European Annals of Otorhinolaryngology, Head and Neck diseases 132 (2015) 317–320

Available online at ScienceDirect www.sciencedirect.com

Elsevier Masson France EM|consulte www.em-consulte.com/en

Original article

# Speech perception, production and intelligibility in French-speaking children with profound hearing loss and early cochlear implantation after congenital cytomegalovirus infection

L. Laccourreye*, V. Ettienne, I. Prang, V. Couloigner, E.-N. Garabedian, N. Loundon

*Service d'oto-rhino-laryngologie pédiatrique, hôpital Necker-Enfants malades, université Paris-Descartes, Sorbonne Paris-Cité, Assistance publique–Hôpitaux de Paris, 149, rue de Sèvres, 75015 Paris, France*

ARTICLE INFO

*Keywords:*
Child
Hearing loss
Cytomegalovirus
Cochlear implant
Speech

ABSTRACT

*Objective:* To analyze speech in children with profound hearing loss following congenital cytomegalovirus (cCMV) infection with cochlear implantation (CI) before the age of 3 years.
*Material and methods:* In a cohort of 15 children with profound hearing loss, speech perception, production and intelligibility were assessed before and 3 years after CI; variables impacting results were explored.
*Results:* Post-CI, median word recognition was 74% on closed-list and 48% on open-list testing; 80% of children acquired speech production; and 60% were intelligible for all listeners or listeners attentive to lip-reading and/or aware of the child's hearing loss. Univariate analysis identified 3 variables (mean post-CI hearing threshold, bilateral vestibular areflexia, and brain abnormality on MRI) with significant negative impact on the development of speech perception, production and intelligibility.
*Conclusion:* CI showed positive impact on hearing and speech in children with post-cCMV profound hearing loss. Our study demonstrated the key role of maximizing post-CI hearing gain. A few children had insufficient progress, especially in case of bilateral vestibular areflexia and/or brain abnormality on MRI. This led us to suggest that balance rehabilitation and speech therapy should be intensified in such cases.



## 1. Introduction

A number of studies focusing on speech perception, production and intelligibility following cochlear implantation (CI) in children with profound hearing loss following congenital cytomegalovirus (cCMV) infection have been published in the scientific medical literature over the last 15 years [1–10]. Analyzing them (Table 1) found a great heterogeneity in numbers included, age at CI and follow-up: 4 had more than 3 years' follow-up [3,6,8,9], 2 were based on cohorts of at least 15 children [2,5], and 1 focused on children with CI earlier than the theoretical age of speech acquisition (3 years) [6], but none associated all 3 criteria. Likewise, some included both profound and severe hearing loss and/or were based on parent surveys [1–10]. None, finally, systematically analyzed speech perception, production and intelligibility, and some set the significance threshold at $P < 0.1$ rather than $< 0.05$ [1–10].

Given these various limitations in the reported studies and the lack of any data for cohorts of children brought up in a French-speaking environment, we performed a retrospective study of a cohort of 15 children with cCMV profound hearing loss, assessing speech perception, production and intelligibility 3 years post-CI, exploring variables impacting results and analyzing results in the light of the scientific medical literature.

## 2. Material and methods

Between 2001 and 2010, 51 cCMV children with profound hearing loss received CI in the ENT departments of the Trousseau and Necker hospitals (Paris, France). In this cohort, 18 received CI before the age of 3 years. The 15 children with a minimum of 3 years' follow-up after CI were included in the present retrospective analysis.

cCMV infection was diagnosed on the basis of maternal CMV seroconversion during pregnancy, associated with neonate viruria and/or positive PCR on Guthrie card. Table 2 documents clinical (gender, mean pre-CI hearing loss (dB) according to the

* Corresponding author.
*E-mail address:* laurence.laccourreye@nck.aphp.fr (L. Laccourreye).

http://dx.doi.org/10.1016/j.anorl.2015.08.020
1879-7296/

**Table 1**
Published series analyzing the impact of cochlear implantation on speech perception, production and/or intelligibility in cCMV children with profound hearing loss.

| Authors | N | Age | Follow-up | Results |
|---|---|---|---|---|
| Pyman et al.[a] [1] | 7 | < 5 yrs | 5 mo–12 yrs | Results poorer than non-cCMV |
| Ramirez Inscoe and Nikolopoulos[a] [2] | 16 | M: 3.9 yrs | 1–5 yrs | Results identical to non-cCMV |
| Lee et al. [3] | 13 | 2–9.2 yrs | 4 yrs | Good results |
| Yoshida et al.[a] [4] | 4 | 2–3.4 yrs | 2.6–3.7 yrs | Results identical to non-cCMV after 2nd month post-CI |
| Ciorba et al.[a] [5] | 16 | 2–7 yrs | 6 mo–18 yrs | Results poorer than non-cCMV |
| Iwasaki et al.[a] [6] | 2 | 1.9–2.9 yrs | 36 mo | Results identical to non-cCMV |
| Malik et al.[a] [7] | 14 | 2–8.3 yrs | 1 –11.5 yrs | Results poorer than non-cCMV |
| Viccaro et al. [8] | 6 | 2–10 yrs | > 10 yrs | Poor results |
| Matsui et al.[a] [9] | 5 | 2.1–5.8 yrs | > 4 yrs | Results identical to non-cCMV if isolated hearing loss |
| Birman et al.[a] [10] | 5 | 1–16 yrs | 12 mo | Results poorer than non-cCMV, especially in non-isolated hearing loss |

M: median.
[a] Case control study.

International Bureau for Audiophonology scale [11], progressive hearing loss, bilateral vestibular areflexia, ophthalmic abnormality, abnormal cerebral MRI) and treatment data (age at CI, mean hearing loss 12 months post-CI, CI model, contralateral aid or not, type of communication, type of schooling).

Language assessment was performed for each child ahead of and during the 36th month after CI by a specialized speech therapist, at our clinic. Perception of the therapist's speech was assessed on closed- and open-list word recognition scores on a 0-to-10 scale. Speech production was assessed on a 1-to-6 scale (meaningless vocalization, words, incomplete sentences, correct simple sentences, complex sentences, structured ordinary language). The intelligibility of the child's speech production was assessed on the 1-to-5 Nottingham Speech Intelligibility Rating [12] (unintelligible, a few words intelligible in context, intelligible for listeners attentive to lip-reading, intelligible for listeners aware of hearing loss, intelligible to all). Data were entered in a PC. Statistical analysis, on StatView software (SAS Institute), comprised two parts. Firstly, development of perception, production and intelligibility were analyzed by comparing scores before and 36 months after CI. Secondly, associations were sought between these developmental data and the clinical and therapeutic variables documented in Table 2. Analysis used Mann-Whitney non-parametric U test and simple regression ANOVA. The significance threshold was set at 0.05.

## 3. Results

### 3.1. Speech perception

None of the 15 cCMV children had perception of the therapist's speech before CI (Table 3). Post-CI, median word recognition on closed-list was 74% and 48% on open-list (Table 3). On closed-list testing, 13% (2/15) of children failed to develop speech perception, and 53% (8/15) on open-list testing. Median post-CI word recognition in children developing speech perception was 100% (range, 30–100%) on closed-list and 90% (range, 50–100%) on open-list testing.

On univariate analysis (Table 4), 2 variables significantly impacted development of speech perception. The lower the mean hearing loss 12 months post-CI, the better the closed-list development (Fig. 1; $P = 0.007$). Median closed-list recognition varied from 60% with associated bilateral vestibular areflexia, to 100% without ($P = 0.04$).

**Table 2**
Clinical and therapeutic data.

| Variables | Population |
|---|---|
| Gender (female/male) | 12/3 |
| Age (months) at implantation: (range) median | (14–36) 24 |
| Mean hearing loss (dB) pre-implantation: (range) median | (95–120) 119 |
| Progressive hearing loss (yes/no) | 5/10 |
| Vestibular areflexia (no/unilateral/bilateral) | 3/4/8 |
| Ophthalmic involvement (yes/no) | 3/12 |
| Brain abnormality on MRI (yes/no) | 8/7 |
| Schooling (normal/specialized) | 8/7 |
| Type of communication (oral, bilingual) | 5/10 |
| Implantation (unilateral, bilateral) | 14/1 |
| Implant model (Cochlear/Advanced Bionics) | 14/1 |
| Mean hearing loss (dB) 12 months post-implantation: (range) median | (30–70) 35 |
| Conventional contralateral aid (yes/no)[a] | 7/7 |

MRI: magnetic nuclear resonance imaging.
[a] 1 child with bilateral cochlear implants.

**Table 3**
Test scores.

| Variables: (range) median | Pre-CI | 36 months post-CI |
|---|---|---|
| *Speech perception* | | |
| Word recognition (closed-list) | (0) 0 | (0–100) 74 |
| Word recognition (open-list) | (0) 0 | (0–100) 48 |
| *Speech production* | (1) 1 | (1–6) 3 |
| *Speech intelligibility* | (1) 1 | (1–5) 3 |

CI: cochlear implantation.

### 3.2. Speech production

Before CI, children produced only meaningless vocalizations (Table 3). After CI, 20% (3/15) remained at that level (level 1). Forty-two percent (5/12) of the children who acquired speech production produced words or incomplete sentences (levels 2 and 3), 50% (6/12) simple or complex sentences (levels 4 and 5) and 8% (1/12) structured ordinary language (level 6).

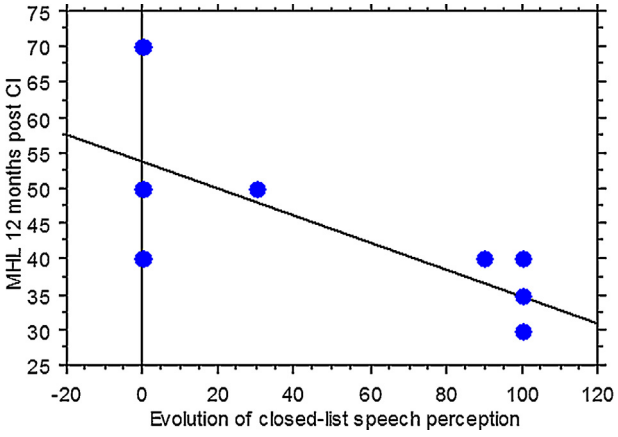


**Fig. 1.** Linear regression (ANOVA): impact of mean hearing loss (MHL) after cochlear implantation (post-CI) on speech perception development.

**Table 4**
Statistical analysis (*P* values) of impact of various clinical and therapeutic variables on development of speech perception, production and intelligibility.

| Variables | Perception | | Production | Intelligibility |
|---|---|---|---|---|
| | Open-list | Closed-list | | |
| *Age (months) at implantation* & | 0.9 | 0.7 | 0.9 | 0.6 |
| *Mean hearing loss (dB)*& | | | | |
| Pre-implantation | 0.7 | 0.1 | 0.2 | 0.2 |
| 12 months post-implantation | 0.007 | 0.08 | 0.01 | 0.007 |
| *Progressive hearing loss (yes/no)*§ | 0.1 | 0.1 | 0.07 | 0.8 |
| *Bilateral vestibular areflexia (yes/no)*§ | 0.04 | 0.08 | 0.09 | 0.03 |
| *Ophthalmic involvement (yes/no)*§ | 0.2 | 0.3 | 0.4 | 0.1 |
| *MRI brain abnormality (yes/no)*§ | 0.2 | 0.6 | 0.08 | 0.04 |
| *Schooling (normal/specialized)*§ | 0.09 | 0.08 | 0.03 | 0.08 |
| *Type of communication (O, bi)*§ | 0.6 | 0.7 | 0.5 | 0.1 |
| *Contralateral aid*[a] *(yes/no)*§ | 0.6 | 0.9 | 0.9 | 0.9 |

O: oral; bi: bilingual; tests: §: Mann-Whitney U; &: ANOVA.
[a] Analysis on 14 children, 1 child with bilateral cochlear implant.

On univariate analysis (Table 4), 2 variables significantly impacted development of speech production. The lower the mean hearing loss 12 months post-CI, the better was the development of speech production (Fig. 2; $P = 0.01$). Development of speech production was better in children receiving normal schooling ($P = 0.03$).

### 3.3. Speech intelligibility

Before CI, children's speech production was unintelligible (level 1; Table 3). After CI, 40% (6/15) of children remained at level 1 or produced only a few words intelligible in context (level 2). Sixty-seven percent (6/9) of the children who acquired speech production were intelligible for listeners attentive to lip-reading or aware of hearing loss (levels 3 and 4) and 33% (3/9) for all listeners (level 5).

On univariate analysis (Table 4), 3 variables significantly impacted development of intelligibility. The lower the mean hearing loss 12 months post-CI, the better the development of intelligibility (Fig. 3; $P = 0.007$). Development of intelligibility was better when there was no bilateral vestibular areflexia ($P = 0.03$) or brain abnormality on MRI ($P = 0.04$).

## 4. Discussion

Worldwide, CMV infection (cCMV) affects 0.5% to 2.5% of neonates, making it the most frequent congenital viral infection [13]. Ninety percent of cCMV infections are asymptomatic [14]. Sensorineural hearing loss is the most common clinical manifestation, noted in 60% of symptomatic cases [14]. In France, 720–2400 of the 1200–4000 cCMV neonates born yearly show sensorineural hearing loss [15]. CMV is thus, after genetic disease, the second most frequent etiology of congenital childhood hearing loss [15].

Cochlear implantation offers a therapeutic solution adapted to the difficulties children encounter when hearing loss is profound [16]. Various studies [2,4,8,9] highlighted CI as the optimal means of restoring hearing in cCMV children with profound hearing loss. Ramirez Inscoe et al. [2] reported that mean tonal hearing loss in cCMV children with profound hearing loss (> 90 dB) ranged from 60 dB with conventional hearing aids to 26–45 dB with CI. With median hearing loss of 119 dB before and 35 dB 12 months after CI (Table 2), the present cohort confirmed that CI restores hearing in cCMV children with profound hearing loss.

In the light of these good auditory results, several teams [1–10] assessed the contribution of CI to speech perception, production and intelligibility in cCMV children with profound hearing loss. Analysis of these studies highlights the heterogeneity of their results (Table 1) and the difficulty of assessing the impact of disorders associated with sensorineural hearing loss. These studies [1–10], which document the results achieved in cohorts of children brought up in English, Japanese or Italian, are fraught with methodological limitations described in the introduction. The selection of 15 cCMV children with profound hearing loss who received CI before the age of 3 years and who were followed up for a minimum 3 years post-CI, allowed us to avoid these methodological limitations. Our findings can be analyzed along 3 main axes: the benefit of CI for speech perception, production and intelligibility, the limitations of CI in terms of speech perception, production and intelligibility, and the detection of variables that impact the

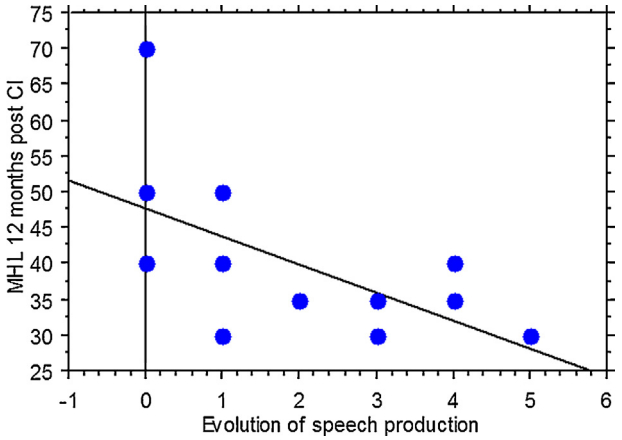


**Fig. 2.** Linear regression (ANOVA): impact of mean hearing loss (MHL) after cochlear implantation (post-CI) on speech production development.

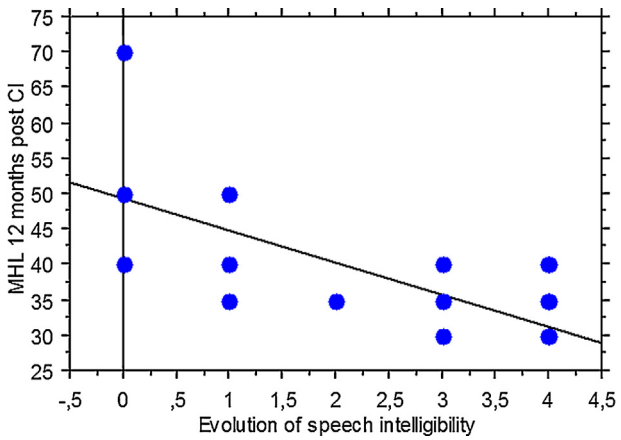


**Fig. 3.** Linear regression (ANOVA): impact of mean hearing loss (MHL) after cochlear implantation (post-CI) on speech intelligibility development.

development of speech perception, production and intelligibility (difference between scores pre-CI and 36 months post-CI).

Regarding the benefit of CI, our study found significant improvement in speech perception, production and intelligibility in cCMV children with profound hearing loss (Table 3). Three years post-CI, the median number of words pronounced by the speech therapist and recognized by the children was 74% on closed- and 48% on open-list testing, confirming the improvement in speech perception reported by Lee et al. [3]. Likewise, CI improved speech production: before CI children produced only meaningless vocalizations (Table 3), but after CI 80% showed progress. This progress, however, was highly variable: 42% produced words or incomplete sentences, 50% simple or complex sentences, and 8% structured normal language. This is in agreement with Matsui et al.'s report [9] which noted that, 3 years post-CI, 4 of the 5 cCMV children in their Japanese cohort progressed in speech production. Finally, in terms of intelligibility, in our series, speech was unintelligible before CI (Table 3); post-CI, 60% of the children had some progression, with 33% having speech intelligible to all listeners. This is in agreement with the report by Ciorba et al. [5], who, in a cohort of 16 Italian children with profound hearing loss, underscored the improvement of speech intelligibility after CI.

However, the apparent efficacy of CI in terms of speech perception, production and intelligibility in cCMV children with profound hearing loss is to be taken with caution. In the current study, 53% of the children did not develop speech perception on open-list testing, which provides more thorough analysis than closed-list testing. Likewise, 3 years post-CI, 20% of the children were still producing meaningless vocalizations and in 20% speech remained unintelligible. We are not the first team to report such difficulties in speech development after CI in cCMV children with profound hearing loss [1–3,5,7–10]. Lee et al. [3] underscored the slow development of speech perception in cCMV children, with a positive evolution on open-list testing 4 years post-CI. Several reports also pointed to a negative impact of associated motor or cognitive disorder [1,2,5,7,9]. Cognitive levels were not assessed in the present cohort. Although we therefore cannot assess the role played by this variable, we consider it to be of major interest, as in univariate analysis cerebral abnormalities on MRI were found to significantly impair the development of speech intelligibility (Table 4). Likewise, bilateral vestibular areflexia was significantly correlated with the development of speech perception and intelligibility (Table 4). We take this, like brain abnormality on MRI, as indicating more severe impact of cCMV. In such cases, we consider the intensification of psychomotor rehabilitation and physiotherapy to be of utmost importance. Finally, the smaller the post-CI mean hearing loss, the better the development of speech perception, production and intelligibility (Table 4; Figs. 1–3). This should encourage us to seek optimal CI setting in order to achieve optimal hearing gain, and to consider early contralateral CI. It is also to be stressed that neither mean pre-CI hearing loss, a progressive profile of hearing loss, nor contralateral hearing aid had an impact on the development of speech perception, production and intelligibility (Table 4). However, speech production developed significantly better (Table 4) if the child was in normal schooling. The value of this finding calls for discussion, as children may have been schooled in normal classes because their level of speech production was already deemed satisfactory by the speech therapist.

## 5. Conclusion

The present study confirms the positive impact of CI before the age of 3 years on hearing and speech in cCMV children with profound hearing loss and the importance of maximizing post-CI hearing gain. It also highlighted the fact that certain children do not progress sufficiently. Limiting factors identified were bilateral vestibular areflexia and brain abnormalities on MRI: such findings doubtless correspond to more severe cCMV impact. They should prompt us, on the one hand, to prepare the parents for difficulties in post-CI speech acquisition, so that their expectations may be realistic, and, on the other hand, to reinforce balance rehabilitation and speech therapy.

## Disclosure of interest

The authors declare that they have no conflicts of interest concerning this article.

## Acknowledgments

The authors thank the Progrès 2000 association for technical support in conducting the study.

## References

[1] Pyman B, Blamey P, Lacy P, et al. The development of speech perception in children using cochlear implants: effects of etiologic factors and delayed milestones. Am J Otol 2000;21:57–61.
[2] Ramirez Inscoe JM, Nikolopoulos TP. Cochlear implantation in children deafened by cytomegalovirus: speech perception and speech intelligibility outcomes. Otol Neurotol 2004;25:479–82.
[3] Lee DJ, Lustig L, Sampson M, et al. Effects of cytomegalovirus related deafness on pediatric cochlear implant outcomes. Otolaryngol Head Neck Surg 2005;133:900–5.
[4] Yoshida H, Kanda Y, Takahashi H, et al. Cochlear implantation in children with congenital cytomegalovirus infection. Otol Neurotol 2009;30:725–30.
[5] Ciorba A, Bovo R, Trevisi P, et al. Rehabilitation and outcome of severe profound deafness in a group of 16 infants affected by congenital cytomegalovirus infection. Eur Arch Otorhinolaryngol 2009;266:1539–46.
[6] Iwasaki S, Nakanishi H, Misawa K, Tanigawa T, Mizuta K. Cochlear implant in children with asymptomatic congenital cytomegalovirus infection. Audiol Neurootol 2009;14:146–52.
[7] Malik V, Bruce IA, Broomfield SJ, et al. Outcome of cochlear implantation in asymptomatic congenital cytomegalovirus deafened children. Laryngoscope 2011;121:1780–4.
[8] Viccaro M, Filipo R, Bosco E, et al. Long term follow up of implanted children with cytomegalovirus-related deafness. Audiol Neurootol 2012;17:395–9.
[9] Matsui T, Ogawa H, Yamada N, et al. Outcome of cochlear implantation in children with congenital cytomegalovirus infection or GJB2 mutation. Acta Otolaryngol 2012;132:597–602.
[10] Birman CS, Elliott EJ, Gibson WP. Pediatric cochlear implants: additional disabilities prevalence, risk factors, and effect on language outcomes. Otol Neurotol 2012;33:1347–52.
[11] http://www.biap.org/recom02-1.htm
[12] Allen C, Nikolopoulos TP, Dyar D, O'Donoghue G. Reliability of a rating scale for measuring speech intelligibility after pediatric cochlear implantation. Otol Neurotol 2001;22:631–3.
[13] Hagay ZJ, Biran G, Ornoy A, Reece EA:. Congenital cytomegalovirus infection: a long-standing problem still seeking a solution. Am J Obstet Gynecol 1996;174:241–5.
[14] Kenneson A, Cannon MJ. Review and meta-analysis of the epidemiology of congenital cytomegalovirus infection. Rev Med Virol 2007;17:253–76.
[15] Avettand-Fenoël V, Marlin S, Vauloup-Fellous C, et al. Congenital cytomegalovirus is the second most frequent cause of bilateral hearing loss in young French children. J Pediatr 2013;162:593–9.
[16] El Bakkouri W, Loundon N, Thierry B, et al. Cochlear implantation and congenital deafness: perceptive and lexical results in 2 genetically pediatric identified population. Otol Neurotol 2012;33:539–44.