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Developmental delays assessed using the Enjoji Scale in children with cochlear implants who have intellectual disability with or without autism spectrum disorder

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ABSTRACT

Objective: Intellectual disability (ID) and autism spectrum disorder (ASD) are common among children who are candidates for cochlear implants. However, the implications of these comorbidities for cochlear implant placement have been not fully established. This study sought to identify these implications by comparing developmental delays among children with these conditions.

Methods: Participants were children who were followed up at least every 6 months for 24 months after cochlear implant surgery. Developmental delays were assessed using the Enjoji Scale of Infant Analytical Development (Enjoji Scale) and compared in three groups with hearing loss: those with ID (ID group, n = 4); those with ASD and ID (ASD + ID group, n = 4); and those with typical development (control group, n = 5). Developmental delay was evaluated longitudinally before and after cochlear implant placement for 18 months.

Results: Among the six subscales that make up the Enjoji Scale, language development and intelligence development were significantly delayed in all three groups and were exacerbated over time except for language development in the control group. Emotional development and social behavior were significantly delayed only in the ASD + ID group. Comparison of intergroup differences revealed delays in language development in the ID and ASD + ID groups compared with the control group.

Conclusion: The Enjoji Scale successfully demonstrated developmental delays characteristic to the underlying comorbidities of ID with or without ASD in children with cochlear implants. The Enjoji Scale can be a useful diagnostic tool for screening children with cochlear implants for ID with or without ASD.

1. Introduction

The criteria for cochlear implant candidacy differ between countries [1] but does not routinely preclude children with multiple disabilities or children with syndromes and conditions associated with disability such as blindness, reduced cognitive ability, mental retardation, global learning difficulties, and attention deficit disorder [2]. Indeed, multiple disabilities are relatively common among children with cochlear implants for profound hearing loss. For example, between around 15% to 40% of children with prelingual hearing loss who have had cochlear implants are reported to have co-occurring disability [3]. These patients in general received significant auditory benefit post-implant placement, although progress was

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typically slow [4]. The difficulty in obtaining audiologic or speech and language development benefits from cochlear implant placement in children with multiple disabilities depends on the features and severity of the co-occurring disability, which varies for each child [3].

Among the co-occurring disabilities, intellectual disability (ID) and autism spectrum disorder (ASD) are relatively common entities. For example, moderate and severe ID co-occurred in 5% of children with cochlear implants [5] and ASD in approximately 4% [5]. Due to the biology of developmental disorders, children with these conditions have developmental limitations not only in socio-communicative skills but also in diverse aspects of other skills. For example, in children with ASD, language development correlates with atypical behavior [6] and impaired joint attention [7]. In children with ID, motor function and cognitive function correlate with each other [8]. Besides, children with cochlear implants have associated difficulties with morphological and syntactic rules and inefficient narrative skills [9]. Therefore, for children with ID with or without ASD, we need to provide intervention that focuses on socio-communicative skills as well as comprehensively assess and support all other aspects of their development in order to mitigate risk and ensure the well-being of the children and their families [10,11].

Comprehensive assessment of development and intelligence in children with cochlear implants has been carried out with a number of tests that are widely used in the clinical setting. The advantage of these tests is that they provide a structured assessment of a child's development that clarifies the quantity and quality of the child's developmental status [12]. The Enjoji Scale of Infant Analytical Development (Enjoji Scale) is a concise, interview-based questionnaire requiring no special tools or devices to detect developmental delay (Table 1). The test evaluates developmental milestones in 6 categories (language development, intelligence development, emotional development, social behavior development, manual activity development and locomotor activity development) [13] and covers skill categories for the assessment of typical development (e.g., hearing, speech and language, intellectuality, emotion, social behavior, gross motor function, fine motor function, and vision) [12]. It takes only 15–20 min to complete and is suitable for following the developmental time course. It is widely used in Japan to assess children with atypical development [13–15].

Despite the growing number of children with cochlear implants who have multiple disabilities, the literature on developmental delay remains sparse to date. Therefore, in this study, we investigated longitudinal changes in developmental delays in children with hearing loss after receiving a cochlear implant, with or without co-occurring ID and ASD.

2. Materials and methods

2.1. Patients

We performed a retrospective chart review of 55 children (age ≤16 years) who underwent cochlear implant placement at Nagoya City University Hospital between January 2001 and

Table 1
The Enjoji Scale of Infant Analytical Development (excerpted from Enjoji et al. [13] and reworked).

Categories	Language (utterance ability)	Intelligence (language perception ability)	Emotional (interpersonal emotional ability)	Social behavior (development in living ability)	Manual activity (development in skilled motor activities)	Locomotor activity (development in trunk movement)
Months	Milestones					
66–72 months	Tells his age, address and name of parent	Answers correctly 3 simple questions	Shy of nakeness	Plays cooperative exercise games	Ties bow-knot	Stands on one foot for 10 s steadily
54–60 months	Doesn't talk baby talk	Understands number notion of 4	Uses threatening words	Puts on dress unaided	Imitates square and cross both	Skips on alternate feet
42–48 months	Gives full name	Distinguishes 3 grams from 15 g of weight	Shows jealousy for his younger brother or sister	Urinates unassisted	Buttons coat	Stands on one foot for 2 s
30–36 months	2 words sentences (put 2 words together)	Knows the meanings of upper, inner, under, front and back part, 2 of 5	Express his pleasure with exaggerated action	Uses chopsticks	Turns pages singly	Jumps with both feet
18–24 months	Uses verb	Identifies one picture	Shows affection to his doll or toy	Asks to toilet	Builds tower of three cubes	Runs

December 2014 and were recruited through a tertiary referral center, the otolaryngology clinic at Nagoya City University Hospital, Japan. All of the children had prelingual profound hearing loss and became candidates for cochlear implant placement after having shown no auditory or linguistic development after placement of a hearing aid. We included those whose auditory skills and development were evaluated before and after surgery and excluded those whose development did not follow for more than 18 months. This left 13 patients for analysis in this study: 3 boys and 10 girls with a mean age of 25.2 (±7.5) months and age range of 18–44 months at the time of cochlear implant placement. Cochlear implants were activated approximately 2 weeks after surgery, with habilitation and education for patients and their family being provided every 2 or 4 weeks for 3–6 months until cochlear implant mapping was optimized. After the optimization, these services are provided every 3 to 6 months. Concurrently, all the enrolled patients attend local public schools for deaf and hard of hearing children in the same region and are educated using similar programs.

The study was explained to the children's guardians, and both the children and their guardians were given the opportunity to opt out of participating. The study protocol was approved by the institutional review board of Nagoya City University, with a waiver of informed consent for the retrospective medical records review (Approval No.: 60-18-0068).

2.2. Determining co-occurring disabilities

Additional disabilities were diagnosed by a pediatric psychiatrist and pediatrician with expertise in pediatric psychiatry, based on the Diagnostic and Statistical Manual of Mental Disorders 5th edition. All 55 children were evaluated by pediatric psychologists prior to the surgery, and were referred to experts in pediatric psychiatry when necessary. Of the 55 identified children, 12 (21.8%) were diagnosed as having comorbid psychiatric disorder(s). Among these, 10 children were diagnosed as having ID and 4 of the 10 were diagnosed with co-existing ASD (i.e., all the children with ASD had accompanying ID; ASD + ID group). From among these 10 children, 8 children for whom consent was obtained to undergo periodic assessment using the Enjoji Scale were enrolled in the study (4 children from the ID group and 4 children from the ASD + ID group). Intelligence quotient was assessed in patients in the ID and ASD + ID group whenever possible, using the Tanaka–Binet Test (Japanese version of the Stanford-Binet Test, standardized by the Tanaka Education Institute, 1970, Table 2). From among children who had undergone cochlear implant surgery during the study period, control patients were enrolled who did not have any disability in addition to hearing loss and for whom consent was obtained to assess their development periodically using the Enjoji Scale.

2.3. Hearing and developmental evaluation

Hearing thresholds were measured using play audiometry performed by speech therapists. Scale out was calculated as 120 dB. Developmental delay was assessed using the Enjoji Scale [13] (Table 1). In this test, development is assessed by checking the child's performance on a chart that shows standard developmental milestones in language, social, and motor skills at 1-month intervals from 1 month to 12 months old, at 3-month intervals from 15 months to 18 months old, and at 6-months intervals from 24 months to 84 months old. There are six subscales: language development (utterance ability), intelligence development (language perception ability), emotional development (interpersonal emotional ability), social behavior development (development in living ability), manual activity development (development in skilled motor activities), and locomotor activity development (development in trunk movement) (Figs. 1–6, respectively). Developmental delay was calculated as the difference between actual developmental age (months) and chronological age (months). Cases where developmental age lags behind chronological age are indicated by the minus sign "-"; "delay" was defined as a lag of more than 4 months.

2.4. Statistical analysis

Unless otherwise noted, all data are presented as mean ± SEM. Developmental delays in each group were evaluated using the one-sample t-test against typical development, denoted 0 (SigmaPlot; Wavemetrics, Inc., Lake Oswego, OR). Statistical comparisons between each group were performed using the one-way ANOVA repeated measures and post hoc Holm–Šídák test; a P-value of $P < 0.05$ was considered statistically significant. For data that were not distributed normally, non-parametric analysis was performed using the Mann–Whitney U test rank sum test (SigmaPlot).

3. Results

3.1. Developmental delays in the control group

In the control group, preoperative assessment revealed significant a developmental delay in language of -15.4 ± 2.9 months. However, the delay was recovered over time to reach -6.4 ± 3.5 months at 18 months after implant placement, which was not significantly different from typical development (Fig. 1, Table 3). Intelligence development demonstrated the same tendency. In the control group, the delay of -14.0 ± 3.9 months preoperatively was improved at 18 months after placement to -13.8 ± 6.3 months. Locomotor activity development showed a small developmental delay of -2.4 ± 0.7 months preoperatively, which became insignificant from 6 months after placement (Fig. 6, Table 3).

3.2. Developmental delays in the ID group

Children with ID showed significant delay in language development of -18.5 ± 1.6 months preoperatively. Unlike the control group, the delay was exacerbated by 4.3 months at 6 months after implant placement and this delay was not recovered but was maintained with a slight exacerbation of 1.5 months over the next 12 months (Fig. 1, Table 3). Intelligence development at 12 months after implantation showed an exacerbation of 13.0 months from -11.5 ± 1.3 months delay

Table 2
Backgrounds of the enrolled children with cochlear implants.

Group	Sex	Comorbidity	Age at implant placement (mo)	Age at diagnosis as ID or ASD + ID (mo)	Pre CI HL unaided (dB)[a]	Post CI HL aided (dB)[a]	Pre-operative DQ	IQ (age, months)[b]	Note
Control group	F	N/A	18	N/A	110	30	98	N/A	Auditory neuropathy, Bilateral implants
	M		18		110	44	91		Bilateral implants
	F		21		113	26	83		
	F		19		108	28	77		
	F		33		110	33	81		
ID group	M	Moderate ID	19	53	120	55	54	N/A	Inner ear malformation (Incomplete partition type II)
	F	Moderate ID	18	49	120	34	58	61 (66 mo)	Bilateral implants
	F	Severe ID/ severe motor disability	26	24	120	45	38	N/A	
	F	Mild ID	23	32	110	40	67	74 (35 mo)	Bilateral implants
ASD + ID group	F	Mild ID	28	59	120	35	69	N/A	Bilateral implants
	F	Moderate ID	28	70	88	42	58	70 (148 mo)	Low birthweight infant
	M	Severe ID	32	56	110	30	34	56 (80 mo)	Waardenburg syndrome
	F	Severe ID	44	42	110	44	39	N/A	

ID: intellectual disability; ASD: autism spectrum disorder; DQ: development quotient; IQ: intelligence quotient; CI: cochlear implant; HL: hearing loss.
[a] Hearing level indicates the average of hearing thresholds at 0.5, 1, 2, and 4 kHz. Patients with no response at the maximum (115 dB) were arbitrarily assigned thresholds 5 dB above the limit (120 dB).
[b] Determined by Tanaka–Binet Test.

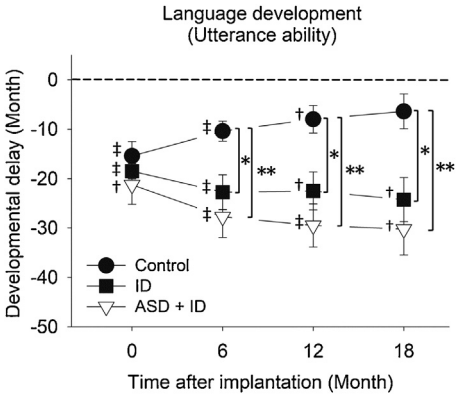

Fig. 1. Temporal changes in language development (utterance ability).

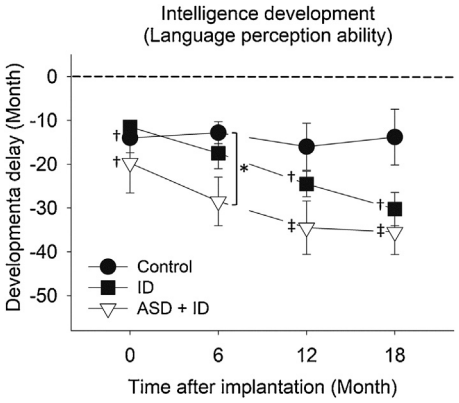

Fig. 2. Temporal changes in intelligence development (language perception ability).

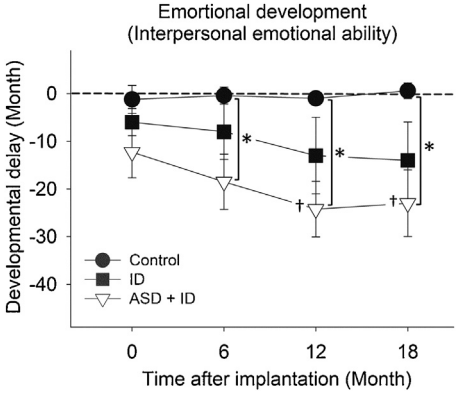

Fig. 3. Temporal changes in emotional development (interpersonal emotional ability).

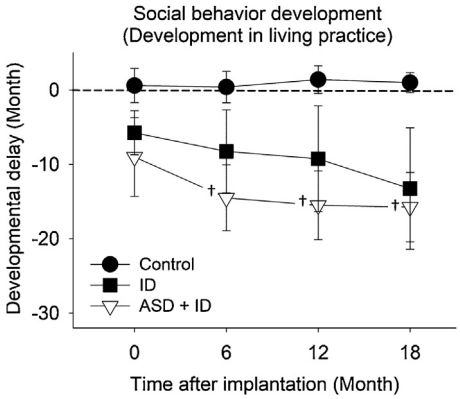

Fig. 4. Temporal changes in social behavior development (development in living ability).

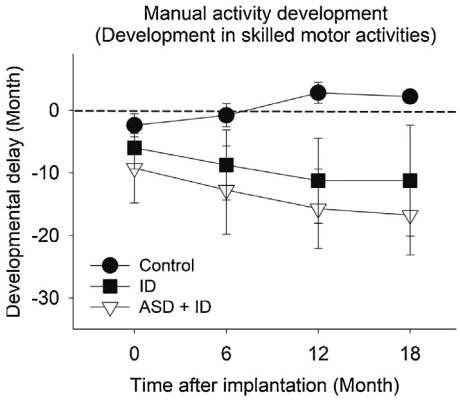

Fig. 5. Temporal changes in manual activity development (development in skilled motor activities).

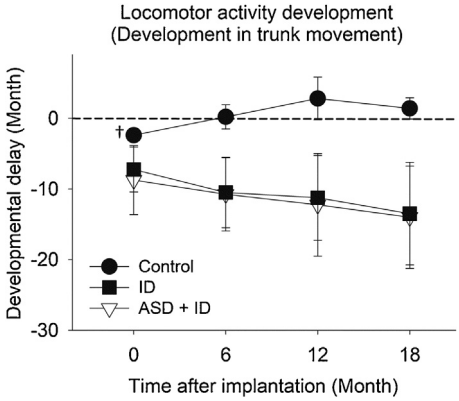

Fig. 6. Temporal changes in locomotor activity development (development in trunk movement).

preoperatively. The delay was significant from 12 months after implantation (Fig. 2, Table 3).

3.3. Developmental delays in the ASD + ID group

As with the ID group, the ASD + ID group showed a significant delay in language development. Analogous to the ID group, the exacerbation of the delay reduced over time, showing 6.5-month exacerbation in the first 6 months and a further delay of 2.5 months in the following 12 months (Fig. 1, Table 3).

Intelligence development demonstrated the same tendency but the exacerbation persisted a little longer. In the first 12 months after implantation, developmental delay was exacerbated by 14.7, but presented with a 1.0-month exacerbation in the following 6 months (Fig. 2, Table 3).

Also, this group showed a significant delay in emotional development, unlike the control and ID groups. From 12 months after implant placement, emotional developmental delay reached −24.3 months, which is significantly longer than that of typical development (Fig. 3, Table 3).

Table 3
Developmental delays in the three groups of children with cochlear implants compared with typical development.

Category	Group	Preoperative value	After 6 months	After 12 months	After 18 months
Language development (utterance ability)	Control	−15.4 ± 2.9	−10.4 ± 2.0	−8.0 ± 2.8	−6.4 ± 3.5
		P = 0.006[b]	P = 0.007[b]	P = 0.045[a]	P = 0.144
	ID	−18.5 ± 1.6	−22.8 ± 3.5	−22.5 ± 3.9	−24.3 ± 4.5
		P = 0.001[b]	P = 0.008[b]	P = 0.010[a]	P = 0.012[a]
	ASD + ID	−21.3 ± 3.9	−27.8 ± 4.2	−29.5 ± 4.4	−30.3 ± 5.2
		P = 0.013[a]	P = 0.007[b]	P = 0.007[b]	P = 0.010[a]
Intelligence development (language perception ability)	Control	−14.0 ± 3.9	−12.8 ± 1.8	−16.0 ± 5.4	−13.8 ± 6.3
		P = 0.022[a]	P = 0.113	P = 0.279	P = 0.641
	ID	−11.5 ± 1.3	−17.5 ± 3.5	−24.5 ± 2.9	−30.3 ± 3.8
		P = 0.085	P = 0.052	P = 0.022[a]	P = 0.010[a]
	ASD + ID	−19.8 ± 6.8	−28.5 ± 5.5	−34.5 ± 6.1	−35.5 ± 5.1
		P = 0.019[a]	P = 0.125	P = 0.004[b]	P = 0.001[b]
Emotional development (interpersonal emotional ability)	Control	−3.5 ± 2.1	−0.4 ± 1.8	−1.0 ± 0.9	0.6 ± 1.6
		P = 0.508	P = 0.833	P = 0.351	P = 0.732
	ID	−6.0 ± 2.9	−8.0 ± 5.8	−13.0 ± 8.0	−14.0 ± 8.0
		P = 0.127	P = 0.261	P = 0.204	P = 0.180
	ASD + ID	−12.3 ± 5.4	−18.5 ± 5.8	−24.3 ± 5.8	−23.0 ± 7.0
		P = 0.108	P = 0.050	P = 0.025[a]	P = 0.046[a]
Social behavior development (development in living ability)	Control	0.6 ± 2.3	0.4 ± 2.1	1.4 ± 1.9	1.0 ± 1.3
		P = 0.807	P = 0.859	P = 0.494	P = 0.497
	ID	−5.8 ± 2.9	−8.3 ± 5.6	−9.3 ± 7.1	−13.3 ± 8.2
		P = 0.147	P = 0.235	P = 0.235	P = 0.204
	ASD + ID	−9.0 ± 5.3	−14.5 ± 4.4	−15.5 ± 4.6	−15.8 ± 4.7
		P = 0.188	P = 0.047[a]	P = 0.044[a]	P = 0.043†
Manual activity (development in skilled motor activities)	Control	−2.6 ± 1.9	−0.8 ± 1.9	2.8 ± 1.7	2.2 ± 1.1
		P = 0.266	P = 0.688	P = 0.172	P = 0.108
	ID	−6.0 ± 3.4	−8.8 ± 5.6	−11.3 ± 6.8	−11.3 ± 8.9
		P = 0.175	P = 0.216	P = 0.197	P = 0.294
	ASD + ID	−9.3 ± 5.6	−12.8 ± 7.1	−15.8 ± 6.4	−16.8 ± 6.4
		P = 0.196	P = 0.168	P = 0.090	P = 0.079
Locomotor activity (development in trunk movement)	Control	−2.4 ± 0.7	0.2 ± 1.7	2.8 ± 3.0	1.4 ± 1.5
		P = 0.024[a]	P = 0.913	P = 0.407	P = 0.404
	ID	−7.3 ± 3.2	−10.5 ± 5.0	−11.3 ± 6.0	−13.5 ± 7.3
		P = 0.106	P = 0.125	P = 0.158	P = 0.160
	ASD + ID	−8.8 ± 4.9	−10.8 ± 5.2	−12.3 ± 7.3	−14.0 ± 7.3
		P = 0.170	P = 0.130	P = 0.190	P = 0.149

ID: intellectual disability, ASD: autism spectrum disorder.
[a] $P < 0.05$.
[b] $P < 0.01$.

Social behavior had the same tendency with emotional development; Significant developmental delay was observed only in this group; a 14.5-month delay at 6 months after implant placement was subsequently maintained (Fig. 4, Table 3).

Development inmanual activity and locomotor activity were delayed but not significantly so (Figs. 5 and 6, Table 3).

3.4. Between-group comparison

To analyze differences between the three groups, we tested intergroup differences with ANOVA. Language development in the ID and ASD + ID groups and emotional development in the ID group were statistically different from the control group (Table 4). Language developmental delay was significant at 6 months and later after surgery in both the ID and ASD groups. Similarly, emotional developmental delay in the ID and ASD group was significant within the same timeframe, from 6 months onward, versus the control group (Table 4).

Intelligence development in the ID group was significantly delayed versus that of the control group at only 6 months, not at other timepoints examined in this study.

4. Discussion

The Joint Committee on Infant Hearing recommends that early intervention of hearing impairment is crucial for good outcomes in verbal communication [16]. Owing to the international prevalence of the newborn hearing screening program [17,18], the number of children with early diagnosis of hearing impairment and early intervention including cochlear implantation is increasing.

In contrast, the mainstay for early detection of child psychiatric problems remains developmental surveillance and screening at preventive health care visits [19], regardless of the importance of early intervention [10,20]. Although various early stage screening methods are proposed, early diagnosis of these diseases is still a challenge. For example, data from the 2002 multi-site Autism and Developmental Disabilities Monitoring Network indicated the

Table 4
Difference in developmental delays between the three groups.

Category	Group	Pre-operative value	After 6 months	After 12 months	After 18 months
Language development (utterance ability)	Control vs ID	P = 0.401	P = 0.039*	P = 0.032*	P = 0.029*
	Control vs ASD + ID		P = 0.009**	P = 0.005**	P = 0.009**
	ID vs ASD + ID		P = 0.313	P = 0.216	P = 0.371
Intelligence development (language perception ability)	Control vs ID	P = 0.443	P = 0.156	P = 0.158	P = 0.04*,[a]
	Control vs ASD + ID		P = 0.046*		
	ID vs ASD + ID		P = 0.403		
Emotional development (interpersonal emotional ability)	Control vs ID	P = 0.162	P = 0.252	P = 0.249	P = 0.185
	Control vs ASD + ID		P = 0.047*	P = 0.03*	P = 0.042*
	ID vs ASD + ID		P = 0.265	P = 0.177	P = 0.310
Social behavior development (development in living ability)	Control vs ID	P = 0.189	P = 0.066	P = 0.106	P = 0.074
	Control vs ASD + ID				
	ID vs ASD + ID				
Manual activity development (development in skilled motor activities)	Control vs ID	P = 0.872	P = 0.247	P = 0.056	P = 0.097
	Control vs ASD + ID				
	ID vs ASD + ID				
Locomotor activity development (development in trunk movement)	Control vs ID	P = 0.667	P = 0.119	P = 0.122	P = 0.115
	Control vs ASD + ID				
	ID vs ASD + ID				

ID: intellectual disability; ASD: autism spectrum disorder.
* $P < 0.05$.
** $P < 0.01$.
[a] All Pairwise Multiple Comparison Procedures (Holm–Šídák method) did not yield significant difference (Overall significance level = 0.05).

median age of ASD diagnosis to be 5.7 years [21] and this was similar even among those with profound hearing loss or total hearing loss (5.5 years) [5]. Indeed, the number of children diagnosed with ASD or ID is increasing with the prevalence of cochlear implant placement and is more common in cochlear implant centers. However, the full range of developmental delay in these populations is still largely unknown, regardless of the importance of a comprehensive approach.

In patients with multiple disabilities, each disability develops in a different way based on the pathology of the entity. Moreover, the resultant symptomatology is a reflection of the components of each disability. Consequently, failure to attain developmental milestones is caused by a combined etiology, which makes it difficult to speculate the exact underlying pathology manifesting as the developmental delay. Likewise in the present study, the delay in language development can be caused not only by hearing impairment, but also by ID [22] and ASD [23].

To elucidate this point, here we compared children with three different types of disability: hearing impairment (control group); ID + hearing impairment (ID group); and ASD + ID + hearing impairment (ASD + ID group). It is highly likely that differences in developmental delays between each group reflect differences in the disabilities and can therefore be a clue to the underlying etiologies in children with cochlear implants, which is important but sometimes hard to deduce.

In this study, the control group showed developmental delay not only in language development but also in intelligence development. The tasks in the intelligence development category on the Enjoji Scale include steps requiring the child to understand the request by the examiner, such as “understands simple requests” for development at 15 months. Thus, developmental delay in verbal communication would underlie the two categories of language and intelligence.

Children with ID in addition to hearing impairment (ID group) had developmental delay in both of these categories as did the control group, but the delay was exacerbated over time. ID, formerly called mental retardation, is a term used to describe general mental disability that develops due to abnormalities in brain structure or function. ID is characterized by significant limitations in both intellectual functioning and adaptive behavior as expressed in conceptual, social, and practical skills [24]. Exacerbation in these domains is assumed to reflect the pathology of the ID. In addition, a more recent study in cochlear implant recipients with ID compared with age- and sex-matched children with cochlear implants but without such disability showed different effects depending on the severity of ID. Children with mild disability showed significant benefit and children with moderate and more severe disability showed limited and minimal benefit, respectively [25]. The degree of exacerbation in developmental delay could therefore correlate with the severity of ID.

In our study, the group with ASD in addition to hearing impairment and ID (ASD + ID group) had developmental delay in emotional and social behavior development, aside from the language and intelligence development delay shown by children in the control and ID groups. This seems to be a reflection of the features of ASD, which is characterized by impaired social interaction, atypical communication, and repetitive, restrictive behaviors. Improvements in speech and language skills in hearing-impaired children with ASD have been reported [5], however, these children improved at half the rate of those of children with typical development, in both receptive and expressive language skills [26]. Increases in externalizing behaviors were also observed at 3 years after implant placement in the ASD group [5,27], although a study of hearing-impairment in ASD showed a generally poor effect on post-implant social development [28]. Although improvements were found in

language listening tasks and vocabulary examinations, communication with spoken language became possible only in those with mild ASD. These non-uniform outcomes in language skills seemed to be partly because of the difficulty in early identification of ASD among children who are deaf or hard of hearing. The median age of ASD identification was 66.5 months [5], which is older than the optimal age for cochlear implantation. During the period when ASD is yet unidentified, routine educational and curative programs for children without ASD may not be enough to achieve best outcomes in children with ASD. Moreover, the natural course of ASD might potentially impact the outcome. Indeed, linguistic ability among children with mild ASD tends to develop markedly around the age of 5 years, so it is difficult to judge whether the resolution of the delay in social skills was really due to the cochlear implant or was a result of naturally improved linguistic ability. Consistent with another study on children with ASD [29], our data suggest that the cochlear implant does not directly improve the characteristic features of ASD itself.

Based on previous reports on children with multiple disabilities, development was also stimulated by communication skills, social interaction, and connection with the environment after cochlear implantation [6,30]. Further benefits were reported such as deeper emotional ties in the family due to the child being able to hear the parents' voices [31]. In contrast, another report showed that children with cochlear implants, 90.5% of whom had cognitive disabilities, had significantly lower social skills compared with controls [32]. Inherently, children with congenital hearing impairment may have both impoverished linguistic and social environments, which may adversely affect their socio-cognitive development [33]. In addition, children with preoperative social immaturity and reduced social competence showed a negative association with improvement in auditory perception and speech production after cochlear implant placement [34]. Therefore, the benefits for sociality among these children may be fully demonstrated thanks to the improved comprehension following improved hearing loss.

On the other hand, due to the low prevalence of intellectual disability (∼1% of the population based on the DSM-5 diagnostic criteria [35,36]) and the small number of patients who undergo cochlear implant surgery for profound congenital hearing loss, a relatively small sample size was employed in the study. Besides, the patients with intellectual disability have a short attention span that is just long enough to perform at most two developmental or other hearing tests in most cases, and therefore, we could not perform other tests to validate the outcomes. These points can be considered as potential limitations of the findings.

In conclusion, the Enjoji Scale appears to be sufficiently informative for comprehensive developmental evaluation even among children with cochlear implants with possible underlying ID and ASD.

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