Exploring the capabilities of modern cochlear implants : from electrophysiology to quality of life

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Chapter 3

Cochlear implant outcomes and quality of life in adults with prelingual deafness

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Abstract

Objectives: To evaluate sound and speech perception and quality of life in prelingually deafened adults implanted with state of the art devices. To investigate which patient factors influence postoperative performance.

Study design: Prospective intervention study.

Methods: Eight prelingually deafened subjects participated (onset of severe hearing impairment before the age of four and functioning in an oral-aural setting). Subjects were implanted at a mean age of 36 (range: 21-55) with a CII or 90K cochlear implant (Advanced Bionics Corp.). All subjects completed standard speech perception tests, as well as quality of life measures (Health Utility Index Mark-II, Nijmegen Cochlear Implant Questionnaire, Visual Analogue Scale for subjects' hearing and health) at different moments in time. Post-operative scores were compared with each other and with the baseline, pre-operative scores. The relationship between nine patient variables and the postoperative CVC phoneme score was also investigated.

Results: Significant improvement was measured for CVC word and phoneme scores and several quality of life measures. Post-operative speech perception correlated with a new and promising factor, defined as the quality of a patients' own speech production (QoSP)

Conclusion: With state-of-the-art implants, speech perception and quality of life do improve in prelingually deafened adults. More importantly, the prognostic value of QoSP should be investigated further.

Introduction

It is well known that speech recognition improves considerably in congenitally and prelingually deafened children who undergo early cochlear implantation (CI).[1] In postlingually deafened adults, the speech perception results with CI have also improved in the last decade, in quiet, as well as in background noise.[2] Thus, fairly good oral-aural communication is possible with CI, even allowing many CI-recipients to use the telephone again.[3] These promising results can be ascribed to improved speech processing strategies and the development of state-of-the-art, multichannel cochlear implants which optimize the tonotopic organisation of the cochlea. Because of these developments, CI is now a viable option for a wider range of patients. A patient category, which might benefit from these advances in CI technology, is the prelingually deafened population undergoing implantation at a later age. Until the mid nineties these patients were considered poor CI candidates, because improvement in speech recognition was limited.[4] However, several recent studies have suggested that the latest implant technology has resulted in open speech understanding although variability among individuals was great and performance lagged behind that of postlingually deafened adults.[5] The first aim of this study was to evaluate the effect that late implantation with a state-of-the-art device has on speech perception in prelingually deafened adults. It is widely recognized that medical interventions must be evaluated by looking at a broad range of health domains, including psychological and social domains. Evaluation of quality of life (QoL) is multidimensional and includes even areas not likely to be affected by therapeutic interventions. The paradigm that is focused on the comprehensive measurement of health outcomes on the subjective level is referred to as health related quality of life (HRQoL). HRQoL issues become even more relevant as an alternative to measure cochlear implant benefit when speech recognition results are limited. Quality of life evaluation in the recent literature of the prelingually deafened adult population is limited. One study described satisfaction with the devices while speech recognition demonstrated little improvement.[4] The second aim of this study was to evaluate quality of life changes induced by implantation with a state-of-the-art device. The outcome of cochlear implantation is affected by several factors. Preoperative factors that make a significant contribution to postoperative hearing performance are duration of deafness, communication mode (children), speech perception performance and residual hearing.[6-8] However, *pre*lingually deafened adults differ in at least two aspects from their postlingually deafened counterparts. First, they have not experienced normal hearing and thus their neural system lacks spatial and structural organisation for auditory processing.[9] Second, (verbal) linguistic development depends on early auditory input. This means that main part of patients' speech intelligibility will be determined by the auditory input during the first years of life.[10] Recently, the role of auditory input on cochlear implant performance in the prelingually deafened adult population was further explored and it was concluded that all factors in past and present, contributing to oral communication, might be important to the effectiveness of CI in this specific group.[11] The third aim of this study was to investigate which factors influence post-implant performance in prelingually deafened adults implanted at a later age.

Materials and Methods

Patients

Eight native Dutch adults (three male, five female) were included in this study (Table 3.1). All patients have been implanted in the Leiden University Medical Centre (LUMC). The procedure was uncomplicated and full insertion of the electrode array was achieved. The average age at implantation was 36 [21-55].
Patients met the criteria developed in the LUMC for postlingually deafened adults, which means (aided) pure tone thresholds exceeding 90dB HL, phoneme recognition less than 40% (phonemes correct on CVC words), no medical contra-indications, normal anatomy of the ear, realistic expectations and the presence of a social support system in an oral/aural setting, where they functioned mainly with lip-reading. No formal lip-reading tests were performed, but a Visual Analogue Scale-score (anchored by "never" and "frequent" use of lip-reading) was taken in the outpatient clinic as part of the general work-up. All patients reported to be heavily dependent on lip reading in quiet and noise (seven subjects rated 100%,

Table 3.1: Demographic information on 8 subjects.

Patient	Gender	Onset (yr)	Etiology	Audiometric evaluation (HFI) at 3 years	Duration of deafness	Communication mode	QoSP	Hearing aid use	Educational background	Hearing loss implanted ear (FI, dB HL)	Pre-implant CVC-phoneme (%)	Implanted ear	Implant
A	M	3	Progressive	not available	55	oral	24	0	secondary school: basic	100	0	AD	CII, HiFocus I with positioner
B	F	0	Meningitis	>100	22	total	7	2	on-the-job-learning	130	0	AS	CII, HiFocus I
C	M	0	Congenital	91	21	oral	60	2	Secondary school: advanced	113	23	AD	CII, HiFocus I
D	M	0	Hepatitits	87	31	oral	68	2	on-the-job-learning	108	14	AS	CII, HiFocus I
E	F	2	Congenital rubella	105	27	oral	36	2	secondary school: advanced	108	20	AS	CII, HiFocus I
F	F	3	Unknown	not available	36	oral	not available	2	secondary school: advanced	107	24	AS	hiRes 90K HiFocus I
G	F	2	Ototoxic medication	not available	36	oral	42	2	Secondry school: basic	113	19	AD	HiRes90K HiFocus I
H	F	0	Congenital	not available	47	oral	74	1	on-the-job-learning	110	14	AS	HiRes90K HiFocus I

one rated 85%). Additional criteria for the study group were: (1) prelinguistic severe hearing loss (all patients attended the pre-primary school for the deaf from the age of four, which implies a hearing loss of at least 80dB); (2) age 16 years or older at the time of implantation; (3) implantation with “state of the art” device; (4) auditory-oral environment; (5) motivation.
Initial fitting was performed at approximately 4-6 weeks after surgery. The speech perception rehabilitation program commenced immediately after the fitting. This intensive program starts with twenty 30-minute sessions in the first two weeks after which the frequency gradually decreases. During the rehabilitation period, attention is paid to the psychological and social aspects of implant use. All patients had audiometric and QoL follow-up for at least two years.

Speech perception

Speech perception scores were obtained in a free-field condition using the Dutch Society of Audiology standard CVC (monosyllabic) word list at 65dB HL, as described elsewhere.[2] In the Netherlands, these tests are used regularly, in routine clinical practice, as well as with cochlear implant users. Pre-operative tests were performed with adequately fitted hearing aids (when in use by the patient), and post-operative tests with the CI.

Quality of life

Quality of Life was evaluated in all patients, at four test moments: pre-operatively, and postoperatively at 4-5 months (shortly after completion of the initial training), 12 months and 30 months. Three different instruments (two questionnaires and a VAS) were used to measure QoL: Health Utility Index (HUI-Mark II)[12], Nijmegen Cochlear Implant Questionnaire (NCIQ)[13] and a VAS for hearing and health. The HUI-Mark II is a generic, multi-attribute, preference based classification system, and is administered as a measure of general health status. The preferences that underpin the scoring of the questionnaire were obtained from a sample of the Canadian public (that may differ from the Dutch public). It is focused on the more functional concepts of HRQoL, such as disabilities (dysfunction) and resulting dependencies. The HUI-Mark II encompasses seven domains or attributes (sensation, mobility, emotion, cognition, self-care, pain and fertility). The last domain can be safely omitted if it is not relevant, without prejudicing the interpretation of the

questionnaire. Three to five levels of functioning are defined in each domain. Any specific combination of functioning on the applicable domain levels constitutes a unique health state. Each health state has an associated weight that indicates the subjective assessment of the state in question. This weight utility, on the scale from 0 (=death) to 1.0 (=perfect health), is obtained by applying a multi-attribute utility function, predetermined from the general population. Utilities express the overall valuation of a specific health state, and can be multiplied by expected life years to compute quality adjusted life years (QALYs). Cost-utility was determined as follows: cost-utility = costs (in euro) / Δ(life years x health utility). In our patient group without significant co-morbidity, life years were calculated to be 45 years on average based on a life expectancy of 83 years for women and 79 years for man. Future benefits of the cochlear implant were discounted at a discount rate of 3% per annum. Costs of providing a cochlear implant in the Netherlands including counseling, operation and rehabilitation were established at 40.768€ on the basis of current data from Dutch health authorities. Furthermore, 2.007€ per year were added for follow-up expenses (calculated as the difference between costs for maintaining implants minus costs for hearing aids). When the costs were discounted annually by 3% to account for the time value of money, the calculated costs for implantation and follow-up were 88.133€.
The NCIQ is a validated, disease specific instrument to measure hearing-related quality of life.[13] Questionnaires comprise three general domains: physical, psychological, and social functioning. Each domain can be divided further in sub-domains. The sub-domain consists of 10 items, formulated as statements with a five-point response scale. There is also a sixth response category if the item is not considered relevant. Final scores for the sub-domains range from 0 (poor) to 100 (optimal). Questionnaires were administered to the patients after an explanation was given. On a 100 mm VAS, subjects rated hearing (anchored by deafness and perfect hearing) and health (anchored by death and perfect health).

Patient factors

The influence of nine preoperative factors on post-implant speech perception was examined at 24 months follow-up. Per patient, team members used the history and preimplantation evaluation to assign a categorical or continuous level. The categorical level comprises gender (male/female), communication mode (sign/

total/ oral), hearing aid use (none/ one/ two) and educational background (primary school/ on-the-job learning/ secondary school basic/ secondary school advanced). The continuous level comprises preoperative hearing thresholds, preoperative CVC scores, duration of deafness and age at implant. Given the fact that linguistic development depends on early auditory input, a ninth factor was introduced: the quality of the patients' own speech production (QoSP). Two experienced speech and language therapists rated the preoperative audiovisual recordings of each patient's spontaneous speech on a VAS (the anchors are given in Table 3.2). Patient F's QoSP is missing, because the preoperative video recording was unavailable.

Table 3.2: Scoring items to qualify the patient's spontaneous language (QoSP) on a VAS.

quality of a patients own speech production (QoSP) (spontaneous speech measured with VAS)	**definition borders**	**VAS**
The pronunciation of this speaker is	unnatural natural	0 100
To listen and understand this speaker is	hard easy	0 100
The speech intelligibility of this speaker be characterized as	unintelligible intelligible	0 100
The language capabilities of this speaker can be characterized by	grammatical incorrect grammatical correct	0 100

Data analysis

Data were analysed using SPSS version 12.0. Per patient, the post-implant CVC phoneme and word scores were compared to the pre-implant scores, using a two-sided *t*-test for paired values ($p<0.05$). Pair wise comparison was also applied to evaluate QoL changes between the various test moments. Correlation analysis was calculated using Pearson's (continuous variables), or Spearman's (categorical variables) correlation.

Results

Speech perception

Post-implant speech perception scores improved over time (figure 3.1). The improvement in phoneme scores is more notable than the word scores, as each CVC word consists of 2.2 independent phonemes.

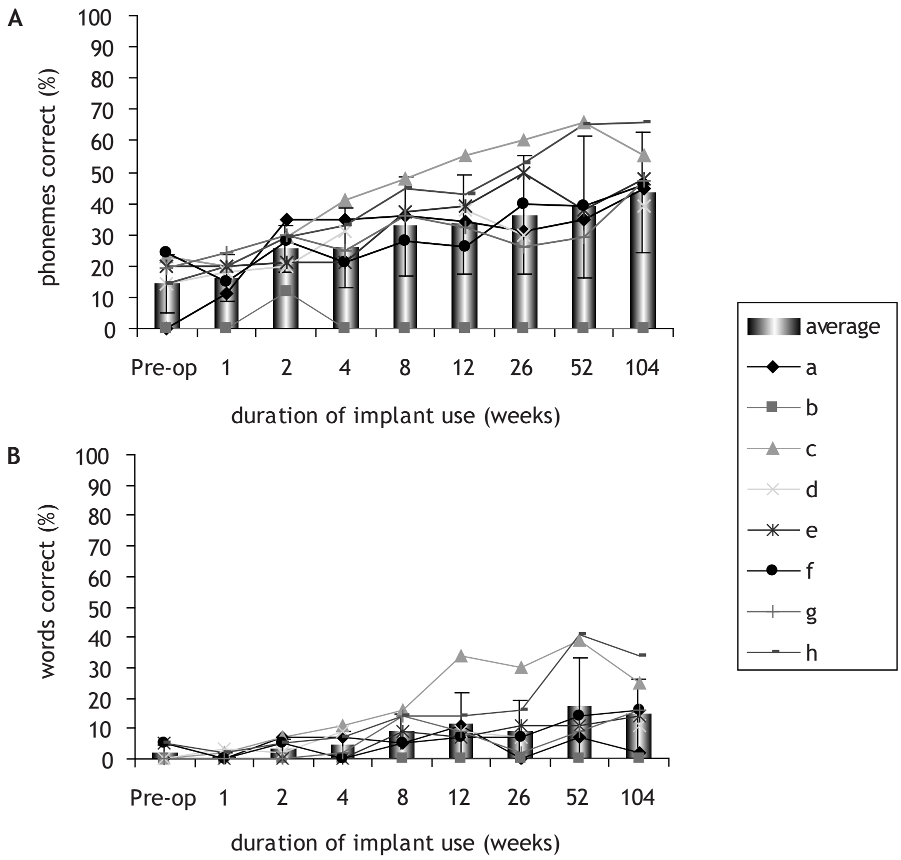

Figure 3.1: Phoneme and word scores. **A**: Phoneme score on a CVC word test in quiet (free field sound only, 65 dB SPL) as measured pre-operative, and at 1, 2, 4, 12, 26, 52 and 104 weeks for the 8 patients in this study. The individual scores are shown as lines, the average scores as bars. **B**: Word scores on the same CVC word test in quiet as in A.

Figure 3.1A gives the average, as well as individual percentages correct phoneme scores, pre- and post-operatively. The mean CVC phoneme score after 24 months of CI use improved significantly from 14% (SD: 9.5%) pre-implant to 43% (SD:19.2%; p=0.001) post-implant. However, individual scores differed considerably. Figure 3.1B shows that mean word score improved significantly from 2% pre-implant to 15% post-implant (p=0.009). As with the phoneme scores, there was considerable variation among the implantees, but only patients C and H reached a word score exceeding 25%.

Quality of life

Different aspects of QoL had improved significantly 4-5 months after implantation, but no significant changes occurred thereafter (figure 3.2). The increase from preoperative to four months postoperative of the "mean" HUI-Mark II was 0.19 (SD: 0.08; paired *t* test: p=0.003; figure 3.2A). Closer inspection of individual attributes in the HUI-Mark II questionnaire revealed that the only significant (paired *t* test, p=0.002) improvement occurred in the domain "sensation" (containing the subdomains vision, hearing and speech). The improvement of 0.19 in health-utility as measured with the HUI-Mark II resulted in a projected accumulated gain of 4.67 QALYs and a cost-utility ratio of 18.872€ per QALY (95% CI: 14.599-26.538€). Figure 3.2B shows the mean scores on the six domains of the NCIQ pre-operatively, at 4, 12 and 30 months. At 4 months, the sub-domain "basic sound perception" improved significantly from 25.0 (SD: 23.9) to 68.1 (SD: 20.9; p=0.002), "advanced sound perception" from 36.6 (SD: 13.9) to 55.9 (SD: 13.3; p=0.01) and "social interaction" from 66.4 (SD: 10.0) to 81.9 (SD: 13.8; p=0.006). No significant improvement occurred thereafter. The improvement in hearing acuity that patients rated on the VAS was significant (p=0.009), while general health did not change (figure 3.2C).

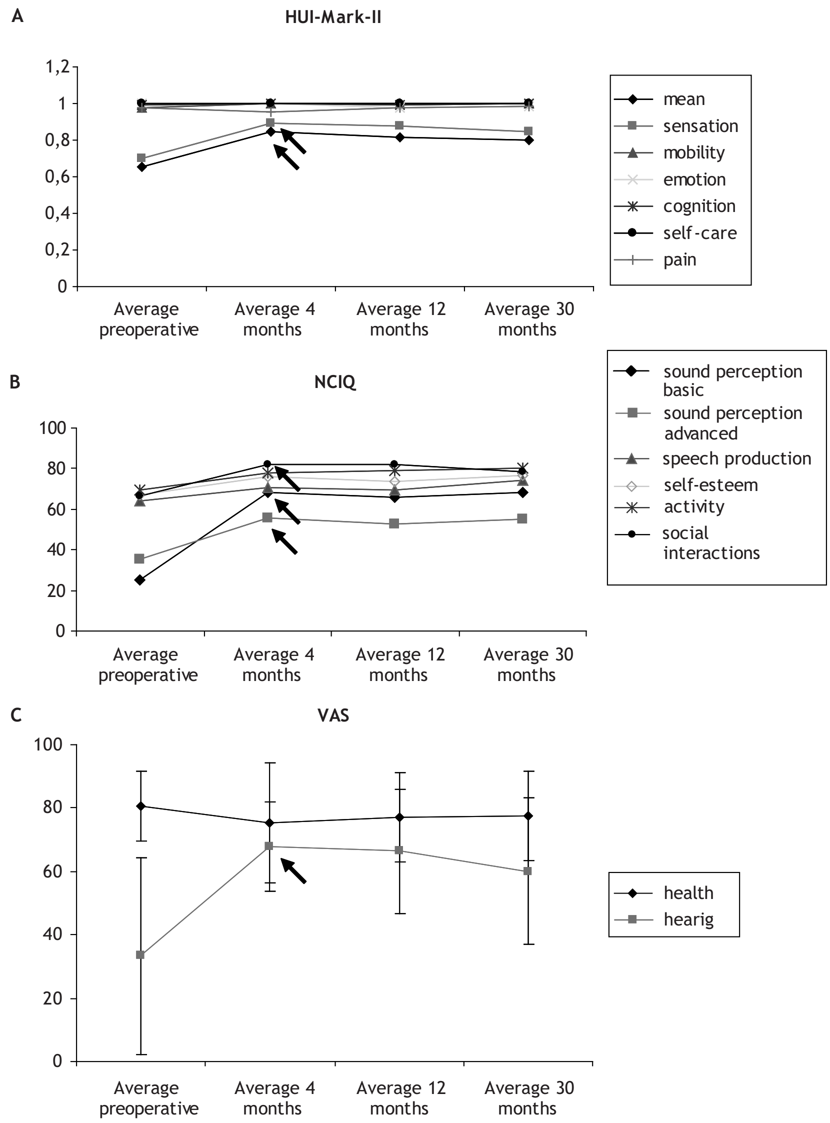

Figure 3.2: Average pre- and postoperative quality of life scores measured preoperatively and at 4, 12, and 30 months concerning: Health Utility Index Mark II (HUI-II) (A); Nijmegen Cochlear Implant Questionnaire (NCIQ) (B); and visual analogue scale (VAS) (C). Standard deviations are shown as error bars (C), or as outlined in the text. Improvement is measured using the Student *t* test (baseline vs. 4 months; 4 months vs. 12 months; 12 months vs. 30 months). Significant change to prior measurement is pointed out by the arrow (➘).

Relation between patient factors and post-implant speech perception

Table 3.3 shows calculated correlation coefficient "r" with 95% confidence interval. The patient cohort was very small, which may have influenced the strength of the correlations and caused non-normal distributions. For example, communication mode appears to have a moderately positive correlation (r=0.577), but this is due to only one patient not having complete oral communication. A strong negative correlation was found for pre-operative hearing threshold (r=-0,725, p=0.042), but the correlation is influenced by one outlier. Moderate to strong positive correlations were found for preoperative CVC scores (r=0.594) and QoSP (r=0.754, p=0.050). After applying a Bonferroni-correction, none of the correlations was significant.

Table 3.3: 2-years CVC phoneme scores as a function of pre-implant patient factors.

Factor	Correlation coefficient "r"	95% Confidence interval
	(Spearman)	
gender	0,169	(-0,601) - (0,781)
communication mode	0,577	(-0,215) - (0,911)
hearing aid use	-0,171	(-0,782) - (0,607)
education	0,379	(-0,445) - (0,855)
	(Pearson)	
preoperative hearing thresholds	-0,725	(-0,946) - (-0,041)
preoperative CVC scores	0,594	(-0,190) - (0,916)
duration of deafness	0,395	(-0,429) - (0,860)
age at implant	0,39	(-0,433) - (0,859)
quality of a patients own speech production	0,754	(0,003) - (0,961)

Discussion

This paper illustrated that individual prelingually deafened patients can reach open set speech perception after implantation. Furthermore, all subjects experienced improvement in several health related quality of life items.

The CVC phoneme scores after implantation range from 0 to 66% in this study. Patients experienced a mean improvement in CVC phoneme scores of 29%, but progress was very variable. In fact, some patients' performance deteriorated (temporarily) during follow up. Our poorest performer (patient B) did not show any improvement on the speech recognition test. This patient attended the school for the deaf at the age of two and was trained in Dutch sign language. She was fitted with hearing aids only at the age of six. It is interesting that this patient demonstrates the lowest QoSP score, which might be a reflection of the lack of auditory input at younger age. Patients' quality of life measured with HUI-II and VAS improved significantly postoperatively, but decreased to preimplant values after one year. The same pattern was noticed with the NCIQ scores, except that the sub-domains sound perception and social interaction remained significant higher. Nevertheless, she reported regular use of the device and improvement of the communication skills. Two of our patients (C and H) could be classified as good performers. A recent report from Teoh et al.[10] described 10 out of 63 prelingually deafened adults, with individual "Hearing in Quiet Test" scores between 40-100%. The same study reports a mean test score of 16% on CVC monosyllabic words after 6 months. They found comparable results for all three cochlear implant devices (Advanced Bionics, Cochlear, and MedEl). Thus, the present study is in line with the findings by Teoh et al., not only with respect to the CVC monosyllabic word scores but also in respect to the variability in speech perception. The present study demonstrates that adult patients with early onset, profound hearing impairment improved significantly in various quality of life dimensions. These changes occurred in the first four months post-implantation, but not thereafter.
On average, the generic HUI-Mark II questionnaire, showed a significant change in quality of life, but this could be solely attributed to the domain "sensation". An increase in "mean" HUI-Mark II scores of 0.19 (4 months) is comparable to that reported in prior studies of postlingual patient cohorts.[14,15] The other five domains did not change significantly compared to the baseline. Nevertheless, QoL measures such as HUI-Mark II remain important because it allows comparison of different studies and provides a method to calculate cost-effectiveness of CI. The calculated cost-utility of 18.872€ per QALY can be regarded as acceptable value for money for a medical intervention.[15]

Disease specific questionnaires are of utmost importance, because the patient's perception of his/her problem is the gold standard in determining patient satisfaction. As could be expected from a disease specific questionnaire, it appears to be much more sensitive than the HUI-Mark-II. This hearing related questionnaire measured important improvements in six domains. Significant improvements were seen in the domains "basic sound perception, advanced sound perception, and social interaction". These scores compare well to the NCIQ scores of a group of postlingually deafened native Dutch speakers[14], except that our prelingual group rated its own pre-operative sound perception considerably higher. This can be explained by the general observation that valuation of health status may differ according to illness experience.
Correlations between post-implant speech perception and pre-operative patient factors were not significant. Nevertheless, an interesting positive correlation was found between postoperative speech perception and QoSP. This is in line with the hypothesis that postoperative hearing capacities are depending on the viability of the central auditory system in long-term deafness. In our opinion, QoSP could be considered an index representing the extent to which patients were able to hear speech in the past. With this in mind, our study population (mean QoSP of 44 [7-74]) might not be truly "prelingually" deafened, given that linguistic development depends on auditory input.[10]
In fact, it is questionable if one should (*and* is able to) differentiate between pre-, peri and postlingual for adult patients that were deafened at an early age. For example, in our older patient group, the moment of being profoundly deaf (hearing loss >80-90dB) is sometimes not established accurately. Furthermore, the prelingual period the period in which the most important aspects of speech and language development are learnt is usually between 0 and 4 years. However, this period cannot be demarcated accurately either. There is, also, individual variation with regard to speech and language development: onset and duration of development, and progression all play a role. With this in mind, we propose the term "early-deafened adult" and recommend focusing on the expected performance rather than classifying the patient as pre-, peri- or postlingually deafened. Further research is necessary to identify factors that predict performance, so that patient selection can be effective. A prospective study

with a larger number of patients, which is currently ongoing in our institute, will increase the statistical power and should reveal whether QoSP might be such a factor.

Conclusion

Early-deafened adults can reach open set speech perception after cochlear implantation. Quality of life is enhanced with a CI, although especially hearing and hearing-related quality-of-life items improve after cochlear implantation. Measures such as the HUI-Mark II and the NCIQ have an additional value in the evaluation of clinical outcome in patients that are generally considered to be poorer candidates. QoSP can be considered as an index of the extent to which patients had been able to hear speech in the past and therefore as an interesting factor in patient selection.

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