Auditory Processing Deficits in a Teenager with Landau–Kleffner Syndrome

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Abstract

This study explores the auditory processing skills of a 14-year-old boy (William) with an acquired aphasia, diagnosed as Landau–Kleffner syndrome. A single case study design was implemented, with the use of both chronological age-matched and receptive-language-matched control participants. Tasks were designed to investigate non-linguistic and linguistic auditory processing. The results indicated that William had intact peripheral hearing and gap detection abilities. However, William was significantly impaired at detecting a tone presented before or during some masking noise. William also showed significant impairments in three auditory processing tasks, requiring discrimination of both word and non-word stimuli. Findings suggest that William has an auditory processing deficit affecting the perception and discrimination of linguistic and some non-linguistic stimuli.

Introduction

Landau–Kleffner syndrome (LKS) is a rare acquired paediatric language disorder. It is also referred to as 'acquired aphasia with convulsive disorder' and 'acquired receptive aphasia' (Paquier *et al.*, 1992; Lees, 1993). In addition to a language deficit, the syndrome is associated with abnormal EEG results and, sometimes, seizures. The clinical presentation is one of normal development, interrupted by the loss of language skills. The child often appears to have a hearing loss, with a reduced response to speech and environmental sounds (McAllister and Greathead, 1991). Pure-tone audiogram and auditory brain stem responses (ABR) are reported to be normal (Paquier and van Dongen, 1993). Symptoms may suggest an auditory verbal agnosia, with accompanying deterioration in expressive language.

Neurological investigations of cases with the syndrome have not found any consistent lesion site (Gordon, 1990; Deonna, 1991). PET scan and SPECT studies, measuring cerebral blood flow, have revealed metabolic disturbance over the temporal lobes (Maquet *et al.*, 1990; DaSilva and Chugani, 1995; Intenzo *et al.*, 1996). These metabolic disturbances have been found to be long term, persisting after recovery from epilepsy (Metz-Lutz *et al.*, 1996). EEG investigations reveal a characteristic bilateral spike wave pattern during sleep, with the focus of discharge in the temporal and/or parietal regions (Deonna, 1991). Magneto-encephalography (MEG) studies have allowed more precise

location of epileptiform discharges. These locate the source of the activity in the superior temporal gyri and sylvian fissure (Morrell and Lewine, 1994; Paetau, 1994; Morrell *et al.*, 1995). Klein *et al.* (1995) describe 'an abnormality within the secondary auditory cortex in the lateral surface of the temporal lobes' (p. 383) observed in cortical event related potentials in young adults. This range of findings implicates temporal lobe and auditory cortex dysfunction in LKS. Kolb and Wishaw (1990) suggest agnosia for sounds may arise from bilateral temporal lobe damage, and studies suggest that even where epileptiform discharges are unilateral, they also seem to suppress contralateral auditory function (Paetau *et al.,* 1991).

Paetau (1994) found that 1 kHz, 50 ms tones, delivered at 80 dB SPL, triggered spikes identical to spontaneously occurring spiking activity in three of six participants with LKS, and these were cases who still had spontaneous epileptiform activity. Paetau (1994) hypothesizes that these triggered spikes are 'disinhibited evoked responses of auditory-related cortical areas' and suggests the epileptiform activity may be produced by sound-responsive neurons in the auditory cortex. In contrast, Boyd *et al.* (1996) found that sounds did not evoke such spikes in the LKS case they presented. Further investigation of Paetau's (1994) finding is required, as it has implications for the management of LKS cases. In particular, clarification would be needed for whether all sound triggers

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spiking activity or only certain sounds, and in which cases such activity occurs. In practice, most individuals with LKS do show some improvement in their ability to recognize sounds over time, and epileptiform activity reduces and eventually ceases as the child matures, despite being exposed to speech and other environmental sound.

Auditory processing deficits in Landau–Kleffner syndrome

A series of case studies have illustrated some aspects of the nature of the auditory processing difficulties in LKS. Denes *et al.* (1986) reported the case of CS, who, at age 11 years, had a severe auditory comprehension deficit, although pure-tone audiometry and auditory evoked potentials were within normal limits. He recognized environmental sounds. However, CS was impaired in his performance on a range of tasks using linguistic stimuli, including same/different judgement of syllables and words, auditory lexical decision, auditory lexical decision with picture support and picture identification. His best performance was in the last two tasks where support for discrimination was available from pictures.

An LKS case presented by Zardini *et al.* (1995) showed changes in auditory discrimination abilities over a 3 year period. At 12 years of age, this individual had marked difficulties in detecting whether CV syllables were the same or different and was 80% correct in responding to a picture-pointing auditory discrimination task. By 15 years of age, performance had improved to 95% correct in discriminating syllables and 100% correct in responding to the picture-pointing task. However, he continued to be impaired in his understanding of complex sentences and verbal production of language was at a 6–7 year level. EEG investigations showed a left mid-temporal focus for spiking activity.

A 27-year-old woman with LKS was investigated by Baynes *et al.* (1998). PET scanning revealed bilateral temporal lobe hypometabolism. Recognition of some environmental sounds, and same/different judgement of pitch and duration of tones, were impaired, suggesting that auditory processing difficulties were not specific to speech. She also showed difficulties with minimal pair word discrimination in making same/different judgements, although vowel changes, which would be longer in duration, were easier to detect than consonant changes. Changes in word final consonants were the most difficult to detect, leading Baynes *et al.* (1998) to suggest that speech processing was easily 'disrupted by the sound that comes before'.

Stefanatos (1993) found deficits in steady-state cortical evoked responses to frequency modulated tones in cases of LKS, but not in children with specific language impairment (SLI) or normal language development. Stefanatos (1993) concludes that LKS results in a disturbance of the auditory analysis of acoustical features necessary for speech perception, and that this underlies the individual's auditory language comprehension difficulties. Responses to frequency modu-

lated tones were depressed bilaterally, supporting the clinical view of bitemporal involvement in LKS.

Case report

The case presented here has previously been reported by Vance (1991, 1997). William had a history of normal development until age 3:6 when his speech and language deteriorated unexpectedly. He no longer responded to environmental sounds or speech and became mute. At age 4:2, speech reappeared in the form of unintelligible jargon, but he was unable to comprehend a spoken language assessment and relied solely on non-verbal communication. William passed a free field distraction hearing test and EEG recordings showed evidence of characteristic spiking, prompting a diagnosis of LKS. He received appropriate drug treatment for a short period of time. Further EEG investigations at age 6:9 showed no signs of a focal lesion or any paroxysmal features in sleeping state, and MRI scan of the brain was entirely normal. William has been in specialist educational provision since the age of 5:2. Teaching and speech and language therapy have focused on modelling language through the visual channel (Vance, 1991, 1997). Meaningful verbal expression and comprehension skills began to re-emerge at the age of 6 years. At the time of the current study, William was 14:6. He could communicate effectively with spoken language, but continued to show residual symptoms of speech and language difficulty with verbal comprehension still affected.

This study aims to explore William's auditory processing skills and to identify the levels of breakdown in auditory processing that underlie his continued speech and language difficulties. Use is made of a psycholinguistic assessment framework for the investigation of speech and language difficulties (Stackhouse and Wells, 1997). This delineates between bottom-up input processing skills that require analysis of the speech signal and top-down speech processing in which stored linguistic knowledge (e.g. lexical representations) influences discrimination and perception of the speech signal. Stackhouse and Wells (1997) advocate the comparison of performance on a range of auditory tasks to allow level(s) of an auditory processing deficit to be identified. Deficits may potentially arise at a level of peripheral, or non-linguistic, processing (discrimination of non-speech sounds), in the discrimination of non-lexical speech stimuli (i.e. non-words), discrimination of lexical speech stimuli (i.e. words), or in the accuracy of phonological representations in the lexicon (as required for lexical decision or picture-pointing auditory discrimination). However, a deficit at one level may have consequences for processing at other levels.

Tasks were selected to investigate linguistic auditory processing, involving stimuli which are used systematically to convey a message (i.e. speech sounds), and non-linguistic auditory processing, the perception of non-speech sounds (i.e. tones and noise). Use of non-linguistic stimuli investigates whether any auditory processing deficits are specific to the

identification of speech or not. The use of non-words for auditory discrimination allows the investigation of William's ability to discriminate speech stimuli without support from the lexicon. The use of word stimuli will determine whether existing lexical knowledge can support discrimination. Auditory discrimination tasks using picture material allows the accuracy of William's phonological representations to be investigated as he will have to compare the speech heard with his own representations in order to respond appropriately.

Tasks that have been used to investigate the non-linguistic auditory processing deficits of children with SLI were included in this study. Tallal and Piercy (1973) found SLI children to be impaired at identifying two tones and detecting whether two tones were the same or not, when the gap between them was brief. Lincoln *et al.* (1992) found that impaired performance persisted into adolescence and young adulthood, although this was only clear for sequences of three or more tones. Recent research has indicated that SLI children have difficulty 'separating a brief sound from a rapidly following sound of similar frequency' (backward masking; Wright *et al.*, 1997, p. 177). Wright *et al.* (1997) also found that SLI children could not use the difference in frequency content between a brief tone and a longer co-occurring or preceding masking sound to aid its detection, suggesting that there may be a spectral component to their perceptual deficit. Poorer performance has also been found in auditory fusion for children with reading and learning difficulties (McCroskey and Kidder, 1980). Here we chose to measure listener's abilities for such fine temporal acuity using a closely related gap detection task, to avoid the possibility of response bias inherent in the one-interval fusion procedure.

It is hypothesized that (i) whilst having an intact peripheral hearing mechanism, William will be impaired in some aspects of non-linguistic auditory processing, and (ii) William will perform significantly less well than receptive-language-matched controls on linguistic auditory processing tasks.

Phase 1 investigations

Method

The first phase of the study assessed William's current audiological and speech and language skills.

1. *Standard audiometric testing.* Pure-tone audiometry, acoustic reflex and ABR testing was carried out by an audiological scientist.

2. *Same/Different Speech-Perception Task.* In this computer-controlled task, two words were presented over headphones on each trial, and the listener was required to indicate whether they were the same or different. At least one word of the six test word pairs contained an onset consonant cluster. The other word in the pair was created by second consonant deletion (e.g. blow/bow) or substitution (e.g. scar/star). The items were specifically chosen to be maximally difficult to differentiate, on the basis of results from Adlard

and Hazan (1998). The test items were presented at a normal speaking rate and in a lengthened condition using the SOLA technique (Roucos and Wilgus, 1985) to double their duration. Practice and control pairs were also included. Each pair occurred twice as a 'same' and four times as a 'different' pair. Two tokens of each word were recorded (in an anechoic chamber by a female speaker of Southern British English) so that 'same' pairs were not physically identical. The listener made his same/different judgement by clicking with the mouse on one of two computer buttons. A 'same' response was indicated by a box of two red circles, whilst a 'different' response was represented by a picture of a red circle and a green square.

3. *Auditory Discrimination and Attention Test (Morgan-Barry, 1988).* A standardized minimal pair identification task was administered.

4. *Clinical Evaluation of Language Fundamentals – Revised (CELF- R) (UK) (Semel et al., 1987).* This standardized assessment of receptive and expressive language skills was administered.

5. *Speech Sample.* Phonetic transcription of William's responses in a conversation and a discussion of the 'What's Wrong?' pictures (Winslow Press, 1994) provided data for an analysis of William's speech errors.

Results

1. *Audiological investigations.* These revealed that William's pure-tone audiometry, acoustic reflexes and ABR were normal.

2. *Same/Different Speech Perception Task.* William correctly judged all of the practice and control minimal pairs as same or different. Of the test stimuli, William responded correctly to 69.4% of the word pairs presented at a normal speaking rate and to 80.6% of the lengthened stimuli.

3. *Auditory Discrimination and Attention Test.* William's performance yielded a *z* score of –1.4 when compared to normative data for 11 year olds (the oldest group for whom such data were available). This performance was, therefore, below normal limits for his age.

4. *Clinical Evaluation of Language Fundamentals – Revised (UK).* The following standard scores were obtained: Receptive Language Score: 83; Expressive Language Score: 64; Total Language Score: 72; Age equivalent: 8 years 8 months. William continues to have significant difficulties with receptive and expressive language.

5. *Speech Sample.* Analysis of the speech sample revealed that William continues to make some speech errors. These included some phoneme substitutions, e.g. /t/ and /d/ used for /k/ and /g/, weak syllable deletion, final consonant deletion and cluster reduction.

The difficulty with initial clusters was also evident in William's auditory discrimination in the Same/Different Speech Perception Task (2). Other errors occurring in William's speech were not represented in the stimuli for the Auditory Discrimination and Attention Test (3).

Phase 2 investigations

The second phase of the study compared William's performance on some experimental non-linguistic and linguistic auditory processing tasks with that of control participants.

Method

Control group participants. Written consent was obtained for all participants in the study. Two groups of normally developing children were invited to take part in this study. Selection criteria were: no previous history, or apparent, hearing, speech, language, literacy, emotional or behavioural difficulties; English as a first language. A group of 10 age-matched controls were aged between 12 and 16 years (with a mean age of 13:3). They were recruited from the families of university staff and students. A group of 10 receptive-language-matched controls were aged between 7 and 9 years (mean age 8:8) and on the CELF-R (UK) (Semel *et al.*, 1987) obtained a receptive language score within the normal range for 8 year olds (i.e. the standardized receptive language score fell between 91 and 117). They were recruited from classes within mainstream primary schools.

The age-matched control group provided a comparison for William's performance on some non-linguistic auditory tasks, to examine whether his non-linguistic sound processing skills were age appropriate or not. The receptive-language-matched control group provided a comparison for William's performance on linguistic auditory processing tasks. This allowed investigation of whether William's receptive language skills and speech discrimination skills were developing in line with each other, or whether he was experiencing persisting difficulties with speech discrimination.

Procedure

1. *Non-linguistic auditory processing tasks.* These tasks were presented to William and to the age-matched control group. They included identifying two tones presented sequentially and detecting whether they were the same or not (Tallal, 1980); backward and simultaneous masking (Wright *et al.*, 1997), and gap detection. An IBM-compatible PC controlled all aspects of the experiments, including sound generation, sound presentation, timing and response collection. All tasks were administered in a soundproofed room through Sennheiser HD 414 headphones. Full details can be found in Rosen *et al.* (1997).

(a) Two Tone Identification and Same/Different Discrimination. These tasks were closely modelled on the stimuli and procedure used by Tallal (1980), with minor differences. The two stimuli were 75 ms complex tones generated by software synthesis, differing only in fundamental frequency (100 and 305 Hz). Each trial consisted of one of the four possible pairings of the two tones (low–low, high–high, low–high, high–low) with varying interstimulus intervals, presented diotically. Responses were made using a computer

mouse. The listener was required to (i) identify which two tones were heard with two button presses or (ii) discriminate whether the stimulus pairs were the same or different. Listeners were first trained to criterion for each task with an interstimulus interval of 428 ms. For test items, there were six interstimulus intervals, varying between 8 and 305 ms. Each of the four orders were paired with each of the six interstimulus intervals to make a total of 24 trials, presented in a random order without feedback.

(b) Gap Detection. Stimuli consisted of two 100 ms Gaussian noise bursts (generated with a sampling frequency of 50 kHz, and rise/fall times of 0.2 ms) between which a gap was inserted, or not. Gap durations were between 0.5 and 15 ms. For each trial, two bursts of noise were presented, one containing a gap. Sounds were presented monaurally. The order of presentation was to right ear, left ear, left ear then right ear, to control for practice effects. Participants used a mouse to click one of two response boxes on the computer screen to indicate which noise burst contained a gap. Starting with a 7 ms gap, an adaptive procedure used a geometric spacing of gap durations to track performance at the level of 79% correct.

(c) Backward and Simultaneous Masking. These tasks replicated the stimuli and procedure used by Wright *et al.* (1997), with minor modifications to the tracking procedure. Stimuli were generated digitally and presented monaurally to the right ear. Participants responded by pressing buttons on a response box to indicate whether a sound signal had occurred in or before the first or second of two 300 ms noise bursts of masking noise with a spectrum level of 40 dB. Masking noises were either bandpass (0.6–1.4 kHz) or notched (0.4–0.8 kHz and 1.2–1.6 kHz). The signal was a 1 kHz tone. A short signal (20 ms) was presented either before the onset of the masker (backward masking condition) or simultaneously with the masker, 200 ms after its onset (simultaneous masking condition). The intensity of the signal was manipulated to find thresholds at which the participant could accurately detect the signal in the masking noise 90% of the time. The order of presentation was signal with simultaneous masking followed by signal with backward masking.

2. *Linguistic auditory processing tasks.* Three linguistic auditory processing tasks were developed. These tasks were completed by William and the receptive-language-matched control group. Verbal stimuli were recorded onto chrome cassette tapes, in a soundproofed room, using a Marantz CP230/CP430 stereo cassette recorder and a Panasonic Dynamic Microphone RP-VK60. The stimuli were presented to subjects using the same cassette recorder. Stimuli for each task were randomly allocated to one of three lists, one list from each task being presented on each of three testing sessions.

(a) Word Discrimination. Forty-six word pairs that reflected some of William's speech errors were selected (see Appendix 1). Contrasts included /s/ + plosive clusters (e.g. <u>stick – sick</u> and <u>mask – mass</u>). There were 27 different-word pairs and

19 same-word pairs The word pairs were recorded with a 2 s pause between each word. Participants were asked whether the two words sounded the same or different. Three practice items were used, with feedback provided.

(b) Non-word Discrimination. Each word pair was used to create a phonologically matched non-word minimal pair, by substituting the vowel in each word, to provide 46 non-word pairs (see Appendix 2), e.g. <u>stick – sick</u> became /stek – sek/. These were recorded with a 2 s pause between each item. The same procedure was followed as for word discrimination, except that participants were told that the words would sound silly as they were made up.

(c) Auditory Lexical Decision with Picture Support. Twenty-two words with initial or final /s/ clusters or fricatives were chosen that reflected the pattern of speech errors found in William's speech. An incorrect pronunciation was created for each word by substituting the onset or final phoneme (see Appendix 3), e.g. <u>skate</u> and /steɪt/. The total of 44 items were allocated randomly to one of three groups and recorded. Pictures were drawn with black ink onto 14×9 cm postcards, duplicated and arranged in the same order as the word stimuli recorded on the tape. The pictures were initially presented for naming to verify that each participant recognized them. Participants were then asked to judge whether the tester had named each picture correctly as each item was played back from the tape. Three practice items were given, with feedback, after which the recorded test items were presented.

Results

1. *Non-linguistic auditory processing tasks.* (a) Two Tone Identification and Same/Different Discrimination. William reached the training criterion for both tone identification and discrimination tasks in the minimum number of trials. He correctly discriminated all but one of the tone pairs as being the same or different. In his first attempt at the identification task, William made one error in each of the four trials at interstimulus intervals of 8–150 ms. Re-training to criterion at an interstimulus interval of 428 ms was carried out, and when William was retested, he made no errors. These stimuli were not presented to the age-matched controls as William's performance showed no evidence of impairment (although ceiling effects may have precluded this possibility).

(b) Gap Detection Task. William's performance was compared to that of the age-matched control group by calculating a z score for each condition (see Table 1). The z scores indicated that for the first condition in which the stimuli were presented to the right ear, William required a significantly longer gap, as compared to the controls. On subsequent conditions, including the second presentation to the right ear, William performed as well as the control group. Interestingly, a recent study by Schulte-Korne *et al.* (1998) found no deficits in gap detection for dyslexic children or adults.

(c) Backward and Simultaneous Masking. For each of the masking conditions, thresholds were accepted as long as at least two measures were within a 10 dB range. William's

Table 1. Performance on gap detection task for LKS subject and age-matched controls

Condition	Mean duration of gap detected in ms (SD)		William's z score
	William	Control group	
1. Right ear	6.0	3.1 (0.93)	3.12 ($P < 0.0013$)
2. Left ear	2.4	3.1 (0.89)	−0.75 ($P > 0.05$)
3. Left ear	3.4	3.2 (1.00)	0.16 ($P > 0.05$)
4. Right ear	4.3	3.1 (0.92)	1.24 ($P > 0.05$)

performance was compared to that of the age-matched control group by calculating a z score for each set of stimuli. The z scores indicated that William was significantly poorer at detecting the signals in all masking conditions than the control group (see Table 2). In fact, his thresholds were higher than every control listener in each of the three conditions.

2. *Linguistic auditory processing tasks.* William's performance on the three linguistic auditory processing tasks was compared to that of the language–age-matched control group by calculating a z score for each task (see Table 3). For the word and non-word discrimination tasks and the auditory lexical decision task with picture support, William discriminated the stimuli significantly less well than the control group. The control group performance was near ceiling. However, William's scores for all three tasks fell well below the lowest score obtained by any control group participant, as shown in the range of scores (see Table 3).

(a) In the Word Discrimination Task, William made 10 errors. He incorrectly judged eight different word pairs to be the same, and two same pairs to be different.

(b) In the Non-word Discrimination Task, William made 15 errors. He incorrectly judged 10 different word pairs to be the same, and five same pairs to be different.

(c) In the Auditory Lexical Decision Task with picture support, William made 16 errors. He accepted 13 inaccurate pronunciations of the words as being correct and three accurate pronunciations as being incorrect.

Discussion

Non-linguistic auditory processing

Pure-tone audiometry, ABR and acoustic reflex responses indicate that William has intact peripheral hearing, at least up to the brain stem. Auditory processing, as reflected in the tone identification, same/different judgements and gap detection tasks, also appears to be intact. However, for the tone identification task, it was found that William benefited from additional trials, improving performance at identifying the two tones at shorter interstimulus intervals. Tomblin and Quinn (1983) also report improved performance with further practice on this task in 5- to 6-year-old children. This finding

Table 2. Performance on masking tasks for LKS subject and age-matched controls

Masking condition	Masking noise type	Mean minimum intensity level for detection (dB-SPL) (SD)		William's z score
		William	Control group	
Backward	Bandpass noise	62.4	48.6 (5.71)	2.42 (P = 0.008)
Simultaneous	Bandpass noise	84.1	79.1 (2.88)	1.74 (P = 0.041)
Simultaneous	Notched noise	70.4	56.9 (3.53)	3.82 (P < 0.001)
Simultaneous	Difference between bandpass and notched	13.7	22.2 (5.78)	1.47 (P = 0.071)

Table 3. Number of correct responses on linguistic auditory processing tasks by LKS subject and receptive-language-matched controls

Task	No. of items	William's no. correct	Controls' no. correct			William's z score
			Mean	SD	Range	
Word discrimination	46	36	45.2	1.5	42–46	−6.23 (P < 0.0005)
Non-word discrimination	46	31	43.7	2.72	39–46	−4.55 (P < 0.0005)
Auditory lexical decision with picture support	44	28	43.4	0.84	42–44	−18.27 (P < 0.0005)

leads them to suggest that the task is a measure of perceptual learning rather than of the temporal resolution of auditory signals, with normally developing children needing fewer learning sets than SLI to reach maximum performance.

William performed as well as an age-matched control group in detecting minute gaps in noise bursts presented to his left ear and to the second presentation to the right ear. He was significantly impaired at the detection of the gap in the noise bursts for the first condition, presented to his right ear first. The reliability of this result is almost certainly undermined by the fact that there was no practice trial for this task and the initial difference in performance in the right ear condition may therefore reflect lack of practice rather than a breakdown in right ear stimulus individuation ability. Improved performance on re-testing in the tone identification task and the marked improvement in gap detection for the second right ear presentation may also indicate that William requires more practice to 'tune' in to the processing demands of these auditory tasks, as compared to the control group.

William showed a significantly greater effect of backward and simultaneous masking as compared to the age-matched control group. He required a higher signal intensity for detection in all bandpass noise conditions, where the noise burst contained energy in the frequency region of the signal, as was found for Wright *et al.*'s (1997) SLI group. However, William's performance was not as impaired as that of Wright's SLI group. The SLI group reported by Wright *et al.* (1997) had more difficulty detecting the tone presented before the noise (backward masking—mean signal intensity detection level 89 dB SPL), rather than with the noise (simultaneous masking—mean signal intensity detection level 83 dB SPL). In contrast, William was better able to detect the tone when it was presented before the noise (backward masking—mean signal intensity detection level about 62 dB SPL) rather than with the noise (simultaneous masking—mean signal intensity detection level about 77 dB).

Wright *et al.* (1997) also found that the mean threshold difference between the notched and bandpass conditions was smaller for the SLI group than for the controls (11 dB compared to 19 dB on the simultaneous masking task). The control group in this study demonstrated a 22 dB difference between the mean short signal detection levels in the notched and bandpass simultaneous masking tasks, whereas William only exhibited a 13.7 dB difference. Therefore, not only does William find it harder to detect a signal in noise that has spectral energy in the frequency region of the signal, he is somewhat less able than the control group to take advantage of the difference in frequency spectrum between a short signal and its noise environment to support detection. (One control listener showed a smaller difference than this at 13.5 dB.) This suggests that William has impaired auditory spectral processing. Unfortunately, we do not know whether this would also hold true for long tones, which are typically used in studies of frequency analysis, or whether such an effect is specific to the use of short tones.

Susceptibility to non-simultaneous masking effects may explain some aspects of William's difficulty in processing speech stimuli. Discrimination of fricative + plosive + vowel onset pairs, e.g. star – scar, may be impaired as the plosive is masked by the surrounding sounds. This could explain his difficulty in determining the place contrasts of the plosive in the consonant clusters, as these plosive place contrasts are revealed in the short time before the vowel, by the second formant transition and the frequency of the burst relative to the vowel (Borden and Harris, 1984; Perkins and Kent, 1986). We would hypothesize that such masking effects would also affect discrimination of plosive + vowel onset pairs, e.g. key – tea, and discrimination of final vowel +

plosive + fricative pairs, e.g. oats – oaks, and that William will find it more difficult to process speech in a noisy environment.

Linguistic auditory processing

The non-word discrimination task assessed William's ability to perceive and compare speech stimuli without support from his lexicon. William showed significant difficulty with this task, as compared to language–age-matched controls. The word discrimination task attempted to determine whether the use of existing lexical knowledge (in particular phonological representations) would support his discrimination. William was significantly less able to make accurate discriminations as compared to the language–age-matched control group.

Auditory discrimination of word minimal pairs was also investigated using the Same/Different Speech Perceptual Task. William was impaired at discriminating word minimal pairs (with initial clusters) when they were presented at a normal speaking rate. His discrimination skills were improved by lengthening the duration of the words. This improvement in performance may have been the result of a reduction of the masking effects present for the normal length pairs, and/or the increase in processing time reducing the auditory processing demands. Lengthening speech stimuli has also been shown to improve the speech perceptual capabilities of SLI listeners (Tallal *et al.*, 1996), whilst normal children (aged 5–12 years) showed no advantage of lengthening even in background noise (Walker, 1998). Improved performance with lengthened stimuli may, therefore, be evidence for an auditory processing deficit.

The accuracy of William's phonological representations was initially investigated using the Auditory Discrimination and Attention Test (Morgan-Barry, 1988). The recognition of the word spoken relied on a comparison being made between the word spoken and William's own phonological representations to create a match. William was impaired in this task as compared to the normative data. The auditory lexical decision task with picture support also relies on accurate phonological representations for correct judgements, as again the spoken stimuli must be compared with the listener's own phonological representations. William demonstrated a significant difficulty with this task, as compared to the language–age-matched controls. He also performed markedly more poorly on this task than the word and non-word discrimination tasks, suggesting that stimuli are not making the expected match or mis-match with his phonological representations, which may be inaccurate or insufficiently specified. Given that the stimuli selected reflected the type of errors in William's own speech production, the findings suggest that some of his speech production errors could be arising at the level of inaccurate central representations of words [see Vance (1997) for further discussion of this point]. Ongoing auditory processing difficulties, from the onset of LKS, will have inhibited the development of accurate and well-specified phonological representations.

However, it is possible that errors in responding to the auditory visual lexical decision task are also arising due to more peripheral auditory processing difficulties, before comparison is made with the representations, as suggested by performance on the masking tasks and the word and non-word discrimination.

This study indicates that 10 years after the onset of LKS, William continues to have significant auditory processing difficulties affecting the development of speech and language skills. These auditory processing difficulties are pervasive, affecting perception and discrimination of linguistic and non-linguistic sound signals. This would support the suggestion that the neurological disturbances found in LKS may disrupt functioning in the auditory cortex. The findings would also validate the use of auditory training programmes in cases of LKS, not only of the traditional kind (Vance, 1991, 1997), but also those that incorporate synthetic sounds that can be manipulated and controlled by computer (Merzenich *et al.,* 1996).

Acknowledgements

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