



Measuring health-related quality of life after pediatric cochlear implantation: A systematic review

Frank R. Lin^{*}, John K. Niparko

Department of Otolaryngology-Head and Neck Surgery, Johns Hopkins University School of Medicine, Baltimore, Maryland, United States

Received 20 March 2006; received in revised form 5 May 2006; accepted 14 May 2006

KEYWORDS

Health-related quality of life;
Pediatric cochlear implantation;
Outcomes

Summary

Objective: The measurement of health-related quality of life (HRQL) in children presents conceptual and methodological challenges owing to the multidimensionality of the required information and limitations in patient self-report. HRQL results provide a broad measure of treatment impact from the patient and family perspective and are crucial to guiding clinical and policy decisions. The objective of this study was to evaluate how HRQL in children with cochlear implants has been measured in published studies in order to draw conclusions that could inform future investigations of this area of clinical research.

Methods: We searched PubMed, EMBASE, CINAHL, PsychInfo, and Web of Science databases using a defined search string and hand-searched reference lists of relevant articles and personal files. Retrieved citations were reviewed in two stages, a title and abstract screen followed by review of the full-length article. Inclusion criteria for studies were: (1) original peer-reviewed research article; (2) enrolled subjects <18 years old with cochlear implants; (3) use of a HRQL instrument that incorporated components of physical, mental, and social health; and (4) in English. Data from full-length articles were extracted by a single-investigator.

Results: We retrieved 671 citations with our search strategy, and 10 citations were found to be eligible for inclusion. All studies used a cross-sectional design, and three types of HRQL instruments were used: generic questionnaires, ad hoc instruments designed specifically for the purposes of the study, and the parents views and experiences with pediatric CI questionnaire. Heterogeneity in study design and instruments prevented a quantitative, meta-analysis of the data.

^{*} Corresponding author at: Department of Otolaryngology-Head and Neck Surgery, Johns Hopkins Outpatient Center, Room 6163, 601 North Caroline Street, Baltimore, MD 21287, United States. Tel.: +1 410 955 3401; fax: +1 410 614 9444.

E-mail address: flin1@jhmi.edu (F.R. Lin).

Conclusions: Studies that used well-validated, generic HRQL instruments supported conclusions that were less subject to potential bias from the perspective of the clinician investigator. Most studies did not use well-defined cohorts with respect to age at implantation and duration of implant use, and conclusions in these studies were also subject to potential bias. No well-validated, deafness-specific HRQL instruments are currently available. Future research should be done with existing, generic HRQL instruments and with strict study inclusion criteria. Suggested generic HRQL instruments are discussed.

© 2006 Elsevier Ireland Ltd. All rights reserved.

1. Introduction

Prelingual deafness carries cascading consequences for oral language development and related experiential learning that affects behavioral and social development [1–5]. Over the past 15 years, cochlear implantation (CI) has evolved as a successful treatment option for eligible deaf children. Early restoration of auditory input provided by a CI enables children to substantially improve their verbal language-learning trajectory [6,7]. However, there still exists wide variability in outcomes after CI [8]. While many children with CI have been able to fully interact with normal-hearing peers and participate in the mainstream, others have experienced limitations in their ability to communicate verbally.

Previous studies of pediatric CI have investigated which factors lead to better outcomes. In the vast majority of these studies, outcome measures were based on clinical tests of speech, hearing, or language. However, given the broad impact of deafness

on a child’s development, it is unclear how clinical measures of efficacy (e.g. hearing and speech measures) manifest in actual effectiveness (i.e. performance in home, school, and social settings). Empirical clinical experience often suggests otherwise, with some children performing well in structured clinical settings but with parents reporting poor results at home. Some studies have also demonstrated that outcomes in clinical settings do not correlate with performance in unstructured settings [9].

The need for a more comprehensive outcome measure has spurred interest in using measures of quality of life to assess the impact of cochlear implantation. Quality of life can be broadly defined as an individual’s contentment or satisfaction with life. Numerous factors may, therefore, contribute to this perception (sense of well-being, financial status, spirituality, living environment). Health-related quality of life (HRQL), on the other hand, represents those domains of overall quality of life that are

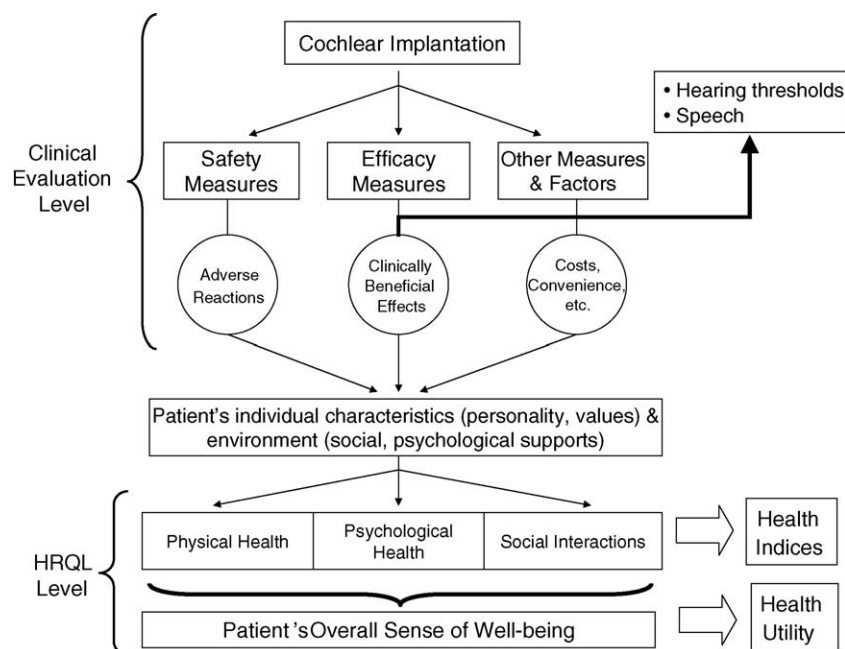


Fig. 1 Conceptual model of the relationship between cochlear implantation and health-related quality of life (adapted from Spilker [47]).

affected by one's health, and we will focus on this concept in our review [10].

A schematic model of the role of HRQL in evaluating a medical intervention is depicted in Fig. 1. Studies of the impact of CI have typically focused on the "clinical evaluation level" and clinical measures of efficacy (auditory skills, hearing thresholds, and speech) in order to evaluate treatment effect. However, these measures only represent a small portion of the effect that a cochlear implant has on a child's life. For instance, these tests would not measure a child's ability to consistently communicate needs and wants or the child's improved self-confidence when interacting with normal-hearing peers. Ideally, a valid HRQL measure would be able to capture such downstream effects.

Instruments that measure HRQL are broadly divided into two categories: health index surveys and health utility instruments. Health index surveys (Fig. 1) comprise batteries of items organized into scales, each of which measures a dimension of HRQL. In some instruments, the domains of HRQL are strictly defined as physical, mental, and social well being in accordance with the World Health Organization's definition of health [11]. Other instruments acknowledge that perceptions of HRQL may not be amenable to definition according to these components. For example, the Child Health and Illness Profile, a validated measure of child HRQL, is comprised of domains of satisfaction, comfort, risk avoidance, resilience, and achievement [12]. Within each of these domains, items query physical, mental, and/or social components of health. With health indices, scores are compiled across questions querying each different domain, and a single summary score or domain-specific score can be reported. These metrics can be generic or disease-specific, and they provide a rich source of information for approaching clinical questions or understanding the health status of patients.

Health utility metrics focus on a subject's broader conceptualization of HRQL through approaches that differ from health index surveys (Fig. 1). In trying to capture a patient's overall sense of well-being, patients are asked to assign a value to their current health state. Respondents are prompted to think about their health state across different domains of HRQL, and the respondent's assessment of their health state is measured on a continuum of 0–1. Health utility is most often measured with one of three commonly used utility metrics, the visual analog scale, time trade-off, or standard gamble [13,14]. Although, health utility metrics do not provide detailed clinical information, they are useful for assessing the cost-utility of medical treatments since the scales are

universally anchored on perfect health and death [13,15].

Measuring HRQL in children is complicated by several conceptual, methodological, and developmental issues that have been reviewed extensively elsewhere [16–19]. With respect to pediatric CI, two major issues arise. First, there are challenges in conceptualizing, in a comprehensive manner, what the salient domains of HRQL would be for a child and how these domains would change through development. Domains that comprise HRQL for a 5-year-old girl who is just beginning to start school should be different than those for a 16-year-old boy who is just starting to drive. A valid study would, therefore, have to choose an appropriate instrument that measures the domains of HRQL relevant for the target population. Second, HRQL measures in children are often completed by parental-proxy rather than by the child. Previous studies have demonstrated poor correlation between parent and child scores for mental- and social-related domains of HRQL and better correlation for physical-related domains [20]. The interpretation of HRQL results must, therefore, take into consideration the choice of survey-respondent, and when possible, both parent and child-reported scores should be used.

Given the inherent difficulties in conceptualizing and applying HRQL measures to children, our objective was to evaluate how HRQL has been assessed in previously published pediatric CI studies in order to determine whether conclusions could be drawn that would inform future investigations. In this review, we present a systematic review and summary of these studies. Specifically, we address: (1) the characteristics of studies using HRQL to assess outcomes of pediatric CI, and (2) the types of HRQL instruments that have been used and the key results and conclusions generated.

2. Methods

2.1. Search strategy

Our search strategy was based on a focused clinical question of "How has HRQL been measured in children with cochlear implants?" To identify all eligible studies, we searched PubMed (1953–2005), EMBASE (1974–2005), CINAHL (1982–2005), PsycInfo (1887–2005), and Web of Science (1945–2005) from June 20 to July 7, 2005. We attempted to capture all relevant studies by searching databases that included biomedical journals (PubMed and EMBASE), journals relevant to audiology and the communication sciences (CINAHL), and psychology journals (PsycInfo and Web of Science). We used variations

of the following search string for all databases: “(Cochlear implant*) AND (quality of life OR effectiveness OR cost utility OR cost effectiveness OR psychology OR health status OR program evaluation OR parent*) AND (pediatric OR child* OR infant OR toddler)”. We also hand-searched personal literature files and the reference lists of all articles found to be eligible for our systematic review [21–30].

2.2. Study selection

Potential studies were reviewed in two stages by a single investigator. In the first stage, titles and abstracts of all retrieved citations were reviewed for eligibility. Citations were selected if they met all the following inclusion criteria: (1) original peer-reviewed research article; (2) enrolled subjects <18 years; (3) children had cochlear implants; (4) study used a patient or proxy measure of multidimensional functional status, HRQL, or health status; and (5) in English. In the second stage, full-length articles of selected citations were reviewed. Articles were accepted for inclusion in our study if they met the previous inclusion criteria in addition to all of the following: (1) separated HRQL results for CI and non-CI children if both groups were included in the study; (2) used a HRQL instrument that incorporated components of physical, mental, and social health; and (3) the HRQL instrument was published with the article if an ad hoc instrument was used.

2.3. Data extraction

For each eligible article, data were extracted using custom-designed forms by a single investigator.

Collected data included: study demographics (including age at time of CI and at study), study purpose, HRQL instrument used and method of administration, and qualitative/quantitative HRQL results.

3. Results

3.1. Search results and study characteristics

Our search query of five databases yielded 667 retrieved citations (Fig. 2). An additional four, non-duplicate, citations were found from hand-searching personal literature files and the reference lists of relevant articles. The initial primary screen that consisted of reviewing titles and abstracts and eliminating duplicate articles yielded 31 potentially relevant citations. The secondary screen involved reviewing the full-length articles of these 31 citations of which 21 were excluded. The most common reason for exclusion was that a HRQL instrument incorporating components of physical, mental, and social health was not used. Our search strategy yielded 10 articles that were found to be eligible for inclusion in our systematic review (Table 1).

All studies used a cross-sectional design to capture HRQL data, and, therefore, no studies captured longitudinal changes in HRQL. Kelsay and Tyler [21] attempted to document changes in HRQL at sequential cross-sectional time points after CI (1–3 years after CI). However, different subjects were enrolled at each time point. Cheng et al. [22] measured HRQL by having parents rate their child’s HRQL an average

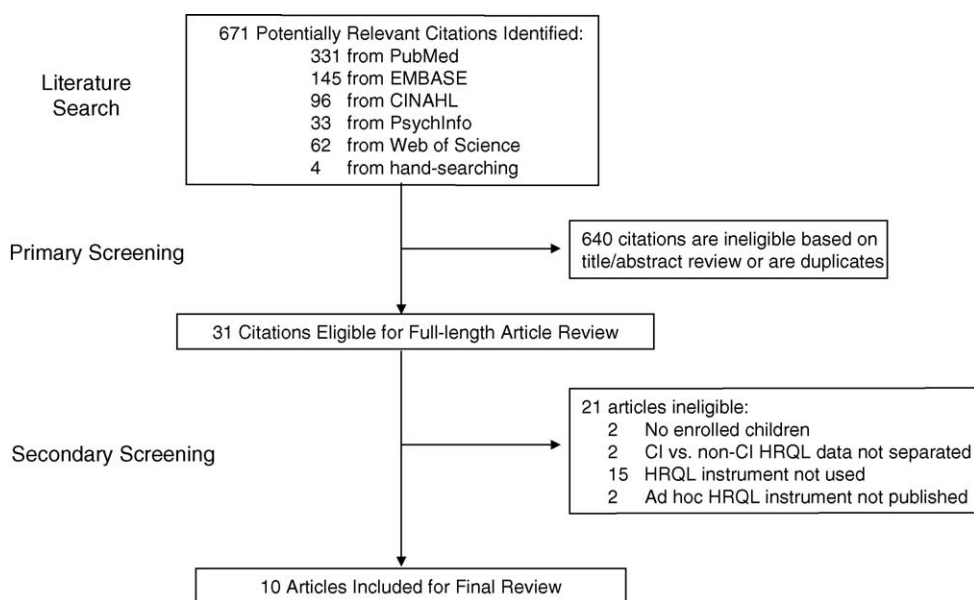


Fig. 2 Screening and selection process of retrieved citations.

Table 1 Evidence table of reviewed articles

Author	N	Prelingually deaf or deaf <age 3 (%)	Age at CI ^a	Years from CI to study ^a	Age of subjects at study ^a	Respondent	Main points
Generic Cheng et al. [22]	78	93	5.7 (4.2)	1.9 (2.0)	7.5 (4.5)	Parent	<i>Objective:</i> to determine the QoL and cost-consequences for deaf children who receive a CI (1) Used generic health utility metrics (VAS, TTO, HUI) ^b (2) Change in health utility from immediately before to after CI: VAS 0.59 → 0.86; TTO 0.75 → 0.97; HUI 0.25 → 0.64
Huber [23]	29	90	Children: 4.3 (2.0) Adolescent: 7.6 (1.8)	>3	8–16	Parent and Child	<i>Objective:</i> to obtain initial, fundamental information on HRQL of children with a CI with reference to their hearing counterparts (1) Used KINDLr generic HRQL survey (2) For CI children (8–12): self-ratings of HRQL were significantly lower than hearing children scores. Parent-proxy ratings of HRQL were significantly higher than their child's self-ratings (3) For female CI adolescents (13–16): self-ratings, parent-proxy ratings, and normal hearing scores were not significantly different from each other
Ad hoc Kelsay and Tyler [21]	<25 ^c	100	2–16	1–3	2–13	Parent	<i>Objective:</i> to document advantages and disadvantages that parents associated with their children's use of a multichannel CI (1) Used an ad hoc open-format, qualitative questionnaire asking parents to list benefits/disadvantages of CI for their child (2) At 1–3 years post-CI, the most common benefits to CI children were awareness of environmental sounds, speech perception/production, lifestyle (activities, independence, friendships) (3) Psychological benefits (increased self-confidence and self-esteem) only identified by 10% parents at 3 years post-CI

Table 1 (Continued)

Author	N	Prelingually deaf or deaf <age 3 (%)	Age at CI ^a	Years from CI to study ^a	Age of subjects at study ^a	Respondent	Main points
Beadle et al. [25]	17	N.S.	2–10	>0.5	N.S.	Parent	<p><i>Objective:</i> to examine the relationship between QoL of the child as reported by parental proxy and parental perceptions about the process of implantation, outcomes, and support received</p> <p>(1) Used an ad hoc, quantitative 10-point HRQL scale (2) Mean HRQL score on 10-pt scale: 7.9 (range 2–10) (3) HRQL scores correlated with parental assessment of outcome as measured by ad hoc 46-item survey ($r = 0.77, p < 0.001$)</p>
Archbold et al. [24]	30	100	2–10	3	5–13	Parent	<p><i>Objective:</i> to explore the perceptions of parents without limiting them to prescribed issues</p> <p>(1) Used an ad hoc open-format, qualitative questionnaire with 17 prompt headings (2) Major themes throughout all responses: (a) improvement in child's confidence and communication ability; (b) increasing dependency on technology and need for long-term educational/implant center support (3) Results formed basis of developing closed-format questionnaire</p>
Vlahovic and Sindija [26]	11	N.S.	2–6	1–5	4–11	Parent	<p><i>Objective:</i> to assess the influence of potentially limiting factors on post-implant outcome</p> <p>(1) Used an ad hoc, quantitative 13-item closed-format questionnaire with children with "potentially limiting factors" (additional disabilities, cochlear malformation/ossification, reimplantation, or cochlear nerve pathology) (2) "Benefit" seen for all children with limiting factors except for child with cochlear nerve pathology</p>
Parents views and experiences with pediatric CI (PVEIQ) Incesulu et al. [29]	27	100	N.S.	1–3	2–13	Parent	<p><i>Objective:</i> to evaluate parental perspectives regarding CI and the child's progress after a minimum of 1 year after CI</p> <p>(1) Authors adapted the PVEIQ to include 11 domains and 58 items</p>

Nicholas and Geers [28]	181	100	<5	4–6	8–9	Parent	(2) Parents reported improvements in communication skills, social relationships, and self-confidence <i>Objective:</i> to document the psychosocial adjustment of a representative sample of school-aged deaf children who have had a CI since their preschool years (1) Authors used 46-items of the PVEIQ, rescaled items to same scale (negative to positive), and scored surveys as percent of potential maximal agreement. HRQL scores correlated with speech perception/production, oral/total language, and reading (2) Parents expressed “a generally positive view of CI”
O’Neill et al. [27]	17	100	N.S.	>2	N.S.	Parent	<i>Objective:</i> to test the reliability of a validated closed-format questionnaire assessing parental views following CI of the child (1) The authors assessed the test-retest reliability of the 107-item questionnaire (2) 35 of the 40 items in HRQL-related domains had Pearson correlations >0.5
Nunes et al. [30]	61	N.S.	N.S.	>3	5–16	Parent	<i>Objective:</i> to carry out an independent assessment of a parent outcome questionnaire for pediatric CI designed by Archbold et al. (1) The authors’ objective was to assess the validity of the 74-item questionnaire (2) Content validity was adequate. The authors also identified missing themes (3) Criterion validity was acceptable. For 8 of 10 themes, extreme questionnaire scores converged with semistructured interview results. Only four themes were suitable for correlation against interview scores of which two had acceptable correlation (4) The scaling properties of the items/domains and internal construct validity were not assessed with a factor analysis

N.S.: not stated in article.

^a In years: range, mean (standard deviation), or no. of years.

^b VAS: visual analog scale; TTO: time-trade off; HUI: health utilities index mark III.

^c This study enrolled 25 subjects who were 1 year post-CI, 21 subjects at 2 years post-CI, and 22 subjects at 3 years post-CI. Different subjects were enrolled in each time period.

of 2 years after their child's surgery. At this time point, the authors also had parents retrospectively rate what their child's HRQL *had been* 1-year and immediately before implantation. The authors argue that this approach to measuring changes in HRQL can be justified on the basis that parents and children regularly revisit the state of deafness when the implant is removed or if there are malfunctions. They also observed that scores gathered from a small group of parents immediately before their child's implantation were "nearly identical" to the retrospectively gathered scores used in their study.

The sample size of the 10 studies ranged from 11 to 181 subjects (Table 1). Seven of the 10 studies explained the subject selection process and the number of non-respondents [22–25,27,29,30]. Two studies accounted for the characteristics of the non-respondents and addressed potential selection bias in measured HRQL scores [22,23].

There was substantial inter- and intra-study heterogeneity in the age of the children at time of CI, the duration of implant use, and the age of the subjects at the time of survey. Several studies did not include these descriptors of their study population [25,27,29,30]. The greatest range in age at implantation was seen in the Kelsay and Tyler study [21] where the children ranged from 2 to 16 at the time of implantation. Most studies only reported a minimum number of years from implantation to survey without stating an actual range. One study [28] had a well-defined study population where all children were implanted at <5 years, and HRQL was measured when all children were 8–9 years old.

3.2. HRQL instruments

The HRQL metrics used in these 10 studies can be broadly divided into three categories: generic HRQL instruments, ad hoc instruments created specifically for the purposes of the study, and the parents views and experiences with pediatric CI questionnaire (PVEIQ) developed by Archbold et al. [24].

3.2.1. Generic

Generic HRQL instruments used included health utility metrics and the KINDLr. Cheng et al. [22] used three different types of health utility metrics to measure HRQL: visual analog scale (VAS), time trade off (TTO), and the health utilities index mark III (HUI). Each of these methods results in a summary HRQL score between 0 and 1 for the subject. An extensive discussion of these instruments is beyond the scope of this article, but excellent reviews can be found elsewhere [13]. Briefly, the VAS presents the subject with a line with grid marks from 0

("death") to 100 ("perfect health"), and the subject is asked to draw a line corresponding to their HRQL. The TTO [31] presents the subject with two alternatives: living in their current state of health for time t (remaining life expectancy) or living in "perfect health" for time x . The duration of x is varied until the subject is ambivalent between the two options; the subject's HRQL is then expressed as x/t . Finally, the HUI was developed by Torrance and co-workers as a population-based measure of HRQL [32]. This instrument divides health status into eight different attributes (e.g. hearing, mobility) with five or six levels per attribute. Subjects specify which attributes best describe themselves, and the subject is then mapped onto one of 972,000 possible health states. A utility score is assigned to the person's health state based on a multiattribute utility function that was derived from the preferences of a sample of adults in Ontario, Canada.

One study [23] used the KINDLr, a generic 24-item HRQL instrument, that was developed in Germany and that has been validated for use in 8–16 years old children [33]. The instrument is composed of six scales: physiological and psychological well-being, self-esteem, family, friends, and functioning. Both child-report and parent-proxy versions are available, and normative values have been established for comparison.

3.2.2. Ad hoc

Ad hoc HRQL instruments that are designed for the purposes of an individual study can be qualitative or quantitative and were used in four reports [21,24–26]. Ad hoc qualitative measures provide rich detailed information and are useful to understand the broad impact of a treatment on HRQL. These instruments typically rely on using open-ended questionnaires and possibly semi-structured interviews to clarify and focus responses. In contrast, ad hoc quantitative instruments allow for simple numerical summary scores but are difficult to interpret since the instruments have not been validated and normative scores are not available. Two studies [21,24] used qualitative, open-ended questionnaires, and two studies developed and used a quantitative instrument [25,26].

3.2.3. Parents views and experiences with pediatric CI questionnaire (PVEIQ)

Archbold et al. developed the PVEIQ, a closed-format questionnaire, based on qualitative research using parental responses to an open-ended questionnaire [24]. This closed-format questionnaire contains 10 domains that broadly capture parental perceptions of their child and the implantation process. Five of these domains are related to the

HRQL of the child: communication, general functioning, self-reliance, well-being and happiness, and social relationships. The earliest reference to this survey contained 107 items [27] but has now been reduced to 74 items [30].

O'Neill et al. studied the test-retest reliability of the survey and found it to be adequate with 35 of the 40 items comprising the five HRQL domains having Pearson correlation coefficients >0.5 [27]. Nunes et al. (25) assessed the validity of the survey. Content validity was found to be adequate, but the authors identify missing concepts based on their interviews with CI children and parents. A factor analysis was not performed to assess the scaling properties (e.g. unidimensionality) of the items assigned to each domain. External construct validity of the PVEIQ (whereby this instrument is compared to other validated measures based on *a priori* hypotheses) was not performed [34]. The authors suggest making further refinements to the instrument.

Heterogeneity in HRQL instruments used in the 10 studies prevented a quantitative analysis (meta-analysis) of HRQL data. The results of individual studies are, therefore, qualitatively summarized by type of HRQL instrument (generic HRQL instruments, ad hoc survey instruments, and the PVEIQ), and key conclusions from each study are presented in Table 1.

4. Discussion

The necessity of studying HRQL outcomes in CI children is highlighted by the importance of measuring patient-reported outcomes [35] and patient-defined problems [36]. HRQL outcomes can also inform policy, and long-standing and slow to evolve policy regulations have had a direct effect on access to CI and CI-related services [37]. In order to understand the needs of deaf children and to study the real-world impact of different treatment factors (e.g. unilateral versus bilateral CI, age at implantation), instruments that measure progress from the child and family perspective are needed [38]. This approach is also underscored by economic forces and the requirements of health-technology assessments that necessitate the collection of HRQL data [39]. This systematic review of the pediatric CI literature revealed a diversity of studies, each using heterogeneous study populations and HRQL instruments. Several conclusions based on a qualitative review of the data emerge that are informative to future investigations.

We observed that studies that used a generic HRQL instrument resulted in stronger conclusions with greater scientific-validity. This is a result of the

generic instruments having had previous validation and the availability of normative scores or a framework for interpreting the scores from these instruments. For example, the objective of the study by Huber [23] was to evaluate the HRQL of CI children with respect to normal hearing children. By applying the KINDLr, a generic instrument for which batteries of age-specific normative scores were available, the author was able to demonstrate that 8–12 years old CI children had lower self-rated HRQL than hearing children and that these scores were discrepant from parent-reported scores. In contrast, the objective of the Vlahovic and Sindija study was to evaluate whether children with additional disabilities (e.g. cochlear malformation) would benefit from a CI. They used an *ad hoc* 13-item HRQL instrument, and the authors concluded that benefit with a CI was observed in these children. In this case, benefit was defined as a score that improved on their 13-item scale. However, without prior validation, the clinical significance of the score change and the external validity of the 13 items in assessing benefit are arbitrary and subject to the authors' bias.

A similar interpretation applies to studies using the PVEIQ. Since the PVEIQ was designed to be a measure of parental perceptions and not a direct measure of HRQL, only five of the instrument's domains are relevant to pediatric HRQL. Normative scores or a framework for interpreting scores are also unavailable. The result was that in studies such as those by Incesulu et al. [29] and Nicholas and Geers [28], authors had to choose to include only a subset of items guided by their clinical judgment, and they were also forced to develop *ad hoc* methods of interpreting scores. Incesulu et al. [29] qualitatively summarized results from the instrument, while Nicholas and Geers [28] rescaled scores and expressed total scores as a percent of maximal agreement. The lack of availability of a well-validated HRQL instrument results in conclusions that are subject to personal judgment about which items to include and how to interpret scores.

The design of a clinical study with well-defined inclusion criteria is fundamental to the validity of the scientific results. In 9 of the 10 studies included in this review, there was substantial heterogeneity in the ages of the children at CI, the duration of CI use, and the ages of the children at the time of the survey. The association between speech/language abilities and age at implantation and duration of implant use is well-established, and therefore, heterogeneity in these factors could result in study populations with widely variable language skills [8]. Since conceptual models of HRQL posit that a person's functional status (i.e. language, speech skills) will have a direct effect on perceptions of

health and HRQL [40], heterogeneity in age and duration of use also leads to variability in HRQL scores. For example, in one study [22], children were on average 5.7 ± 4.2 years old at time of CI and the duration of implant use among subjects had mean 1.9 ± 2 years. HRQL was then reported as an average of the group's scores. However, this mean score could have been higher or lower if a different cohort of children with different ages and duration of implant use was enrolled. The simplest approach to dealing with this potential source of bias is to include strict inclusion criteria in the study design. For example, in Nicholas and Geers study, all subjects were 8–9 years old and had received their implant 4–6 years ago.

4.1. Limitations of present review

This systematic review has several limitations. First, this review was restricted to the English language, and it is possible that some studies were excluded. However, this may be minimal given that many authors from non-English language countries still often publish in English-language journals. Three of the studies in our review were from groups located in non-English language countries [23,26,29]. Our search strategy was also intentionally narrow in order to capture only those articles that have specifically used a HRQL instrument to evaluate children with cochlear implants. Other studies that have applied HRQL instruments to deaf children without cochlear implants were specifically excluded since the conceptual basis of HRQL may be different in these children. For example, since medical versus sociological models of deafness arise from widely different perspectives [41], families who choose to forgo CI may not see deafness as being a health issue and may have a different perspective on HRQL than families electing for a medical intervention such as CI. Our data extraction and screening process was also only carried out by a single investigator, and therefore, there is a slightly greater likelihood that a reference may be inadvertently excluded or data abstracted incorrectly. This may be minimal, though, given the precise study inclusion criteria used, the primarily qualitative data extracted, and the modest scope of this systematic review.

4.2. Future studies

Future studies investigating HRQL should use well-validated HRQL instruments and employ strict inclusion criteria in the study design. At present, well-validated HRQL instruments are limited to generic measures that are not deafness-specific. The PVEIQ

has the potential to be a valid, deafness-specific questionnaire; further refinement of the content and scaling properties and the development of normative scores will strengthen investigators' ability to interpret results and formulate clinically relevant conclusions when using this instrument. While the future validation or development of a deafness-specific HRQL instrument may allow for more specific clinical questions to be addressed (e.g. social functioning specifically related to hearing), currently available generic instruments have already demonstrated sufficient sensitivity given the broad impact that deafness has on a child [22,23,42]. In addition to the generic instruments discussed in this current review, other instruments such as the Child Health and Illness Profile [43,44], the PedsQL [45], and the Child Health Questionnaire [46] could all be applied to CI children. The advantages of these three instruments are their comprehensive approach to conceptualizing HRQL, the availability of both parent and child-report forms, and established normative scores. A previous population-based study of children with congenital hearing loss has successfully used the Child Health Questionnaire to demonstrate HRQL decrements in 7–8 years old deaf children relative to hearing peers [42]. Extensive reviews of other validated, pediatric HRQL instruments are also available [16–19]. In a current multi-site cohort study of CI outcomes being conducted at our institution and five other sites throughout the United States, we plan to explore the use of the Child Health and Illness Profile as a longitudinal measure of HRQL in parallel with other speech and language measures.

Future well-conducted studies that systematically evaluate the HRQL of cochlear-implanted children will allow us to assess the impact of CI during critical phases of early development in terms that are relevant to families and society. Ideally, HRQL data will also lead to a better understanding of criteria for selecting childhood candidates for implantation, the rehabilitative needs of CI children, and enable access to services that allow deaf children to use implantable technologies to their fullest potential.

References

- [1] J. Holt, Classroom attributes and achievement test scores for deaf and hard of hearing students, *Am. Ann. Deaf.* 139 (1994) 430–437.
- [2] A.E. Carney, M.P. Moeller, Treatment efficacy: hearing loss in children, *J. Speech Lang. Hear. Res.* 41 (1998) S61–S84.
- [3] L.S. Schilling, E. DeJesus, Developmental issues in deaf children, *J. Pediatr. Health Care* 7 (1993) 161–166.
- [4] M. Marschark, Origins and interactions in the social, cognitive, and language development of deaf children, in:

- M. Marschark, M.D. Clark (Eds.), *Psychological Perspectives on Deafness*, Lawrence Erlbaum Associates, Hillsdale, NJ, 1993, pp. 7–26.
- [5] C. Vaccari, M. Marschark, Communication between parents and deaf children: implications for social-emotional development, *J. Child Psychol. Psychiatry* 38 (1997) 793–801.
- [6] J.B. Tomblin, L. Spencer, S. Flock, R. Tyler, B. Gantz, A comparison of language achievement in children with cochlear implants and children using hearing aids, *J. Speech Lang. Hear. Res.* 42 (1999) 497–509.
- [7] M.A. Svirsky, S.W. Teoh, H. Neuburger, Development of language and speech perception in congenitally, profoundly deaf children as a function of age at cochlear implantation, *Audiol. Neurootol.* 9 (2004) 224–233.
- [8] E. Thoutenhoofd, S. Archbold, S. Gregory, M.E. Lutman, T.P. Nikolopoulos, T. Sach, Paediatric cochlear implantation: evaluating outcomes, Elsevier (2005).
- [9] S. Vidas, R. Hassan, L.S. Parnes, Real-life performance considerations of four pediatric multi-channel cochlear implant recipients, *J. Otolaryngol.* 21 (1992) 387–393.
- [10] B. Spilker, D. Revicki, Taxonomy of Quality of Life, in: B. Spilker (Ed.), *Quality of Life and Pharmacoeconomics in Clinical Trials*, Lippincott Williams & Wilkins, Baltimore, 1996, pp. 25–31.
- [11] World Health Organization, *The First 10 Years of the World Health Organization*, WHO, Geneva, 1958.
- [12] B. Starfield, M. Bergner, M. Ensminger, A. Riley, S. Ryan, B. Green, P. McGauhey, A. Skinner, S. Kim, Adolescent health status measurement: development of the child health and illness profile, *Pediatrics* 91 (1993) 430–435.
- [13] D.G. Froberg, R.L. Kane, Methodology for measuring health-state preferences. II. Scaling methods, *J. Clin. Epidemiol.* 42 (1989) 459–471.
- [14] G.W. Torrance, Measurement of health state utilities for economic appraisal, *J. Health Econ.* 5 (1986) 1–30.
- [15] D.G. Froberg, R.L. Kane, Methodology for measuring health-state preferences. I. Measurement strategies, *J. Clin. Epidemiol.* 42 (1989) 345–354.
- [16] S. Petrou, Methodological issues raised by preference-based approaches to measuring the health status of children, *Health Econ.* 12 (2003) 697–702.
- [17] M. De Civita, D. Regier, A.H. Alamgir, A.H. Anis, M.J. Fitzgerald, C.A. Marra, Evaluating health-related quality-of-life studies in paediatric populations: some conceptual, methodological and developmental considerations and recent applications, *Pharmacoeconomics* 23 (2005) 659–685.
- [18] L.S. Matza, A.R. Swensen, E.M. Flood, K. Secnik, N.K. Leidy, Assessment of health-related quality of life in children: a review of conceptual, methodological, and regulatory issues, *Value Health* 7 (2004) 79–92.
- [19] C. Eiser, R. Morse, The measurement of quality of life in children: past and future perspectives, *J. Dev. Behav. Pediatr.* 22 (2001) 248–256.
- [20] C. Eiser, R. Morse, Can parents rate their child's health-related quality of life? Results of a systematic review, *Qual. Life Res.* 10 (2001) 347–357.
- [21] D.M. Kelsay, R.S. Tyler, Advantages and disadvantages expected and realized by pediatric cochlear implant recipients as reported by their parents, *Am. J. Otol.* 17 (1996) 866–873.
- [22] A.K. Cheng, H.R. Rubin, N.R. Powe, N.K. Mellon, H.W. Francis, J.K. Niparko, Cost-utility analysis of the cochlear implant in children, *JAMA* 284 (2000) 850–856.
- [23] M. Huber, Health-related quality of life of Austrian children and adolescents with cochlear implants, *Int. J. Pediatr. Otorhinolaryngol.* 69 (2005) 1089–1101.
- [24] S.M. Archbold, M.E. Lutman, S. Gregory, C. O'Neill, T.P. Nikolopoulos, Parents and their deaf child: their perceptions 3 years after cochlear implantation, *Deaf. Educ. Int.* 4 (2002) 12–40.
- [25] E.A. Beadle, A. Shores, E.J. Wood, Parental perceptions of the impact upon the family of cochlear implantation in children, *Ann. Otol. Rhinol. Laryngol. Suppl.* 185 (2000) 111–114.
- [26] S. Vlahovic, B. Sindija, The influence of potentially limiting factors on paediatric outcomes following cochlear implantation, *Int. J. Pediatr. Otorhinolaryngol.* 68 (2004) 1167–1174.
- [27] C. O'Neill, M.E. Lutman, S.M. Archbold, S. Gregory, T.P. Nikolopoulos, Parents and their cochlear implanted child: questionnaire development to assess parental views and experiences, *Int. J. Pediatr. Otorhinolaryngol.* 68 (2004) 149–160.
- [28] J.G. Nicholas, A.E. Geers, Personal, social, and family adjustment in school-aged children with a cochlear implant, *Ear Hear* 24 (2003) 695–815.
- [29] A. Incesulu, M. Vural, U. Erkam, Children with cochlear implants: parental perspective, *Otol. Neurotol.* 24 (2003) 605–611.
- [30] T. Nunes, U. Pretzlik, S. Ilicak, Validation of a parent outcome questionnaire from pediatric cochlear implantation, *J. Deaf Stud. Deaf Educ.* (2005).
- [31] G.W. Torrance, W.H. Thomas, D.L. Sackett, A utility maximization model for evaluation of health care programs, *Health Serv. Res.* 7 (1972) 118–133.
- [32] D. Feeny, G.W. Torrance, W. Furlong, Health utilities index, in: B. Spilker (Ed.), *Quality of Life and Pharmacoeconomics in Clinical Trials*, Lippincott Williams & Wilkins, Baltimore, 1996, pp. 239–252.
- [33] U. Ravens-Sieberer, M. Bullinger, Assessing health-related quality of life in chronically ill children with the German KINDL: first psychometric and content analytical results, *Qual. Life Res.* 7 (1998) 399–407.
- [34] E. Juniper, G. Guyatt, P. Jimenez, How to develop and validate a new health-related quality of life instrument, in: B. Spilker (Ed.), *Quality of Life and Pharmacoeconomics in Clinical Trials*, Lippincott Williams & Wilkins, Baltimore, 1996, pp. 49–56.
- [35] E.A. Zerhouni, US biomedical research: basic, translational, and clinical sciences, *JAMA* 294 (2005) 1352–1358.
- [36] B. Starfield, Quality-of-care research: internal elegance and external relevance, *JAMA* 280 (1998) 1006–1008.
- [37] N.M. Kane, P.D. Manoukian, The effect of the medicare prospective payment system on the adoption of new technology. The case of cochlear implants, *N. Engl. J. Med.* 321 (1989) 1378–1383.
- [38] R.E. Stein, B. Stanton, B. Starfield, How healthy are US children? *JAMA* 293 (2005) 1781–1783.
- [39] A.Q. Summerfield, D.H. Marshall, Paediatric cochlear implantation and health-technology assessment, *Int. J. Pediatr. Otorhinolaryngol.* 47 (1999) 141–151.
- [40] I.B. Wilson, P.D. Cleary, Linking clinical variables with health-related quality of life. A conceptual model of patient outcomes, *JAMA* 273 (1995) 59–65.
- [41] I.M. Munoz-Baell, M.T. Ruiz, Empowering the deaf. Let the deaf be deaf, *J. Epidemiol. Commun. Health* 54 (2000) 40–44.
- [42] M. Wake, E.K. Hughes, C.M. Collins, Z. Poulakis, Parent-reported health-related quality of life in children with congenital hearing loss: a population study, *Ambul. Pediatr.* 4 (2004) 411–417.
- [43] A.W. Riley, C.B. Forrest, G.W. Rebok, B. Starfield, B.F. Green, J.A. Robertson, P. Friello, The child report form of the chip-child edition: reliability and validity, *Med. Care* 42 (2004) 221–231.

- [44] B. Starfield, M. Bergner, M. Ensminger, A. Riley, S. Ryan, B. Green, P. McGauhey, A. Skinner, S. Kim, Adolescent health status measurement: development of the child health and illness profile, *Pediatrics* 91 (1993) 430–435.
- [45] J.W. Varni, M. Seid, C.A. Rode, The PedsQL: measurement model for the pediatric quality of life inventory, *Med. Care* 37 (1999) 126–139.
- [46] J.M. Landgraf, E. Maunsell, K.N. Speechley, M. Bullinger, S. Campbell, L. Abetz, J.E. Ware, Canadian-French, German and UK versions of the child health questionnaire: methodology and preliminary item scaling results, *Qual. Life Res.* 7 (1998) 433–445.
- [47] B. Spilker, Introduction, in: B. Spilker (Ed.), *Quality of Life and Pharmacoeconomics in Clinical Trials*, Lippincott Williams & Wilkins, Baltimore, 1996, pp. 1–10.

Available online at www.sciencedirect.com

