


# Cochlear implantation in children with white matter lesions

## Prediction of hearing outcomes by multiple regression analysis

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### Abstract

Brain magnetic resonance imaging (MRI) white matter lesions have been reported in some preoperative cochlear implant children. However, the role of white matter lesions in predicting the hearing outcome is yet unclear. The present study investigated the outcomes of cochlear implantation (CI) in 40 children with white matter lesions.

The data from children with white matter lesions were reviewed in this retrospective study. Based on brain MRI, the patients were divided into 3 groups: mild, moderate, and severe. The children were treated with unilateral CI and monitored for a follow-up period of at least 3 years. The main outcome measures were category of auditory performance (CAP) and speech intelligibility rating (SIR). MRI white matter lesions, age at implant, gender, physical impairment, and cognitive impairment were obtained from a research database to assess the correlation with long-term CAP and SIR outcome by multiple regression analysis.

The data of children with white matter lesions were reviewed (18 females and 23 males). The mean age at implantation was 31.6 months. Strikingly, all children obtained better CAP and SIR scores. The age at implantation, brain white matters lesions on MRI, and cognitive and physical disabilities were associated with CAP and SIR scores. Multiple regression established a weak correlation between the degree of white matter lesions on brain MRI and long-term CAP and SIR, while cognitive impairment strongly accounted for long-term CAP and SIR outcome.

The majority of the children with brain white matter lesions obtained a satisfactory postoperative effect. The cognitive impairment before CI is a major factor, and such factor should be considered.

**Abbreviations:** AVT = post-implantation rehabilitation therapy, ABR = auditory brainstem response, ANSD = auditory neuropathy spectrum disorder, ASSR = auditory steady state response, CAP = category of auditory performance, CI = cochlear implantation, CMV = congenital cytomegalovirus infection, CT = computed tomography, HIE = hypoxic-ischemic encephalopathy, H-NTLA = Hiskey-Nebraska Test of Learning Aptitude, IEMs = inner ear malformations, MRI = magnetic resonance imaging, NRT = neural response telemetry, OAEs = Otoacoustic emissions, SEM = standard error of the mean, SIR = speech intelligibility rating, SNHL = sensorineural hearing loss.

**Keywords:** CAP, cochlear implantation, cognitive hearing loss, MRI, SIR, white matter lesions

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## 1. Introduction

The brain central nervous system findings by preoperative brain MRI scans on cochlear implant candidates are common. Typically, white matter lesions are shown to be a possible determination of the abnormal neurodevelopmental outcome in some children.<sup>[1–3]</sup> According to the literature, most of the candidates for cochlear implantation (CI) with white matter lesions are not in agreement with the characteristics of hereditary leukoencephalopathy.<sup>[4]</sup> The white matter lesions are related to previous conditions/insults, including infection, ischemia, hypoxia, and prematurity.<sup>[5]</sup> Among these, congenital cytomegalovirus infection (CMV) can not only cause preterm birth and white matter abnormality but also non-genetic sensorineural deafness in children.

CI has been successful in children suffering from sensorineural hearing loss (SNHL).<sup>[6]</sup> The continual improvement in the technical development the diverse individual demands and the indications have been extended. The large-scale use of CI has made it possible to assess the functional outcomes as well as complications, and identify various prognostic factors.<sup>[2–6]</sup> MRI brain central nervous system findings have been reported in 20% to 40% patients in preoperative radiological work and the most common abnormality detected was white matter lesions that

determine the abnormal neurodevelopmental outcome in some children.<sup>[1–3]</sup> However, the role of white matter lesions in predicting hearing outcomes in cochlear implant children is yet unclear, and few studies have evaluated the outcomes following implantation in these children.

Overall, the functional outcome of CI is satisfactory and depends on the duration of deafness before CI, age at implantation, cause of deafness, co-morbidity including auditory neuropathy spectrum disorder (ANSD), and the presence of inner ear malformations (IEMs).<sup>[7–9]</sup> Only a few studies have investigated the outcomes following implantation in children with complications. Several studies have reported CI in patients with MRI brain abnormalities, but the majority focus on the incidence of abnormal brain MRI findings including white matter lesions,<sup>[5]</sup> especially, long-term results of category of auditory performance (CAP) test and the speech intelligibility rating (SIR) are sparse.

In summary, the presence of white matter lesions poses several questions about operation time decision, identification of prognostic factors, impact of co-morbidity, and outcome of cochlear implantation. The present study aimed to ascertain the importance of abnormal brain white matter MRI findings in predicting the outcome after CI and identify various prognostic factors in pediatric CI candidates.

## 2. Subjects and methods

### 2.1. Case inclusion criteria

The application for the cochlear implant was referred to the Department of Otolaryngology of Children's Hospital Affiliated to Zhengzhou University through Provincial Disabled Persons' Federation or foreign hospital, according to the following criteria:

1. Severe-to-profound SNHL was confirmed by audiological and audiological examinations conducted by professional ear institutes;
2. According to the time of onset, the child was identified as prelingual;
3. Inner ear malformation, dysplasia of the internal auditory canal, and ANSD were eliminated;
4. Unilateral cochlear implant
5. MRI showed mild and moderate abnormal white matter signals, and genetic leukoencephalopathy was excluded by genetic testing for severe abnormal white matter signals.

This study was approved by the Medical Ethics Committee of Children's Hospital Affiliated to Zhengzhou University.

All parents of children had signed the informed consent form before initiation of the study protocol.

### 2.2. Materials, data, and analysis

This study is a retrospective observational review of cochlear implant outcomes among severe-to-profound hearing-impaired children who present brain white matter lesions in MRI. To fulfill the demand of long-term follow-up (at least 3 years), the study cohort was identified among patients operated from January 2012 to December 2015 at the Department of Otolaryngology, Affiliated Children's Hospital of Zhengzhou University.

Otoacoustic emissions (OAEs), auditory brainstem response (ABR), auditory steady state response (ASSR), computed

tomography (CT), and MRI were performed preoperatively. Among patients with severe brain white matters lesions, 7 cases were excluded from hereditary leukoencephalopathy and 1 patient was diagnosed with Pelizaeus-Merzbacher disease by gene testing. Preoperative MRI was performed at least 2 times (more than 6 months apart), and the brain white matter disorder was found to be stable, shrunk, or disappeared before surgery (Fig. 1).

All children were implanted on the unilateral side. The same senior surgeon performed the cochlear implantation surgeries using soft surgical principal and round window insertion. The surgeries included all 3 cochlear implant companies available in the MED-EL, Cochlear, and Advanced Bionics. The type and make of cochlear implant contained MED-EL (SONATA, CONCERTO), Cochlear (CI422, CI522, CI512), and Advanced Bionics (HiFocus 1j, HiFocus Helix). And, the cochlear processor included MED-EL (Opus2, OpusXs, SONNET), Cochlear (CP802, CP900, N7), and Advanced Bionics (Harmony). The post-implantation rehabilitation therapy (AVT) of 40 children were in Henan Deafness Rehabilitation Center. All children had established consistent use of an Auditory-Verbal (AV), Oral Communication (OC), or Total Communication (TC) habilitative approach. The methods of rehabilitation training with expert guidance included family training and school training. The training time is at least 4 hours a day.

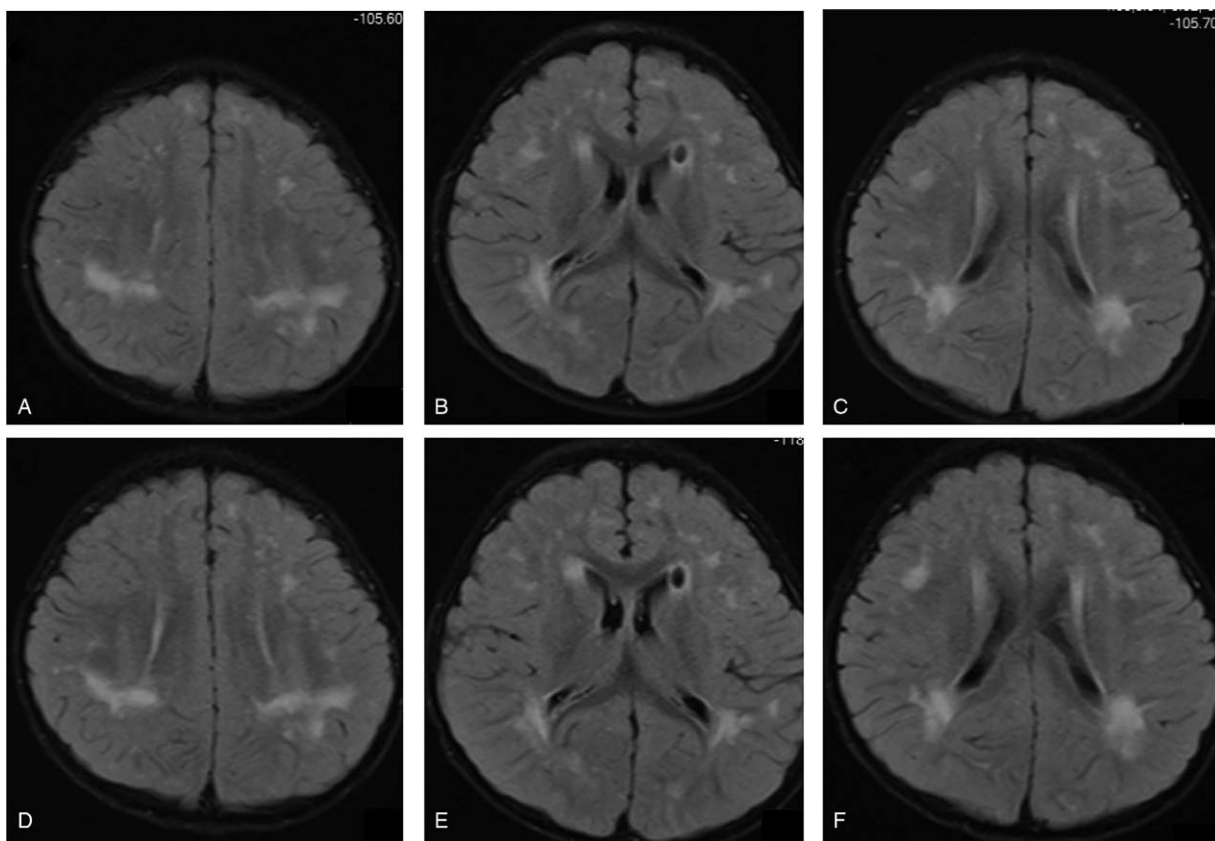
All children with brain white matter lesions were identified. The demographic data in terms of age at implantation, duration of follow-up, information about pre- or post-lingual deafness, situation of brain white matters lesions with MRI, and cognitive and physical disabilities were assessed by members of the cochlear implant team to facilitate analysis.

Cognitive ability was assessed in children by performance in Griffith Cognitive Development Scale and Hiskey-Nebraska Test of Learning Aptitude (H-NTLA).<sup>[10,11]</sup> Physical disabilities were assessed by performance in Gesell development scale.<sup>[12]</sup> Since the clinical characteristics of the nervous system damage were mild, the cognitive and physical disabilities were categorized into “none” and “present” based on assessment. The development quotient was evaluated by the scales and  $\leq 80$  was presented.

The brain white matters lesions were graded as “mild,” “moderate”, or “severe”, according to the scoring criteria.<sup>[13]</sup> Mild: involved the periventricular posterior horn, anterior horn, temporal horn, or the body respectively; Moderate: involved the above 2 to 3 joint lesions; Severe: more than 3 extensive sites (Fig. 2).

Numerous methods are available for the assessment of functional outcome after CI. Some of the most commonly applied tests are CAP and SIR.<sup>[14–16]</sup> The CAP consists of 8 performance categories arranged in the order of increasing difficulty with high inter-user agreement making it a reliable tool in measuring the auditory capacity after CI. The SIR test consists of an index 1–5 with improved language and is available for testing the children's speech intelligibility after CI. Some studies have shown that normal hearing children obtain maximum CAP and SIR scores within the first 3 years of life. The CAP and SIR tests were conducted by the speech therapist at 36 months postoperatively, according to the standard procedures.

SPSS17.0 was used for data analysis. The data that fit the normal distribution were expressed as mean  $\pm$  standard error of the mean (SEM). Correlations between each CAP and SIR index and the prognostic factors were examined by simple and multiple regression analyses. *P* values  $< .05$  were considered statistically significant.



**Figure 1.** The second brain MRI of the same patient with congenital SNHL combined with white matter lesions (fluid-attenuated inversion recovery). A-C: Brain MRI scan slices of one patient at 24 months of age; D-F: Brain MRI scan slices of the same patient at 19 months of age; Brain MRI of one patient retaken after 6 months later demonstrated that 3 white matter lesions were still or narrowed.

### 3. Results

Demographic data showed that 23/40 patients were male. In the majority of the children, brain white matter lesions were associated with prematurity, hypoxia, hypoxic-ischemic encephalopathy (HIE), maternal infection, intrauterine growth retardation, low birth weight, and meningitis. Other etiologies are unknown. The average age at implantation was 31.6 (range, 11–84) months with a similar mean age for both males and females (29.9 and 33.9 months, respectively). All the children were prelingual deaf (Table 1).

The white matter disorders in the brain MRI showed dots or flakes with sharp boundaries. The intensity of T1-weighted imaging, T2-weighted imaging, and fluid-attenuated inversion recovery decreased. In addition, the brain MRI showed that the bilateral periventricular white matter was involved, primarily in the posterior horn, followed by the anterior horn and temporal horn, and then in the body with lesions. The white matter lesions in all children who underwent cranial MRI were found to be still, narrowed, or even disappeared. Eight patients had severe brain white matter lesions, and 16 had moderate lesions. The remaining patients presented mild brain white matter disorder. In addition to these abnormalities, 11 children had physical impairment, and 4 showed cognitive impairment.

Normal preoperative NRT (neural response telemetry) values were obtained in all cases, including those with severe brain white matter lesions. Follow-up from implantation of the first CI ranged from 36 to 72 (median, 52.74) months.

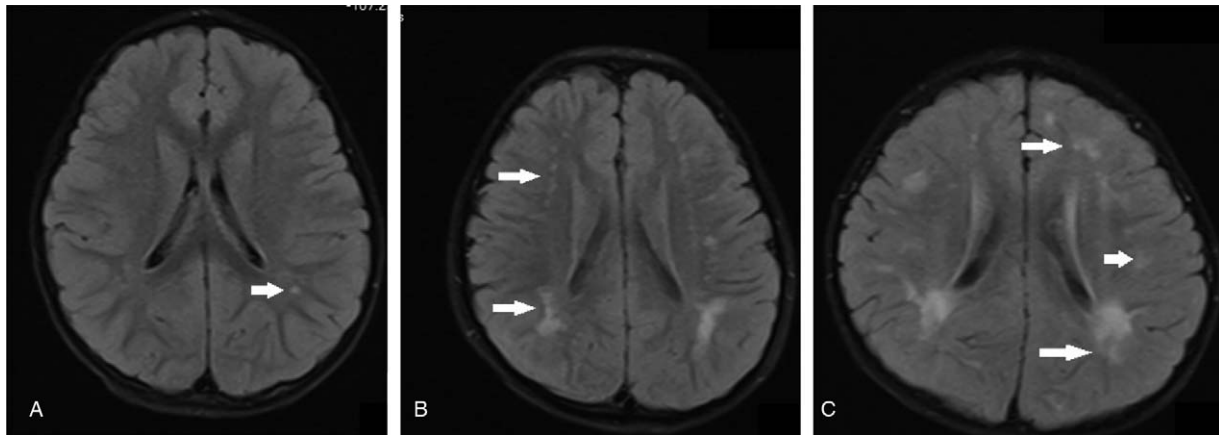
At 3 years after implantation, the median CAP score was 6.4 (range, 2–7), and the median SIR score was 3.5 (range, 1–5). Strikingly, all children obtained better CAP and SIR scores than before. The age at implantation, brain white matter lesions on MRI, and cognitive and physical disabilities were associated with the CAP scores. Similar results were found by correlation with the SIR scores (Tables 2 and 3). Multiple regression showed that the degree of white matter lesions on brain MRI was weakly correlated with long-term CAP and SIR, while cognitive impairment accounted for long-term CAP and SIR outcomes (Tables 4 and 5).

### 4. Discussion

Children with brain white-matter lesions detected by preoperative brain MRI scans on cochlear implant candidates are not rare (9.1%) and obtain a satisfactory postoperative effect in the long term. The most critical factor determining the outcome is cognitive impairment before CI.

Among 441 children with CI in our hospital from 2012 to 2015, 40 cases with abnormal white matter were identified by preoperative brain MRI, and the incidence was 9.1%. These findings were similar to those from related studies.

Jonas et al found that 22% had different degrees of white matter changes in a retrospective study consisting of 162 deaf children.<sup>[5]</sup> Another study showed that brain MRI before CI revealed white matter lesions in 57 children, accounting for 10% of the study cohort.<sup>[14]</sup> Among 426 children with cochlear



**Figure 2.** Brain MRI of children with congenital SNHL combined with white matter lesions (fluid-attenuated inversion recovery, white arrow). A: mild white matter lesions involved the periventricular posterior horn. B: moderate white matter lesions involved the above 2 to 3 joint lesions. C: severe white matter lesions involved more than 3 extensive sites.

implants studied by Busi et al, 74 cases showed abnormal brain changes before the operation. The most common abnormal brain changes were white matter lesions, accounting for 70% of the abnormal brain changes detected by MRI.<sup>[17]</sup> These studies suggested that the incidence of white matter abnormalities in children with CI is not low. Notably, hereditary leukoencephalopathy was excluded by gene testing in all cases with severe abnormal changes in the white matter. The clinical and imaging characteristics of all cases were significantly different from those of hereditary leukoencephalopathy caused by a gene mutation. Continuous T2W1 high signal was observed around the bilateral periventricular on MRI, and the intensity of T1-weighted imaging, T2-weighted imaging, and fluid-attenuated inversion recovery decreased. These findings suggested that this is a delayed area of normal development, and the abnormal white matter of the brain does not conform to the normal myelination process related to the age.

It has been found that acquired factors can influence the development of myelination in white matter. Frequently, white matter lesions were related to previous conditions/insults and included infection, ischemia, hypoxia, and prematurity.<sup>[4]</sup> Neonatal HIE is a major cause of white matter lesions. Because the white matter around the ventricle mainly supplies blood to the terminal branch of cerebral artery, the region is prone to be damaged first under the condition of ischemia-hypoxia, leading to the formation of corresponding nerve fiber myelin sheath or myelin sheath disorder.<sup>[18]</sup> HIE is also the most common cause of non-hereditary sensorineural deafness in children. In addition, cytomegalovirus infection not only causes premature delivery but also abnormalities in the white matter in the brain. Manara et al found 14 cases of congenital cytomegalovirus infection in children, 12 with sensorineural deafness, and 11 with different degrees of white matter abnormalities.<sup>[19]</sup> Haginoya et al reported 2 cases of congenital cytomegalovirus deafness, of which 1 had clinical and imaging features consistent with those of a previous study.<sup>[20]</sup> We found that the white matter lesions were caused by maternal infection during pregnancy, perinatal ischemia, hypoxia, intrauterine growth retardation, and premature birth. Etiological investigation showed that premature birth, ischemia, hypoxia, and viral infection were prominent, and could be speculated as the main factors causing white matter lesions.

In the present study, white matter lesions were shown to be a major outcome that might help to predict future problems, such as seizures and intellectual impairment in certain patients.<sup>[3]</sup> However, the role of white matter lesions in predicting hearing outcomes in cochlear implant patients is yet unclear.

Theoretically, the outcome of CI might be impeded in patients with brain white matter lesions due to putative disorganization or misplacement of the remaining neural structures and abnormality findings that are associated with poor neurodevelopmental outcomes;<sup>[21]</sup> such children might have decreased secondary CI outcomes. Actually, the children had obtained reasonable long-term results since the median CAP score was 6.4, and median SIR score was 3.5. The results were related to younger age at implantation and mild to moderate brain white matter disorders as observed by MRI. Also, no cognitive or physical disabilities were detected, and gender and prelingual deafness seemed insignificant. Hierarchical regression analysis showed that cognitive impairment accounted for long-term CAP and SIR outcome.

Edwards concluded that in children with cochlear implants and complex needs, cognitive impairment is a strong predictor of the outcome.<sup>[22]</sup> The current study also confirmed that cognitive function is the main predictor of outcome, which could be satisfactory, especially in those with no cognitive impairment even when brain white matter disorder was moderate or severe. However, in individuals with physical impairment, speech patterns may still be perceived. Hierarchical regression showed that only cognitive impairment accounted for long-term CAP and SIR outcome. Since cognitive impairment has been shown to adversely influence the outcomes of the cochlear implant, early identification of those with poor prognosis could lead to the early intervention by specific education programs to improve these outcomes.

Herein, we selected CAP and SIR for assessment of implantation outcomes, as these are used worldwide.<sup>[23]</sup> Nonetheless, the present results are limited by a ceiling effect, since the majority of the children present superior scores. To distinguish between children with high scores, other tests are recommended. These tests (e.g., behavioral tests) compare the results obtained elsewhere and are in a native language and designed specifically for the country in question.

**Table 1****Demographic data.**

Patient	Gender	Etiology	Age at first CI (m)	Follow-up (m)	Brain white matter lesions (MRI)	Physical impairment	Cognitive impairment	CAP*	SIR*
1	F	Unknown	14	42	Severe	Yes	No	7	3
2	M	Intrauterine Growth retardation	51	45	Severe	Yes	Yes	5	2
3	F	Hypoxia	30	36	Severe	Yes	No	6	5
4	F	Unknown	16	38	Severe	Yes	No	6	2
5	F	Hypoxia	24	71	Moderate	No	No	6	3
6	M	Unknown	36	68	Severe	Yes	Yes	2	1
7	M	HIE	30	39	Severe	Yes	No	7	3
8	F	Premature	30	45	Moderate	No	No	7	2
9	M	Unknown	18	47	Moderate	No	No	7	2
10	M	Maternal infection	24	70	Moderate	No	No	6	4
11	F	Intrauterine growth retardation	78	48	Moderate	No	No	7	4
12	M	Premature, Low birth weight	36	36	Moderate	No	No	7	3
13	F	Premature	51	38	Moderate	No	No	6	4
14	M	Premature	14	39	Moderate	No	No	7	4
15	M	Unknown	52	51	Moderate	No	No	7	5
16	M	Premature	32	63	Moderate	No	No	8	4
17	F	Unknown	16	66	Mild	No	No	7	4
18	M	Unknown	24	68	Mild	No	No	8	4
19	M	HIE	36	72	Mild	No	No	6	3
20	M	Low birth weight	32	55	Mild	No	No	7	4
21	F	Unknown	48	48	Mild	No	No	7	4
22	M	Maternal infection	30	65	Mild	No	No	7	4
23	M	Unknown	36	39	Mild	No	No	7	4
24	F	Hypoxia	52	64	Mild	No	No	5	4
25	F	Unknown	24	42	Mild	No	No	7	4
26	M	Premature	36	63	Mild	No	No	7	5
27	M	Unknown	26	64	Mild	No	No	7	3
28	M	Maternal infection	16	52	Mild	No	No	7	4
29	F	Maternal infection	11	55	Mild	No	No	7	4
30	F	Unknown	26	48	Mild	No	No	7	4
31	M	Hypoxia	51	38	Severe	Yes	No	5	4
32	M	Unknown	30	37	Severe	Yes	No	4	3
33	M	Hypoxia	16	59	Moderate	Yes	No	7	3
34	F	Intrauterine growth retardation (twisted twist of the umbilical cord)	24	67	Moderate	Yes	Yes	3	1
35	F	HIE	36	61	Moderate	No	Yes	7	4
36	M	Unknown	30	54	Mild	No	No	6	3
37	M	Unknown	30	62	Moderate	No	No	6	4
38	F	Meningitis	48	42	Mild	Yes	No	5	4
39	F	Premature	35	44	Mild	No	No	7	5
40	M	Low birth weight	84	45	Moderate	No	No	7	4

\* 36 months postoperative.

Children with additional disabilities and brain white matter lesions on MRI pose many challenges for cochlear implant teams. There may also be a delay in referral for the assessment of CI due to the misconception that a child with this combination of disabilities

may not benefit from a cochlear implant. It is well known that the earlier a patient receives intervention for their hearing impairment, the better the outcome.<sup>[24]</sup> The current study shows a trend towards a better outcome if CI is carried out at an earlier age.

**Table 2****Correlation coefficients between CAP/SIR and prognostic factors.**

	CAP	P value
Simple correlation coefficient		
Age	-0.089	.575
Gender	-0.051	.749
MRI	-0.406*	.008
Physical impairment	0.599*	.000
Cognitive impairment	0.575*	.000

\*  $P < .1$ .**Table 3****Correlation coefficients between CAP/SIR and prognostic factors.**

	SIR	P value
Simple correlation coefficient		
Age	0.245*	.117
Gender	-0.021	.897
MRI	-0.392*	.010
Physical impairment	0.418*	.006
Cognitive impairment	0.492*	.001

\*  $P < .1$ .

**Table 4**  
Multiple regression model of CAP related to 6 prognostic factors.

Prognostic factor	Regression coefficient	Standardized regression coefficient*	Partial correlation coefficient (P value)
Age ( $x_1$ )	-0.010	-0.143	.229
MRI( $x_3$ )	0.167	0.102	.527
Physical impairment ( $x_5$ )	1.489	0.538	.002
Cognitive impairment ( $x_6$ )	1.687	0.407	.002

\* Standardized regression coefficient is a regression coefficient on the condition that all explanatory and dependent variables are converted into those with a mean of 0 and a standard deviation of 1.

**Table 5**  
Multiple regression model of SIR in relation to 6 prognostic factors.

Prognostic factor	Regression coefficient	Standardized regression coefficient*	Partial correlation coefficient (P value)
Age ( $x_1$ )	0.016	0.271	.048
MRI ( $x_3$ )	-0.317	-0.233	.206
Physical impairment ( $x_5$ )	0.191	0.084	.655
Cognitive impairment ( $x_6$ )	1.368	0.399	.007

\* Standardized regression coefficient is a regression coefficient on the condition that all explanatory and dependent variables are converted into those with a mean of 0 and a standard deviation of 1.

Assessing cognitive ability in these children poses some challenges. In this study, cognitive ability was assessed using nursery/school performance. This might be difficult in children in early age but highlights the difficulty of assessment and predicting the outcome as implantation is undertaken at an increasingly early age. Thus, standardized and unified assessment training and new methods of assessment will be required.

Another challenge is a contraindication of MRI after CI. Various studies have shown that MRI scans can be safely performed with CI. This does not imply that it is generally safe to perform MRI in CI patients because of the type of implant, and MRI units and sequences may vary. Even if it can be performed safely, the distortion caused by the implanted magnet is ascribed to sub-optimal interpretation.<sup>[25]</sup> MRI is essential in the diagnostic work-up for a number of neurological conditions, especially for a patient with brain white matter disorder. Typically, if an MRI scan is required in a patient with cochlear implant, the magnet has to be removed from the implanted package prior to scanning and then replaced. In a pediatric patient, the magnet removal and replacement usually require a general anesthetic making this a critical undertaking.

Assessing outcomes in children following CI allows parents to decide on the subsequent operation. Children with severe white matter abnormalities are observed for more than 6 months, and then surgery is performed if the white matter lesions are stationary or improved. However, implantation may not be beneficial at an earlier age. In children with brain white matter disorders, assessment before implantation presents some difficulties. Herein, we suggested that success is often judged using clinical characteristics (including brain MRI and etiology), and gene testing excludes hereditary leukoencephalopathy. Also, we

excluded 1 child suffering from Pelizaeus-Merzbacher disease in the study.

The current results are in accordance with the test systems about auditory and speech performance in cochlear-implanted patients with brain white matter lesions and follow-up time described in the international literature. Thus, we will advocate using international standardized tests to obtain valid and reliable tools for detailed and accurate results. Furthermore, long-term results are required in all CI patients to comply with the possibility of deterioration of the outcome due to progressive degeneration of brain white matter disorders.

## 5. Conclusions

Children with brain white-matter lesions detected by preoperative brain MRI scans on cochlear implant candidates are not rare (9.1%) and obtain a satisfactory postoperative effect in the long term. The most critical factor determining the outcome is cognitive impairment before CI. However, additional studies are required to ascertain the cognitive impairment and help guide candidacy; each case must be considered individually by an experienced cochlear implant team.

## Author contributions

**Conceptualization:** Sufang Wang, Fugen Han.

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**Visualization:** Yanyan Wei, Hongbo Ren.

**Writing – original draft:** Yanhong Cui.

**Writing – review & editing:** Fugen Han.

## References

- Trimble K, Blaser S, James AL, et al. Computed tomography and/or magnetic resonance imaging before pediatric cochlear implantation? Developing an investigative strategy. *Otol Neurotol* 2007;28:317–24.
- Lee DJ, Lustig L, Sampson M, et al. Effects of cytomegalovirus (CMV) related deafness on pediatric cochlear implant outcomes. *Otolaryngol Head Neck Surg* 2005;133:900–5.
- Perlman JM. White matter injury in the preterm infant: an important determination of abnormal neurodevelopment outcome. *Early Hum Dev* 1998;53:99–120.
- Lyon G, Fattal-Valevski A, Kolodny EH. Leukodystrophies: clinical and genetic aspects. *Top Magn Reson Imaging* 2006;17:219–42.
- Jonas NE, Ahmed J, Grainger J, et al. MRI brain abnormalities in cochlear implant candidates: how common and how important are they? *Int J Pediatr Otorhinolaryngol* 2012;76:927–9.
- O'Donoghue GM. Cochlear implants in children: principles, practice and predictions. *J R Soc Med* 1996;89:345–7.
- Papsin BC. Cochlear implantation in children with anomalous cochleovestibular anatomy. *Laryngoscope* 2005;115(1 Pt 2 Suppl 106):1–26.
- Walton J, Gibson WPR, Sanli H, et al. Predicting cochlear implant outcomes in children with auditory neuropathy. *Otol Neurotol* 2008;29:302–9.
- Black J, Hickson L, Black B, et al. Prognostic indicators in paediatric cochlear implant surgery: a systematic literature review. *Cochlear Implants Int* 2011;12:67–93.
- Griffiths R. *The Abilities of Young Children. A Study in Mental Measurement.* London: University of London Press; 1970.
- Hiskey MS. *Manual H-N TLA.* Nebraska. USA: Lincoln; 1968. 1–22.

- [12] Hou S, Yuan L, Jin P, et al. A clinical study of the effects of lead poisoning on the intelligence and neurobehavioral abilities of children. *Theor Biol Med Model* 2013;10:13–13.
- [13] Fedrizzi E, Inverno M, Bruzzone MG, et al. MRI features of cerebral lesions and cognitive functions in preterm spastic diplegic children. *Pediatr Neurol* 1996;15:207–12.
- [14] Archbold S, Lutman ME, Nikolopoulos T. Categories of auditory performance: inter-user reliability. *Br J Audiol* 1998;32:7–12.
- [15] Allen C, Nikolopoulos TP, Dyar D, et al. Reliability of a rating scale for measuring speech intelligibility after pediatric cochlear implantation. *Otol Neurotol* 2001;22:631–3.
- [16] Percy-Smith L. Associations between auditory capacity, speech and language, level of communication and parental assessment of children with cochlear implant. *Cochlear Implants Int* 2010;11:50–62.
- [17] Busi M, Rosignoli M, Castiglione A, et al. Cochlear implant outcomes and genetic mutations in children with ear and brain anomalies. *Biomed Res Int* 2015;2015:696281–1696281.
- [18] Deguchi K, Oguchi K, Matsuura N, et al. Periventricular leukomalacia: relation to gestational age and axonal injury. *Pediatr Neurol* 1999;20:370–4.
- [19] Manara R, Balao L, Baracchini C, et al. Brain magnetic resonance findings in symptomatic congenital cytomegalovirus infection. *Pediatr Radiol* 2011;41:962–70.
- [20] Haginoya K, Ohura T, Kon K, et al. Abnormal white matter lesions with sensorineural hearing loss caused by congenital cytomegalovirus infection: retrospective diagnosis by PCR using Guthrie cards. *Brain Dev* 2002;24:710–4.
- [21] Lapointe A, Viamonte C, Morriss MC, et al. Central nervous system findings by magnetic resonance in children with profound sensorineural hearing loss. *Int J Pediatr Otorhinolaryngol* 2006;70:863–8.
- [22] Edwards LC. Children with cochlear implants and complex needs: a review of outcome research and psychological practice. *J Deaf Stud Deaf Educ* 2007;12:258–68.
- [23] Nikolopoulos TP, Archbold SM, Gregory S. Young deaf children with hearing aids or cochlear implants: early assessment package for monitoring progress. *Int J Pediatr Otorhinolaryngol* 2005;69:175–86.
- [24] Nikolopoulos TP, O'Donoghue GM, Archbold S. Age at implantation: its importance in pediatric cochlear implantation. *Laryngoscope* 1999;109:595–9.
- [25] Vincent C, Ruzza I, Vaneeckloo FM, et al. Magnetic resonance imaging with the Digisonic SP Neurelec cochlear implant. *Eur Arch Otorhinolaryngol* 2008;265:1043–6.