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Auditory Remediation for a Patient with Landau-Kleffner Syndrome: A Case Study

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Landau-Kleffner Syndrome (LKS) is a rare, childhood neurological disorder characterized by a sudden or gradual development of acquired aphasia. This case study offers a unique opportunity to assess the changes in the auditory processing ability of a 12 year old male with LKS after two distinct auditory training programs, Fast ForWord® and Dichotic Interaural Intensity Difference (DIID) training. Improvement in the electrophysiological recordings and the behavioral scores from the Dichotic Digits Test are evidence of the plasticity of the central auditory nervous system and may indicate a viable auditory remediation therapy for persons with LKS.

Introduction

Landau-Kleffner syndrome (LKS) is a rare childhood neurological disorder characterized by a sudden or gradual development of the inability to understand or express language. LKS is often referred to as acquired epileptic aphasia, acquired aphasia with convulsive disorder, or acquired receptive aphasia (Lees & Urwin, 1991; Paquier, Van Dongen, & Loonen 1992) characterized by an abnormal electroencephalogram (EEG) typified by abnormal spike activity in the temporal and/or parietal regions (Deonna, 1991). The abnormal EEG activity predominately occurs in the left temporal lobe, but may be present in both temporal lobes (Deonna, 1991) with nocturnal seizures occurring in over 80% of patients with LKS (Patry, Lyagoubi, & Tassinari, 1971).

The onset of LKS usually occurs between three and seven years of age, affecting males more often than females (Miller, Campbell, Chapman, & Weismer, 1984). The deterioration in language may be rapid, or may decline over a few months (Miller et al., 1984). Often, because a child with LKS fails to respond to language and environmental sounds, the child is thought to have acquired a hearing loss (Tharpe, Johnson, & Glasscock, 1991). A child with LKS may also be misdiagnosed as having autism or other developmental delays (Tharpe et al., 1991). LKS is known to be heterogeneous with varying symptoms, pathophysiology, degree of impairment, and prognosis. Furthermore, behavioral disturbances, such as aggression, attention, autistic-like behaviors, and withdrawal may be present and related to frustration from the communication breakdown (Tharpe et al., 1991).

Previous researchers have reported significant auditory discrimination deficits, as well as electrophysiological evidence, suggesting neural coding problems. Specifically, Vance, Dry, and Rosen (1999) reported on one 14 year old male with auditory discrimination deficits for syllables and words. Baynes, Kegl, Brentari, Kussmaul and Poizner (1998) reported on a 27 year old female with linguistic and non-linguistic auditory discrimination deficits, as well as a deficit in the discrimination of frequency and duration. Stefanatos (1993) has provided electrophysiological evidence suggesting an inability to phase lock to frequency-modulated (FM) steady-state cortical evoked response in a group of children with LKS, but not in a group of children with language impairment. The inability of the auditory system to phase-lock provides objective evidence for the underlying neurophysiological basis of the acoustical analysis of temporal cues, which is important for speech understanding (Kraus & Nicol, 2005).

While medical treatment for LKS usually consists of anticonvulsants to treat the seizure activity, there is very little information about the clinical management for the language disorder or acquired (central) auditory processing disorder ([C]APD). Hungerford, Coppens, and Clarke (1998) reported successful implementation of a computer-based language program for one patient with LKS. In addition, Pedro and Leisman (2005) reported significant improvement in the auditory, language, and motor skills in a case study of a 14 year old female after completing auditory integration therapy (Interactive Metronome).

Treatment of (C)APD generally focuses on three areas: 1) environmental changes to ease communication difficulties, 2) introduction of compensatory skills and strategies for the disorder; and 3) remediation of the auditory deficit. One type of direct remediation of (C)APD is an auditory training program that takes advantage of the brain's lifelong capacity for plasticity and adaptive reorganization, which may be at least partially reversible through a deficit-specific training program (Musiek, Chermak, & Weihing, 2007). Brain reorganization is reflected in an increase in the number of synapses, increased neural density, and improvement in auditory evoked responses (Elbert, Pantev, Wienbruch, Rockstroh, & Taub, 1995; Merzenich, Schreiner, Jenkins & Wang, 1993; Raconzone, Schreiner, & Merzenich, 1993). Changes in both behavioral and electrophysiological measures of auditory processing after auditory training reflect plasticity of the central auditory nervous system (CANS; Russo, Nicol, Zecker, Hayes, & Kraus, 2005; Warrier, Johnson, Hayes, Nicol, & Kraus, 2004) and may be used to monitor the effectiveness of an auditory training program.

To that end, we present a case report which monitors the changes in the auditory processing ability of a 12 year old male with LKS after two distinct auditory training programs: Fast ForWord® followed by Dichotic Interaural Intensity Difference Training (DIID; Musiek & Schochat, 1998). This report provides objective evidence of improvement in the central auditory processing ability following these forms of training.

Rationale for Fast ForWord®

Fast ForWord® is a commercially available software program that purports to capitalize on the plasticity of the auditory system. The term plasticity refers to "physiological changes in the central nervous system in response to sensory experiences" (Tremblay, 2003). Fast ForWord® is based upon the research of Tallal, Miller, Bedi, Byma, Wang, Magarajan, et al., (1996) and Merzenich, Jenkins, Johnston, Schreiner, Miller, and Tallal (1996). The Fast ForWord® program is designed to develop temporal and acoustic skills to detect rapid transitions of speech. It has been reported that (C)APD or auditory-based learning difficulties (language and reading disorders) may be remediated through intensive training provided by the Fast ForWord® program (Tallal et al., 1996; Merzenich et al., 1996). The exercises in the Fast ForWord® program use five levels of acoustically modified speech. At the beginning of the program, the exercises prolong and emphasize the sounds so that they are easier to distinguish. As the listener progresses, speech sounds approach normal speech. As the listener improves, the exercises become more challenging, and the participant develops enhanced language awareness and comprehension. The Fast ForWord® program was administered to our subject by his local school district and focused on sound blending, fine motor skills, hand-eye coordination, pattern recognition, and color-shape identification.

Rationale for Dichotic Interaural Intensity Difference (DIID) Training

Dichotic listening tasks are routinely used to assess CANS function, as they are known to be sensitive to central auditory dysfunction (Musiek & Pinheiro, 1985). In normal listening conditions, auditory information is conducted to the auditory cortex by both ipsilateral and contralateral auditory pathways; however, during controlled dichotic listening, the ipsilateral pathway is suppressed by the dominant contralateral pathway. This dominate contralateral model was first purposed by Kimura (1961), who (based on her data) hypothesized that the contralateral pathway must have greater neural innervations, which enables it to be dominant over the ipsilateral pathway.

Early work in the 19th century noted that most aphasia is the result of left cerebral hemisphere lesions with the conclusion that most people are left hemisphere dominant for expressive and receptive language (Webster, 1995). Furthermore, a language related auditory signal presented to the right ear travels from the dominant contralateral right auditory pathway directly to the left hemisphere. Conversely, a language related auditory signal directed to the left ear is conducted to the right cortex, but must be transferred to the left hemisphere via the corpus callosum in order for the person to repeat what was heard in the left ear. Thus, it is not surprising that there is a slight right ear advantage for neurologically normal listeners when listening to dichotic tasks (Berlin, Lowe-Bell, Cullen & Thompson, 1973; Kimura, 1961; Lowe, Cullen, Berlin, Thompson, & Willett, 1970).

When there is damage or a lesion in the auditory temporal lobe, the ear contralateral to the lesion will be affected in dichotic listening tasks, as the contralateral pathway is the dominant pathway (Berlin, Lowe-Bell, Jannetta, & Kline, 1972). Importantly, in most cases of LKS, the left temporal lobe is the lesioned area; therefore, a right ear deficit on dichotic testing is expected. Conversely, left ear deficits have been sporadically reported in children with specific characteristics of (C)APD (Bellis, 2003). In fact, Moncrieff (2006) reported 84% of children with (C)APD had left ear dichotic listening deficits.

DIID training is an innovative therapy for the remediation of the compromised central auditory pathway (Musiek et al., 2007). DIID training utilizes dichotic listening tasks whereby the signal intensity presented to the unimpaired pathway is first decreased and then slowly increased over time as the weaker, impaired pathway grows stronger. DIID training purports to specifically target the deficit ear; thus, activating brain regions that receive auditory sensory input on the side of the lesion. Previous investigations have shown behavioral and electrophysiological evidence of improvement of the central auditory nervous system after DIID training (Hurley & Billiet, 2008; Musiek, Baran, & Shinn, 2004). For a complete review of the DIID procedure, see Musiek et al. (2007).

Case Study Report

"JP" was the product of a normal pregnancy and birth with all developmental milestones being appropriately reached until the age of three. At that time, it was noted by his parents that JP's speech and language skills began to decline. Initially, this decline in speech and language was attributed to sibling jealousy, as it coincided with the birth of a younger sibling. Because of the decline in speech and language performance, JP was sent for a hearing evaluation, which established normal peripheral hearing.

As JP had a history of otitis media, the lack of progression in speech and language was next related to his history of ear infections. Subsequently, autism and pervasive developmental disorder were also erroneously diagnosed. With the onset of seizure activity at the age of three and a half years, the diagnosis of LKS was made based on the characteristic LKS spiking EEG. Nocturnal seizure activity continued until JP was eleven years old even though anticonvulsants were prescribed. At age 11, JP had his first normal EEG.

JP has attended speech/language therapy and occupational therapy through early intervention programs, in order to address an expressive and receptive language delay and verbal apraxia. He sporadically uses sign language as needed when he experiences difficulty with word finding or speech production. He is currently receiving speech/language therapy at school two times per week, thirty minutes per session.

Prior to beginning of DIID training at LSU Health Sciences Center (LSUHSC), a comprehensive speech-language evaluation was completed. This assessment indicated a moderate to severe receptive and expressive language disorder characterized by moderately impaired receptive language skills, severely impaired expressive language skills, and severely impaired language memory skills. Expressive language skills were significantly weaker than receptive language skills. JP also presented with severely impaired articulation skills consistent with a diagnosis of verbal dyspraxia. Errors were characterized by oral scanning/groping during attempts to execute oral movements, vowel distortions, inability to perform oral diadochokinesis, inconsistent errors with multiple attempts at production, and increased errors with increased linguistic complexity. Speech intelligibility was fair in known contexts and fair to poor in unknown contexts. Fluency and voice were within normal limits, although JP did demonstrate inconsistent hyper-nasality. Attention and concentration skills were adequate with redirection to tasks, as needed.

Investigational Methods

Timeline. Our involvement with JP began when he was 12.5 years of age, as he was a subject in a study examining pre- and post-behavioral and electrophysiological measures after Fast ForWord® training. The eight week Fast ForWord® computer mediated program was provided to qualified students through his school system. Permission to participate was obtained from a parent according to the policies of the LSUHSC Institutional Review Board.

It is important to note this case study is a retrospective review, rather than prospective study. Behavioral tests of (C)APD and electrophysiological recordings, described in a later section, were administered pre- and post-DIID training. A post-language assessment was not completed after DIID training because JP did not receive language therapy at the LSUHSC clinic.

Approximately four weeks after JP completed the Fast ForWord® program, one of the authors (AH) began weekly DIID training sessions for one hour per week at the LSUHSC. Although JP demonstrates a severe language deficit, it was decided that JP would begin motor speech therapy to address verbal apraxia at the LSUHSC Speech and Language Clinic, as this was not addressed in language therapy at his school. He received DIID therapy for two semesters and attended a total of twenty-two sessions, with eight therapy sessions cancelled by JP's mother. Additional therapy sessions could not be scheduled, due to the patient's geographical distance from the clinic. JP continued to receive language therapy two times per week for thirty minutes per session at his school.

Peripheral auditory assessment. Audiometric air conduction thresholds were within normal limits (<15 dB HL), bilaterally. Immittance audiometry indicated normal tympanograms with ipsilateral and contralateral acoustic reflexes present at normal intensity levels. Transient evoked otoacoustic emissions (TEOAEs) were obtained using the ILO system (Version 5.0). TEOAEs were present (>3 dB) at all frequencies (0.8, 1.6, 2.4, 3.2, and 4.0 kHz), suggesting normal outer hair cell function. All of these tests indicate normal peripheral hearing.

Behavioral auditory processing assessment. Due to JP's severe verbal apraxia, speech intelligibility was significantly impaired; therefore, the test battery was limited to non-linguistic or low- linguistically-loaded tests. Therefore, the Dichotic Digits Test (DDT) was administered. In this test, two numbers (numbers one through nine, with exception of seven) are presented to the right ear and two numbers are presented to the left ear. The listener must repeat all four numbers. Forty pairs of numbers are presented and the test is scored on the percentage correct for each ear.

JP's responses to temporal resolution thresholds on the Random Gap Detection Test were inconsistent at every frequency. Each pure tone in the Random Gap Detection Test is 17 msec in duration with inter-stimulus intervals (gaps) of 0, 2, 5, 10, 20, 25, 30, and 40 msec randomly presented. For example, JP would respond that he heard two tones when the silent interval was 2 msec, but only one tone for tones with a larger silent interval, such as 10, 20, or 40 msec. A Three-Interval Forced Choice Gap Detection Test (3-IFCGD; Davis & Hurley, 2002) was administered. This test is a variation of the Random Gap Detection Test, and is used as a temporal resolution screening tool, when the listener has difficulty on the Random Gap Detection Test. In this test, three tones are presented with one of the tones having a silent interval that varies from 2 to 40 msecs in length. The listener must tell which tone in the series has the silent interval, by indicating "1, 2, or 3."

The Frequency Pattern Test (FPT) and Duration Pattern Test (DPT) were administered. These tests require auditory discrimination, temporal ordering, and pattern recognition and are similar in composition. Each tone in the FPT test is 200 msec in duration with a 10 msec rise-fall time. The inter-toneburst interval is 150 msec, with a 7 second inter-pattern interval.

For the FPT, three low- and high-frequency tones are presented to the listener, two are the same and one is different. The listener must repeat the pattern of the tones by verbalizing, for example, “low, low, high” or “high, low, low.” The frequency of the low tone is 880 Hz, and the frequency of the high tone is 1430 Hz.

For the DPT, the tones are either “short” (250 msec) or “long” (500 msec). Forty items are given for each test. The listener must respond by verbalizing the pattern, for example, “long, long, short” or “short, long, long.” The test is scored on the percentage correct for each ear.

Electrophysiologic Recordings

Electrophysiologic recordings were obtained while the subject rested comfortably in a reclined position and watched silent videos. These recordings were employed to assess the integrity of the CANS from the brainstem through the auditory cortex.

Auditory brainstem response (ABR). A one channel ABR was collected using the Bio-logic Navigator Pro System (Bio-logic Systems Corporation). Test stimuli consisted of 100 µsec condensation clicks with a rate of 27.7 /sec, presented at 80 dB nHL via insert ER3A earphones. Two stimulation sequences consisting of 2000 click presentations were recorded for each test condition.

Recordings were made with three surface electrodes attached to the skin at the vertex (non-inverting), and each ipsilateral mastoid (inverting). Electrode impedance was below 5000 Ohms. The response was amplified and filtered (bandpass 10-3000 Hz) and averaged over a 12 msec window. Artifact rejection was employed. Peak-to-following trough amplitude and latency of Waves I, III, and V were measured.

Auditory middle latency response (AMLR). The AMLR was obtained for each auditory pathway using the Nicolet Spirit evoked potential system. A three-channel recording was obtained with non-inverting electrodes located at Cz, C3, and C4 and the inverting electrode at the earlobe of the stimulus ear. Click stimuli at a rate of 6.7/sec were presented at 75 dB nHL. The time window for recording was 100 msec. The response was amplified and filtered (bandpass 5 - 100 Hz). Electrode impedance was below 5000 Ohms. A total of 500 click presentations for each run and four repetitions for each condition were obtained and added off- line. The amplitude of the Na-Pa wave complex was obtained for the summed waveform, as amplitude measures may be more sensitive than latency measurements (Chermak & Musiek, 1997; Kraus, Ozdamar, Hier, & Stein, 1982; Scherg & Von Cramon, 1986).

Auditory late evoked response (ALER)/P300. The ALER/P300 recording was obtained for each auditory pathway using the Nicolet Spirit system. A two-channel recording and an “odd-ball” paradigm were used, with the non-inverting electrode site being Cz and the inverting electrode being at the earlobe of the stimulated ear. The ‘frequent’ or ‘standard’ stimulus was a 500 Hz rarefaction toneburst; the ‘infrequent’ or ‘odd’ tone was a 2000 Hz rarefaction toneburst. Each toneburst had a 20 cycle plateau and a 5 cycle rise-fall. The time window for the recording was 750 msec. The response was amplified and filtered (bandpass 1-30 Hz). Electrode impedance was below 5000 Ohms. The software selection for oddball paradigm ratio was 80/20, indicating the frequent stimulus would be presented 80% of the time, and the rare tone would be presented 20% of the time. However, the actual responses collected were to a 75/25 paradigm. This error in the software should not have significantly affected the data, as actual percentages are very close to the ideal percentage paradigm (Hall, 1992). The same paradigm was used for all within-subject comparisons. Responses to approximately 215 “frequent” stimuli were averaged, and responses to approximately 70 “infrequent” stimuli were likewise averaged. Recordings were replicated and collected for the right and left ears.

BioMARK. Responses were collected using the Bio-logic Navigator Pro System (Bio-logic Systems Corporation). Responses were obtained for the right and left ears to an alternating polarity 80 dB SPL, 40 msec CV (da). The response was amplified and filtered (100-2000 Hz). Three thousand stimulus repetitions were collected at a rate of 11.1/sec. Two repeatable recordings were obtained and then added together for a grand average BioMARK response. The grand average is compared to a normative recording for an algorithmic, numeric score that can be interpreted from normal, borderline, to abnormal.

DIID Training

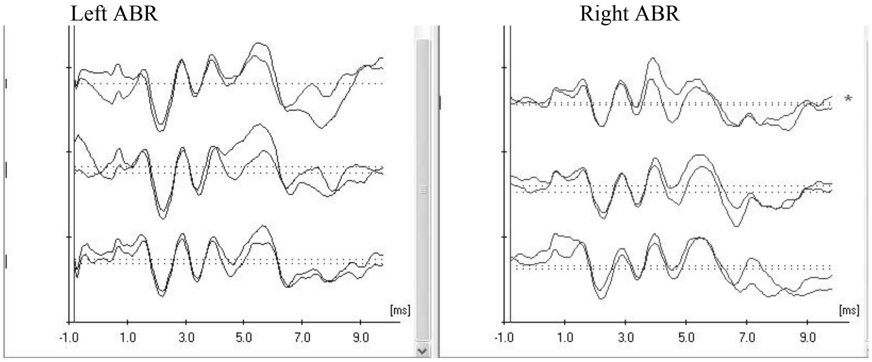
JP demonstrated a right ear deficit on the Dichotic Digits Test. In this training, dichotic materials are presented at different interaural intensities with a higher intensity directed to the poorer (right) ear and lower intensity directed to the better (left) ear. Initial presentation level was 55 dB HL to the right ear and 0 dB HL to the left ear. When the individual right ear score was >70%, the intensity level delivered to the right ear was increased by 1, 2, or 5 dB. This continued until there were equal intensities presented to the right and left ears. Motivation was difficult to maintain throughout the training sessions. Therefore, various dichotic listening materials, including digits, words, sentences, CVs and short stories were used in the training sessions.

At the beginning of training exercises, JP was instructed to ignore the signal (digits) in his left ear and repeat only the numbers that he heard in his right ear; thus, a directed listening task working

Table 1. Dichotic Training Progress Summary

	Left Intensity	Right Intensity	Right Binaural Separation Score
Pre Fast ForWord	55	55	0%
Post Fast ForWord	55	55	0%
DIID TRAINING			
Session 1	0 dB	55 dB	100%
	5 dB	55 dB	65%
	10 dB	55 dB	56%
	10 dB	55 dB	52%
	20 dB	55 dB	56%
Session 6	40 dB	55 dB	50%
	42 dB	55 dB	34%
	42 dB	55 dB	40%
Session 15	50 dB	55 dB	56%
	52 dB	55 dB	68%
	55 dB	55 dB	68%
Post DIID Training	55 dB	55 dB	94%

Figure 1. A normal auditory brainstem response (ABR) was recorded pre- and post-training.



on binaural separation. Subsequently, JP was required to listen and repeat numbers from both ears; thus, an exercise of binaural integration. In addition to intensity differences, dichotic digits that were separated in time (25, 50, 100, 200, 500 and 900 msec) were used to train binaural integration.

Results (Pre and Post): Behavioral Tests

During the initial assessment, JP could not repeat any numbers presented to the right ear on the DDT, but had a left ear score of 92%. The right ear deficit is not surprising since the contralateral left temporal lobe is the lesioned area with abnormal EEG spiking and presumed to be responsible for the "ear" deficit. The left ipsilateral pathway travels to the right cortex and theoretically may then cross to the left temporal lobe, where auditory language information is believed to be processed.

Post-testing after Fast ForWord® training showed no difference in the right ear score. After DIID training, individual right ear scores were consistently above 90%, when JP was instructed to ignore the left ear (binaural separation). JP's scores for the DDT test are shown in Table 1 as training progressed. The individual left ear scores remained within normal limits (>90%). A binaural integration deficit remained, which was demonstrated when JP was required to repeat all of the numbers from both ears. When repeating all numbers, JP's score for the left ear was consistently in the 80-90% range, while the right ear score stabilized in the 60-70% range.

Prior to Fast ForWord® training, normal temporal resolution was found as JP was able to discriminate tones with a 2 msec silent interval on the 3-IFCGDT. In addition, JP scored 100% for the individual right and left ears on the FPT and the DPT. JP's performance on these non-linguistic auditory processing tests indicates normal temporal processing ability. Therefore, additional testing after Fast ForWord® and DIID training was not of clinical value.

Results (Pre and Post): Electrophysiological Tests

ABR. A normal ABR was obtained for the right and left ears (see Figure 1). Wave latency and amplitude values are recorded in Tables 2 and

Table 2. Latency of the ABR in msec.

Ear	Wave	Pre-therapy Latency in msec	Post Fast ForWord Latency in msec	Post DIID Latency in msec
Left	I	1.62	1.57	1.57
Left	III	3.95	3.99	3.95
Left	V	5.74	5.74	5.65
Right	I	1.66	1.62	1.57
Right	III	3.91	3.99	3.99
Right	V	5.61	5.53	5.53

Table 3. Amplitude of the ABR in µV.

Ear	Wave	Pre-therapy Amplitude in µV	Post Fast ForWord Amplitude in µV	Post DIID Amplitude in µV
Left	I	.40	.41	.41
Left	III	.17	.24	.26
Left	V	.27	.27	.45
Right	I	.30	.30	.31
Right	III	.22	.22	.22
Right	V	.26	.26	.36

3. There was no difference in the ABR after Fast ForWord® or DIID training. The ABR is interpreted to be within normal limits. This is not surprising as the ABR may not be sensitive for patients with (C)APD (Hall, 1992).

AMLR. Pre- and post-AMLR recordings are shown in Figures 2a and 2b. Amplitude measures are shown in Table 4. Unfortunately, post-auricular muscle artifact occluded the C4 recording when stimulating the left ear. Greater amplitude was shown in the post recordings for electrode C3 when stimulating the left ear, and C4 when stimulating the right ear.

Auditory late evoked response (ALER) /P300. There was no discernable P300 response for either the right or left recordings. This might be due to our patient's lack of attention to the rare stimulus. Pre- and post-ALER responses for the "standard" tone are shown in Figure 3. Pre- and post-latency and amplitude N1-P2 amplitude measures are recorded in Table 5. An improvement in N1-P2 amplitude was noted for the right ear.

BioMARK. Pre- and post-BioMARK recordings are shown in Figure 4. The latency values of Waves V and A, and the V/A slope are shown in Table 6. A decrease in the latency of Waves V and A are noted after training. Initially, JP's BioMARK recording was abnormal for both the right and left ears. These recordings did improve after Fast ForWord®. Both the right and left BioMARK recordings are now within normal limits after DIID training, using age norms 5-12, as there are no normative data for children 13-17 years of age.

Discussion

This case presents an opportunity to assess objective changes in the auditory system after two auditory training programs. Many of the non-

Figure 2a. . Pre- auditory middle latency response (AMLR) recordings with left ear stimulation from Cz and C3 are shown in tracings 1 and 2, respectively. Recordings after Fast ForWord® training are shown in traces 3 and 4, and post-recordings after Dichotic Interaural Intensity Training (DIID) training are shown in tracings 5 and 6 for electrodes Cz and C3, respectively.

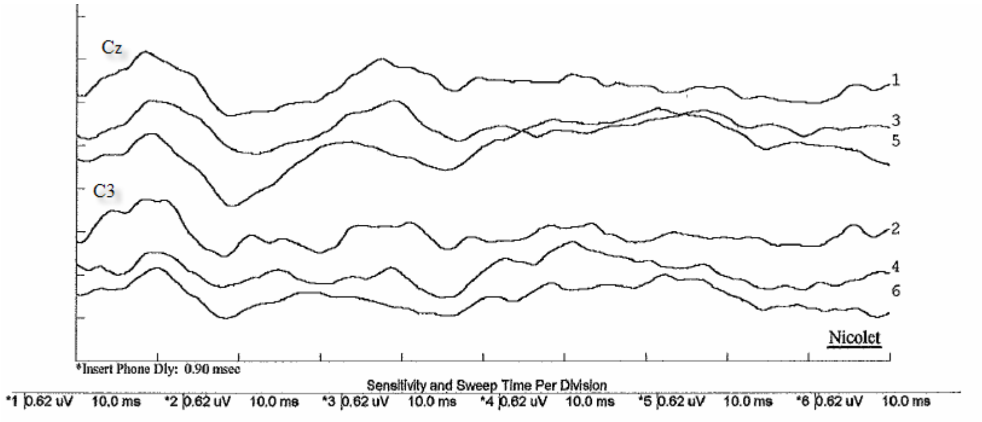


Figure 2b. Pre-auditory middle latency response (AMLR) recordings with right ear stimulation are shown for electrodes Cz, C3 and C4 in tracings 1, 2, and 3. Post-Fast ForWord® recordings are shown in tracings 4, 5 and 6, and recordings after Dichotic Interaural Intensity Difference (DIID) training are shown in tracings 7, 8 and 9.

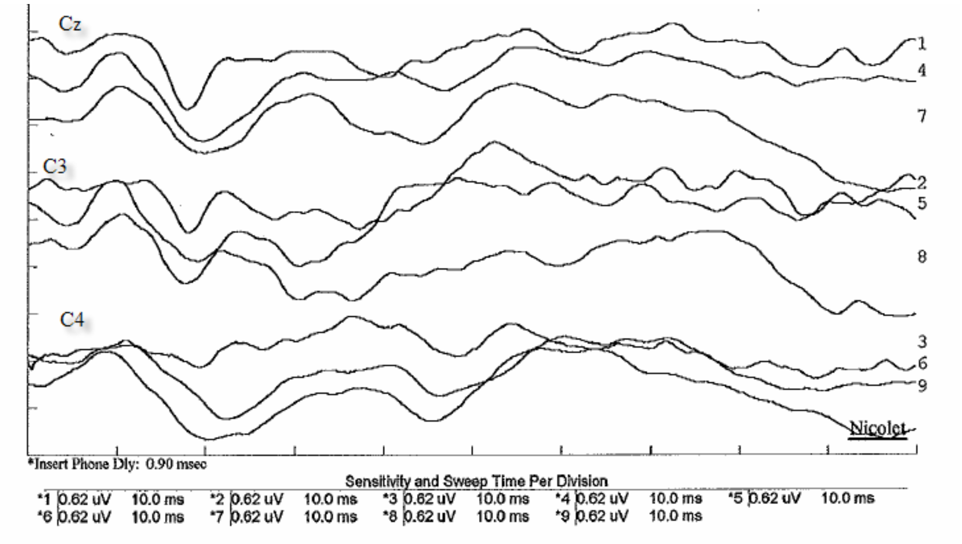


Table 4. Amplitude of the AMLR.

Ear	Electrode	Pre-therapy Na-Pa amplitude (µV)	Post-Fast ForWord Na-Pa amplitude (µV)	Post-DIID Na-Pa amplitude (µV)
Left	Cz	.82	.78	.93
Left	C3	.32	.11	.46
Right	Cz	.67	.86	.66
Right	C3	.54	.40	.44
Right	C4	.36	.64	.62

Figure 3. Pre- and post-Auditory Late Evoked Response (ALER) for the left and right ears are shown. Pre-recordings are shown in tracings 4 and 1, post-Fast ForWord® recordings are shown in tracings 5 and 2, and post-Dichotic Interaural Intensity Difference (DIID) training recordings are depicted in tracings 6 and 3.

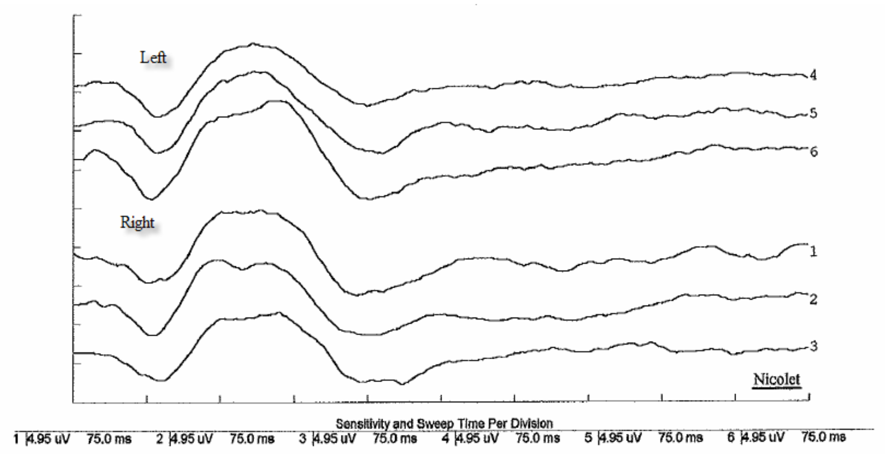
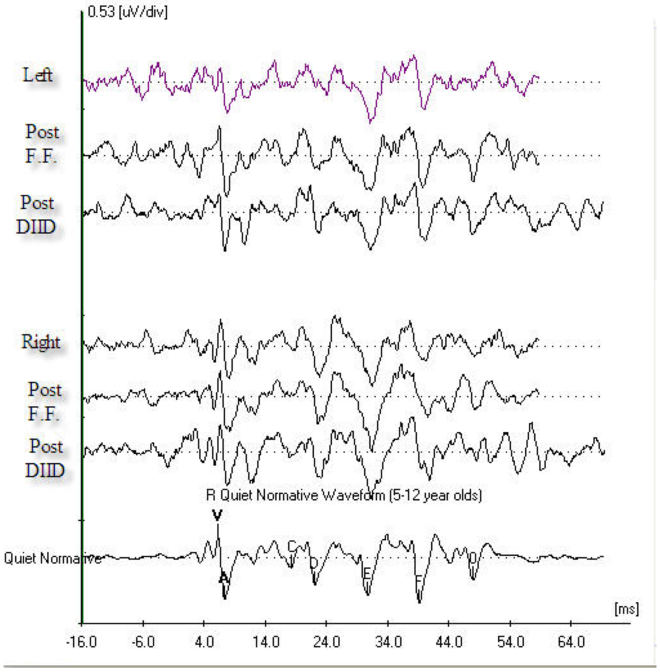


Table 5. Latency and Amplitude of the ALER.

Ear	Measure	Pre-therapy	Post-Fast ForWord	Post-DIID
Left	N1 Latency (in msec)	91.5	81	82.5
Left	P2 Latency (in msec)	172.5	174	169.5
Left	N1/P2 Amplitude (in µV)	8.61	8.64	8.14
Right	N1 Latency (in msec)	85.5	85.5	79.5
Right	P2 Latency (in msec)	181.5	182.5	177
Right	N1/P2 Amplitude (in µV)	9.55	10.57	11.35

Figure 4. Pre and Post BioMARK waves are depicted for the left and right ears. Also depicted is a normative waveform.



linguistic (C)APD tests were within normal limits and were not repeated after each auditory training program.

The right ear score for the DDT was initially abnormal and did not improve after Fast ForWord®. We have previously reported improvements in right ear scores for the DDT after Fast ForWord® training, but the improvement did not reach statistical significance (Hurley, Hurley, & Cook, 2007). The lack of improvement in this case may be due to the anatomically abnormal left temporal lobe (0% right ear score). On the other hand, after DIID training, the individual right ear score improved with binaural separation scores being consistently within the >90% range (although difficulty with binaural integration remains). Therefore, based on behavioral testing, the appropriate training to target the impaired auditory pathway was DIID training.

The improvements in the electrophysiologic recordings are objective evidence of plasticity or changes in the CANS after auditory training. The ABR is a test that reflects the auditory synchrony of the brainstem (Hall, 1992). The ABR was within normal limits for both pre- and post-recordings with latency and amplitude and showed no significant changes after Fast ForWord® training ($p > .05$). This is consistent with our previous reports of no changes in the ABR after Fast ForWord® (Hurley et al., 2007; Hurley, Hurley, & Homer, 2008).

AMLR is useful in assessing plasticity of the central auditory nervous system, as previous investigators have reported increased amplitude of the Na-Pa complex after subjects have completed

Table 6. Latency of Waves V and O for the BioMARK recording.

Measure	EAR	Pre-therapy	Post-Fast ForWord	Post-DIID
BioMARK Algorithm	Left	12	10	*3
Wave V latency (in msec)	Left	6.78	6.62	6.53
Wave A latency (in msec)	Left	7.83	7.70	7.39
BioMARK Algorithm	Right	15	11	*6
Wave V latency (in msec)	Right	6.87	6.70	6.70
Wave A latency (in msec)	Right	8.03	7.70	7.70

Note: *Denotes within normal limits.

an intensive auditory training program (Morlet, Norman, Ray, & Berlin, 2003; Musiek et al., 2004). Greater Na-Pa amplitude was recorded from the C4 electrode when stimulating the right ear after Fast ForWord® and DIID training, but not for the Cz and C3 electrodes. The Na-Pa amplitude at C4 increased from 0.36µV to 0.62µV - an improvement considered to be clinically significant (Musiek, Charlette, Kelly, Lee, & Musiek, 1999). There was no significance difference in the amplitude of Na-Pa with left ear stimulation for the Cz and C3 electrodes. Unfortunately, a post-auricular muscle artifact obscured the analysis of the C4 responses for left ear stimulation.

The ALERs have been employed to assess auditory plasticity and reflect neural activity of the auditory cortex (Hall, 1992). Investigations have shown significant amplitude changes in the N1-P2 complex after auditory training and attribute this enhanced amplitude to changes in neural activity (Tremblay, Kraus, McGee, Ponton, & Otis, 2001; Tremblay & Kraus, 2002). Previously we have reported a decrease in N1 and P2 latencies and an increase in N1-P2 amplitude in a group of subjects after Fast ForWord® training (Hurley et al., 2007; Hurley et al., 2008). In JP's recording, greater amplitude in the N1-P2 complex for the right ear was seen after Fast ForWord®, and continued to improve after DIID training, suggesting an improvement in JP's "lesioned" area of the brain. No significant change was seen in the recordings when stimulating the left ear.

The BioMARK is a neurophysiological test assessing auditory neural timing for a speech stimulus. Previous research has shown that approximately 30% of individuals with a language, reading, or learning disorder will have an abnormal BioMARK recording (Banai, Nicol, Zecker, & Kraus, 2005). Previous research has shown an improvement in the BioMARK recording after auditory training (Hurley et al., 2008; Russo, Nicol, Zecker, Hayes, & Kraus, 2005). For JP, the latency of Waves V and A for both the right and left ears decreased after Fast ForWord® training and continued to decrease after DIID training. The BioMARK algorithm was abnormal pre auditory training, and progressed to within normal limits after DIID training. Again, it is important to note that this normative data was based upon the age norms provided for ages 5-12.

Summary

The improvement of the Na-Pa amplitude for the C4 electrode for the AMLR recordings, the improvement of the N1-P2 amplitude for the right ear, and the improvement of the BioMARK recordings provide evidence for plasticity of the CANS. However, it is not possible in this retrospective review to separate the effects of each language therapy, motor training, DIID training, Fast ForWord® training, maturation, or a possible synergistic effect of all therapies on the post-test results demonstrated by JP. Furthermore, we believe that it would have been unethical to withhold any of the other therapies during the time JP was receiving Fast ForWord® and DIID training.

In addition to objective evidence of improvement in the auditory function, antidotal evidence (provided by unsolicited parental reports) was positive, as were reports from extended family members and JP's teachers. Each commented that JP's speech and language were improving and that he was speaking in complete sentences that conveyed organized thoughts, rather than in his previous telegraphic-type speech. They also reported that, post-training, JP rarely used sign language to communicate.

Unfortunately, there has been limited research on the auditory processing deficits in children with LKS or the success or failure of remedial therapies. This case study demonstrates positive changes in auditory processing ability after two distinct auditory training programs and the effectiveness of a deficit-specific auditory training program (DIID). We do not contend that we have 'cured' JP of all academic difficulty. JP has a severe language disorder and continues to receive therapy at his school. He will participate in additional Fast ForWord® training programs offered through his school in the future. Future controlled investigations incorporating language measures for the treatment of children with LKS would be beneficial.

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