

Original article

Auditory processing disorder in perisylvian syndrome

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Abstract

We hypothesized that the processing of auditory information by the perisylvian polymicrogyric cortex may be different from the normal cortex. To characterize the auditory processing in bilateral perisylvian syndrome, we examined ten patients with perisylvian polymicrogyria (Group I) and seven control children (Group II). Group I was composed by four children with bilateral perisylvian polymicrogyria and six children with bilateral posterior perisylvian polymicrogyria. The evaluation included neurological and neuroimaging investigation, intellectual quotient and audiological assessment (audiometry and behavior auditory tests). The results revealed a statistically significant difference between the groups in the behavioral auditory tests, such as, digits dichotic test, non-verbal dichotic test (specifically in right attention), and random gap detection/random gap detection expanded tests. Our data showed abnormalities in the auditory processing of children with perisylvian polymicrogyria, suggesting that perisylvian polymicrogyric cortex is functionally abnormal. We also found a correlation between the severity of our auditory findings and the extent of the cortical abnormality.

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1. Introduction

Perisylvian syndrome comprises a variety of clinical manifestations due to lesions on the perisylvian or opercular regions [1]. Congenital malformations such as polymicrogyria may be implicated as etiological factors. Bilateral perisylvian polymicrogyria is a malformation of cortical development characterized by excessively small gyri seen around the Sylvian fissure on magnetic resonance imaging [2,3].

Clinical features consist of pseudobulbar signs such as drooling, poor swallowing and palatal dysfunction in neurological examination, as well as dysarthria, epilepsy, specific language impairment and reading disabilities [4–7].

The severity of the clinical manifestations correlates with the extent of the lesion. Therefore, the term bilateral perisylvian polymicrogyria is applied when the cortical malformation spreads around the entire extent of the sylvian fissure, and bilateral posterior perisylvian polymicrogyria is applied when polymicrogyria occurs only in the posterior part of the parietooccipital regions. The latter appears to be associated with a genetic predisposition and softer clinical features (such as speech delay) when compared to the bilateral perisylvian poly-

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microgyria (which often presents with pseudobulbar palsy and epilepsy) [4,7].

Auditory processing is defined as the mechanism and process of the auditory system that enables localization and sound lateralization, auditory discrimination, pattern recognition, temporal processing and auditory performance in competitive speech or degraded acoustic signals [8]. Temporal auditory processing is the perception of the temporal characteristics of a sound or of a sound change within a restricted time interval. It is considered a fundamental ability required for the auditory perception of verbal and non-verbal sounds [9]. This ability is important to identify the sequence of phonemes in the speech. Problems with this processing can interfere with temporal aspects of speech, such as perception and recognition of phonemes [10]. Children with auditory processing disorders may have difficulties listening in background noise, following oral instructions and understanding temporal aspects of speech in the presence of normal peripheral hearing [11].

We hypothesized that the processing of the auditory information by the polymicrogyric cortex is different from the normal cortex. Therefore, the objective of this study was to characterize the findings of the evaluation of the auditory processing in individuals with perisylvian syndrome.

2. Subjects and methods

This work was carried out in the Clinical Hospital of the University of Campinas (Unicamp), Campinas, SP, Brazil after being approved by the Ethics Committee of the same university (protocol 196/2003). Parents agreed to their children participating in this study, having read and signed the informed consent form. The study was conducted from March 2007 to July 2008.

Inclusion criteria were the presence of either bilateral perisylvian polymicrogyria or bilateral posterior perisylvian polymicrogyria on magnetic resonance imaging, performance intelligence quotient >80 and specific language impairment. Specific language impairment refers to inadequate oral language acquisition in the absence of sensory or intellectual deficits, pervasive developmental disorders or severe environmental deprivation. It occurs at least 12 months behind chronological age and nonverbal cognitive skills are usually normal. These children may present with school difficulties [4,5].

The assessment included: neurological examination, neuroimaging investigation, audiological assessment and intellectual quotient score. We evaluated 10 children with bilateral and symmetrical perisylvian polymicrogyria (Group I) and compared them with seven normal children (Group II). Ages ranged from eight to 16 years (mean = 11.6 years in Group I and mean = 11.4 years in Group II).

In the neurological examination, four children in Group I had pseudobulbar signs. They had abnormal history of drooling, choking, and sucking difficulty during the first years of life. There were no neurological changes in the other children.

To obtain the intellectual quotient we utilized the Wechsler intelligence scale for children III (WISC-III) [12]. For this study we included patients with performance intelligence quotient >80. Since language delay was required for inclusion in the study, our patients frequently presented verbal intelligence quotient scores inferior to performance intelligence quotient scores. Low verbal scores jeopardized full scale intelligence quotient scores, therefore we decided to include the performance intelligence quotient score as it better represents the cognitive ability of this type of patient.

Neuroimaging investigation was performed in a 2.0 T scanner (Elscent Prestige) with posterior multiplanar reconstruction and curvilinear reformatting in 3D magnetic resonance imaging. Neuroimaging evaluation allowed a subdivision of Group I: four children had bilateral perisylvian polymicrogyria and six children had bilateral posterior perisylvian polymicrogyria.

The participants underwent pure tone audiometry. For the peripheral audiological evaluation an acoustic cabin, AC-30 audiometer (Interacoustics) was used. In children with normal peripheral hearing (threshold <25 dB), behavior tests of auditory processing were applied.

Taking into account age and auditory maturation, the behavior auditory tests were: the adapted digits dichotic test with binaural integration [13], the nonverbal dichotic test [14], and the random gap detection test or random gap detection test expanded [10], with stimuli through a two channel audiometer connected to a Philips CD player. These were all conducted in an acoustic cabin.

The digits dichotic test consists of 20 stimuli presentations (40 per ear). The test is presented at 50 dB based on the average of pure tone thresholds at frequencies of 500, 1000 and 2000 Hz. The patient hears two numbers simultaneously in each ear and repeats all of them. The digits list consists of the digits 4, 5, 7, 8 and 9, which form dissyllable words in Portuguese. This test, in the binaural integration task, evaluates the ability to combine components of the acoustic signal with competitive sounds and turn them into verbal terms. After 20 presentations, the phones are reversed to avoid interference of the phones calibration [13].

The nonverbal dichotic test consists of six different nonverbal sounds, with three onomatopoeic sounds and three environmental sounds, combined in pairs and presented simultaneously in each ear in three stages. In the stage of free attention, the individual points only to a figure corresponding to one of the two sounds presented. In the right and left attention, he points to the

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