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Cognition in Children With Sensorineural Hearing Loss: Etiologic Considerations

Suzanne K. Pierson, PhD, **Susan E. Caudle, PhD**, **Kevin R. Krull, PhD**, **Jody Haymond, MD**, **Ross Tonini, AuD**, and **John S. Oghalai, MD**
Learning Support Center for Child Psychology (S.K.P., S.E.C., K.R.K.), Texas Children's Hospital, Houston, Texas, U.S.A.; Allied Health Sciences and Department of Pediatrics (S.E.C., K.R.K.), Baylor College of Medicine, Houston, Texas, U.S.A.; Texas Children's Hearing Center (J.H., R.T., J.S.O.), Houston, Texas, U.S.A.; the Department of Otorhinolaryngology (J.H., R.T.), Baylor College of Medicine, Houston, Texas U.S.A.; and the Bobby R. Alford Department of Otolaryngology, Head and Neck Surgery (J.S.O.), Baylor College of Medicine, Houston, Texas U.S.A.

Abstract

Objectives—A considerable amount of literature has documented the impact of hearing impairment on spoken language skills in deaf children referred for cochlear implantation. Critical areas of neurocognitive development in the acquisition of visual (manual) language also appear to be impacted, although the evidence is less robust. The present study focused on the development of visual and fine motor skills in a sample of preschool-age children diagnosed with sensorineural hearing loss with no known neurologic conditions (n = 36).

Study Design—Analysis of data collected as part of a standardized screening process for cochlear implantation at an academic medical center.

Method—Children underwent a standardized neuropsychological assessment battery. Children were classified into three groups based on the etiology of their deafness (Connexin = 15, Structural Malformation = 11, and Unknown = 10).

Results/Conclusions—Correlational analyses replicated previous research on the reduction in visual reception and fine motor skills as deaf children age. Children with genetic (Connexin) etiology exhibited a significant reduction in fine motor skills with age, whereas those with an etiology of Structural Abnormality exhibited a significant reduction in visual reception skills with age. Results of planned comparisons conducted as part of a multivariate analysis of variance (Skill × Group) indicated that the Connexin group was significantly better than the Unknown group with regard to fine motor skills. Implications for these findings and future studies are discussed.

Keywords

Cognitive development; fine motor skills; intelligence; sensorineural hearing loss; cochlear implant

Send correspondence to Dr. Susan E. Caudle, Texas Children's Hospital, 6621 Fannin Street, CC1630.00, Houston, TX 77030-2399. secaudle@texaschildrenshospital.org.

INTRODUCTION

The neurocognitive functioning of children diagnosed with sensorineural hearing loss (HL) has been investigated by multiple studies. An increased interest in this area has occurred since the 1990 Food and Drug Administration approval of cochlear implantation in children ages 2 through 17, which allowed investigation of the impact of a return of auditory stimulation on children's development. Investigation of neurocognitive functioning in deaf children is considered an important part of candidacy for cochlear implantation to better inform postimplant status.

There are several known causes of deafness in humans, including genetic, prematurity, structural abnormalities, diseases affecting the ear, ototoxic drugs, and physical trauma. Knowledge regarding the etiology of a child's deafness is important for several reasons. First, such information helps to predict whether the level of HL will remain stable or progressively worsen over time. Second, ascertaining the nature of damage to the hearing system may help determine the extent a cochlear implant or hearing aid may facilitate a return of auditory stimulation. Third, should the etiology be genetic in nature, the HL may be a symptom of a larger syndrome. Determining whether the etiology is genetic is also important information for family planning.

Estimates indicate that approximately half of all childhood deafness is of hereditary causes, of which up to 30% is syndromal.[1] In syndromal deafness, the HL may be present at birth or may develop in childhood. In deafness syndromes, the HL may be a fundamental part of the syndrome, or deafness may be one of many factors included in the syndrome. Often, syndromal deafness involves additional disabilities, such as progressive blindness as seen in Usher syndrome or heart problems as seen in Jervell and Lange-Nielson syndromes. In nonsyndromic deafness, however, the only obvious medical problem is a loss of hearing. Mutations in the Connexin 26 gene (on chromosome 13) are the most common genetic cause of deafness in children and are thought to be responsible for up to half of all recessive nonsyndromic HL.[2] Unlike other forms of congenital deafness, Connexin-related deafness has no known comorbidity or associated neurologic problems.[3] In addition, there is evidence of generally intact cognitive functioning in comparison with other etiologies of deafness.[4]

Additional causes of deafness in children include prematurity, with sensorineural HL diagnosed approximately 5% of the time in these children. There are many known diseases that can affect the developing auditory system, including viruses (e.g., rubella, herpes, cytomegalovirus, toxoplasmosis) and infections (e.g., meningitis, measles, mumps). Some medications have been found to cause irreversible damage to the ear, most notably the aminoglycosides. Structural abnormalities of the hearing system, such as malformation of the cochlea, are also an established cause of deafness. Such structural abnormalities can occur in the presence of normal brain structure and without any resulting neurologic impairment.

The negative impact of sensorineural HL on spoken language development is clear and not surprising given the evidence on the importance of auditory stimulation for communication skills.[5–7] The impact of auditory deprivation on the development of nonverbal skills in children has been less studied. Nonverbal skills, including visual and fine motor functioning, are especially important in this population given their frequent reliance on sign language for communication. Although proposed via the "crowding hypothesis," in which compromise of one modality (e.g., spoken language) results in accentuation of a separate modality (e.g., visual), there is no clear evidence supporting the idea of visual/visual-perception enhancement in the deaf population.[8] For example, in a study of prelingually deaf children who were not fluent or consistently exposed to sign language and normal hearing same-age

peers, there were no group differences on measures of visual-spatial skill.[9] Prelingually deafened children free of neurologic impairment have been found to perform significantly more poorly than same-age normal hearing children on measures of nonverbal reasoning, planning, and visual perception.[10] In a study of 18 prelingually deafened preschool children who underwent neuropsychological assessment, correlational analyses indicated that visual reception scores declined with age.[11] Many factors, however, complicate the observed development of visual-spatial skills in deaf children, including their exposure to and consistent use of sign language, as well as the severity and age of HL.

With regard to motor development in children diagnosed with HL, there is evidence of reduced gross motor skills. In a study comparing deaf children with no neurologic impairment with normal-hearing peers, deaf children were found to have reduced balance and complex motor movements.[10] In regard to fine motor skills, Kutz et al.[11] found a trend for a decline with age. This study is one of few studies of deaf children that investigated fine motor skill development.

Few studies have considered the etiology of deafness when investigating neurocognitive functioning of deaf children. In part, this is because of relatively recent advances in medical technology, including improved genetic testing and brain imaging capabilities. As mentioned, etiology that is genetic in nature can include either syndromic or nonsyndromic forms of deafness. There is evidence of many forms of syndromic deafness also being linked to neurocognitive deficits, with the deficits varying with the specific syndrome.[12] Other causes of deafness, such as prematurity and herpes viral infection, are well known to have associated neurocognitive deficits involving such areas as attention and memory, regardless of hearing status.[13,14]

The present study investigated the neurocognitive functioning, primarily the relatively understudied areas of visual and fine motor skills, of children diagnosed with sensorineural HL. The study focused on both visual and fine motor skills because of their importance for communication via sign language. As such, we were interested in replicating our previous results with a larger sample size and determining the impact of etiology of HL on visual and fine motor skills in cochlear implant candidates.[11] As earlier outlined, there are several known causes of deafness. We chose to investigate visual reception and fine motor skills in three etiologic groups, Connexin, Structural Abnormality, and Unknown, in an effort to identify neurocognitive deficits present in children without any known medical conditions or neurologic impairment other than HL.

Thus, the three primary research questions were as follows. 1) Among deaf toddlers/preschoolers, does a significant decline in visual reception or fine motor skills occur with advancing age? 2) If there is a significant decline in visual reception or fine motor skills with age, does this relationship hold true regardless of the etiology of deafness? 3) What, if any, etiologic group differences are there with regard to visual reception and fine motor skills?

METHODS

Participants and Procedure

Thirty-six children (20 girls and 16 boys) with profound, bilateral sensorineural HL underwent neuropsychological testing as part of their candidacy for cochlear implantation. The average age of the children at the time of testing was 17.6 (SD 7.3) months, with the youngest being 7 months and the oldest being 33 months old. The average age of diagnosis of HL was 4.2 (SD 6.4) months, with half (51.3%) diagnosed as deaf at the time of their newborn hearing screening. The mean maternal educational level was 13.7 (SD 3.5) years,

and the mean paternal educational level was 15.1 (SD 4.2) years. No children were considered fluent in sign language or had evidence of neurologic impairment. All children, as part of their candidacy for cochlear implantation, underwent the following medical testing in an effort to identify the etiology of their deafness: thyroid, renal, liver, and immunologic function tests, assessments for toxoplasmosis and cytomegalovirus, genetic testing, magnetic resonance imaging, and computed tomography of the temporal bone. For the current study, the children were divided into three groups based on the etiology of their deafness: Connexin (n = 15), Structural Abnormality (n = 11), and Unknown (n = 10). Children within the Connexin group (40% female) had been identified with genetic etiology consistent with mutations of Connexin 26 or a Connexin variant. Children grouped into the Structural Abnormality group (72% female) were those who were identified via magnetic resonance imaging/computed tomography as having normal brain imaging but temporal bone abnormality, bilateral cochlear malformation, or malformation of the internal auditory canals. Children were grouped into the Unknown group (60% female) if all other known etiologies, including genetic, structural abnormality, viral, infectious, prematurity, and exposure to ototoxic drugs, had been ruled out. There were no significant group differences with regard to subject age, handedness (if established), or parental education level. Children with medical histories of prematurity, viruses and infections known to cause deafness, as well as children with genetic etiologies other than those involving a Connexin mutation were excluded. Four of the children in the Connexin group and one subject from each of the Structural Abnormality and Unknown groups were also included in our previous study.[11]

All children who were considered to be cochlear implant candidates were referred to the Learning Support Center–Child Neuropsychology Program at Texas Children's Hospital (Houston, TX) for neuropsychological testing. The evaluation included parent interview and standardized administration of the Mullen Scales of Early Learning by an examiner experienced in working with children with sensorineural HL. At least one of the first three authors was present during all portions of the evaluation.

Materials

All participants were administered the Mullen Scales of Early Learning as a measure of cognitive/developmental functioning.[15] It comprises five scales, including Gross Motor, Fine Motor (FM), Visual Reception (VR), Receptive Language, and Expressive Language. The latter four scales make up the Early Learning Composite (ELC). The ELC has been found to be a valid measure of global cognitive ability.[15] The Mullen is standardized from birth to age 68 months (33 mo for the Gross Motor scale). The raw score from each of the scales is transcribed into a *t* score that is a standardized score age-corrected for the child. The mean normative *t* score for each scale is 50 with an SD of 10. Performance was considered to be average if the *t* score fell between 43 and 56. The main focus of the current study was the FM and VR scales, and, thus, the ELC was not considered.

RESULTS

Based on correlational analyses for the VR and FM scales with age, a significant decline occurred for the overall sample in both VR skills ($r = -.48$, $P < .01$) and FM skills ($r = -.37$, $P < .05$) with advancing age. The significant decline in VR with age is consistent with our previous findings. Furthermore, the trend for a decline in fine motor skills with age found by Kutz et al.[11] was found to be a significant relationship in the present study.

To further investigate the previously identified relationship between VR and FM with age, correlational analyses were conducted individually for each of the three etiologic groups by age. Results are outlined in Table I. It can be seen that there is a significant decline in FM skills with age for children with a Connexin etiology and a significant decline in VR with

age for children with a Structural Abnormality. There was also a trend for a decline in VR with age for children with a Connexin etiology. There was no significant relationship between age and VR or FM for the Unknown group.

The possible implication of cognitive factors based on the etiology of the sensorineural HL was next investigated. Table II shows the mean scores and standard deviations of the VR and FM scales for each of the three etiologic groups. It can be seen that among each of the groups, VR scores consistently fell within the average range. FM scores fell within the average range for the Connexin and Structural Abnormality groups but below average (low average range) for the Unknown group.

A multivariate analysis of variance (MANOVA) was conducted to investigate whether there were differences among the groups on the measures of visual reception and fine motor skills. The results suggest that judgment be suspended with regard to whether there were significant differences between groups as a result of the significance level for the model falling at $P = .06$.[16] Therefore, the analysis of potential group differences was limited to an a priori planned comparison. For the planned comparison analysis, the Unknown group was used as the reference group. This was because children within the Unknown group had the same functional difference (i.e., hearing impairment) as the comparison groups but as a result of a wide variety of etiologic mechanisms. The results of the univariate test for fine motor skills approached significance $F(2,34) = 2.96$, $P = .066$. An examination of the planned comparisons indicated that there was a significant difference between the Unknown group and the Connexin group for the measure of fine motor skills (favoring the latter group). This difference was significant to $P = .021$, with the contrast difference between the means equal to 11.700 and the SD 4.812.

An analysis of the size of the effect associated with the significant tests of differences indicated above was conducted. Wilk's Lambda was used to estimate the size of the effect for the MANOVA comparison. The results indicated that approximately 15% of the variance was a result of the variance between groups on the two measures $\eta^2 = 0.153$. The proportion of the variance estimated to be a result of the difference between groups as a result of the differences in the performance on the VR scale is estimated to be 3% as indicated by the $\eta^2 = 0.0385$. The proportion of variance estimated to be a result of the difference between groups as a result of the differences in the performance on the FM scale is estimated to be 15% as indicated by the $\eta^2 = 0.152$. These results indicate support for the significance of the difference between groups on the FM scale and indicate that the majority of the difference between groups on the MANOVA is not caused by the differences on the VR scale.

DISCUSSION

Among deaf toddlers/preschoolers, does a significant decline in visual reception or fine motor skills occur with advancing age? Consistent with our previous study, correlational analyses indicated that preschool-age children diagnosed with sensorineural HL had a significant decline in visual reception skills with advancing age. Fine motor skills were also found to significantly decline with advancing age, confirming the trend reported previously.

If there is a significant decline in visual reception or fine motor skills with age, does this relationship hold true regardless of the etiology of deafness? Repeat correlational analyses of the neurocognitive areas of visual reception and fine motor skills when considering etiologic status indicated a significant reduction in fine motor skills with age in the Connexin group and a trend for a reduction in visual reception with age in this group. A significant reduction in visual reception with age was found in the Structural Abnormality

group. There was no significant relationship between age and visual reception or fine motor skills in the Unknown group.

What, if any, etiologic group differences are there with regard to visual reception and fine motor skills? When considering the etiologic status of deaf children, the Connexin group consistently performed the best on the Mullen VR and FM scales, followed by the Structural Abnormality group. Mean group performance on both visual reception and fine motor fell within the average or age-expected range, with the exception of the performance of the Unknown group on the fine motor scale, which fell in the low average range. A planned comparison found a significant difference with regard to fine motor skills between the Connexin and Unknown group, favoring the Connexin group.

Overall, the results of the present study support the importance of considering nonverbal skill development in children diagnosed with sensorineural HL. Although the previously mentioned “crowding hypothesis” would suggest enhancement of visual skills over time as a result of reduced stimulation to spoken language areas of the brain, our results did not support this theory across the general sample of children with HL or when etiology was considered. In fact, visual skills were found to significantly decline with age for the Structural Abnormality group, and there was a trend for this pattern among the Connexin children. There was no such relationship for the children whose etiology of HL was unknown. It is possible, however, that the “crowding hypothesis” is supported at a later developmental stage. Emmorey et al.[17] found evidence that myelination within auditory cortices depends on auditory input during development. Because myelination occurs with projection pathways prior to association pathways and there is evidence of a pathway from the primary auditory to the primary visual cortex, it is possible that the current findings are indicative of reduced pathway connectivity. Over time, however, this reduced interaction between auditory and visual domains may result in the effects proposed via the “crowding hypothesis.” With regard to fine motor coordination, development corresponds to myelination, and it is possible that myelination in this domain is similarly affected. This hypothesis could be investigated further via additional measures of fine motor and visual processing speed.

This study also supports the importance of identifying the etiology of the deafness to determine the possible neuropsychological profile to better inform treatment planning. Children with a genetic etiology of a Connexin mutation appear to fare the best on nonverbal measures of visual and fine motor functioning, as consistent with previous studies. Children with an etiology of Structural Abnormality also performed in the age-expected range on measures of visual and fine motor skills. The final group, in which all known etiologies of deafness had been ruled out, performed within the average range with regard to visual reception but mildly below average with regard to fine motor skill. It will be important that research and medical advances continue to focus on improving the ability to determine etiologic cause so as to better identify the etiology of children currently categorized within the Unknown group because it appears that these children experience increased difficulty compared with those with a Connexin etiology. Regardless, the results support intervention in the areas of visual skills as well as fine motor development rather than the traditional targeting only of spoken language development in these children. As such, ensuring that these children receive occupational therapy as well as participate in activities to enhance visual and fine motor skills (such as puzzles) is warranted.

With a larger sample size, future studies may include item analysis of the Mullen scales to identify whether there are certain patterns as to the items that children with HL experience success with or more difficulty with compared with normative samples (for example, short-term visual memory, visual-sequential tasks). The current study could also be furthered

through use of a comparison group of deaf children who are fluent in, and continually exposed to, sign language. This group would help identify any benefit sign language has on the development of visual and fine motor skills, possibly because of the consistent use and "practice" of these neurocognitive areas. Follow-up studies of these children after cochlear implantation would also be beneficial to ascertain to what extent a return of auditory stimulation impacts development of nonverbal skills and the impact of etiologic status on the level of benefit these children experience with cochlear implantation.

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TABLE I

Correlations Matrix for Visual Reception and Fine Motor *t* Score With Age (mo) for Etiologic Groups.

	VR *t* Score	FM *t* Score
Connexin		
Pearson correlation	−.50	−.58*
Significance (2-tailed)	.059	.023
Structural Abnormality		
Pearson correlation	−.63*	−.38
Significance (2-tailed)	.038	.250
Unknown		
Pearson correlation	−.28	.01
Significance (2-tailed)	.437	.975

*Significant at .05 level.

†Significant at .01 level.

TABLE II

Mean *t* Scores for Visual Reception (VR) and Fine Motor (FM) Scales by Etiologic Groups.

Mullen Scale	Etiology	Mean	(SD)	*df*	F	Sign
VR *t* Score				2	0.66	.523
	Connexin	49.80	(11.84)			
	Structural	47.18	(12.13)			
	Unknown	44.00	(13.42)			
FM *t* Score				2	2.96	.066
	Connexin	52.60	(11.01)			
	Structural	48.27	(10.80)			
	Unknown	40.90	(13.84)			