

Improved Health and Development of Children who are Deaf and Hard of Hearing Following Early Intervention

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Abstract

Infants begin to learn language in the earliest months of life. In the absence of early identification and intervention, infants who are deaf or hard-of-hearing experience significant and lasting deficits in language learning, academic achievement, social-emotional development, and quality of life. Evidence is mounting that early identification of infants who are deaf or hard of hearing through newborn hearing screening and intervention by the age of 6 months improves developmental outcomes for these children, especially in the area of language proficiency. Newborn screening programmes, including newborn hearing screening, are typically public health activities aimed at the early identification of infants who are affected by certain congenital disorders: including genetic, metabolic, haematologic, and infectious diseases. Early identification of these conditions is critical, as timely intervention can lead to a significant reduction of morbidity, mortality and associated disabilities in affected infants. For infants with hearing loss, the goal of early identification is to provide early intervention leading to language development and academic achievement commensurate with cognitive ability, and ultimately an improved quality of life. For newborn hearing screening, the definition of early intervention is initiation of intervention by the age of 6 months. Initiatives for newborn hearing screening have spread to every continent and many countries now have well-developed, comprehensive programmes of screening, diagnosis, and early intervention for childhood hearing loss. Although no exact statistics currently exist, tens of millions of infants probably receive newborn hearing screening each year. Because the developmental effects of early intervention for hearing loss are improved and sustained language development, large-scale studies documenting the language outcomes in early-identified children take decades to collect. Furthermore, because full-scale implementation of universal newborn hearing screening has occurred only within the last 5 years in most countries, many early-identified children are still at preschool-age. Thus, documenting these youngsters' language development is a work in progress. Despite these limitations, evidence is mounting that early intervention for childhood hearing loss improves the developmental outcome of these children.

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Language Development in the Earliest Months of Life

Although normally-developing infants appear to learn their mother tongue effortlessly, they are actively engaged in language-learning from the birth through early childhood. Linguists, social scientists, and developmental psychologists have extensively studied language development in 2 domains: the infant's perception of the acoustic stream of speech which comprises language, and the infant's production of non-speech and speech-like sounds and words. During the first year of life, rapid changes in both perception and production are observed.

For perception, research has shown that, at birth, babies can distinguish virtually all the phonetic units used in languages; they are language universalists. Of the 600 consonants and 200 vowel sounds in the world's languages, babies can appreciate all these sounds. However, because each language uses a unique set of only about 40 specific language units, or phonemes, by 6 months of age, the infant begins to develop phonetic perception specific to his or her mother tongue.¹ By 8 months of age, babies demonstrate knowledge of the supersegmental features of speech, such as stress patterns, and by 9 months of age, normally-hearing infants recognise language-specific sound combinations.

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By 1 year of age, perception of acoustic distinctions of non-native language consonants declines, and babies have become language specialists.

Production of language also progresses rapidly during the first year of life. Up until about 3 months of age, babies produce only non-speech sounds; cries and grunts. At 3 months of age, vowel-like sounds are produced which rapidly evolve into consonant-vowel repetitive babbling with syllabic timing patterns, termed canonical babbling. This represents an important milestone linking perception and production. By the age of 10 months, infants combine sounds into mother-tongue specific speech sounds, usually resulting in the infant's first native language word at the age of 12 months.

Recent neuropsychological and brain imaging research has shown that language acquisition involves *neural commitment*. Exposure to language produces dedicated neural networks that code speech patterns: the "native language neural commitment" theory of language development. As the distribution of speech patterns that a baby hears stabilises in neural representation, the period of learning novel speech patterns begins to close, resulting in a sensitive language-learning period. As summarised by Kuhl,² "...an absence of early exposure to patterns that are inherent in natural language ~ whether spoken or signed ~ produces life-long changes in the ability to learn language..."

Effect of Congenital Hearing Loss on Language Development

Congenital hearing loss has a devastating effect on this normal process of speech perception and speech production. Children who do not hear from birth do not experience the normal stimulation needed to develop neural networks specific to language learning. Research has shown that deaf infants are significantly delayed in onset canonical babbling.³ Furthermore, there is a clear correlation between age of onset of canonical babbling and age at which auditory amplification is provided to hearing-impaired infants⁴ proving the importance of hearing to speech development.

If mastery of the rules of language does not occur within the "sensitive period," the probability that the child can adequately develop this skill is significantly diminished. Research from national surveys of deaf children in the United States has shown that, on the average, children with hearing loss have a 20 to 40 point discrepancy between verbal and performance intelligence scores. In addition, only 40% of deaf children at high school graduation demonstrate reading levels at the fourth grade equivalent or above.⁵ Thus, the lifetime effects of congenital hearing loss cascade from delayed language development into (i) poor literacy skills, (ii) decreased academic success, (iii) limited

job opportunities, especially in a world economy highly dependent on oral and written communication, (iv) social-emotional problems, (v) lowered lifetime economic success, and ultimately, and (vi) reduced quality of life.

Interventions for Congenital Hearing Loss

There are multiple effective interventions for congenital hearing loss. The 3 principle interventions are (i) parent education and support, (ii) amplification and cochlear implants, and (iii) early intervention.

Parent education and support includes training the family on the effects of congenital hearing loss, facilitating development of a language-rich environment in the home setting, helping families advocate for and obtain the resources needed for their child's optimum development, and training parents to recognise developmental milestones. Research has shown that high levels of family involvement in children's intervention programmes correlate with positive outcomes for these children.⁶ In addition, Calderon⁷ documented that maternal communication skills are predictive of language development and literacy competency for children who are deaf.

Conventional hearing aids provide infants who are hard of hearing, that is, infants with residual hearing, access to sound. Contemporary hearing aids are digitally-programmable, flexible, electroacoustic devices with adaptive listening environment features such signal-to-noise reduction capabilities. Hearing aids may be fitted to infants as young as 2 to 3 months of age.

For infants with minimal residual hearing, cochlear implants provide important, albeit limited, access to sound. A cochlear implant consists of a stimulating electrode array that is surgically implanted into the cochlea to provide direct electrical stimulation of the auditory (eighth cranial) nerve, and an external receiver/transmitting device that extracts critical speech elements from the spoken word and converts this acoustic stream into an electrical stream for activating the stimulating electrode array. For infants and young children, cochlear implants are typically considered after a trial with conventional hearing aids has demonstrated very limited benefit, and usually after about 1 year of age. Multiple studies have shown that children who receive early implantation obtain better language-learning results than children who are implanted at older ages.^{8,9}

Early intervention for childhood hearing loss is customised to the family's desired language mode, whether native sign language or auditory-oral, spoken language. It is typically initiated as soon as the infant is confirmed with permanent hearing loss, and provided in the home-setting with guidance for developing a language rich environment. Families of children who are deaf and hard of hearing must commit to intervention throughout the child's toddler and preschool

years. Research demonstrates that early identification and early intervention improve language outcomes for children who are deaf or hard-of-hearing.^{6,10-12} For example, Colin Kennedy and his colleagues in the U.K. studied language and speech outcomes in one hundred and twenty 8-year-old children with bilateral permanent hearing impairment identified from a large birth cohort in southern England. Of these 120 children, about half of them were born during periods of universal newborn hearing screening and the other half were born during periods without newborn hearing screening. Using standardised assessment tools to measure language competency, the authors found that children who were exposed to universal newborn hearing screening or who had confirmation of hearing loss by 9 months of age had significantly higher scores for language, but not for speech, in mid-childhood than those who were not exposed to newborn hearing screening or whose impairment was confirmed after 9 months of age. For the early identified group, the difference in language was equivalent to 10 to 12 points in the verbal as compared to the nonverbal intelligence quotient.

How Early is “Early Enough?”

“Early intervention” for childhood hearing loss is considered intervention by the age of 6 months. Assuming high-quality intervention and optimum family commitment, is intervention by age 6 months early enough to ensure language development commensurate with cognitive ability? The medical geneticists, Morton and Nance¹³ argue that newborns who fail initial screening should move immediately into audiological confirmation of hearing loss before discharge from the birth admission. By their proposal, infants who fail newborn hearing screening receive confirmatory diagnosis before age 1 month and are enrolled in early intervention by the age of 2 months. If successful, this acceleration of the identification and intervention process might: (i) reduce loss-to-follow up, (ii) improve specification of etiology of hearing loss, especially for babies with congenital cytomegalovirus, and (iii) normalise access to sound for hard of hearing infants through prompt hearing aid fitting.

For babies for whom otological and audiological diagnosis of hearing impairment is possible immediately after birth, intervention by 2 months of age should be possible. Whether families are comfortable with an accelerated diagnostic process is unknown; however, reducing that period of uncertainty that worries families today would seem beneficial.

Summary

Children who are deaf or hard-of-hearing will enjoy improved health and developmental outcomes only if early

identification and intervention are effective. Evidence is emerging from well-developed and implemented programmes of universal newborn hearing screening that early identification and intervention improve language development in these youngsters, at least through mid-childhood. Through universal newborn hearing screening, the goal of language competence should be within reach for all deaf and hard-of-hearing children.

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