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A warning to the Brazilian Speech-Language Pathology and Audiology community about the importance of scientific and clinical activities in primary progressive aphasia

Alerta à comunidade fonoaudiológica brasileira sobre a importância da atuação científica e clínica na afasia progressiva primária

Keywords

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Dementia
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Descritores

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ABSTRACT

This article aims to warn the Brazilian Speech-Language Pathology and Audiology scientific community about the importance and necessity of scientific and clinical activities regarding Primary Progressive Aphasia. This warning is based on a systematic literature review of the scientific production on Primary Progressive Aphasia, from which nine Brazilian articles were selected. It was observed that there is an obvious lack of studies on the subject, as all the retrieved articles were published in medical journals and much of it consisted of small samples; only two articles described the effectiveness of speech-language therapy in patients with Primary Progressive Aphasia. A perspective for the future in the area and characteristics of Speech-Language Therapy for Primary Progressive Aphasia are discussed. As a conclusion, it is evident the need for greater action by Speech-Language Pathology and Audiology on Primary Progressive Aphasia.

RESUMO

Este artigo tem como objetivo alertar a comunidade científica fonoaudiológica brasileira sobre a importância e necessidade da atuação científica e clínica a respeito da Afasia Progressiva Primária. Esse alerta é fundamentado em um levantamento bibliográfico sistemático da produção científica brasileira sobre Afasia Progressiva Primária, a partir do qual foram encontrados nove artigos brasileiros. Percebe-se que há uma evidente escassez de estudos sobre o tema, pois todos os artigos encontrados foram publicados em periódicos da área médica e boa parte deles foi composta por amostras pequenas, sendo que dois desprezaram a efetividade da reabilitação fonoaudiológica de pacientes com Afasia Progressiva Primária. São discutidas perspectivas futuras na área, bem como características da terapia fonoaudiológica para Afasia Progressiva Primária. Concluiu-se que é evidente a necessidade de uma maior atuação da fonoaudiologia nas Afasias Progressivas Primárias.

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INTRODUCTION

Primary Progressive Aphasia (PPA) is a dementia syndrome characterized by progressive deterioration of language because of neurodegenerative processes in the left hemisphere. The disease begins in adulthood and compromises language, causing significant functional impairment^(1,2). Patients with PPA go through difficulties ranging from the delay in receiving the correct diagnosis⁽³⁾ to the absence of effective pharmacological treatments, which are only symptomatic⁽⁴⁾. Speech-Language Therapy is usually indicated⁽⁴⁾, but there are few therapeutic strategies specifically for PPA described, and the few studies described previous cover only case reports⁽⁵⁻⁷⁾.

It is believed that there will be an increase in prevalence and incidence rates of diseases related to aging, such as PPA, owing to the growth of the elderly population that has occurred in Brazil in recent years. Thus, there is concern in preparing health systems for the appropriate detection and management of such diseases.

This article aims to warn the Brazilian Speech-Language Pathology and Audiology scientific community about the importance and necessity of scientific and clinical activities regarding PPA. This warning is based on a systematic literature review of the scientific production on PPA.

METHODS

A systematic search for scientific articles was conducted in Pubmed, Scielo, and Lilacs database. All scientific articles produced by Brazilian research groups, developed in Brazilian institutions, and whose objective was to study PPA were included. Studies that were not published in the scientific article format, literature reviews, and studies in other clinical groups outside of PPA were excluded. Repeated articles were included in the first moment and excluded in the following searches. In addition, an open search in Google Scholar was performed to check for possible studies that were not obtained by the systematic search.

For Scielo and Lilacs, the term used was “Primary Progressive Aphasia” for a search in all fields. In Pubmed, the term “Primary Progressive Aphasia” was used in conjunction with the terms “Brasil” OR “Brazil” OR “Brazilian” in all the fields in order to refine the search, because of the large number of publications on the subject in the database.

RESULTS

Figure 1 shows a flow chart of the search for articles, and Table 1 summarizes the nine Brazilian studies on PPA included in the review.

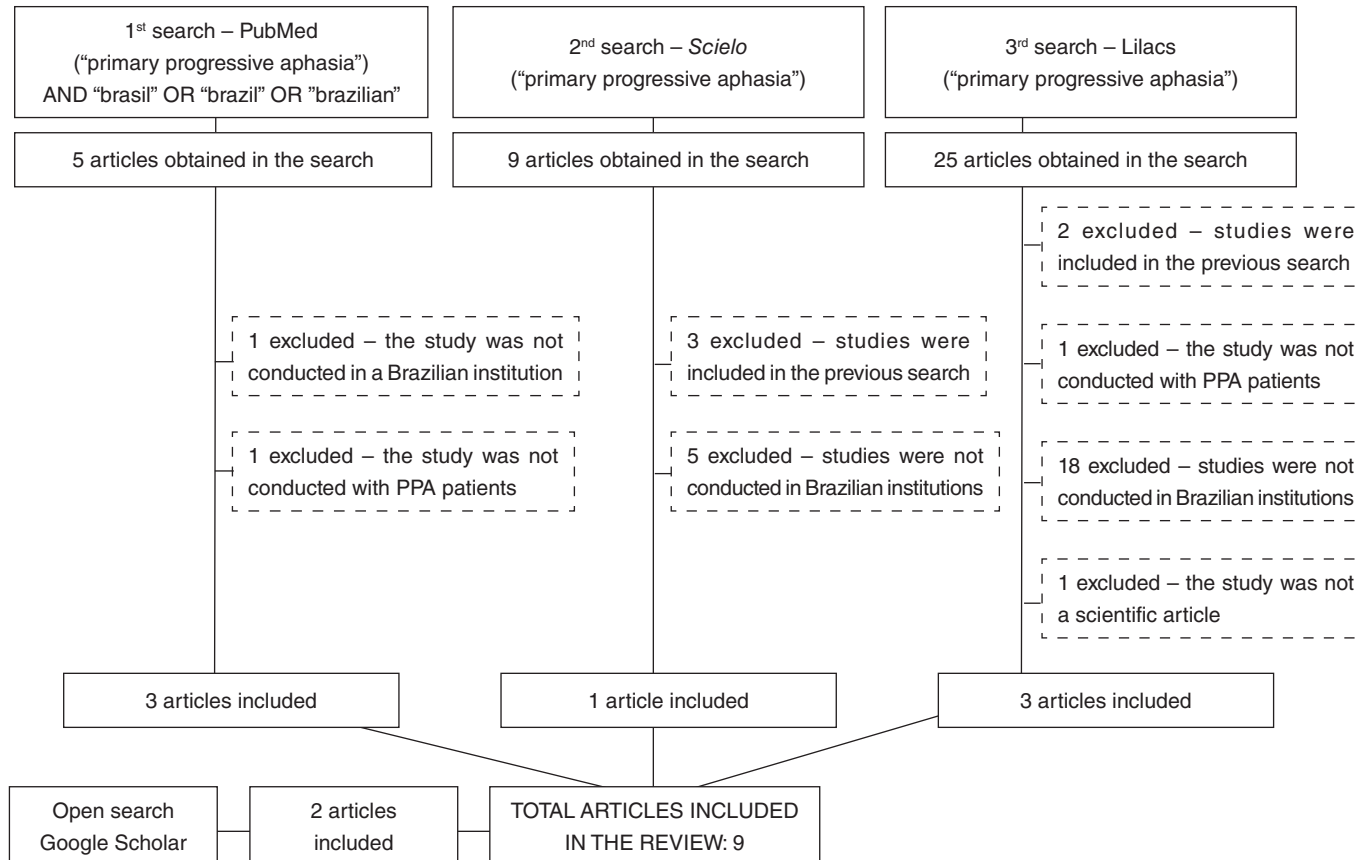


Figure 1. Flowchart of the systematic search for articles

DISCUSSION

It is known that the study of language as a domain of communication is one of the leading fields in Speech-Language Pathology and Audiology. Considering that language is the main aspect affected by PPA, such clinical entity should be widely exploited by the Speech-Language Pathology and Audiology community. However, there is still a scarce production of scientific knowledge on PPA and a timid participation of Speech-Language Pathology and Audiology in this field. These facts are evident by the results obtained in this literature review.

The current systematic review of studies on PPA in Brazil allowed us to reach the following conclusions: there is an

obvious lack of studies on the subject; all articles found were published in medical journals; except for one study, all the rest were composed of small samples; and two articles described the effectiveness of speech-language therapy for patients with PPA.

The findings demonstrate that the theme in question is still little explored in Brazil and that, when it is explored, seems to have a limited participation from the Speech-Language Pathology and Audiology community and to have a limited range, because there are no publications in periodicals in this field.

PPA is less prevalent than other neurodegenerative diseases such as Alzheimer and Parkinson's diseases. This is reflected by the size of the samples used in the articles. However, the sample

Table 1. Articles included in the systematic review of literature

Study	Journal field	Objective	Methodological design	Sample size	Main findings/conclusions
Radanovic et al. (2001) ⁽¹⁰⁾	Medicine - Neurology	To describe the clinical and neuroimaging characteristics of 16 cases with PPA.	Case study	16	Anomia was the earliest symptom of PPA, and the sensitivity of SPECT as a diagnostic method is emphasized.
Caixeta and Mansur (2005) ⁽¹³⁾	Medicine - Neurology	To report a case of the semantic variant of PPA.	Case report	1	The differential diagnosis should be made with AD and other variants of FTD.
Senaha et al. (2010) ⁽⁹⁾	Medicine - Neurology	To analyze the effectiveness of rehabilitation for lexical reacquisition in the semantic variant of PPA.	Case report	3	It is possible to regain lost vocabulary despite the progression of semantic PPA.
Caixeta and Caixeta (2011) ⁽¹¹⁾	Medical Sciences	To report a case of PPA with initial presentation of panic attacks.	Case report	1	PPA can manifest initially through behavioral symptoms.
Oliveira et al. (2011) ⁽¹²⁾	Medicine - Neurology	To identify, through neuroimaging, brain areas that lead to dysphasia when compromised.	Cross-sectional study	4	PPA patients tend to have losses at cortical and subcortical levels.
Vaz et al. (2012) ⁽¹⁴⁾	Medicine - Neurology	To report a case of the semantic variant of PPA.	Case report	1	Language impairment may be the initial characteristic of AD. There is a need to recognize not only FTD and semantic dementia, but also dementia by AD as a result of the evolution of PPA.
Senaha et al. (2013) ⁽²⁾	Medicine - Neurology	To analyze demographic data and classify the variants from a sample of 100 PPA cases.	Cross-sectional study	100	It was possible to classify the language impairment in 80% of the sample into one of three variants of PPA. It would be interesting to have adjustments in diagnostic recommendations to deal with cases that do not fit into any classification and to avoid cases that overlap in more than one classification.
Beber et al. (2014) ⁽¹⁵⁾	Medicine - Neurology	To describe a case in which the diagnosis was divided between Alzheimer's and the logopenic variant of PPA.	Case report	1	It is possible that atypical cases of AD exhibit the logopenic variant of PPA in the early stages of the disease.
Machado et al. 2014 ⁽⁸⁾	Medicine - Neurology	To report a short Speech-Language Therapy treatment in a patient with the nonfluent variant of PPA.	Case report	1	There was improvement and generalization of the trained strategies.

Caption: PPA = primary progressive aphasia; AD = Alzheimer's Disease; DTI = diffusion tensor imaging; FTD = frontotemporal dementia; SPECT = single-photon emission tomography.

size of the studies and their scarcity may reflect the difficulty of attracting patients who were able to participate in research. PPA patients often experience difficulties in diagnosis, causing them to go through many different medical specialties and diagnostic errors in the early stages of the disease, hence taking a long time to receive a proper diagnosis⁽³⁾. In many of the cases, when they are sent to specialized reference centers and receive the correct diagnosis, they are already in advanced stages of the disease, making it difficult for them to participate in studies and therapeutic intervention.

Two studies found in the literature showed cases where Speech-Language Therapy promoted positive results in patients^(8,9), which has also been described in the literature. The international trend refers to the lexical-semantic approaches that use multiple cues (phonological, semantic, and orthographic) and approaches, which focus on the training of oral reading to benefit fluency⁽⁵⁻⁷⁾. There are still no effective pharmacological treatments for PPA, and speech therapy comes as an important option, because it shows to be effective, although it does not prevent the progression of the disease. In addition, the preservation of other cognitive domains and the possibility of compensatory activation of the right hemisphere also suggest that the Speech-Language Therapy may be useful, especially in the early stages⁽¹⁾.

However, there are few descriptions of therapeutic strategies for PPA and their effectiveness. It is known that the neuropathophysiology of aphasia arising from neurodegenerative diseases, such as PPA, is different from aphasia arising from nondegenerative diseases, such as those resulting from strokes⁽⁷⁾. Thus, it is believed that differentiated therapeutic approaches may be necessary for these two types of aphasia.

Faced with this evidence, this article was intended as a warning to the scientific community of the Speech-Language Pathology and Audiology area on the need for clinical actions and the construction of scientific knowledge in this field. The following actions are suggested to improve the performance of Speech-Language Pathology and Audiology in this field:

- increased focus of academic education on PPA;
- educational work with other health professionals, in order to advise on the identification of PPA, giving priority to health professionals working in the early stages of the disease-recognition process;
- insertion of the Speech-Language Pathologist in multi- or interdisciplinary teams to work in the neuropsychological assessment of language and participate in the diagnostic process;
- insertion of the Speech-Language Pathologist in multi- or interdisciplinary research groups to undertake studies on language in PPA and to develop evidence-based therapeutic techniques;
- the role of the Speech-Language Pathologist in the therapeutic treatment of patients with PPA in the public and the private institutions.

CONCLUSION

The scientific production about PPA in Brazil is still scarce. The need for greater action by the Speech-Language Pathology and Audiology field on this aspect is evident in order to contribute to scientific knowledge and to improve the recognition, diagnosis, and therapeutic treatment of these patients.

**BCB conceived the idea for the manuscript; systematically reviewed the articles and wrote the manuscript; LB contributed in the drafting and review of the manuscript; MLFC reviewed the manuscript and supervised the research.*

REFERENCES

1. Mesulam MM. Primary progressive aphasia. *Ann Neurol*. 2001;49(4):425-32.
2. Senaha MLH, Caramelli P, Brucki S, Smid J, Takada L, Porto C, et al. Primary progressive aphasia: classification of variants in 100 consecutive Brazilian cases. *Dement Neuropsychol*. 2013;7(1):110-21.
3. Beber BC, Chaves MLF. Evaluation of patients with behavioral and cognitive complaints: Misdiagnosis in frontotemporal dementia and Alzheimer's disease. *Dement E Neuropsychol*. 2013;7(1):60-5.
4. Tsai RM, Boxer AL. Treatment of frontotemporal dementia. *Curr Treat Options Neurol*. 2014;16(11):319.
5. Henry ML, Rising K, DeMarco AT, Miller BL, Gorno-Tempini ML, Beeson PM. Examining the value of lexical retrieval treatment in primary progressive aphasia: two positive cases. *Brain Lang*. 2013;127(2):145-56.
6. Henry ML, Meese MV, Truong S, Babiak MC, Miller BL, Gorno-Tempini ML. Treatment for apraxia of speech in nonfluent variant primary progressive aphasia. *Behav Neurol*. 2013;26(1-2):77-88.
7. Tsapkini K, Hillis AE. Spelling intervention in post-stroke aphasia and primary progressive aphasia. *Behav Neurol*. 2013;26(1-2):55-66.
8. Machado TH, Campanha AC, Caramelli P, Carthery-Goulart MT. Brief intervention for agrammatism in Primary Progressive Nonfluent Aphasia: a case report. *Dement neuropsychol*. 2014;8(3):291-6.
9. Senaha MLH, Brucki SMD, Nitri R. Rehabilitation in semantic dementia: Study of the effectiveness of lexical reacquisition in three patients. *Dement Neuropsychol*. 2010;4(4):306-12.
10. Radanovic M, Senaha ML, Mansur LL, Nitri R, Bahia VS, Carthery MT, et al. Primary progressive aphasia: analysis of 16 cases. *Arq Neuropsiquiatr*. 2001;59(3-A):512-20.
11. Caixeta L, Caixeta M. Primary progressive aphasia beginning with a psychiatric disorder. *Clin São Paulo Braz*. 2011;66(8):1505-8.
12. Oliveira FP de, Costa JC da, Marroni SP, Silva AMM da, Barreiro SH, Maeda FK et al. Primary progressive aphasia patients evaluated using diffusion tensor imaging and voxel based volumetry-preliminary results. *Arq Neuropsiquiatr*. 2011;69(3):446-51.
13. Caixeta L, Mansur LL. Semantic dementia: clinical and neuroimaging evaluation. Case report. *Arq Neuropsiquiatr*. 2005;63(2A):348-51.
14. Vaz TS, Bertolucci PHF, Oliveira FF de. A patient with primary progressive aphasia developing dementia due to Alzheimer's disease. *Arq Neuropsiquiatr*. 2012;70(7):551-2.
15. Beber BC, Kochhann R, Silva BM, Chaves MLF. Logopenic aphasia or Alzheimer's disease. *Dement Neuropsychol*. 2014;8(3):302-7.