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Should You Follow the Better-Hearing Ear for Congenital Cytomegalovirus Infection and Isolated Sensorineural Hearing Loss?

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Abstract

Objective.—To describe the progression of sensorineural hearing loss (SNHL) in the better- and poorer-hearing ears in children with asymptomatic congenital cytomegalovirus (CMV) infection with isolated SNHL.

Study Design.—Longitudinal prospective cohort study.

Setting.—Tertiary medical center.

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Author Contributions

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Subjects and Methods.—We analyzed hearing thresholds of the better- and poorer-hearing ears of 16 CMV-infected patients with isolated congenital/early-onset or delayed-onset SNHL identified through hospital-based CMV screening of >30,000 newborns from 1982 to 1992.

Results.—By 12 months of age, 4 of 7 patients with congenital/early-onset SNHL developed worsening thresholds in the poorer-hearing ear, and 1 had an improvement in the better-hearing ear. By 18 years of age, all 7 patients had worsening thresholds in the poorer-hearing ear and 3 patients had worsening thresholds in the better-hearing ear. Hearing loss first worsened at a mean age of 2 and 6 years in the poorer-and better-hearing ears, respectively. Nine patients were diagnosed with delayed-onset SNHL (mean age of 9 years vs 12 years for the poorer- and better-hearing ears), 6 of whom had worsening thresholds in the poorer-hearing ear and 1 in both ears.

Conclusion.—In most children with congenital CMV infection and isolated SNHL, the poorer-hearing ear worsened earlier and more precipitously than the better-hearing ear. This study suggests that monitoring individual hearing thresholds in both ears is important for appropriate interventions and future evaluation of efficacy of antiviral treatment.

Keywords

cytomegalovirus; sensorineural hearing loss; infection; hearing; asymptomatic

Congenital cytomegalovirus (CMV) is the most common infectious cause of sensorineural hearing loss (SNHL) in developed countries.[1] With a prevalence of 4.5 per 1000 live births, it is estimated that up to 20,000 congenitally infected infants are born in the United States annually.[2] About 10% to 15% of these infants present with symptomatic congenital CMV disease at birth, 0.5% die in the neonatal period, and 20% develop permanent disabilities,[3] the most common of which is SNHL. Congenital CMV infection accounts for approximately 15% to 20% of bilateral moderate to profound SNHL in young children.[4] Hearing loss, especially its delayed and progressive nature, has detrimental effects on prompt detection, speech, and language development and adds to the overall healthcare cost associated with congenital CMV infection. Among the much larger group of infants who are asymptomatic at birth, almost 10% will develop SNHL,[5,6] 5% will require hearing amplification and rehabilitation, and more than 2% will potentially be candidates for cochlear implantation.[6]

A recent clinical trial by the National Institute of Allergy and Infectious Disease Collaborative Antiviral Study Group suggested that a 6-month course of valganciclovir (VGC) antiviral treatment initiated in the first month of age improved hearing and developmental outcomes in infants with symptomatic congenital CMV disease with central nervous system involvement.[7] The primary outcome of the trial was the change in better-ear hearing from baseline to 6-month follow-up.[7] Whether VGC treatment is effective for infants with asymptomatic congenital CMV infection and isolated SNHL is unknown. At least half of children with asymptomatic congenital CMV infection and isolated SNHL will initially be diagnosed with unilateral loss.[6] Despite their increased risk of developing bilateral loss, hearing thresholds in the better-hearing ear may remain normal in about 80% of those children by 12 to 14 months of age.[6]

The authors of this article, who are affiliated with the University of Utah School of Medicine, are about to embark on a National Institutes of Health–funded multi-institutional clinical trial to determine the efficacy and safety of VGC for infants with asymptomatic congenital CMV infection with isolated SNHL (ValEAR trial: ClinicalTrials.gov identifier NCT03107871). A key question for the clinical trial design is which ear should be evaluated as the primary end point (ie, the better- or poorer-hearing ear). Determining which ear is more likely to worsen over time can inform the designation of clinical trial end points so that any improvement from VGC can be assessed. This study describes SNHL progression in the better- and poorer-hearing ears in children with asymptomatic congenital CMV infection with isolated SNHL.

Methods

Enrollment of newborns into the Houston Congenital CMV Longitudinal Study has been described in prior publications.[6,8] Briefly, from 1982 to 1992, 32,543 newborns delivered at Women's Hospital of Texas were screened for congenital CMV infection. Of 135 (0.4%) CMV-positive newborns, 92 (68%) were enrolled in a longitudinal study as asymptomatic patients (ie, they had no CMV-related signs or symptoms at birth). The Institutional Review Board for Human Subject Research for Baylor College of Medicine and Affiliated Hospitals approved the study protocol.

Unaided audiologic assessments were conducted by audiologists blinded to the patient's CMV status and included auditory brainstem response (ABR), behavioral audiometry(0.25–8 kHz), and tympanometry.[6,8] ABR testing included click and frequency-specific tone-burst stimuli with correction factors for comparison with behavioral thresholds.[9] For ABR assessments, hearing loss was defined as threshold responses >25 dB for click or for any of the tone burst frequencies at 1, 2, or 4 kHz.[10,11] For behavioral testing, hearing loss was defined as any threshold >20 dB at 1, 2, or 4 kHz. Hearing threshold results obtained when patients had middle ear fluid were excluded from the analysis.

We included in this analysis asymptomatic patients with isolated SNHL, classified for the purpose of this analysis as either congenital/early-onset or delayed-onset. We defined a patient with congenital/early-onset SNHL as having unilateral or bilateral hearing loss detected in the first ABR assessment at age ≤6 months and confirmed in at least 1 subsequent assessment by 24 months of age and delayed-onset SNHL when the initial ABR assessment was normal for both ears; a second test was done by 24 months of age and hearing loss was detected after 1 year of age.

We analyzed SNHL by laterality; the categorization of the poorer- and better-hearing ears was based on the first hearing assessment when hearing loss was diagnosed. The ear with the worse threshold was defined as the poorer-hearing ear. If both ears were found to have hearing loss with identical thresholds, the assessment when the hearing in both ears first diverged in severity was used to determine the poorer- or better-hearing ear. This situation occurred in only 1 patient who had congenital/early-onset SNHL (patient 5; Figure 1). We also categorized SNHL severity for each ear based on click ABR or an average of 3 frequencies from behavioral testing: 1, 2, and 4 kHz. Severity was defined as mild for 30 to

45 dB (ABR) or 21 to 45 dB (behavioral), then for all testing methods moderate for 46 to 70 dB, severe for 71 to 90 dB, and profound for >90 dB.

For patients with congenital/early-onset SNHL, the hearing change in the better- and poorer-hearing ears was assessed between the first hearing test and 2 time points, the one closest to 12 months of age (0.75–1.25 years) and at 18 years of age. For patients with delayed-onset SNHL, the hearing change was assessed between the first hearing test and at 18 years of age. The hearing change was calculated using the threshold obtained in the first click ABR as baseline and subsequent click ABR or average of 1, 2, 3 and 4 kHz from behavioral testing. To limit the effects of fluctuations in thresholds within the normal range, any threshold determined to be under 15 dB was set at 15 dB prior to analysis. This would allow a change to be clinically significant for hearing loss onset (ie, 15–25 dB in behavioral testing) but not within normal hearing thresholds (ie, −10 to 20 dB). The average and range of change by ear were reported in decibels and do not necessarily reflect clinically significant change. A clinically significant worsening hearing was defined as a threshold increase of greater than a 10-dB change in a click ABR result, greater than a 15-dB change in 1 frequency, or greater than a 10-dB change in more than 1 frequency in behavioral testing. This categorization included changes at any frequency (0.25, 0.5, 1, 2, 4, and 8 kHz), although hearing loss was defined based only on 1, 2, and 4 kHz.

Results

Among 20 asymptomatic patients with isolated SNHL in the longitudinal study,[6] 4 did not meet criteria of congenital/early-onset or delayed-onset SNHL considered for this study. Two of 9 patients previously reported with congenital/early-onset SNHL were excluded because 1 did not have the first ABR assessment at ≤6 months of age, and the other did not have a second assessment around 24 months of age. Two of 11 patients previously reported with delayed-onset SNHL were excluded; 1 did not undergo any hearing assessment by 24 months of age, and the other had SNHL at 8 kHz only. Demographic characteristics of the 16 asymptomatic patients with isolated SNHL are shown in Table 1.

Among 7 patients with congenital/early-onset SNHL included in this study, the mean number of audiologic assessments was 13 (range, 10–17). The mean age of the first ABR evaluation was 0.2 years (range, 0.1–0.4 years). The mean age of the last hearing assessment was 17 years (range, 13–18 years). Individual hearing thresholds over time are depicted in Figure 1. By 12 months of age, 4 of 7 patients developed worsening thresholds in the poorer-hearing ear, while there was no change in the better-hearing ear; 2 patients had no change in their hearing for either ear (Patients 2 and 6); and 1 patient (5) had an improvement in the better-hearing ear while the poorer-hearing ear remained unchanged. By 12 months of age, the average change in the poorer-hearing ear was 17 dB (range, 0–44 dB) and −2 dB (range, −20 to 10 dB) in the better-hearing ear. Three patients had a unilateral profound loss by 12 months of age. When evaluated through 18 years of age, all 7 patients with congenital/early-onset SNHL had worsening thresholds in the poorer-hearing ears (Table 2). Three of the 7 patients had worsening in the better-hearing ears, and 4 had no change in hearing. The average change in the poorer-hearing ear was 43 dB (range, 18–71 dB). The average change

in the better-hearing ear was 20 dB (range, −5 to 68 dB). Three of 7 patients developed a profound hearing loss in only the poorer-hearing ear.

Among the 9 patients with delayed-onset SNHL included in this study, the mean age of diagnosis of delayed-onset SNHL was 9 years for the poorer-hearing ear (n = 9) compared to 12 years for the better-hearing ear (n = 2). The mean number of audiologic assessments was 11 (range, 5–17). The mean age of the first ABR evaluation was 0.2 years (range, 0.1–0.4 years). The mean age of last hearing assessment was 17 years (range, 9–19 years). Individual differences in hearing thresholds over time are depicted for each patient in Figure 2. Patients 11 and 15 had worsening hearing thresholds bilaterally (Table 3). Six patients (8, 9, 10, 12, 13, 14) developed worsening thresholds in the poorer-hearing ear while the better-hearing ear remained unchanged. One patient (16) had no significant hearing threshold changes. The mean line plots for all the patients' hearing thresholds by ear are shown in Figure 2. The average change in the poorer-hearing ear was a worsening of 24 dB (range, 5–100 dB). The change in the better-hearing ear was not clinically significant (range, 5–10 dB). Clinically relevant hearing loss was variable, ranging from normal hearing bilaterally due to slight or mild loss at isolated frequencies (patients 9, 13, 15, and 16) to 1 patient who developed unilateral profound hearing loss (patient 8). There were no patients with bilateral profound hearing loss. Table 3 details the change in hearing severity for patients with delayed-onset SNHL throughout childhood.

Discussion

This study included children with asymptomatic congenital CMV infection with isolated SNHL who were identified by a hospital-based newborn CMV screening during the 1980s and 1990s to describe the natural history of disease. By age 12 months, most patients with congenital/early-onset SNHL had worsening hearing thresholds in the poorer-hearing ear and no change in the better-hearing ear. The results of this study suggest that assessing individual hearing thresholds in both ears is important to track hearing worsening in the poorer-hearing ear in children with asymptomatic congenital CMV infection with isolated SNHL.

Describing which ear is more likely to worsen from this "natural history study" will directly inform selection of the appropriate primary outcome measure and follow-up duration for clinical trials and long-term hearing monitoring. In clinical trials for children with symptomatic congenital CMV disease, the primary outcome was a change in the better-hearing ear within a 6-month follow-up period.[7,12] We found that over half of asymptomatic patients with isolated SNHL had worsening hearing thresholds in the poorer-hearing ear and no child had worsening in the better-hearing ear by 12 months of age. A previous meta-analysis including 95 children with asymptomatic congenital CMV infection and isolated SNHL found that 57% had unilateral loss, although the follow-up duration in each study was variable. In our cohort, the median interval from unilateral to bilateral SNHL was 4 years.[6] Designating the better-hearing ear (which might remain with normal thresholds) as a primary end point would result in smaller differences between study groups and therefore lower likelihood to detect statistically significant differences. When the primary end point is negative, secondary end points that are positive will usually be considered hypothesis

generating.[13] The results of this analysis suggest there is greater statistical power for using either the poorer-hearing ear or the ear with worse hearing progression as a primary endpoint to assess an interventional efficacy.

In the follow-up period through 18 years of age, the poorer-hearing ear worsened earlier and more precipitously than the better-hearing ear in most patients with congenital/early-onset SNHL. However, in 1 patient (5) with bilateral symmetrical loss and improved hearing thresholds in the better-hearing ear by 12 months of age, hearing deterioration occurred in both ears, such that the patient had profound hearing loss bilaterally by 18 years of age. This finding is consistent with the observation that patients with fluctuating thresholds will typically develop progressive loss, although we did not assess this characteristic separately for the better- and poorer-hearing ears.[6] Patients with delayed-onset SNHL also demonstrated a similar pattern of hearing progression in that the poorer-hearing ear appeared to worsen earlier than the better-hearing ear. However, the overall progression was slower and the degree of loss was less severe than for children with congenital/early-onset SNHL; only 2 poorer-hearing ears ever reached severe or profound hearing loss, and all but 1 better-hearing ear retained normal hearing.

Defining guidelines and recommendations for hearing monitoring of children with congenital CMV infection has become increasingly important as more hospitals and states have mandated early CMV screening. Currently, 5 states—Connecticut, Illinois, Iowa, New York, and Utah—have legislated some form of hearing-targeted CMV screening. This approach was first instituted on a large scale at 1 birthing hospital in Dallas, Texas.[14] Any infant who did not pass an automatic ABR study before discharge was tested for CMV using urine culture. Over a 5-year period, 24 (5%) of 483 infants who failed the newborn hearing screen, including 16 (6%) of 256 infants diagnosed with hearing loss, had congenital CMV infection. Other studies in the era of universal hearing screening found that 3% to 28% of children with confirmed SNHL had congenital CMV infection.[15–23] The province of Ontario, Canada, is transitioning from a hearing-targeted CMV program to a universal one using neonatal dried blood spot samples.

In 2015, a panel of experts on congenital CMV, the International Congenital Cytomegalovirus Recommendations Group, recommended audiological testing at 6-month intervals for the first 3 years and annually thereafter through adolescence (ages 10–19).[24] Goderis et al[5] recommended long-term audiological follow-up of both asymptomatic and symptomatic children with congenital CMV infection for ≥6 years. Kadambari et al[25] cited the National Deaf Children's Society guidelines recommending hearing assessment for babies with congenital CMV infection every 3 to 6 months in the first year until age 3 and then annually until 6 years. Despite these recommendations, there are no widely adopted guidelines and none of these studies addressed which ear to assess.

Sound field behavioral testing assesses both ears together and would not be appropriate to track hearing worsening in individual ears. Assessing hearing thresholds in each ear among infants and young children is not trivial; 8.2% to 45.9% of behavioral testing has been reported as unsuccessful.[26] Some patients may require multiple visits to establish hearing thresholds through behavioral testing, and others may require sedated ABR testing. Thus,

when monitoring individual hearing thresholds, many audiologists choose to test the better-hearing ear only due to time constraints and young children's short attention spans. Nonetheless, the results of this study suggest that monitoring individual hearing thresholds in both ears is important for appropriate interventions.

This study has several limitations. The sample size is small, although this series is the largest to date, with children with asymptomatic congenital CMV infection identified through screening of more than 30,000 newborns and followed through 18 years of age. Although other potential causes of SNHL were possible, we were unable to investigate for the presence of such causes. Although we excluded 2 patients with congenital/early-onset SNHL from this analysis, they also had earlier and more precipitous worsening hearing thresholds in the poorer-hearing ear.

Conclusion

In our cohort, for most children with congenital CMV infection and isolated SNHL, either congenital/early-onset or delayed-onset, the poorer-hearing ear worsened earlier and more precipitously than the better-hearing ear. In children with initially symmetric hearing loss, it may be difficult to predict which ear will experience a greater loss of hearing over time. This study suggests that monitoring individual hearing thresholds in both ears is important for appropriate interventions and future evaluation of efficacy of antiviral treatment. These results will be relevant to audiologists and otolaryngologists as more and more children are diagnosed with asymptomatic congenital CMV infection and isolated SNHL.

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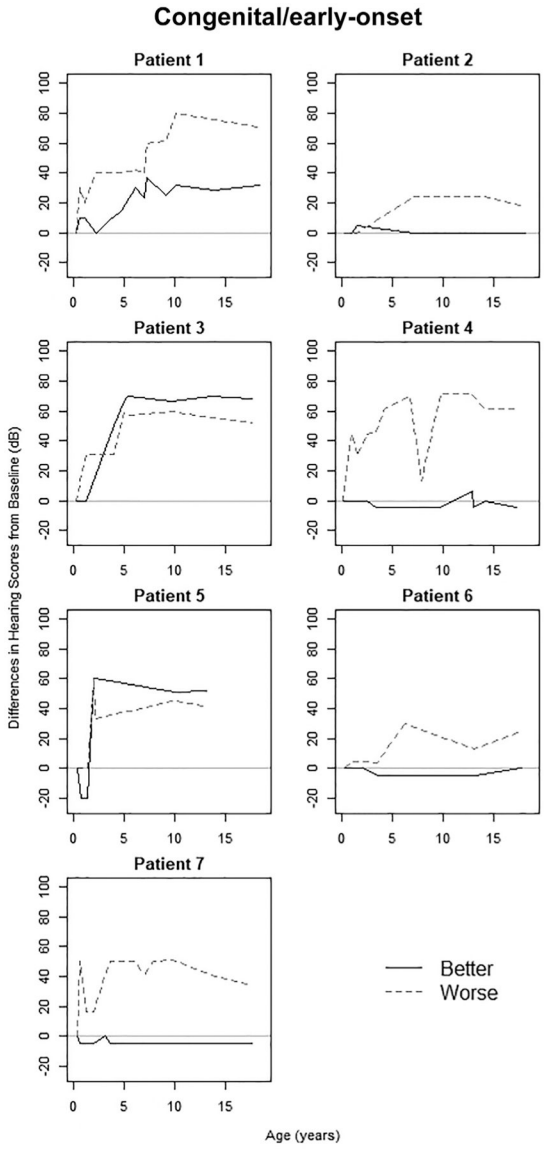

Figure 1.
Hearing changes over time in patients with congenital/early-onset sensorineural hearing loss.

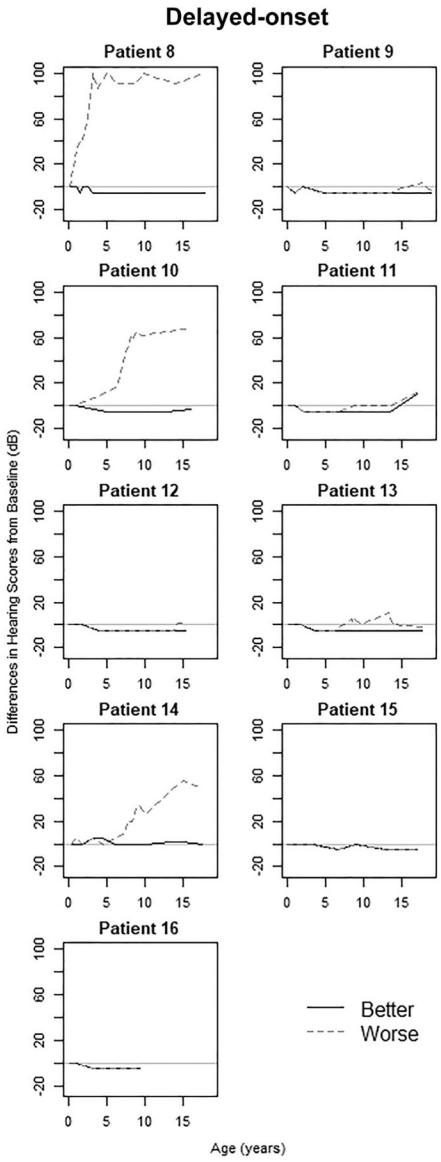

Figure 2.
Hearing changes over time in patents with delayed-onset sensorineural hearing loss.

Table 1.

Demographic Characteristics of Asymptomatic Patients with Sensorineural Hearing Loss.

Demographic Characteristics	No. (%)
Sex	
Male	13 (81)
Female	3 (19)
Mother's age, y	
<20	2 (12)
20–29	7 (44)
30–39	6 (38)
40–49	1 (6)
Mother's race/ethnicity	
Non-Hispanic white	16 (100)
Mother's marital status	
Married	15 (94)
Mother's education	
Some college	7 (44)
Graduated college	6 (38)
Postgraduate degree	3 (18)
Health insurance	
Private/health maintenance organization	11 (69)
Medicaid	1 (6)
Unknown	4 (25)
Socioeconomic status	
Low	1 (6)
Medium	7 (44)
High	8 (50)

Table 2.

Hearing Change from First Assessment to 18 Years of Age among Children with Congenital Sensorineural Hearing Loss.

Patient No.	Age at First Hearing Assessment, y	Age at Last Hearing Assessment, y	Change in Poorer-Hearing Ear, dB	Change in Better-Hearing Ear, dB	Clinical Change in the Poorer-Hearing Ear	Clinical Change in the Better-Hearing Ear
1	0.1	17	51	22	Worse	Worse
2	0.1	18	18	0	Worse	None
3	0.2	18	21	68	Worse	Worse
4	0.2	18	17	−5	Worse	None
5	0.3	18	42	72	Worse	Worse
6	0.4	13	20	0	Worse	None
7	0.2	18	18	0	Worse	None
Average	0.2 ± 0.1	17 ± 2	27 ± 14	22 ± 34		

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Table 3.

Hearing Change from First Assessment to 18 Years of Age among Children With Delayed-Onset Sensorineural Hearing Loss.

Patient No.	Age at First Hearing Assessment, y	Age at Last Hearing Assessment, y	Change in Poorer-Hearing Ear, dB	Change in Better-Hearing Ear, dB	Clinical Change in the Poorer-Hearing Ear	Clinical Change in the Better-Hearing Ear
8	0.2	18	100	−5	Worse	None
9	0.1	19	−3	−5	Worse	None
10	0.2	16	68	−3	Worse	None
11	0.2	17	12	10	Worse	Worse
12	0.1	15	2	−5	Worse	None
13	0.2	18	−2	−5	Worse	None
14	0.4	18	48	0	Worse	None
15	0.3	14	−5	−5	Worse	Worse
16	0.1	17	−5	−5	None	None
Average	0.2 ± 0.1	17 ± 2	24 ± 39	−3 ± 5		