AUDITORY PROCESSING DISORDERS IN TWINS WITH PERISYLVIAN POLYMICROGYRIA

Mirela Boscariol[1], Vera Lúcia Garcia[2], Catarina A. Guimarães[3], Simone R.V. Hage[4], Maria Augusta Montenegro[5], Fernando Cendes[6], Marilisa M. Guerreiro[7]

Bilateral perisylvian polymicrogyria is a malformation of cortical development due to abnormal late neuronal migration or abnormal cortical organization around the sylvian fissure[1].

The severity of the clinical manifestations correlates with the extent of the lesion. Therefore, the term diffuse polymicrogyria is applied when the cortical malformation spreads around the entire sylvian fissure, and restricted polymicrogyria is applied when polymicrogyria occurs only in the posterior part of the parietal region. The restricted form is also called bilateral posterior parietal polymicrogyria and appears to be associated with a genetic predisposition and soft clinical features (such as speech delay and dysarthria) when compared to the diffuse form of polymicrogyria. Studies with families showed correlation of specific language impairment (SLI) and learning disabilities with perisylvian polymicrogyria[2].

Referencing cited studies, this paper describes the language, learning and audiology findings in dizygotic twins and associates them with the neuropsychological, neurological and neuroimaging findings.

CASE

This study was approved by the Ethics Committee of our University Hospital (protocol 196/2003). Parents gave permission for their children to participate in this research and signed the informed consent form. The study was conducted from March 2007 to May 2008.

Two 14-year-old, male, dizygotic twins were evaluated. The assessment included: neurological examination, neuroimaging investigation, neuropsychological, language and learning assessment and audiological evaluation. In childhood, the twins had been diagnosed as having SLI.

The neuropsychological assessment utilized the Wechsler Intelligence Scale for Children III (WISC-III). Neuroimaging investigation was performed in a 2.0 T scanner (Elscint Prestige) with posterior multiplanar reconstruction and curvilinear reformatting in 3D magnetic resonance imaging (MRI).

The language assessment considered the following aspects: phonological, morphosyntactic, semantic and pragmatic production. Standard and non-standard speech protocols were used: sample of free speech; ABFW – Children Language Test with phonological and vocabulary tests[3]. Reading/writing evaluation included: sample of free writing, Phonologic Skill Test[4], School Performance Test[5], non-words reading and writing, oral speed reading, and text understanding.

The peripheral audiological capability was assessed with audiometry, speech reception thresholds and acoustic impedance tests. An acoustic cabin was used with an AC-30 audiometer (Interacoustics) with phone TDH-39P and impedance audiometer AT235h (Interacoustics).

After establishing normal peripheral hearing, we applied behavioral auditory processing tests. The measures of short and middle latency auditory evoked potentials were also tested with GSI-Audera.

The auditory processing tests were: Random Gap Detection Test[6], Digits Dichotic Test with binaural integration[7], and Nonverbal Dichotic Test[8] with stimuli through a two channel audiometer connected to a Phillips CD player, using an acoustic cabin.

The Random Gap Detection Test is a temporal resolution test that determines the time interval in which the gap detection thresholds exist. It includes the time interval between zero and 40 milliseconds[6].

The Digits Dichotic Test consists of 20 stimuli presentations (40 per ear). The patient hears two numbers simultaneously in each ear and repeats all of them[7].

The Non-verbal Dichotic Test consists of six different nonverbal 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

DISTÚRBIO DO PROCESSAMENTO AUDITIVO EM GEMELARES COM POLIMICROGIRIA PERISYLVIANA

Department of Neurology, Faculty of Medical Sciences, University of Campinas (Unicamp), Campinas SP, Brazil: [1]PhD student; [3]PhD; [5]Assistant Professor; [6]Associate Professor; [7]Full Professor. Interaction between University Service and Community Program, Botucatu Medical School, State University of São Paulo "Júlio de Mesquita Filho": [2]Assistant Professor. Department of Speech Therapy, University of São Paulo (USP), Bauru SP, Brazil: [4]Assistant Professor. Mirela Boscariol received a scholarship from CNPq (grant number # 132461/2007-2) and FAPESP (grant number # 07/00806-4). Catarina A. Guimarães received a scholarship from FAPESP (grant number # 06/56257-6).

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Dra Marilisa M. Guerreiro – Department of Neurology / FCM / Unicamp - PO Box 6111 - 13083-970 Campinas SP - Brasil. E-mail: mmg@fcm.unicamp.br

Table. Characteristics of language, reading and written and auditory processing data.

Data	First twin	Second twin
Language		
Phonology	Normal	Impairment
Syntax	Impairment	Impairment
Semantic	Normal	Normal
Pragmatic	Normal	Normal
Reading and written		
Phonological awareness (PST)	Low performance	Low performance
SPT written/reading	Low performance	Low performance
SPT arithmetic	Low performance	Low performance
Non-word written /reading	Low performance	Low performance
Oral speed reading	Abnormal	Abnormal
Text understanding	Abnormal	Abnormal
Auditory processing		
Digits Dichotic – BI	RE: normal	RE: abnormal
	LE: normal	LE: normal
Nonverbal dichotic		
Free attention	RE: normal	RE: normal
	LE: normal	LE: normal
Right attention	RE: normal	RE: abnormal
	LE: abnormal	LE: abnormal
Left attention	RE: abnormal	RE: abnormal
	LE: normal	LE: abnormal
RGDT (ms)	Normal	Abnormal

PST: phonologic skill test; SPT: school performance test; BI: binaural integration; RE: right ear; LE: left ear; RGDT: Random gap detection test.

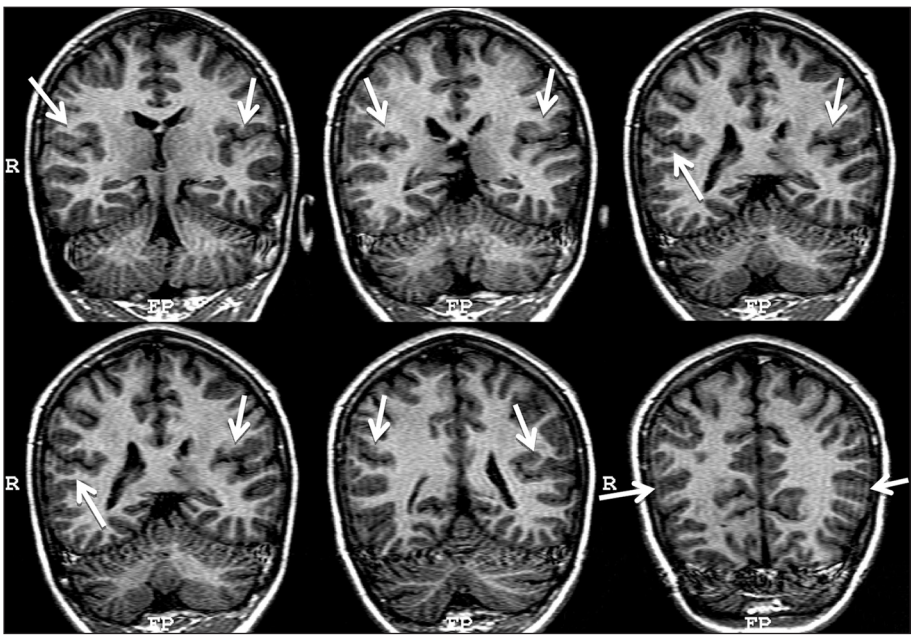

Fig 1. T1 image. MRI study of first twin with bilateral posterior parietal polymicrogyria.

two sounds presented. In the right and left attention, he points to the figure on the right and left ears, respectively[8].

The auditory brainstem response and the middle latency response were evaluated following the Audera's protocols.

With respect to the neurological and neuropsychological findings, the first twin showed normal neurological examination, verbal IQ=76, performance IQ=87 and full scale IQ=80. The second twin showed pseudobulbar signs in the neurological examination, verbal IQ=76, performance IQ=86 and full scale IQ=80. Neuropsychological and reading assessments showed learning disabilities in both patients.

The Table shows the evaluation of language, reading assessment, and auditory processing in both twins.

Despite the previous language impairment in childhood,

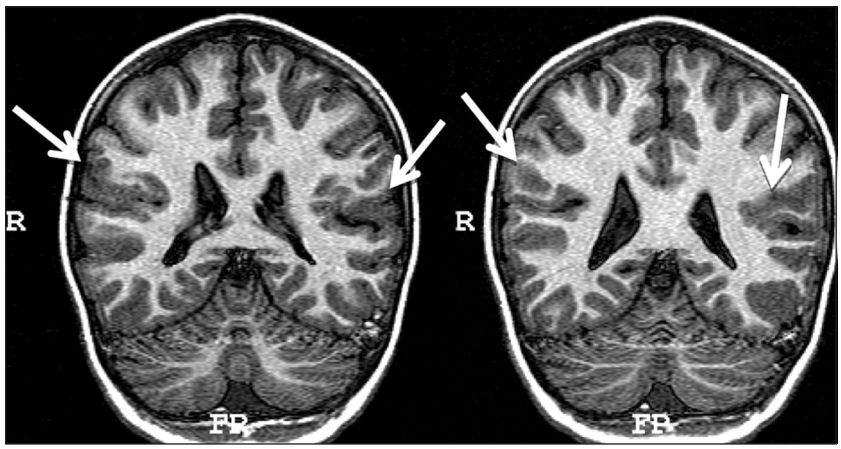

Fig 2. T1 image. MRI study of second twin with bilateral posterior parietal polymicrogyria.

the first twin showed no change in the current phonological assessment.

The Figures 1 and 2 show MRI studies of both twins demonstrating bilateral posterior parietal polymicrogyria.

DISCUSSION

The proximity of the sylvian fissure to the auditory region and its association with areas responsible for language and learning means that a change in the former area can affect functions in the latter area. As described above, the twins had a SLI in childhood, which did not improve over the study period in the second twin, and evolved into a learning disorder in both. Language and learning disabilities have been associated with brain changes, such as malformations of cortical development[2]. Those changes may also cause phonological processing abnormalities and auditory processing disorder[9]. Perisylvian areas harbor the auditory processing function which explains our findings and reinforce the association as perisylvian polymicrogyria was seen in the high-resolution MRI in our patients.

In our study there was a discrepancy between verbal and performance IQ. Previous data[2] are in agreement with this study which found the same discrepancy between verbal and performance IQ in children with SLI and perisylvian polymicrogyria.

The auditory processing was abnormal in some aspects (non-verbal auditory processing in both twins and temporal resolution in the second twin). A failure in auditory processing can cause damage in the phonological system resulting in language and learning impairment[9].

Electrophysiological studies have also been used to investigate auditory processing disorders. Purdy et al.[10] found increased latency Pa waves in the middle latency response in children with learning impairment when compared to a control group. We did not find changes in auditory brainstem response or middle latency response in the investigated twins.

Our data suggests that structural changes in the sylvian region can lead to changes in auditory processing and, therefore, in language and learning. The correlation of speech and auditory findings, neuropsychological and neuroimaging results is important for diagnostic and therapeutic purposes.

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