Cochlear implant in prelingually deafened oralist adults: speech perception outcomes, subjective benefits and quality of life improvement

Summary

The aim of this study is to report our results in a group of prelingually deafened adults, who followed an oralist rehabilitation programme, and submitted to cochlear implant at our institution. We evaluated 30 prelingually deafened adult patients, 18 males and 12 females, median age 35 years, of a group of 36 prelingually deafened adult patients consecutively submitted to unilateral cochlear implantation at the ENT Unit of the University of Pisa. After implantation, patients achieved significant benefits in terms of speech perception skills, including the ability to have telephone conversations in some cases, quality of life and their own perception of disability. According to literature data, the results herein reported are quite variable but generally satisfactory. Procedures other than traditional speech perception measures should be used to evaluate the benefits of cochlear implant in such patients, to compressively evaluate the global benefits, not only in terms of speech perception, but also in terms of quality of life and daily life.

Key words: Prelingual deafness • Cochlear implant • Quality of life

Introduction

Cochlear implantation is the treatment of choice for patients affected with severe to profound sensorineural hearing loss. Until the mid 1990s, these patients were considered poor CI candidates, because improvement in speech perception was limited. However, several recent studies have suggested that the latest implant technology results in open-set speech perception abilities, although variability among individuals was great and performance lagged behind that of post-lingually deafened adults. These conclusions are mainly based on results obtained in small and heterogeneous samples. It is useful to underscore that in Italy since the sixties oralism has been the main rehabilitative choice for patients suffering with preverbal deafness. As a consequence, in Italy the vast majority of adult patients with preverbal deafness have received oral rehabilitation and thus the population is largely different from those included in western rehabilitation and implant programmes.
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Perception test 13 without lip-reading with hearing aids, comprehensive audiological evaluation, including a speech perception test. Pre-operatively, all patients were submitted to unilateral cochlear implantation at the ENT Unit of the University of Pisa (Italy), during the period from July 1999-July 2011. All prelingually deafened adult patients implanted in our institution were oral language users and consistently used hearing aids before implantation. The 30 patients enrolled in the study are those who gave their consent to participate to the study. Among the 6 implanted patients not included in the study sample, one is a non-user, one has not come for follow-up at our centre for many years and the others do not come frequently for follow-up visits, as they live far from the centre.

Pre-operatively, all patients were submitted to comprehensive audiological evaluation, including a speech perception test 13 without lip-reading with hearing aids, neuroradiological evaluation by petrous bone high resolution computed tomography (CT) and brain and inner ear magnetic resonance (MR). Aetiology of hearing loss was also investigated by molecular analysis of the connexin 26 and 30 genes, and mitochondrial DNA. Genetic (Connexin 26 mutation), and mitochondrial DNA A1555G mutation analysis in all cases, and PDS gene mutation analysis in patients with a large vestibular aqueduct. The most salient features of patients are summarised in Table I.

Post-operatively, during follow-up visits, all patients were assessed by pure tone audiometry in a free field with the CI in use and to a speech perception test 13 without lip-reading with the CI in use. Pure-tone audiometry was conducted with an Interacoustics Clinical Audiometer AC40. When measuring the hearing threshold, both without HA and with HA, we assigned a value of 125 dB at any frequency threshold over the maximum output limit of the audiometer (105 dB for 0.25 KHz and 125 dB for 0.5 and 1 KHz, 120 dB for 2 KHz). Any vibrotactile sensation was excluded. Speech perception was assessed using a speech perception test in Italian language 13 both before (with hearing aids) and after implantation (with CI) in free field, by the same speech therapist in all the patients to avoid bias, with live voice, and without lip-reading. We evaluated the di-syllabic words recognition score using lists of 20 disyllabic Italian words at a level of 65 dB.

In order to obtain information about personal factors and subjective benefits from CI, at the moment of the study setup we administered a questionnaire “University of Pisa Questionnaire” (UPQ) to all enrolled patients. This questionnaire was specifically developed by our research team and is a 40 question survey, divided into 3 sections. In the first section, pre-operative information, such as type of hearing aid, ability to have telephone conversations, perception of music and rehabilitation are collected; in the second section, the same aspects after implantation are investigated; in the third section, information about social life, education level and working life are recorded. Patients were also submitted to the APHAB questionnaire to assess improvement in the patients’ own perception of the disability and in quality of life derived from CI procedure.

### Materials and methods

The sample was composed of 30 prelingually deafened adult patients, 18 males and 12 females, median age 35 years (IQR (interquartile range): 28-42, range: 16-54) of a group of 36 prelingually deafened adult patients (83%) consecutively submitted to unilateral cochlear implantation at the ENT Unit of the University of Pisa (Italy), during the period from July 1999-July 2011. All prelingually deafened adult patients implanted in our institution were oral language users and consistently used hearing aids before implantation. The 30 patients enrolled in the study are those who gave their consent to participate to the study. Among the 6 implanted patients not included in the study sample, one is a non-user, one has not come for follow-up at our centre for many years and the others do not come frequently for follow-up visits, as they live far from the centre.

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The questionnaire was administered both before and after implantation at the moment the study was initiated. Quality of life (QoL) was measured in all the patients after implantation and at the moment of study initiation by using the Italian version of the Short Form-36 Health Survey, version 1 (SF36 v1). Authorisation from the company was obtained. The SF36 is a generic QoL questionnaires that, through a 36-item short form survey, assesses eight health scale scores in relation to physical functioning, role-physical, physical pain, general health, vitality, social functioning, role-emotional, mental health) 14. The SF36 scores estimated from the studied sample were compared to that reported in the Italian literature for normal hearing subjects and to that reported for hearing impaired subjects 15.

Finally, we studied the relationship between the results, in terms of speech perception with CI, and age at the first hearing aid fitting, progression of hearing loss, pre-operative PTA without hearing aids and pre-operative PTA with hearing aids in order to find any possible associations.

The mean follow-up duration after surgery was 6.2 years, ranging from 1 to 13 years.

### Statistical analysis

Statistical analysis was performed with the aim to investigate differences between the pre-operative and the post-operative speech perception abilities and health conditions of patients. The distribution of numeric variables was assessed by the Shapiro-Wilk W test for normality. Comparisons between pre- and post-CI intervention patients data were conducted using the Wilcoxon matched-pairs signed-ranks test for non-parametric distribution of continuous variables and the McNemar’s chi-squared test for categorical variables. The difference between paired data was considered statistically significant with a p < 0.05. Using the quantile regression model, we examined the relationship between the post implantation open-set speech recognition score and selected patient characteristics (covariates) such as as age at first hearing aid fitting, pre-operative PTA with hearing aids and progression of hearing loss, as well as the APHAB scales and SF36 scales for the 50th percentile. Compared with the traditional linear model, quantile regression is robust for departures from normality and heteroscedasticity assumptions in the response variable 16.

Statistical analyses have been performed using the statistical software Stata 12.1 (Stata Corp, College Station, Texas USA).

### Results

The mean pre-operative hearing threshold in the implanted ear (pure tone audiometry - PTA between 0.5-1-2 KHz) was 111.08 dB (range 93 dB-125 dB). The mean pre-operative PTA in free field with hearing aids was 54.6 dB (range 31.6 dB-125 dB).

With regards to speech perception skills, the mean post-operative open-set words recognition score was 60% (range 0-100%). Two of 30 patients scored 0%. The improvement in terms of speech perception was statistically significant (p = 0.000).

After implantation, the mean pure tone audiometry (PTA between 0.5-1-2 KHz) in free field with the speech processor on was 34.7 dB (range 25-51.6 dB).

The mean pre-operative speech perception score was 15.7% (range 0-50). Before implantation, 7% of the patients were able to have telephone conversations. After implantation, 60% of patients are able to have telephone conversations. The difference between the proportion of patients able to have telephone conversations before and after implantation was significant (p = 0.000).

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The most significant data, regarding subjective benefits after implantation, collected at the UPQ questionnaire are shown in Table II.

### Results of APHAB scales are reported in Table III.

The norm-based scores of the SF36 eight scales measured in prelingual deafened patients revealed a significant improvement after CI intervention compared to the before situation in terms of general health, social functioning and mental health as reported in Table IV.

In the post-intervention framework, a significant as-
association was found between disyllabic word recognition scores and the Ease of Communication scale of the APHAB questionnaire (p = 0.030). Considering the other scales of the APHAB questionnaire and the eight scales of the SF36 questionnaires, we found no significant associations between each scale and disyllabic word recognition scores after CI intervention.

From the univariate quantile regression model, there are no evidence of statistically significant associations between post-implantation open-set speech recognition score and age at first hearing aid fitting, pre-operative PTA without hearing aids and pre-operative PTA with hearing aids (p > 0.05). Nevertheless, we found a significant association between post-implantation open-set speech recognition score and progression of hearing loss (p < 0.05).

Discussion

In the present paper, we report the results achieved in a group of 30 oralist prelingually deafened adults, submitted to CI.

Cochlear implantation in prelingual adult patients is still a true challenge since it requires a difficult decision making process for both clinicians and patients due to the uncertainty of the outcome and the risk of obtaining poor or no improvement in speech perception and stopping to use the implant; nevertheless, an increasing number of recent studies attest the efficacy of CI even in these cases, albeit with variable results. 

In a recent systematic review of the literature on effectiveness and cost-efficacy of CI in children and adults, we reported that cochlear implantation is both clinically- and cost-effective even in adults with prelingual deafness.

In the sample herein, the results in terms of speech perception skills are quite satisfactory. After implantation, we recorded a significant improvement in open-set speech recognition scores: before implantation the mean open-set speech recognition score was 15.7% and after implantation it increased to 63.6%. Moreover, before surgery 7% of the patients were able to have telephone conversations, while after implantation 60% of patients were able, even if at various degrees of ability.

Most of the studies reporting post-implantation results in prelingual deafness have focused on speech perception results and only a few have dealt with patient satisfaction, subjective benefits and impact of CI on the quality of life. Studies investigating the impact of cochlear implantation on the quality of life in prelingually deaf adults found that, even when the results are poor in terms of speech perception abilities, participants reported satisfaction with the CI procedure. These observations suggest that measures other than speech perception test should be used to evaluate the benefit of a CI in this population, and therefore benefit and performance should be viewed as two separate outcomes.

In the present study, at the UPQ questionnaire we recorded an improvement in the ability to carry out some daily life activities such as the use of the telephone and the abil-

### Table III. Results of APHAB scales before and after the cochlear implant intervention.

<table>
<thead>
<tr>
<th>Scale</th>
<th>Pre CI</th>
<th>Post CI</th>
<th>p*</th>
</tr>
</thead>
<tbody>
<tr>
<td>EC scale</td>
<td>Median (IQR)</td>
<td>54.2 (17.7-74.7)</td>
<td>37.5 (16.5-46.2)</td>
</tr>
<tr>
<td>BN scale</td>
<td>Median (IQR)</td>
<td>64.3 (18.8-78.2)</td>
<td>47.8 (33.7-66.3)</td>
</tr>
<tr>
<td>RV scale</td>
<td>Median (IQR)</td>
<td>70.7 (45.2-82.7)</td>
<td>54.2 (33.7-66.3)</td>
</tr>
<tr>
<td>AV scale</td>
<td>Median (IQR)</td>
<td>83.1 (1-26.8)</td>
<td>31.7 (8.3-49.8)</td>
</tr>
</tbody>
</table>

Note: *Wilcoxon signed-rank test, CI, Cochlear implant. IQR, interquartile range (25° percentile-75° percentile), EC, ease of communication; BN, background noise; RV, reverberation; AV, aversiveness

### Table IV. Comparison between the norm-based scores of the 8 SF36 domains in the sample of prelingually deafened patients before and after the cochlear implant intervention.

<table>
<thead>
<tr>
<th>Domain</th>
<th>Pre CI Median</th>
<th>IQR</th>
<th>Post CI Median</th>
<th>IQR</th>
<th>p*</th>
</tr>
</thead>
<tbody>
<tr>
<td>Physical Functioning (PF)</td>
<td>57.2</td>
<td>55.1-57.2</td>
<td>57.2</td>
<td>55.1-57.2</td>
<td>0.516</td>
</tr>
<tr>
<td>Role-Physical (RP)</td>
<td>56.2</td>
<td>49.2-56.2</td>
<td>56.2</td>
<td>49.2-56.2</td>
<td>0.415</td>
</tr>
<tr>
<td>Body Pain (BP)</td>
<td>62.8</td>
<td>47.3-62.8</td>
<td>62.8</td>
<td>51.6-62.8</td>
<td>0.271</td>
</tr>
<tr>
<td>General Health (GH)</td>
<td>57.2</td>
<td>50.9-61.7</td>
<td>60.3</td>
<td>54.6-61.7</td>
<td>0.003</td>
</tr>
<tr>
<td>Vitality (VT)</td>
<td>53.8</td>
<td>46.7-60.9</td>
<td>56.2</td>
<td>51.4-63.3</td>
<td>0.107</td>
</tr>
<tr>
<td>Social Functioning (SF)</td>
<td>46.3</td>
<td>35.4-57.2</td>
<td>49.0</td>
<td>40.9-57.1</td>
<td>0.026</td>
</tr>
<tr>
<td>Role-Emotional (RE)</td>
<td>55.3</td>
<td>46.8-55.3</td>
<td>55.3</td>
<td>55.3-55.3</td>
<td>0.138</td>
</tr>
<tr>
<td>Mental Health (MH)</td>
<td>46.9</td>
<td>39.1-55.0</td>
<td>52.7</td>
<td>45.9-55.0</td>
<td>0.004</td>
</tr>
</tbody>
</table>

IQR, Interquartile Range (25° percentile-75° percentile)

* Wilcoxon signed-rank test
ity to listen to television and music. After implantation, 60% of the patients (n = 18) were able to have telephone conversations and 28% of these (n = 5) were able to have free telephone conversations even with unknown persons. In addition, 30% of patients were able to listen to television without reading subtitles, compared to only 3% before implantation. Furthermore, 77% of patients reported that after implantation the perception of music improved and 70% of patients indicated that with CI the perception of voice was more natural than with hearing aids (see Table I).

Concerning the results of the APHAB questionnaire, we recorded significant improvement after surgery in terms of median frequency of troubles while hearing in each of the four situations investigated by the questionnaire, such as ease of communication, background noise, reverberation and averseness (p < 0.05). Substantial improvements due to CI interventions were observed for ease of communications and background noise: median benefit 24.8 (IQR: 0-49.5, p = 0.003) and 16.2 (IQR: 3.7-40.5, p = 0.000).

With regards to the SF 36 questionnaire, our patients reported very high scores after CI intervention. The patient’s own quality of life perception after CI intervention was very similar to that of the normative sample of healthy Italian subjects, and even better for some aspects. Patients scored as well as the normative healthy sample for physical health, in terms of Physical Activity, Role and Physical Health and Physic Pain, and for mental health in terms of Role and Emotional Status. Implanted prelingually deafened patients did not experience limitations or pain due to their health condition in work or domestic activities nor problems resulting from emotional status. The general health and the mental health subjective perception was better in the sample of patients with CI than that of healthy patients. Implanted patients experienced lower scores for Vitality and Social Activities as compared to healthy subjects (median score: 70 (IQR: 60-85) versus 75 (IQR: 65-85) and 81.3 (IQR: 63-100) versus 87.5 (IQR: 75-100).

When comparing the scores recorded at SF 36 in our sample to the normative sample of Italian hearing impaired patients, our sample scored better than the normative sample in every aspect except for Role and Emotional State, where they achieved the same score (see Fig. 1). The SF36 scores obtained in our sample of prelingually deafened patients have to be interpreted with much caution, as they are related to subjective evaluations on their own perception of quality of life. Moreover, the sample is small and in some subscales we recorded a high variability in answers. The comparison between our sample and the Italian normative one has some limitations due to the self-reported conditions identifying the Italian healthy group (mean age 34.8 years, age > 65 years 3.7%, female gender 46.8%) and the Italian hearing impaired group (mean age 63 years, age > 65 years 50.7%, female gender 45%).

According to literature data, even in the reported group, patients who did not achieve improvements in terms of open set speech recognition abilities reported substantial benefits in daily life activities and in the QoL.

It is well known that the outcome after CI in prelingually deafened adults is quite variable and is best shown in the work by Klop et al (2007) 8. This is likely to be related to the wide variability of the characteristics of patients. Post-implantation benefits both in adults and children is related to several individual factors, and this is mostly true in the case of pre-lingually deafened adults. Pre-operative factors that seem to make a significant contribution to post-operative hearing performance are age at onset of hearing loss and at first hearing aid fitting, speech perception performance with hearing aids 6 7 11 12, residual hearing 21, communication mode 21, type of rehabilitation, educational environment 21, motivation and psychological aspects 21 6 7 11 12.

Moreover, Dijkhuizen et al. in 2011 reported that speech intelligibility is predictive of post-implantation hearing results in a group of prelingually deafened adults, using a specifically developed test battery 22 and in a recently published paper has confirmed this 23.
Cochlear implant in prelingually deafened oralist adults

thus their neural system lacks the spatial and structural organisation for auditory processing. The literature suggests that the colonisation of the auditory cortex by other sensory modalities is the main limiting factor in post-implantation performance, and not the pathological degenerative changes of the auditory nerve, cochlear nucleus, or auditory midbrain. Consequently the educational programs that stress oral communication as the preferred educational modality could potentially reduce the cortical colonisation of the central speech and language processing centres, and consequently the use of oral communication should be an important candidacy criterion in cochlear implantation of patients with long-term prelingual deafness. Moreover, some recent studies have explored the role of early auditory input on CI performance in prelingually deafened adults, and it was concluded that the availability of effective auditory input in early life may be the fundamental factor underlying the potential for speech perception with a CI in later life. Considering the sample herein reported, a relatively high percentage of the patients (63%) experienced a progression of hearing loss. These patients presumably had a better auditory input early in life. We believe that this factor, associated with oralism as an exclusive mode of rehabilitation, played an important role in achieving satisfactory results reported, also in terms of open-set speech recognition abilities and consequently in the ability to have telephone conversations. In this regard, we recorded a statistically significant correlation between post-operative dysyllabic word recognition score and progression of hearing loss (p < 0.05) in our sample; nevertheless, we did not find a significant correlation between post-operative results and pre-implant hearing threshold either with or without hearing aids and age of first hearing aid fitting.

Conclusions
In conclusion, our results demonstrate that cochlear implantation is an effective option for prelingually deafened adult patients. In accordance with the literature data, the results reported herein are quite variable but generally satisfactory, both in terms of improvement of speech perception abilities and in terms of improvement of the quality of life and patients’ perception of their disability. Indeed, some individual factors significantly affect the results, and among these effective auditory input in early life and oralist rehabilitation mode seem to be the fundamental factors underlying the potential for speech perception with a CI later in life. In this regard, we found a significant correlation between the progression of hearing loss and post-operative results. Such patients had experienced a relatively good and effective auditory input early in life, and this allowed their auditory cortex to develop properly, which in turn allowed them to gain satisfactory results in terms of speech perception after implantation.

Therefore, the indications to CI and prognostic factors must be discussed on a case by case basis, taking into account mainly the progression of hearing loss, hearing aid use and results before implantation, rehabilitation mode, motivations and psychological aspects, and it is important to perform accurate counselling to provide patients with realistic expectations.

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