



ELSEVIER

Contents lists available at ScienceDirect

## Pediatric Neurology

journal homepage: [www.elsevier.com/locate/pnu](http://www.elsevier.com/locate/pnu)

Original Article

## Language Impairment in Adolescents With Sydenham Chorea



Estefânia Harsányi OT, PhD<sup>a</sup>, Janaina Moreira PhD, MD<sup>b</sup>,  
Arthur Kummer PhD, MD<sup>b,\*</sup>, Zilda Maria Alves Meira PhD, MD<sup>c</sup>,  
Francisco Cardoso PhD, MD<sup>d</sup>, Antonio L. Teixeira PhD, MD<sup>b</sup>

<sup>a</sup> Fundação Hospitalar do Estado de Minas Gerais, Belo Horizonte, Brazil

<sup>b</sup> Laboratório Interdisciplinar de Investigação Médica, Faculdade de Medicina, Universidade Federal de Minas Gerais (UFMG), Belo Horizonte, Brazil

<sup>c</sup> Pediatric Cardiology Clinic, University Hospital, UFMG, Belo Horizonte, Brazil

<sup>d</sup> Movement Disorders Clinic, Neurology Unit, University Hospital, UFMG, Belo Horizonte, Brazil

## ABSTRACT

**BACKGROUND:** Neuropsychiatric comorbidities are frequent in Sydenham chorea. However, cognitive impairment in Sydenham chorea has not been sufficiently described. The objective of this study was to evaluate expressive and receptive language deficits in adolescents with Sydenham chorea. **METHODS:** Twenty patients with Sydenham chorea were compared with 20 patients with rheumatic fever without chorea and 20 healthy controls. Participants were matched for age and gender. Participants were assessed with verbal fluency tasks (phonemic and semantic) and with verbal comprehension tasks (Token Test). Patients with Sydenham chorea were also assessed with the Universidade Federal de Minas Gerais Sydenham Chorea Rating Scale. **RESULTS:** Performance in verbal fluency and in verbal comprehension tasks differed significantly ( $P < 0.01$ ) among the three groups. Patients with Sydenham chorea performed significantly worse than healthy control group in phonemic and semantic verbal fluency tasks as well as in the Token Test. The group with rheumatic fever also performed worse than healthy controls in phonemic verbal fluency. Severity of motor signs in Sydenham chorea inversely correlated with performance in phonemic verbal fluency (words beginning with letter S, and total sum of words beginning with letters F, A, and S). **CONCLUSIONS:** Adolescents with Sydenham chorea show difficulties in verbal fluency and in verbal comprehension. Patients with rheumatic fever also have some degree of language impairment. Future studies must investigate language impairment in different stages of Sydenham chorea (acute, persistent, and remission) and putative biological markers.

**Keywords:** Sydenham chorea, rheumatic fever, language, cognition, neuropsychiatry

Pediatr Neurol 2015; 53: 412–416

© 2015 Elsevier Inc. All rights reserved.

## Introduction

Rheumatic fever is a systemic autoimmune inflammatory disorder following streptococcal  $\beta$ -hemolytic infection. When this autoimmune reaction affects the central nervous

system, the patient may develop a neuropsychiatric disorder called Sydenham chorea. The pathophysiology of Sydenham chorea points to a basal ganglia disorder with evidence derived from magnetic resonance imaging studies that have shown focal striatal enlargement.<sup>1</sup> Some studies have shown that antibodies induced by group A  $\beta$ -hemolytic streptococcus cross-react with basal ganglia antigens.<sup>2</sup> Husby et al. showed that 46% of patients with Sydenham chorea have antibodies reactive to caudate and subthalamic nuclei.<sup>3</sup> In another study, Kotby et al. confirmed the presence of antibasal ganglia antibodies in 100% in acute and 93% in chronic Sydenham chorea patients.<sup>4</sup> Church et al. showed that these antibodies decrease in persistent

## Article History:

Received April 16, 2015; Accepted in final form June 26, 2015

\* Communications should be addressed to: Dr. Kummer; Laboratório Interdisciplinar de Investigação Médica; Universidade Federal de Minas Gerais - Faculdade de Medicina; Av. Prof. Alfredo Balena, 190; 2o andar, sala 281; Belo Horizonte, MG, Brasil 30130-100.

E-mail address: [r2kummer@hotmail.com](mailto:r2kummer@hotmail.com)

Sydenham chorea (63%) and occurs in only 13% of patients with rheumatic fever without Sydenham chorea.<sup>5</sup>

Sydenham chorea is one of the major symptoms of rheumatic fever occurring in 20% to 30% of cases and affecting children between 5 and 15 years.<sup>6–8</sup> Patients with Sydenham chorea often display a combination of motor and behavioral signs and symptoms. The motor features of the disease are chorea, motor impersistence, gait disturbance, oculomotor abnormalities, dysarthria, and tics. The most commonly reported neuropsychiatric syndrome in Sydenham chorea is obsessive-compulsive behavior, but there are also descriptions of attention deficit, irritability, hyperactivity, and depression.<sup>7,9–11</sup>

Despite the significant volume of published articles on the behavioral and motor alterations in Sydenham chorea, there are few systematic studies on cognitive impairment, especially with children and adolescents. Considering that previous studies have not found major cognitive differences (i.e., IQ impairment) between adults with rheumatic fever with and without chorea and that children and adolescents with Sydenham chorea and rheumatic fever fall behind in school,<sup>12</sup> we hypothesized that these patients could have subtler cognitive impairment, although this educational issue could also be explained by absenteeism because of the disease.

Verbal fluency and verbal comprehension are cognitive functions of utmost importance for the execution of daily life activities and for academic performance. The former is defined as the quantity and facility with which words are produced by a person. Verbal fluency could be measured by counting the amount of words produced within a restricted stimulus and within a time limit. There are two components in the spoken verbal fluency that can be evaluated: phonological clusters, where the patient is asked to speak words with a given letter, and semantic clusters, where the person is asked to speak words from the same semantic category.<sup>13</sup> Verbal comprehension is defined as the ability to use verbal reasoning and understanding of the language through the receptive mode and includes at least two components: core grammatical processing, and executive resources such as working memory.<sup>14</sup> There are several studies supporting verbal fluency and verbal comprehension reduction in other disorders that affect basal ganglia, such as Parkinson disease and Huntington disease.<sup>15–18</sup> Current neuropsychological and neuroanatomical data suggest that both neocortical regions (Broca and Wernicke areas) and subcortical regions are the neural bases for language.<sup>19,20</sup> In these cases, the verbal fluency reduction is associated with left dorsolateral and/or striatal lesions<sup>21</sup> and the verbal comprehension reduction results from the disruption of several different parts of frontostriatal circuits.<sup>15</sup> However, language impairment in Sydenham chorea remains unclear. Until now, only two studies investigated verbal fluency in cohorts of Sydenham chorea. Both of them agreed that phonemic verbal fluency is impaired, but they disagreed concerning semantic verbal fluency.<sup>22,23</sup> Furthermore, there are no studies about verbal comprehension in that group. The aim of the present study is to evaluate the hypothesis that Sydenham chorea causes both expressive and receptive language impairment, which will be assessed with tasks of verbal fluency and verbal comprehension.

## Methods

### Participants

This is a cross-sectional study which enrolled 20 (male/female: 11/9) participants with Sydenham chorea, 20 (male/female: 11/9) participants with rheumatic fever, and 20 (male/female: 11/9) healthy controls. All participants were adolescents matched by gender and age in years. Patients with Sydenham chorea were consecutively recruited in the Movement Disorders Clinic at the University Hospital of the Universidade Federal de Minas Gerais. Patients with rheumatic fever without Sydenham chorea were recruited from the Ambulatory of Pediatric Cardiology at the same hospital. Those patients were initially selected from the database of the ambulatory according to their gender and age to match patients with Sydenham chorea. Participants from the healthy group were students from the local public schools also matched for gender and age. Patients were also students of local public schools. Public schools are usually attended by children and adolescents with lower socioeconomic status; therefore, participants probably had similar socioeconomic and cultural backgrounds.

The study was approved by the local ethics committee. All participants and their parents signed the informed consent forms to participate in the study. Participants were excluded from the study if they had intellectual disability, language impairment or other developmental delay, or neurological disorder.

### Assessment

Diagnosis of Sydenham chorea and rheumatic fever was made according to the revised Jones criteria.<sup>24–26</sup> All patients with Sydenham chorea underwent evaluation with the Universidade Federal de Minas Gerais Sydenham Chorea Rating Scale (USCRS),<sup>27</sup> which is divided into three sections. The first section (USCRS-I) assesses behavioral problems related to Sydenham chorea, such as obsessive-compulsive symptoms and hyperactivity. USCRS-II quantifies impairment in activities of daily living. USCRS-III measures motor symptoms. The higher is the score, the more impaired the individual.

All participants underwent a cognitive assessment, which consisted of the Token Test and semantic and phonemic verbal fluency. The Token Test evaluates the patient's ability to understand 36 commands by pointing to objects arranged in an array.<sup>28</sup> The Token Test is an important measure of verbal comprehension (receptive language). During semantic verbal fluency task, participants were asked to produce as many animal names as they could in 1 minute. During phonemic verbal fluency task, they should produce as many words as they could starting with the letters F, A, and S. For each letter, the participant was given 1 minute. The verbal fluency tasks are also used worldwide for the assessment of expressive language, although executive functions are also particularly recruited during these tasks.

Considering that these cognitive tests were not standardized for Brazilian adolescents, the 10th percentile of Brazilian children aged 10 years was used as a conservative cutoff point for the Token Test and semantic verbal fluency to assess whether performance in these tasks had clinical significance.<sup>29</sup> The phonemic verbal fluency tasks do not have norms for Brazilian children or teens.

### Statistical analysis

Fisher exact test was used to compare proportions. Normality was assessed with Shapiro-Wilk test and by visual inspection of histograms. The comparisons between the sociodemographic and clinical variables of the three groups were made by Kruskal-Wallis test. Mann-Whitney tests were used as post hoc tests to pairwise comparison. Correlations were assessed with Spearman correlation coefficient. All tests were two-tailed and *P*-values <0.05 were considered significant for Kruskal-Wallis and correlation tests. For Mann-Whitney tests, *P*-values <0.017 (Bonferroni corrected alphas) were considered significant. Statistical analysis was performed using Statistical Package for Social Sciences, version 17.0.

Download English Version:

<https://daneshyari.com/en/article/3084408>

Download Persian Version:

<https://daneshyari.com/article/3084408>

[Daneshyari.com](https://daneshyari.com)