupation

During the past 20 years, many evaluations on the long-term success of cochlear implant in children have been performed and published. Reports about schooling and professional development, however, are rarely found in literature. In order to determine the effectiveness of cochlear implantation also with regard to education and graduation over a longer period, the hearing results of 933 CI users (mean age at the time of assessment: 23.6 years; mean age at implantation: 5.4 years) were retrospectively analyzed and the patients were asked about their school and professional career [123]. Auditory performance was classified into the categories of auditory performance (CAP, ▶ **Table 9**). The reply rate was 18.65 %. The answers were categorized into international standards (ISCED: International Standard Classification of Education; ISCO: International Standard Classification of Occupation; ALLBUS: allgemeine Bevölkerungsumfrage in Deutschland), compared with national population data and evaluated statistically. The results show that 86 % of the participants use their CI more than 11 hours per day, only 2 % of the participants state that they do no longer use their CI (▶ **Fig. 57**). The mean CAP value is 4.63 (0–8) which means that speech can be differentiated without lip-reading (▶ **Table 9**). Certain parameters have impact on the auditive development like the age at the time of implantation that correlates significantly with the CAP values ($r = 0.472$; $p = 0.0$).

The graduations from school and professional qualifications of CI users and normal hearing peers are significantly different ($p = 0.001$). CI users achieve high school graduation allowing the access to university education in a lower percentage compared to normal hearing peers (▶ **Fig. 58**). Therefore, also professional qualifications requi-



▶ **Fig. 56** Quality of life with CI in different age groups of postlingually deafened adults.

ring university degrees are underrepresented among CI users. Most CI users reach professional qualifications in the so-called skill level 2 which summarizes service employees, salespeople, employees in agriculture and fishing, car mechanics and others as well as employees in the horticulture sector (▶ **Fig. 59**).

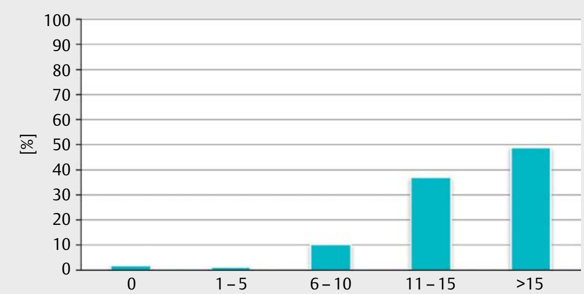
Maternal school degree correlates positively with the education of the CI recipients. 94 percent of the patients report that their CI was necessary for communication in school. According to the data, 83 percent used spoken language as most important communication during their school career. 64 percent of the patients visited special schools during their professional training. About 70 percent work in the job they have learned. Overall, about 40 percent are employed compared to 85 percentin the normal hearing population. [166]. Among the CI users, a major percentage is still in professional training. More than 60 percent of the CI users who returned their replies work in permanent positions, one in five (20 percent) has a limited employment contract. Nearly half of the CI users has been unemployed for a certain time compared to 30 percent of the normal hearing people [166].

70 percent stated that they could learn the job they wanted and nearly two third (62 percent) report that their job is as they expected and wanted it to be. Nine in ten CI users get along well with their work, their tasks, and the responsibility they have in their jobs, nearly 60 percent are satisfied with their salary. The majority of CI users (97 percent) use their device permanently at work but only in 18 percent specific arrangements have been made at work for better hearing or comprehension. 68 percent state that the CI is necessary for the communication at work, 28 percent state that it is sometimes necessary because spoken language is used. Most CI users do not use a FM device at work (95 percent).

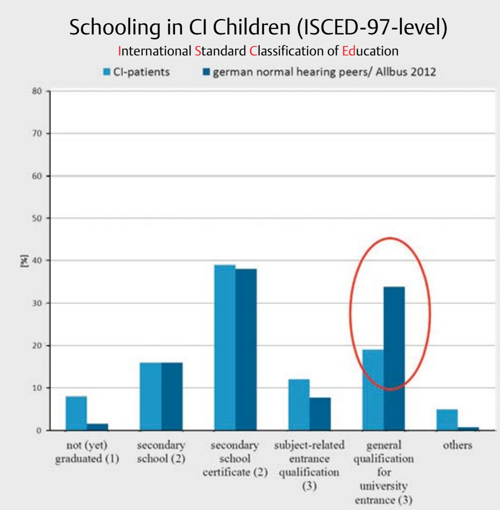
The analysis of the data showed a positive linear correlation between auditory performance and professional degrees. Therefore, there is still the hypothesis that low auditive abilities lead to poorer education and professional development. Early diagnostics and cochlear implantation open the best chances for school education and professional career. Cochlear implants do not only improve the quality of life in hearing-impaired children, but they are also economically reasonable because they can save costs (tax payers instead of tax consumers). In comparison to normally hearing people, the professional situation of CI users shows a higher rate of unemployment, the employment contracts are more frequently limited. In the job market, CI users do not have the same possibilities as normally hearing people. It remains open how jobs for hearing impaired people have to the designed. This aspect has to be assessed and evaluated in the future. In the future, it may be expected that the school and professional situation of CI users further improves and equals the ones of normally hearing people because an always younger age of implantation and technical developments contribute to better speech comprehension so that children may be treated early in an optimal way and the educational opportunities further increase.

7.5 Non-users

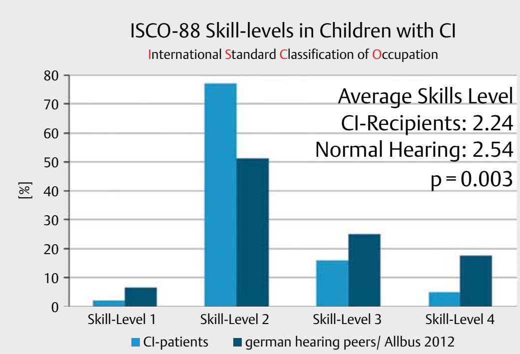
Date on non-users are difficult to assess because these patients do no longer appear in the CI clinics. From the CI database of the Department of Otolaryngology at Hannover Medical School , Germany, 9,949 datasets have been analyzed retrospectively regarding history, comprehension of monosyllables, and datalog protocols of the audio-processors [168]. 104 non-users (duration of use < 1 hour per day) (1.04 %) were identified, 83 (0.83 %) as partial users (duration



▶ **Fig. 57** Daily use of CI in hours. Non-User rate 2 %.



▶ **Fig. 58** Schooling in CI implanted children in comparison to normal hearing peers (ISCED(Illg et al., 2017).



▶ **Fig. 59** Skill-Levels of CI recipients implanted in childhood compared to normal hearing peers (Illg et al. 2017).

of use = 1.5 hours). Among these 187 patients, compliance problems became obvious mainly in cases of early and long-term hearing loss (32.6 %) and of asymmetric hearing performance and single-sided deafness (13.4 %). Further reasons were discomfort during use or deception about the course and the results of hearing rehabilitation. The mean, time of daily use was 1.73 hours (0–5.4, n = 115). The mean monosyllable ward score was 15.5 % (0–100; n = 170), the mean age at time of investigation was 34.4 years (1–84; n = 187). The reasons for a reduced or nonuse of a cochlear implant may be manifold. Compliance problems in general are difficult to predict, to identify, and finally also to solve which makes a multiprofessional approach very important. The analysis of individual courses and the identification of risk groups are significant for prevention of nonuse. High-risk patients in this context are people suffering from early and long-term hearing loss as well as asymmetric hearing. Non-users being asked about the permanent refuse of the CI reported about multiple reasons [169]. The main reasons were poor auditory performance and the development of an identity as deaf person. In cases of children with single-sided deafness and CI, non-users have a significantly longer duration of hearing loss compared to users [170]. Children with additional disabilities are prone.to non use. About 27 percent of the children who have autism spectrum disorders in addition to the hearing impairment do not continue using their CI later on [171]. Adolescent hearing impaired people do not want to receive a CI during puberty. Furthermore, false expectations and false promises, social pressure for example in school, or psychological problems play a crucial role regarding the use.

8. Future developments

The major progress in cochlear implant technology led to good results in hearing rehabilitation. However, the results show a significant variability which is due to several factors. Besides the well-known demographic factors, comorbidities and additional disabilities play an important role. The information transmission capacity at the electrode-nerve interface is of importance which is determined by the individual functional condition of the auditory nerve as well as the number of effective electrode channels.

The focus of future development will be the realization of the bionic ear (▶ **Fig. 60**) with a best possible restoration of hearing through reconstruction of physiological hearing based on technology solutions.

Significant components of this bionic ear are an improved electrode-nerve interface for restoration of a near to normal physiological stimulation pattern of the auditory nerve, the regeneration of the peripheral auditory system by biological therapies, and the adequate use of the information transmission channels by an physiology based speech processing strategy.

The way to bionic hearing will be characterized by numerous intermediate steps and includes the following areas (▶ **Table 10**):

8.1 Improvement of hearing preservation

It is generally possible to preserve the residual hearing in cochlear implantation and to use it for hybrid hearing systems. However, the rate of good hearing preservation is still far poorer than interventions like stapes surgery. This objective implies that the mechanics of electrode array as well as the surgical technique and the post-traumatic biological reaction in the cochlea have to be addressed. The last-mentioned aspect is already approved by drug-eluting electrodes. Currently, dexamethasone-eluting electrode arrays are under clinical evaluation. First results indicate a significant reduction of the trauma reaction as indicated by lower impedances and better hearing preservation.

The mechanical properties of the electrode arrays can be improved in order to exactly adapt to the individual cochlear anatomy. For this purpose, additive manufacturing of electrode carriers is one possible way of realization of mechanically active electrodes.

The third factor focuses on improving the surgical technique by applying robotic insertion systems that insert the electrodes exactly in the cochlea with low speed along precalculated insertion trajectories.

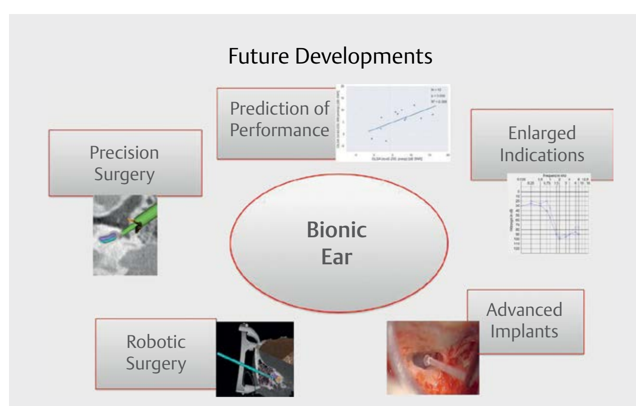
8.2 Improvement of the electrode-nerve interface

By positioning the electrode carrier at the modiolus, the required stimulus intensities can be significantly reduced which leads to a reduction of the electric spread of excitation. This may be achieved by adaptive electrode systems that change their shape after insertion and are able for example by uptake of perilymph via polymer biomorphs to bend and thus to adapt exactly to the modiolus. Nitinol-based systems are another potential solution. A different approach is the so-called auditory nerve implant consisting of an electrode pad with many electrode pins (up to 96) that penetrate directly into the auditory nerve (▶ **Fig. 61**). In this way, a direct stimulation of auditory nerve fibers is possible with simultaneous reduction of the stimulation current and improvement of channel separation.

Advanced implants additionally use intracochlear biological factors like growth factors or stem cells to improve the electrode-nerve- interface.

8.3 Regeneration of the auditory nerve

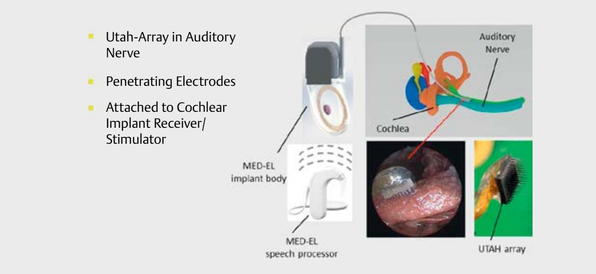
Another element for improvement of the information transmission capacity is the regeneration of the auditory nerve. By releasing nerve growth factors as well as the application of electrodes with cell linings for autoproduction of these nerve growth factors, peripheral dendrites from the spiral ganglion cells will grow onto the functionalized electrode. This direct nerve connection significant-



▶ **Fig. 60** Areas of future CI development towards Bionic Hearing.

► **Table 10** Future developments – Bionic Hearing.

1. Improvement of hearing preservation rates
2. Improvement of the electrode-nerve interface
3. Regeneration of the auditory nerve
4. Development of hybrid stimulation systems
5. Speech processing strategies
6. Closed-loop systems and brain-computer interfaces
7. Hearing device of the future with integrated multi-sensor technology
8. Cochlear implants as personal communicator
9. Invisible hearing – totally implantable CI (TICI)

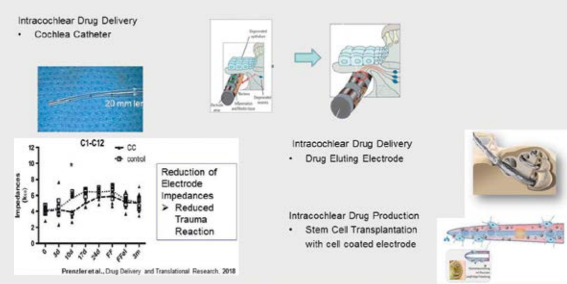


► **Fig. 61** Auditory Nerve Implant to improve electrode-nerve interface through direct intraneural electrical stimulation.

ly improves the specificity and selectively of electrical stimulation and thus the channel separation so that electrode systems with higher numbers of channels may be realized (► **Fig. 62**).

8.4 Development of hybrid stimulation systems

For electroacoustic stimulation, alternatively electromechanical and electrooptical systems are possible that are integrated into the implant system. In this way, universal stimulators can be realized for the inner ear allowing an optimal use of the peripheral residual hearing and the functional properties of the auditory nerve at any time, depending on the future development of hearing loss.



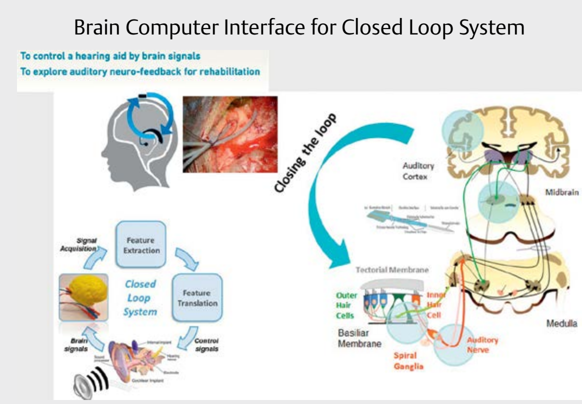
► **Fig. 62** Advanced Cochlear Implants with added biological components to reduce trauma reaction, improve hearing preservation and induce regeneration of dendrites of the auditory nerve.

8.5 Speech processing strategies

The improved electrode-nerve interface allows far and better speech processing strategies, e. g. the algorithm used to translate the acoustic signal into a logical sequence of electrical pulses for the cochlear implant system. The improved electrode-nerve interface with a higher number of electrically separated channels allow to use advanced speech processing strategies to increase the information transmitted through the electrode-nerve interface , such as spectral contrast, and an imitation of physiological stimulation patterns of the auditory nerve. By means of suitable modelling of the individual current spread in the cochlea, the best combinations of electrode contacts for stimulation may be determined.

8.6 Closed-loop systems and brain-computer interfaces

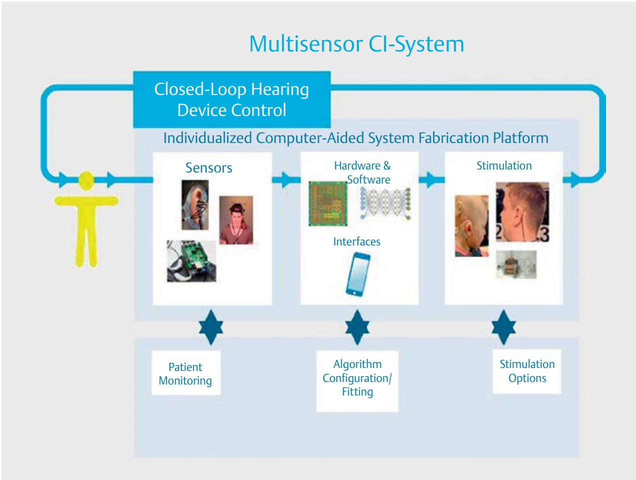
The central processing of the auditory information coded by the peripheral electrical stimulation can be recorded appropriately via integrated EEG electrodes of the CI system. These signals can then be used by the implant (► **Fig. 63**) to optimize speech coding algorithms for best possible auditory performance depending on the individual hearing situation.



► **Fig. 63** CI as brain-computer-Interface: Closed-Loop Systems with EEG-Feedback for automated optimization of speech coding strategy, control of auditory performance and speech and language development in children.

8.7 Hearing device of the future with integrated multi-sensor technology

The assessment of numerous additional parameters by means of the hearing implant opens the possibility for example to identify and diagnose movement disorders associated with vestibular lesions. Additional electrodes can be placed in the vestibular system to compensate balance disorders as well as to register biochemical changes in the perilymph for example in connection with inflammatory processes. These multisensory systems further allow the measurement of numerous parameters like pulse rate, oxygen saturation etc. Thus, the cochlear implant becomes a health monitoring system (► **Fig. 64**).



▶ **Fig. 64** Multisensor CI System for recording of additional biomarkers such as vestibular responses (permission by cluster of excellence Hearing4All).

8.8 Cochlear implants as personal communicators

By integrating the cochlear implant in a universal communication system, the potentials of audiotechnology and telecommunication can be fully used through the bionic ear.

8.9 Invisible hearing – Totally Implantable CI (TICI)

Due to progress in battery and microphone technology, totally implantable hearing systems are meanwhile possible. The power is supplied by transcutaneously chargeable batteries. The sound is registered via subcutaneous microphones. If needed, an external speech processor may be coupled in addition. Currently, battery running times of about 10–15 years are considered as realistic. The patients gain further freedom of action and lose the stigma of disability. However, hardware upgrades are only possible by reimplantation

Conclusion

Today, cochlear implants are a key method for auditory rehabilitation of severe and profound hearing impaired patients. The rapid development of implant technology led to significant improvement of the hearing results. About 80 percent of the patients are able to use the telephone and children achieve a near to normal hearing and speech development. All this led to a broader spectrum of indications including patients suffering from high frequency hearing loss and single-sided deafness. Currently, however, only about 60,000 of approximately one million possible CI candidates in Germany are implanted. In the future, multimodal universal hearing implants for combined electro-mechanical stimulation will be available allowing a continuous adaptation of the stimulation strategy to the functional status of the inner ear and the auditory nerve, especially in cases of progressive hearing loss. Brain-computer interfaces give input for the automated adaptation of speech coding to the hearing situation and an optimization of the signal processing to achieve the best possible auditory performance. Binaural hearing systems will improve directional hearing and speech in noise understanding. Advanced implants are composed of additively manufactured individualized electrodes that actively adapt to the anatomy of the cochlea after atraumatic insertion using robotic assistance. Depending on the pathophysiology, they are equipped with integrated biological components, support the preservation of residual hearing and induce the regeneration of neural elements to improve the electrode-nerve interface. In this way, the general limits of CI technology may be overcome and shifted towards physiological hearing. The bionic ear is within reach. Due to the consequent development towards simplification of the therapy, hearing preservation implantation under local anesthesia, and application of robotic systems, even more patients will benefit from the new physiological hearing option.

Conflict of interest

The authors declare that they have no conflict of interest.

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