

# Characterization of language and phonological working memory in patients with myoclonic astatic epileptic syndrome

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## ABSTRACT

The course of myoclonic astatic epileptic syndrome (MAES) is variable and little information is available about cortical functions in the presence of the disease. The objective of the present study was to assess the phonological working memory (PWM) and the verbal language of six patients between 8 and 18 years old, on treatment for at least 5 years, and good control of seizures, diagnosed in the Service of Epilepsy of Hospital of Clinics of Ribeirão Preto, São Paulo University in Brazil. The Test of Repetition of Meaningless Words was used to assess PWM. Video-recorded of language samples were collected during spontaneous and directed activities for the study of verbal language and pragmatics. A qualitative analysis showed that all patients presented deficits in the execution of the PWM test and only one patient showed poor mastery of all aspects studied. These finds contribute to strategies of treatment for language problems of patients with MAES, focusing on PWA. **Key words:** working memory, language, epilepsy, myoclonus.

## Caracterização da linguagem e memória de trabalho em pacientes com síndrome epiléptica mioclônica astática

## RESUMO

A evolução da síndrome epiléptica mioclônica astática (SEMA) é variável e há poucas informações sobre funções corticais. Este estudo pretendeu avaliar a memória de trabalho fonológica (MTF) e a linguagem verbal de pacientes com SEMA. Foram avaliados seis pacientes entre 8 e 18 anos em tratamento há mais de 5 anos e bom controle das crises, do Serviço de Epilepsia do Hospital das Clínicas da Faculdade de Medicina de Ribeirão Preto da Universidade de São Paulo. A Prova de Repetição de Palavras Sem Significado foi utilizada para MTF, e coletaram-se amostras de linguagem, gravadas em vídeo, durante atividades espontâneas e dirigidas para estudo da linguagem verbal e pragmática. Análise qualitativa mostrou que todos apresentaram deficiência na MTF e, exceto um, mostraram domínio dos aspectos verbais estudados. Os achados contribuem para estratégias de tratamento de problemas de linguagem de pacientes com SEMA, com maior enfoque na MTF. **Palavras-chave:** memória de trabalho, linguagem, epilepsia, mioclonia.

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Myoclonic astatic epileptic syndrome (MAES) or Doose syndrome occurs in 0.2% among epileptic patients, higher among males (3:1) and seizures normal-ly start between two and five years of age.

Myoclonic attacks occur in a characteristic manner, with successive bent of knees, showing a slow fall and the MAES can course with other types of seizures. Electroencephalographic (EEG) finds are gen-

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eralized, theta and irregular polyspike waves in awake, and hypersynchronous polyspike activity in the sleep. The course is variable, with the possible occurrence of both spontaneous remission and a more severe impairment of cognition<sup>1-3</sup>.

Dysfunction or poor structuring of cerebral connections impair memory skill and consequently language development depending on the characterization of brain involvement, and from this viewpoint, in epilepsy, the temporary dysfunction of a set of neurons, may be an aggravating factor regarding language acquisition<sup>1</sup>.

The objective of the present study was to assess and characterize the verbal language and to determine the phonological working memory (PWM) skill of patients with MAES without spontaneous remission.

## METHOD

The study was approved by the Ethics committee of the University Hospital, Faculty of Medicine of Ribeirão Preto-USP: (n° 12406/2006 – document n° 644/2007) and informed consent was obtained from patients.

The patients were evaluated and diagnosed on the basis of clinical characteristics and EEG patterns, and according to ILAE classification of epilepsy, by the Service of Childhood Epilepsy of the University Hospital, Faculty of Medicine of Ribeirão Preto, University of São Pau-

lo (HCFMRP-USP). Among 16 patients diagnosed since 1990, six participated in the study (Table 1). Inclusion criteria were: MAES patients on treatment with good control for at least five years - free of seizures in the last month during awake period, and without seizures in the night before the evaluation (nocturnal generalized tonic-clonic seizures); absence of visual and hearing impairment and mental diseases, mainly depressive disorders. A model TK Missouri otoscope, a model MA-41 Maico audiometer and a model Az-7 Interacoustics imitancíometer were used for the acoustic meatus exam and audiologic evaluation. The Test of Repetition of Nonsense Words was used for the evaluation of PWM<sup>4</sup>. The test consists of 30 nonsense words with a simple syllable structure following the phonological structure of the Portuguese language of the consonant-vowel type.

For verbal language evaluation, language samples were collected from each patient and videotaped during directed and spontaneous activities with the examiner. A model NV-VZ185LB-5 Panasonic video camera and a VHS-C cassette tape were used to record the samples.

To insure the emission of all phonemes of the Portuguese language during each directed activity, the words were used for imitation and figures were used for the ABFW<sup>5</sup> naming test. The ABFW was used in order to analyze the occurrence of the phonological processes.

**Table 1.** Epilepsy data of the patients.

Patient / Age	Age at the first seizure	Types of seizures*	Intervals – out of seizure –	Anticonvulsants – sequency of changes to control	Last seizure
A / 18	5 mo	1, 4, 5	1 y within first years	Phb, Phb+DPH, Phb+VPA, Phb+CBMZ, VPA+CNZ, VPA+LMT (seizures control)	18 mo
B / 12	4 y 6mo	1, 2, 4, 5	1 mo within first year Last 6 years out of seizures Recent relapse	Phb+DPH, VPA+Phb+CLBZ (decompensation) VPA+ Phb+CLBZ+LMT (1 month without crises; but relapsed after taking off LMT due to a cutaneous rash), Phb (IV) for GTC VPA+CLBZ+LMT (GTC on the sleep)	24 hours
C / 10	2 y 2mo	1, 3, 4	1 mo after 7mo with daily seizures	Phb +CBMZ+VPA+VGB (2 months) Phb, Phb+CBMZ+VPA (myoclonias improved), Phb+VPA+LMT (seizures control)	45 days
D / 9	2 y 4mo	1, 2, 4, 3, 7	2 mo within first year	VPA+ Phb+ CNZ VPA+Phb+ LMT (seizures control)	2 y 6 mo
E / 8	2 y 7mo	6, 4	Without intervals	Phb, Phb+VPA (Phb tapered off) VPA	5 y
F / 18	3 y 2 days	1, 3, 4, 5, 7	Without intervals	CBMZ + Phb + PRI VPA+NTZ (myoclonic improved, GTC persists) VPA+ZRT+ CNZ (status) VPA+CNZ (only GTC on sleep) VPA+ LMT (seizures control)	7 y

\*Sequence of appearance of the seizures. Types of seizures: (1) generalized tonic-clonic (GTC) on sleep; (2) generalized tonic-clonic awake; (3) absence with blinking; (4) myoclonic-astatics; (5) myoclonic generalized; (6) febrile - clonic generalized; (7) tonic on sleep. Phb: phenobarbitone; DPH: diphenil-hydantoin; VPA: sodium valproate; CLBZ: clobazam; CNZ: clonazepam; LMT: lamotrigine; PRI: primidone; y: years; old mo: months.

**Table 2.** Phonological working memory test (PWM). Number of correct answers.

PWM	Patient					
	A	B	C	D	E	F
1 syllable	4	5	5	5	5	5
2 syllables	3	5	5	5	5	5
3 syllables	0	3	2	5	5	5
3 syllables	0	3	3	4	5	5
4 syllables	0	0	0	2	5	5
5 syllables	0	0	0	4	0	2
Total	7	16	15	25	25	27

The patients were first submitted to audiologic evaluation and then to PWM evaluation. For the Test of Repetition of Nonsense Words<sup>4</sup>, the patient was asked to repeat the word immediately after the model given by the examiner without visual clues. The attempt was considered incorrect when the patient omitted, substituted or did not produce phonemes or when he was unable to reproduce the item presented to him by the examiner. The data regarding the number of correct and wrong repetitions of nonsense words were described qualitatively.

For the evaluation of verbal language, the examiner proposed directed and spontaneous activities. For the evaluation of the pragmatic aspects, the capacity of each patient for communicative autonomy, coherence and im-

provisation in new topics was considered<sup>6</sup>. Regarding the morphosyntactic aspect, the types of syntactic constructions correctly used were considered, as well as the sentences containing errors or syntactic omissions, and the variety of structures used during the spontaneous activities with the examiner<sup>7</sup>. Regarding the semantic aspect, evaluation of the following meanings was performed: lexicon, relational-abstract, contextual, and figurative language<sup>8</sup>. Analysis of the evaluation and characterization of the verbal language of the patients was conducted according to qualitative criteria by means of the observation of behavior and of phonetic transcription (based on Brazilian Portuguese language) of the speech of each patient.

### RESULTS

All patients presented deficits in the execution of the PWM test (Table 2), with differences among individuals, and the best performance was observed in the patients D, E and F. The analysis in each verbal aspect of language (Tables 3 and 4) pointed that only patient A showed difficulties in the most of them, pragmatics and dysarthria as well.

### DISCUSSION

To date this is the first work that details language aspects of patients with MAES. Regarding the analysis of performance in the PWM, in the comparison of the six patients evaluated, the increase in chronological age tend-

**Table 3.** Performance of language evaluation: phonological and pragmatic aspects.

Aspects of verbal language	Patient					
	A	B	C	D	E	F
Phonological						
• Mastery of the phonological component	Yes	Yes	Yes	Yes	Yes	Yes
• Distinction of individual sound elements		Yes	Yes	Yes	Yes	
• Clearly perceives distinctive traits of the phonemes	Yes	Yes	Yes	Yes	Yes	
• Demonstrates comprehension of all speech sounds presented by means of naming and imitation tests of the ABFW	Yes	Yes	Yes	Yes	Yes	Yes
Phonetic						
• Coherence in the production of all speech sounds presented by means of naming and imitation tests of the ABFW	Yes	Yes	Yes	Yes	Yes	Yes
• Expression with clarity	No	Yes	Yes	Yes		
Pragmatic						
• Mastery of the understanding of the rules that govern how language is used	No	Yes	Yes	Yes	Yes	Yes
• Communicative autonomy (able to start and change subject, ceded and initiated turns)	No, stimulation needed	Yes	Yes	Yes	Yes	Yes
• Satisfactory latency between the question and the response of the patient	No, large latency	Yes	Yes	Yes	Yes	Yes
• Improvisation in new subjects	No	Yes	Yes	Yes	No	Yes
• Conversation with coherence, regarding subject approached with predominance	Yes	Yes	No	Yes		
• Comprehension within the contexts used during spontaneous activity (sang, imitated a TV anchor, a reporter, animals)	Yes, but with short periods of abstraction	Yes	Yes	Yes		

**Table 4.** Performance of morphosyntactic and semantic aspects of language.

Language aspects	Patient					
	A	B	C	D	E	F
Morphosyntactic						
• The types of phrasal constructions were performed properly during spontaneous conversation	No				Yes	Yes
• The sentences followed a temporal order of subject, verb and predicate	Yes, but asystematic and omissions of subject, verbs, pronouns and connecting elements in a few moments	Yes, Simple phrases	Yes, Simple phrases	Yes, Simple phrases	Yes, Simple phrases	Yes, Complex phrases
• The sentences were constructed in a straightforward manner with good organization of the words	No. Difficulty in the organization of words in different times and limited vocabulary	Yes	Yes	–	–	–
• The sentences were constructed in a complex way and with good organization of words in different times	No	No	No	Yes	Yes	Yes
• There was a variety of phrasal constructions, ie complex structures subject	No	No	No	Yes	Yes	Yes
• The average size of production	Short	Good	Good	Good	Good	Good
• Satisfactory agreement with the markers of gender and number in relation to classes of words used	Yes	Yes	Yes	Yes	Yes, but asystematic	Yes
Semantic						
• Understanding of words	Simple and semi-complex words	Yes	Yes	Yes	Yes	Yes
• Mastery of the lexical meanings of complex words	No	Yes	Yes	Yes	Yes	Yes
• Mastery of the contextual meaning	No	Yes	Yes	Yes	Yes	Yes
• Clarity and understanding on relational-abstract	No	Yes	Yes	Yes	Yes	Yes
• Good use of adverbs	No	Yes	Yes	Yes	Yes	Yes
• Good use of figurative language	No	Yes	Yes	Yes	Yes	Yes
• Consistency, clarity and understanding within the contexts of spontaneous conversation about your daily life.	No	Yes	Yes	Yes	Yes	Yes

ed to coincide with an increased number of errors of repetition of nonsense words. A possible explanation for this fact could be disease duration, with older children having suffered a larger number of seizures and the side effects of various types of medications along the years, since the age at onset of the disease was in infancy. Another relevant item was the marked difference of the errors made by patient A compared to all other patients, observing the same with the verbal language items. Thus, the earliest age of beginning of MAES observed in patient A, 5 months of age, as opposed to about 30 months for the remaining patients could justify such difference. There is consistent literature about the importance of CNS integ-

rity during the first year of life for an adequate forthcoming development<sup>9</sup>.

It should be pointed out that patient F (18 years of age), who showed the best performance in the PWM test, had presented spontaneous remission of the MAES about 7 years before the study, and although this patient presented the results not fully faultless, the large time free of seizures could favor his best performance.

Regarding dysarthria observed in patient A, since MAES is marked by symmetrical and bilateral myoclonic seizures usually strongly involving the upper limbs, in addition to irregular tension of facial and ocular muscles, which in turn may impair speech articulation. Re-

garding the pragmatic aspect, patient A failed all of the items studied, although he presented interest in the conversation. It is important to point that the language deficits presented by patients may have been influenced by variables related to poor sociocultural exposure, especially regarding the lack of use of words with concordance of gender and number markers.

Patient C presented some inadequacies regarding the pragmatic aspect, such as talking about topics out of context. This fact, however, does not reflect primary language alterations but represents manifestations secondary to deficits in attention and behavior constantly observed in this patient during the period of evaluation.

Another important point to be considered is the fact that patient A, who obtained the lowest number of correct answers in the PWM test, also presented greater deficits in the different language components, especially regarding the morphosyntactic aspect. In contrast, patient E, who showed the best performance in the PWM test, demonstrated mastery of language aspects. Several studies have pointed out the relation between PWM and language development and have observed direct interference of this memory with the learning of new words, with more elaborate grammatical constructions, with language comprehension and with learning to read and write<sup>9-15</sup>. Thus, the performance of the two cited patients agrees with literature findings.

The patients between 8 and 12 years old did not manifest evident alterations in the formal aspects of language although they did present deficits in the performance of the nonsense word repetition task. The results obtained regarding PWM deficits may represent a risk factor for spontaneous conversation, the form of language here analyzed, and may be possible predictors of future disorders or difficulties in reading and writing. It should be considered that other forms of language such as formal narrative, reading and writing could offer a more accurate mastery of PWM<sup>16</sup>.

These finds can contribute to strategies of treatment for language problems of patients with MAES, focusing on PWM.

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