Auditory Processing Disorder and Brain Pathology in a Preterm Child with Learning Disabilities

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Abstract

Background: Auditory processing disorders involve deficits in the processing of information in the auditory domain that are not due to higher order language, cognitive or other related factors.

Purpose: To evaluate the possibility of structural brain abnormalities in preterm children manifesting as auditory processing disorders.

Research Design : A case report of a young girl, preterm at birth, with language difficulties, learning problems at school, and additional listening problems.

Results: A diagnosis of a central auditory processing disorder was made on the basis of severe deficits in three nonspeech temporal tests (the frequency and duration pattern and the random gap detection tests). Her brain MRI revealed large porencephalic cysts and thinning of the corpus callosum.

Conclusions: The observed auditory deficits would be compatible with a pressure effect of the cysts at a brainstem or higher level for the random gap detection test, and with the thinning of the corpus callosum for the pattern tests, the latter requiring interhemispheric transfer of information. The case highlights that preterm children with learning difficulties may suffer from an auditory processing disorder, in the presence of structural brain abnormalities that are due to birth and neonatal complications.

Key Words: Auditory processing disorder, brain pathology, learning disabilities

Abbreviations: (C)APD = (central) auditory processing disorder; EEG = electroencephalography; MRI = magnetic resonance imaging

Sumario

Antecedentes: Los trastornos de procesamiento auditivo involucran deficiencias en el procesamiento de la información en el ámbito auditivo que no se deben a lenguaje de orden superior, a factores cognitivos o a otros factores relacionados.

Propósito: Evaluar la posibilidad de anormalidades estructurales del cerebro en niños de pretérmino manifestados como trastornos de procesamiento auditivo.

Diseño de Investigación: Es un reporte de caso de una niña joven, pretérmino al nacimiento; con dificultades del lenguaje; trastornos del aprendizaje en la escuela y problemas adicionales de audición.

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Resultados: Se realizó el diagnóstico de trastorno central de procesamiento del lenguaje sobre la base de deficiencias en tres pruebas temporales de no lenguaje (Las pruebas de patrones de frecuencia y duración y de detección aleatoria de la brecha). Su MRI del cerebro reveló grandes quistes porencefálicos y adelgazamiento del cuerpo calloso.

Conclusiones: Las deficiencias auditivas observadas serían compatibles con un efecto de presión de los quistes a nivel de tallo cerebral o más arriba, con la prueba de detección aleatoria de la brecha, y con el adelgazamiento del cuerpo calloso en relación a las pruebas de patrones, que se requieren luego de transferencia interhemisférica de información. El caso resalta que los niños de pretérmino con dificultades de aprendizaje pueden sufrir de un trastorno de procesamiento auditivo, en presencia de anormalidades estructurales del cerebro que son debidas a complicaciones al nacimiento o neonatales.

Palabras Clave: Trastorno de procesamiento auditivo, patología cerebral, discapacidad del aprendizaje

Abreviaturas: (C)APD = Trastrono (central) de procesamiento auditivo; EEG = electroencefalograma; MRI = imágenes por resonancia magnética

(Central) auditory processing disorder ([C]APD) may present: (1) in isolation (Musiek et al, 2005), (2) comorbid with other developmental syndromes (Bellis and Ferre, 1999, Moncrieff and Musiek, 2002), or (3) as a result of a neurological condition. Clinicians are aware that (C)APD in adults may be caused by a neurological condition, such as stroke or demyelinating disease, with (C)APD the first or only manifestation of the neurological condition (e.g., Bamiou et al, 2004). However, brain pathology should also be considered and ruled out as a causal factor in pediatric cases with auditory processing disorders. We present the case of a young child referred for a central auditory assessment because of language and learning problems, with listening difficulties a lesser concern, in whom central auditory tests revealed deficits and the brain MRI (magnetic resonance imaging) identified abnormalities of the central auditory pathway.

CASE PRESENTATION

A 13-year-old girl was referred to the audiological laboratory of the AHEPA University Hospital, Aristotle University of Thessaloniki for auditory processing evaluation because of learning problems at school. The parental reported difficulties with sustained attention in the auditory channel, distraction, and minor listening problems. The child constantly experienced difficulty in following multistep directions, which was evident in the classroom. She was not able to write down homework activities as a whole effectively, and managed to write down only part of them. She experienced difficulty in reading and spelling especially in the classroom. Auditory memory was poor; there was a major deficit in confusing the order of the steps in multistep instructions. The child's memory failed when she had to recall instructions an hour later then when they were given to her. Her attention span through the auditory channel appeared to be limited while in a noisy room; she experienced major difficulties when having to pay attention for more than 10 to 15 minutes in the classroom and when she had to attend school in the afternoon.

The child was born at 30 weeks gestational age with normal labor and a weight of 4.85 lb. She spent two days in the special care baby unit as a result of being premature at birth. Her APGAR (appearance, pulse, grimace [reflex], activity, respiration) scores were nine for the first minute and ten for the fifth minute. She had a mild increase in bilirubin (jaundice), which returned to normal with no further treatment. The patient walked when she was two and a half years old. She experienced speech delay; she uttered her first words when two and a half years old and her first two-word sentence at three years old. Patient's visual acuity is reduced; she has myopia in both eyes, and she wears glasses.

Her language and learning testing with Greek-language-validated tools revealed language skills well below her chronological age and documented her learning disabilities. Testing was accomplished by a battery called the Athena test (Paraskevopoulos, 1996), which includes normative data for children from 5 to 15 years of age. The test includes 14 subcategories; normative values are provided according to chronological age for each subcategory separately, and a child's profile is sufficiently documented with his or her strengths and weaknesses highlighted. The test can be administered either as a whole or using some of the subtests. Results for each test produce a raw number and the developmental age for the specific skill of the child tested. The 14 subtests are (1) Linguistic analogies, (2) Copying shapes, (3) Vocabulary (providing descriptive definitions), (4a) Memory for digits, (4b) Common sequences (days of the week, months, numbers), (5) Memory for pictures, (6) Memory for shapes, (7) Completing sentences with one word missing, (8)

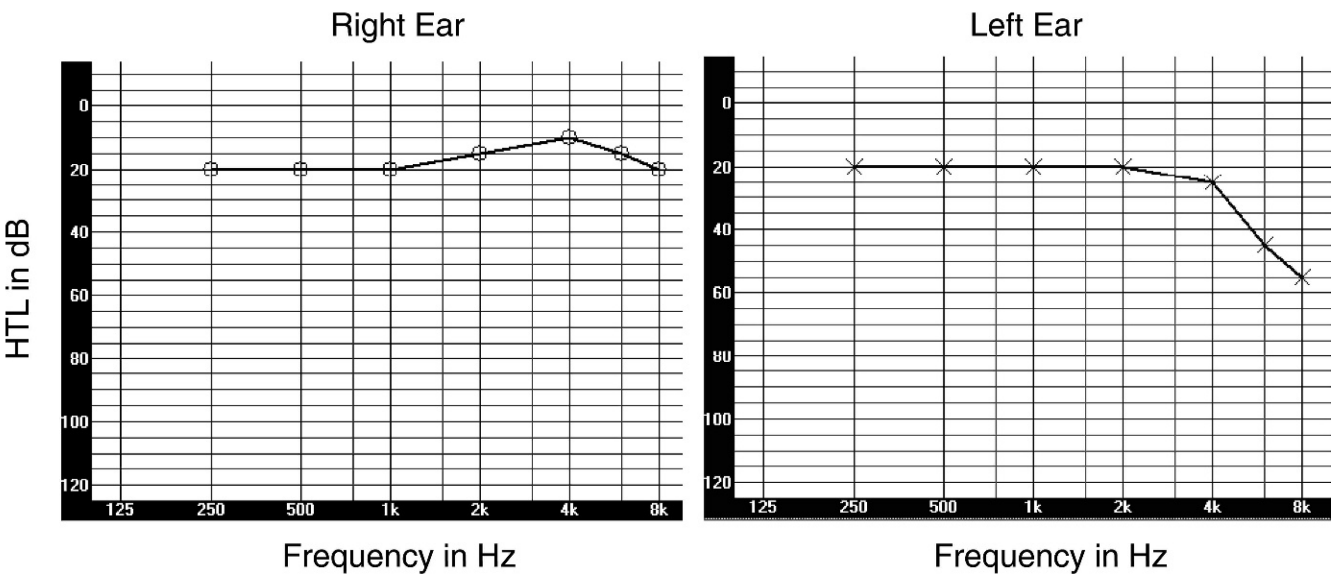

Figure 1. Audiogram during patient's first visit showing unilateral (left ear) high-frequency hearing loss.

Completing words with one phoneme (corresponding to a letter) missing, (9) Visual discrimination of nonwords, (10) Auditory discrimination of nonwords, (11) Phonemic synthesis, (12) Motor and visual coordination, (13) Discriminating right from left, (14) Laterality. Her ability to provide definitions was equivalent to a child with a developmental age of six years old. The child's phonemic synthesis skills were equivalent to those of a child of seven years and eight months, and her skill in discriminating between same and different pairs of nonwords (auditory mode) was equivalent to that of a four-year-old. Her ability to complete words with one phoneme missing was equivalent to a child of eight years and one month, while completing sentences with one word missing was consistent with her age. Her auditory memory for digits was equivalent to a child of the developmental age of six years and seven months. All other subtests were within normal limits according to her age. Intelligence was assessed by administering the Wechsler Intelligence Scale for Children (WISC)-III. This test was standardized on a Greek population in 1997 and includes picture completion, coding, similarities, picture arrangement, arithmetic, block design, vocabulary, object assembly, comprehension, and digital span. (Georgas et al, 1997). Her IQ testing with the WISC-III Greek version gave a low average score for both verbal (95) and nonverbal (85) scales, with the verbal scale being slightly better than the nonverbal one.

AUDITORY PROCESSING EVALUATION

In the first session the child was evaluated otoscopically and audiometrically, by both pure tone and speech audiometry (Figure 1). In her left ear there was a significant hearing loss averaging 35 dB across the high-frequency range from 4000 to 8000 Hz. In her right ear the loss over the same high-frequency range was only 5 dB. Speech discrimination scores at 35 dB above the average threshold across the frequencies 500, 1000, 2000, and 4000 Hz were 97% for the right ear and 87% for the left ear. The child did not report any hearing loss and parents confirmed that hearing was unremarkable in everyday situations.

A second session took place about two months later. In this session basic audiometric testing (pure tone and speech audiometry) showed that hearing thresholds were, at this point in time, both symmetrical and within normal limits (Figure 2). Since peripheral hearing was now normal, we proceeded with the auditory processing testing, which consisted of a speech in babble test, the duration pattern sequence test (Musiek et al, 1990), the pitch pattern sequence test (Musiek and Pinheiro, 1987), and the random gap test (Keith, 2000). Auditory brain stem responses were also recorded, together with tympanometry and acoustic reflexes. EEG (electroencephalography) and MRI were also performed for her memory issues.

Test battery selection was based on the American Speech-Language-Hearing Association's 2005 recommendations for establishing diagnosis and included (1) both verbal and nonverbal materials and (2) testing different aspects of auditory processing. However, there were limitations in the test battery selection used. First, the Greek dichotic digits test was not used, as technical reasons did not permit us to use it in this particular patient. Other limitations included the use of a newly developed speech in babble test with normative data in 32 children over a large age span. However, in this particular case, as results are high (within normal limits) and the child's age is not in the extremities of our sample (neither very "young" nor very "old"), it is our

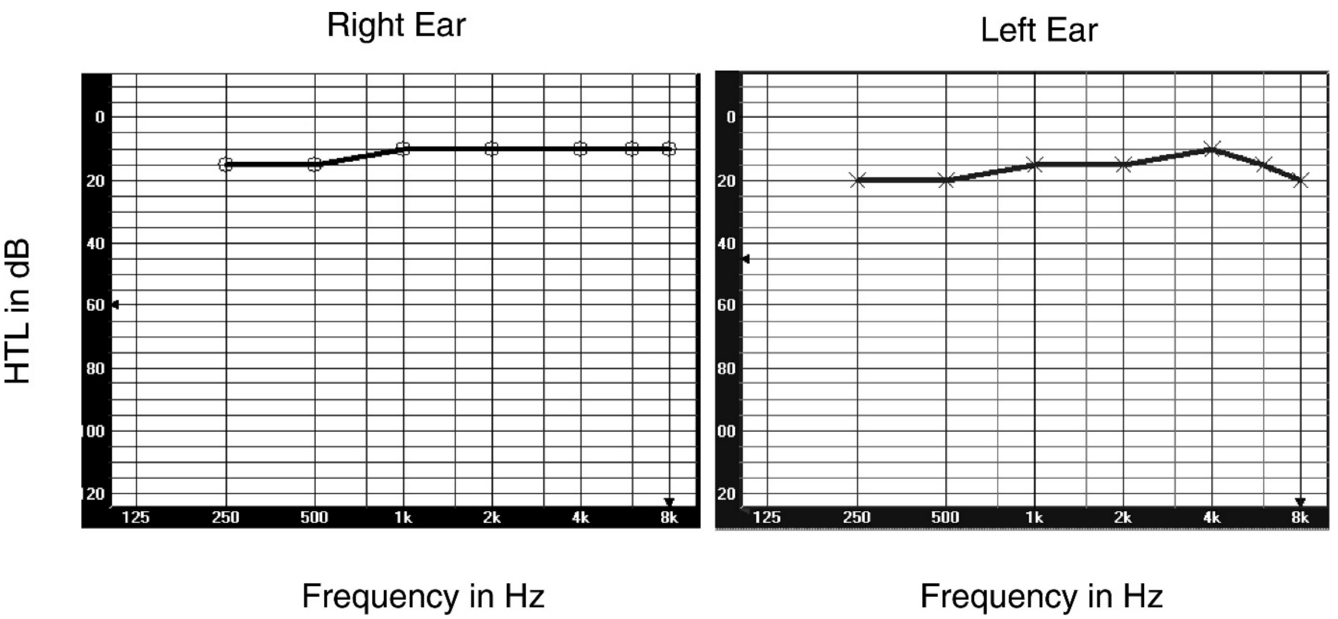

Figure 2. Audiogram during patient's second visit showing normal and symmetrical hearing.

belief that the result is strongly suggestive of normal speech perception in noise (babble), at least for the specific test. Pattern sequence tests were only performed in the labeling condition. The humming condition was not performed: this is the standard procedure in our laboratory. Temporal resolution was tested by the Random Gap Detection Test with nonverbal stimuli but with limited published data showing validity and efficiency (Chermak and Lee, 2005).

The Greek speech in babble test employs an 8 dB signal to noise ratio, bisyllabic words from three recently developed, phonetically balanced and frequent word lists (Iliadou et al, 2006), and natural cafeteria speech as competition. Normative data for this test were obtained on 32 Greek children, between the ages of 7 and 17.5 years, with no known learning or auditory processing deficits. The objective of the test is to identify and repeat the words that are heard in a background of speech babble.

The duration and pitch pattern sequence tests assess the ability to discriminate between, to sequence, and to correctly label different pure tones as either high or low (pitch) or long or short (duration); to some extent, short term memory is also tested. The duration pattern sequence test is more sensitive to cortical as opposed to cochlear lesions. Three consecutive 1000 Hz tone bursts are presented. The durations are either 500 msec (long) or 250 msec (short). Right and left ear are tested separately. The subject indicates the pattern by reporting the combinations of short and long tones according to her or his perception. The pitch pattern frequency test begins with a pitch discrimination practice test in which the child hears pairs of tone bursts. In the actual test three consecutive tone bursts are presented in each trial; each burst is either high (1430 Hz) or low (880 Hz). The child must respond verbally. Right and left ears are tested separately.

The random gap detection test evaluates temporal processing by determining the smallest time interval detectable between two closely approximated stimuli (Greene, 1971). Stimulus intervals vary between 0 to 40 msec, and pairs of the same frequency tones are presented. Frequencies tested are 500 Hz, 1000 Hz, 2000 Hz and 4000 Hz. There is evidence that the test produces results comparable to other tests of temporal resolution (Chermak and Lee, 2005) and that it may provide information on central auditory nervous system involvement (Musiek et al, 2005).

RESULTS

The results of the second session, which included the auditory processing testing, revealed that her previously documented high-frequency sensorineural hearing loss was now absent. Her pure tone audiometry thresholds were both normal and symmetrical in this session. Moreover, tympanometry and acoustic reflex thresholds were normal and symmetrical; these results documented both normal status of the middle ear and normal peripheral hearing sensitivity. Central auditory testing, however, revealed abnormal results on three nonspeech tests (Table 1). The poorest results were observed for the left ear. Auditory brain stem responses revealed a less than robust response (Figure 3) but with normal morphology, wave latencies, and latency intervals from the left ear according to our clinical data based on control subjects matched for age

Table 1. Audiometric Test Results

Tests	Right ear results	Left ear results	Normative data
PTA average	10	15	0–20 dB
Speech audiometry	97%	100%	80–100%
Speech in babble	95%	90%	75–100%
Random Gap Detection Test	>40 msec		<20 msec
Duration pattern sequence	35%	15%	67–100%
Pitch pattern sequence	55%	20%	75–100%

Note: PTA average = Pure-tone audiometry, average of six frequencies ranging from 250 Hz to 8 KHz.

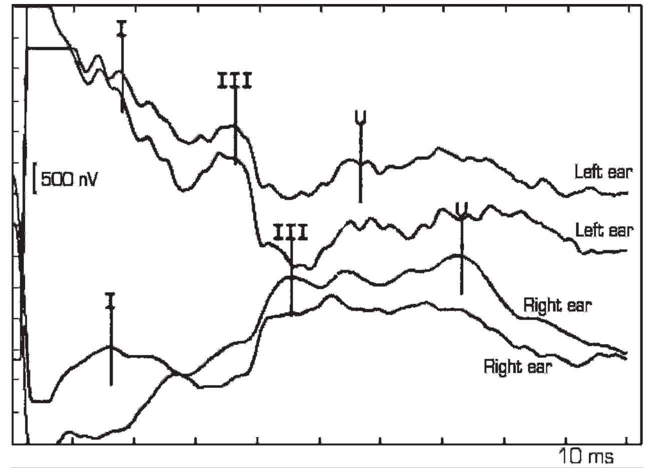

Figure 3. ABR waveform acquired at 130 dB SPL, showing normal wave latencies and latency intervals for the left ear (wave I =1.8 msec, III = 3.64 msec, V = 5.68 msec) and absent wave V for the right ear and prolonged latency for wave III (4.6 msec) for the right ear. Marked delayed wave V for the right ear is not clearly replicable.

and sex. ABR from the right ear revealed a normal wave I, prolonged latency of wave III, and the absence of wave V. The possibility of the presence of wave V at 7.3 msec, although not clearly replicable, has also been considered. In this case there was prolongation of interwave latencies I-III and III-V, which also renders the ABR recording abnormal. EEG revealed an abnormal rhythmic pattern in the left central-temporal area. Brain MRIs revealed large porencephalic cysts in communication with the ventricular system, significantly enlarged ventricles (III and IV) (Figure 4), and considerable thinning of the corpus callosum (Figure 5).

DISCUSSION

Of particular interest in this case is the documented high-frequency sensorineural hearing loss in the patient's left ear. This was not reported to be of concern by either the child or her parents. Oral cortisone was administered in a decreasing dose for 15 days. Follow-up was two months after the first session. Now normal audiometric thresholds were obtained from both ears. This was puzzling. After reviewing the MRI, we hypothesized that the transient asymmetrical high-frequency hearing loss could be the result of a pressure effect from hemorrhagic areas (due to a pre- or perinatal hemorrhage lesion that later brought about the cysts) or alternatively resulting from the same cause that produced the cysts and the significant enlargement of the third and fourth ventricles. (Gosalakkal, 2002; Chao, 2005). Cortisone could have aided in the attenuation of pressure, although this can not be validated. There was high-frequency loss during the first session, but we have no data to confirm or deny its presence for a long or short period of time.

The results of the second session led to the diagnosis of auditory processing disorders. A similar case is cited by Musiek et al (1994) in a 13-year-old boy. In that case the mother had been concerned about his hearing in noise for many years. Interestingly, his right ear had a moderate to severe high-frequency sensorineural hearing loss. Auditory processing disorder was diagnosed and a large subarachnoid cyst in the right hemisphere, present at birth, was thought to be the basis for the learning disability. In our case, the abnormal result for the random gap test could be due to pressure effects at the brainstem level or above, and the same is most probably true for the absent wave V (or, alternatively, prolonged) in her right ear, which would indicate pathology at the level of the inferior colliculus/lateral lemniscus on the right side. ABR pathology has been previously documented in a small percentage, less than ten percent, of pediatric (C)APD cases (Hall and Mueller, 1997). The possibility that memory problems may interfere with reliable (central) auditory processing testing should also be considered.

In our case, in addition to the porencephalic cysts, there was a significant reduction in the thickness of the corpus callosum. The child was premature at birth. One of the most common brain abnormalities in preterm babies is thinning of the corpus callosum. In a recent study (Nosarti et al, 2004), it was documented that the total surface area of corpus callosum in very preterm individuals was 7.5% less than in controls and that this correlated with their verbal fluency tests, without showing in the overall IQ score. The corpus callosum is extremely vulnerable to ischemia and hemorrhage occurring in preterm births; it can also be influenced by its position adjacent to the periventricular area, a common site of hemorrhage in infancy. In a study by Santhouse et al 2002, preterm adults with thinner or athrophic corpus callosum were able to perform, at a reduced level, timbre and bilateral field comparison tasks. We similarly propose that this child's deficits in the sequencing tasks were to a large

Figure 4. Large (occipital left and frontal right) porencephalic cysts in communication with the ventricular system.

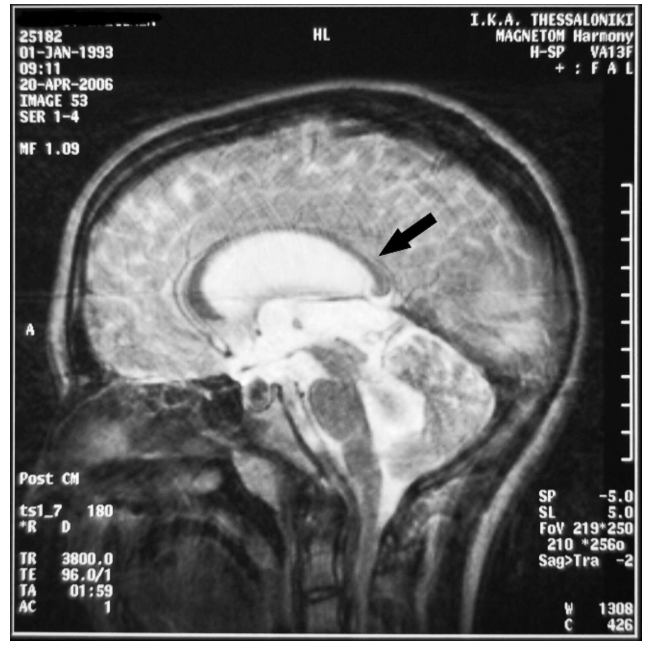

Figure 5. Significant thinning of the corpus callosum.

extent due to the thinning of the corpus callosum. It has been proposed that for a sequence of sounds, the right hemisphere determines the pattern of the sequence as a gestalt, but the labeling of the sequence happens in the left (language) hemisphere (Pinheiro and Musiek, 1985). Thus, tasks that require labelling of sound sequences depend on transfer of information from the right to the left hemisphere via interhemispheric commissures (Pinheiro and Musiek, 1985). After complete section of the corpus callosum, subjects show abnormal results for both tested ears for orally reported frequency and duration patterns, while scores for a hummed response will remain normal or near normal (Musiek et al, 1990; Musiek et al, 1994). Similarly, children with a reduction in the size of the corpus callosum, due to the presence of a mutation in the PAX6 gene, give abnormal scores in the frequency and duration pattern tests (Bamiou et al, 2007). This is consistent with the findings in our case.

In addition, there was a marked deficit in the left ear with slightly better but still abnormal results in the right ear. The marked deficit in the left ear could be attributed to the additional pathology at a high brainstem level on the right side, as indicated by the absence of wave V on the ABR. Pathology at a high brain stem level can lead to contralateral deficits in auditory pattern tests (Baran and Musiek, 1999). In the case presented, thinning of the corpus callosum appears to play a role in the manifestation of the auditory processing disorder but not the sole/exclusive one. The limitations of our test battery mentioned in the auditory processing evaluation section should be taken under consideration and may interfere or worsen the auditory processing abilities of the subject tested.

It is interesting that when testing language abilities, her lowest scores were for vocabulary and nonwords, whereas her ability to complete sentences with one missing word was normal for her chronological age. These findings could indicate phonological-auditory decoding difficulties rather than specific language or cognitive ones. Her language abilities appear to be sufficient to overcome auditory perceptual difficulties when dealing with sentences, which are more linguistically redundant than words that are out of context and for perceiving and understanding words out of context, basic auditory perception is more essential. Her IQ score was low average, but in those tests the auditory modality is mostly used, and we could not rule out the possibility that these scores may be influenced by an auditory processing disorder and possibly linked to it. If low IQ results are considered to be independent of the underlying auditory processing disorder, there is always the possibility of IQ influencing the central auditory processing battery tests. Our lab has cases of children with matching age and IQ scores who give normal scores in the auditory processing tests. In the auditory processing battery used, all the nonspeech tests were abnormal, as opposed to speech in babble which was normal. This points to a form of auditory processing deficit at a "sensory" as opposed to a "linguistic" level and is in accordance with both the ASHA and the UK definitions of auditory processing disorders (British Society of Audiology APD Interest Group, 2008).

CONCLUSION

This case of an ex-preterm child highlights the need to consider the possibility that disordered auditory processing may underlie language and learning difficulties and the possibility that auditory processing deficits, as well as language and learning difficulties in a child, may be due to structural brain abnormalities. Clinicians who come across preterm children ought to investigate these children accordingly.

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