Cochlear Implants for Deaf Children With Early Developmental Impairment

John S. Oghalai, MD,[a] Heather Bortfeld, PhD,[b] Heidi M. Feldman, MD, PhD,[c] Niharika Chimalakonda, MD,[d] Claudia Emery, AuD,[e] Janet S. Choi, MD, MPH,[a] Shane Zhou, MD[a]

abstract

BACKGROUND AND OBJECTIVES: Infants with profound hearing loss are typically considered for cochlear implantation. Many insurance providers deny implantation to children with developmental impairments because they have limited potential to acquire verbal communication. We took advantage of differing insurance coverage restrictions to compare outcomes after cochlear implantation or continued hearing aid use.

METHODS: Young children with deafness were identified prospectively from 2 different states, Texas and California, and followed longitudinally for an average of 2 years. Children in cohort 1 ($n = 138$) had normal cognition and adaptive behavior and underwent cochlear implantation. Children in cohorts 2 ($n = 37$) and 3 ($n = 29$) had low cognition and low adaptive behavior. Those in cohort 2 underwent cochlear implantation, whereas those in cohort 3 were treated with hearing aids.

RESULTS: Cohorts did not substantially differ in demographic characteristics. Using cohort 2 as the reference, children in cohort 1 showed more rapid gains in cognitive, adaptive function, language, and auditory skills (estimated coefficients, 0.166 to 0.403; $P \leq .001$), whereas children in cohort 3 showed slower gains (-0.119 to -0.243; $P \leq .04$). Children in cohort 3 also had greater increases in stress within the parent-child system (1.328; $P = .02$), whereas cohorts 1 and 2 were not different.

CONCLUSIONS: Cochlear implantation benefits children with deafness and developmental delays. This finding has health policy implications not only for private insurers but also for large, statewide, publicly administered programs. Cognitive and adaptive skills should not be used as a "litmus test" for pediatric cochlear implantation.

Full article can be found online at www.pediatrics.org/cgi/doi/10.1542/peds.2021-055459

[a]*Caruso Department of Otolaryngology–Head and Neck Surgery, University of Southern California, Los Angeles, California;* [b]*Department of Psychological Sciences, University of California, Merced, Merced, California;* [c]*Department of Pediatrics, Stanford University, Stanford, California;* [d]*The Doctors Clinic, Port Orchard, Washington; and* [e]*Texas Children's Hospital, Houston, Texas*

Dr Oghalai conceptualized and designed the study, designed the data collection instruments, performed the analyses, and drafted the initial manuscript; Drs Bortfeld and Emery conceptualized and designed the study, designed the data collection instruments, collected data, and reviewed and revised the manuscript; Dr Feldman collected data and critically reviewed the manuscript and analyses for important intellectual content; Drs Chimalakonda, Choi, and Zhou collected data, performed the analyses, and reviewed and revised the manuscript; and all authors approved the final manuscript as submitted and agree to be accountable for all aspects of the work.

This trial has been registered at www.clinicaltrials.gov (identifier NCT01256229).

DOI: https://doi.org/10.1542/peds.2021-055459

Accepted for publication Mar 10, 2022

WHAT'S KNOWN ON THIS SUBJECT: Cochlear implantation has become the standard of care for children with deafness because it permits better speech and language acquisition. However, coexistent severe developmental delays have been used by insurance providers as a rationale to deny coverage for this treatment.

WHAT THIS STUDY ADDS: We demonstrate in this prospective cohort study in children with deafness and severe developmental impairment substantial benefits of cochlear implantation over hearing aids. Cognitive and adaptive skills should not be used as a "litmus test" for cochlear implantation.

To cite: Oghalai JS, Bortfeld H, Feldman HM, et al. Cochlear Implants for Deaf Children With Early Developmental Impairment. *Pediatrics.* 2022;149(6):e2021055459

In the United States, 0.1% to 0.3% of newborn infants have sensorineural hearing loss.[1] A strong consensus is that early detection and appropriate treatment is essential to allow children to maximize their developmental skills, particularly in the domains of language and speech. The first step is always amplification through hearing aids, which should be provided before the children reach age 6 months.[2] If children with profound hearing loss fail to develop early skills in language and speech despite hearing aids, they become eligible for cochlear implantation. Cochlear implantation has become the standard of care for children with deafness and no other developmental disorders because of compelling evidence that children with implants are more likely to acquire good speech and language than those without, enabling them to participate without major assistance in general education programs.[3–6]

Children with profound hearing loss may have associated deficits or impairment in other developmental domains, including nonverbal cognitive skills and adaptive function. Heretofore, we refer to the combination of low cognition and adaptive function as early developmental impairment (EDI). Common causes of both hearing loss and EDI include complications of preterm birth, genetic or chromosomal abnormalities, and hypoxic-ischemic encephalopathy.[7] If these children continue to show cognitive impairment and disorders in adaptive behavior into school age, they may later meet criteria for intellectual disability. The optimal therapeutic regimen for children with profound hearing loss and EDI is not straightforward because even children with normal hearing and EDI may not develop age-appropriate spoken communication skills.[8] Many studies of pediatric cochlear implantation in children with additional disabilities have revealed either no effect or limited improvements in speech perception, speech production, and spoken language.[9–12] However, researchers have recently found other benefits to cochlear implantation in this population, including improvements in receptive language, adaptive behavior, and auditory skills.[13–17] Thus, even though a child with severe EDI may have better hearing through a cochlear implant than through hearing aids, they are likely to have minimal improvement in speech and language.[18]

For this reason, some medical insurance programs may not cover cochlear implantation for a child with EDI. For example, the publicly administered California Children's Services program has excluded these children from payment for cochlear implantation, and only recently administrators have agreed to cover implantation for select patients.[19,20] In contrast, the analogous Children's Health Insurance Program in Texas typically covers cochlear implantation for most children with EDI. In both California and Texas, some private insurance plans also do not cover cochlear implantation for children with severe developmental delays. This study was motivated by our impression from providing cochlear implantation to selected patients with EDI that these children also benefited from early cochlear implantation.[21,22] Although authors of many other studies have compared outcomes after cochlear implantation versus hearing aids, the data regarding children with additional needs are sparse.[18] Here, we took advantage of insurance coverage differences to compare the outcomes of deaf children with EDI who underwent cochlear implantation versus those who continued to use hearing aids. We hypothesized that deaf children with EDI would benefit from cochlear implantation in highly meaningful ways not exclusive to auditory and language development.

METHODS

Study Design

A prospective longitudinal cohort study was performed that included all children identified with severe to profound sensorineural hearing loss managed by 2 large pediatric cochlear implant centers: Texas Children's Hospital (Baylor College of Medicine, Houston, TX) and Lucile Packard Children's Hospital (Stanford, CA). Institutional review boards at both institutions approved the protocol, and the study was registered with ClinicalTrials.gov (identifier NCT01256229; start date September 2009; completion date September 2017).

A baseline assessment was completed by clinicians in audiology, speech-language pathology, and neuropsychology. We used validated tests to assess cognition (Mullen Scales of Early Learning [MSEL]), adaptive behavior (Vineland Adaptive Behavior Scales [VABS]), language skills (Preschool Language Scale [PLS]), auditory skills (LittlEARS Auditory Questionnaire [LEAQ]), and stress within the parent-child system (Parenting Stress Index [PSI]). Subject recruitment procedures and assessment measures are described in the Supplemental Information.

All children started with hearing aids. When performed, the cochlear implantation surgery was ~1 month after the baseline assessment. The only exceptions were children younger than the minimum age approved by the US Food and Drug Administration for cochlear implantation (12 months). For these

patients, implantation was performed within the month after the children's first birthday. Follow-up evaluations were scheduled annually using the same assessment protocol.

Definition of EDI

Developmental impairment was defined on the basis of scores on nonverbal cognitive skills and adaptive functioning. To estimate cognition, we used the visual reception domain from the MSEL. To assess adaptive behavior, we used the daily living skills domain from the VABS. The other domains were excluded because of the possibility that results may not reflect actual capability due to either reduced receptive and expressive language or motor delays. Previously, we validated these 2 measures in a patient population with hearing loss and EDI, and we defined meaningful thresholds for EDI guided by standard definitions of intellectual disability.[21,23,24] Children who scored 2 SDs below the mean for their age in the visual reception domain of the MSEL met the criterion for low nonverbal cognitive skills. Children who scored 1 SD below their age in the daily living skills domain of the VABS met the criterion for low adaptive behavior. Normal nonverbal cognitive skills and normal adaptive behavior were defined as being above these thresholds. Thus, children who met our definition of EDI had severe developmental delays.

Statistical Analyses

For comparing the patient characteristics, which were categorical variables (Table 1), we used the Fisher's exact test for proportions. For comparing the developmental outcomes, which were continuous variables (Table 2), we used analysis of variance followed by Tukey's post hoc subgroup comparisons. In both cases, adjusted *P* values were used to account for multiple comparisons. To perform linear fits for each developmental outcome for each cohort (Fig 2), we used least squares regression. To compare developmental trajectories among the 3 cohorts (Table 3), we used hierarchical linear modeling. Testing age and cohort number were used as fixed-effects predictors, and the subject identifier code was used as a random effect predictor. This statistical approach was chosen for these longitudinal data because the resulting models provide estimates for growth rate, which has less error than each individual data point for each child. The method is also advantageous because it is permissive regarding missing data and can account for random variations in follow-up testing age between subjects. For all analyses, we adjusted for simultaneous inference by term, and the threshold for statistical significance was set at $P < .05$. Confidence intervals (CIs) were calculated at 95%. Analyses were conducted in R, and figures were

TABLE 1 Patient Characteristics

	Cohort 1	Cohort 2	Cohort 3	*P*
Site				
Baylor College of Medicine	100 (72)	32 (86)	24 (83)	.16
Stanford University	38 (28)	5 (14)	5 (17)	
Sex				
Female	67 (49)	25 (68)	14 (48)	.11
Male	71 (51)	12 (32)	15 (52)	
Birth				
Preterm	26 (19)	20 (54)	17 (59)	<.001
Full term	103 (75)	15 (41)	10 (34)	
Unknown	9 (7)	2 (5)	2 (7)	
Household income, $K	64 ± 29	72 ± 30	65 ± 32	.34
Rural-urban continuum code				
1	100 (72)	33 (89)	28 (97)	.02
2	23 (17)	3 (8)	1 (3)	
3–8	15 (11)	1 (3)	0 (0)	
Ethnicity				
Not Hispanic	69 (50)	25 (68)	15 (52)	.009
Hispanic	67 (49)	12 (32)	10 (34)	
Unknown	2 (1)	0 (0)	4 (14)	
Race				
White	113 (82)	28 (76)	17 (59)	.14
American Indian	7 (5)	2 (5)	1 (3)	
Asian	5 (4)	3 (8)	3 (10)	
Pacific Islander	1 (1)	0 (0)	0 (0)	
Black	5 (4)	3 (8)	3 (10)	
Mixed	1 (1)	0 (0)	1 (3)	
Unknown	6 (4)	1 (3)	4 (14)	
Preferred language				
English	93 (67)	32 (86)	20 (69)	.006
Spanish	28 (20)	2 (5)	3 (10)	
Bilingual	17 (12)	3 (8)	3 (10)	
Unknown	0 (0)	0 (0)	3 (10)	
Mother's education				
No high school diploma	10 (7)	2 (5)	1 (3)	.93
High school diploma	26 (19)	7 (19)	7 (24)	
College degree	25 (18)	10 (27)	4 (14)	
Advanced degree	6 (4)	2 (5)	1 (3)	
Unknown	71 (51)	16 (43)	16 (55)	

Data are presented as No. (%) or mean ± SD. Cohort 1, normal cognitive skills and adaptive behavior, underwent cochlear implantation ($n = 138$); cohort 2, EDI, underwent cochlear implantation ($n = 37$); and cohort 3, EDI, hearing aids only ($n = 29$). $K, thousands of dollars.

TABLE 2 Developmental Outcomes

	Cohort, Mean ± SD			Adjusted *P* (95% CI)[a]		
	1	2	3	Cohorts 2–1	Cohorts 3–1	Cohorts 3–2
Evaluations						
No. (range)	2.43 ± 1.45 (1 to 9)	2.27 ± 1.04 (1 to 7)	1.38 ± 0.73 (1 to 4)	.77 (−0.7 to 0.4)	<.001 (−1.7 to −0.4)	.02 (−1.7 to −0.1)
Chronological age						
Initial, mo	19 ± 10	24 ± 16	37 ± 19	.04 (0.3 to 11.6)	<.001 (12.0 to 24.5)	<.001 (4.7 to 19.8)
Final, mo	39 ± 16	51 ± 21	57 ± 7	.04 (0.6 to 24.4)	.20 (−7.3 to 44.3)	.85 (−20.6 to 32.7)
MSEL						
Initial developmental age, mo	13 ± 6	8 ± 5	7 ± 6	<.001 (−8.0 to −2.8)	<.001 (−9.1 to −3.5)	.81 (−4.3 to 2.5)
Final developmental age, mo	18 ± 10	11 ± 7	8 ± 8	<.001 (−10.6 to −2.4)	<.001 (−14.5 to −5.4)	.31 (−8.9 to 2.1)
VABS						
Initial developmental age, mo	19 ± 13	10 ± 7	12 ± 10	.001 (−14.9 to −3.2)	.13 (−13.7 to 1.3)	.72 (−5.8 to 11.6)
Final developmental age, mo	27 ± 20	18 ± 22	12 ± 10	.09 (−18.0 to 1.0)	.02 (−26.7 to −1.7)	.61 (−20.2 to 8.7)
PLS						
Initial raw score	28 ± 13	22 ± 8	22 ± 15	.06 (−13.1 to 0.3)	.13 (−13.8 to 1.4)	1.00 (−9.1 to 9.5)
Final raw score	50 ± 30	40 ± 24	27 ± 17	.18 (−23.7 to 3.3)	.003 (−39.6 to −6.5)	.27 (−32.5 to 6.8)
LEAQ						
Initial raw score	7 ± 9	7 ± 7	2 ± 3	.97 (−4.8 to 5.9)	.28 (−11.4 to 2.5)	.30 (−13.1 to 3.0)
Final raw score	15 ± 13	13 ± 10	2 ± 3	.76 (−8.6 to 4.7)	.002 (−21.9 to −4.3)	.02 (−21.1 to −1.2)
PSI						
Initial total score	196 ± 33	226 ± 45	252 ± 42	<.001 (11.4 to 48.8)	<.001 (31.4 to 80.5)	.08 (−2.4 to 54.1)
Final total score	200 ± 41	227 ± 46	252 ± 41	.007 (6.2 to 47.7)	<.001 (24.1 to 79.2)	.16 (−6.8 to 56.2)

Cohort 1, normal cognitive skills and adaptive behavior, underwent cochlear implantation, not lost to follow-up (n = 99); cohort 2, EDI, underwent cochlear implantation, not lost to follow-up (n = 32); cohort 3, EDI, hearing aids only, not lost to follow-up (n = 29).

[a] Tukey post hoc subgroup comparisons.

produced using the package ggplot2.[25,26]

RESULTS

Stratification of Cohorts

From the 297 children enrolled in this longitudinal study, 3 subgroups were formed (Fig 1). Cohort 1 included children who received a cochlear implant and had normal cognitive skills and normal adaptive behavior (n = 138). Cohort 2 included children who received a cochlear implant and who met criteria for EDI based on both low cognitive skills and low adaptive behavior scores (n = 37). Cohort 3 included children who continued to be treated with hearing aids and who also met criteria for EDI based on both low cognitive skills and low adaptive behavior scores (n = 29). The remaining 93 children who had low cognitive skills or low adaptive behavior, but not both, were excluded from further analysis. Although these 93 children are worthy of study, the did not meet the criteria for testing the hypothesis proposed in this manuscript.

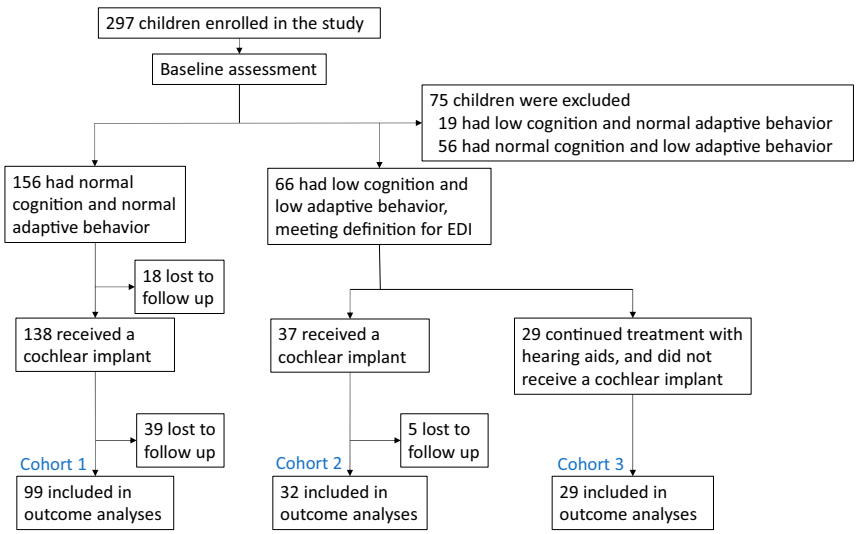

FIGURE 1
Enrollment, cohort assignment, and developmental outcome assessment.

We found no differences among the cohorts on the basis of study site,

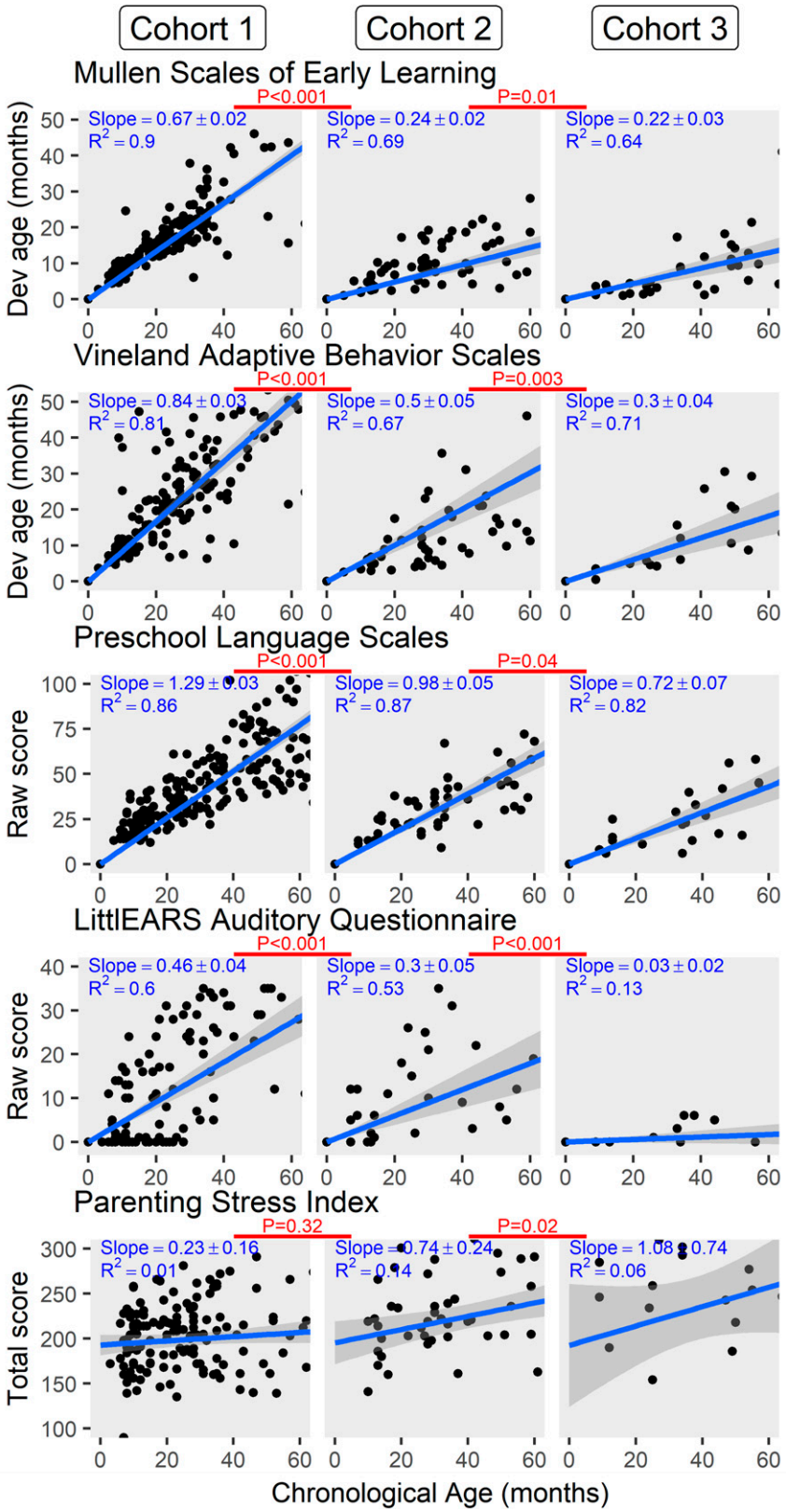

FIGURE 2

Developmental trajectories. Cohort 1 included children with normal cognitive skills and normal adaptive behavior who underwent cochlear implantation. Cohort 2 included children with early developmental impairment who underwent cochlear implantation. Cohort 3 included children with early

sex, household income, race, or mother's education level (Table 1). As expected, there was a higher rate of preterm births in both cohort 2 and cohort 3 compared with cohort 1. There were also differences among the cohorts in the rates of living in an urban or rural environment, ethnicity, and preferred language, with higher rates of urban Hispanic, Spanish-speaking families in cohort 1. Because we had no hypotheses about the interaction of these demographics with our outcome data, we assumed that the significant *P* values represented random statistical variability and did not include any demographic data in further analyses.

Developmental Outcomes

Baseline data were collected at the time of enrollment, with repeated evaluations performed approximately annually over the next 1 to 5 years. The final evaluation was defined as the last data point we collected before the grant ended. During the study, 39 of the children in cohort 1 and 5 of the children in cohort 2 were lost to follow-up, typically because their family moved out of town. Interestingly, none of the children in cohort 3 were lost to follow-up. The rates of attrition were acceptable for a study of this duration. Children in cohorts 1 and 2 had more follow-up evaluations than children in cohort 3 (Table 2). This difference is not surprising because insurers who were willing to cover cochlear implantation were also willing to cover repeated neurocognitive examinations.

At baseline, cohort 1 was the youngest and cohort 3 was the oldest. There were no differences in cognition, adaptive behavior, language, auditory skills, or stress within the parent-child system between cohorts 2 and 3. Because

TABLE 3 Hierarchical Linear Modeling of Predictors

	Estimated Coefficient	SE	95% CI	*P*
MSEL				
Intercept	0.384	0.274	−0.153 to 0.921	.16
Age	0.306	0.032	0.244 to 0.368	<.001***
Cohort 1–2	−0.096	0.31	−0.704 to 0.511	.76
Cohort 2–3	−0.378	0.428	−1.217 to 0.462	.38
Age:cohort 1–2	0.403	0.037	0.332 to 0.475	<.001***
Age:cohort 2–3	−0.119	0.048	−0.213 to −0.026	.01*
VABS				
Intercept	−1.72	1.489	−4.639 to 1.198	.25
Age	0.513	0.039	0.437 to 0.589	<.001***
Cohort 1–2	3.28	1.683	−0.018 to 6.579	.05
Cohort 2–3	2.054	2.385	−2.621 to 6.729	.39
Age:cohort 1–2	0.281	0.049	0.186 to 0.377	<.001***
Age:cohort 2–3	−0.219	0.074	−0.365 to −0.073	.003**
PLS				
Intercept	1.369	2.241	−3.023 to 5.761	.54
Age	0.935	0.068	0.801 to 1.068	<.001***
Cohort 1–2	2.718	2.501	−2.183 to 7.619	.28
Cohort 2–3	−0.955	3.479	−7.773 to 5.863	.78
Age:cohort 1–2	0.263	0.075	0.116 to 0.410	<.001***
Age:cohort 2–3	−0.234	0.115	−0.460 to −0.008	.04*
LEAQ				
Intercept	1.514	1.02	−0.486 to 3.513	.14
Age	0.268	0.041	0.188 to 0.347	<.001***
Cohort 1–2	−0.668	1.15	−2.923 to 1.587	.56
Cohort 2–3	−1.283	1.588	−4.396 to 1.830	.42
Age:cohort 1–2	0.166	0.049	0.071 to 0.261	<.001***
Age:cohort 2–3	−0.243	0.061	−0.363 to −0.124	<.001***
PSI				
Intercept	61.561	10.622	40.742 to 82.380	<.001***
Age	3.005	0.283	2.450 to 3.561	<.001***
Cohort 1–2	−7.356	11.998	−30.872 to 16.160	.54
Cohort 2–3	−33.579	17.142	−67.177 to 0.019	.05
Age:cohort 1–2	0.357	0.356	−0.341 to 1.055	.32
Age:cohort 2–3	1.328	0.572	0.208 to 2.448	.02*

Cohort 1, normal cognitive skills and adaptive behavior, underwent cochlear implantation, not lost to follow-up (n = 99); cohort 2, EDI, underwent cochlear implantation, not lost to follow-up (n = 32); cohort 3, EDI, hearing aids only, not lost to follow-up (n = 29); cohort 1–2 and cohort 2–3, cohort 2 is the baseline, cohorts 1 and cohort 3 compared against it; and age:cohort 1–2 and age:cohort 2–3, interaction between age and cohorts 1–2 or cohorts 2–3. * $P < .05$, ** $P < .01$, *** $P < .001$.

the children were all deaf, cohort 1 had language and auditory skills like cohorts 2 and 3. However, cohort 1 had less stress within the parent-child system relative to the 2 other cohorts. By the time of final testing, average developmental scores were highest for cohort 1 and lowest for cohort 3.

Developmental Trajectories

Next, we compared the developmental trajectory for each cohort by calculating a linear fit for all evaluations from birth, where all developmental ages and raw scores would be 0, with the final evaluation (Fig 2). However, to compare the growth trajectories, we did not simply compare these linear fits. Instead, we performed hierarchical linear modeling (Table 3). This analysis permitted us to control for age of testing, differences in the number of follow-up evaluations, and random differences between individuals. The estimated coefficient values reflect the differences in the slope of either cohort 1 or 3 compared with cohort 2. Positive values mean that the slope is steeper than for cohort 2; negative values mean that the slope is shallower.

To analyze growth in cognitive skills and adaptive behavior, we plotted the developmental ages for the MSEL and VABS versus chronological age. A perfectly normal developmental trajectory would have a slope of 1. Cohort 1 performed best, with near-normal trajectories in intelligence and adaptive behavior. In addition, the R^2 values were high (>0.8), demonstrating that most children in this cohort fit our regression model. In contrast, the developmental trajectories of cohort 2 were shallower and more variable, and cohort 3 underperformed cohort 2.

To analyze language and auditory skills outcomes, we plotted raw PLS and LEAQ scores versus chronological age. Cohort 1 had the best outcomes and high R^2 values, demonstrating that raw scores and chronological age were strongly correlated. In contrast, the developmental trajectories for cohort 2 were not as steep as those in cohort 1, and cohort 3 underperformed cohort 2.

To assess for changes in stress within the parent-child system, we plotted the PSI score versus chronological age. There were no differences between cohorts 1 and 2, but cohort 3 demonstrated more stress in the parent-child system over time.

developmental impairment who did not receive a cochlear implant and continued to use appropriately fitted hearing aids. Each dot is the test result for 1 child, the blue lines are the linear fits, and the gray-shaded region is the 95% CI. The slopes and R^2 of the fit lines are shown in the upper left quadrant of each analysis. The *P* values come from the interaction term between age and either cohort 1 versus 2 (age:cohort 1–2 in Table 3) or cohort 2 versus 3 (age:cohort 2–3 in Table 3) in the hierarchical linear modeling analysis. Dev, developmental.

Hierarchical Linear Modeling Analysis

To test our hypothesis that cochlear implantation was better than hearing aids for children with EDI, we compared developmental trajectories between cohorts 2 and 3. We found that cohort 3 did worse than cohort 2 in all assessments, including cognition (95% CI, −0.213 to −0.026; $P = .01$), adaptive behavior (95% CI, −0.365 to −0.073; $P = .003$), language (95% CI, −0.460 to −0.008; $P = .04$), and auditory skills (95% CI, −0.363 to −0.124; $P < .001$). In addition, parental stress was higher in cohort 3 (95% CI, 0.208 to 2.448; $P = .02$), indicating larger increases in stress within the parent-child system.

To verify that site did not affect these findings, we repeated the hierarchical linear modeling analysis, including site as a random effect. This produced only slight changes in the values given in Table 3, and the statistical significances for every finding did not change.

DISCUSSION

In this study, we provide the most compelling data available that cochlear implantation in children who are born deaf and who have EDI is associated with better developmental outcomes and faster rates of development in multiple domains than continued use of hearing aids. Our experience in assessing children with EDI at 2 large pediatric hospitals reveals that cochlear implantation is associated with improved scores on measures of nonverbal cognitive skills, adaptive functioning, language, auditory skills, and reduced stress in the parent-child system compared with treatment with hearing aids alone. On the basis of our data, we argue that cochlear implantation should be considered for all children with severe to profound hearing loss, regardless of their cognitive skills and adaptive behavior. Because many studies in children who are deaf with normal development have revealed that cochlear implantation at a younger age is associated with better developmental outcomes[4,27–30] than delaying implantation, we argue that this decision should be made as early as possible before a critical window for rapid development has passed.

It is unfortunate and inequitable that, in our society, children with EDI may be excluded from receiving potentially beneficial interventions because their developmental outcomes are below that of children who function in the normal range in terms of cognition and adaptive behaviors. Our study reveals that this problem is seen for children who need cochlear implantation. Besides the obvious problem that EDI has been used as a reason to deny insurance coverage, a more complex hurdle to overcome is that providers may also fail to refer these patients for cochlear implant evaluation on the basis of the notion that their cognitive delays render them ineligible for insurance coverage for the procedure. For example, we found that children with EDI who continued with hearing aid use were older and had lower auditory skills at initial evaluation than children who went on to receive a cochlear implant. This finding might be explained by differences in pediatricians' approaches to referring these children. Although the types of developmental testing paradigms in use today are wide ranging and highly valid, it remains difficult to accurately predict the ultimate cognitive outcome of any individual child from tests performed when they are infants and toddlers, like those included in this study. To give all children the best opportunity to fully develop their maximum potential, cochlear implantation should be carefully considered regardless of the presence of severe developmental delays.

We recognize the several limitations of this study. Most importantly, a randomized controlled trial was not feasible given the current state of clinical practice. When we initially proposed this study, we planned a prospective, randomized controlled trial of children with EDI in which some children would receive a cochlear implant and some would continue with hearing aid use. In the early 2000s, pediatric cochlear implantation in children with normal cognition was still considered a novel procedure, and it was rarely performed in any child with EDI. However, by the time we were awarded National Institutes of Health funding for this trial, nearly all children with normal cognition and most children with EDI were undergoing cochlear implantation. This change occurred because the evidence for the benefit of cochlear implantation in children with normal cognition had become clear, and parents and medical professionals considered it clinically unwise and unethical to withhold the "best" treatment of the child with EDI. However, equipoise had not been lost, as the question of whether cochlear implantation is beneficial to children who may not have the cognitive ability to use the auditory information provided by the cochlear implant remained unclear.

We attempted to overcome our inability to perform a controlled clinical trial by capitalizing on differences in insurance coverage to create randomization. However, we could not determine with certainty the underlying reasons for the decisions made by families of

children with deafness and EDI in opting for cochlear implants or hearing aids. For most of the families in cohort 3, the decision regarding cochlear implantation was taken out of their hands by the reimbursement agency. Nevertheless, it was impossible to quantify how differing eligibility policies affected the implantation rate because neither pediatric cochlear implant team determined the care plan for any patient solely on the basis of insurance coverage. Instead, according to best practice, the teams described the risks, benefits, and typical outcomes for hearing aids and for cochlear implantation. The parents then decided how they wished to proceed. Interestingly, none of the families whose child was denied by insurance was adamant about receiving a cochlear implant. Over time and repeated visits, most parents ultimately determined for themselves that cochlear implantation would not be best for their child. In contrast, parents who did not have this eligibility issue tended to make the decision to proceed with implantation.

Another caveat is that these children were only followed for an average of 2 years after their initial evaluation. We know that in children with normal intelligence, cochlear implantation produces better long-term outcomes than hearing aids.[31] Although we found that children with EDI treated with hearing aids have a slower developmental trajectory than those treated with cochlear implantation, it is possible that both cohorts will ultimately end up with similar developmental outcomes.

To conclude, our data are supportive of the value of cochlear implantation in children with multiple disabilities/developmental delays and provide an argument against insurance plans requiring that certain developmental milestones be met prior to authorization. Developmental impairments will adversely affect a child's outcome after cochlear implantation because the combination of hearing loss and EDI synergistically affect other developmental outcomes, with the loss of auditory input creating a cascade of higher-order neurocognitive deficits.[32] This outcome should be discussed carefully with the parents to ensure that their expectations are appropriate. However, EDI should not be the deciding factor alone. Every child is an individual and has unique health and developmental limitations, together with unique family, home, and school environments. All these factors should be considered during the process of cochlear implantation candidacy.

ACKNOWLEDGMENTS

We thank E. O'Brian Smith, Lane Strathearn, Ann Stark, Jody Haymond, Sue Caudle, Dian Baker, Ross Tonini, Homer Abaya, Jody Winzelberg, Barbara Bentley, Ryan Lucash, Laurie Eisenberg, and Karen Johnson for their contributions. Deidentified individual subject data (including data dictionaries) will be made available, in addition to study protocols, the statistical analysis plan, and the informed consent form. The data will be made available upon publication to researchers who provide a methodologically sound proposal for use in achieving the goals of the approved proposal. Proposals should be submitted to oghalai@usc.edu.

ABBREVIATIONSS

CI: confidence interval
EDI: early developmental impairment
LEAQ: LittlEARS Auditory Questionnaire
MSEL: Mullen Scales of Early Learning
PLS: Preschool Language Scale
PSI: Parenting Stress Index
VABS: Vineland Adaptive Behavior Scales

Address correspondence to John S. Oghalai, MD, Caruso Department of Otolaryngology–Head and Neck Surgery, University of Southern California, Healthcare Center 4, 1450 San Pablo St, Ste 5800, Los Angeles, CA 90033. E-mail: oghalai@usc.edu

PEDIATRICS (ISSN Numbers: Print, 0031-4005; Online, 1098-4275).

FUNDING: Funded by National Institutes of Health (NIH) grant R01 DC010075. The NIH had no role in the design and conduct of this study.

CONFLICT OF INTEREST DISCLOSURES: The authors have indicated they have no potential conflicts of interest to disclose.

REFERENCES

1. Centers for Disease Control and Prevention. Research and tracking of hearing loss in children: determining how many children have hearing loss. Available at: https://www.cdc.gov/ncbddd/hearingloss/research.html. Accessed December 13, 2021
2. The Joint Committee on Infant Hearing. Year 2019 Position Statement: Principles and Guidelines for Early Hearing

Detection and Intervention Programs. *J Early Hear Detect Interv.* 2019;4(2):1–44

3. Fink NE, Wang NY, Visaya J, et al. Childhood Development after Cochlear Implantation (CDaCI) study: design and baseline characteristics. *Cochlear Implant Int.* 2007;8(2):92–116
4. Niparko JK, Tobey EA, Thal DJ, et al. Spoken language development in children following cochlear implantation. *JAMA.* 2010;303(15):1498–1506
5. Svirsky MA, Robbins AM, Kirk KI, Pisoni DB, Miyamoto RT. Language development in profoundly deaf children with cochlear implants. *Psychol Sci.* 2000;11(2):153–158
6. Cejas I, Mitchell CM, Hoffman M, Quittner AL; CDaCI Investigative Team. Comparisons of IQ in children with and without cochlear implants: longitudinal findings and associations with language. *Ear Hear.* 2018;39(6):1187–1198
7. Gallaudet Research Institute. Annual Survey of Deaf and Hard of Hearing Children & Youth: 2011-12 Regional and National Summary. Available at: https://research.gallaudet.edu/Demographics/2012_National_Summary.pdf. Accessed June 29 , 2021
8. Abbeduto L, Kover ST, McDuffie A. Studying the language development of children with intellectual disabilities. In: Hoff E, ed. *Research Methods in Child Language: A Practical Guide.* 1st ed. Hoboken, NJ: Wiley-Blackwell; 2012: 330–346
9. Johnson K, Wiley S, Meinzen-Derr J. Cochlear implantation in children with multiple disabilities. In: Eisenberg LS, ed. *Clinical Management of Children With Cochlear Implants.* 2nd ed. San Diego, CA: Plural Publishing; 2017: 737–789
10. Eze N, Ofo E, Jiang D, O'Connor AF. Systematic review of cochlear implantation in children with developmental disability. *Otol Neurotol.* 2013;34(8):1385–1393
11. Waltzman SB, Scalchunes V, Cohen NL. Performance of multiply handicapped children using cochlear implants. *Am J Otol.* 2000;21(3):329–335
12. Cupples L, Ching TYC, Leigh G, et al. Language development in deaf or hard-of-hearing children with additional disabilities: type matters! *J Intellect Disabil Res.* 2018;62(6):532–543
13. Cruz I, Vicaria I, Wang NY, Niparko J, Quittner AL; CDaCI Investigative Team. Language and behavioral outcomes in children with developmental disabilities using cochlear implants. *Otol Neurotol.* 2012;33(5):751–760
14. Hang AX, Kim GG, Zdanski CJ. Cochlear implantation in unique pediatric populations. *Curr Opin Otolaryngol Head Neck Surg.* 2012;20(6):507–517
15. Corrales CE, Oghalai JS. Cochlear implant considerations in children with additional disabilities. *Curr Otorhinolaryngol Rep.* 2013;1(2):61–68
16. Nasralla HR, Montefusco AM, Hoshino ACH, et al. Benefit of cochlear implantation in children with multiple-handicaps: parent's perspective. *Int Arch Otorhinolaryngol.* 2018;22(4):415–427
17. Beer J, Harris MS, Kronenberger WG, Holt RF, Pisoni DB. Auditory skills, language development, and adaptive behavior of children with cochlear implants and additional disabilities. *Int J Audiol.* 2012;51(6):491–498
18. Killan CF, Hoare DJ, Katiri R, et al. A scoping review of studies comparing outcomes for children with severe hearing loss using hearing aids to children with cochlear implants. *Ear Hear.* 2021;43(2):290–304
19. State of California–Health and Human Services Agency, Department of Health Care Services. Cochlear implant updated candidacy criteria and authorization procedure (NL 12-0818). Available at: https://www.dhcs.ca.gov/services/ccs/Documents/CCS.NL.12-0818.Cochlear.Implant.pdf. Accessed September 7, 2021
20. California Children's Services. Child Cochlear Implant Profile (CHIP) rating (NL 12-0818, attachment 3). Available at: https://www.dhcs.ca.gov/services/ccs/Documents/Attach3Form.pdf. Accessed September 7, 2021
21. Oghalai JS, Caudle SE, Bentley B, et al. Cognitive outcomes and familial stress after cochlear implantation in deaf children with and without developmental delays. *Otol Neurotol.* 2012;33(6):947–956
22. Pierson SK, Caudle SE, Krull KR, Haymond J, Tonini R, Oghalai JS. Cognition in children with sensorineural hearing loss: etiologic considerations. *Laryngoscope.* 2007;117(9):1661–1665
23. Caudle SE, Katzenstein JM, Oghalai JS, Lin J, Caudle DD. Nonverbal cognitive development in children with cochlear implants: relationship between the Mullen Scales of Early Learning and later performance on the Leiter International Performance Scales-Revised. *Assessment.* 2014;21(1):119–128
24. American Association on Intellectual and Developmental Disability. Defining criteria for intellectual disability. Available at: https://www.aaidd.org/intellectual-disability/definition. Accessed August 2, 2021
25. The R Foundation. The R Project for Statistical Computing. Available at: https://www.r-project.org. Accessed March 2, 2022
26. Wickham H. *Ggplot2: Elegant Graphics for Data Analysis.* New York, NY: Springer-Verlag; 2016. Available at: https://ggplot2.tidyverse.org. Accessed March 2, 2022
27. Dettman SJ, Pinder D, Briggs RJS, Dowell RC, Leigh JR. Communication development in children who receive the cochlear implant younger than 12 months: risks versus benefits. *Ear Hear.* 2007;28(2 suppl):11S–18S
28. Chweya CM, May MM, DeJong MD, et al. Language and audiological outcomes among infants implanted before 9 and 12 months of age versus older children: a continuum of benefit associated with cochlear implantation at successively younger ages. *Otol Neurotol.* 2021;42(5):686–693
29. Geers AE, Strube MJ, Tobey EA, Pisoni DB, Moog JS. Epilogue: factors contributing to long-term outcomes of cochlear implantation in early childhood. *Ear Hear.* 2011;32(1 suppl):84S–92S
30. Geers AE, Mitchell CM, Warner-Czyz A, Wang NY, Eisenberg LS; CDaCI Investigative Team. Early sign language exposure and cochlear implantation benefits. *Pediatrics.* 2017;140(1):e20163489
31. Sharma SD, Cushing SL, Papsin BC, Gordon KA. Hearing and speech benefits of cochlear implantation in children: A review of the literature. *Int J Pediatr Otorhinolaryngol.* 2020;133:109984

32. Kral A, Kronenberger WG, Pisoni DB, O'Donoghue GM. Neurocognitive factors in sensory restoration of early deafness: a connectome model. *Lancet Neurol.* 2016;15(6):610–621