Research

Hearing and speech outcomes in children with cochlear nerve hypoplasia who underwent cochlear implantation

Rindy Yunita Pratamisiwi*, Semiramis Zizlavsky*, Harim Priyono*, Indrati Suroyo, Setyo Handryastuti***, Joedo Prihartono******

*Department of Otorhinolaryngology Head and Neck Surgery, Faculty of Medicine, Universitas Indonesia/RSUPN dr. Cipto Mangunkusumo, Jakarta

**Department of Radiology, Faculty of Medicine, Universitas Indonesia/RSUPN dr. Cipto Mangunkusumo, Jakarta

***Department of Pediatric, Faculty of Medicine, Universitas Indonesia/RSUPN dr. Cipto Mangunkusumo, Jakarta

****Department of Community Health, Faculty of Medicine, Universitas Indonesia/RSUPN dr. Cipto Mangunkusumo, Jakarta

ABSTRACT

Background: Data on hearing and speech outcomes following cochlear implantation in prelingual sensorineural hearing loss (SNHL) children with cochlear nerve hypoplasia, especially in Indonesia, is still limited. **Purpose:** To evaluate the hearing and speech function of prelingual SNHL children who had undergone cochlear implantation. **Method:** A retrospective cross-sectional study was conducted on 28 children who had undergone cochlear implantation procedures at the Department of Otorhinolaryngology and Head and Neck Surgery, Dr. Cipto Mangunkusumo National General Hospital, Jakarta, Indonesia, for more than 12 months. The hearing and speech function of the subjects were assessed using the modified Categories of Auditory Performance II (CAP-II) and the speech intelligibility rating (SIR) scores. **Result:** Children with cochlear nerve hypoplasia showed an increase in CAP-II and SIR scores after using cochlear implants for more than 12 months (versus pre-surgery, high CAP score [≥5]: 89.3% [25/28 children] vs 3.6% [1/28 children] and high SIR score [≥3]: 82.1% [23/28 children] vs 7.1% [2/28 children]; both $p<0.001$). Post-operative CAP-II and post-operative SIR scores were highly positively correlated ($r=0.705$, $p<0.001$). Types of cochlear nerve hypoplasia (unilateral vs bilateral) were not significantly associated with post-operative CAP-II and SIR scores ($p=0.382$ and $p=0.459$, respectively). **Conclusion:** Prelingual SNHL children with cochlear nerve hypoplasia may still get the benefit from cochlear implantation, as shown by a remarkable improvement in hearing and speech function as assessed with the CAP-II and SIR scores.

Keywords: auditory perception, cochlear implants, cochlear nerve hypoplasia, prelingual sensorineural hearing loss, speech intelligibility

ABSTRAK

Latar belakang: *Data mengenai kemampuan auditori dan wicara pasca implan koklea pada anak dengan gangguan pendengaran sensorineural (GPSN) prelingual, yang disertai hipoplasia nervus koklearis, terutama di Indonesia, masih sangat terbatas.* ***Tujuan:*** *Untuk mengevaluasi fungsi auditori dan wicara pada anak dengan GPSN prelingual yang telah menjalani implan koklea.* ***Metode:*** *Studi potong lintang retrospektif dilakukan pada 28 subjek (56 telinga) yang telah menjalani operasi implantasi koklea di RSUPN Cipto Mangunkusumo, selama lebih dari 12 bulan. Fungsi auditori dan wicara dinilai dengan menggunakan skor* modified Categories of Auditory Performance II (CAP-II) *dan* Speech Intelligibility Rating (SIR). ***Hasil:*** *Anak dengan kondisi hipoplasia nervus koklearis pada evaluasi menunjukkan peningkatan nilai* CAP-II *dan* SIR *setelah menggunakan implan koklea lebih dari 12 bulan (dibandingkan dengan nilai pra-operasi, nilai* CAP-II *tinggi [≥5]: 89,3% [25/28 anak]* *versus 3,6% [1/28 anak] dan nilai* SIR *tinggi [≥3]: 82,1% [23/28 anak] versus 7,1% [2/28 anak]; keduanya p<0,001). Nilai* CAP-II *dan* SIR *pasca implan koklea memiliki hubungan korelasi positif yang kuat (r=0,705, p<0,001). Tipe dari hipoplasia nervus koklearis (unilateral versus bilateral) tidak memiliki hubungan yang signifikan dengan nilai* CAP-II *dan* SIR *pasca operasi (p=0,382 dan p=0,459).* ***Kesimpulan:*** *Anak dengan GPSN yang disertasi hipoplasia nervus koklearis dapat memperoleh manfaat dari pemasangan implan koklea, ditandai dengan adanya peningkatan kemampuan auditori dan wicara berdasarkan penilaian skor* CAP-II *dan* SIR.

Kata kunci: *persepsi auditori, implan koklea, hipoplasia nervus koklearis, tuli sensorineural prelingual, kemampuan wicara*

Correspondence address: Rindy Yunita Pratamisiwi, MD. Department of Otorhinolaryngology Head and Neck Surgery (ORL-HNS), Faculty of Medicine, Universitas Indonesia, Jakarta, Indonesia.
Email: rindyyunita@yahoo.com

INTRODUCTION

Language ability is one of the most important indicators in assessing a child's development, in which a child is diagnosed with speech delay when their speech ability and/or patterns are inferior to those of other children their age. The single primary cause of speech delay in children is hearing loss, whether it is conductive, sensorineural, or mixed type hearing loss; which highlights the crucial role of early habilitation and rehabilitation of hearing function in these children to mitigate potential developmental delays.[1,2]

Cochlear implantation, which works by amplifying the sound stimuli and converting them to electrical stimuli, is an effective method to improve the hearing function of patients with sensorineural hearing loss (SNHL). The procedure is usually indicated for children aged more than 12 months with bilateral severe-to-profound hearing loss refractory to conventional hearing aids.[3] The success of cochlear implantation may vary widely and is influenced by various factors, including patient characteristics such as age at implantation, progression of hearing impairment, and cochlear nerve morphology; implant device factors such as the number of electrodes and channels and the precision of electrode placement in the cochlea; as well as environmental and family factors.[4] Among these factors, the morphology and function of the cochlear nerve is one of the most important predictors of cochlear implantation success considering its essential function to transmit auditory impulses from the receptor to the central nervous system. Previous studies have found that cochlear nerve size, which may be measured by magnetic resonance imaging (MRI),[5] is inversely associated with the duration and degree of hearing loss.[3,6] This suggests the importance of pre-operative screening for cochlear nerve abnormalities to enhance the outcomes of cochlear implantation in potential patients.

The evaluation of cochlear implantation outcomes should utilize objective and user-friendly instruments that can be easily accessed and used by both healthcare providers and patients, including children and their caregivers. This may be achieved by using the Categories of Auditory Performance (CAP) index and the Speech Intelligibility Rating (SIR) score. CAP-II, a modified version of the original CAP index invented by Archbold et al.[7] in 1995, aimed to assess the auditory perceptive ability of cochlear implant recipients.[8] On the other hand, SIR was developed by Cox and McDaniel in 1989 to evaluate the speech ability of cochlear implant users by measuring their daily spontaneous speech.[9]

While the hearing and speech outcomes following cochlear implantation had been studied in postlingual SNHL children with cochlear nerve hypoplasia,[3] data on prelingual SNHL children, especially in Indonesia, is still limited. Therefore, this study aimed to evaluate the hearing and speech function of prelingual SNHL children who had undergone cochlear implantation.

METHOD

This retrospective cross-sectional study included 28 cochlear implanted children with bilateral prelingual severe-to-profound SNHL presenting to the Ear, Nose, and Throat (ENT) clinic of the Department of Otorhinolaryngology Head and Neck Surgery, Dr. Cipto Mangunkusumo National General Hospital, Jakarta, Indonesia between July and December 2022. The children underwent the cochlear implantation procedures between 2018-2021, and had used the implants for more than 12 months prior to the study. All children were diagnosed with cochlear nerve hypoplasia based on re-measurement of the cross-sectional area (CSA) of the cochlear nerve on pre-operative MRI. Cochlear nerve hypoplasia was diagnosed when the CSA of the cochlear nerve was below 1.12±0.08 mm.[10]

The hearing and speech function of the children was assessed using the CAP-II index and the SIR score, respectively. Briefly, the CAP-II index consists of a hierarchical scale ranging from 0 ('displays no awareness of environmental sounds') to 9 ('ability to call an unfamiliar person with unpredictable speech context'),[8] while the SIR score comprises of five performance categories ranging from 1 ('pre-recognizable words in spoken language') to 5 ('connected speech is intelligible to all listeners').[11] The scores were assessed subjectively based on the observations by the children's caregivers, speech therapists, and direct observation by the authors using video recordings. The results of hearing function based on the CAP-II index is categorized into low (score 0-4) and high (score ≥5), while the results of speech ability based on the SIR score is categorized into low (score 1-2) and high (≥3).[8]

The collected data were presented in frequencies and proportions. The pre-operative and follow-up CAP-II and SIR scores were compared using McNemar tests, while the association between potential factors including type of cochlear nerve hypoplasia (unilateral versus bilateral), history of prenatal TORCH infection, age at diagnosis, age at intervention, and age at implantation, the duration of hearing aid use, age of cochlear implantation, and frequency of auditory verbal therapy (AVT); and the CAP-II and SIR scores were tested using Fisher's exact tests. All statistical analysis were performed using SPSS 24.0 (SPSS Inc., Chicago, IL), and a p-value of ≤0.05 denotes statistical significance.

RESULT

Among 28 children with cochlear nerve hypoplasia, 14 were boys (50.0%), 12 had prenatal TORCH infection (42.9%), and 4 had birth defect (14.3%). None of the children were diagnosed with behavioral disorder or physical disability. Re-measurement of cochlear nerve CSA based on pre-operative MRI showed that 25 children (89.3%) had bilateral cochlear nerve hypoplasia, while 3 subjects (10.7%) had unilateral hypoplasia. Characteristics of the included children were summarized in Table 1.

Table 1. Characteristics of the study population (N=28)

Characteristics	N	%
Gender		
Boys	14	50.0%
Girls	14	50.0%

Place of residence		
Jabodetabek[a]	15	53.6%
Others	13	46.4%
Birth defect		
Yes	4	14.3%
No	24	85.7%
Behavioral disorder		
Yes	0	0.0%
No	28	100%
Physical disability		
Yes	0	0.0%
No	28	100%
Prenatal TORCH infection[b]		
Yes	12	42.9%
No	16	57.1%
Cochlear nerve hypoplasia		
Unilateral	3	10.7%
Bilateral	25	89.3%
Age at diagnosis (years)		
0-3.5	27	96.4%
≥3.5	1	3.6%
Age at intervention (years)		
0-3.5	27	96.4%
≥3.5	1	3.6%
Duration of hearing aid use (months)		
<6	7	25.0%
≥6	21	75.0%
Age at implantation (years)		
0-3.5	11	39.3%
3.5-7	13	46.4%
>7	4	14.3%
Duration of implant use (years)		
1	1	3.6%
2	9	32.1%
3	10	35.7%
>3	8	28.6%
Side of implantation		
Unilateral	17	60.7%
Bilateral	11	39.2%
Frequency of AVT		
<1x/week	3	10.7%
1-2x/week	24	85.7%
>2x/week	1	3.6%

Pre-operative CAP-II score		
Low (<5)	27	96.4%
High (≥5)	1	3.6%
Pre-operative SIR score		
Low (<3)	26	92.9%
High (≥3)	2	7.1%

[a]Jabodetabek is a metropolitan area in Indonesia consisting of Jakarta, Bogor, Depok, Tangerang, and Bekasi. [b]TORCH infection includes toxoplasmosis, rubella, cytomegalovirus, herpes simplex, and other agents (syphilis and hepatitis-B). AVT: auditory verbal therapy; CAP-II: Category of Auditory Performance II index; SIR: speech intelligibility rating.

About 25 children (89.3%) had a high score of post-operative CAP-II score (≥5). Among them, 24 showed an increase in CAP-II score after cochlear implantation, while one patient had a high CAP-II score even before surgery. Meanwhile, 23 children (82.1%) had a high post-operative SIR score: 21 of them showed an increase in SIR score after cochlear implantation, and 2 subjects had a high SIR score even before surgery. The association between pre- and post-implantation CAP-II and SIR scores were statistically significant (both p<0.001; Table 2). In addition, post-operative CAP-II and post-operative SIR scores were highly positive correlated (r=0.705, p<0.001), indicating that children with high post-operative CAP-II score were more likely to have high post-operative SIR score.

Table 2. Distribution of CAP-II and SIR scores before and after cochlear implantation (N=28)

Score	Before implantation	After implantation	p-value
CAP-II			
Low (<5)	27 (96.4%)	3 (10.7%)	<0.001
High (≥5)	1 (3.6%)	25 (89.3%)	
SIR			
Low (<3)	26 (92.9%)	5 (17.9%)	<0.001
High (≥3)	2 (7.1%)	23 (82.1%)	

p-values were derived from McNemar test. CAP-II: Category of Auditory Performance II index; SIR: speech intelligibility rating.

In the study population, we did not find a significant relationship between type of cochlear nerve hypoplasia (unilateral versus bilateral) and pre- and post-operative CAP-II (p>0.999 and p=0.382), and SIR scores (p>0.999 and p=0.459). Moreover, prenatal TORCH infection was not statistically significant to post-operative CAP-II score (p=0.613), while none of the sought factors including age at diagnosis (p>0.999), age at intervention (p>0.999), age at implantation (p=0.574), the duration of hearing aid use (p=0.574), the duration of implant use (p=0.410), and frequency of auditory verbal therapy (p=0.694) were significantly related to SIR score. (Table 3)

Table 3. Factors associated with post-operative CAP-II and SIR scores

Variables	CAP-II; N (%)			SIR; N (%)		
	Low	High	p-value	Low	High	p-value
Prenatal TORCH infection[a]			0.613			
Yes						
No						

Cochlear nerve hypoplasia						
Unilateral	1 (33.3)	2 (66.7)	0.382	3 (100)	0 (0.0)	0.459
Bilateral	3 (12.0)	22 (88.0)		23 (92.0)	2 (8.0)	
Age at diagnosis (years)						
0-3.5						>0.999
≥3.5						
Age at intervention (years)						
0-3.5						>0.999
≥3.5						
Duration of hearing aid use (months)						
<6						0.574
≥6						
Age at implantation (years)						
0-3.5						0.574
3.5-7						
>7						
Duration of implant use (years)						
1						
2						0.410
3						
>3						
Frequency of AVT						
<1x/week						0.694
1-2x/week						
>2x/week						

p-values were derived from Fisher's exact tests. CAP-II and SIR scores were categorized into low (<5 and <3, respectively) and high (≥5 and ≥3, respectively). [a]TORCH infection includes toxoplasmosis, rubella, cytomegalovirus, herpes simplex, and other agents (syphilis and hepatitis- B). AVT: auditory verbal therapy; CAP-II: Category of Auditory Performance II index; SIR: speech intelligibility rating.

DISCUSSION

Cochlear nerve deficiency, either in the form of aplasia or hypoplasia, may be incidentally found in cochlear implant candidate patients with bilateral SNHL. This abnormality, if left untreated, may affect the hearing and speech outcomes of the children after implantation; which highlights the importance of regular screening and prompt treatment of cochlear implant candidate patients with cochlear nerve deficiency. Such regular screenings are usually performed with CAP-II and SIR scores as instruments for the evaluation of hearing and speech function. Those are easily accessible and readily available to healthcare providers and the children's caregivers. However, these methods are not yet widely implemented in Indonesia for children who have undergone cochlear implantation.

The hearing and speech function of children with cochlear nerve deficiency after cochlear implantation may vary greatly, ranging from limited increase in environmental sounds perception to achieving open-set speech recognition and fluent language abilities. A study by Vincenti et al.,[12] evaluating the speech perception of five children with cochlear implants using SIR score at 6, 12, and >12 months after surgery, revealed that only one child showed a significant improvement in speech abilities (SIR score 5) after >12 months post-implantation. This finding was corroborated by Kutz et al.,[13] who found that, although cochlear implanted children with cochlear nerve hypoplasia showed a better improvement in sound and speech awareness according to speech awareness threshold and speech perception categories index than children with cochlear nerve aplasia, the hearing and speech function of these children were still limited and suboptimal compared to children without cochlear nerve abnormalities. Previous reports had consistently shown that the hearing and speech outcomes of children with cochlear nerve deficiency who underwent cochlear implantation were poorer than those without cochlear nerve abnormalities, as tested using CAP and SIR scores.[13,14] This renders the morphology of cochlear nerve (i.e. diameter and number of nerve fibers) are significant predictors of hearing and speech outcomes in cochlear implant candidate patients with cochlear nerve deficiency.[14] On the other hand, a study by Birman et al.[15] proved otherwise, a considerable number of children (73.2%, 30/41 children) with cochlear nerve deficiency were able to use some spoken language. In their study, 89% and 47% of the children with hypoplastic and aplastic cochlear nerve had a fair CAP index (score of 5-7), respectively. This was similar to our study where more than 80% of the children with cochlear nerve hypoplasia could achieve high CAP-II scores. In addition, Wu et al.[16] also found that children with hypoplastic cochlear nerve were also able to achieve similar hearing and speech outcomes to children without cochlear nerve deficiency. This indicated that cochlear nerve deficiency was not an absolute contraindication of cochlear implantation, and that children with cochlear nerve hypoplasia could still benefit from cochlear implantation, which was proven in this study where CAP-II and SIR scores of the children improved following cochlear implantation.

The hearing and speech function of patients with cochlear implants may change over time. According to Lyu et al.,[17] the CAP index, which was used to evaluate hearing function, increased significantly between 6 to 24 months after implantation, and then the levels dropped off until 60 months post-surgery. This suggests that the first 2 years after cochlear implantation are critical for improving a cochlear implant user's auditory function. Meanwhile, the SIR score, which assesses speech ability, improved notably between 12 to 24 months after implantation and then plateaued, and reaching a stable value at 48 months post-implantation. This indicates that speech ability generally improves more slowly than hearing function, and that regular evaluations of both hearing and speech function are important, especially in the first 2 years after implantation. The results of these evaluations can also be used to guide speech therapy programs.[18,19]

Further analysis revealed that none of the factors examined in this study (i.e. type of cochlear nerve hypoplasia, prenatal TORCH infection, age at diagnosis, age at intervention, and age at implantation, duration of hearing aid use, duration of implant use, and frequency of auditory verbal therapy) were found to be significant predictors of post-operative CAP-II and SIR scores. This was in contrast with a previous study conducted in the same setting, which found that the frequency of auditory verbal therapy was a significant predictor of a high post-operative CAP-II index.[8] A possible explanation for the lack of significant predictors in our study was the small sample size, which might had restricted the analysis.

In this study, only 3 children had low post-operative CAP-II scores. Upon investigation, we found that these children did not use their implants consistently, using them for less than 6 hours a day due to discomfort, and also failing to attend follow-up mapping sessions. Similarly, of the 5 patients with low post-operative SIR scores, 3 children did not use their implants consistently, 1 child had used the implant for less than 1 year, and 1 child had stopped receiving auditory verbal therapy as their caregivers were unable to attend follow-up visits. The findings highlighted the crucial role of caregivers in supporting these children, who were not yet capable of performing these tasks independently. This finding was in line with a study by Silva et al.[6] which showed that cochlear implanted children who received adequate support from their families had a better quality of life and demonstrated better social and emotional independence.

In conclusion, our study showed that prelingual SNHL children with cochlear nerve hypoplasia might still benefit from cochlear implantation, as shown by a remarkable improvement in hearing and speech function as assessed with the CAP-II and SIR scores. CAP-II and SIR scores should be widely and regularly used to evaluate the hearing and speech function of children who had undergone cochlear implantation in Indonesia.

REFERENCE

1. McLaughlin MR. Speech and language delay in children. Am Fam Physician. 2011 May 15;83(10):1183–8.
2. Sunderajan T, Kanhere S V. Speech and language delay in children: Prevalence and risk factors. J Family Med Prim Care. 2019;8(5):1642.
3. Kim BG, Chung HJ, Park JJ, Park S, Kim SH, Choi JY. Correlation of cochlear nerve size and auditory performance after cochlear implantation in postlingually deaf patients. JAMA Otolaryngology–Head & Neck Surgery. 2013 Jun 1;139(6):604–9.
4. The Joint Committee on Infant Hearing. Year 2019 Position Statement: Principles and Guidelines for Early Hearing Detection and Intervention Programs. The Journal of Early Hearing Detection and Intervention. 2019;4(2):1–44.
5. Alam-Eldeen MH, Rashad UM, Ali AH. Radiological requirements for surgical planning in cochlear implant candidates. Indian J Radiol Imaging. 2017 Jul 1;27(3):274–81.
6. Silva J de M, Yamada MO, Guedes EG, Moret ALM. Factors influencing the quality of life of children with cochlear implants. Braz J Otorhinolaryngol. 2020 Jul 1;86(4):411–8.
7. Archbold S, Lutman ME, Nikolopoulos T. Categories of auditory performance: inter-user reliability. Br J Audiol. 1998;32(1):7–12.
8. Arief W, Zizlavsky S, Priyono H, Kurnia Wahyuni L, Endyarni Medise B, Prihartono J, et al. Gambaran persepsi auditori CAP-II pada anak tuli prelingual bilateral 6–12 bulan pasca implantasi koklea. Oto Rhino Laryngologica Indonesiana (ORLI). 2018 Jun 28;48(1):1–10.
9. Cox RM, McDaniel DM. Development of the Speech Intelligibility Rating (SIR) test for hearing aid comparisons. J Speech Hear Res. 1989;32(2):347–52.
10. Lou J, Gong WX, Wang G Bin. Cochlear nerve diameters on multipoint measurements and effects of aging in normal-hearing children using 3.0-T magnetic resonance imaging. Int J Pediatr Otorhinolaryngol. 2015 Jul 1;79(7):1077–80.
11. Bakhshaee M, Ghasemi MM, Shakeri MT, Razmara N, Tayarani H, Tale MR. Speech development in children after cochlear implantation. Eur Arch Otorhinolaryngol. 2007;264(11):1263–6.
12. Vincenti V, Ormitti F, Ventura E, Guida M, Piccinini A, Pasanisi E. Cochlear implantation in children with cochlear nerve deficiency. Int J Pediatr Otorhinolaryngol. 2014 Jun 1;78(6):912–7.
13. Kutz JW, Lee KH, Isaacson B, Booth TN, Sweeney MH, Roland PS. Cochlear implantation in children with cochlear nerve absence or deficiency. Otol Neurotol. 2011 Aug;32(6):956–61.
14. Wei X, Li Y, Chen B, Gong Y, Fu QJ, Liu T, et al. Predicting auditory outcomes from radiological imaging in cochlear implant

patients with cochlear nerve deficiency. Otol Neurotol. 2017 Jun 1;38(5):685–93.

15. Birman CS, Powell HRF, Gibson WPR, Elliott EJ. Cochlear implant outcomes in cochlea nerve aplasia and hypoplasia. Otol Neurotol. 2016 Jun 1;37(5):438–45.
16. Wu CM, Lee LA, Chen CK, Chan KC, Tsou YT, Ng SH. Impact of cochlear nerve deficiency determined using 3-dimensional magnetic resonance imaging on hearing outcome in children with cochlear implants. Otol Neurotol. 2015 Jan 20;36(1):14–21.
17. Lyu J, Kong Y, Xu TQ, Dong RJ, Qi BE, Wang S, et al. Long-term follow-up of auditory performance and speech perception and effects of age on cochlear implantation in children with pre-lingual deafness. Chin Med J (Engl). 2019 Aug 8;132(16):1925.
18. Degirmenci Uzun E, Batuk MO, D'Alessandro HD, Sennaroglu G. Auditory perception in pediatric cochlear implant users with cochlear nerve hypoplasia. Int J Pediatr Otorhinolaryngol. 2022 Sep 1;160:111248.
19. Phillips L, Hassanzadeh S, Kosaner J, Martin J, Deibl M, Anderson I. Comparing auditory perception and speech production outcomes: non-language specific assessment of auditory perception and speech production in children with cochlear implants. Cochlear Implants Int. 2009 Jun;10(2):92–102.