Cytomegalovirus (CMV) virus infection is the most common intrauterine viral infection. Although approximately 90% of congenitally infected infants remain asymptomatic, the remaining 10% develop neurologic deficits, chorioretinitis, or sensorineural hearing loss (SNHL).

CMV infection is the cause of around 13–22% of all cases of neonatal hearing loss. The GJB2 mutation accounts for 30–50% of all cases of profound nonsyndromic hearing loss in many populations, and is observed in about 25% of Japanese patients with congenital hearing loss.

Cochlear implantation has been shown to be effective in the treatment of SNHL associated with congenital CMV infection and GJB2 mutation.

In this study, after cochlear implantation in children with hearing loss associated with either congenital CMV infection or GJB2 mutation, their hearing and speech and language development was examined. The effectiveness of cochlear implantation for patients with multiple disorders including mental retardation was also evaluated.

Among 43 children with congenital hearing loss who underwent cochlear implantation at our hospital, 12 children in whom hearing loss was associated with either congenital CMV infection or GJB2 mutation were included in this study.

The audiological and subjective characteristics of our study group are given in Table 1. After cochlear implantation, the hearing level and speech and language development of the patients were evaluated using the Infant-Toddler Meaningful Auditory Integration Scale (IT-MAIS), the Meaningful Use of Speech Scale (MUS), and the Test for Language Retardation Based on Sign-Significate Relations (S-S method).

Full electrode insertion was possible in all cases of cochlear implantation, and none of the children experienced postoperative complications such as meningitis, facial palsy, or wound infection.

In this study, favorable surgical outcomes and the effectiveness of cochlear implantation for improving hearing and speech development were confirmed. Children with multiple disorders showed low levels of development in hearing, language comprehension, and speech production following cochlear implantation.

In our study, the fact that 3 of the 4 children with MR in both groups (cases C5, G5, and G6) showed development, albeit slow, following cochlear implantation demonstrates the effectiveness of the surgical procedure.

The one child with GJB2 mutation and AD/HD (case G7) also performed well on the IT-MAIS, MUS, and S-S method tests.

Thus, the presence of multiple disorders, especially severe PDD or MR, may preclude development following cochlear implantation, but should not contraindicate the use of the surgical procedure by itself. In other words, the effect of cochlear implantation depends on the presence or absence of coexisting multiple disabilities rather than having congenital CMV infection or GJB2 mutation as the cause.

Accurate diagnosis is the future goal since it is expected that the age for cochlear implantation will be lowered and the diagnosis of multiple disabilities will remain difficult.

The present results suggest that the use of cochlear implantation is appropriate for patients with SNHL associated with asymptomatic CMV infection or GJB2 mutation. In the case of multiple disorders, the surgery will likely be approved by the guardians after full awareness of the outcomes of cochlear implantation.

Cochlear implantation was effective in improving development in hearing, language comprehension and speech in children with hearing loss associated with congenital CMV infection or GJB2 mutation.

The children with MR tended to show low levels of improvement in language comprehension and speech. Although our patients with developmental disorder showed poor auditory performance and speech and language skills after cochlear implantation, SNHL with developmental disorder should not be a contraindication for the procedure.

For children with multiple disorders, the surgery may achieve only a limited level of development following surgery, informed consent and aural habilitation post-implantation should be given on a case-by-case basis.

REFERENCES