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S2k Guideline

Cochlear Implantation

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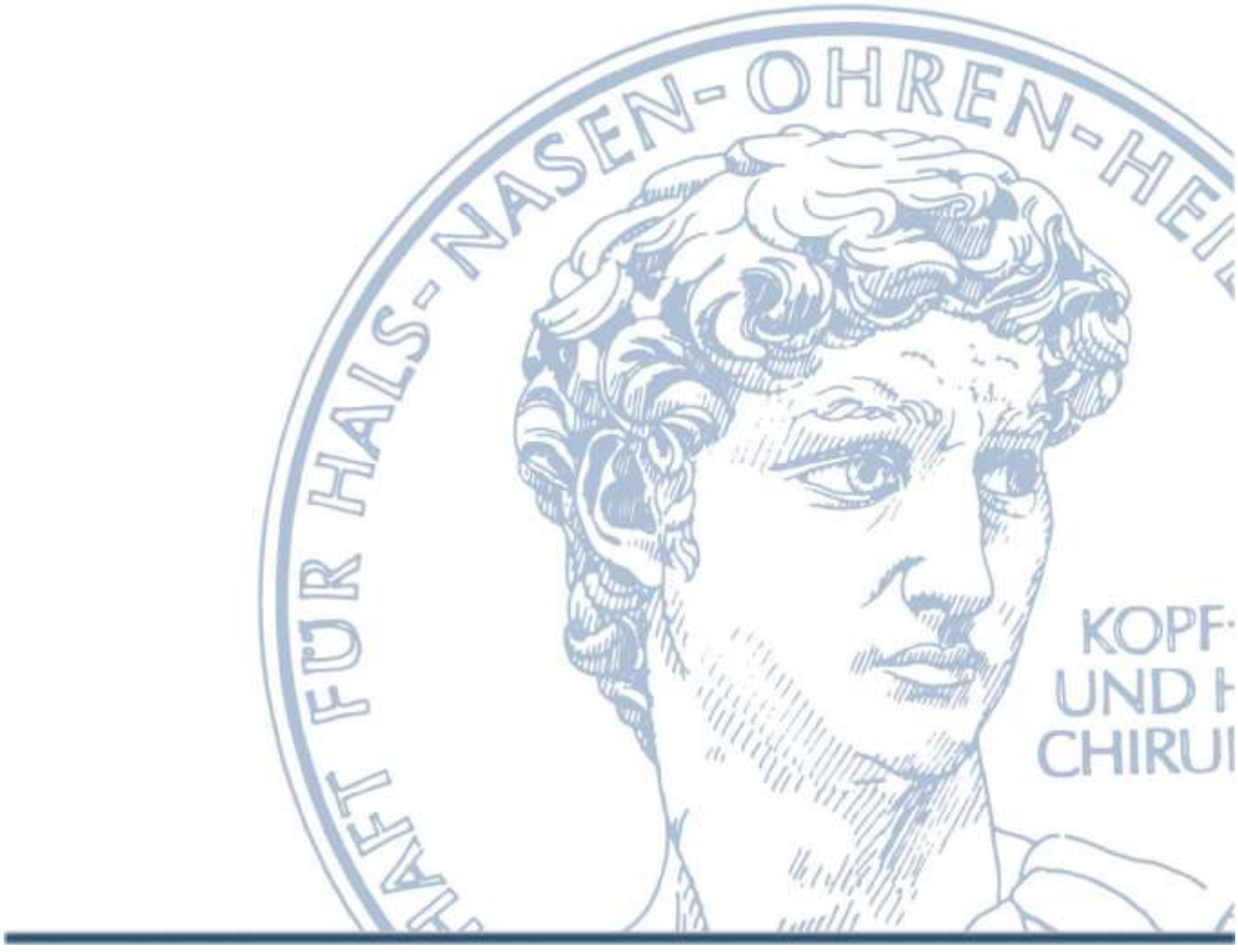
German Society of Oto-Rhino-Laryngology, Head and Neck Surgery.



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in collaboration with

- Arbeitsgemeinschaft für Cochlear Implant Rehabilitation (Working Group Cochlear Implant (Re)Habilitation, ACIR)
- Berufsverband Deutscher Hörgeschädigten-Pädagogen (Professional Association of German Hearing Impaired Educators, BDH)
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Contents

1 Abstract

The objective of this S2 Guideline is to promote high-quality care for people with profound congenital and acquired hearing loss or deafness. The therapeutic objective in adults is to restore hearing with cochlear implants when sufficient hearing for spoken communication cannot be achieved with conventional hearing aids, bone conduction hearing aids, or implantable hearing aids. The therapeutic goal objective in children is to initiate auditory development and hence to bring about the conditions to acquire spoken language through hearing.

The Guideline establishes quality assurance criteria for the entire process of cochlear implantation under the guidance of centres qualified for this purpose (CI-implanting institutions). Preparation of a separate guideline on the implantation process for central auditory implants is currently being planned.

2 Objectives and target groups of the Guideline

The goal of this Guideline is to promote high-quality specialist care for people with congenital and/or acquired hearing loss and deafness to restore hearing, where a comprehensive ENT specialist assessment has found that better speech understanding and hearing is likely to be achieved with cochlear implants than with other hearing systems.

The "Cochlear Implantation" Guideline is committed to the ideal of a respectful interaction between physicians, technical experts, audiologists, therapeutic specialists and patients "at eye level". The target groups therefore are all professions involved in the diagnosis and implantation process, as well as persons affected[1].

The "Cochlear Implantation" Guideline covers pre-operative diagnostics, indication, contraindications, surgical phase, basic therapy (initial fitting phase), follow-up therapy (CI rehabilitation) and long-term follow-up in children, adolescents and adults. At the same time, the required prerequisites for the quality of structure, process and results are described.

The following contains recommendations for cochlear implantation. The prerequisites and processes necessary for quality-assured interdisciplinary care in normal cases and those useful in particular cases will be described.

This should lead to a rational use of the procedure based on indication, highly specialized surgery, postoperative basic and follow-up therapy, including rehabilitation and aftercare.

[1] ** The wording for professions and occupational groups includes all genders (m/f/d). This applies throughout the entire Guideline. The masculine form is used here as well as in the following for better readability only.*

The Guideline was revised under the guidance of the German Society of Oto-Rhino-Laryngology, Head and Neck Surgery for improved, integrated and life-long cochlear implantation, care including all in- and outpatient parts on level S2 and has undergone an interdisciplinary consensus process, with patient associations also involved in all work phases. A total of five consensus conferences were convened for this purpose. The consensus conferences produced an evidence-based guideline, with a uniform recommendation based on a systematic literature search and the own experience of participating experts. It addresses all disciplines and fields involved and provides guidance for persons affected and their families. The Guidelines is seeking to use generally understandable language as far as possible. All professions are encouraged to explain technical terms that are not part of commonly language, to patient associations upon request, if needed.

3 Methodology and literature research

Guidelines are systematically developed recommendations that form the basis for joint decisions by physicians and their patients on sensible health care in particular cases.

The Cochlear Implantation Guideline has been prepared in line with the methodological guidelines for the development of guidelines for diagnostics and therapy of the Association of the Scientific Medical Societies in Germany (AWMF) and according to the AWMF's 3-stage concept corresponds to a S2k Guideline. Studies registered in PubMed or in the Cochrane Library between January 1990 and May 2018 were taken into account.

These classifications refer exclusively to the demonstrability of recommendations with the help of published studies. The aspect of the practical significance of a recommendation must be strictly separated from this.

It is necessary to reach a consensus so as to generate acceptance for a guideline where existing "evidence" is limited and to support dissemination and implementation. A combined procedure of multiple nominal group processes and Delphi method was used as the consensus procedure. The participants in the further consensus process are identical with the authors of the Guideline (see Appendix).

3.1 Procedure in detail - protocol of the Guideline revision

The German Society of Oto-Rhino-Laryngology, Head and Neck Surgery (DGHNO-KHC) commissioned the coordinator (Prof. Dr. med. Thomas Zahnert) in his function as chairman of ADANO (Working Group of German-speaking Audiologists, Neurootologists and Otologists) to update the 2012 Cochlear Implant Guideline.

This Guideline is an update of the S2k Guideline "Cochlea- Implantat Versorgung einschließlich zentral-auditorischer Implantate" (Cochlear implantation, including central auditory implants), published by DGHNO-KHC in 2012.

On 1 March 2018, the coordinator invited the guideline group to submit recommendations on the revision of the current Guideline. The individual working groups then prepared editorial recommendations for a revision of the current Guideline in an initial group process, taking into account the selected literature. These recommendations were submitted to the guideline coordinator and initially fully incorporated into the new guideline manuscript. In addition, following the incorporation of the editorial suggestions, specific questions were defined by the guideline coordinator and on 30 July 2018, individual experts were assigned to work on them. The recommendations made were sent to the coordinator and were also incorporated in the Guideline manuscript. This was used as a basis to issue general recommendations for cochlear implantation by the guideline coordinator.

The preliminary manuscript was discussed and revised in consensus conferences under neutral moderation by Dr. Sitter on 3 September 2018, 23 November 2018, 17 January 2019, 21 March 2019, and 25 April 2019, and were reconciled in a subsequent Delphi process (completed on 1 April 2020).

In addition, declarations of interest were collected according to AWMF specifications, and none of the members of the consensus group had to be excluded from participation due to conflicts of interest.

At the consensus conferences, all recommendations were discussed, modified as necessary, and subsequently adopted. Each proposed change was discussed, a draft prepared, and finally voted on.

It was generally possible to introduce proposed amendments into the procedure even after the prepared version had been sent out. These were given to all participants in the consensus conference for their information and reconciliation. Reconciliation generally took place during the meetings.

The final edited version was sent to all participants for written reconciliation with the participating organizations and a final written vote was requested.

Once all votes had been received, a final version was sent to AWMF for evaluation and, where required, publication, taking into account and, where required, citing divergent positions of the participating organizations.

3.2 Selection criteria and value judgements citing scientific evidence.

The recommendations represent a synthesis of the publications identified by the authors. The literature search in MEDLINE was performed via PubMed, the website of the US National Library of Medicine (http://www.ncbi.nlm.nih.gov/PubMed/), until and including Nov. 2018. The Cochrane Library (status: Nov.

2018) was also searched for relevant reviews. Guidelines from other countries and societies from GIN (guidelines international networks), evaluated according to DELBI.

Recommendations: The recommendations based on consensus are indicated as linguistic recommendations in accordance with AWMF guidelines and are shown together with the respective consensus strength (see Fig. 1).

Consensus strength	Description
>95%	strong consensus
>75%	consensus

ling. recommendation	Description
should/should not	strong recommendation
should/should not	moderated recomm.
may be considered/may be dispensed with	open recommendation

Fig. 1: AWMF consensus strength and linguistic recommendations

4 Definitions

4.1 Sensorineural hearing loss

Sensorineural hearing loss means hearing disorders caused by damage to the inner ear and/or auditory nerve.

In this Guideline, deafness and severe hearing loss are defined as hearing disorders that, according to the current state of science and technology, can be expected to be treated more successfully with a cochlear implant than with other forms of therapy.

4.2 The cochlear implant (CI) and CI implantation

The cochlear implant (CI, inner ear prosthesis) enables auditory perceptions by way of electrical stimulation of the still functioning auditory nerve. Usually, the hearing achieved in this way is better compared to other therapy methods in cases of severe sensorineural hearing loss or deafness. It can be used on one or both sides.

A microphone is used to record signals, which are processed in the CI processor and currently sent wirelessly transcutaneously to the implant (receiver and stimulator). The decoded signal effects the stimulation of different auditory nerve sections via a defined electrical stimulation.

Electric-acoustic stimulation (EAS) or hybrid implantation combines acoustic stimulation of a hearing aid, typically in the apical region, with electrical stimulation of the basal portions of the cochlea by a CI.

Since stimulation conditions vary greatly between patients, the CI processor must be fitted individually and repeatedly according to hearing development.

The cochlear implant usually is implanted behind the ear, under the skin in a bed of bone. The electrode array is inserted into the cochlea.

4.3 Implantation process

The CI implantation process covers the phases of pre-operative evaluation and consultation, surgery (implantation) as well as initial fitting phase (basic therapy), follow-up therapy (CI rehabilitation), and culminates in (long-term) follow-up care, which is provided by the implanting institution. The treating fields (otolaryngology, neuroradiology, phoniatrics and paediatric audiology, neuropediatrics, etc.) have the relevant professional responsibility for the technical aspects of the respective partial components of the CI implantation process. The medical director (usually an otolaryngologist) of the CI implanting institution is in charge of process coordination and has the final overall responsibility.

4.3.1 Diagnostics

The diagnosis of hearing loss, its classification by degree and type, requires special expertise and the use of appropriate methods. In addition to an otolaryngological and, as the case may be, paediatric audiological examination, this includes subjective and objective audiometric procedures, audiological recording and assessment of the current situation, imaging diagnostics and - if required - an educational, logopedic and psychological assessment of the potential for rehabilitation, including the psychosocial situation. Special characteristics in children and adults are considered separately

4.3.2 Surgical Phase

Due to technological development and increasing experience, cochlear implantation today has reached a level of complexity where implantation is allowed only at appropriately qualified institutions with an interdisciplinary team of experts and adequate technical equipment. The quality indicators for the quality of structure, process and results are of special significance (see Ch. 11).

4.3.3 Basic therapy/initial fitting phase

Basic therapy includes the postoperative phase of medical treatment as well as the audiological phase of starting up the CI processor. A phase of hearing therapy and, as the case may be, of speech therapy may be added as part of the basic therapy for adults. In children, basic hearing and speech therapy is obligatory. To guarantee a high level of quality results in the interdisciplinary implantation process, the initial fitting should be done at the CI-implanting institution. While the basic audiological therapy is provided by the CI-implanting institution, the basic hearing therapy and speech therapy for adults can be provided in cooperation with CI rehabilitation institutions.

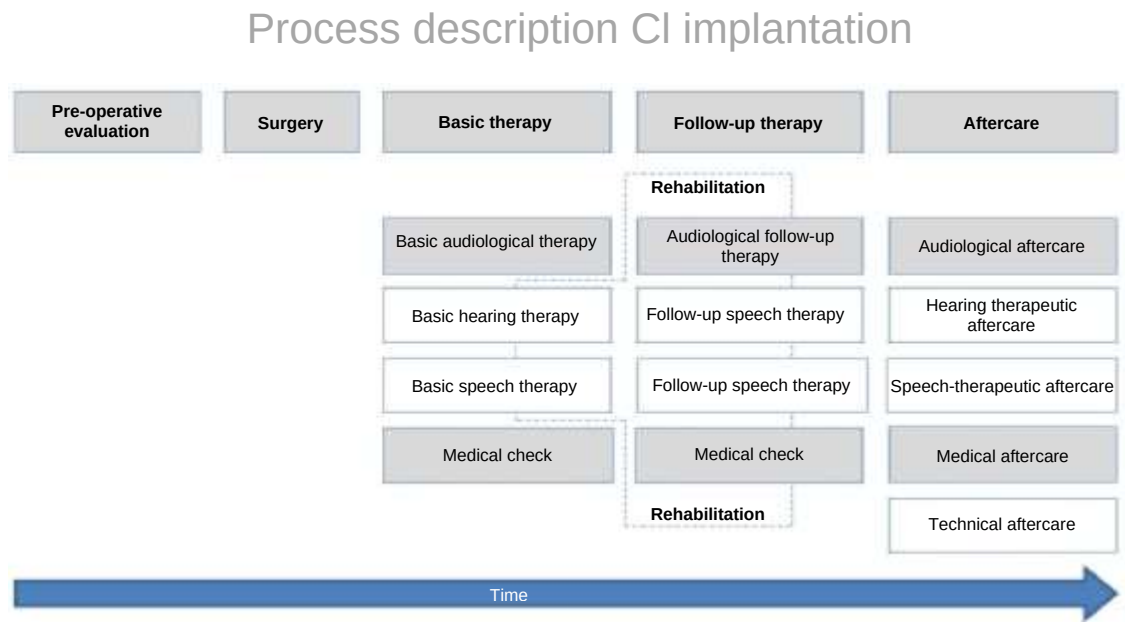
4.3.4 Follow-up therapy/CI rehabilitation

The basic therapy (initial fitting phase) is followed by the follow-up therapy phase. This phase aims at the optimal use of the implant and it comprises a range of fields, e.g. at least audiological, hearing therapy, speech therapy and medical therapy. This process may take place as a follow-up therapy or as a rehabilitation measure. The fitting of the CI processor in combination with a comprehensive auditory and speech therapy forms the critical basis for the success of treatment. Special expertise, comprehensive diagnostic procedures and an adequate spatial situation are indispensable for this purpose. Out- or inpatient forms of care also come into play. The choice is made based on the required or expedient intensity of treatment and on local as well as social factors.

Following rehabilitation measures, further therapeutic parts of the follow-up therapy phase may be indicated (see Fig. 2).

4.3.5 Aftercare

Current CI implantation requires lifelong follow-up care (long-term follow-up) under the responsibility of a CI-implanting institution. Aftercare begins after completion of follow-up therapy. Aftercare is designed to ensure the function of the CI system and hearing result, as well as social inclusion and participation. Where possible, this should be done close to the patient's residence, if the relevant expertise is available there. It is used for medical-technical and audiological checks and consultation as well as for pedagogical-therapeutic assessment and consultation, including documentation with the objective of stabilizing and optimizing the individual's ability to communicate. If indicated, the patient is returned to the follow-up therapy phase for renewed therapeutic measures (e.g. follow-up speech therapy) or renewed CI rehabilitation. Follow-up care must be provided for as long as the implant is used. This also includes required technical upgrades and checks and, as the case may be, re-implantations at the CI-implanting clinic.



Gray background: Non-delegable process parts of CI implantation at a CI implanting institution
- White background: Potentially delegable process parts of CI implantation at a CI implanting institution (with cooperation agreement)

Fig. 2: CI implantation process, terminology, also see Weißbuch (White Paper) of DGHNO-KHC on CI implantation

Patients receiving a cochlear implant are entitled to pre-diagnosis, therapy and usually, lifelong aftercare at highly specialized and qualified institutions. To ensure and maintain continuity of the implantation process, these qualified institutions should comply with the following requirements: They

- undertake to comply with this Guideline,

- participate in a national or European CI registry yet to be established (DGHNO- KHC) and submit an annual report, including statistics on surgical results and complications,

- maintain a comprehensive, complete, up-to-date range of implantation options to allow for an informed choice of the procedure most suitable for the individual to be made together with the patient,

- throughout the entire implantation process, work as part of a multidisciplinary team of experts, using adequate technical equipment,

- ensure for the entire team - e.g. by way of continuing education and participation in pertinent conferences - that everyone involved in CI implantation is at all times at the cutting edge of science,

- seek and maintain cooperation with patient self-help groups, and

- maintain an active, transparent and effective quality management.

To describe the institutions involved in the CI implantation process, a definition is provided below:

4.3.6 CI implanting institution

The term a CI-implanting institution generally is understood to mean a main ENT inpatient ward with own beds, where pre-operative diagnostics and indication, surgery, at least basic audiological therapy and follow-up therapy, as well as medical checks are performed as non-delegable services in the implantation process (see Fig. 2). Follow-up audiological therapy/CI rehabilitation and aftercare also take place at CI-implanting institutions, but may also be provided at specialized rehabilitation facilities and aftercare centres (see Chapters 9.3 and 9.4 as well as Ch. 10). The institution has the necessary specific structural, spatial, personnel, technical equipment and devices to be able to guarantee the CI implantation process.

4.3.7 CI rehabilitation institutions

The CI rehabilitation institution is an institution which provides the CI rehabilitation measures/follow-up therapy in the implantation process according to structural and substantive criteria (see chapters 9.3, 9.4, 10).

4.3.8 Combined institutions

Some CI-implanting institutions have an internal organizational unit for CI rehabilitation.

5. Pre-operative diagnostics and preparation for surgery

Children, adolescents and adults each require different consultation and therapy concepts.

Before and after surgery, detailed consultation with the affected persons and their relatives takes place under the responsibility of the CI-implanting institution. The detailed contents for children and adults are explained in detail in the following chapters.

For the individually required components of interdisciplinary diagnostics, the treating fields (otolaryngology and, if necessary, neuroradiology, audiology, phoniatrics and pedaudiology, neuropediatrics; therapeutic specialists, etc.) bear the respective professional responsibility. Interdisciplinary diagnostics are performed according to the cutting edge of science and form the basis for the indication by the CI-implanting institution. The medical director of this institution has the final overall responsibility for the implantation process.

5.1 Pre-diagnostics and surgery preparation in adults

For pre-diagnostics in adults, the following findings should be collected:

5.1.1 Medical history and clinical examinations

- General status, medical history, including ENT-specific history,
- ENT status, including eardrum microscopy.

5.1.2 Sound and speech audiometry

- Sound audiometry with air and bone conduction hearing threshold (separate for each side, with documentation of masking noise levels, according to DIN EN ISO 8253-1),
- Speech audiometry (Freiburg speech intelligibility test, according to DIN 45621) measured via air conduction transducers (headphones or plug-in earphones) (separate for each side, with documentation of the masking noise levels, according to DIN EN ISO 8253-3). Determination of:
    - single syllable understanding from 65 dB SPL up to tolerance limit (mEV/dBopt),
    - hearing loss for numerals
    - as the case may be, maximum understanding of numerals.
    - as the case may be, measuring at levels 60 dB, 80 dB, 100 dB SPL.

5.1.3 Speech audiometry in noise

Selection of at least one of the test procedures listed below, depending on the patient's residual hearing:

- Hochmair-Schulz-Moser (HSM) sentence test in quiet and with noise,
- Oldenburg sentence test in quiet and in noise,
- Göttingen sentence test,
- matrix test in the patient's respective native language for comparative determination of speech understanding.

5.1.4 Hearing aid check

- Technical check:
  - visual inspection of hearing aids and otoplastics, as the case may be,
  - simple function check by way of listening,
  - checking the amplification setting of the main or everyday program against the target curves determined based on the key audiological data at different input levels (ISTS signal) as a probe-style microphone/in-situ measurement or with a measurement box.

- Audiometric check in the free field, measured monaurally or separate for each side:
  - Freiburg monosyllabic test at 65 dB SPL,
    - ♦ as the case may be, for discrimination function: measuring at 50 dB SPL and 80 dB SPL,
  - as the case may be, sentence test in speech simulating ambient noise (S0N0),
  - as the case may be, sentence test in noise in different spatial hearing situations,
  - as the case may be, directional hearing (S0N0),
  - as the case may be, categorical loudness scaling with special narrowband noise pursuant to DIN ISO 16832 with mid-range frequencies at 0.5, 1, 2 and 4 kHz,
  - as the case may be, effective resting hearing threshold (inflation curve) and discomfort threshold with hearing aid,
  - if required: hearing aid optimization

5.1.5 Extra-cochlear electrical stimulation of the auditory nerve

Performing the promontory test (Kuo and Gibson, 2002; Lee et al. 2007; Nikolopoulos et al. 2000; Lesinski-Schiedat et al. 1997; Kileny et al. 1994) if medical reasons exist to suspect damage to the auditory nerve or no residual hearing can be audiometrically detected. If retro-cochlear and/or central hearing disorders is suspected, E-BERA and, if possible, E- CERA should be performed (see "Objective hearing tests").

5.1.6 Evaluation of hearing-related quality of life

- Evaluation of hearing-related quality of life (e.g. structured medical history) or with standardized questionnaire inventories (e.g. Nijmegen-Fragebogen, Abbreviated Profile of Hearing Aid benefit (APHAB), Speech, Spatial and Qualities of Hearing Scale (SSQ)).

- For patients with tinnitus: tinnitus diagnostics (matching, masking, recording the level of tinnitus impairment, tinnitus questionnaire, scaling).

- As the case may be, measuring the listening efforts (e.g. SSQ, Listening Effort Questionnaire).

5.1.7 Objective hearing tests

- Impedanceometry:
  - tympanogram,
  - ipsi- and contralateral stapedius reflex thresholds at 0.5, 1, 2, 4 kHz.

- Otoacoustic emissions (DPOAE at least at f2 = 1; 2; 3; 4; 6 kHz, L1 = 65 dB, L2 = 55 dB).

- Electric response audiometry (ERA):
  - Brainstem ERA (BERA), stimulus clicking sequence (Click):
    - ♦ threshold determination,
    - ♦ suprathreshold retrocochlear diagnostics (determination of conduction times according to Jewett.

  In case of EAS/hybrid indication:
  - low-frequency BERA (500 Hz range) with band-limited chirp signals, alternatively ASSR.

  In case of irregular hearing curve and/or suspected psychogenic hearing loss:
  - auditory steady-state response (ASSR) to estimate hearing threshold response; or another frequency-specific electrophysiological method with test frequencies at 0.5, 1, 2, 4 kHz.

- To rule out auditory neuropathy/synaptopathy (e.g. incongruence of sound and speech audiogram):
  - electrocochleography (ECochG).

- In case of suspected damage to the auditory nerve (e.g. after traumatic brain injury with fracture of the petrous bone, after meningitis where the auditory nerve or facial nerves are affected, if a vestibular schwannoma is detected and, as the case may be, after therapy) or in case of a suspected congenital malfunction or reduced function of the auditory nerve (aplasia, hypoplasia):

- E-BERA with needle electrode, placement on promontory is possible.

5.1.8 Balance diagnostics

- Labyrinth function testing, including caloric testing and/or V-KIT (video head impulse test),
- As the case may be, cVEMPs,
- Additional tests, depending on the vertigo history, clinical and diagnostic findings.

5.1.9 Imaging diagnostics

Imaging diagnostics are used to determine the pathologies of cochlea, mastoid and middle ear, auditory nerve, cerebellopontine angle, and, as the case may be, the auditory pathway. Currently, high-resolution petrous bone CT/DVT and MRI generally are performed in coordination with the neuroradiology department. To assess cochlea and the inner auditory canal, the smallest possible layer thicknesses should be selected, and in any case less than 1 mm (Biller 2007).

Multiplanar reconstructions (MPR) should be created in the three spatial directions. The MRI examination should prepare an axial flair sequence of the entire cerebrum as well as T1-weighted images before and after paramagnetic contrast enhancement with a slice thickness not exceeding 2 to 3 mm. Moreover, a heavily T2-weighted 3-D sequence should be created so as to visualize the neural structures of the inner auditory canal as well as the fluid-filled structures of the inner ear. Depending on the further technical development of the method, the sequences best suited to answer the diagnostic question are to be used in each case. Examinations should be assessed by a radiologist experienced in petrous bone imaging, ideally in the field of neuroradiology. An interdisciplinary case conference is desirable.

Patients who are deaf due to meningitis are at risk of connective tissue or osseous obliteration of the cochlea within a few months. Such cases require CI implantation or close follow-up of the fluid-filled cochlea by way of MRI in the short-term. In particular cases, it may be sensible to use functional imaging techniques to demonstrate that auditory nerve and pathways are functioning, with simultaneous functional activation by way of acoustic and / or electrical stimulation of the auditory nerve. This includes functional MRI, positron emission tomography (PET), and near-infrared spectroscopy (NIRS).

5.1.10 Further examinations and pre-operative measures

Obligatory:

- Examination of speech, speech development, voice (e.g. by phoniatricians and pediatric audiologists, otolaryngologists, in collaboration with speech therapists), expectations, motivation, learning ability, and the patient's psychosocial situation as criteria for the use of the cochlear implant,

- Vaccination recommendation in line with the current STIKO recommendations.

Optional:

- As the case may be, pedagogical and psychological, possibly gerontopsychiatric examinations regarding the ability to communicate.

- As the case may be, consultative examinations regarding clarification risk factors (ability to undergo surgery, possibly prognostically relevant comorbidities).

- As the case may be, neurological consultative examinations regarding clarification partially impaired brain capacity and rehabilitation capability in case of neurological disorders.

5.1.11 In-depth personal consultation and information

In pre-operative consultation and diagnostics, the individual prerequisites, predispositions, possibilities and needs of each individual patient must be considered. In so doing, it must be taken into account at all times that the patient has independently decided to have a CI implant and that they support the general decisions of the implantation process in an informed manner. This also means that all parties involved promote the patient's ability to consent and competence. They clearly show in consultation and therapy the scope for the patient in the various phases of implantation.
Patients generally undergo consultation and receive information on:

- explanation of the implantation process (pre- and post-operative phase, including rehabilitation),
- function and mode of action of a CI,
- different CI systems: the consultation is non-proprietary. The patient is thus provided with criteria to make a selection for their individual situation,
- presentation of alternative forms of treatment,
- differences from hearing aids and option to integrate in existing hearing systems,
- operative approach and risks in surgery,
- prospects of success of CI implantation,
- clarification and, as the case may be, adjustment of expectations,
- individual therapy planning,
- provision of contacts with self-help groups of CI-users or parents of CI-users,
- limitations and risks of CI implantations.

5.1.12 Basic in-/outpatient conditions for pre-diagnostics

Preliminary examinations in adults may be performed on an out- or inpatient basis. The need of admission as an inpatient results from the foreseeable intensity of the examinations, medical necessities and from local and social circumstances.

5.2 Pre-diagnostics and surgery preparation in children

Compared to adults, there are differences in pre-diagnostics and surgery preparation in case of children, which must be taken into account by way of phoniatric, pediatric audiological diagnostics and in the planning of surgery.

In this regard, reference is generally made to the S2k Guideline "Periphere Hörstörungen im Kindesalter" (Peripheral childhood hearing disorders) (http://www.awmf.org/leitlinien/aktuelle-leitlinien/ll-liste/deutsche-gesellschaft-fuerphoniatrie-und- paedaudiologie-ev.html).

The subsequent core modules must be applied specifically, depending on the child's age and developmental stage:

5.2.1 Medical history and clinical examination

Depending on the age of the child, a detailed medical history is taken, including family history, by interviewing the parents. The clinical diagnosis is performed in an interdisciplinary manner and includes the complete ENT findings, which potentially may need to be supplemented by further clinical examinations in the case of syndromal hearing disorders. Ear microscopy is of particular importance, since pathological middle ear processes and their causes, e.g.

otitis media with effusion in case of enlarged adenoid vegetations, should be remediated prior to CI implantation. Depending on the child's age and ability to cooperate, it may be necessary or expedient to perform the examination in parts or in its entirety in one session under anaesthesia and, as the case may be, supplement it with paracentesis as well as adenotomy. Where possible, this should be done in one session in case of infants and toddlers.

5.2.2 Subjective hearing test procedures

- Determining the frequency-dependent auditory threshold (if possible, separate for each side, (Dettman et al. 2004), according to age and developmental stage by way of reflex audiometry, response audiometry, deflection audiometry, with visual conditioning or play audiometry,
- Speech audiometry (according to the stage of speech development in older children with peri- or postlingual hearing loss), using headphones or plug-in earphones, if possible separate for each side:
    - in quiet (e.g. Mainz child speech test, Göttingen child speech test, Oldenburg child speech test [OlKi] or Oldenburg child sentence test [OlKiSa]),
    - possibly in noise (by way of OlKiSa).

5.2.3 Objective hearing and balance tests

- Impedanceometry (stimulus adapted to age):
  - tympanogram,
  - ipsi- and contralateral stapedius reflex thresholds at 0.5, 1, 2, 4 kHz.

- Otoacoustic emissions (TEOAE and DPOAE at least at f2 = 1; 2; 3; 4; 6 kHz, L1 = 65 dB, L2 = 55 dB).

- Electric response audiometry (ERA):
  - frequency-specific BERA (e.g., notched-noise, chirp, ASSR) with test frequencies at 0.5, 1, 2, 4 kHz and click ABR to determine auditory thresholds and conduction times.

- To exclude auditory neuropathy/synaptopathy (e.g. in the presence of detectable OAE):
  - electrocochleography (ECochG).

- In case of suspected damage to the auditory nerve (e.g. after traumatic brain injury with fracture of the petrous bone, after meningitis where the auditory nerve or facial nerves are affected, if a vestibular schwannoma is detected and, as the case may be, after therapy) or in case of a suspected congenital malfunction or reduced function of the auditory nerve (aplasia, hypoplasia): Damage to the auditory nerve (e.g. after traumatic brain injury with fracture of the petrous bone, after meningitis where the auditory nerve or facial nerves are affected, in case of a suspected a congenital malfunction or reduced function of the auditory nerve (aplasia, hypoplasia):
  - E-BERA with needle electrode, placement on promontory is possible.

- Balance diagnostics depending on age and developmental stage in case of abnormalities in sensory-motor development (e.g. cVEMPs, oVEMPs, KIT or V-KIT).

5.2.4 Imaging diagnostics

See Chapter 5.1.9. Diagnostic imaging of children usually is performed with sedation/under anesthesia.

5.2.5 Evaluation of hearing system wear test

In case of residual hearing and severe hearing loss, a wear test with conventional hearing aids or bone conduction systems is usually first required, and the results have to be assessed within 3 months.

- Objective evaluation:

- visual inspection of hearing aids and otoplastics,
- simple function check by way of listening,
- checking the amplification setting against the target curves determined based on the key audiological data at different input levels (ISTS signal) as a probe-style microphone/in-situ measurement or with a measurement box.

- Subjective evaluation in the free sound field, where, possible separate for each side (Eisenberg et al. 2004; Steffens and Hacker, 2009; Zichner, 2012):

  - threshold for reaction ("functional gain"),
  - as the case may be, speech audiometry, adapted to development:

    - in quiet (e.g. by way of Mainz child speech test, Göttingen child speech test, Oldenburg child speech test [OlKi], Oldenburg child sentence test [OlKiSa]),
    - possibly in noise (by way of OlKiSa).

5.2.6 Speech development diagnostics and communication competence

All children and adolescents generally are assessed in terms of speech development and communication competence by way of standardized and normalized procedures adapted to their age and developmental stage, and in the case of children previously fitted with hearing aids, according to their hearing age (Kim et al. 2010; Lamprecht-Dinnesen et al. 2002). Depending on the developmental stage, pre-linguistic developmental stages as well as the level of speech must be assessed development systematically receptive as well as expressive on all linguistic levels (phonetic-phonological, lexical-semantic, morphological-syntactic, communicative-pragmatic) using standardised and normalised methods and deviations from the age norm must be documented.

Among other things, the following is to be used to record the child's overall situation:

- Parent questionnaires to record pre-speech and early speech development, especially in the first or second year of life, e.g.:

  - LittlEARS (Kühn-Inacker et al. 2003),
  - Parent questionnaires for the early identification of children at risk (ELFRA; Grimm and Doil 2006),
  - Functioning After Cochlear Implantation (FAPCI, German version; Lin et al. 2007, Grugel et al. 2009),
  - Early Speech Production Questionnaire (LEESPQ; Keilmann et al. 2018).

- Direct procedures for assessing speech development, starting at 24 months, e.g.:

- Sprachentwicklungstest für zweijährige Kinder (speech development test for children at the age of two; SETK-2; Grimm, 2006),
- Sprachentwicklungstest für drei- bis fünfjährige Kinder (speech development test for children at the age of three to five; SET3-5; Grimm, 2010),

- Professional assessments of the fields involved in care and support of a child (e.g., early intervention, education for the hearing impaired, speech therapy (de Raeve 2010a).

In case of deficits in communication skills, the parents or adolescents concerned must be offered alternative communication aids. For further information, please refer to the S2k guideline "Diagnostik von Sprachentwicklungsstörungen (SES) unter Berücksichtigung umschriebener Sprachentwicklungsstörungen (USES)" (Diagnostics of developmental speech disorders (SES) considering specific speech development disorders (USES)" (de Langen-Müller et al. 2011).

5.2.7 Developmental-psychological status, assessment of socio-familial situation and rehabilitation capacity

In addition, the developmental status should be recorded as part of the holistic treatment in interdisciplinary cooperation (e.g. paediatrician, early intervention centres, SPC, kindergarten, schools, etc.) and evaluated by the CI-implanting institution/CI rehabilitation institution with regard to the significance for hearing rehabilitation. Since cognitive development is a significant predictor of speech outcome (Edwards et al. 2006) and complex interactions between neurocognitive processes, auditory, speech, and articulatory development (Xueman, 2016), as well as psychosocial factors (Theunissen et al. 2014) affect a child's overall development, a psychometric testing algorithm is useful as part of the preoperative diagnostic process and throughout the rehabilitation process with the following objectives.

Possible aspects here are:

- early detection of developmental risks,
- identifying developmental disorders (e.g. partially impaired brain capacity),
- creating a support concept adapted to the development profile and circumstances of the child/adolescent,
- initiating support measures,
- as the case may be, initiating additional neuro-developmental examinations,
- as the case may be, initiating accompanying support measures.

Depending on the underlying problem as well as on age or developmental stage, possible instruments are listed for recording for the following developmental areas:

- Psychomotorics:
    - Direct methods, e.g., Entwicklungstest für Kinder von 6 Monaten bis 6 Jahren, revidierte Fassung (developmental test for children 6 months to 6 years, revised edition; ET 6-6 R; Petermann & Macha, 2013), Bayley Scales of Infant and Toddler Development, 3rd edition (Bayley-3; Renner & Rosenkranz, 2014).
    - Indirect methods, e.g. Elternfragebögen zur ergänzenden Entwicklungsbeurteilung bei den kinderärztlichen Vorsorgeuntersuchungen (parent questionnaires for supplemental developmental assessment at paediatric checkups; EEE U6-U9; Petermann & Macha, 2003).

- Cognition:
    - Direct methods, e.g. Bayley-III-scales (1-42 months), Snijders-Oomen Non-verbaler Intelligenztest, revidierte Fassung (Snijders-Oomen nonverbal intelligence tests, revised edition; SON-R 2-8; Tellegen et al. 2018), SON-R 6-40 (Tellegen et al. 2012), Kaufman-Assessment Battery for Children-II (K-ABC-II, Kaufman, 2015), Wechsler Preschool and Primary Scales of Intelligence-IV (WPPSI-IV; Hrsg. Petermann et al. 2018), Wechsler Intelligence Scales for Children-V (WISC-V, ed. Petermann, 2017).
    - Indirect methods, e.g. Fragebogen zur Erfassung kognitiver Prozesse bei 4- bis 6Jährigen (Questionnaire for the assessment of cognitive processes in 4- to 6-year-olds; KOPKI 4-6; Gleissner et al. 2011).

- Social-emotionale development:
    - Direct methods, e.g., behavioral observation.
    - Indirect methods, e.g. Child Behavior Checklist, Elternfragebogen für Klein- und Vorschulkinder (Parent questionnaire for toddlers and pre-school children; CBCL 1 ½-5; Achenbach et al. 2000), CBCL, Eltern- und Lehrerfragebogen über das Verhalten von Kindern und Jugendlichen (parent and teacher questionnaires on the behavior of children and adolescents; CBCL 6-18R or TRF-R; Achenbach et al. 2014).
- The child's parents/close caregivers coping with the illness:
    - Direct methods: standardized psychopathological assessment.
    - Indirect methods, e.g. Disabkids zur Selbst- und Fremdbeurteilung der gesundheitsbezogenen Lebensqualität (4-7 Jahre) (Disabkids for self- and third-party assessment of health-related quality of life (4-7 years)), Kindl-R, Selbst- und Fremdbeurteilung der gesundheitsbezogenen Lebensqualität (3-17 Jahre) (Kindl-R, self- and third-party assessment of health-related quality of life (3-17 years)), KID Screen, Selbst- und Fremdbeurteilung der gesundheitsbezogenen Lebensqualität in

verschiedenen Sprachen (8-18 Jahre) (KID Screen, self- and other-assessment of health-related quality of life in different languages (8-18 years)); www.kidscreen.org.

5.2.8 In-depth personal consultation with parents, children and adolescents regarding CI

In-depth personal consultation with parents, children and adolescents should include the following points:

- process of CI implantation,
- presentation of different CI systems
- (re-)habilitation (concept: inpatient vs. outpatient), importance of parent-child interaction (Rüter 2011, Szagun and Stumper 2012, Reichmuth 2017),
- realistic prospects of success (Boons et al. 2010; Geers, 2006; Niparko et al. 2010),
- individual therapy planning (duration, sequence),
- information and contact with affected persons,
- reference to alternatives to CI implantation (Kim et al. 2010),
- consultation regarding issues of social law (e.g. disabled person's card).

The situation of deaf parents must be given special consideration in consultation. It must be ensured that communication is as barrier-free as possible. Especially, it must also be made clear to deaf parents that they retain responsibility for development of their children and that they relinquish this responsibility to a degree they can understand when it comes to promoting their children's speech development.

6. Indications for surgery

The indication for CI surgery is made by the surgeon, taking into account all pre-diagnostic findings. Cochlear implantation is an option for patients where cochlear implants are likely to provide better hearing and speech understanding than with conventional hearing aids or other implantable hearing systems.

The indication is determined separately for each ear affected by the hearing disorder.

Based on preliminary examinations it should be assumed that auditory nerve and pathway are functioning. Bilateral implantation should be aimed for in case of bilateral indication (Laszig et al. 2004). A suspected obliterative labyrinthitis is an indication for urgent CI implantation.
The indication for CI implantation in children and adults takes into account both audiological criteria as well as the expected improvement in communicative abilities and social inclusion.

According to the diagnostic possibilities in children and adults, these two groups should generally be distinguished.

6.1 Postlingually deaf (after speech acquisition) children, adolescents, and adults with residual hearing

Based on fitting hearing aid (optimized setting) or alternatively, of another hearing system, the literature supports a CI indication from an audiological perspective for one ear if the measured monaural single syllable understanding (Freiburg Speech Intelligibility Test according to DIN 46521) in the free sound field at a speech level of 65 dB SPL is ≤ 60% (Hoppe et al. 2015, Hoppe et al. 2017, Leigh et al. 2017, Plant et al. 2015).

Reason:

Even though the single syllable understanding of CI users varies widely between individuals and ranges only between 50% and 70% at 65 dB SPL (Hoppe et al. 2017, Krüger et al. 2008, Leigh et al. 2016, Plant et al. 2015, Zeh and Baumann 2015), in case of ears with speech understanding that is still measurable pre-surgery (mEV > 0%), above-average CI hearing compared to the general population can be expected (Gifford et al. 2010, Holden et al. 2013, Hoppe et al.2017, Lenarz et al. 2009, Hoppe et al. 2019). Moreover, the result can be expected to be worse if the deprivation period of an ear significantly exceeds more than five years (Dowel 2016). Expert assessment of the quality of fitting a hearing aid is of great importance in the pre-surgery evaluation of CI candidates.

When assessing the fitting of hearing aids, monaural single-syllable understanding in the free sound field at a speech level of 65 dB SPL (EV65) should not deviate significantly from maximum single-syllable understanding (mEV). However, if this is the case, an expert (HG acoustician, CI audiologist) should check whether the setting of the hearing aid is sufficient or whether individual reasons are responsible for the underperformance (Müller-Deile and Hoppe 2018). (Hoppe et al. 2014, McRackan et al. 2016, McRackan et al. 2018, Müller et al. 2016).

Statement: In postlingually deaf patients, there is no fixed time limit with regard to the general option of cochlear implantation. Even after decades of deafness, successful implantation is possible in postlingually deaf patients (strong consensus).

RECOMMENDATION: In children, adolescents, and adults who are postlingually deaf or have residual hearing, the audiologic CI indication should be met if monaural single-syllable understanding measured with a hearing aid in the free sound field at a speech level of 65 dB SPL is ≤ 60% (strong consensus).

CI implantation may be recommended even after many years of deafness (strong consensus).

6.2 Prelingually (before speech acquisition) deaf adults

In selected cases, implantation may be indicated in prelingually deaf adults (bilaterally if necessary).

It has been assumed already since the 1980s that CI implantation in prelingually deaf adults is beneficial and promotes individual communication skills (Eisenberg 1982). Even in a group of pre- and postlingually deaf patients studied by Hinderink et al., 1995, nearly all prelingual CI users (n = 8) were subjectively satisfied with their achieved communication improvements; even though the moderate speech discrimination attained did not reach the level of postlingually deaf patients (n = 11).

In a more recent paper studying 43 prelingually deaf CI users, Eisenberg's assessment is conformed that CI implantation in prelingually deaf patients can be useful: 88% of the studied prelingually deaf CI users showed significant improvement in speech understanding, whereby a shorter time interval until implantation and some residual hearing were shown to be beneficial (Rousset et al. 2016). In another study, nearly 80% of patients diagnosed as prelingually deaf before the age of 2 who were fitted with an CI implant as adults benefited after surgery from an albeit limited, yet still improved speech understanding of up to 25% monosyllables (Lammers et al. 2018). Only 21% of the prelingually deaf CI users did not show any improvement in daily communication. This subgroup is feared to be at increased risk of discontinuing CI use and becoming "non-users."

Expressive speech also improves in prelingually deaf patients who received an implant at a late stage (Splitthoff et al. 2013). This draws attention to the fact that in this group of patients, not only the best possible post-surgery speech understanding should be considered a "success", but improved speech production, improved acoustic orientation and improvements in communication also contribute to individual patient satisfaction and therefore to individual "success" and compensation for disability.

The indication in this group of patients requires a complex evaluation of interdisciplinary findings, including as key aspects an assessment of hearing ability, hearing history, description of the speech developmental status and of the speech/speaking status, of communicative and speech competencies, as well as of general communication skills. Socio-familial aspects, including the assessment of the intention to use speech and the viability of hearing/speech rehabilitation are additional criteria to be considered for the indication of CI implantation in this group of patients.

RECOMMENDATION: Cochlear implantation may also be indicated in prelingually deaf adults (strong consensus).

6.3 Prelingually deaf as well as perilingually (during speech acquisition) deaf children and children with residual hearing

Children and adolescents with unilateral or bilateral severe or complete loss of hearing, usually of cochlear origin, who are likely to achieve better hearing and speech understanding with CI than with conventional hearing aids or implantable hearing systems are eligible for CI implantation. The objective of implanting CI in children is to achieve successful communication skills using speech and open speech understanding up to age-appropriate receptive and expressive speech performance.

The indication requires a complex phoniatric, pediatric audiological evaluation of interdisciplinary findings, including as key aspects a frequency-specific assessment of the hearing ability as well as a description of the level of speech development, communicative competencies and general developmental state. Socio-familial aspects, including the assessment of the parents' intention to use speech and the viability of hearing/speech rehabilitation are additional key criteria for the assessment of the prospects of success of CI implantation in children.

With regard to the hearing ability, the threshold for a possible CI indication has evolved from a complete loss of hearing to a hearing threshold > 70 dB HL, whereby these findings should be evaluated in synopsis with the other factors already described (Leigh et al. 2011; Vickers, Summerfield, and Lovett 2015).

RECOMMENDATION: In children with a hearing loss > 70 dB HL, the prerequisite for CI implantation should be assessed; in addition to audiological criteria, the evaluation of the stage of speech development, communicative competencies, the general developmental status, as well as socio-familial aspects should also be considered (consensus).

In case of bilateral hearing, if the indication criteria for cochlear implantation are met, simultaneous bilateral implantation should be aimed for in children, to allow for the potential of binaural hearing with regard to auditory processing and perception to develop as early as possible (Sparreboom et al. 2015). Simultaneous bilateral CI implantation requires increased effort compared to unilateral implantation, both in terms of surgery and also in the subsequent implantation.

Where the hearing ability differs in both sides and in case of severe hearing disorders with residual hearing abilits in both ears and stagnation of speech acquisition, usually, implantation is first limited to one site so as to check in the further course whether optimal implantation is achieved with bimodal implantation with unilateral CI and a contralateral conventional hearing system or whether implantation on the other ear should be carried out in the near term in the sense of a sequential bilateral implantation. Other reasons for sequential implantation include, inter alia, anesthesiologic risks, cochlear abnormalities, multiple disabilities with limitations in the child's cognitive performance, or parental concerns. In principle, the indication for CI implantation should be assessed separately for each ear and, in the case of primarily bimodal implantation, should be periodically re-evaluated (Sadadcharam et al. 2016).

RECOMMENDATION: Bilateral implantation (simultaneous or sequential) should be performed for bilateral CI indications (strong consensus).

According to the current state of knowledge, one or two cochlear implants in congenitally deaf children is favoured as early as possible after completion of the paediatric audiological confirmation diagnostics (Bruijnzeel et al. 2016; Ching et al. 2017; Kral et al. 2017). The prerequisite is that the results of the frequency-specific confirmation diagnostics performed in the context of an abnormal hearing screening in newborns are reliable and that retardations, e.g. in case of premature babies or central co-morbidities are taken into account. In case of a borderline CI indication (relevant residual hearing), a sufficient wearing test with conventional hearing instruments is necessary before a decision is made regarding CI implantation so as to ascertain whether method is sufficient for adequate hearing/speech development. In addition, in the course of decision-making, the parents' acceptance of the diagnosis, their informed consent to the further course of treatment, and socio-familial aspects, in particular the assessment of the parents' intention to use speech in primarily sign language-oriented environments and other cultural groups, as well as the viability of hearing/speech rehabilitation, must be considered (Bruijnzeel et al. 2017). In addition, when planning implantation in the first year of life, the anesthesiologic risk must also be assessed and an appropriate anesthesiologic assessment must be obtained (Kim et al 2017).

In case of infants with confirmed bilateral deafness, the overall assessment of all those aspects may allow CI implantation in the second half of the first year of life. In case of children with severe hearing disorders, but with relevant residual hearing and speech acquisition initiated by conventional hearing aids, rigorous paediatric audiological follow-up examinations are required in early childhood, so as to initiate a pathway for bimodal or bilateral CI implantation in the near term, as the case may be, depending on the development of hearing and speech.

RECOMMENDATION: In case of infants with confirmed bilateral deafness, CI implantation should take place in the first year of life if indicated (strong consensus).

In children with unilateral deafness and mild to moderate loss of hearing on the opposite side, a bimodal pathway with unilateral CI implantation of the deaf ear and contralateral hearing system usually is indicated. In many cases, conventional hearing systems fitted in the better ear indeed achieve speech understanding in quiet. However, in situations with high levels of ambient noise, which is common in the daily lives of children, speech understanding deteriorates rapidly, with the effect that these children with only conventional hearing systems often develop communication distress and social limitations (Sadadcharam et al. 2016).

In case of existing and stable residual hearing in the low frequency range, special implant systems with suitable electrode arrays are used in childhood, analogous to CI implantation of adults, for both electric and acoustic stimulation, when residual hearing is preserved (EAS). These children indeed initially in many cases benefit from fitting conventional hearing systems due to their ability to hear in the low

frequency range. However, the EAS option should be considered early in cases of stagnating speech acquisition and articulation of disorders due to loss of hearing in the high frequency range.

Studies on children using EAS regarding the prognosis of low frequency hearing in the long-term do not yet allow for a conclusive assessment, but based on smaller groups of patient, data for post-surgery hearing preservation and proportional hearing deterioration for the first months post surgery are available already (Rader et al. 2018). Studies of adult CI users demonstrate the potential for medium- and long-term hearing preservation in the low-frequency range (Helbig et al. 2016, Mertens et al. 2014, Roland et al. 2018). Surgical techniques preserving the ability to hear (hearing preservation cochlear implantation = HPCI) are generally recommended in children, regardless of residual hearing, in order to preserve the inner ear structures for the option of electroacoustic stimulation or for future regenerative treatment methods (Rajan et al. 2017).

RECOMMENDATION: In children, atraumatic surgical techniques that may preserve residual hearing should be used to preserve the possibility of electroacoustic stimulation (consensus).

More than 40% of severely hearing-impaired children with sensorineural hearing loss have additional disorders or additional disorders can be expected during childhood development. CI implantation can be supported where the child's communicative competence can be expected to improve and there are no contraindications. In the case of multiple disabilities, a modified objective of CI implantation must be formulated in interdisciplinary consensus, based on the individual developmental prerequisites of the child. The focus is a positive impact on the quality of life and social involvement as parameters of success, since open speech comprehension and successful speech development cannot always be expected in children with multiple disabilities.

In cases of bacterial meningitis, a rigorous assessment of the loss of hearing and a neuroradiologic follow-up of the cochlea are required in childhood as well, so as to implant in the near term, if possible before the onset of labyrinth obliteration, and thus to achieve the most complete electrode insertion possible.

6.4 Single-sided deafness (SSD), asymmetric hearing loss (AHL)

Another indication is for single-sided deafness (SSD) with CI indication with and without tinnitus (van de Heyning et al 2008; Arndt et al. 2011b). In adults, the indication should be determined based on extensive consultation, audiological evaluation (van de Heyning et al. 2016) and testing of treatment alternatives, such as CROS/BiCROS. In patients with SSD following labyrinthitis (Hassepass et al. 2013) with incipient obliteration/ossification, there is an urgent indication for CI implantation in analogy to postmeningitic bilateral deafness.

In children, detailed consultation, audiologic evaluation, and, in particular, MRI examination are also necessary, since in up to 50% of cases with congenital unilateral deafness, hypo or aplasia of the auditory nerve may be present (Arndt et al. 2015, Usami et al. 2017), making CI implantation impossible or questionable. In these cases, CROS may be used, as the case may be, also together with bone-anchored hearing systems. A trial of CROS/BiCROS hearing aids is not usually performed in children and is not indicated in the context of hearing and speech development (Beck et al. 2017). Sufficiently reflective use of CROS/BiCROS hearing aids usually is conceivable from adolescence, at the earliest.

In adults, the study situation regarding treatment success after CI implantation in SSD, but also AHL (asymmetric hearing loss, with single-sided deafness and hearing loss on the other side) overall is positive. Significant positive effects were established for sound localization and speech understanding in noise by a binaural integration of electrical stimulation by the CI and acoustic stimulation of the normal hearing or low-to-moderate hearing-impaired ear of the other side (Arndt et al. 2011a and b, Jakob et al. 2011, Tavora-Vieira et al. 2015, Grossmann et al. 2016, Hoth et al. 2016, Arndt et al. 2017, Rahne et al. 2016, Holder et al. 2017, Mertens et al. 2017, Döge et al. 2017, Finke et al. 2017a and b, Louza et al. 2017, Buss et al. 2018, Litovsky et al. 2018).

In a large proportion of patients, electrical stimulation by the CI may lead to permanent tinnitus suppression as a side effect of auditory rehabilitation (van de Heyning et al. 2008, Punte et al. 2011, Arndt et al. 2011a and b, Sladen et al. 2017).
Likewise, recent studies show an improvement in quality of life after CI implantation (Rösli et al. 2015, Härkönen et al. 2015, Mertens et al. 2015, Friedmann et al. 2016, Dillon et al. 2017, Finke et al. 2017b, Arndt et al. 2017).

Special cases of single-sided deafness such as Meniere's disease, post-acoustic neuroma resection or intralabyrinthine schwannomas are also successfully treated with cochlear implants (Hansen et al. 2013, Doobe et al. 2015, Hassepass et al. 2016, Aschendorff et al. 2017, Schipper et al. 2017, Plontke et al. 2018).

To date there are only a few studies on children with congenital SSD, and comprehensive long-term experience are not yet available. However, the available publications all show a positive effect of CI implantation (Arndt et al. 2015, Rahne et al. 2016, Friedmann et al. 2016, Beck et al. 2017, Greaver et al. 2017, Thomas et al. 2017, Sladen et al. 2017). The outcome has to be assessed based on the age at the time of implantation (Polonenko et al. 2017), with the effect that, also based on animal studies (Kral et al. 2013, Gordon et al. 2015), implantation at an early stage analogous to bilateral congenital deafness must be recommended. In these cases, differentiated consultation with parents should be provided with information about limitations of monaural hearing and sensitive time windows for speech acquisition for adequate cortical representation of speech and offering single-sided CI implantation as a treatment option. In case of SSD acquired in childhood, results are comparable to those in adults (Arndt et al. 2015). Communicative skills can be expected to improve with single-sided CI implantation in the near term.

RECOMMENDATION: For adults and children with SSD (single-sided deafness) or AHL (asymmetric hearing loss with single-sided deafness and hearing loss on the other side), cochlear implantation should be recommended. The indication should not be made without prior imaging diagnostic of the auditory pathway and consultation in cases of proven significant limitations in communication in everyday life (consensus).

In patients with SSD and tinnitus, CI implantation may be recommended for tinnitus suppression (consensus).

6.5 Auditory synaptopathy/neuropathy

In case of a pathological disorder beyond the outer hair cells, in the area of the inner hair cells, synapses or spiral ganglia in the sense of an auditory synaptopathy/neuropathy, CI implantation may be useful if no positive effect can be realised with hearing aids in corresponding hearing loss (Walger et al. 2011). In addition to the objective hearing examination by means of OAE, FAEP and electrocochleography, diagnostics also include the precise imaging evaluation of the inner ear and auditory pathway structures (among other things, to exclude aplasia of the auditory nerve). A diagnostic algorithm is proposed by Harrison et al. (2015).

7. Contraindications

Absolute contraindications include abnormalities of cochlea, auditory nerve, and auditory pathway that preclude electrical stimulation. Other absolute contraindications include structural or patient-related barriers to continuing the CI implantation process after surgery. Relative contraindications include individual medical characteristics that complicate the implantation process. A relative contraindication means that the indication is critically assessed in the particular case.

7.1 Absolute contraindications for CI implantations

- Evidence of a missing cochlea or missing auditory nerve.
- Inability of the patient to participate in the CI implantation process in its entirety (among other things, basic therapy; rehabilitation, aftercare).
- No opportunity or no access to initial fitting, rehabilitation or aftercare (patient- or institution-based, see structural process).

7.2 Relative contraindications for CI implantation

- Middle ear infections (implantation possible after rehabilitation)
- Limited rehabilitation ability with CI implantation.
- Negative subjective promontory test depending on the results of further audiological diagnostics.

- Severe comorbidities that significantly interfere with the implantation process.
- Lack of evidence of the auditory nerve on imaging.

7.3 Age limits

In adults, there are no age-related limits as long as the conditions for successful completion of the overall implantation process are met, including CI rehabilitation. The recommended age for surgery in children is from 6 months of age (except for urgent medical indications, such as impending obliteration of the cochlea).

8. Surgical phase

8.1 Requirements of the surgeon and the CI-implanting institution

Prerequisites for the surgeon include many years of continuous experience in special microsurgery of the ear and previous surgical activity at a clinic specialized in CI surgery (ENT main department pursuant to chapter 4.3.6 of this Guideline) with an adequate number of CI surgeries under the supervision of an experienced CI surgeon.

The implanting clinic should publish the number of surgeries and complication statistics as part of a quality report and through data maintenance in a (yet to be established) national CI registry (see Chapter 0).

A sufficient number of implantations per year per surgeon should be performed at the CI-implanting institution so as to ensure routine and quality. While at this stage, there is no clear scientific evidence of a correlation between the number of surgeries and quality, it can be assumed that, in addition to other factors, the number of surgeries performed by the CI-implanting institution also affects the quality and outcome of cochlear implant surgery (Schulze-Gattermann 2002, Aschendorff et al. 2011).

At least two experienced surgeons should be available at the CI-implanting institutions, who meet the following requirements:

- Many years of continuous experience in special microsurgery of the ear.
- Previous surgical activity at a clinic specialized in CI surgery.
- Performance of an adequate number of CI surgeries under the supervision of an experienced CI surgeon.
- Use and knowledge of implants from different manufacturers.

The implanting institution should publish the number of surgeries as well as the number of surgeries per surgeon and complication statistics as part of a quality report. In addition, it must show that surgeons regularly undergo further training on CI implants from different manufacturers.

8.2 Special equipment features in the ENT OR

- Special surgical instruments for cochlear implantation,
- cochlear implants as well as reserve and special implants for at least 3 manufacturers,
- measuring station for intraoperative functional testing of the implant and of the auditory pathway of different manufacturers,
- intraoperative EMG monitoring of the N. facialis,
- intraoperative imaging options: intraoperative radiological position control of the electrode is not required in every case. It is sufficient if the technical and organizational prerequisites for such intraoperative control are available. Postoperative position control generally is not indicated if intraoperative control has already taken place (radiation hygiene),
- immediately and continuously available specialist consultant services in the following departments, usually main departments: anaesthesiology and intensive care medicine, neurosurgery, radiology, including neuroradiology, paediatrics; paediatric anaesthesiology and intensive care medicine.

8.3 Inpatient stay/length of stay

Implantation takes place during a clinic stay as an inpatient in a main ENT department. The length of stay is determined according to medical as well as social criteria relating to the patient, with checks for complications being required, in particular for wound healing (see chapter 8.4 and 8.5.2).

8.4 Risks of the procedure and complication management

CI surgery, performed in a CI-specialized clinic, is a low-risk surgical procedure. Any complications of the procedure should be manageable by the implanting clinic in interdisciplinary management (see chapter 4.3.2). Examples include:

- Middle ear infections (possible spread towards the inner ear and cerebrospinal fluid space, with the risk of meningitis, obliteration of the inner ear, possible damage to the afferent auditory nerve fibers),
- wound healing disorder,
- vertigo and balance disorders,
- facial nerve palsy, also permanent,
- taste disorder,
- tinnitus,

- loss of any residual hearing,
- technical implant failure,
- medical complications due to the implant,
- need to replace the implant,
- deterioration of electrical stimulability, e.g. due to progressive ossification of the cochlea after meningitis,
- incorrect positioning of electrodes,
- adverse stimulation effects of other cranial nerves (e.g., facial nerve, vestibular nerve),
- intolerance to materials of the implants,
- bleeding; dura and brain injury, cerebrospinal fluid fistula,
- failure or expected hearing outcome not realised,
- neuralgias, scar pains.

8.5 Intra-, peri- and postoperative measures

8.5.1 Intraoperative checks

- checking the implant function by way of telemetry,
- monitoring of the N. facialis,
- measurement of physiological responses to electrical and, as the case may be, acoustic stimulation (e.g. stapedius reflexes),
- neural responses (e.g., E-CAP measurements, etc.), especially in children,
- as the case may be, radiological check of the electrode position.

8.5.2 Postoperative checks

- radiological check of the electrode position,
- clinical, inpatient monitoring of the patient so as to detect and treat complications at an early stage (damage to the facial nerve, vertigo, wound healing disorder).

8.5.3 Perioperative antibiotic prophylaxis (PAP) in CI implantation

Cochlear implantation is an aseptic, surgical implantation with a risk of infection of the associated cerebrospinal fluid space due to the opening of the inner ear. This should be taken into account in consideration of the current AWMF-Leitlinie zur perioperativen Antibiotikaprophylaxe(AWMF guideline on perioperative antibiotic prophylaxis; AWMF register no. 029/022).

Specifically, this means that (regardless of age) prophylaxis penetrating the CNS should be given in the so-called "prophylaxis window". The first dose of antibiotic therefore should be administered well in

advance of the start of surgery (e.g. for parenteral administration of cephalosporins, 30-60 minutes before the start of surgery) A single course of antibiotics is usually sufficient for effective prophylaxis when the surgery takes less than 2 hours and is not inferior to multiple courses of antibiotics beyond that.

In case of additional risks, therapeutic antibiotics should be administered according to the literature (Anne et al. 2016), e.g.

- Chronic middle ear infection or mastoiditis.
- Patient risk factors:
    - age >70 years, colonization with staphylococcus aureus and MRSA, immunosuppression, underlying diseases such as diabetes mellitus,
    - dialysis requirement, reduced or poor general health reflected by a high ASA score (ASA score 4-5).
- Postoperative risk factors (repeat surgery or implant replacement, local wound infections).

Post-surgery, prolonged antibiotic use should be reviewed daily and, as the case may be, adjusted, depending on local wound conditions, general condition, and risk factors.

RECOMMENDATION: At least single administration of an antibiotic penetrating the CNS before surgery as preoperative antibiotic prophylaxis (strong consensus).

8.5.4 Systemic steroid therapy

Numerous studies exist on preserving hearing after CI implantation and on the use of steroids.

It is not yet possible to make a general decision for a specific corticosteroid. Overall, according to available studies, intracochlear and/or intratympanic administration seems to be preferable to systemic administration.

However, there are also studies that show a benefit of systemic corticosteroid therapy. There is indeed a high degree variability on the timing of use, the duration of therapy, and the corticosteroid used.

For example, Skarzynska et al. have described that a combined oral and intravenous corticosteroid therapy before, during, and after cochlear implantation has a significant effect on postoperative hearing preservation (Skarzynska et al. 2018).

Other research groups have shown in animal models (Kuthubuteen et al. 2018) that single-shot administration of dexamethasone has a significant effect on preserving hearing. Sweeney et al. 2015 find a significant effect on preserving hearing with oral administration of prednisolone if this is started as early as 3 days prior to surgery and is continued until two weeks after.

Prednisolone, hydrocortisone, triamcinolone and dexamethasone, for example, are used in various concentrations for both local and systemic administration.

Due to the multitude of available corticosteroids, a direct comparison as part of a (randomised) trial seems almost impossible.

A further limitation is that since the use of glucocorticoids for the treatment of acute sensorineural loss of hearing is not approved in Germany, the systemic application of glucocorticoids in the context of CI care as well as the local application is an "off-label".

According to the research, the level of evidence for the use of steroids peri- and/or postoperatively in CI surgery corresponds to level IIb to III.

It is not possible to infer from the currently available literature adequate evidence regarding the dose or the duration of therapy. As a general rule, it should be aimed for a high dosage for systemic application and a high concentration of corticosteroid for intratympanic application.

RECOMMENDATION: Preoperative, intraoperative, and postoperative systemic steroid therapy may be considered (strong consensus).

8.5.5 Local steroid therapy

Some CI centres argue that an intratympanic and intracochlear application of cortisone during CI surgery reduces adverse immune reactions and fibrosis and minimize intracochlear insertion trauma (Plontke et al. 2016, Lyu et al. 2018, Cho et al. 2016). Potentially, the stimulation of nonauditory neural structures may also be reduced, and the tonality and dynamics of auditory implants improved. The main advantages of direct application are considered to be the bypassing of the blood-brain barrier and the enhanced inner ear concentration with a smaller amount of drug.

Various methods were described for the application of cortisone and other medication in the middle and inner ear. In addition to manual intratympanic cortisone instillation, hydrogels containing cortisone (lactic acid-based polymer) and nanoparticles are available. The latter are also suitable for the intratympanic transport of proteins and nucleic acids for the purpose of activating intracochlear signalling pathways. Furthermore, the CI electrode as such may be used as a "drug delivery device" (Douchement et al. 2015, Stathopoulos et al. 2014, Bas et al. 2016) (coating or incorporation of drugs, application channel).

Individual studies were able to show that cortisone concentrations in the perilymph are higher following intratympanic administration than after systemic cortisone administration (Lyu 2018). Since due to pharmacokinetic reasons, the maximal steroid effect occurs with a time latency, it was recommended to apply cortisone 8-12 hours prior to CI surgery.

The vast majority of studies are prospective randomized studies in animal models. No meta-analyses of intraoperative cortisone administration in the context of CI implantation are available. A small number of non-randomized studies in humans exist. They do not justify drawing the conclusion that the intratympanic application of cortisone has either a positive or negative effect on the implantation outcomes. Hence there currently is no sufficient evidence to recommend or reject intratympanal cortisol administration in the context of CI implantation.

RECOMMENDATION: intravenous or intratympanic steroid therapy may be considered (strong consensus).

8.6 Reimplantation

Reimplantations and sequential, bilateral implantations in this regard equate to initial implantations. Bilateral implantation usually is associated with an increased effort. Reimplantation requires a renewed basic therapy (initial fitting phase) and subsequent follow-up therapy (CI rehabilitation).

9. Basic therapy (initial fitting phase) and follow-up therapy (CI rehabilitation)

In addition to the intensive preoperative diagnostics described above, basic therapy (initial fitting phase) and follow-up therapy (CI rehabilitation) are of great importance for the success of CI implantation. They are an integral component of CI implantation.

Cochlear implantation overall is a multidisciplinary approach that must be performed in institutions with adequate expertise. Basic therapy (initial fitting) should be performed in the implanting clinic.

For follow-up therapy (CI rehabilitation), the CI-implanting institution or appropriately qualified CI rehabilitation centers should also be available to provide CI rehabilitation in close cooperation with the CI-implanting institution. The service must be available all year-round and if required, in clinic for inpatients. The basic therapy is indicated and follow-up therapy and aftercare follow the structure of an interconnected process in a multidisciplinary team (see Chapter 10.2, Structural framework, also see Fig. 2). All disciplines are involved both in the daily processes of basic and follow-up therapy and also in seeing the patient as part of the aftercare and treat the patient together.

With successfully completed CI surgery, the CI-implanting institution initiates basic therapy, follow-up therapy, and aftercare.

9.1 Basic therapy (initial fitting phase) in adults

9.1.1 Start and duration

The basic therapy (initial fitting phase) starts in the period between the first day and 6 weeks post-surgery. The basic therapy in adults usually is performed on an inpatient basis (3 - 5 days). Basic therapy may also be provided on an outpatient basis under favourable social conditions and close structuring. Next, 40 days of follow-up therapy (CI rehabilitation) are usually required over a period of 6 - 24 months. In individual cases, there may be a need for a longer period. The therapy for adults can vary greatly in terms of the time required, depending on the individual medical history.

9.1.2 Contents of basic therapy (also see Fig. 2, page 13)

- medical aftercare
- initial setting of the CI processor and gradual optimization of the CI processor setting,
- initial hearing/speech therapy,
- as the case may be, comprehensive speech therapy diagnostics,
- speech therapy measures,
- technical and audiometric controls,
- audiometric hearing and speech tests (in quiet and, as the case may be, in noise),
- as the case may be, psychological support (including to resolve conflicts impeding the therapy),
- documentation and evaluation of the results,
- training in handling the CI system (care, maintenance, detection of faults) and in using accessory devices (e.g. telephone adapter, wireless transmission system, inducer or T-coil),
- relaying safety-related information (e.g. MRI, electrical equipment, diving),
- for further contents see: "Weißbuch CI-Versorgung der DGHNO" (DGHNO White Paper on CI implantation)

9.2 Basic therapy in children

9.2.1 Start and duration

The basic therapy (initial fitting phase) starts in the period between the first day and 6 weeks post-surgery. The subsequent follow-up therapy (CI rehabilitation) may continue until the age of 18 and usually comprises a period of 60 treatment days. Rehabilitation should continue at least until speech acquisition and written language acquisition have been completed. The follow-up therapy may overlap with the initial fitting phase.

There may also be a need for rehabilitation again later in life (e.g. adolescence, testing a new CI processor model, ...). In case of children with additional disabilities, more and longer rehabilitation effort is expected to be required.

9.2.2 Contents of basic therapy (initial fitting phase) (also see Fig. 2, page 13)

The basic therapy involves medical follow-up examinations, transmission of neurophysiological data determined intraoperatively as a basis for programming the CI processor, as well as initial hearing and speech therapy. The initial fitting and optimisation of CI processor settings require general conditions that are child- and age-appropriate (usually two people: audiologist and therapist) as well as the cooperation of audiologists, therapists, and parents/caregivers. The basic therapy (initial setting phase) and the follow-up therapy in infants usually take place under inpatient conditions with the involvement of the caregiver (usually a parent). An initial fitting phase may also be provided on an outpatient basis under favourable social conditions and similarly close structuring.

Contents and objectives:

- Medical checks: wound checks and follow-up by ENT specialists and phoniatrics/pediatric audiologists,

- Basic audiological therapy:

    - initial setting of the CI processor, taking into account the parameters determined intraoperatively,
    - gradual adjusting of the CI processor setting,
    - technical checks of CI processor and implant: visual inspection, functional check, impedance measurement, electrically evoked summation action potentials (ECAP),
    - checks of the subjective parameters (determination of threshold of minimum stimulation intensity (T-level), balancing of electrical stimulation of different electrodes (i.e. loudness balancing), possibly bilateral loudness balancing (Keilmann et al. 2009),

- relaying safety-related information (e.g. MRI, use of electrical equipment, diving),
- audiometric checks: (age-adjusted) sound hearing test (i.e. functional gain, separate for each side), age-adjusted speech audiometry in quiet and in noise (in case of bilateral fitting also separate for each side), possibly test of spatial hearing ability,
- training relatives in handling the CI system (operation, function and maintenance, care, detection of faults) and accessories (e.g. charger, drying device, other accessories of the basic equipment),
- training relatives in using accessory devices, wireless digital and inductive transmission equipment, wired use of external audio sources, use in water), if already relevant.

9.2.3 Initial hearing/speech therapy

If required, the hearing/speech therapy adapted to age and developmental stage also includes initiating communicative precursor skills (eye contact, turn-taking, shared focus, triangulated eye contact...) and the relevant guidance of parents / caregivers. The latter will be advised on their own behaviour to promote communication and speech. Parents/caregivers are informed and advised about hearing/speech and other areas of development in children. If required, where the child is not yet receiving early intervention or other support at home, this should be initiated. Criteria for the appropriate location for providing such support may be discussed already, if required.

9.3 Follow-up therapy/CI rehabilitation in adults

The main objective of follow-up therapy and rehabilitation of adult CI users is occupational and social inclusion, as well as the active possibility of spoken communication within a reasonable period of time. Consultation, surgery, initial fitting, follow-up therapy, and rehabilitation in adults usually should take place within 24 months to maintain patient motivation, reduce the psychological strain on patients and accelerate patient reintegration into work and society.

The therapy for adults can vary greatly in terms of the time required, depending on the individual medical history. The auditory perception subsequently must adapt to the new artificial stimulation or habituate with the effect that, depending on the achieved progress, a timely and closely interconnected combination of therapy and recurrent adjustments of the CI processor as well as audiometric checks are necessary.

The CI rehabilitation and follow-up therapy take place in institutions that are qualified for this purpose and are carried out by the CI-implanting clinic or by a CI rehabilitation centre. It includes careful checking of the implant and of all parts of the CI processor. The therapeutic measures started in the initial fitting phase are continued in CI follow-up therapy and CI rehabilitation, depending on the individual therapeutic progress. The follow-up CI therapy and CI rehabilitation may be performed on an outpatient, inpatient or day-patient basis. This will be decided based on the complexity of the treatment required and on the patient's social and health situation.

9.3.1 Contents of CI follow-up therapy and CI rehabilitation (see also Fig. 2, page 13)

- additional medical care,
- technical checks, gradual optimisation of the CI processor setting,
- intensive hearing/speech therapy,
- logopedic, phoniatric, educational and psychological diagnostics,
- audiometric hearing and speech tests (in quiet and in noise),
- consulting the patient and their social environment,
- psychological support,
- additional training in handling the CI system (care, maintenance, detection of faults) and in using accessory devices,
- documentation and evaluation of results during weekly team meetings,
- consultation with the social service on disability law and occupational integration.
- For further contents see: "Weißbuch CI-Versorgung der DGHNO-KHC" (DGHNO-KHC White Paper on CI implantation)

9.4 Follow-up therapy/CI rehabilitation in children

One main objective following a cochlear implantation in children is to establish hearing as an integral part of their lives, to develop and constantly, over years, improve their communication and speech skills, and to bring those skills as closely as possible to those of children with normal hearing.

The follow-up therapy (CI rehabilitation) is provided at CI rehabilitation centres that cooperate with CI-implanting institutions. The structural framework is, in particular, the multiprofessional environment. The child/adolescent will initially attend CIRehabilitation at shorter intervals. The further distribution of rehabilitation days must be determined depending on the individual development and may also require more intensive phases at a later stage. The rehabilitation in young children should continue at least until primary speech acquisition has been completed and should continue at regular intervals into school age. There are a minimum of 3 treatment sessions per (treatment) day, plus breaks.

The therapy concepts must ensure that the natural developmental conditions of hearing and spoken language are stimulated, supported and promoted. The hearing and speech therapy is based on the principles of hearing-based speech acquisition with an interdisciplinary approach in the sense of the bio-psycho-social model of the WHO so as to enable each child with an CI implant to gain the best possible (re-)habilitation success in terms of social participation and integration. Based on the International Classifications of Functioning, Disability and Health (ICF-CY classifications), the treatment objective includes the elimination of negative consequences of illnesses and the improvement in health-related quality of life.

The intensive involvement of parents and caregivers as well as of therapists and educators from supporting institutions in the therapeutic process is indispensable. The objective of hearing/speech therapy is to develop comprehensive hearing and speech skills.

9.4.1 Contents and objective of follow-up therapy/CI rehabilitation (see also Fig. 2, page 13)

Medical checks

- inspection of the scar and implant site,
- checking coils and fit of the magnet,
- reviewing all findings for plausibility.

Audiological follow-up therapy

- technical checks of the CI system (CI processor and implant):
    - visual inspection, functional check,
    - impedance measurement,
    - measurement of electrically evoked summation action potentials (ECAP).

- checking the subjective parameters:

    - determining the threshold value of the minimum stimulation intensity (T-level),
    - balancing the electrical stimulation of different electrodes (i.e. loudness balancing),
    - possibly bilateral loudness balancing (Keilmann et al. 2009).

- documentation of usage (if recorded by CI processor) and consultation,
- reviewing the wearing option,
- as the case may be, neurophysiological audiological diagnostics (auditory and cranial nerve potentials),
- replacement of defective components of the CI processor,
- children with residual hearing: regular monitoring of residual hearing as well as of the hearing ability in the contralateral ear,
- adjusting the CI processor setting based on audiological diagnostics,
- audiometric checks (age-adjusted),
- age-appropriate audiological diagnostics to test CI processor settings,

- repeating safety-related instructions and documentation of the instruction.

9.4.2 Follow-up hearing/speech therapy

The objective of hearing/speech therapy is to develop comprehensive hearing skills to initiate speech skills.

The therapy is carried out from a holistic perspective and is reliant on the child's parents or close contacts being involved in hearing and speech therapy. The discussion of the therapy and the guidance of the parents (legal guardians/relatives) is of particular importance for the success of the therapy for the therapy to continue in the home environment. The parents and guardians and/or caregivers are instructed to behave in a manner that supports hearing and speech appropriate to developmental stage and situation and hearing tactics.

They receive ongoing training in handling the CI processor, recognizing sources of faults and, as the case may be, eliminating them. If the CI system is not used throughout the entire "awake" day, parents are consulted on the importance of wearing the CI processor continuously.

Support to build auditory memory in very young children and monitoring progress of the ability of auditory discrimination before and after fitting in older children depending on speech acquisition are also components of hearing/speech therapy.

9.4.3 Consideration of pedagogical aspects in audiology

Auditory processing is tested using sounds, syllables, and words to classify the ability to discriminate. The development of phonetic-phonological progress in a child's speech development can be classified in this way against the background of its hearing experience. Moreover, the extent of auditory memory performance can be placed in the context of general speech development.

In case of children with additional disabilities or with more complex support needs, this may be used to classify auditory processing if standard evaluation procedures are not effective.

<u>Rhythmic-musical and mototherapeutic services:</u>

Gross and fine motor skills and rhythmic skills are the basis for speech development. The continued difficult hearing and speech conditions hence make it necessary to include rhythmic-musical and rhythmic-motor offers in addition to the specific hearing-speech therapy in children with CI implants.

<u>Promoting sign language:</u>

In particular cases, additional support in sign language may also be useful, especially in case of children of deaf parents who are using sign language, or if speech acquisition stagnates or if the parents explicitly

request both types of support. Parents will be referred to support groups for parents of children with CI implants/hearing-impaired children.

9.4.4 Cooperation within the network

A main component of a successful therapy is the cooperation of the CI-implanting institution with early intervention of hearing-impaired children, pre-schools, regular or special schools and other therapists working with the child or in the family (e.g. speech therapists, occupational therapists, physiotherapists, socio-pedagogical family help). In case of children with additional disabilities, coordination of the interdisciplinary therapy team (for example, cooperation with the social paediatric centres close to home) is required.

Special educational support (usually institutions focusing on hearing and communication in the form of early intervention and mobile services) is also required for children with CI and is the task of the school. CI rehabilitation does not replace - and cannot be replaced by - any special education support that may potentially be necessary. This is usually the task of the educators in the area of hearing impairment.

The basic therapy (initial fitting phase) is the task of the CI-implanting institution. The subsequent follow-up therapy (CI rehabilitation) should take place in the relevant specialized CI rehabilitation centres. Speech therapy close to the home and qualified for hearing-impaired children is a sensible addition to CI rehabilitation and to support provided by special educators. The institutions collaborate in an interdisciplinary and cooperative manner.

Children of deaf parents

In case of children of deaf parents it must be made sure that the children receive adequate communication and support in spoken language for their hearing and speech development.

If required, sign language interpreters should be involved to communicate with the parents during basic and follow-up therapy as well as long-term follow-up care.

10. Aftercare

CI implantation requires lifelong aftercare (long-term follow-up) under the responsibility of a CI-implanting institution. Aftercare should be offered as part of ongoing care for as long as the implant is used (see Richtlinien GBA Leistungen zur Rehabilitation (GBA guidelines: Benefits for rehabilitation)). It usually is performed by the CI-implanting institution, but may also be delegated in parts. While the audiological and medical aftercare must continue to be provided at the CI-implanting institution, hearing therapy, speech therapy and/or technical aftercare may be delegated to cooperating CI (rehabilitation) centres or other

qualified, cooperating institutions (e.g. hearing aid acousticians for technical aftercare) (see also Fig.2, page 13).
Aftercare is used for medical and audiological-technical checks and consultation as well as for pedagogical-therapeutic assessment and consultation, including documentation with the objective of stabilizing and optimizing the individual's ability to communicate. Aftercare furthermore serves to ensure the therapeutic result and quality assurance, as well as to establish indications for supplementary diagnostic, therapeutic (or, as the case may be, rehabilitative) measures.

It involves recording the long-term effects of CI implantation, possible complications, informing the patient on adaptation to the state of the art, and should assist in the use of additional communication devices and assistance systems. Aftercare thus also serves to determine any potential renewed therapeutic need (e.g. medical, audiological, hearing therapy, speech therapy, socio-medical). This must then be performed or initiated by a CI implanting institution. If required, the patient therefore will be returned to the phase of "follow-up therapy" with the effect that, as the case may be, renewed therapeutic measures can also be initiated, e.g. renewed CI rehabilitation. Aftercare may also involve determining the need for and performing of a technology upgrade and/or update.

10.1 Providing aftercare

Aftercare may be provided on an inpatient, day-patient or outpatient basis. In case of adults, it should be performed at least once annually.

Aftercare for children usually requires shorter examination intervals (usually twice annually) and more time should be scheduled.

In case of bilateral implants, the follow-aftercare effort required is necessarily greater. Since additional therapeutic needs may be identified as part of the aftercare, follow-up therapy measures may be required again, including any necessary rehabilitation measures.

10.1.1 Medical aftercare

Medical aftercare involves:

- specialist ENT examination,
- as the case may be, specialist phoniatric/paediatric examination,
- ear microscopy,
- as the case may be, radiological examination,
- as the case may be, laboratory testing,
- consultation,

- evaluation of the therapeutic outcome,
- as the case may be, prescribing additional diagnostic and therapeutic measures,
- ensuring collection and documentation of quality assurance measures.

10.1.2 Technical aftercare

A CI-qualified hearing care professional may be involved in the technical aftercare on site. If the technical aftercare detects sings of a need for audiological aftercare, this should be done primarily at a CI implanting institution.

Technical aftercare comprises:

- checking the stimulation parameters,
- objective measurements:
    - testing the electrode impedance,
    - ECAP measurement.
- hearing tests to assess CI processor setting (e.g., loudness scaling, functional gain),
- as the case may be, pure tone audiogram,
- speech audiogram/speech tests,
- technical consultation and evaluation of the CI system's functionality,
- supply of spare parts/accessories/additional equipment.

10.1.3 Audiological aftercare

Audiological aftercare comprises:

- hearing tests to assess CI processor setting (e.g., loudness scaling, functional gain),
- speech test in quiet and in noise at defined control time points: 3, 6, 12 months, thereafter annually,
- technical and audiometric controls,
- testing the electrode impedance, ECAPs,
- optimisation of CI processor settings,
- documentation and evaluation of the results,
- as the case may be, correcting the bimodal fitting,
- in case of hybrid systems: fitting a hearing aid component in addition to the cochlear implant and/or technological upgrade/update.

10.1.4. Speech-therapeutic aftercare in children

- Testing speech development (if appropriate tests exist) and/or spontaneous speech samples to assess articulation, vocabulary, grammar, pragmatics, auditory perception or auditory discrimination, auditory memory, including documentation,
- Exchange with treating speech therapists and, if required, initiating therapy or endorsing the continuation of ongoing hearing/speech therapy.

10.1.5 Involvement of hearing care professionals

Hearing care professionals involved in CI implantation must complete further training based on the DGA (German Society of Audiology) recommendations of the "CI Audiologist".

The contents of such further training should be based on the respective current version of the "Weißbuch CI-Versorgung in Deutschland" (White Paper CI Implantation in Germany) of DGHNO-KHC.

The work of the hearing care professional in the context of CI implantation is focused on the technical aftercare of patients with CI implants. Work in the context of basic and follow-up therapy for processor fitting and setting should not be carried out. These are tasks of the CI-implanting institution. Processor setting in the context of aftercare by the acoustician should be performed exclusively within a cooperative agreement with the CI-implanting institution. Processor setting during basic and follow-up therapy is not the task of hearing care professional, of the CI-implanting institution.

The basic prerequisite for the accompanying implant aftercare is proof of theoretical training and of practical training at a CI-implanting institution as well as proof of product-specific training by the CI manufacturers used at the CI-implanting institution.

10.2 Structural framework

Quality assurance of CI implantation requires adequate and expedient year-round space, equipment, and staffing. It can only take place in CI-implanting institutions being centres with the relevant equipment and with a sufficient number of staff that is to be qualified on an ongoing basis. This is to be distinguished from pure CI rehabilitation institutions, which may be managed under both medical and pedagogical direction. Likewise, combination models of clinical institution with attached rehabilitation institutions exist. In addition to the general structural requirements, the personnel, spatial and equipment structures of a CI implanting facility are described below.

As this concerns an active medical implant, the therapy must be under the supervision of a physician throughout the entire process. The centres must have a defined and documented structure, including quality management.

At this time, a recognized form of certification for CI centres does not exist. This Guideline provides an important technical basis along the pathway to the future establishment of a certification system. The equipment features of a CI-implanting institution are based on the current version of the "Weißbuch CI-Versorgung in Deutschland" (White Paper CI implantation in Germany) of DGHNO as well as the certification concept for audiological centres of the German Society of Audiology (DGA; https://www.dga-ev.com/audiologische-zentren/zertifizierungsverfahren/).

10.2.1 General structural requirements of a CI-implanting institution

Mandatory minimum structural requirements for a CI-implanting institution are the provision of the following:

1. Medical expertise and staffing.
2. Audiological expertise and staffing.
3. Technical equipment.
4. Facilities.
5. Interdisciplinary cooperation structure:
    a. Hearing therapy (speech therapy, linguistics, speech therapy, audiotherapy),
    b. Phoniatrics/paediatric audiology,
    c. Neurology,
    d. Neuroradiology,
    e. Psychiatry,
    f. Psychology,

g. (Neuro-) paediatrics.

10.2.2 Minimum facilities, CI-implanting institution

The following facilities are required in addition to the clinical operational requirements of a CI clinic:

- ENT examination room,
- auditory booth for free-field audiometry (sound insulation according to DIN EN ISO 8253-2:2010-07),
- room for fitting and testing hearing instruments,
- paediatric audiometry facility (e.g. "Mainzer Kindertisch" paediatric diagnostic setup[2]) ,
- testing booth for brainstem audiometry or acoustic evoked potentials with adequate electromagnetic shielding such as apparatus for brainstem audiometry and otoacoustic emissions (click-BERA, ASSR and CERA), measurement may also be performed under anaesthesia/sedation,
- set-up for intraoperative implant function testing and performance of intraoperative subsequent testing,
- free-field room for testing directional hearing,
- facilities/equipment for performing neurootological diagnostics,
- space for consultation and meetings with patients,
- therapy room (as the case may be, also for group therapy) in case of hearing or speech therapy,
- waiting area for patients.

The therapy rooms should be adapted to hearing-impaired people. Where required, additional technologies (accessories, inductionloop, wireless transmission system) are available for consultation at the CI centres.

10.2.3 Minimum equipment/methodology, CI-implanting institution

The CI-implanting institution should provide the following minimum equipment/methodology:

- Sound audiometry/speech audiometry with clinical audiometers class 1 (DIN EN ISO 82531:2011-04); paediatric audiometry facility in paediatric care (e.g. Mainzer Kindertisch),
- OAE (DIN EN 60645-6:2010-08), impedance audiometer (DIN EN 60645-5:2005-08), loudness scaling (DIN ISO 16832:2007-07), speech tests in quiet and in ambient noise,
- hearing aids measurement box, probe microphone/in-situ audiometry,
- hardware and software for conventional hearing aids and implantable systems from at least 3 different manufacturers,

[2] *Where children are fitted with implants.*

- measuring system for BERA, ASSR and CERA (DIN EN 60645-7:2010-08),
- free field audiometry / directional hearing,
- neurostimulator for electrical stimulation of the auditory nerve (promontory test),
- VEMP, KIT, caloric testing, as the case may be, swivel chair testing, optokinetic testing,
- neuroradiological intraoperative imaging,
- neuroradiological postoperative imaging with adequate resolution to assess electrode location (e.g. digital volume tomography),
- holding special and replacement implants available (from at least 3 manufacturers),
- special surgical instruments.

10.2.4 Additional aspects of implantation in children

Adequate accommodation options for carers as part of a centre for inpatient treatment or rest and stay facilities for outpatient or day-patient care are required in addition.

10.2.5 Minimum staffing, CI-implanting institution

The structure of the CI-implanting institution must have comprehensive and permanent, i.e. constantly available (weekdays during normal office hours), minimum staffing so as to safely maintain the competence required for patient consultation and treatment. This requires the following positions to be filled on a full-time basis:

2 ENT consultants specialising in CI.

1 phoniatrician and paediatric audiologist (in case of paediatric care), as the case may be, from a cooperating department or clinic for phoniatrics and paediatric audiology.

1 audiologist specialising in CI: (qualification: technical-scientific master's degree (university or university of applied sciences) with additional further education in the area of audiology, e.g. medical physicist, specialising in audiology or master's degree with focus on audiology and practical experience in the area of CI. A medical degree is also a suitable entry requirement for further qualification as an audiologist specialising in CI. The additional qualification for the use of a specialised audiologist in dealing with CI systems should be based on the DGA's further training regulations "CI-Audiologe" (CI audiologist) (https://www.dga-ev.com/ci-audiologe/Z. f. Audiol. 2019 (2) 57 - 60).

1 "hearing technician": (qualification: employee trained in the area of technical hearing aids with a technical vocational degree; e.g. UAS graduate in audiology and hearing technology/hearing aid technology with professional practical experience or master hearing acoustician with advanced training in the area of CI),

2 medical technology assistants for functional diagnostics (MTA-F), with training in basic information on cochlear implants by a CI manufacturer,

2 therapists for basic and follow-up hearing/speech therapy with professional practice experience or with advanced training in the area of CI from speech therapy professions, e.g.:

- educators of the hearing-impaired,
- graduate speech pathologists,
- state-accredited or academic logopedcis therapists,
- state-accredited or academic speech therapists,
- clinical speech scientists,
- state-certified respiration, elocution and voice teachers,
- audio-therapists.

1 medical assistant/administration

10.2.6 Ensuring the overall process responsibility for CI implantation

The process responsibility lies with the medical director of the CI-implanting institution.

10.3 Implant/CI processor replacement as part of aftercare

10.3.1 CI processor replacement

The CI processor is replaced upon prescription by an ENT consultant experienced in CI implantation and is based on the expected improvement in speech understanding. Usually, a CI processor of the latest generation or development version is used as a replacement (so-called upgrade). The documentation of the gain is based on the patient's subjective assessment in day-to-day life and in special hearing situations, and also on the improved hearing performance in quiet and in noise before and after the replacement. A replacement may also be indicated if the patient's circumstances or quality of life can be significantly improved (e.g. BTE processor versus single-unit processor) or in the event of "end-of-support" for the used processor. The outcome of the replacement is checked and confirmed by the CI-implanting institution. The CI processor usually is replaced by the CI-implanting institution. It may also be performed by cooperating institutions.

10.3.2 Implant replacement

In case of medical or technical impairments which cannot be controlled by any other measures, or in case of a probable defect of the implant which will lead to a deterioration of the patient's hearing performance

or has a negative impact in the patient, a replacement (CIReimplantation) is indicated. The respective latest implant and corresponding CI processor should be used for this purpose. Replacement is furthermore indicated if technical support by the manufacturer is no longer guaranteed for a particular implant system or if technical developments required for adequate hearing rehabilitation can no longer be implemented. In case of patients with below-average hearing performance, replacement may be considered if the improved technology of current implants is expected to provide significantly better hearing performance. A risk-benefit balance is required with a view to medical and technical complications versus an expected hearing improvement. When selecting the implant and the electrode array, the patient's individual medical and technical requirements must be taken into account. In case of reimplantation, a renewed basic therapy and subsequent follow-up therapy are required.

11. Quality assurance

To ensure the quality of CI implantation, the organisational, structural, diagnostic and therapeutic standard processes should be recorded and described in a quality management system (e.g. in a QM manual). The processes should incorporate the relevant statutory regulations - Medizinprodukt-Betreiberverordnung (Medical Device Operator Regulation; MPBetreibV), Medizinprodukte-Sicherheitsplanverordnung (Medical Device Safety Plan Regulation; MPSV), Medizinproduktegesetz (Medical Device Act; MPG) - into a QM certification process.

11.1 Documentation of therapy and rehabilitation progress

Documentation of the therapy and rehabilitation progress includes the following process steps:

- preoperative medical, therapeutic and technical consultation,
- surgery report,
- postoperative progress and medical procedures,
- processor fitting,
- educational and therapeutic procedures,
- medical control results in the context of therapy and aftercare,
- audiological evaluation results,
- results of pedagogic-psychological evaluation Results of phoniatric-paediatric and logopedic evaluation of speech, language development and voice,
- medical complications and technical malfunctions.

11.2 Evaluation of the treatment outcome

The evaluation of the CI implantation process (preoperative evaluation, surgery, basic therapy, follow-up therapy (rehabilitation), aftercare) uses qualitative and quantitative survey methods to determine, monitor,

and assess the effects of the therapy. The evaluation is the basis for quality assurance of complete CI implantation. The evaluation relates to auditory skills and hearing, speech and language development (Schulze-Gattermann 2002; Aschendorff et al. 2007, Finley et al. 2008, Aschendorff et al. 2011, also see ch. 5.1, 5.2, 8, 9, 10).

Since the benefits of CI implantation extend beyond the audiological benefits also to social and psychosocial matters, measurement tools to gauge the quality of life are needed for an holistic assessment. Both CI-specific and hearing loss-specific measurement instruments may be used for this purpose (e.g. Nijmegen Cochlear Implant Questionnaire [NCIQ]).

The "Hearing Handicap Inventory for the Elderly" (HHIE), which has been validated in German, and the APHAB ("Abbreviated Profile of Hearing Aid Benefit"), which is established in hearing aid control, are suitable for recording subjective hearing impairment (Cox et al. 1995, Newman et al. 1990). As another international standard measurement instrument, the SSQ "Speech, Spatial and Qualities of Hearing Scale" (SSQ), which now is also available in German, allows for the subjective assessment of speech comprehension, spatial hearing and hearing quality (Noble and Gatehouse 2004).

Moreover, depending on the accompanying symptoms, additional symptom-related questionnaires such as the tinnitus questionnaire according to Goebel and Hiller (Goebel and Hiller 1994) or the "Vertigo Handicap Questionnaire" (VHQ), which has been validated in German, may be used to supplement the pre- and postoperative diagnostics (Tschan et al. 2010).

Current literature has yet to provide a consistent definition of the postoperative measuring intervals. The aim should generally be to perform repeated measurements during the rehabilitation process so as to map the dynamic development, with current studies showing that an initial first postoperative measurement after 6 months is more beneficial.

11.2.1 Specific issues regarding children and adolescents

The diagnostic and evaluation associated with the implantation process often begins already during the preoperative phase or basic therapy and continues during the course of follow-up therapy/CI rehabilitation based on the milestones of the child's development. With regard to the diagnostic inventory, reference is made to Chapter 5.2 (Pre diagnostics and surgery preparation in children).

During therapy, the hearing/speech development is evaluated and, if required, further measures (e.g. diagnostics, therapy) are derived. Where necessary, psychological consultation is initiated.

The evaluation includes an examination f the following skills:

- auditory perception,
- pre-speech and communicative development (precursor skills, alternative forms of communication such as signing),
- speech understanding and production,
- general developmental diagnostics,
- phonological awareness,
- literacy acquisition

For the evaluation of communication competence in the course of rehabilitation, standardised and normalised tests or parent questionnaires should be used according to hearing age and, as the case may be, age, based on a test algorithm with fixed examination times. For further information, reference is made to the S2k Guidelines "Diagnostik von Sprachentwicklungsstörungen (SES) unter Berücksichtigung umschriebener Sprachentwicklungsstörungen (USES)" (Diagnostics of developmental language disorders (SES) with consideration of circumscribed developmental language disorders (USES) (de Langen-Müller et al., 2011) and "Periphere Hörstörungen im Kindesalter“ (Peripheral hearing disorders in children) (http://www.awmf.org/leitlinien/aktuelle-leitlinien/ll-liste/deutsche-gesellschaft-fuerphoniatrie-und-paedaudiologie-ev.html).

11.3 Quality report

CI-implanting institutions and CI centres (rehabilitation institutions) should disclose their structures in a publicly available quality report in line with the contents of the Guideline. A yet to be established national CI registry should also be participated in. Structured internal documentation must be maintained that records the relevant data to this end.

- Type and number of implantations (unilateral, simultaneous bilateral).
- Demographic data (age, gender).

    - The following serious adverse events, indicating their frequency:

- intra- and postoperative facial nerve paresis,
- incorrect positioning of electrodes with need for revision,
- meningitis following CI implantation, o inpatient admission due to CI-related complication, o Death,
- device breakdown, severe technical malfunctions (device failure) according to "European consensus statement on cochlear implant failures and explantations".

- Total number of basic therapies/year and follow-up therapies/year (number of patients and number of therapy) performed at the CI institution,
- Total number of patients treated at the CI institution for aftercare/year,
- Documentation of average number of appointments per patient for aftercare/year,
- Total number of CI-implantation patients, cumulative.

12. Special provisions for implantable medical devices

According to current medical device law, SOPs must be held available at the CI-implanting institutions for handling cochlear implants.

According to the Medizinprodukt-Betreiberverordnung (Medical Device Operator Regulation; MPBetreibV; last amended by Art. 9 V of 29 November 2018), safety checks must be performed regularly on external active components of active implants (CI processor) in accordance with the generally accepted rules of technology (Section 11). According to Section 5, such checks are to be carried out only by persons who:

1. have up-to-date knowledge of the respective activity based on suitable training and relevant professional activity,
2. are not subject to any instructions with regard to the professional assessment, and
3. have the means, in particular premises, devices and other work equipment, such as suitable measuring and testing equipment, required to duly and comprehensibly perform the respective activity.

Furthermore, according to Section 15 para. 1, the person responsible for the implantation (usually the surgeon) is responsible for handing out written or electronic information after completion of the implantation. Such information should contain the behavioural instructions required for the patient's safety after implantation in a generally comprehensible manner. This also includes instructions for performing MRI examinations in line with manufacturer instructions. Furthermore, the time of the subsequent check-ups must be noted. According to Section 15 para. 2, an implant passport must be issued, containing at least the following data:

a. the patient's first and last name,

b. name, type as well as serial number of the medical device,
c. name or company of the medical device manufacturer,
d. date of implantation, and
e. name of the person responsible and the institution that performed the implantation.

According to Section 15 para. 2, the operator of an institution where active medical devices are implanted must retain the documentation on such implants, which can be used to clearly identify and contact patients in the event of corrective measures under the Medizinprodukte-Sicherheitsplanverordnung (Medical Devices Safety Plan Regulation), in such a way that the affected group of patients can be identified within three working days via type and batch or serial number of the implant as well as via the name of the responsible person under Section 5 of the Medizinproduktegesetz (Medical Devices Act). The records must be retained for a period of 20 years after implantation; after the expiry of this period, they must be destroyed without undue delay.

According to Section 3 of the Medizinprodukte-Sicherheitsplanverordnung (Medical Devices Safety Plan Regulation; MPSV), any malfunction, failure, change in features or performance, or improper labelling or user instructions of a CI system must be reported that has led, could have led, or could lead directly or indirectly to the death or serious deterioration of the health of a patient, user, or other person. Malfunctions also include defects in serviceability causing misuse.

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14. Appendix

14.1 Editorial independence

The Guideline has not received any external funding. Travel expenses and facilitation costs to the consensus conference were borne by the respective professional society.

14.2 Declaration of interests and handling of conflicts of interest

All participants declared their direct material and indirect e.g. academic interests via the AWMF form. (Summary table in the extra document "Disclosures on conflicts of interest"). The declarations of all members of the Guideline group were assessed by the Guideline coordinator for thematic relevance in respect of the Guideline topic and for low, moderate, and high-level conflicts of interest and significance for voting, taking into account the pluralistic composition of the Guideline group. Abstentions were not deemed necessary.

14.3. Validity period and updates

This Guideline is valid until 5 years after publication. At that time, at the latest, the content will be reviewed and, as the case may be, updated. Should the Guideline coordinator become aware of findings in the meantime that necessitate revision of the Guideline, such update will be made earlier.

14.4 Adoption of the Guideline

The present version of the Guideline was deliberated and adopted by 31 August 2020 by the steering committees of the participating professional societies and associations.

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