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Monique Ford

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A RETROSPECTIVE LOOK AT COCHLEAR IMPLANTATION OF POST-LINGUALLY DEAFENED CHILDREN

by

Monique Ford

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INTRODUCTION

Speech and Language Deficits

Language can be thought of as a communication system consisting of three basic elements: phonology (sounds the language uses and the rules for their combination, semantics (the meaning of the words of a language), and syntax (rules for constructing sentences (Berko Gleason, 2001, p. 505). While speech can be thought of as that which is spoken, it is not synonymous with language. Being able to recognize, produce, or imitate spoken words or sentences is not the same as having a meaningful use or understanding of the language (Preisler, Ahlstrom, & Tvingstedt, 1997). "Language is a mode of action into which the child grows because the mode is implicit in the human developmental system" (Studdard-Kennedy, 1991, p. 24). There are critical learning periods that exist early in childhood that affect the ability of children to develop normal speech and language and are apparently developmentally interdependent (Novak, Firszt, Rotz, Hammes, Reeder, & Willis, 2000).

Research has shown that the infant begins to learn to distinguish speech sounds almost immediately after birth, between the 1st and 4th months of age. Infants can recognize many speech sounds of language and are able to catalog these different sounds by 6 months in age (Northern & Downs, 2002).

Ruben (1997) summarized the literature and found support for a critical period of phonological development that exists in the first 12 months of life. Following phonological development is semantical and syntactical specialization, which is thought to have critical/sensitive periods of 4 years and the late teens, respectively. From his perspective, phonology is the foundation on which syntax and semantic abilities are built, and if speech development is delayed by the sensory deprivation of deafness, then spoken language will also be delayed. Not only does deafness limit or prohibit access to the phonetic code of spoken languages, it also lessens the child's exposure to the varied language models that normally hearing children receive from listening to those around them talk (Novak et al., 2000). However, deafness does not cause one specific kind of communication problem.

The effects of a hearing loss depend on its severity, configuration, duration, and stability as well as the age of the person at the onset (Northern & Downs, 2002). There are a number of detrimental conditions that affect a child's language learning when she/he has a hearing loss. One such detriment is the lack of constancy of auditory clues when acoustic information fluctuates. "When a child does not hear speech sounds in the same way from one time to another, there is confusion in abstracting the meanings of words due to inconsistent categorization of speech sounds" (Northern & Downs, 2002, p.19). Confusion of acoustic parameters in rapid speech is also a problem for a child with a hearing loss. Frequency, duration, and intensity vary as a result of differences among speakers of varied ages, gender, and personality types. The child with hearing loss will be confused in language learning as a result. He or she will also exhibit confusion in segmentation and prosody. The child with hearing loss may miss linguistic markers such as plurals and tenses, as well as suprasegmental cues such as intonation and stress patterns. These factors are fundamental for meaningful interpretation to speech. A breakdown in early perception of meanings can occur. If a hearing listener misses some unstressed words or sounds, she/he is able to fill in by understanding the context of the message. However, when a hearing loss causes a young child to miss many of these soft or inaudible sounds, there is confusion in word naming, difficulty in developing classes of objects, and misunderstanding of multiple meanings. Lastly, faulty abstraction of grammatical rules and the missing of subtle stress patterns can both result from the hearing loss of a child. When short words are soft, it becomes more difficult for a hearing-impaired child to identify the relationships between words and to understand word order. Also, the emotional content of speech, its rhythm, and its intonation are communicated through the low frequencies, thus this is another condition that impairs the learning of speech and language (Northern & Downs, 2002). Nonetheless, the type of education received by a child who is deaf will determine her/his mode of communication.

Tools of Oral/Deaf Education

Auditory/oral methods are educational methodologies that encourage deaf children to communicate via spoken words. A principal surrounding the auditory/oral methods is that deaf children, if at all possible, should be given the opportunity to communicate by speech. Being a good listener and using one's vision to become a good speechreader are important in being an aural/oral communicator if one is deaf. For this approach, the acoustic channel needs to be developed as the primary mode of spoken language input, and the use and development of the visual channel will come naturally as needed. The main aim of these methods is to make children with hearing impairment or deafness an integral part of the hearing society through good speechreading and hearing aid or cochlear implant use (Northern & Downs, 2002).

Hearing aids, which are electronic amplifiers, are used for rehabilitation of any hearing loss. The device consists of the basic microphone-amplifier-receiver components contained within a single package that is worn in or around the ear (Stach, 1998). The microphone is a transducer that alters acoustical energy into electrical energy. The amplifier increases the level of electrical signal that is sent to the hearing aid. It also controls how much amplification occurs at certain frequencies. (Stach, 1998). Conversely, cochlear implants are used primarily for rehabilitation for severe-profound losses, and are designed to stimulate the auditory nerve directly.

"Cochlear implants have an electrode array surgically implanted into the cochlea. The electrode array is attached to a magnet that is implanted into the temporal bone. Acoustic signals are received via a microphone attached to an amplifier. The amplifier then sends signals to the electrode via the implanted magnet/receiver. When the electrode receives a signal, it amplifies an electrical current to the cochlea, thereby stimulating the auditory nerve" (Stach, 1998, p.508). In analyzing speech, cochlear implants extract frequency, intensity, and temporal cues from speech and translate them to an electrode array in a way in which the residual neurons of the auditory nerve can process them.

The central responsibility of the auditory organ is maintaining equilibrium. The sense of hearing developed from the primordial structures developed for balance (Northern & Downs, 2002). The outer and inner portions of the ear develop from ectodermal tissue, whereas the middle ear ossicles and the bone surrounding the inner ear originate from mesodermal tissue. The ear begins its development during the early life of the embryo. It is during the stage of the neural tube that the earliest beginnings of the ear are seen.

The earliest demarcations of the ear in the human embryo are seen early in the third week as thickenings in the superficial ectoderm on either side of the open neural plate (Northern & Downs, 2002). The empholymphatic duct can be eventually recognized as the future vesticular portion of the labyrinth, while the more slender portion of the vesicle begins to elongate from the saccular area as the future cochlea. Three arch-like out-pockets become visible and are destined to become the semi-circular canals, simultaneously as the utricle and saccule become two definitve areas through a deepening construction of the vestibular portion of the auditory vesicle (Northern & Downs, 2002). During the 8th through 11th week, the 2.5 coils of the cochlear are completed, and the cochlear portion of the eighth nerve follows the elongating and coiling of the cochlear duct, and fans its fibers out to be dispersed along the duct's entire length (Northern & Downs, 2002).

The membranous labyrinth of the inner ear reaches its full adult contour by the early part of the third month. It is the only sense organ to reach full adult size and differentiation by fetal midterm (Northern & Downs, 2002). However, the cochlea is the last inner ear end organ to differentiate and mature.

While the inner ear is developing, the middle ear is developing as well. It begins its development during the third week. A passageway from the pharynx to the outside of the head develops and becomes the external ear canal and Eustachian tube (Northern & Downs, 2002). By the eighth week, the tympanic cavity is present in the lower half of the future middle ear. The malleus, incus, and stapes are the next structures to develop.

The external ear develops during the third or fourth week during the same time that the auditory vesicle is formed in the development of the inner ear. It moves from its medial position to a more lateral one due to the growth of the mandible and face.

(Northern & Downs, 2002). The auricle continues to grow in size until the individual is 9 years of age. The external auditory meatus continues to develop until the 9th year as well.

Cochlear Implants

Cochlear implants have become a widely accepted form of (re)habilitation for some deaf children (Zimmerman Phillips, McConkey Robbins, & Osberger, 2000). One of the main aims of implantation is to give the recipient access to sound so that, in the case of a child, they can procure speech and language (Brinton, 1999). Some feel as if cochlear implantation will allow these children to become independent, participating, and contributing adults in a world in which speech is the chief means of communication. The National Association of the Deaf (NAD) released an updated position statement on cochlear implants in October of 2000. In this paper the group asserted that, "Diversity requires mutual respect for individuals and/or group differences and choices" (NECCI News, 2000). This was a markedly different view than what they shared in April of 1993. Upon the Food and Drug Administration's approval of cochlear implants in children in 1990, the NAD issued a position paper on cochlear implants in children, "deploring the decision of the FDA as being unsound scientifically, procedurally, and ethically" (NECCI News, 2000). In keeping with the most recent NAD statement and largely due to the urging of ENT surgeons, there has been a trend toward earlier implantation. Much of the impetus for this is due to the observed improvements in measures assessing areas of speech perception, speech production and language acquisition (Miyamoto, Kirk, Svirsky, & Sehgal, 1999).

Cochlear implants have been shown to be the treatment of choice in children with sensorineural hearing loss: congenital or acquired, profound to severe (Lenarz, 1997).

They are safe and reliable and also effective in terms of speech perception as well as speech and language acquisition (Lenarz, 1997). Successful cochlear implantation is expected to produce a cascade of benefits to children with severe and profound hearing losses. These would take the form of improvements in auditory, speech, and linguistic skills in the short term. One study indicated that the rate of vocabulary acquisition in profoundly deaf children tended to decrease over long periods of follow-up after implantation (El-Habim, Papsin, Mount, Levasseur, Panesar, Stevens, Y Harrison, **2001**[2]). Miyamoto et al. (1999) noted different findings in the research on speech production and language skills of most prelingually deafened children who received implants before, at, and after the age of 3 years. Significantly higher scores on the Grammatical Assessment of Elicited Language – Pre Sentence Level (GAEL-P), word and sentence scores, and speech intelligibility were received by children implanted before 4 years of age. However, children in all experimental groups showed delayed receptive vocabulary relative to their chronological age when the Reynell Developmental Languages Scales and Peabody Picture Vocabulary Test (PPVT) were administered to them. They obtained a vocabulary age that was somewhere near half of their chronological age. Also, children in this study who used oral communication had significantly better spoken word recognition, speech intelligibility, and expressive language abilities when compared with those who used Total Communication. Miyamota, Svirsky, and Robbins (1997) noted that the cochlear implant permits better perception of speech sounds and increase access to the segmental features of speech. This, in turn, improves spoken language performance.

Implanted children have been shown to acquire a more mature vocabulary and the syntactic structures necessary for communicating through spoken language faster than those who use hearing aids or tactile aids (Geers & Moog, 1994). Rapid improvements in a short amount of time suggest that cochlear implants produce immediate gains in specific aspects of speech production, possibly with slower progress, or even a decline, later on (Te, Hamilton, Rizer, Schatz, Arkis, & Rose, 1996). However, the real effects of a cochlear implant on expressive language development may be hard to determine because the groups studied vary across a large number of factors relevant for language development – age at onset of deafness, duration of deafness, type of language input received (Ouellet, Le Normand, & Cohen, 2001). Ouellet et al. (2001) found that implanted children can gradually increase their vocabulary and rearrange the components of this enlarged vocabulary to build more and more complex sentences. They then acquire more mature syntactic structures and a larger lexicon. They noted that these children also present significant deficits in general language production even with the implanted hearing device and are expected to remain delayed at subsequent evaluations in comparison to both age-matched hearing children and younger ones.

In looking at the speech intelligibility of 18 children with prelingual deafness, Osberger, McConkey Robbins, Todd, & Riley (1994) found that the average speech intelligibility score of children who used oral communication was 48%, which was significantly higher than the average score of 21% of the subjects who used total communication. The range of scores for the subjects who used oral communication was relatively large, with the scores of the subjects with the lowest intelligibility comparable to those of the subjects who used total communication. The authors reported a mean speech intelligibility score of nearly 40% in subjects who had used the Nucleus multi-channel cochlear implant for 3.5 years or longer. Major improvements in intelligibility did not occur, however, until after the subjects had used their multi-channel cochlear implants for two or more years. Before implantation of subjects who previously used hearing aids, Valimaa, Sorri, and Lopponen (2001[1]) found the mean recognition score was 38% for sentences and 17% for words. One year after switching on the implant, the mean recognition score was 84% for sentences and 70% for words. Those with the longest duration of profound hearing impairment scored the lowest on both sentence and word recognition. However, Balkany, Hodges, Miyamoto, Gibbin, & Odabasi (2001) found that implanted children learn language at the same rate as hearing peers of the same age (except for vocabulary, which develops at a faster rate). Nonetheless, implanted children were shown to retain a language delay relative to their hearing peers equal to that present at the time of implantation. El-Hakim et al. (2001[1]) concluded that children with cochlear implants developed their vocabularies at rates that were sufficient to prevent an increase in their gap indices on the PPVT and Expressive One-Word Picture Vocabulary Test (EOWPVT).

Phoneme production accuracy exceeded reported performance levels of children with profound hearing loss who use hearing aids in the research of Tye-Murray, Spencer, and Woodsworth (1995). Overall intelligibility was found to be low, and only 22% of the words in story-retell samples were produced without any phonemic errors. Blamey, Barry, Bow, Sarant, Paatsch, and Wales (2001) found that all nine children observed in their longitudinal study showed substantial gains in the intelligibility, length, and phonemic accuracy of their speech during the 6 years immediately following implantation with a cochlear implant. The plasticity of the brain may play a major role in the changes that occur after implantation.

Plasticity of the Brain

"A major theme in neuroscience is that the brain is not a "rigid" structure but a malleable, "plastic" organ with the capability of reorganizing itself based on sensory and motor input, a phenomenon known as neuroplasticity" (Northern & Downs, 2002, p.128). The neurons in the cortex mature during the first 3 years of life, and there's not much organizational change after this age. From the beginning of life brain cells increase rapidly, making connections that will shape a lifetime of experiences. As the neurons carry electrical signals along the nervous system, systematic pathways are established through coordinated routes that are used over and over again. The stimulated neurons develop long axons that send out multiple branches that connect with a large number of different neurons. Spontaneous bursts of electrical activity have been thought to strengthen some of those pathway connections, while other neurons that are not reinforced by electrical activity begin to atrophy and disappear (Northern & Downs, 2002). During the early years of life, the brain eliminates the excess neurons and connections that are seldom or never used. New sensory experiences cause an increase in cortical and brainstem electrical activity and this aids the brain's circuitry in determining which connections will remain and which will not (Northern & Downs, 2002).

Even though brain cell formation is nearly complete before birth it's maturation is far from this stage of development. Connections among the neurons are made that allow learning to take place.

> Over the first few days, weeks, and months of life, sensory activity stimulates the neuronal connections from the brainstem to the appropriate areas of the cortex. The results of these early sensory experiences are those actions we have come to

> expect as normal infant development: by 2 months the baby is able to grasp objects, by 4 months the complex actions required to locate a sound in space are initiated, by 6 months a baby can recognize and mimic the vowel sounds that are the precursors to speech formation, and by 12 months, we begin to see the results of neural pathways formed to produce the first words that mark the beginning of language expression (Northern & Downs, 2002, p.129).

Also, there is the possibility of the environment having a long-term effect on early development. The prenatal environment affects not only the number of brain cells and their connections, but also the way that these connections are "wired" (Northern & Downs, 2002). The authors also note there is evidence that reintroduction of sensory input after auditory deprivation induces further plastic changes, and deleterious effects may be reversed only during early stages of development. The brain's growth spurts begin to slow down around 10 years of age. By the end of adolescence, around age 18, the brain has declined in plasticity but increased in power. "Although the adult nervous system continues to lay down new synaptic connections as we learn new ideas and skills, never again will the brain be able to assimilate and master new information as readily as during the first 3 years of life" (Northern & Downs, 2002, p.130). It appears therefore, that cochlear implants have their most profound effect during the childhood years.

Postlingually Deafened Children

Children who are born with a profound hearing loss, or who suffer a profound hearing loss in the early stages of spoken language development, find it difficult to hear and understand speech, even with a powerful hearing aid (Blamey et al., 2001). This population frequently has delayed or disordered speech production patterns and delayed development of other aspects of language such as vocabulary and syntax, which has been documented by (Geers, Moog, and Schick, 1984; and Bamford and Saunders, 1992). A consistent finding across studies is that the average intelligibility of profoundly hearing-impaired children's speech is only 20%, although individual scores can range from 0% to 80% approximately (e.g., Smith, 1975). The capacity to incorporate a significant number of words into the existing vocabulary and to use these words to form sentences with more mature syntactic structures over a considerable time course is expected of young children who have undergone a cochlear implantation (Ouellet et al., 2001).

It seems to be the case with most children that expressive language improves with experience of the device (Ouellet et al., 2001). Speech recognition afforded by the cochlear implant should effectively supplement the information least favorably cued through speechreading (NIH Consensus Development Panel on Cochlear Implants in Adults and Children, 1995). Substantial variability is to be expected across children due to age of onset, age of implantation, the nature and intensity of (re)habilitation, and mode of communication. Perceptual performance increases on average each succeeding year after implantation, and performance may be broadly comparable to that of some children with hearing aids (NIH, 1995).

> Children receiving implants at younger ages are on average more accurate in their production of consonants, vowels, intonation, and rhythm. Speech produced by children with implants is more accurate than speech produced by children with comparable hearing losses using vibrotactile devices or hearing aids. One year after implantation, speech intelligibility is twice that typically reported for children with profound hearing impairments and continues to improve (NIH, 1995).

What has been reported for postlingually deafened adults, as well as some children, with immediate implantation is marked improvements in speech perception, often within a few months (Dowell, Mecklenburg, & Clark, 1986). Hildesheimer, Teltelbaum, Segal, Tenne, Kishon-Rabin, Kronenberg, & Muchnik (2001) have reported a significant difference between groups of profoundly deafened adults and children in that between both groups in a short time, within 0.5 to 1 year, the adults reached their maximum performance on open set tests of Hebrew origin, whereas the children may have reached it within 6 months, but as a group, they required on average 2 years. Valimaa et al. (2001^2) have shown ten subjects tested before implantation both with and without a hearing aid, and 3, 6, and 12 months after switching on the implant with recognition scores of 34% for vowels, 28% for consonants, and 13% for syllables to mean recognition scores of 77% for vowels, 66% for consonants, and 46% for syllables. According to phonological analysis, vowels appear to be easier to perceive than consonants during the first stage after implantation. Also, children with shorter durations of deafness have achieved substantial open-set speech recognition without lipreading Children and adults implanted after longer periods of profound deafness have achieved improvements on open-set tests with lipreading and closed-set tests without lipreading as well (Dawson, Blamey, Rowland, Dettman, Clark, Busby, Brown, Doweel, & Rickards, 1992).

Purpose of This Study

The purpose of this study is to examine the benefit of relatively immediate cochlear implantation following post-lingual deafness in the preschool period as compared with initiation and continuation of intervention with traditional hearing aids. Language progress will also be compared with deaf children of the same chronological age who were deaf from birth and used hearing aids. The purpose of the latter comparison group is to show whether or not there is a difference in the language development of hearing aid users who are congenitally or postlingually deafened.

METHOD

Participants

This study includes nine females and nine males who attended Central Institute for the Deaf between the years of 1972 to 2002. Group 1 is composed of nine congenitally deafened children who wore hearing aids. The age range at diagnosis of this group was 5 months to 4 years with a mean age of 1 year, 5 months. The age range for which this group was aided was 8 months to 4 years, with a mean aided age of 2 years. Group 2 consists of four postlingually deafened children who wore hearing aids. The age range for diagnosis was 1 year, 10 months to 3 years with a mean age at diagnosis of 2 years, 3 months. The range in age at the time these children were aided was 1 year 10 months to 3 years with a mean aided age of 2 years, 4 months. Table I summarizes these characteristics for Group 1 and 2. Group 3 consists of five postlingually deafened children who were implanted with cochlear implants. The age range at onset for this group was 1 year, 6 months to 4 years, 9 months with a mean age at time of onset of deafness of 2 years, 1 month. The age range at the time of implantation was 5 years to 11 years 1 month with a mean age at time of implantation of 8 years, 7 months. The cause of deafness for four of the five children was meningitis, while only one was due to ototoxic drugs. Three of the five children in this group had the SPEAK processing strategy while two children had the F0F1F2 processing strategy. Table II summarizes the characteristics of Group 3.

Procedure

Before beginning this archival research study, an attempt to acquire subjects was made by asking veteran teachers of the school to submit student etiologies that were matches for groups 1, 2, and 3. Secondly, the CORKS, Clinical Oriented Records Keeping System, database was searched using variables of age (5 –6 year-olds) and tests administered (either a GAEL P, S, or C) to obtain more subjects. One hundred twenty records were examined and of these only 30 records were matches for groups 1, 2, and 3. Eighteen subjects in the 30 records perused were administered deaf-normed and/or normal hearing-normed tests during 5 and 6 years of age. The tests administered to the children included the Grammatical Assessment of Elicited Language P, S, and C (GAEL-P, S, and C), Scales of Early Communication Skills (SECS), Peabody Picture Vocabulary Test-Revised (PPVT-R), Expressive One Word Picture Vocabulary Test-R (EOWPVT-R), and the Test of Language Development-2 (TOLD), of which the latter three were standardized on normal hearing children and the former on deaf children. The earliest and latest used tests scores were from 1975 to 1994. Due to there being so few children who were administered the same test at the same age, the percentile ranks for all of the deaf-normed tests were averaged for each child, each group and overall.

Variables of Interest

The independent variables were time of deafness and type of sensory device. The percentile ranks received on both the deaf-normed and normal hearing-normed tests were the dependent variables.

RESULTS

The median percentile rank (PR) achieved for each child on deaf-normed tests at age 5 or 6 years is plotted in Figure 1. The range of percentile rank averages for individual children is from 15 to 86.3. The group medians are 73, 66, and 60 for groups 1, 2, and 3, respectively, and are displayed in Figure 2.

Eleven of the eighteen children included in these analyses were also administered language tests that were designed for and standardized on normally-hearing children. The median percentile rank average for all children who took these tests was 21.69 (range was 1- 38). This compares with an overall percentile rank median of 59.16 for the language test scores of these children on the deaf-normed tests. Due to the very small number of children in each comparison group (congenital onset, postlingual with and without CI) it was not possible to compare scores on deaf and normal-hearing tests within comparison group.

DISCUSSION

Due to the difficulty of accessing the records of children who would have been appropriate candidates for inclusion in this study, only a very small number of subjects were found. The group averages presented in the previous section can only be taken as very preliminary data addressing the question of whether or not cochlear implantation is especially beneficial just after onset of deafness. Clearly, the next step for this study is for another student, or a researcher, to continue collecting information on this question from student files until sufficient numbers of subjects are acquired. The inclusion of case history information in every clinical and educational record would greatly facilitate data collection and enable researchers to answer different questions regarding possible studies resourcefully and proficiently.

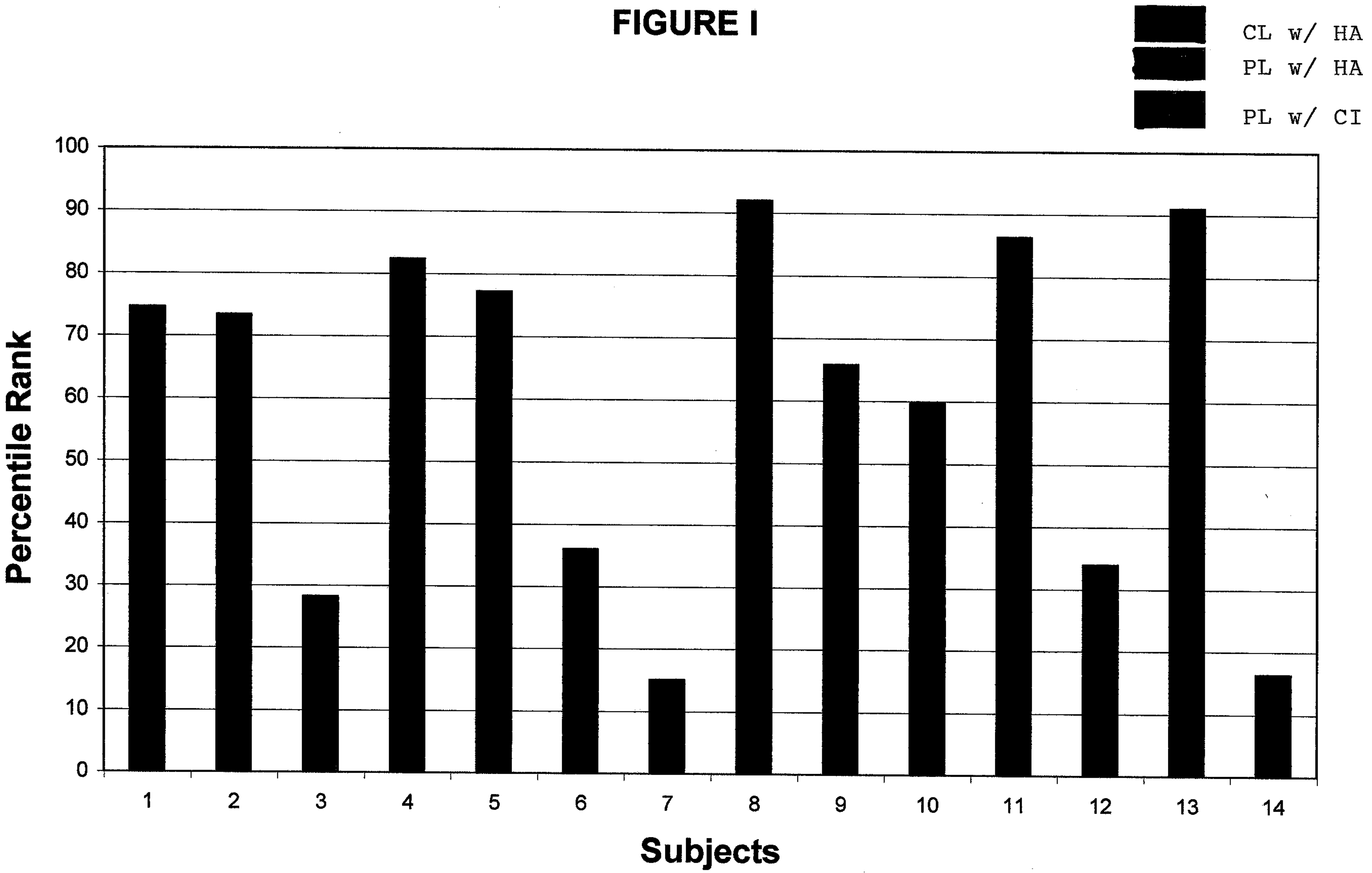
Participants of Group 1 and 2
Table I

Subject ID#	Group #	Age at Onset	Cause	Age Diagnosed	Aided
1	1	Birth	Illness at birth	8 mo	9 mo
4	1	Birth	Maternal Rubella	1 year, 7mo	3 years
5	1	Birth	Reactions to DPT shot	5 mo	8 mo
6	1	Birth	Maternal German Measles	6 mo	3 years
7	1	Unknown	Unknown	2 years, 8mo	3 yrs
8	1	Unknown	Unknown	1 year, 9mo	2 years
10	1	Unknown	Unknown	11 mo	1year, 4mo
12	1	3 ½ mos	Meningitis	6 mo	1year, 1mo
17	1	Unknown	Complications from Prematurity	4 years	4 years
18	2	1 year, 10mo	Meningitis	2 years, 1mo	2 years, 1 mo
19	2	2 years, 1mo	Ototoxic Drugs	2 years, 3mo	2 years, 5 mo
20	2	1 year, 10mo	Meningitis	1 year, 10 mo	1 year, 11mo
21	2	2 years, 3mo	Meningitis	3 years	3 years

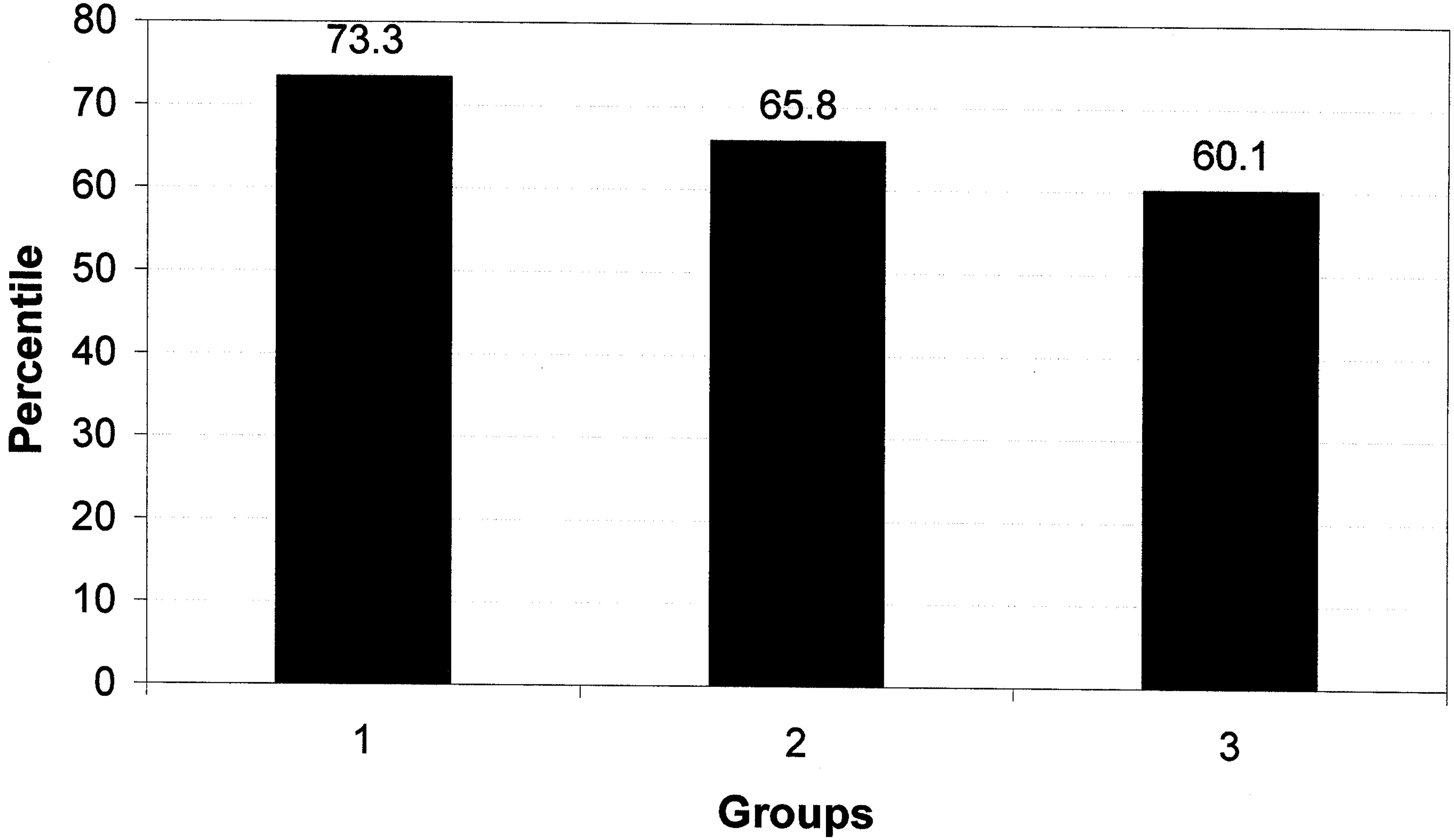
Participants of Group 3
Table II

Subject ID#	Age at Onset	Cause	Aided	Implanted	Processing Strategy
22	1 year, 6mo	Meningitis	1 year, 7mo	7 year, 11mo	SPEAK
23	1 year	Meningitis	1 year, 1mo	6 yeas, 7mo	F0F1F2
28	4 years, 9mo	Meningitis	4 years, 10mo	5 years	SPEAK
29	4 year, 2mo	Meningitis	4 years, 3mo	11 years, 1mo	SPEAK
30	1 year, 6mo	Meningitis	1 year, 7mo	10 years, 2mo	F0F1F2

PERCENTILE RANK AVERAGES FOR DEAF-NORMED TEST
FIGURE I



GROUP MEDIANS
FIGURE II



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